Hypertrophic Cardiomyopathy:

What healthcare providers need to know

PCNA
PREVENTIVE CARDIOVASCULAR NURSES ASSOCIATION

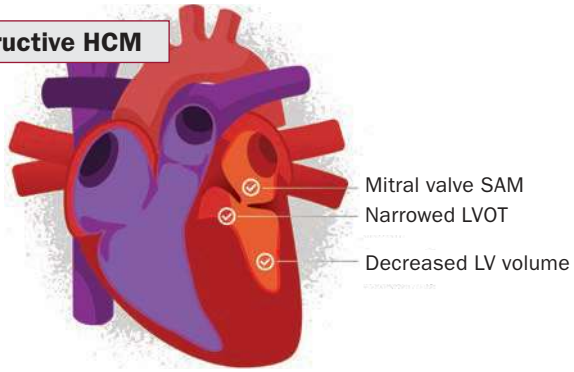
What Is Hypertrophic Cardiomyopathy (HCM)?

HCM is a chronic disease in which the left ventricle (LV) becomes abnormally thick (hypertrophied) in the absence of any underlying health condition. Primarily genetic, but some instances are idiopathic (genesis is unknown).

HCM occurs in approximately 1:500 people of all ages and races.

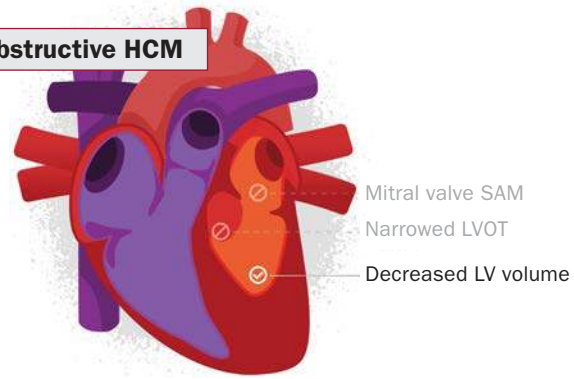
There are diverse clinical presentations of the two subtypes: Obstructive (more common) and Nonobstructive. In both cases, heart muscle will be thickened and stiff.

Obstructive HCM



Obstructive HCM (HOCM) includes reduced or obstructed blood flow from the LV to the aorta, known as left ventricular outflow tract (LVOT) obstruction. This is due to septal hypertrophy and may also involve the narrowing of the systolic anterior motion (SAM) of the mitral valve.

Nonobstructive HCM



In Nonobstructive HCM, the thickened heart muscle does not impede blood flow out of the LV. The hypertrophy is distributed in a variety of ways, and diastolic impairment is a common manifestation.

Common Symptoms of HCM

Patients with HCM may be asymptomatic. Those experiencing symptoms may report variations based on hydration, blood pressure and environment (temperature and humidity). It is important to rule out coronary artery disease (CAD) and exercise-induced asthma, which present with similar symptoms. The following HCM symptoms may occur independently, or in the presence of tachycardia or heart failure (HF):

- Dyspnea (shortness of breath)
- Angina pectoris (chest pain)
- Exercise intolerance
- Palpitations
- Dizziness and/or syncope (fainting)
- Peripheral edema

HCM Complications

HCM can lead to other cardiac conditions and affect a patient's physical and physiological quality of life. Complications may include:

- Arrhythmias (e.g., atrial fibrillation [AFib], ventricular arrhythmia [VA])
- Stroke
- HF

Many patients change their lifestyle to accommodate the disease and side effects of prescribed medications.

Why HCM Diagnosis Is Difficult

- Non-specific symptoms
- May be masked by, or be similar to, other comorbid conditions including: hypertension, obesity, diabetes, obstructive sleep apnea, asthma, or athletic remodeling
- Heterogeneous symptomology; clinical presentation and progression of disease
- Lack of standardized clinical measures to track HCM symptoms and progression
- Difficulty/challenges in imaging (echocardiogram, cardiac magnetic resonance [CMR]) and measuring changes in structure and function that are characteristics of HCM (wall thickness, SAM, LVOT)

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HCM Diagnostic Tools

Sometimes obstruction is not evident when a patient is at rest, but is evident when under activity/at stress. Genetic testing may be used to confirm HCM diagnosis.

