Your Guide to Hypertrophic Cardiomyopathy

Virginia Mason Franciscan Health™
Center for Cardiovascular Health

Welcome! We are here to empower you with the knowledge you need to better understand and manage your heart health.

What is Hypertrophic Cardiomyopathy (HCM)

Hypertrophic cardiomyopathy is a disease in which the heart muscle becomes abnormally thick, and it is usually inherited. The thickening involves the left ventricle, the chamber of the heart that is responsible for pumping blood out of the heart. This abnormal thickening causes the heart to contract harder and the muscle to become stiffer, which can reduce the amount of blood that can be pumped out of the heart into the aorta with each heartbeat.

Often, this thickened muscle can cause a partial obstruction of blood from leaving the heart. Patients with obstructive forms of HCM make up about 2/3 of patients and often are brought to medical attention by a heart murmur picked up on a physical exam or by symptoms related to the obstruction.

Types of HCM

There are several different patterns of left ventricular thickening seen in patients with HCM. These thickening patterns can often be seen using cardiac imaging techniques such as echocardiography (cardiac ultrasound) and cardiac magnetic resonance imaging (MRI).

When classifying HCM, it is important to evaluate if the hypertrophy causes obstruction or not. Patients with both obstructive and non-obstructive forms of HCM can experience similar symptoms, although the treatment of some of these symptoms may differ depending on if your HCM specialist determines the symptoms to be related to obstruction or not.

Symptoms of HCM

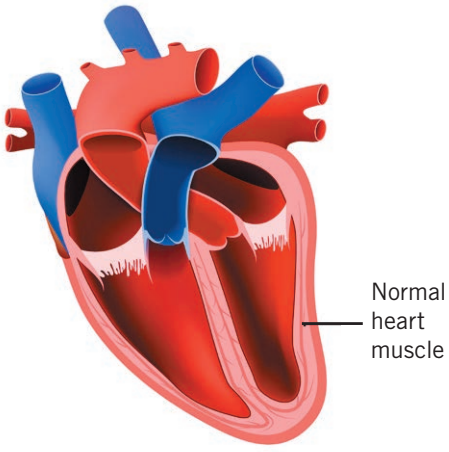
Common symptoms of HCM include dizziness, shortness of breath (especially with exercise), fatigue, palpitations, or chest pain that often occurs during exercise or after meals. Many times symptoms can vary in severity day to day. Sometimes, patients may not have any symptoms at all.

Patients can be undiagnosed for years, being told that they may have anxiety, panic attacks or asthma when they have actually had HCM. The early manifestations of HCM can be very subtle and can be overlooked unless evaluated by a team with experience looking for this condition.

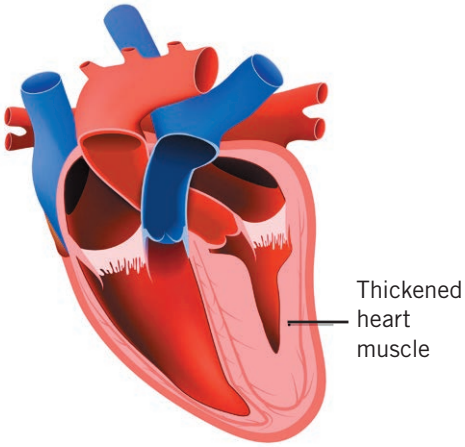
Diagnosing HCM

If you have a family history of HCM or physical exam signs or symptoms suggestive of HCM, your provider may order diagnostic testing. These tests may include:

Hypertrophic cardiomyopathy



Healthy heart



Heart with hypertrophic cardiomyopathy

- Electrocardiogram (ECG) is often performed. Electrodes are attached to your chest, arms and legs to get a quick 'snapshot' look at your heart's rhythm.
- An Echocardiogram is an ultrasound of the heart. It gives a real-time picture of your beating heart,and shows the size and structure of your heart and how well it is working.

