

HYPERTROPHIC OBSTRUCTIVE CARDIOMYOPATHY



**EDITH AND BENSON FORD
HEART & VASCULAR INSTITUTE**

WHAT IS HYPERTROPHIC OBSTRUCTIVE CARDIOMYOPATHY?

THE HEART AND HEART MUSCLE

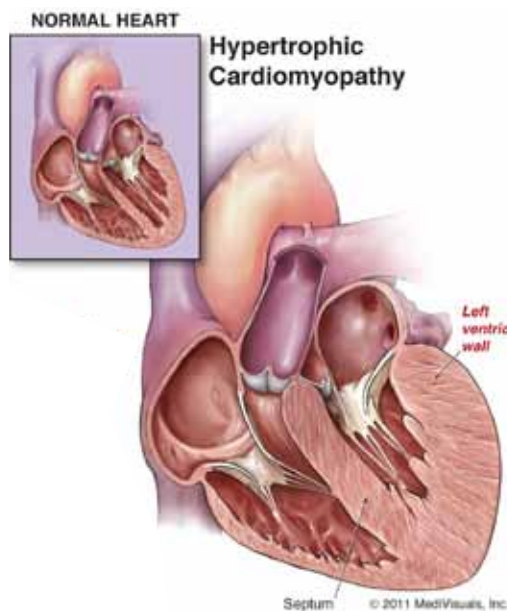
The heart is an organ with two sides, right and left, separated by a wall called the septum. Each side is divided into an upper chamber (atrium) and a lower chamber (ventricle). The walls between the chambers, and the outer walls of the heart, are all made of muscle. When this muscle becomes abnormally thick or bulky, it is known as cardiomyopathy (cardio = heart, myo = muscle and pathy = abnormality). There are several types of cardiomyopathy, one of which is hypertrophic obstructive cardiomyopathy.

CAUSES

Hypertrophic obstructive cardiomyopathy is a genetic condition, although not every family member will develop the disease. There are many genes that code for heart muscle development so there can be various forms of inherited hypertrophic cardiomyopathy. A careful family history is important to determine if genetic counseling is needed.



HYPERTROPHIC OBSTRUCTIVE CARDIOMYOPATHY (HOCM)



In a hypertrophic heart, there is a thickening of some or all of the walls of the heart. The ventricular septum is the inner wall that divides the left and right lower chambers (ventricles). When this wall grows so thick that it intrudes into the passage in which blood flows out of the heart, called the “outflow tract,” it is known as hypertrophic obstructive cardiomyopathy. In this disease, four changes occur:

1. The upper septum thickens. This creates an obstruction that reduces the flow of blood out of the heart and into the body.

2. The muscle cells become disorganized.

When normal heart cells are looked at under a microscope, they are organized like bricks in a wall. In a thickened muscle, the cells are disorganized and chaotic. This disorganized pattern can disrupt the heart’s rhythm resulting in irregular beats.



3. The left ventricle becomes stiff. The disorganized cells prevent the muscle from relaxing and stretching, reducing the amount

of blood that can fill the heart. When less blood is pumped forward into the body, blood can back up all the way into the lungs, where it can lead to breathing problems.

4. The mitral valve function changes. This one-way valve is located between the atrium and ventricle on the left side of the heart. It is made up of two flaps called leaflets, both of which flip open to let blood pass into the left ventricle, and then flip back closed, creating a tight seal. A thickened septum can cause the mitral valve to change shape as it opens and closes. This shape change can result in one of the valve leaflets to be sucked into the outflow tract. This creates a partial blockage on both sides of the passage, the thickened septum on one side and the valve leaflet on the other. This can make it harder for blood to be pumped out of the heart.

The blockage of the outflow tract results in symptoms of shortness of breath, chest pain, fainting and fatigue that occurs with HOCM.

HOW IS HOCM DIAGNOSED?

