



LIFE WITH HYPERTROPHIC CARDIOMYOPATHY

I sing in my
local choir

I really enjoy
performing

I'm still
challenging
myself

I live with an
inherited heart
condition

ELAINE'S STORY,
PAGE 10

**FIGHT
FOR EVERY
HEARTBEAT**

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In association with

Cardiomyopathy^{UK}
the heart muscle charity



This booklet has been developed from the original version inspired by the founder of Cardiomyopathy UK – Carolyn Biro. Published by the British Heart Foundation.

This booklet is not a substitute for the advice your doctor or cardiologist (heart specialist) may give you based on his or

her knowledge of your condition, but it should help you to understand what they tell you.

The illustrations used in this booklet are artistic impressions and are not intended to accurately depict the medical material that they represent.

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You may be reading this booklet because you have been diagnosed with a heart condition called **hypertrophic cardiomyopathy**. Or maybe someone else in your family has been diagnosed with the condition and your doctor has suggested that you should have some tests to find out if you've also inherited it.

Cardiomyopathy is a disease of the heart muscle. It can run in families and can affect one or more members of a family. Some members of a family may be affected more than others. Some family members may not be affected at all.

There are three main types of cardiomyopathy:

- hypertrophic cardiomyopathy (HCM or HOCM)
- dilated cardiomyopathy (DCM)
- arrhythmogenic right ventricular cardiomyopathy (ARVC).

This booklet is about hypertrophic cardiomyopathy. For information on the other types of cardiomyopathy, see the other booklets in this series. See page 59 for details.

Some people with hypertrophic cardiomyopathy may experience significant symptoms, and a small number could be at risk of abnormal heart rhythms (known as arrhythmias) and sudden death. It's important that families affected receive accurate assessment, diagnosis, treatment and support from specialists in a clinic for inherited heart conditions.

This booklet:

- describes how the normal heart works
- explains what hypertrophic cardiomyopathy is and what can happen if you have the condition
- explains why it's important that close blood relatives of someone with the condition should have an assessment to find out if they've inherited the same condition
- describes the tests your doctor may ask you and your family to have
- describes the treatments you may need
- offers advice on how to live a healthy lifestyle if you have hypertrophic cardiomyopathy.

We explain the medical and technical terms as we go along but, if you find a word you don't understand, look it up in the list of Technical terms on page 55.

This booklet has been produced with the help of doctors and other health professionals, and people who have hypertrophic cardiomyopathy. We hope this booklet will help you to understand your condition and to come to terms with what it means for you and your family. This booklet is the result of a joint collaboration between the British Heart Foundation (BHF) and Cardiomyopathy UK.

At the **British Heart Foundation** we're fighting for every heartbeat. The research we fund has helped push the boundaries of our understanding of genetics, and given us tools we can use to find and help people at risk of serious heart conditions like hypertrophic cardiomyopathy. Join the fight at **bhf.org.uk**

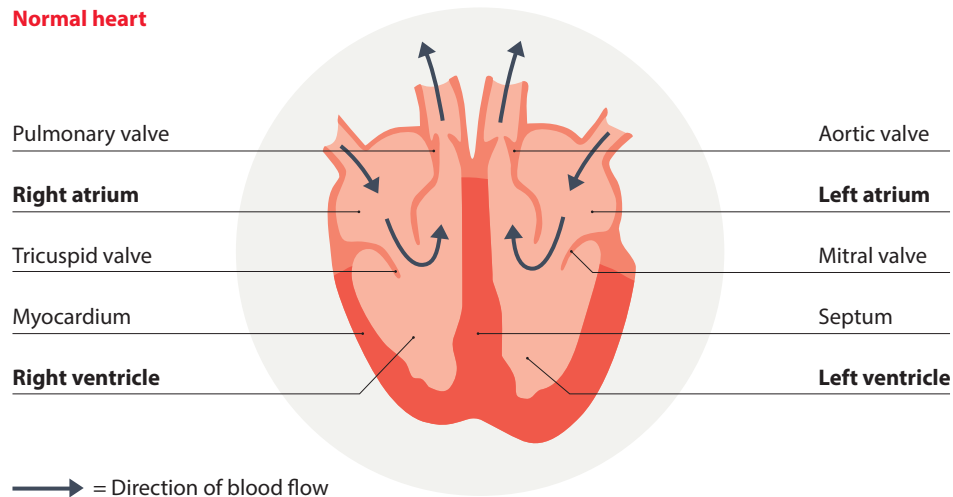
At **Cardiomyopathy UK**, we campaign for more heart checks and gene tests for affected families, to find those at risk and save lives. We help educate doctors about best practice in diagnosing and treating affected families so they get better care. You can help the fight against cardiomyopathy by supporting us at **www.cardiomyopathy.org**

UNDERSTANDING YOUR HEART

THE NORMAL HEART

The heart is a specialised muscle that contracts regularly and continuously, pumping blood to the body and the lungs. It has four chambers – two at the top (the atria), and two at the bottom (the ventricles). See the diagram below.

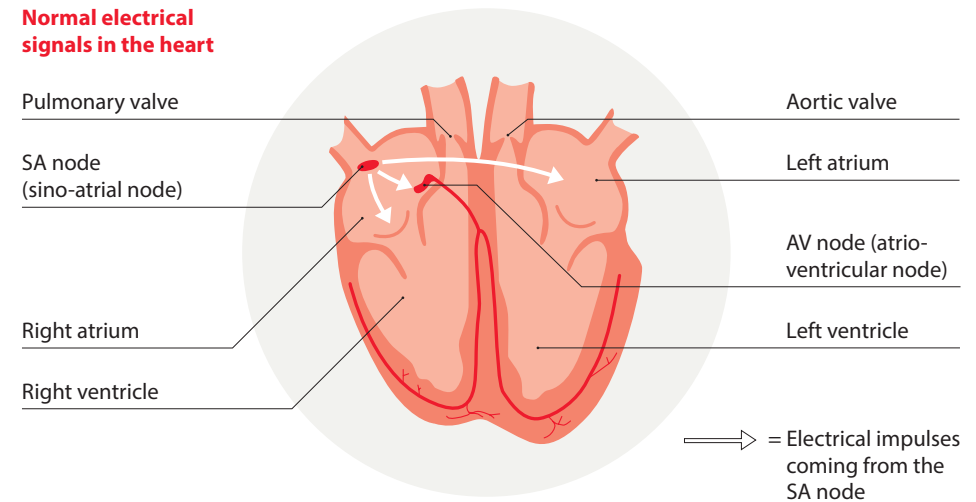
Normal heart



How the heart functions electrically

- The normal trigger for your heart to contract starts in the heart's natural pacemaker, the SA node (sino-atrial node), which is in the right atrium (see the diagram below).
- The SA node sends out regular electrical impulses, which make the atria contract and pump blood into the ventricles.
- The electrical impulses then pass to the ventricles through a form of 'junction box' called the AV node (atrio-ventricular node). This causes the ventricles to contract and pump blood out of your heart.
- The blood from the right ventricle goes through the pulmonary artery to your lungs, and the blood from the left ventricle goes through the aorta and then around your body.

Normal electrical signals in the heart



Structure of the heart

The heart is made up of three layers:

- the endocardium
- the myocardium
- the pericardium.

The endocardium is a thin layer on the inside of the heart, lining the chambers and valves.

The myocardium is the thick, muscular layer of the heart that contracts and squeezes the blood out of the heart. It's this layer that is affected by cardiomyopathy.

The pericardium is a thin, double layer that forms a protective sac around the outside of the heart. It contains a small amount of fluid – called pericardial fluid – which acts as a lubricant when the heart is contracting.

HYPERTROPHIC CARDIOMYOPATHY

ELAINE'S STORY

I was in my mid-30s when I was diagnosed with HCM.

At the time genetic testing wasn't very advanced, and I was told that looking for a faulty gene was like looking for a needle in a haystack. But years later I was offered the chance to have a test. I waited for my results and was told to make an appointment. They had identified one of the most common gene mutations for HCM.

I was worried about my children. I knew they had a one in two chance of inheriting the faulty gene. But if the children were tested, at least we knew which gene to look for.

But I've had positive as well as negative experiences. My cardiologist recommended I have an implantable cardioverter defibrillator fitted, but it doesn't stop me. I've learnt to pace myself. There isn't a cure for my condition and my diagnosis was a huge blow, but I'm living with it safely now and enjoying life.

I sing in my local choir

I really enjoy performing

I'm still challenging myself

I live with an inherited heart condition



WHAT IS HYPERTROPHIC CARDIOMYOPATHY?

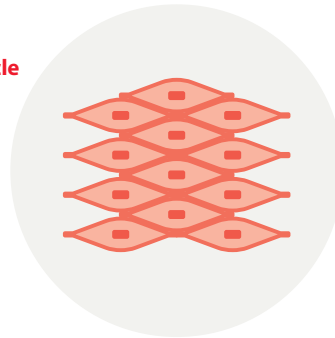
Hypertrophic cardiomyopathy is a disease of the heart muscle. It is often a genetic condition. This means that it's caused by a change (known as a mutation) in one or more genes that can be passed on through families. About 1 in 500 of the UK population has the condition, although most people who have it have few if any symptoms. We explain more about how cardiomyopathy is inherited on page 26.

