

HYPERTROPHIC CARDIOMYOPATHY AND ALCOHOL SEPTAL ABLATION

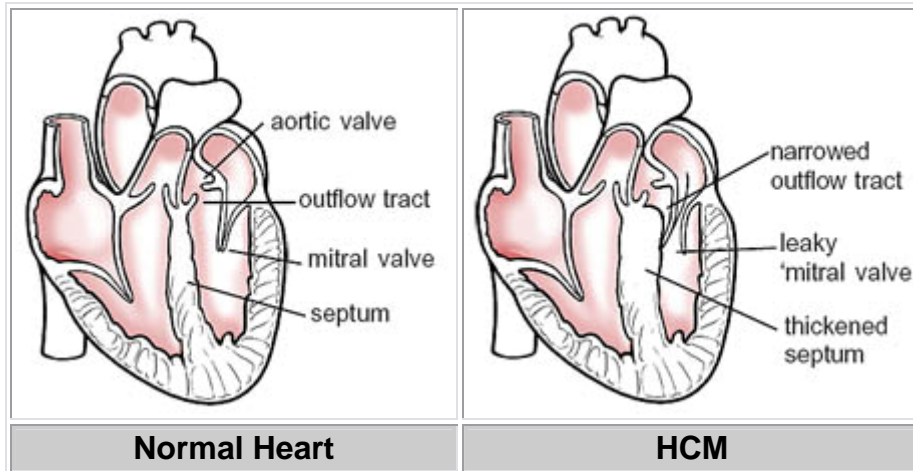
What is hypertrophic cardiomyopathy (HCM)?

Hypertrophic cardiomyopathy (HCM) is a complex type of heart disease marked by thickening of the heart muscle, left ventricular stiffness, mitral valve changes and cellular changes.

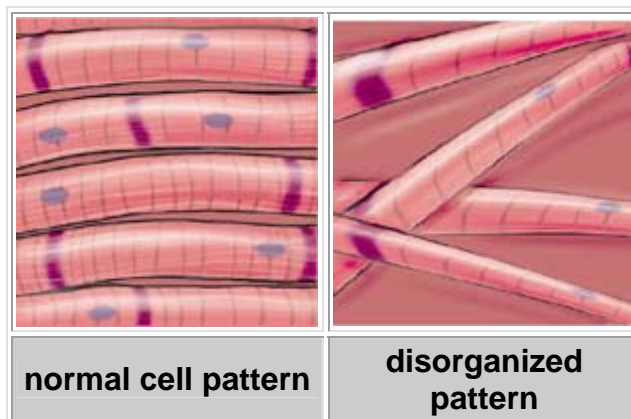
- **Thickening of the heart muscle (myocardium)** occurs most commonly at the septum, just below the aortic valve. The septum is the muscular wall that separates the left and right side of the heart. Problems occur when the septum between the heart's lower chambers or ventricles is thickened. The thickened septum may cause a narrowing that can block or reduce the blood flow from the left ventricle to the aorta - a condition called "outflow tract obstruction." The ventricles must pump much harder to overcome the narrowing or blockage. This type of hypertrophic cardiomyopathy may be called:
 - Hypertrophic obstructive cardiomyopathy (HOCM)
 - Idiopathic hypertrophic subaortic stenosis (IHSS)
 - Hypertrophic Cardiomyopathy (HCM)

Hypertrophic cardiomyopathy may also cause thickening in other parts of the heart muscle, such as the bottom of the heart called the apex, right ventricle, or throughout the entire left ventricle.

- **Stiffness in the left ventricle** is a sign of hypertrophic cardiomyopathy that occurs as a result of the thickened septum. The left ventricle also is unable to normally relax and fill with blood. The stiffness in the left ventricle causes pressure to increase inside the heart.
- **Mitral valve changes:** When the outflow tract is narrowed, blood rushes through the passageway toward the aortic valve (like a tight garden hose nozzle), dragging the leaflets of the mitral valve with it. The mitral valve normally functions to keep blood flowing in one direction from the left atrium (upper heart chamber) to the left ventricle. However, the increased force of blood caused by hypertrophic cardiomyopathy pulls the valve open and may cause blood to leak backward (called regurgitation) into the left atrium.



- Cellular changes** or changes in the cells of the heart muscle cells occur with hypertrophic cardiomyopathy. Through a microscope, the cells appear disorganized and irregular (called "disarray") instead of being organized and parallel. This disarray may impact the electrical signals traveling through the heart and contribute to ventricular arrhythmia (a type of abnormal heart rhythm).



Who is affected by hypertrophic cardiomyopathy?

HCM affects an estimated 600,000 to 1.5 million Americans, or one in 500 people. It is more prevalent than multiple sclerosis, which affects one in 700 people.

HCM is the most common cause of sudden cardiac death in people under age 30. HCM may be best known for its role in cardiac arrest and subsequent death in some young professional athletes.

What causes HCM?

HCM can be:

- **Inherited:** HCM may be inherited, caused by an abnormality in the gene that codes the characteristics for the heart muscle. When the gene defect is present, the type of HCM that develops varies greatly within the family. In addition, some people who have the HCM gene may never develop the disease.
- **Acquired:** HCM may be acquired, the result of high blood pressure or aging.
- **Unknown:** In other instances, the cause of HCM is unknown.

Because the cause of HCM varies, it is frequently difficult to identify a high-risk population.

What are the symptoms of HCM?