SYMPTOMS

Each person with HOCM will have different symptoms, which may occur relatively suddenly or develop so slowly that they are not recognized at all and the



individuals unknowingly limit their lifestyle based on how they feel. Symptoms may include:

- Shortness of breath and fatigue that limit activity. Fatigue may occur with or without breathing difficulties
- Chest discomfort, or angina that usually occurs with activity
- Heart palpitations, or an uncomfortable awareness that there are extra or skipped heart beats
- Lightheadedness, dizziness, fainting or passing out may be due to an irregular heart rhythm, changes in blood pressure or blood flow with activity

DIAGNOSIS

The earliest indicators that HOCM may be present are symptoms, changes on an electrocardiogram (ECG) or a unique heart murmur found by your health care provider. You will then be referred to a cardiologist for a complete evaluation to include:

- A complete history of your symptoms and family health history looking for premature death or heart problems.
- A complete physical to assess for special heart murmurs.
- Electrocardiogram (ECG) to evaluate how the electrical currents are traveling in the heart and shows the heart's rhythm.
- Echocardiogram (Echo) and Doppler ultrasound which use sound waves to view the heart, including showing blood flow, movement of the walls and valves, and thickness of the walls.
- Magnetic Resonance Imaging (MRI) which is a noninvasive technique used to gain a detailed image of your heart's structure and movements.
- Cardiac Catheterization which is a minimally invasive test that measures pressures in your heart and looks at the blood flow in the vessels that feed your heart. Thin tubes called catheters are inserted through major vessels (in either your wrist or groin area), and then threaded up to your heart. The pressures and oxygen content in different chambers of the heart and aorta are measured to define how hard the heart is working due to the thickened wall. The blood flow in the vessels are looked at to see if any blockages may be causing your symptoms.



HOCM AND SUDDEN CARDIAC DEATH

There is a small group of people who are at high risk for sudden cardiac death from HOCM. There are markers which help identify patients that may die prematurely:

- Family history of sudden or unexpected death
- Young patients who have several episodes of fainting
- Abnormal blood pressure response to exercise
- A history of an abnormal heart rhythm with a fast rate
- Severe symptoms with poor heart function
- Excessive thickness of the heart muscle

If you have two or more of these risk factors, your cardiologist may recommend special heart rhythm medications or an implantable cardiac defibrillator (ICD). This device continually monitors the heart and delivers an electrical shock when needed to restore a normal heart rhythm.

HOW IS HOCM MANAGED AND TREATED?

TREATMENT OVERVIEW

Treatment focuses on relieving heart function, managing symptoms and preventing complications. It is important that a cardiologist monitors everyone with HOCM.

MEDICAL MANAGEMENT

Medications are the main treatment used to control symptoms and can include:

- **Beta blockers (metoprolol, atenolol or carvedilol):** Slow the heart beat, reduce the force of the contraction and may decrease the obstruction during exercise
- **Calcium channel blockers:** Help relax the heart and improve the filling of the lower chambers
- **Heart rhythm drugs:** Some patients need drugs to prevent or reduce abnormal heart rhythms called ventricular tachycardia or atrial fibrillation
- **Blood thinners:** If an irregular heart rhythm called atrial fibrillation is present, blood thinners may be used to prevent strokes that can occur when blood clots form in the heart, break loose and travel to the brain.



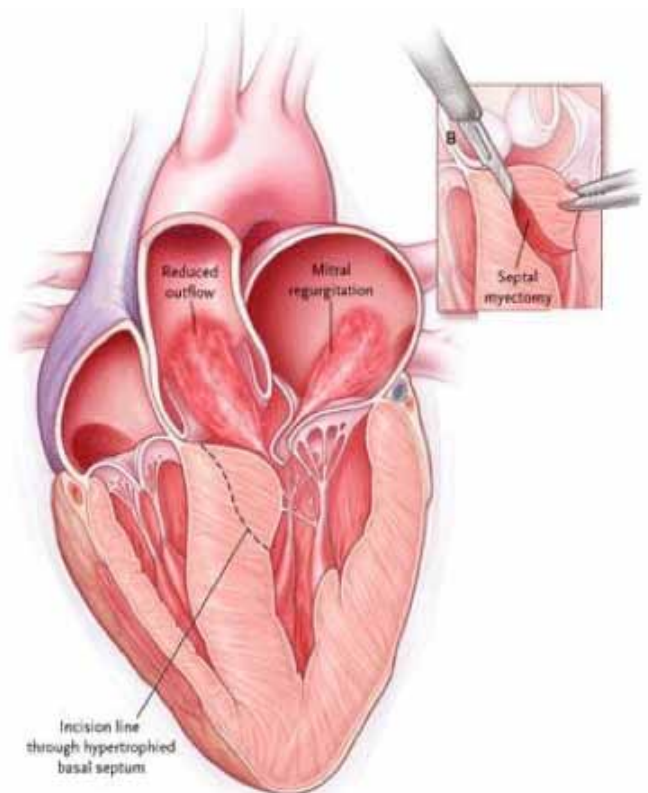
In addition, antibiotics may be recommended before routine procedures such as dental work, which can introduce bacteria into the bloodstream that can lead to heart valve infection.