How does hypertrophic cardiomyopathy affect the heart muscle?

Having hypertrophic cardiomyopathy means that the heart muscle (myocardium) can become excessively thick. How thick the muscle is, and how much of the muscle is affected, can vary from person to person. The left ventricle is almost always affected, and in some people the muscle of the right ventricle also thickens.

In a normal heart, the cells that make up the heart muscle lie in smooth, straight lines, as shown in diagram A below. But in people with hypertrophic cardiomyopathy, the cells lie in disorganised, jumbled layers (known as myocardial disarray), as shown in diagram B below. The heart muscle can also become scarred. The thickening and scarring of the muscle makes the heart muscle stiff. This makes it harder for the heart to pump blood out of your heart and around your body.

A
Normal heart muscle structure



B
Myocardial disarray



The heart muscle can also thicken in people who do not have cardiomyopathy. This can happen because of high blood pressure or intensive athletic training over a long period of time. This type of thickening is not a genetic condition that can be inherited like hypertrophic cardiomyopathy and the thickening is generally mild. The cells of the heart muscle in these people lie in smooth lines as in diagram A on page 12.

The blood vessels that supply the heart muscle (the coronary arteries) are usually normal in people with hypertrophic cardiomyopathy. The valves in the heart are also generally normal, although the mitral valve, which lies between the left ventricle and left atrium, can be affected if certain parts of the myocardium are thickened (see page 15).

How does the muscle thickening vary?

There are four types of heart muscle thickening patterns:

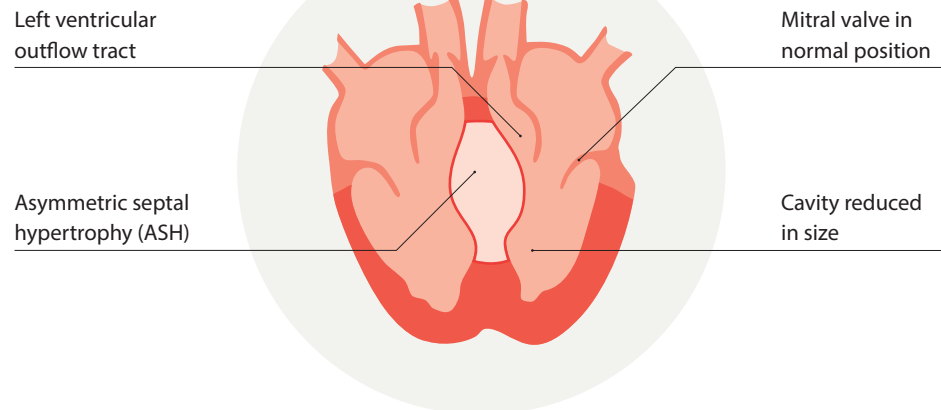
- asymmetrical septal hypertrophy without obstruction
- asymmetrical septal hypertrophy with obstruction
- symmetrical hypertrophy, or concentric hypertrophy
- apical hypertrophic cardiomyopathy.

ASYMMETRICAL SEPTAL HYPERTROPHY WITHOUT OBSTRUCTION

This is the most common heart muscle thickening pattern in people with hypertrophic cardiomyopathy. Here the muscle thickening occurs mainly in the septum – the muscular wall between the right and left sides of your heart (see the diagram below).

The thickening occurs in the centre of the septum but does not restrict the normal flow of blood out of your heart through the left ventricular outflow tract (the short channel below the aortic valve). This is known as asymmetrical septal hypertrophy without obstruction. The mitral valve is not affected and is in a normal position.

Asymmetrical septal hypertrophy without obstruction



ASYMMETRICAL SEPTAL HYPERTROPHY WITH OBSTRUCTION

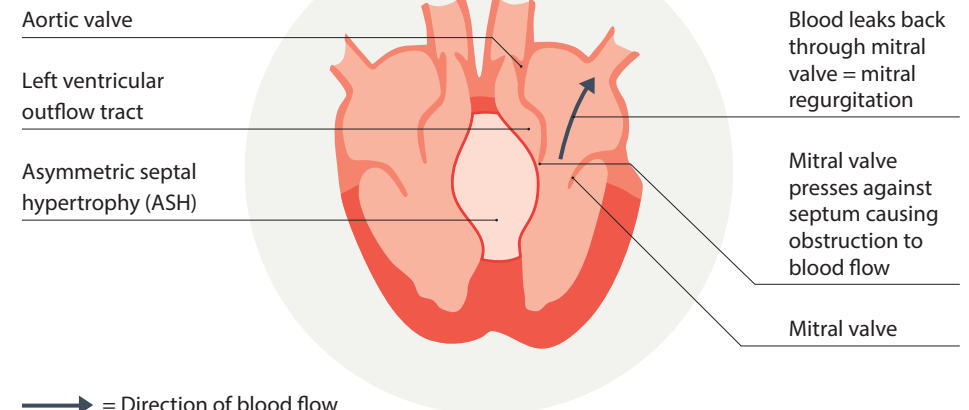
In some cases, the thickening of the septum obstructs the flow of blood through the left ventricular outflow tract when the heart contracts. This is known as asymmetrical septal hypertrophy with obstruction.

The thickening of the septum causes the left ventricular outflow tract to become narrower. This means that when the heart contracts, the mitral valve touches the septum (see the diagram below).

This reduces the amount of blood that is pumped out of the heart and is known as left ventricular outflow tract obstruction. It causes a turbulent blood flow, producing a heart murmur (an unusual heart sound) that can be heard with a stethoscope.

The heart valves are designed to allow blood to move through the heart in one direction only. The mitral valve controls the flow of blood from the left atrium to the left ventricle. However, the obstruction can cause the valve to leak, letting blood flow in a backwards direction from the left ventricle back into the left atrium. This is known as mitral regurgitation.

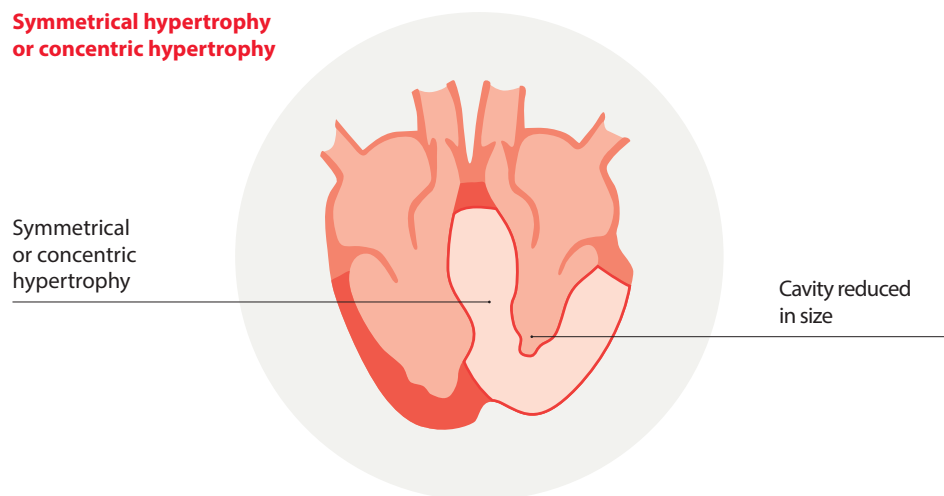
Asymmetrical septal hypertrophy with obstruction



SYMMETRICAL HYPERTROPHY OR CONCENTRIC HYPERTROPHY

This is when the muscle thickening is evenly distributed and equally severe throughout the whole left ventricle. This results in a smaller left ventricular cavity. This condition can also lead to left ventricular outflow tract obstruction as described on page 15.

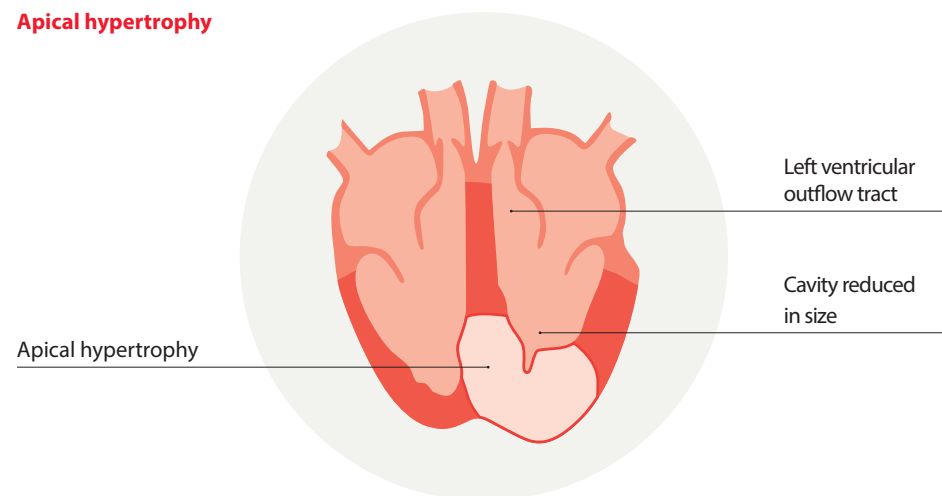
Symmetrical hypertrophy or concentric hypertrophy



APICAL HYPERTROPHIC CARDIOMYOPATHY

In about one in every ten people who have hypertrophic cardiomyopathy, the thickening of the myocardium is mostly at the bottom tip, or apex, of the heart (see the diagram below). This is called apical hypertrophic cardiomyopathy. This thickening does not usually block the left ventricular outflow tract.