  There are two types of echocardiograms, including the standard echocardiogram that involves taking ultrasound images from the surface of the chest. Some patients also need a transesophageal echocardiogram (TEE) that allows better evaluation of certain structures within the heart. A TEE requires an ultrasound probe placed into the esophagus and is done under sedation to optimize comfort.
- Cardiac Magnetic Resonance Imaging (cardiac MRI) uses magnets, radio waves and a computer to create moving images of your heart, including video of your heart beating, allowing for very accurate measurements of heart muscle thickness. It is often combined with an IV contrast agent, which allows for the identification of inflammation or scar tissue in the heart.
- Stress testing shows how your heart works during physical activity and can reveal problems with blood flow through your heart. It involves walking on a treadmill while your rhythm, blood pressure and heart rate are monitored by highly trained staff. If you cannot walk on a treadmill, you may be given a medication that mimics the effects of exercise.In patients with hypertrophic cardiomyopathy, the stress test can unmask problems with the blood leaving the heart which is an important cause of symptoms in patients with HCM.
- A wearable heart monitor is a small device applied to your skin like a patch. It continuously records every heartbeat as you go about daily life. This monitor makes it easier for both you and your doctor to gather the right information and determine if you have an irregular heart rhythm or arrhythmia. It can be worn for up to two weeks.
- Genetic testing involves a sample of your DNA, either with a saliva or blood test, and checking your sample for a mutation that is thought to cause HCM.

Genetic testing and family screening

Hypertrophic cardiomyopathy is usually passed down through families. A patient inherits the gene that causes HCM from one of their parents. This means that a patient with HCM has a 50% chance of passing the mutation on to each of their biological children.

Genetic testing is now available for patients with HCM and their families. We have not discovered all of the gene mutations associated with HCM, but if a patient has a positive genetic test for a gene change known to cause HCM, this can not only help confirm the diagnosis but also allow family members to be tested for this condition through a saliva or blood test.

It is important for all first degree relatives (e.g. children and siblings) of a patient with HCM to be screened with an ECG and an echocardiogram for HCM. Changes in the heart muscle for a patient at risk for developing HCM can occur at any time during a lifetime, so periodic screening is recommended for most patients with family members affected by HCM.

What does living with HCM look like?

Most patients with HCM lead normal lives with minimal or manageable symptoms and normal longevity. A subset of patients are at increased risk for atrial fibrillation, heart failure, or sudden cardiac death. If HCM is diagnosed in a timely fashion, these patients can be identified early and treated to reduce their risks for these conditions.

Atrial fibrillation

The right and left top chambers of the heart are called the atria. The bottom right and left chambers act as the pump of the heart and are called the ventricles.Normally, an electrical impulse starts in the atria and travels to the ventricles. This impulse signals the heart muscle

to contract regularly. The blood is then pumped out of the heart to the rest of the body.

Atrial fibrillation occurs when there are multiple impulses firing in the atria, which causes them to quiver rapidly in a disorganized pattern. The heart is no longer able to follow the normal electrical pattern when this occurs. The ventricles try to follow the erratic beats of the atria, which leads to an irregular heartbeat. This heartbeat can occur at a faster, slower, or even at a normal heart rate.

Atrial fibrillation is the most common arrhythmia (abnormal heart rhythm) associated with HCM. While typically not life-threatening, atrial fibrillation increases the risk of stroke and heart failure. Many patients with HCM do not feel well when their heart is in atrial fibrillation and require additional medications or interventions to restore the heart's electrical rhythm to normal. Patients with HCM have an increased risk of stroke compared to other patients with atrial fibrillation and all patients with HCM should be on medications to thin the blood and reduce the risk of blood clots and stroke.

Heart failure

Heart failure refers to symptoms caused by the heart not being able to adequately pump blood out of the left ventricle to the rest of the body.

In patients with HCM, symptoms such as fatigue, shortness of breath, chest pain, or dizziness may occur. Medications to reduce the intensity of the heart contraction can often help with these symptoms. Sometimes surgical interventions to reduce the thickening of the heart muscle can cure the obstruction to relieve the symptoms of heart failure.

Sudden death

This is frequently due to dangerously fast abnormal heart rhythms (arrhythmias). Your HCM cardiologist will screen for these regularly with rhythm monitors and may obtain additional imaging to evaluate your risk of having abnormal rhythms. If your risk of having sudden cardiac death is elevated, your physician may refer you to a cardiac electrophysiologist to discuss interventions that can help prevent sudden death. A cardiac electrophysiologist specializes in heart rhythms, pacemakers, and defibrillators.

