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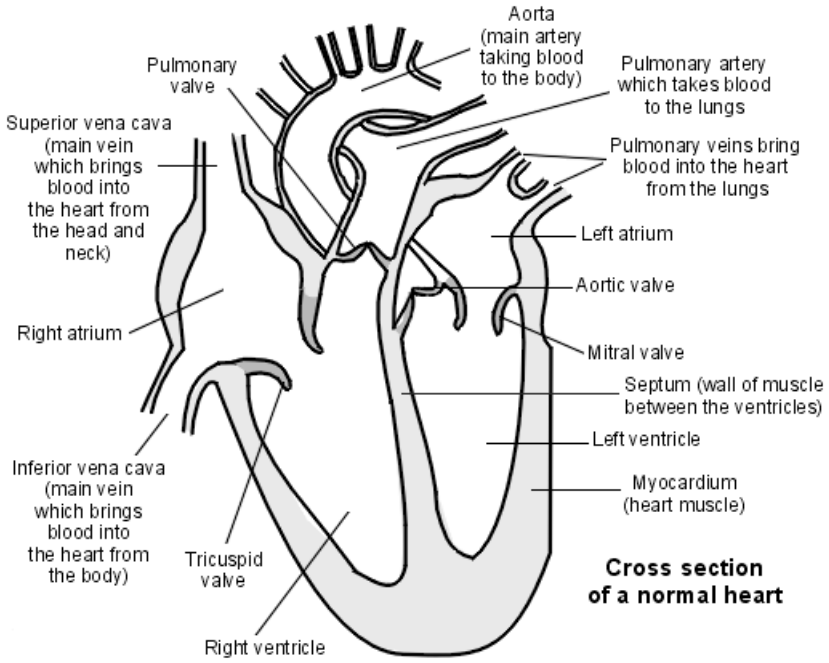
# Hypertrophic Cardiomyopathy

In hypertrophic cardiomyopathy the heart muscle becomes thickened (hypertrophies) in parts of the heart. In the normal heart, the muscle cells are regular and patterned. In hypertrophic cardiomyopathy the cells of the heart muscle become irregular and disordered.




The muscle surrounding the left ventricle is the area commonly affected. Sometimes the muscle around the right ventricle is also affected. The degree of thickening may vary in different places. For example, the wall dividing the right and left ventricle (the septum) is often the area with the greatest thickening. In about 1 in 4 people the muscle thickening is evenly distributed throughout the walls of the left ventricle.

Hypertrophic cardiomyopathy (HCM) may lead to problems which include the following:

- The affected heart muscle (usually around the left ventricle) may become stiff. This can mean that your left ventricle may not fill as easily as normal. Less blood than normal is then pumped out from your heart with each heartbeat.
- The thickening may partly obstruct the flow of blood from your left ventricle into your aorta. This results in less blood being pumped out from your heart. The partial obstruction may also make the blood flow turbulent. Turbulent blood flow can make small blood clots more likely.
- The thickened heart muscle may affect the function of your heart valves. In particular, the mitral valve may become leaky if it does not close properly.
- In some people, the abnormal heart muscle affects the electrical conducting system of the heart. This may cause abnormal heart rates and/or rhythms to develop.

## Symptoms

Most people with this condition do not have any symptoms. Symptoms can range from mild to severe and may not develop straightaway. Possible symptoms include the following:

- **Shortness of breath**. This may develop only when you exercise if the condition is mild. When the condition is more severe, you can be breathless at rest.
- **Chest pain (angina)**. This may develop only when you exercise but it can also occur at rest when it is more severe. The pain occurs because the supply of blood and oxygen to the heart muscle is not sufficient to meet the demands of the thickened muscle.
- **Palpitations**. Sometimes abnormalities of heart rhythm (arrhythmias) develop which can cause palpitations. You may become aware that your heartbeat is fast and/or irregular.

- **Dizziness and fainting attacks**. These occur more commonly when you exercise but they may occur when you are resting. This may be due to reduced output of blood from the heart or because of arrhythmias.

## What causes hypertrophic cardiomyopathy?

Heart muscle can thicken because of something, such as high blood pressure. In HCM the heart muscle thickens without an obvious cause.

In most cases the condition is inherited. If a couple (where one person has HCM) has a child, there is a 1 in 2 chance of the child being affected. This pattern of inheritance is called autosomal dominant. It seems that affected people inherit defective genes which are involved in making parts of the heart muscle cells.

HCM affects about 1 in 500 people. It is sometimes present at birth and can develop in young children. However, it most commonly develops in early adulthood. HCM tends to affect men more often than women.