Apical hypertrophy



WHEN DOES HYPERTROPHIC CARDIOMYOPATHY DEVELOP?

Each one of us has our own genetic information that makes us unique. Your genes make you who you are, for example, what colour your hair is, your blood type and gender. This genetic information is held in your DNA, in the cells of your body. Your genetic information acts as a code from which a system of proteins can be created. This tells all of the cells in your body what their function should be.

If there's a mistake in a gene, your cells may not work as they should do. These mistakes are known as mutations. We explain more about genetic inheritance on pages 26-30.

In hypertrophic cardiomyopathy, a gene mutation leads to the disorganised arrangement of heart muscle cells (as shown on page 12). The genetic mutation responsible for hypertrophic cardiomyopathy is present at birth. However, the condition is not usually diagnosed at this early stage or even in infancy. Hypertrophy usually develops after puberty, as children grow into adults.

Hypertrophic cardiomyopathy is usually diagnosed when an abnormal electrocardiogram (ECG) or heart murmur (an unusual heart sound) is detected. This can be during a routine examination, perhaps for a medical or for insurance reasons, or when seeing a doctor for another unrelated reason. Sometimes people have symptoms that may suggest they have a heart condition (see page 19) and are sent for further tests (see page 20) so that a diagnosis can be made.

In some cases, the hypertrophy may not be detected until much later in life. This form of hypertrophic cardiomyopathy is known as 'late onset hypertrophic cardiomyopathy'.

WHAT ARE THE SYMPTOMS OF HYPERTROPHIC CARDIOMYOPATHY?

Most people with hypertrophic cardiomyopathy have no symptoms, or have a stable condition throughout adult life. A few people develop serious symptoms and some people find that their symptoms get worse in later life. This may be because they develop an arrhythmia (an abnormal heart rhythm – see page 21). Or it may be because their heart muscle has become progressively stiffer, making it more difficult for the heart to pump.

The most common symptoms are:

Shortness of breath (dyspnoea)

If shortness of breath is a symptom of hypertrophic cardiomyopathy, it generally happens when the person is exercising or being active. Most people are only mildly restricted by their shortness of breath. However, a small number of people may be more restricted in the level of exercise they can do. Very occasionally, some people may be short of breath while they're resting.

Chest pain

Chest pain or tightness on exertion (angina) is a common symptom. The pain occurs because the heart muscle is not getting a sufficient blood supply. Although the main coronary arteries that supply the heart muscle are usually normal, the hypertrophic cardiomyopathy may have caused the microscopic coronary arteries within the heart muscle itself to become narrowed, reducing the supply of blood and causing pain.

Palpitations

This is when you experience a sensation of feeling your heart beating. In some cases, palpitations may start suddenly and feel very fast, and may be accompanied by sweating or light-headedness.

Light-headedness and blackouts

This may happen during exercise, or after palpitations, or sometimes for no obvious reason.

Other symptoms

Sometimes, other conditions can develop as a result of hypertrophic cardiomyopathy and may produce other symptoms. We describe these on page 21.

HOW IS HYPERTROPHIC CARDIOMYOPATHY DIAGNOSED?

Your doctor may suspect that you have hypertrophic cardiomyopathy because of your symptoms, or because you have a heart murmur, or because of the results of your electrocardiogram (ECG). You may also be going for tests because someone else in your family has been diagnosed with the condition (this is known as screening).

If your doctor suspects that you may have thickening of your heart muscle, they will send you to the hospital for a series of tests. Your doctor will also refer you to a cardiologist for specialist advice. The most common tests for hypertrophic cardiomyopathy are:

- a physical examination
- an electrocardiogram (ECG)
- an echocardiogram
- exercise testing
- magnetic resonance imaging (MRI).

We explain more about these and other tests on page 30.

IS THERE A CURE FOR HYPERTROPHIC CARDIOMYOPATHY?

At present there is no cure for hypertrophic cardiomyopathy, but treatment aims to improve symptoms and prevent complications. The need for and choice of treatment will be different for everyone. Your treatment may stay the same for many years, or may change more frequently. There are many forms of treatment available which may help to reduce the risk of developing symptoms, or help to relieve your symptoms. We explain more about these on page 33.

WHAT OTHER CONDITIONS CAN OCCUR AS A RESULT OF HYPERTROPHIC CARDIOMYOPATHY?

Some people with hypertrophic cardiomyopathy can develop a number of other conditions. We describe these conditions in this section.

Arrhythmias

The abnormality of the heart muscle in people with hypertrophic cardiomyopathy can sometimes interfere with the normal electrical activity of the heart. In the affected parts of the heart muscle, the electrical impulse may become disrupted as it crosses the areas of disorganised cells and scarring. This can lead to abnormal heart rhythms, known as arrhythmias.

Arrhythmias are a common complication in people with hypertrophic cardiomyopathy. The two most common arrhythmias in people with this condition are atrial fibrillation and ventricular tachycardia.

In **atrial fibrillation**, the two upper chambers of the heart (the atria) beat irregularly and often very fast. This can lead to feelings of palpitations or fluttering in the chest. The condition can usually be controlled with medication (see page 33).

Ventricular tachycardias are arrhythmias that affect the ventricles, the lower pumping chambers of your heart. The ventricles take over the heartbeat independently of the SA node, leading to a rapid heartbeat. Ventricular tachycardias can be controlled with medication (see page 33), but they can sometimes lead to more life-threatening arrhythmias and increase the risk of sudden death.

Heart block

A small number of people with hypertrophic cardiomyopathy may develop heart block. This is when the electrical impulse travels down to the ventricles slowly, or may even be completely blocked. This affects the way that your heart contracts and causes it to beat much slower.

Is there a risk of sudden death?

Hypertrophic cardiomyopathy is a common condition, and the majority of affected people remain well and have few or no symptoms. Research has shown that, with proper treatment and follow-up, most people with the condition live a normal life. However, because there is a very small risk of developing a life-threatening arrhythmia, a small proportion of people with hypertrophic cardiomyopathy are at risk of sudden death.

Some arrhythmias can cause your heart to beat too fast and chaotically, eventually causing your heart to stop beating. This is a cardiac arrest and can lead to sudden death. For information on treatment for people with hypertrophic cardiomyopathy who may be at risk of sudden death, see page 37.

TESTING, TREATMENT AND YOUR FAMILY

I get strength
from my family

I use my
experience to
help others

I'm taking on
new challenges

I live with an
inherited heart
condition



GILBERT'S STORY

When I was younger, I used to get tired and out of breath doing sport. I put this down to being diagnosed with asthma from an early age.

It wasn't until my thirties that I started getting a dull ache in my chest. I called my doctor and went for a check-up. She thought she could hear a heart murmur and sent me for some tests. An ECG showed some signs that my heart wasn't working properly and I was admitted to hospital.

I spent a long weekend attached to a heart monitor with friends and

family visiting and asking what was happening. The next week a consultant came to see me. He told me I had hypertrophic cardiomyopathy and that it was an inherited heart disease. I was sent home after a two-week stay with a host of tablets.

Getting diagnosed with a heart condition after being fit and well up to that point made me think - why me? But my wife and children have been my rock when times have been hard and have given me the strength to take on new challenges.

BEING DIAGNOSED WITH HYPERTROPHIC CARDIOMYOPATHY

How do people get hypertrophic cardiomyopathy?

Your body is made up of trillions of cells. Each cell has a nucleus, which contains information that makes each one of us unique. This information is your genes. We each have between 20,000 and 25,000 different genes. Genes give the instructions that are needed for development and growth of all the cells in your body, and they determine things like hair colour, eye colour, height and blood type.

Genes are arranged end to end along threadlike structures called chromosomes and are made up of a chemical substance called DNA. Each cell usually carries 46 chromosomes arranged in 23 pairs.

You inherit one set of chromosomes from each of your parents. As chromosomes are made up of genes, this means that you inherit one set of genes from each of your parents. This is why you get certain characteristics from your mother and others from your father.



Your body is made of trillions of cells



Each cell has a nucleus



Each nucleus has 46 chromosomes, in 23 pairs



Each chromosome is made up of a long spiral of DNA



The DNA spiral is divided into genes. You have between 20,000 and 25,000 genes.

