

Hypertrophic



Illustration by Science Picture Co/Getty Images ©

cardiomyopathy

A clinical and genetic update

Abstract: This article presents an overview of hypertrophic cardiomyopathy (HCM) and the associated clinical findings, treatment, and management for nurse practitioners. Topics include an overview of the condition, major and minor criteria for diagnosis, clinical course and pathophysiology, genetic inheritance and testing, and dysrhythmias associated with HCM.

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Hypertrophic cardiomyopathy (HCM) is a heterogeneous cardiac disease with varied clinical manifestations, age of onset, pattern and extent of left ventricular hypertrophy (LVH), degree of obstruction, and risk for sudden cardiac death (SCD). Sudden deaths in young competitive athletes are highly visible and catastrophic events. They have become a source of controversy as to whether prescreening of athletes should be performed. Recently, Maron and colleagues examined SCD across 1,866 young athletes in the United States from 1980 to 2006.¹ Sudden deaths were primarily due to cardiovascular disease (n = 1049 [56%]); however, causes also included blunt trauma that resulted in structural damage (n = 416 [22%]), commotio cordis (n = 65 [3%]), and heat stroke (n = 46 [2%]). The most common cardiovascular causes were HCM (36%) and congenital coronary artery anomalies (17%).¹ According to Maron and colleagues, in the national registry, the absolute number of cardiovascular sudden deaths in

Key words: clinical care, genetics, hypertrophic cardiomyopathy, inherited heart disorder

young U.S. athletes was considered low at a rate of less than 100 per year. The data are relevant to the current debate concerning the issue of preparticipation ECG sport screening programs and emphasize the need for mandatory reporting of sudden deaths in athletes to a national registry.¹

HCM is the most common inherited heart condition affecting 1 in 500 individuals in the general population.^{2,3} It is characterized by a thickened left ventricular heart muscle and most frequently involves the interventricular septum, although both ventricles of the heart may be involved.² One recognizable trait of HCM on cardiac imaging is LVH of the heart, usually greater than or equal to 15 mm, and with no underlying or coexisting conditions that could account for the thickening. (See *A comparison of the normal heart structures and HCM*.) This can result in an obstruction of blood flow in the myocardium vasculature and blood flow through the heart chambers, changing the heart's overall contractility and metabolic function over time. The results of HCM are diverse and may include left ventricular outflow tract obstruction, mitral regurgitation, diastolic dysfunction, myocardial ischemia, and cardiac dysrhythmias.²⁻⁴ Treatment and management strategies are directed

at symptom relief and risk stratification for the prevention of SCD.^{5,6}

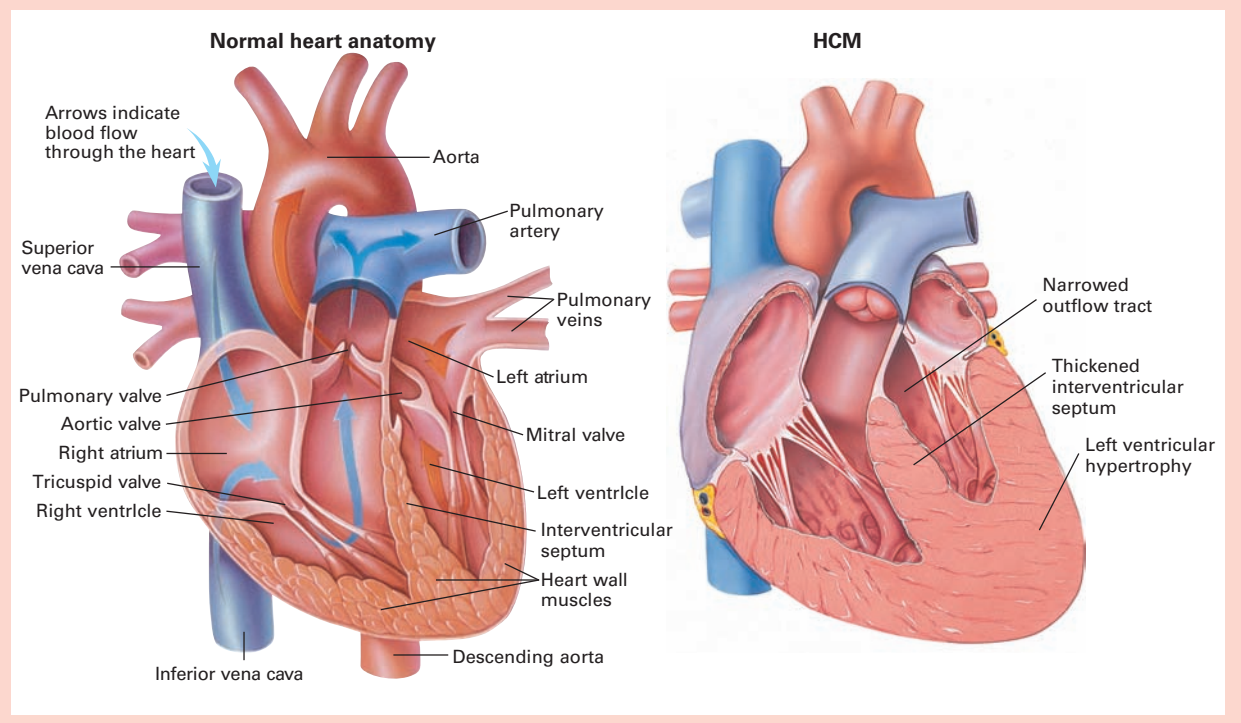
The purpose of this article is to provide nurse practitioners (NPs) with a general overview of HCM and its associated clinical presentation, genetic findings, treatments, and management.