SEPTAL REDUCTION THERAPY: SURGICAL MYECTOMY

When blood flow out of the heart is severely blocked, an open-heart operation called surgical myectomy may be done. This open-heart procedure involves cutting and removing a portion of the thickened part of the septum.

- Before the procedure begins, you are put completely to sleep by anesthesia.
- During the procedure your heart is stopped and a heart-lung bypass machine is used to do all the work of your heart and lungs.
- The surgeon approaches the septum through the aortic valve, which controls the flow of blood leaving the heart.
- The surgeon cuts and removes a portion of the thickened septum, which will heal on its own without stitches.
- Your heart is restarted and a breathing machine is used until all anesthesia has worn off.
- The surgery usually requires staying in the hospital for five to seven days, a period of recovery at home and exercise rehabilitation therapy.

Patients who have this procedure often show significant improvement. If the heart's mitral valve is leaking, an additional surgery may be recommended to repair or replace the valve.

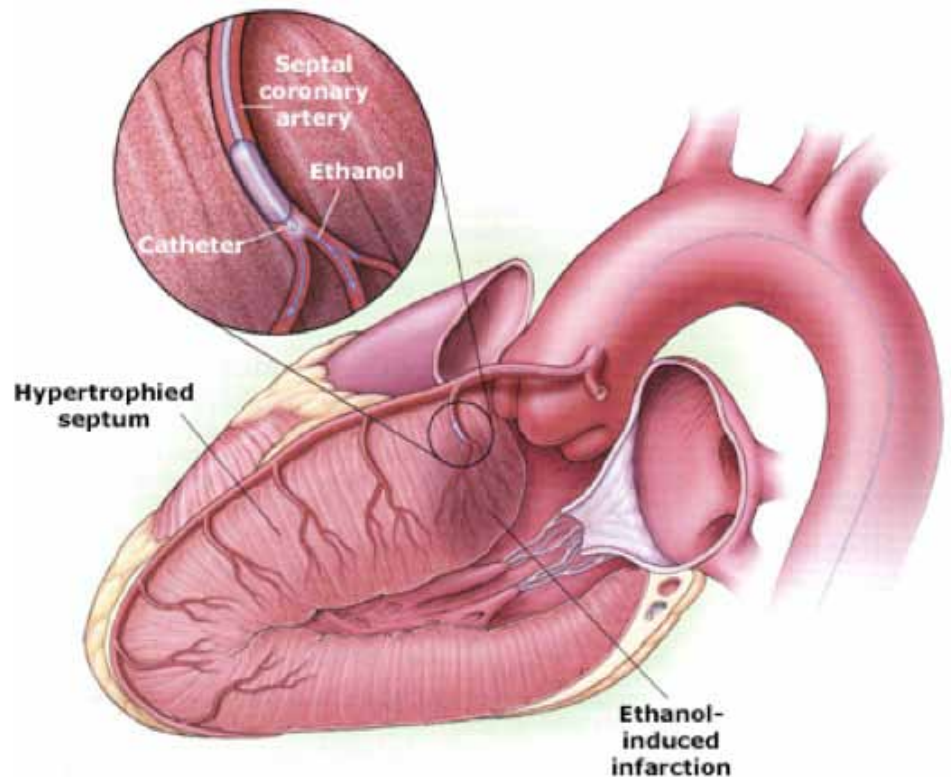


Continued on next page

SEPTAL REDUCTION THERAPY: ALCOHOL SEPTAL ABLATION

In some cases, a minimally invasive technique known as alcohol septal ablation may be used. This is a cardiac catheterization procedure. In this procedure, thin tubes known as catheters are inserted into the major vessels in the groin area and threaded up to the heart. One of these is used to deliver pure alcohol to the thickened part of the septum. The alcohol is toxic to the heart muscle and essentially causes a controlled heart attack. In the months following the procedure, the damaged muscle shrinks back to a normal size, widening the passage used to pump blood out of the heart.

- During the procedure, you are awake but medicines are used to help you relax and for pain.
- A temporary pacemaker wire is placed in the heart through a catheter inserted in a major vein in either the groin, neck or arm
- A dye injection is used to locate the small arteries that supply blood to the septum.
- A balloon-tipped catheter is inserted into one of these arteries and inflated, blocking the artery, and the alcohol is injected beyond the balloon.
- The balloon is deflated and removed.
- The procedure generally involves a hospital stay of about three days.



For more information, visit henryford.com/heart or call (313) 916-1534.



henryford.com/heart