It may help to think of your DNA as a book:

- The chromosomes are the chapters. There are 23 pairs of chromosomes, so 23 chapters.
- Each gene is like a paragraph in the chapter. Genes provide the code for proteins, which decide characteristics like hair and eye colour.
- The code in each of your genes is determined by a string of DNA. The DNA is like the letters in the words. The way these 'letters' are arranged can influence your risk of developing conditions such as high blood pressure or heart disease. For example, the words BARE and BEAR have the same letters, but they're arranged differently so they mean different things. In the same way, if the codes in the DNA are arranged differently, they may work in different ways.

If one of your genes has a 'mistake' in it, a bit like a spelling mistake in a word, it could lead to an abnormal protein being produced in that particular type of cell. The same mistake can be passed on to the next and following generations. This 'mistake' is known as a **genetic variant** or a **gene mutation**. Some gene mutations have little or no effect, but others can result in heart conditions such as hypertrophic cardiomyopathy.

Research has shown that about five or six in every ten families with hypertrophic cardiomyopathy have mutations in one of 12 genes that are important in the development of heart muscle cells. The discovery of these gene mutations has helped us understand how hypertrophic cardiomyopathy develops. If someone is found to have hypertrophic cardiomyopathy and a mutation is identified, close family members can have a genetic test to see if they have the same mutation and are potentially at risk of developing the condition.

How is hypertrophic cardiomyopathy inherited?

Drawing a medical family tree will allow your doctor to see if there is anyone else in your family who may have the same condition. The family tree, known as a pedigree, will show an inheritance pattern.

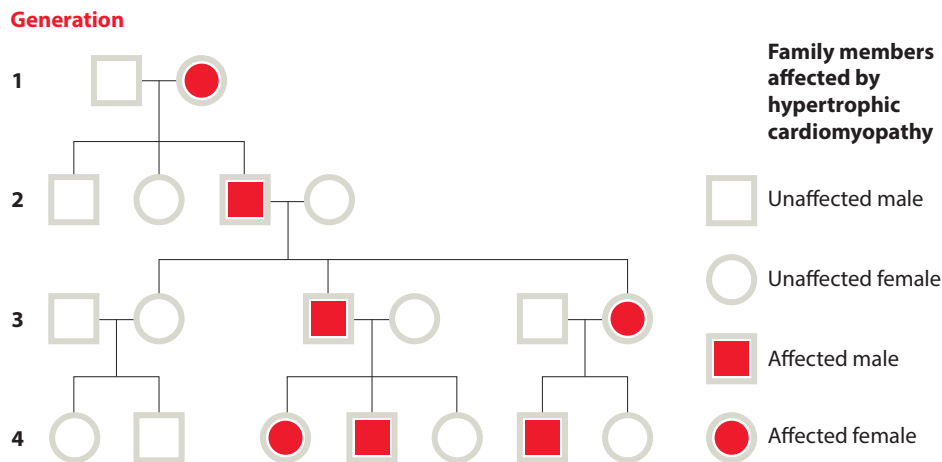
The inheritance pattern for hypertrophic cardiomyopathy is known as autosomal dominant. This means that each child of someone with hypertrophic cardiomyopathy has a 50:50 or one in two chance of inheriting the condition (see the diagram below). The condition may be passed on from an affected male or female and does not skip generations.

A family tree

The family tree below shows four generations affected by hypertrophic cardiomyopathy. Each child of an affected person has a 50:50 chance of inheriting the condition.

If you inherit the condition, there is no way of knowing how severe it might be. Two people in the same family may have different symptoms, and the symptoms may be mild in one person but severe in the other. However, if you don't inherit the mutation, you cannot pass it on to your children, even if other close members of your family have it.

If you don't know your medical family history, or if this is the first time that hypertrophic cardiomyopathy seems to have been diagnosed in your family, any children of the affected person are likely to have a 50:50 or one in two chance of inheriting the condition.



What if something is found in you?

If your doctor thinks that you may have hypertrophic cardiomyopathy, it's important that you have an assessment to find out if you have the condition. We describe all the tests that you may need to have as part of this assessment on page 30. Confirming a diagnosis will help the doctors to decide what treatment is best for you and how often you will need to be followed up. They will also be able to advise you on what you can do to help you to live a normal life.

It may also be possible to have a genetic test to identify the specific gene mutation that has caused your condition. Genetic testing needs to be done at a specialist clinic for inherited heart conditions. Genetic counsellors will explain how likely it is for the abnormal gene to be passed on to your children.

If you don't know where to go for the assessment, call the **BHF Genetic Information Service on 0300 456 8383** to find out where your nearest clinic for inherited heart conditions is, and for information and support about genetic testing.

You should only have genetic testing after you've had advice from a specialist team who can make sure that the right tests are done and that the results are interpreted correctly. Buying genetic tests without the appropriate support and guidance could give you misleading and inaccurate information.

What about screening for your family?

All first-degree relatives (a parent, brother, sister or child) of someone with hypertrophic cardiomyopathy should be provided with information to allow them to decide whether they wish to be tested for the condition. If a family member has a known gene mutation causing hypertrophic cardiomyopathy, it may be possible to test other family members to see if they have the same genetic mutation. We describe the tests for hypertrophic cardiomyopathy on page 31.

For families without a known gene mutation, first-degree adult relatives can be offered clinical screening with an ECG and echocardiogram (see page 31). In most adults where tests show no abnormalities, it's unlikely that the person has hypertrophic cardiomyopathy. However, in a small number of people, a gene mutation may still be present even though there is no physical evidence of the disease. As hypertrophic cardiomyopathy can develop at any age, you may need to be screened again in the future to make sure you haven't developed the disease.

The children of affected parents should be screened about every three years from the age of 10 years onwards. However, children that have symptoms or that are involved in high intensity or competitive physical activity may be screened from a younger age. After the age of 21, it's

recommended that screening is repeated every five years up until the age of 60 and any family members that show symptoms are re-evaluated promptly.

ASSESSMENT AT A CLINIC FOR INHERITED HEART CONDITIONS

In this section we describe what happens during an assessment at a clinic for inherited heart conditions such as hypertrophic cardiomyopathy. These clinics are usually in a hospital.

Medical history

Your doctor will ask you lots of questions about you and your family, such as:

- if any medical conditions affect you or your family, including your parents and possibly your grandparents
- if you've ever had symptoms such as blackouts or palpitations
- if there have been any sudden deaths in your family, including any cot deaths.

If there is a history of sudden death, any coroner's or pathologist's reports that you may have could be helpful.

Medical examination and tests

Most people with hypertrophic cardiomyopathy don't have any visible physical signs of the condition. As part of a medical examination, your doctor will:

- Take your pulse - if this feels 'jerky' or quite strong, it could suggest that the heart muscle is thickened and that your heart is working harder than normal.
- Listen to your heart with a stethoscope - a heart murmur (an abnormal heart sound) is found in three or four in every ten people with hypertrophic cardiomyopathy.
- Do an ECG to look at the electrical rhythm and rate of your heart.
- Arrange an echocardiogram to confirm whether you have hypertrophic cardiomyopathy.

You may also need further tests to find out how thick your heart muscle is, how much of it is affected and to measure how well you can exercise. We explain more about all of these tests on the next pages.

NON-INVASIVE Tests marked with this symbol are 'non-invasive', which means that the test does not involve penetrating the skin or body.

ECG **NON-INVASIVE**

Also called an **electrocardiogram**

This is the most basic test. Small sticky patches called electrodes are put onto your chest, arms and legs and are connected by wires to an ECG recording machine. This picks up the electrical activity that makes your heart beat.

Exercise test **NON-INVASIVE**

Also called an **exercise ECG**

This test is the same as the ECG described above, but is recorded before, during and after a period of time spent exercising on a treadmill or an exercise bike. This allows the doctor to examine any changes in the electrical patterns that occur with exercise, and analyse any abnormalities.

24-hour ECG monitoring **NON-INVASIVE**

Also called **Holter monitoring**

This test involves using a small digital device that you wear on a belt round your waist. Four or six ECG leads from the device are taped to your chest. The device records the electrical activity of your heart for 24 to 48 hours, or for up to seven days. The doctor can then analyse the electrical activity and rhythm of your heart to find out if you have any arrhythmias, such as atrial fibrillation or ventricular tachycardia.

Echocardiogram **NON-INVASIVE**

Also called an **echo**

An echocardiogram uses high-frequency sound waves that reflect against structures in your heart to help build up a detailed picture of your heart. It allows doctors to measure your heart muscle and identify areas of abnormal thickness. It's a similar sort of scan to the ultrasound scan used in pregnancy. An echocardiogram is a safe and easy test, and most people find it's not uncomfortable at all.

Cardiopulmonary exercise test **NON-INVASIVE**

Also called **CPET**

Some hospitals may also ask you to have a cardiopulmonary exercise test – or CPET for short. This test analyses the efficiency of your heart muscle by measuring the amount of oxygen your body uses during exercise. You will be asked to breathe into special equipment while you're exercising. If the efficiency of your heart is low, this may suggest that you have cardiomyopathy.

Coronary angiogram

A coronary angiogram involves having a local anaesthetic in your arm or groin, where a catheter (a thin flexible tube) will be passed into your artery. Using an X-ray machine, the catheter will be directed through your blood vessels and into your heart. A special dye will then be passed through the catheter and a series of X-rays taken. This can show up any narrowed areas or blockages in your artery which could be caused by coronary heart disease.