## How does hypertrophic cardiomyopathy progress?

The thickening of the heart muscle does not tend to progress once you stop growing. This means that, for many people, the symptoms remain stable during adulthood. Unfortunately, the symptoms gradually become worse for some people as the heart muscle becomes more stiff. Sometimes the function of the heart gradually deteriorates and heart failure may develop. See the separate leaflet called Congestive Heart Failure.

## How is hypertrophic cardiomyopathy diagnosed?

A doctor may suspect this condition because of:

- Your symptoms.
- Your family history.
- Changes on your heart tracing (electrocardiogram, or ECG) - this is a tracing of the electrical activity of the heart.
- Changes on your chest X-ray. This may show your heart is large or that there is fluid in your lungs.
- An ultrasound scan of the heart (echocardiogram, or 'echo'). This is a painless test which can measure the thickness of your heart muscle.
- Cardiac catheterisation - a small plastic tube is passed into the heart, usually through a blood vessel in the groin, to measure the pressure in the heart chambers.

Once the diagnosis is confirmed, other tests may be needed to assess the severity of your condition. A Doppler ultrasound scan also looks at blood flow through the heart chambers. This shows how well the heart ventricles are filling and contracting. A Doppler ultrasound scan can also show if there is any turbulent blood flow within the ventricles.

## Family screening

Your first-degree relatives (mother, father, brother, sister, child) should have tests such as an ECG and an echocardiogram. Some people with HCM do not have any symptoms. This is why close relatives should be screened.

In some centres it may be possible to have a genetic blood test. The children of affected parents should be screened every three years until puberty, and then every year until they reach the age of 20 years.

There is currently no UK national screening policy so the tests available may depend on the services offered at your local hospital.

## Treatment

There is no treatment which can reverse the changes of the heart muscle. Treatment aims to ease symptoms if they occur and to prevent complications. If you do not have any symptoms or you only have mild symptoms then you may not need any treatment.

Treatment which may be required includes the following:

### General lifestyle advice

- **Exercise**. Depending on the severity of the condition, some people are advised not to take part in strenuous sports or jobs. Your doctor can advise you about this.
- **Weight**. Try not to become overweight, which can put an extra strain on your heart.
- **Alcohol**. Normal social drinking in moderation should not affect your heart. However, too much alcohol can affect the heart muscle and should be avoided.
- **Don't smoke**. Smoking can cause more damage to your heart.

### Medication

The medicines advised depend on what symptoms or complications develop. For example:

- **Beta-blockers** (such as propranolol) and calcium antagonists (especially verapamil) are the commonly used medicines. These can slow the heart rate and make the heart squeeze (contract) less forcefully. This allows more time for the ventricle to fill with each heartbeat. These medicines may be used to treat chest pain, breathlessness and palpitations.
- **Various other medicines called anti-arrhythmic medicines** (for example, amiodarone) are used to treat and to prevent abnormalities of heart rhythm (arrhythmias). They work by interfering with and helping to correct the electrical impulses in your heart.
- **An anticoagulant medicine** may be advised if you develop atrial fibrillation (a common arrhythmia). With this arrhythmia a blood clot is a possible complication. Anticoagulant medicines help to prevent blood clotting by thinning the blood.

### Surgery

If your cardiomyopathy is severe, an operation may be an option:

- **Myectomy**. This is an operation to remove a segment of thickened muscle from the wall dividing the right and left ventricle (the septum). It is done as open heart surgery. It is not a cure but it can help when the thick septum is causing obstruction to the flow of blood through the aortic valve.
- **Alcohol septal ablation**. Alcohol is injected into the small blood vessels (arteries) which supply the thickened area of heart muscle. This destroys that part of muscle, which then becomes thinner.
- **Valve replacement** may be needed if the mitral valve is affected and does not work properly.
- **A heart transplant** may be needed in a very small number of people.

## What are the complications?

The severity of HCM and the complications caused by HCM vary from person to person. The possible complications include:

- Abnormal heart rhythms (arrhythmias).
- Infective endocarditis.

Sudden collapse and death occurs in a small number of people with HCM. This is probably due to a severe arrhythmia which may develop suddenly.

## Outlook

The thickening of the heart muscle does not tend to progress once you stop growing. This means that, for many people, the symptoms remain stable during adulthood. Unfortunately, the symptoms gradually become worse for some people as the heart muscle stiffens. Sometimes the function of the heart gradually deteriorates and heart failure may develop. See the separate leaflet called Congestive Heart Failure for more details.

## Further reading & references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