Hypertrophic Cardiomyopathy

Hypertrophic cardiomyopathy (HCM) is a genetic disease that causes the heart muscle to grow thicker (hypertrophy). The heart muscle becomes stiff. This causes it to work harder. To understand HCM, it is helpful to know more about the heart.

The heart

The heart is a muscular organ about the size of a fist (Figure 1). It sends blood to the lungs and to all parts of the body. The heart has 4 chambers. The 2 upper thin-walled chambers are the right atrium and left atrium. The 2 more powerful lower chambers are the right ventricle and left ventricle.

The right heart chambers get blood from the body and pump it to the lungs. There, the blood picks up oxygen and gives up carbon dioxide. The left ventricle, the main pumping chamber of the heart, gets this oxygen-rich blood from the lungs and pumps it to all parts of the body.

Figure 1. Normal heart

If you have any questions, please ask your physician or nurse.
HCM

In HCM, the heart muscle, especially the wall between the right and left ventricles (interventricular septum), is affected (Figure 2). The septum thickens due to abnormal growth of the muscle cells and becomes stiff. This reduces blood flow to and from the ventricles. In some people, the thick tissue can greatly reduce the flow of blood out of the heart to the body (outflow tract obstruction). The thickened heart muscle may also cause an abnormal heartbeat.

Figure 2. Heart with HCM

About 1 in 500 people have HCM. It is the most common cause of sudden cardiac death in young people and athletes. HCM is often found in young adults in their teens or in their 20s. In most cases, there is a family history of HCM.

Symptoms

Most people with HCM have no symptoms at all, while some may feel these symptoms:

- Shortness of breath
- Chest pain
- Dizziness or fainting
- Palpitations (feeling that the heart is fluttering)

These symptoms may happen after physical activity.
Tests
Sometimes the 1st signs of HCM are found during a routine physical exam. The physician may find these signs:
- Irregular heart rate
- Heart murmur due to outflow tract obstruction

The physician may check for HCM if a person has a family member with this disease.

The physician may order these tests:
- Electrocardiogram (ECG) – Shows the heart’s electrical activity
- Echocardiogram – Ultrasound of the heart at rest
- Stress echocardiogram – Ultrasound of the heart during exercise on a treadmill
- Magnetic resonance imaging (MRI) – This scan gives the physician the following information:
  - Confirms the HCM diagnosis
  - Identifies the area and extent of muscle thickening
  - Shows if there is any scar tissue in the heart muscle
- 48-hour Holter monitor – Checks for irregular heart rhythms that would increase your risk of sudden death

The care team will explain the tests you need in more detail.

Treatment
Most patients, who have a normal heart rhythm and are without symptoms, do not need a specific treatment. Still, a small number of these patients may be at risk for sudden cardiac death. This often happens at a young age and during competitive sports. People with HCM should never do very heavy physical activity or competitive sports.

The physician will evaluate a person with HCM very carefully for their risk of sudden cardiac death. The evaluation looks at:
- Family history of HCM
- Extent of heart muscle thickening
- Heart rhythm
- The heart’s ability to work normally during exercise
- Current symptoms

Most people with HCM lead normal, active lives. But, they need routine follow-up visits to check for any changes in their heart.
They may go a long time without a change in their condition. However, some people have symptoms that increase over time and need treatment, such as medications.

- Beta blockers and calcium channel blockers can help you manage shortness of breath, palpitations, lightheadedness or chest pain. Although most people will start these medications at low doses, it is quite common to use high doses if symptoms are not completely managed.
- Antiarrhythmic medication can help heart rhythm problems.

In some cases, a person with HCM may need certain procedures or surgery. Your physician will explain these in more detail.

- Cardioversion – This procedure can control heart rhythm problems that last and are bothersome.
- Pacemaker – This device can relieve outflow tract obstruction.
- Implantable cardioverter defibrillator (ICD) – This device that can protect against dangerous abnormal heart rhythms that put you at risk of sudden death. The ICD treats these rhythms by pacing or shocking the heart back to a normal rhythm.
- Septal myectomy surgery – The surgeon removes some of the thickened heart muscle that is blocking the flow of blood out of the heart.
- Mitral valve surgery – The surgeon can repair a leaking mitral valve caused by the thickened muscle.
- The Maze procedure – This treats atrial fibrillation, a fast heart rhythm that is common in HCM.
- Heart transplant – This may be an option for people with HCM who have advanced heart failure that cannot be treated in other ways.

**Family Screening**

If you have HCM, we recommend HCM screening for your parents, siblings and children (your first-degree relatives). This includes getting an ECG and echocardiogram every 5 years.

Genetic testing may be an option. We offer genetic testing (at an outside lab) for people with HCM who are interested in learning whether their children may inherit this condition.

Your physician can give you more information. Check with your insurance company for specifics about coverage for this testing.

**More information**

To learn more about HCM, please go to [nm.org/conditions-and-care-areas/cardiovascularcare/center-for-heart-failure/hypertrophic-cardiomyopathy](nm.org/conditions-and-care-areas/cardiovascularcare/center-for-heart-failure/hypertrophic-cardiomyopathy).

If you have any questions, contact us at:

Northwestern Medicine Bluhm Cardiovascular Institute HCM Program
Galter Pavilion
675 North Saint Clair Street, Suite 19-100
Chicago, Illinois 60611
Phone: 312.695.4965 • TTY: 711