Cardiac CT scans

CT stands for 'computerised tomography'. A CT scan is a sophisticated type of X-ray. It is useful for looking at the internal organs in your body, such as your heart or lungs.

A CT coronary angiogram shows the blood flow through your coronary arteries. If this test shows that your coronary arteries are narrow, you will need to go on to have a standard coronary angiogram, to find out if you need to receive treatment.

Magnetic Resonance Imaging (MRI)

An MRI scan uses a magnetic field to create images of the heart. For this test, you lie in a short 'tunnel', around which there is a large magnet. At the start of the test you may need to have a special dye (contrast medium) injected into a vein in your arm.

During the test, short bursts of magnetic fields and radio waves from the MRI scanner allow images to be created, processed and analysed. You must lie still while the scan is done. The whole test takes about an hour. An MRI is not painful or uncomfortable. However, some people with claustrophobia (fear of enclosed spaces) may find they cannot cope with having this type of scan.

An MRI is very good at:

- showing the structure of your heart and blood vessels
- showing the condition of your heart muscle
- identifying any scarring (fibrosis) within your heart.

An MRI scan can interfere with the way a pacemaker or ICD (see page 37) works. If you have one of these devices, you can't have an MRI scan unless you have an MRI-safe device.

For more detailed information about these and other tests, see our booklet *Tests*. (To order our booklets see *More information* on page 59).

TREATMENTS FOR HYPERTROPHIC CARDIOMYOPATHY

Hypertrophic cardiomyopathy is not curable, but most symptoms caused by the condition can be controlled by using medicines. Some people may need to have other treatments as well. You will find information about these on page 37. If you have hypertrophic cardiomyopathy with obstruction you may need additional treatment (see page 38).

Medicines

Below we describe the medicines most commonly used for people with hypertrophic cardiomyopathy. Different people need to take different medicines, depending on their condition and symptoms.

Some medicines may cause unwanted side effects, but these are often temporary and disappear over time. Not everyone experiences side effects and you may have none at all. You should not stop taking your prescribed medicines suddenly without medical advice, as this could make your symptoms or condition worse.

Beta-blockers

Beta-blockers act by slowing the heart rate. This reduces the amount of work your heart has to do, so that it needs less oxygen, blood and nutrients. Some beta-blockers can also help control abnormal heart rhythms.

Many preparations of beta-blockers are available, but they differ only slightly from each other.

Unwanted effects

Minor side effects include tiredness, fatigue, and cold hands and feet. Other less frequent effects include feeling sick, diarrhoea, skin rashes, impotence, nightmares and dizziness.

Calcium channel blockers (calcium antagonists)

The heart needs a regular flow of calcium for the muscle cells in the heart to work properly. Calcium channel blockers reduce the amount of calcium entering the muscle cells of the arteries and cause them to relax and widen. As a result of this, the 'resting phase' of the heart's pumping cycle lasts longer. (The resting phase is when the heart rests in between heartbeats and the coronary arteries fill up and supply the heart muscle with blood.) This means that your heart receives a better supply of blood and has to do less work to pump enough blood around your body.

Some calcium channel blockers may increase or reduce your heart rate while you are resting, while others may have no effect on your heart rate. So different calcium channel blockers are used for different heart conditions.

Unwanted effects

Minor effects include constipation, flushing, headache, dizziness, feeling faint or fainting, swollen ankles, indigestion, feeling sick and vomiting.

Anti-arrhythmic medicines

Anti-arrhythmic medicines are medicines for controlling the rhythm of the heart. Beta-blockers, and the calcium channel blocker verapamil, are often used to treat heart rhythm disturbances (arrhythmias), but there are several other medicines that are used almost exclusively for this purpose. One of these is amiodarone.

Amiodarone is very effective in controlling heart rhythm disturbances, especially atrial fibrillation (see page 21).

Unwanted effects

At low doses, amiodarone is well tolerated, but it does have side effects. It may produce headache, flushing, dizziness and stomach upsets. More seriously, and more rarely, it may cause disorders of the thyroid gland, lungs and liver. To avoid these complications, you will need to have regular blood tests. You may also have chest X-rays and tests to see how your lungs are working.

Amiodarone tends to make the skin very sensitive to sunlight so, if you're taking this medicine, you should use a strong sunscreen cream when you're in strong daylight or bright sunshine, and wear a hat.

Anticoagulants

Some people with hypertrophic cardiomyopathy develop atrial fibrillation (see page 21). Atrial fibrillation carries a risk of blood clots forming in the heart which can travel to the brain and cause a stroke. Blood clots are made up of platelets (tiny blood cells) clumped together, and a protein called fibrin. Anticoagulants help prevent fibrin from forming and so prevent clots from forming too. However, in doing so, they may cause internal bleeding or make bleeding from a minor injury worse. Two types of anticoagulants that are often used are heparin and warfarin.

Heparin is injected into a vein and has an immediate effect in preventing further blood clots from forming. This is known as intravenous heparin and is only given in this way if you are in hospital. Another form of heparin called 'low molecular weight heparin' may be given by injection just under the skin, over a longer period of time. This can be given to you either in hospital, or by a nurse at home.

Warfarin (or another oral anticoagulant) is given when long-term prevention of clotting is needed. This medicine is most often used for people with disease of the heart valves, especially those who have been given an artificial heart valve, or for some people who have an abnormal heart rhythm such as atrial fibrillation.

Because the desired effect of anticoagulants like heparin and warfarin is to make the blood thinner so that it doesn't clot so easily, you will need regular blood tests to make sure that the clotting activity of your blood is within safe but effective levels. At the start, these tests may be carried out almost every day, but later on they will be done only every four to eight weeks. If you've been taking warfarin for a long time and your condition is stable, you may be able to get a home-testing kit and do the tests yourself.

If you are taking oral anticoagulants, you should check with your doctor or pharmacist before taking any other medicines. This is because oral anticoagulants interact with many medicines.

Alcohol increases the effect of warfarin, so it is important to avoid excessive drinking or binge drinking.

If you are taking oral anticoagulants, you should avoid drinking cranberry juice as this can affect the amount of time it takes for your blood to clot. Your anticoagulation clinic should be able to give you advice on what changes you may need to make to your diet.

If you're taking anticoagulants, you should always carry an **Anticoagulant card** and remember to tell any doctors, dentists and nurses who are treating you that you are taking anticoagulants.

Any of the following symptoms could mean that your dose of anticoagulants may be too high:

- prolonged bleeding from cuts
- bleeding that does not stop by itself
- nose bleeds that last for more than a few minutes
- bleeding gums
- severe bruising
- red or dark brown urine
- red or black stools
- for women, heavier bleeding during periods, or other vaginal bleeding.

If you're worried, contact your GP or anticoagulant clinic or the emergency department at your local hospital. Make sure that you have your dosage record card and any other medications with you.

Diuretics

Diuretics, or water tablets, increase the output of water and salt in the urine. They reduce the workload of your heart by making sure that the body doesn't hold too much water or salt.

Diuretics can also help to control your blood pressure. If you have too much fluid in your body, your heart has to work harder to pump it around the body. This extra workload increases your blood pressure.

There are three main types of diuretic:

- thiazide diuretics
- loop diuretics
- potassium sparing diuretics.

Thiazide diuretics (such as bendroflumethiazide) and loop diuretics (such as furosemide and bumetanide) can cause you to lose potassium, so your doctor will arrange a blood test a few weeks after you start taking your tablets, to check the potassium level in your blood. If this is getting low, you may be given potassium supplements or a potassium sparing diuretic instead, to correct the problem with the potassium level.

If you're taking a diuretic, you shouldn't have too much salt in your food, as this will counteract the effects of the diuretics. Don't add any salt to food during cooking or at the table, and avoid salty foods. Many processed foods and ready meals contain high levels of salt. It's also important to avoid using salt substitutes as these contain potassium which may have an effect on your blood test results.

Unwanted effects

People with diabetes may find that diuretics raise their blood sugar. People with gout may find that diuretics make their condition worse.

For information about medicines, see our booklet *Medicines for your heart*. (To order our booklets see More information on page 59).

Other treatments for hypertrophic cardiomyopathy

Implantable devices

If you're at high risk of sudden death (for example, if you've already had a cardiac arrest), or if medicines haven't controlled your symptoms, your doctor may advise you to have a pacemaker or an ICD fitted, as well as taking your medication. ICD stands for 'implantable cardioverter defibrillator'.

Both a pacemaker and an ICD consist of:

- a small box called a pulse generator, containing a battery
- one, two or three electrode leads that deliver electrical impulses to the heart.

Pacemaker

A pacemaker can prevent your heart from beating too slowly by 'pacing' your heart to make your heart rate faster.

A pacemaker works by:

- monitoring and storing information about your heart rhythm and heart rate
- sending electrical impulses to your heart that stimulate it to contract. This helps to control your heart rate and stop any excessive slowing of your heart that could trigger an arrhythmia.