HCM Diagnosis

Diagnostic Tool*	Potential Results
Medical history	Symptoms and/or conditions associated with HCM (see previous page)
Family history	Family member with sudden, unexplained cardiac death, known arrhythmia, HF
Cardiac auscultation	Heart murmur, irregular rhythm
Electrocardiogram: static or ambulatory (Holter)	Atrial tachycardia (AT) or ventricular tachycardia (VT), irregular rhythm
Echocardiogram	Thickening of the LV walls and septum, decreased LV volume, LVOT obstruction, SAM of the mitral valve
Stress testing	Reduced functional capacity; symptoms of HCM with activity; abnormal blood pressure response
Genetic testing	Gene mutation; familial inheritance

*Additional cardiac diagnostics, labs and imaging may be performed to evaluate symptoms, measure functional capacity and stratify risk. These may include cardiac angiography, positron emission tomography (PET) and cardiac MRI (cardiovascular magnetic resonance [CMR]).

HCM Management

Management is individualized according to patient symptoms, age, activity level, and the presence of LVOT obstruction, HF or any arrhythmia. The goal is to prevent or reduce symptoms and risk of complications. Current management and treatment options may include lifelong lifestyle modifications and pharmacotherapies. For patients with cardiac dysfunction and more advanced disease, surgical techniques such as implantable cardioverter-defibrillator (ICD), myectomy/alcohol septal ablation (ASA), or heart transplant may be warranted.

Screening and Surveillance

Echocardiogram screening for HCM is recommended for all first-degree family members, those diagnosed with HCM, or those who had a sudden, unexplained cardiac death.

Regular surveillance is essential to monitor symptoms (worsening or improving), evaluate heart function (including patient-reported changes in behavior/activity), support healthy lifestyle habits, and evaluate the need for and/or response to medications.

HCM Resources

For healthcare providers

- 2011 ACCF/AHA Guidelines for the Diagnosis and Treatment of Hypertrophic Cardiomyopathy
  https://professional.heart.org/professional/ScienceNews/UCM_433410_Hypertrophic-Cardiomyopathy-Guidelines.jsp
- 2014 ESC Guidelines on the Diagnosis and Management of Hypertrophic Cardiomyopathy
  escardio.org/Guidelines/Clinical-Practice-Guidelines/Hypertrophic-Cardiomyopathy
- SCD and ICD Risk Calculator (Ages 17-79) HCM Risk-SCD
  https://doc2do.com/hcm/webHCM.html

For patients and families

- HCM Association, **4hcm.org.** Improving the lives of those with HCM through support, education, advocacy and research.

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REFERENCES:

- Epstein AE, DiMarco JP, Ellenbogen KA, et al. ACC/AHA/HRS 2008 Guidelines for Device-Based Therapy of Cardiac Rhythm Abnormalities. *Circulation*. 2008;117;e350-e408. doi:10.1161/CIRCUALTIONAHA.108.189742.
- Gersh BJ, Maron BJ, Bonow RO, et al. 2011 ACCF/AHA Guideline for the Diagnosis and Treatment of Hypertrophic Cardiomyopathy. *Circulation*. 2011;124:e783-e831. doi:10.1161/CIR.0b013e318223e2bd.
- Maron BJ, Edelson JE, Bonow RO, et al., Eligibility and Disqualification Recommendations for Competitive Athletes With Cardiovascular Abnormalities: Task Force 3: Hypertrophic Cardiomyopathy, Arrhythmogenic Right Ventricular Cardiomyopathy and Other Cardiomyopathies, and Myocarditis: A Scientific Statement from the American Heart Association and American College of Cardiology. *Circulation*. 2015;132(22):e273-80. doi: 10.1161/CIR. 0000000000000239.