Many people with HCM have no symptoms or only minor symptoms, and live a normal life. Other people develop symptoms, which progress and worsen as the heart function worsens. Symptoms can occur at any age and may include:

- **Chest pain or pressure** that usually occurs with exercise or physical activity, but also may occur with rest or after meals.
- **Shortness of breath (dyspnea) and fatigue**, especially with exertion. These symptoms are more common in adults with HCM and are most likely caused by backup of pressure in the left atrium and to the lungs.
- **Syncope (fainting or passing out)** affects about 15 to 25 percent of HCM patients. Syncope with HCM may be caused by irregular heart rhythms, abnormal responses of the blood vessels during exercise, or no cause may be found.
- **Palpitations (fluttering in the chest)** due to abnormal heart rhythms (arrhythmias), such as atrial fibrillation or ventricular tachycardia. Atrial fibrillation occurs in about 25 percent of those with HCM, and increases risk for blood clots and heart failure.
- **Sudden death** occurs in a small number of patients with HCM.

How is HCM diagnosed?

The diagnosis of HCM is based on:

Medical history: Your doctor will ask you many questions about your symptoms and family history.

Physical exam: Your doctor will listen to your heart and lungs. Patients with severe hypertrophic obstructive cardiomyopathy (HOCM, or outflow tract obstruction) may have a heart murmur.

Tests: Echocardiogram is the most common test used to diagnose HCM, as the characteristic thickening of the heart walls is usually visible on the echo. Other tests may include blood tests, electrocardiogram, chest x-ray, echocardiogram, exercise stress test, cardiac catheterization and magnetic resonance imaging (MRI).

How is Hypertrophic Cardiomyopathy treated and What is Alcohol Septal Ablation

In the past there have been three ways to treat hypertrophic cardiomyopathy:

- 1) Medications
- 2) Pacemaker
- 3) Surgery

Now there is a fourth treatment option available which is called alcohol septal ablation.

Medications are usually given to slow the heart rate and increase the filling of the heart, which helps alleviate some of the obstruction and relax the heart. These medications include beta-blockers, calcium channel blockers, and other drugs such as disopyrimide. In many patients the symptoms are well-controlled on medications. However, some patients have symptoms that are refractory to medical therapy.

In the last ten years, much attention has been given to placing pacemakers in patients with hypertrophic cardiomyopathy. In this particular case, the pacemaker is not used to treat a slow or abnormal heartbeat, which is the usual indication for a pacemaker. It is thought that the pacemaker may change the pattern of contraction of the heart and may help alleviate the outflow tract obstruction. There is controversy over whether or not this is an effective treatment.

Until recently surgery has been the primary way to treat hypertrophic cardiomyopathy. This is an open chest, open heart procedure where the surgeon makes an incision into the heart and cuts out the area of overgrown heart muscle. This is a very effective procedure, with an operative mortality of less than 5%, and is quite effective in reducing symptoms once the patient has healed from the operation. Now there is a new treatment option available called alcohol septal ablation.

The procedure itself involves injecting 100% absolute alcohol into one of the branches of a heart artery that goes to the enlarged septum. Patients are routinely sedated during the procedure. By injecting the alcohol the heart muscle cells are killed. The result is that the thickened heart muscle which has been killed is replaced with thin scar tissue, and thus the obstruction to blood flow out of the left

ventricle is reduced. This also appears to improve the mitral valve abnormalities. It is effective in reducing the left ventricular gradient.

To perform this procedure an interventional cardiologist must perform a cardiac catheterization procedure and identify the anatomy of all the coronary arteries. Next the artery which supplies the septum is blocked with a balloon and alcohol is injected into the artery. The patient is constantly monitored with x-ray fluoroscopy and with echocardiography. The alcohol is left in the artery for few minutes, and then the balloon is removed and follow-up pictures are taken.

The alcohol septal ablation procedure essentially creates a heart attack in the area of the enlarged heart muscle. A scar replaces the thick heart muscle with thin tissue. However, this all occurs due to the injury to the heart muscle that is created by the alcohol injection. Patients will begin to feel better almost immediately, and symptoms will continue to improve over the next several weeks and months.

Typically, patients are kept in the hospital for three to five days following the procedure so that they can be monitored. The most common side effect of this procedure is an irregular heart rhythm called "complete heart block". The electrical system that controls the heart beat travels through the septum of the heart. Therefore when alcohol injures this area, it can also injure the electrical system of the heart, and cause the patient to have an extremely slow heart beat. This often requires placement of a permanent pacemaker to keep the heart rate from being too slow. The patient is monitored in the ICU for several days following the procedure. If the patient has complete heart block then the patient remains in the hospital until a permanent pacemaker can be implanted. All patients having the alcohol ablation must be agreeable to having a pacemaker placed if indicated.

Patients will begin to feel better almost immediately and will usually experience less chest pain, less shortness of breath, less edema, and better exercise tolerance. Symptoms will continue to improve over the next weeks and months and often patients can reduce or discontinue their medications. Patients should be followed by their interventional cardiologist as well as their primary care physician for the next several years. Echocardiograms to evaluate the healing of the septum, the left ventricular gradient, and the mitral valve should be done at three months after the procedure and then yearly.

Most patients have a successful procedure. In some patients the procedure is only partially successful and may need to be repeated. Other potential complications of the procedure include heart attack, irregular heart rhythms, infection, bleeding, allergic reaction to the x-ray dye, kidney failure from the dye, cardiac perforation, need for emergency surgery, and ineffective procedure. All of these complications are extremely rare.