Most pacemakers are set to work on demand – they monitor your heart and only deliver an electrical impulse if your heart has missed a beat, or if it's beating too slowly. Other pacemakers send out impulses all of the time and are known as fixed rate pacemakers.

A pacemaker is usually implanted just under your left collarbone. This procedure usually takes about an hour and is normally done with a local anaesthetic and mild sedation. The pacemaker battery usually lasts between six and ten years (and sometimes even longer), but you should receive regular check-ups.

For more information on pacemakers, see our booklet *Pacemakers*. To order our booklets see More information on page 59.

ICD

An ICD monitors your heart rhythm through electrodes placed into your heart. If it detects a dangerous arrhythmia it can deliver a small electrical shock to restore your heart's normal rhythm. This is called shock therapy. An ICD can deliver the following treatments:

- pacing your heart to correct your heart rhythm
- cardioversion – one or more small electric shocks to restore your heart's normal rhythm
- defibrillation – one or more larger electric shocks to get your heart back into a normal rhythm.

An ICD is slightly larger than a pacemaker and is usually positioned under your chest wall muscle below your left shoulder. The procedure may take between one to three hours. Most people have a local anaesthetic and mild sedation, but some may have a full (general) anaesthetic. The ICD battery lasts between four and eight years, but you should have regular check-ups at an ICD clinic.

A new type of ICD called a subcutaneous ICD – or S-ICD for short – is suitable for some people. An S-ICD works in the same way as an ICD, but the leads are inserted under the skin next to the breast bone and not inside the heart as in a normal ICD.

For more on ICDs, see our booklet *Implantable cardioverter defibrillators (ICDs)*. To order our booklets see More information on page 59.

Heart transplant

For a very small number of people, a heart transplant may be considered. As many as 200 heart transplants are carried out on adults in the UK each year. For more information, see our booklet *Heart transplant*. To order our booklets see More information on page 59.

TREATMENT FOR HYPERTROPHIC CARDIOMYOPATHY WITH OBSTRUCTION

About one in every four people with hypertrophic cardiomyopathy has an obstruction to the outflow of blood from the heart (see page 15). This is known as left outflow tract obstruction. It can cause chest pain, breathlessness and fainting. It can also mean you're more likely to develop arrhythmias, in particular atrial fibrillation.

Treatment with medications

Medicines are used to reduce the force of the contraction of your heart. The medicines that are often used are beta-blockers and calcium channel blockers (see page 33).

Disopyramide

Disopyramide was developed mainly for treating abnormal heart rhythms, but is also very useful for treating symptoms relating to hypertrophic cardiomyopathy with obstruction, such as chest pain and shortness of breath. If you need to take this medicine, your doctor will start you on a low dose, and gradually increase the dose while keeping you under medical supervision. Disopyramide should not be taken with amiodarone or the beta-blocker sotalol.

Unwanted effects

Common side effects of disopyramide include dry mouth, blurred vision, difficulty passing urine, and constipation. Once you've been taking this drug for a short while, you'll need to have an ECG to monitor the effect of disopyramide on your heart rhythm, as a small number of people can develop arrhythmias as a result of taking this medicine.

Other treatments

A small number of people who have hypertrophic cardiomyopathy with obstruction will still have symptoms even though they're taking the maximum amount of medicines. These people may need further treatment to help reduce their symptoms and improve their heart muscle function.

There are two main types of treatment:

- surgical myectomy
- alcohol septal ablation.

Both these treatments carry a small risk, and are therefore only used for people who have hypertrophic cardiomyopathy with obstruction, and who have symptoms that are difficult to manage.

Surgical myectomy

A myectomy is an open-heart surgical procedure to remove part of the thickened muscle that is causing the obstruction, allowing the blood to circulate more freely. It involves removing part of the thickened muscle from the septum to widen the outflow tract and relieve the obstruction (see the diagram on page 15). In some people the mitral valve may need to be repaired or replaced. If this is necessary, it is done during the same operation.

What are the risks?

As with any open-heart surgery, there are risks associated with the surgery itself and with having an anaesthetic. You should discuss the specific risks of this procedure with your cardiologist. For more information, see our booklet *Having heart surgery*. (To order our booklets see More information on page 59).

Alcohol septal ablation

This procedure reduces the thickening of the top part of the septum without the need for open-heart surgery. It involves inserting a catheter into the groin in the same way as for an angiogram (see page 32). A small amount of alcohol solution is then injected into a branch of the coronary artery that supplies blood to the upper part of the septum. The alcohol destroys the part of the thickened heart muscle that is causing the obstruction. This makes the muscle thinner and relieves the obstruction.

What are the risks?

In a small number of cases, the alcohol injection may damage the electrical system of the heart. If this happens, you may need to have a pacemaker fitted after this procedure (see page 37). You should discuss this with your cardiologist.

EVERYDAY LIFE

MELISSA'S STORY

My hypertrophic cardiomyopathy was diagnosed when I was admitted to hospital with something else. While I was there, some student doctors came and listened to my heartbeat. One of them realised something was wrong, so they gave me an echo and that's how I found out.

Now I take medicine daily and go for regular check-ups at hospital. Once a year I'm also given a 24-hour heart monitor.

My condition is genetic, so all my family have been tested. I've been told by my cardiologist that if I have children then I could pass it down to them too.

My condition has prevented me from doing some stuff that all my friends can do, but I have good days and bad days. A bad day is when I can just about walk from here to there and then have to rest. My condition will never go away, but now I'm studying at college and I'm looking forward to my future.

I'm learning
new skills

I want to work
with children

I can't wait to
get started

I live with an
inherited heart
condition



LIVING WITH HYPERTROPHIC CARDIOMYOPATHY

Making lifestyle changes are an essential part of managing hypertrophic cardiomyopathy. If you have this condition you should:

- Avoid strenuous exercise – especially intense, competitive sport and heavy weight lifting.
- Ask your doctor about any fluid and dietary guidelines you may need to make to manage your condition.
- Have regular check-ups to monitor your condition.

Who should be involved in routine medical care?

Your GP should be involved with your general everyday care. They can discuss your condition with your cardiologist and make any changes to your treatment. You will probably see your cardiologist once a year, but this will depend on your overall condition and your symptoms.

Do I have to make any changes to my everyday life?

For many people, hypertrophic cardiomyopathy does not interfere with their lifestyle. However, there are some things that could make your symptoms and condition worse. On the next few pages we talk about the everyday things that concern people with hypertrophic cardiomyopathy.

Exercise

Your heart is a muscle and needs exercise to stay healthy. However, some people with hypertrophic cardiomyopathy become short of breath or get chest pain (angina) when they are physically active. Most people with hypertrophic cardiomyopathy will be advised not to take part in competitive sports or other strenuous physical effort. This can be difficult for younger people who have previously taken part in this type of activity. You should always discuss with your doctor the type and level of physical activity that you should do.

Diet, alcohol and weight

Being overweight places an extra strain on the heart. Eating a healthy, balanced diet will help you to maintain a healthy weight. See our booklet *Keep your heart healthy* for more information. (To order our booklets see More information on page 59).

Drinking more than the recommended amount of alcohol can damage your heart health and can lead to high blood pressure. Alcohol is also a depressant, so it can make feelings of anxiety or depression worse. Talk to your doctor to find out what is a sensible limit for you. If you have left ventricular outflow tract obstruction, you may find that alcohol makes your symptoms worse.

Weight gain

If you're eating a healthy diet but you notice that you've put on weight over a few days (about a half to one kilo, or one or two pounds) you'll need to see your doctor. The weight gain could be due to fluid retention which can be treated by adjusting your medicines.

Smoking

Although tobacco smoking is not directly associated with hypertrophic cardiomyopathy, smoking will increase your risk of developing coronary heart disease and reduces the flow of blood to your heart muscle. It also reduces the amount of oxygen being carried in the blood and makes your heart work harder. For information on giving up smoking, see our booklet *Stop smoking*. (To order our booklets see More information on page 59).

Sex

Some people with hypertrophic cardiomyopathy will have concerns about sex. Knowing your limitations and what brings on your symptoms can help you enjoy a full and active sex life. Like any other physical activity, having sex can increase the heart rate and blood pressure. This increases the work of the heart and, for some people with a heart condition, sex may bring on symptoms such as breathlessness or chest discomfort. However, sex is just as safe as other equally energetic forms of physical activity or exercise.

To reduce the chance of having angina symptoms during sex, avoid having sex after a heavy meal, and try not to be too energetic at the start of your sexual activity.

Loss of sex drive is not uncommon in people with a heart condition. Some men may experience impotence. This may be the result of:

- taking certain medicines, such as beta-blockers, which can affect your sex drive
- the emotional stress you may be feeling
- poor blood circulation or diabetes.

Impotence is a common problem so, if you're having difficulties, talk to your doctor about it. Talking to your partner can also help to lessen your concerns and fears and help your partner to understand your situation.

Other common causes of loss of sex drive are:

- depression
- concerns and fear about how safe it is for you to have sex
- anxiety about the possibility of your ICD delivering an electrical shock during sexual activity
- lack of communication between you and your partner.

