

Hypertrophic Cardiomyopathy

The aim of this fact sheet is to explain what Hypertrophic Cardiomyopathy is, what effect it will have on a child and how it can be treated.

What is Hypertrophic Cardiomyopathy?

Hypertrophic means thickening. Cardio means of the heart. Myopathy is any disease of muscles. Hypertrophic Cardiomyopathy (HCM) means a thickening of the heart muscle. HCM is caused by 'disorganised' (not arranged in the normal way) heart muscle cells. It causes the muscle to thicken.

How your child may be affected

When the heart beats, it contracts to pump blood out to the body and lungs, and relaxes to fill with blood again. HCM can have the following effects:

- ♥ The ventricle (pumping chamber) does not pump enough blood. There are two reasons for this. Firstly, the thickened muscle around it makes the ventricle smaller. This means that it does not hold as much blood as in a normal heart. Secondly, the thickened muscle cannot relax very much so less blood is able to enter the ventricle.
- ♥ The thickening can happen in the dividing wall (septum) between the left and right ventricles. This makes the pathway of the blood (the outflow tract) into the aorta narrower, reducing the amount of blood that can be pumped into the aorta.
- ♥ The thickened heart muscle may affect the function of the heart valves. In particular, the mitral valve may become 'leaky' if it does not close properly.

- ♥ The electrical pathways of the heart may be damaged causing the heart beat to become fast or erratic – arrhythmia.

Figure 1- Hypertrophic Cardiomyopathy

Hypertrophic Cardiomyopathy

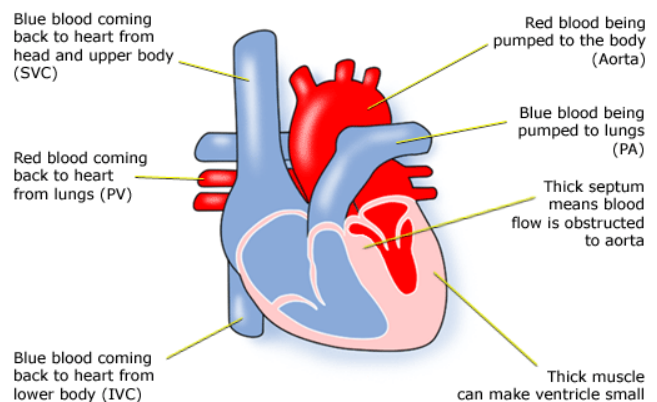
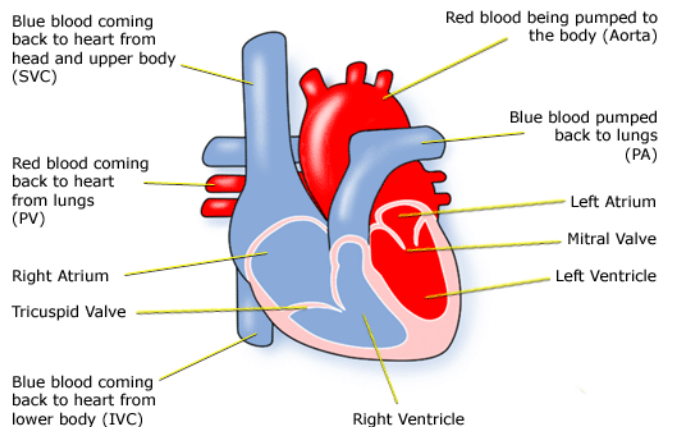


Figure 2 – Normal Heart

Normal Heart



A young person may not necessarily have symptoms as a result of having HCM. The symptoms have and how serious they are will depend on how thick their heart muscle is, and where the thickening happens in their heart. Symptoms may develop at any stage in life. Possible symptoms include:

- ♥ Breathlessness (shortness of breath).
- ♥ Chest pain (angina). The pain happens because not enough blood and oxygen is getting to the thickened heart muscle.
- ♥ Palpitations. Arrhythmias (fast heart beats) can cause these palpitations.
- ♥ Dizziness and fainting attacks. This can happen because of the smaller than normal quantity of blood being pumped by the heart or because of an arrhythmia (fast heart beat).

Diagnosis

Hypertrophic Obstructive Cardiomyopathy (HOCM) may be suspected in your child because a relative has been diagnosed. About half the children with HOCM have a family member affected.

HCM may occur in those with other illnesses such as muscular dystrophy, or in Noonans Syndrome.

When a heart problem is suspected the tests used can be:

- ♥ Measuring the pulse, blood pressure, temperature, and number of breaths taken per minute;
- ♥ listening with a stethoscope for changes in the heart sounds;
- ♥ an oxygen saturation check to see how much oxygen is getting into the blood;
- ♥ a chest x-ray to see the size and position of the heart;
- ♥ an electrocardiogram (ECG) to check the electrical activity of the heart;

- ♥ checks for chemical balance in blood and urine;
- ♥ a 24 hour ECG to monitor the electrical activity of the heart throughout an entire day;
- ♥ an ultrasound scan (echocardiogram) to see the thickness of the muscle, how blood moves through the heart, and how the ventricles fill;
- ♥ a catheter test to look at the changes inside the heart, to measure the pressures in the heart chambers, and perhaps to check the electrical activity that regulates heartbeat;
- ♥ an MRI scan to show the anatomy and function of the heart; and
- ♥ a biopsy – taking small pieces of the heart under general anaesthetic.

Monitoring

As your child gets older their condition may be stable, could deteriorate, or could improve. They will need regular monitoring. Tests could be the same as for diagnosis, depending on the symptoms your child has, or may include exercise testing and 24 hour ECGs.

Contact the cardiologist, if your child experiences any of the following symptoms:

- ♥ Increased breathlessness that is interfering with their normal activities.
- ♥ Light-headedness caused by arrhythmia or a drop in blood pressure.
- ♥ Chest pain – which could be angina caused by the heart muscle needing more oxygen than can be supplied.

Treatment

HCM can be improved and controlled by treatment including:

- ♥ drugs to control dangerous heart rhythms and relax the heart, allowing it to fill more easily;

- ♥ fitting a pacemaker to allow the heart to work in a more efficient manner; and
- ♥ implanting a defibrillator to prevent arrhythmias and sudden death.

Surgery may be necessary:

- ♥ to remove part of the septum that is blocking the outflow tract; or
- ♥ to repair or replace a damaged mitral valve.

For most children this surgery is low risk, but it depends on how well your child is otherwise. The doctors will discuss risks with you in detail before asking you to consent to the operation.

For open heart surgery the length of time in hospital will usually only be 10 to 12 days, of which one or two will be spent in the intensive care and high dependency unit. Of course, this depends on how well your child is before and after the surgery, and whether any complications arise.

How it affects your child

If the open heart surgery is straightforward, and they do not have

other health problems, your child should be well soon after surgery.

There will be a scar down the middle of the chest, and there may be small scars where drain tubes were used. These fade very rapidly in most children, but they will not go altogether. Smaller scars on the hands and neck usually fade away to nothing.

If the child has been given an artificial valve, they will need to take an anticoagulant medicine to stop blood clots forming. Aspirin and warfarin are anticoagulant medicines. If your child is on warfarin, they will need to have their blood tested regularly. Read more in our fact sheet 'Warfarin' - order by calling our freephone infoline 0808 808 5000 or download from our website www.chfed.org.uk.

Heart transplant

If your child's condition cannot be controlled using the treatments described above, they may be assessed for a heart transplant.