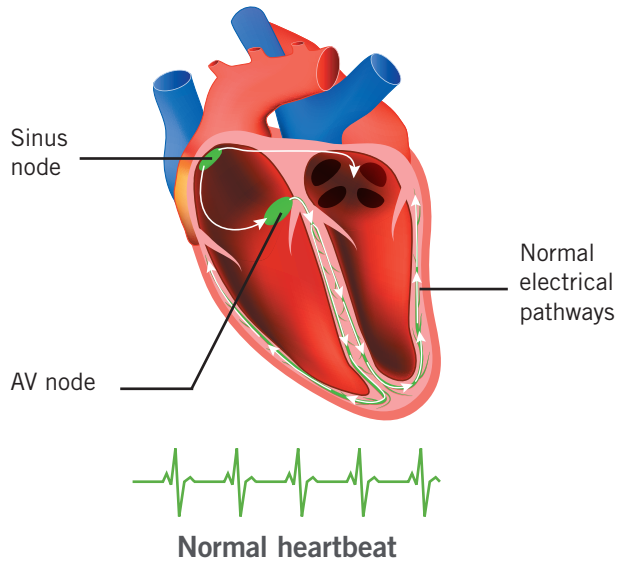
Treatment options for HCM

Medications

Beta blockers

These medications are often prescribed to slow the heart rate, treat arrhythmia and also reduce the degree of exercise associated left ventricular outflow tract obstructions.

Atrial fibrillation



Calcium channel blockers
Often prescribed as an alternative or second line agent to beta blockers. Calcium channel blockers can help reduce the degree of left ventricular outflow tract obstruction at rest, and with exercise in obstructive HCM.

Cardiac myosin inhibitors
FDA-approved medication specifically designed to treat more severe symptoms related to obstructive hypertrophic cardiomyopathy. This medication is prescribed through a special drug and safety program through our HCM center in patients.

Disopyramide
This is an antiarrhythmic medication that can help treat irregular heart rhythms, but also reduce the degree of left ventricular outflow tract obstruction if beta blockers and/or calcium channel blockers are not effective.

Anticoagulants
Often referred to as 'blood thinners'. Anticoagulants help thin the blood to make it less likely to clot, thereby reducing the risk of stroke.

Catheter ablation for arrhythmias

Some arrhythmias can be treated effectively with a minimally invasive procedure called a catheter ablation. An ablation catheter is then used to burn or freeze a tiny amount of targeted heart tissue to prevent abnormal electrical signals from moving through the heart. Arrhythmias including atrial fibrillation and ventricular tachycardia may be treated this way.

Implantable cardiac defibrillator

A defibrillator (ICD) is a battery powered device placed under the skin that keeps track of your heart rate. Thin wires connect the ICD to your heart. If a dangerous abnormal heart rhythm is detected, the device will deliver an electric shock to restore a normal heartbeat if your heart is beating chaotically and/or too fast.

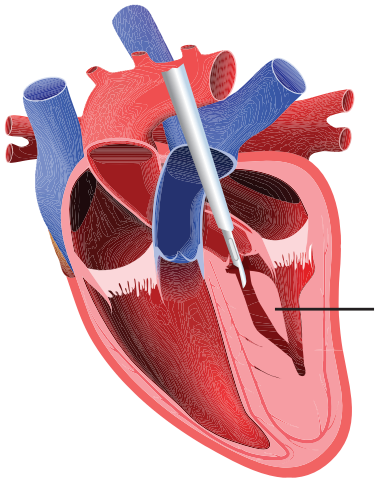
Septal reduction therapy

Some patients have life-impacting symptoms related to left ventricular obstruction despite taking medications. These patients may benefit from septal reduction therapy, which includes both a surgical procedure (septal myectomy) or a catheter based procedure (alcohol septal ablation).

A surgical myectomy is an open heart surgery that involves careful surgical removal of the thickened heart muscle causing the obstruction. This effectively is a cure for left ventricular obstruction. With careful patient selection, this surgery can be very safe and provide life-long benefits.

For patients who are not candidates for open-heart surgery, or prefer a minimally invasive procedure, alcohol septal ablation may be an option. Catheters are placed into the heart through a small incision at the wrist or groin.Your doctor then injects alcohol through the catheter and into the area where the heart muscle is too thick. The alcohol is toxic and causes some of the thickened heart muscle cells to shrink and die. The remaining scar tissue is thinner than the heart muscle, which will improve blood flow through your heart and out to your body.

Septal myectomy



For more information, contact your cardiologist or go to:

vmfh.org/cardiology

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