If you have a heart condition, you should be cautious about taking PDE-5 inhibitors such as Viagra. You should not take these medications if you are taking GTN or any other medication containing nitrates. PDE-5 inhibitors can also interfere with other medicines for your heart, so always check with your doctor beforehand.

If you have hypertrophic cardiomyopathy with left ventricular outflow tract obstruction you should not take this type of medication as it can make your symptoms worse.

General health

Do I need to have a flu vaccination?

Having a flu vaccination will not prevent you from getting a flu virus, but it will reduce the severity of the flu if you do get it. Every year there is a different strain of the flu virus and a new vaccine is developed. If your doctor recommends that you have the flu vaccine, you will need to have a vaccination each year.

What if I need an anaesthetic?

In most people with hypertrophic cardiomyopathy, having a general or local anaesthetic is not associated with any additional risk. Your anaesthetist will monitor your blood pressure and heart rate carefully during your surgery or procedure. Some spinal blocks or epidurals can cause a drop in blood pressure, so they should be used with caution.

Anxiety and depression

Finding out that you have a diagnosis of cardiomyopathy can be distressing. In the early days it is normal to feel shock, anger and disbelief. We all have different ways of adjusting, and there is no right or wrong way. Stress, fear and anxiety can sometimes help us to re-examine and to change our lifestyles. However, when emotions like this persist for long periods of time, or begin to overwhelm you, they can lead to anxiety and depression that may need treatment.

Talking to someone can be very helpful. Friends and relatives may be able to understand how the condition is affecting you, but sometimes it's easier to talk to someone who's not so closely involved. Talking to other people with cardiomyopathy may help you come to terms with your condition and help you to see that you're not alone. There are also many organisations and charities that can help you to cope. See page 60 for details.

Recognising and understanding the cause of your anxiety can help you to deal with it. Using relaxation techniques can help. If simple techniques don't help, or if you feel overwhelmed with your anxiety, you should speak to your doctor who will be able to decide if you need medical treatment.

Depression

It's normal to feel fed up or miserable from time to time, and feelings like this usually don't last for long or interfere significantly with our lives. However, if these feelings persist and severely interfere with your life, you may have depression. Depression can last for months and can affect you in a variety of ways, both physically and psychologically. You may be unable to sleep properly, lose your appetite, or frequently think that life is not worth living. Depression usually comes on gradually. Some people may not recognise that they are becoming depressed because they are paying more attention to their physical symptoms than their psychological state.

Having a routine and remaining active can help to deal with mild depression. It can help you to feel in control and get back to a natural sleeping pattern. Although hypertrophic cardiomyopathy often restricts vigorous exercise, it shouldn't stop you being active. (See page 44 for more on physical activity.)

If you have severe or long-term depression, you may benefit from taking anti-depressants. They help by altering the transmission of chemical substances in the brain, thereby improving your mood. It can take several

weeks to for you to feel the benefits of anti-depressants. Your doctor may also suggest that you would benefit from speaking to a clinical psychologist.

Driving

You may continue to drive if your symptoms are well controlled and your doctor says that you're fit to do so. You may need to notify the DVLA (Driver and Vehicle Licensing Agency) about your heart condition or about a treatment you've had for it. For more information, contact the DVLA:

- visit www.gov.uk/health-conditions-and-driving
- call the DVLA on 0300 790 6806
- write to them at DVLA, Swansea SA99 1TU.

Work

Most people who are diagnosed with hypertrophic cardiomyopathy will be able to continue in their normal job. However, if you have a heavy manual job which involves strenuous activity, you might not be able to do as much and you should discuss this with your doctor.

For some occupations – such as airline pilots and heavy goods vehicle drivers – there are strict guidelines about whether you can continue in your job if you have hypertrophic cardiomyopathy. Whatever your job, your doctor may be able to provide additional information for your employer which could mean that you're able to continue in your job. If your employer (or potential employer) asks you to fill out a medical questionnaire, you must tell them about your condition.

Holidays and travel insurance

If you have hypertrophic cardiomyopathy, there are no restrictions on flying provided that your symptoms are well controlled, but make sure you plan ahead.

- If you have heavy bags, make sure there is someone who can lift them for you.
- Remember to leave enough time to get to the departure gates without rushing.
- If you get breathless easily, you may also want to organise in advance to have help at the airport. Airlines can often provide transport to a departure gate or a wheelchair for you to use.
- If you think you might need oxygen during the flight, speak to your doctor about this and then to your travel company or airline.

If you want to travel within the European Union, you should get a European Health Insurance Card (EHIC) and take it with you when you're travelling. This card ensures EU nationals obtain the same level of health care as a local resident in a member country when travelling. You can apply for this card online through the NHS website or by phoning the automated service on 0300 3301350.

If you're diagnosed with hypertrophic cardiomyopathy, an insurance company may charge more for your travel insurance. For a list of insurance companies who are sympathetic to people with heart conditions, contact the British Heart Foundation or Cardiomyopathy UK. Their contact details are on pages 59-60.

Life insurance and mortgages

If you have hypertrophic cardiomyopathy, you may have difficulty obtaining life insurance or a mortgage.

The Association of British Insurers (ABI) says that insurers will take into account a family history of all medical conditions. They will not ask people to take genetic tests when applying for life insurance. However, if you wish to take out a new life insurance policy, you may be required to report the results of any genetic tests you have already had, unless you're told otherwise by your life insurance company.

Financial support

Some people with hypertrophic cardiomyopathy may be able to apply for Personal Independence Payment (PIP). This is a social security benefit that helps towards some of the extra costs arising from a long-term illness or disability. Between April 2013 and October 2017, PIP will start to replace Disability Living Allowance (DLA) for people aged 16 to 64. If you already receive DLA, you'll need to check how this change affects you. There are also other benefits and allowances available for those people whose symptoms result in severe restrictions. For more information, search online for GOV.UK.

PREGNANCY AND CHILDBIRTH

Pregnancy is usually safe for the majority of women with hypertrophic cardiomyopathy. However, as pregnancy increases the workload of the heart, it can carry an additional risk for women with hypertrophic cardiomyopathy. The level of risk will vary depending on the nature of your cardiomyopathy and how well your heart is pumping. If you know you have hypertrophic cardiomyopathy, you may also be concerned about taking medications during pregnancy and the effect that they may have on your baby. If you're planning a family, you should talk to your doctor before becoming pregnant.

If you're thinking of having an epidural during your labour, you should discuss this with your doctor early on in your pregnancy, as an epidural can cause a significant fall in blood pressure.

For some women, becoming pregnant produces symptoms of cardiomyopathy for the first time. If this is the case, your obstetrician will liaise closely with your cardiologist to look after you and your baby.

Should I have children?

If you have hypertrophic cardiomyopathy, there's a risk of passing on the condition to your children. For more information see page 26. The decision about whether to have children is one that you should make only after discussing it with your partner and at a clinic for inherited heart conditions. To find out where your nearest clinic is, call the **BHF Genetic Information Service** on **0300 456 8383**.

For more details about living with cardiomyopathy, see the booklet *Living with Cardiomyopathy*. To order our booklets see More information on page 60.

LOOKING FORWARD

I'm training as a cardiac nurse

I've got the support of my family

I want to help others going through what I did

I live with an inherited heart condition



STEPHEN'S STORY

My diagnosis came as an incredible shock – completely out of the blue.

I was diagnosed with hypertrophic cardiomyopathy at the age of 19. At the time I was applying to do a dance degree at university and needed a medical. They picked it up then. My GP referred me to the cardiac unit at my local hospital for some tests. I'd always been fit and into sport, and I'd never had any symptoms, so I couldn't believe my diagnosis.

It probably took me two years to get over it mentally.

I've now got an ICD and I go back to hospital every six months to have it checked. It's strange at first but you wouldn't know it's there now. My wife certainly feels happier, knowing there's a back-up in case something goes wrong.

I'm training to be a nurse now because of the people I've met along my journey. I'd like to help people going through what I've been through – people like me.

THE FUTURE

Diagnosis and treatment of hypertrophic cardiomyopathy has improved recently, and many people who get an early diagnosis and treatment remain well and have a good quality of life. Further research into better and more effective treatment and screening options means that the quality of life, treatment and monitoring of people with hypertrophic cardiomyopathy is likely to improve further in the future.

New areas of research and development include:

- Faster and cheaper technologies that allow more of the gene mutations that cause cardiomyopathy to be identified. A new project is looking at the genes of 100,000 people with inherited heart conditions and is expected to inform care and lead to new treatments for these conditions.
- Very small pacemakers that don't use electrode leads and are inserted directly into the heart through a vein in the groin.
- Smaller ECG recorders that can send information straight from a patient to their doctor through email.
- New drugs aimed at improving cardiomyopathy symptoms, improving exercise capacity, and preventing disease progression.
- A sticking plaster like patch to stick on the chest and record heart rhythms.
- 3D electronic membranes that fit over the outside of the heart and use sensors and electrodes to monitor the heart's electrical activity. In the future, these membranes may replace pacemakers and ICDs.
- Research on the use of stem cells to reduce scarring in the heart, increase the number of healthy, working heart cells and improve the heart's ability to pump properly.
- Gene therapies - drugs to treat the actions of particular gene mutations causing cardiomyopathy.
- New tools to help cardiologists identify people who are at high risk of complications and help target life saving treatment.

TECHNICAL TERMS

A

Angina

Heaviness or tightness in the centre of your chest, which may spread to your arms, neck, jaw, back or stomach. Or it may affect just your neck, jaw, arms or stomach.

Angiogram

An X-ray picture of your blood vessels which shows whether the arteries are narrowed and, if so, how narrow they have become. An angiogram can be used to examine the coronary arteries (a coronary angiogram) or other arteries in your body.

Anticoagulation

Treatment with medicine, to reduce the risk of blood clots.

Aorta

The large artery (blood vessel) leading out of the left side of your heart and supplying your whole body with blood.

Apical hypertrophic cardiomyopathy

Thickening of the heart muscle at the tip of the left ventricle known as the apex. 'Apical' means 'to do with the apex'.

Arrhythmia

An abnormal heart rhythm.

Atrial fibrillation

A fast, irregular heart rhythm.

Atrium

One of the two top chambers of your heart. (The plural of 'atrium' is 'atria'.)

Autosomal inheritance

Where a condition is passed on in a family from one generation to the next without skipping any generations.

Autosomal dominant inheritance

In this type of inheritance, if one of your parents has a faulty gene, there's a 50:50 chance you could inherit it.

C**Cardiac arrest**

When a person's heart stops pumping blood around the body and they stop breathing normally. This is fatal if the heart's normal rhythm is not restored within a few minutes.

Cardiologist

A doctor specialising in diseases of the heart.

Cardiomyopathy

A disease of the heart muscle.

Cardioversion

A procedure to restore a regular heart rhythm.

Chromosome

A threadlike fibre which is in all cells and which carries genetic information.

Concentric hypertrophy

A condition where thickening occurs equally throughout the wall of the ventricle.

D**Diuretics**

Also known as 'water tablets'. Diuretics increase the output of water and salt in your urine.

DNA

The genetic code from which proteins 'the building blocks of life' are made. We all receive a copy of half of each of our parents' DNA when the egg and sperm meet to conceive a new human being.

G**Gene**

A segment of DNA responsible for the production of a specific substance such as a protein, which in turn is essential for a particular characteristic or function in your body.

H**Heart block**

When the electrical impulses sent by the atria to the ventricles are delayed or are blocked.

Hypertrophy

Thickening.

I**Implantable cardioverter defibrillator (ICD)**

A metal electronic device similar to a pacemaker. It is implanted under the chest wall muscle below your left shoulder. It can regulate the rhythm of your heartbeat and, if a dangerous arrhythmia occurs, it can deliver an electrical shock to your heart to restore the normal heart rhythm.

L**Left ventricular outflow tract**

The short channel below the aortic valve, through which the blood flows from the ventricle into the aorta.

M**Mitral regurgitation**

When your blood flows in a backward direction through the mitral valve.

Mitral valve

The valve on the left side of your heart, between the atrium and the ventricle.

Murmur

The sound of turbulent blood flow in the heart which may be caused by an underlying problem such as a heart valve defect. It can be heard through a stethoscope.

Mutation

A mutation or 'mistake' in the DNA code that causes its eventual product (usually a protein) to function abnormally, which in turn is responsible for a disease.

Myectomy

A surgical operation to remove thickened muscle and relieve outflow tract obstruction.

Myocardial disarray

When the cells of the heart muscle are lined up in a disorganised way.

Myocardium

Your heart muscle.

P**Pacemaker**

An electronic device which takes over the role of your heart's natural pacemaker and regulates the rhythm of your heartbeat. It is usually implanted just under your left collarbone.

Palpitation

This is the sensation of feeling your heart beating - whether it is beating normally, quickly, slowly or irregularly.

Pulmonary artery

The artery carrying blood from the right side of your heart to your lungs.

S**Septum**

The thick, muscular wall between the left and right sides of your heart.

Side effects

Unwanted effects of a medicine.

T**Tachycardia**

A fast heart rate.

V**Ventricles**

The two bottom chambers of your heart.

Ventricular

From, or belonging to, the ventricle.

FOR MORE INFORMATION

For information on your nearest clinic for inherited heart conditions

BHF Genetic Information Service

Greater London House
180 Hampstead Road
London NW1 7AW

Phone: 0300 456 8383

Website: bhf.org.uk

The BHF GIS can provide you with information and support if someone in your family has been diagnosed with, or has died from, a suspected inherited heart condition. This includes supporting you and your family by helping you get an expert assessment in a specialist clinic that deals with inherited heart conditions.

BHF publications

You can find out more about the topics covered in this booklet in some of our other resources:

Atrial fibrillation (HIS24)**Heart rhythms (HIS14)****Heart transplant (HIS13)****Implantable cardioverter defibrillators (ICDs) (HIS19)****Losing someone to heart disease (G419)****Medicines for your heart (HIS17)****Pacemakers (HIS15)****Stop Smoking (G118)****Sudden arrhythmic death syndrome (M111A)****Tests for heart conditions (HIS9)**

We also have a range of booklets that cover other inherited heart conditions, how to have a healthy lifestyle and how to keep your heart healthy. Visit our website bhf.org.uk for more information.

To order any of these booklets:

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

Our resources and services are free of charge, but we rely on donations to continue our vital work. If you'd like to make a donation, please call our donation hotline on **0300 330 3322** or visit our website at bhf.org.uk/donate

FOR MORE ON CARDIOMYOPATHY

Cardiomyopathy UK

Unit 10 Chiltern Court
Asheridge Road
Chesham
Bucks HP5 2PX

Freephone Helpline: 0800 018 1024
(Monday to Friday, 8.30am to 4.30pm)

Website: www.cardiomyopathy.org

Email: info@cardiomyopathy.org

Cardiomyopathy UK is a registered charity that helps people who are affected by cardiomyopathy.

Cardiomyopathy UK provides information and support to families affected by cardiomyopathy. It has cardiomyopathy support nurses, patient information days, support groups and a network of affected volunteers who provide support to others by telephone and email. To become a member of the charity, call the helpline or see the website.

Other Cardiomyopathy UK publications

- Living with cardiomyopathy
- Cardiomyopathy in children and young people
- Hearty – children's guide to cardiomyopathy

For support on coping as a family where there has been a sudden cardiac death

Cardiac Risk in the Young – CRY

Unit 7, Epsom Downs Metro Centre
Waterfield
Tadworth
Surrey KT20 5LR

Phone: 01737 363222

Email: cry@c-r-y.org.uk

Websites:

www.c-r-y.org.uk

www.sads.org.uk

www.cry-csc.org.uk

CRY offers help, support and counselling to families where there has been a sudden cardiac death of an apparently fit and healthy young person.

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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 British Heart Foundation, please call our donation hotline on **0300 330 3322**, visit **bhf.org.uk/donate**, or post it to us at the address below. Thank you for supporting our fight.

There are lots of other ways that you can help us. Go online at **bhf.org.uk** to find out how.

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** or write to us at the address below.

British Heart Foundation

Greater London House
180 Hampstead Road
London NW1 7AW
bhf.org.uk

Heart Helpline

0300 330 3311
(a similar cost to 01 and 02 numbers)
For information and support on anything heart-related.

Genetic Information Service

0300 456 8383
(a similar cost to 01 and 02 numbers)
For information and support on inherited heart conditions.

About Cardiomyopathy UK

Cardiomyopathy UK is a registered charity that provides information and support to families affected by the heart muscle disease cardiomyopathy. We provide information on the different types of cardiomyopathy and help people to understand cardiomyopathy, reducing their fears and promoting independence. We provide a free helpline, information booklets, cardiomyopathy support nurses, information days, support groups and a network of volunteers, called key contacts, who provide one-to-one support over the telephone or by email.

Cardiomyopathy UK Website

Cardiomyopathy UK is the only UK charity dedicated to providing support and information to families affected by cardiomyopathy. It is supported by supporter donations, gifts in wills and fundraising. For more information about the charity and what it does, please call **0800 018 1024** or visit our website **www.cardiomyopathy.org**

Freephone Helpline

We have a freephone helpline (**0800 018 1024**) that is manned from 8.30am to 4.30pm on weekdays. Callers can usually speak to one of our cardiomyopathy support nurses.



**British Heart
Foundation**

Cardiomyopathy^{UK}
the heart muscle charity

At the British Heart Foundation, we've pioneered research that's transformed the lives of people living with heart and circulatory conditions. Our work has been central to the discoveries of vital treatments that are changing the fight against heart disease.

Cardiomyopathy UK campaigns for more heart checks and gene tests for affected families to find those at risk and save lives. We educate doctors about best practice in diagnosing and treating affected families so they get better care.

But so many people still need our help.

Join the British Heart Foundation and Cardiomyopathy UK in our fight for every heartbeat in the UK. Every pound raised helps to make a difference to people's lives.