Prevalence, symptoms, and clinical findings

HCM affects all age and ethnic groups and is a major cause of SCD in those under 50.^{2,3} Many individuals with HCM are unaware they have it until they experience symptoms, suffer a cardiac event, or have a family relative diagnosed with the condition. Other individuals may not develop symptoms until late in life or not at all. For example, even within the same family, individuals with the same genotype may have varying onset, symptoms, and clinical manifestations of HCM (phenotype). Thus, it is critical to recognize those who remain silently at risk within a family and provide proper screening, follow-up, education, and treatment as indicated. Most individuals living with a diagnosis of HCM achieve a normal life expectancy, although some experience significant symptoms, and consequently, undergo medical

A comparison of the normal heart structures and HCM

On the left is a normal septum thickness without chamber dilatation. On the right is an image of a heart showing HCM with extreme septal thickness, noted in both the right and left ventricle.



Source: The Anatomical Chart Company.

or surgical management to alleviate symptoms and improve quality of life and survival.^{3,5-7} Therefore, the clinical heterogeneity of HCM calls for individualized treatment. For those with severe symptoms of HCM, the treatment and management will be focused on alleviating symptoms and improving quality of life. In other individuals without symptoms, an approach of vigilant monitoring with regular echocardiograms (ECHOs) and clinical evaluations may be appropriate.^{2,3,5}

■ Risk stratification

The following criteria can be used by the NP to guide risk stratification.⁸ Patients with a prior cardiac arrest or spontaneous sustained ventricular tachycardia (VT) are at high risk and should be considered as candidates for prophylactic implantable cardioverter defibrillator (ICD) therapy.⁴ In the absence of such a history, risk is probably best assessed on the basis of the total number of the following risk factors:

- Maximum LV wall thickness greater than 30 mm.
- Systolic BP response, measured at 1-minute interval during maximal upright exercise in patients 40 years of age. An abnormal BP response is generally failure to increase by greater than 25 mm Hg or a fall from peak values during continued exercise of greater than 15 mm Hg.
- Nonsustained VT (NSVT) during ambulatory hour ECG monitoring.
- A history of at least one SCD in a relative before the age of 45 years, together with a history of syncope.
- A resting peak LV outflow tract gradient of greater than 30 mm Hg.

Patients with two or more risk factors are considered high risk and should be further evaluated and considered for prophylactic therapy with an ICD.^{2,8}

■ Common symptoms of HCM

Patients may experience symptoms of HCM, including dyspnea, angina, syncope, and palpitations.² Dyspnea, or shortness of breath, is the most commonly reported symptom of HCM in otherwise young, healthy adults without any underlying cardiac conditions.² This symptom is often associated with fatigue, limiting lifestyle or exercise capacity, and with some patients reporting that they have experienced severe shortness of breath while lying supine. Angina, or chest pain or pressure, is another common symptom of HCM, usually occurring after exertion, such as running or climbing stairs. Chest pain results from the substantial oxygen demands of thickened heart muscle, which impedes the circulation of blood and oxygen in the heart. Syncope,

or temporary loss of consciousness, may be caused by a drop in BP, which occurs in about 15% to 25% of HCM patients; septal hypertrophy that obstructs and distorts the outflow tract may be present and may result in an underlying dysrhythmia, such as VT.^{3,9}

Another common symptom is palpitations, usually attributed to a change in the underlying heart rhythm. Palpitations may make patients feel like they have an increased awareness of their heartbeat, can be associated with dizziness or lightheadedness, and may occur at rest or with exercise.^{2,10} Evaluation of symptoms, such as palpitations, is essential because it may reveal the presence of significant structural changes or result from underlying atrial or ventricular dysrhythmias. Identifying the associated cause of symptoms will help guide specific therapies (such as beta-blockers or ICD therapy).²

■ Murmurs on cardiac exam

Murmurs are common on physical exam and may be the initial finding that makes an NP suspect the presence of HCM. The point of maximal impulse felt on cardiac exam

Identifying the associated cause of symptoms will help guide specific therapies (such as beta-blockers or ICD therapy).



will be forceful and sustained, and a palpable S_4 gallop may be present. The classic HCM murmur is a crescendo-decrescendo systolic murmur along the left sternal border that increases with the Valsalva maneuver (usually by closing one's mouth and pinching one's nose shut while pressing out as if blowing up a balloon). Almost all cardiac murmurs decrease in intensity during Valsalva, with the exception of HCM, which increases the murmur, making this maneuver an important part of the cardiac exam. Aortic stenosis, or left ventricular outflow tract obstruction, can also occur at supra-ventricular, valvular, or subvalvular levels. (See *Types of aortic stenosis and their associated murmur locations.*) NPs must be able to recognize these differences on physical exam and refer appropriate patients for further evaluation.

■ HCM findings on ECHO

An ECHO is a useful diagnostic tool for evaluating the presence of HCM and can be used to measure the dimensions and shape of the heart, determine the degree of obstruction or pressure gradients within the heart, evaluate valve function, visualize blood flow, and identify abnormal blood flow patterns. ECHO findings consistent with the major

diagnostic criteria for HCM include anterior septum or posterior wall thickness greater than or equal to 13 mm, posterior septum or free wall thickness greater than or equal to 15 mm, and/or systolic anterior motion (SAM).^{4,11} SAM of the mitral valve may occur if the mitral valve leaflets are pulled or dragged in an anterior motion toward the ventricular septum and can cause mitral regurgitation. Consequently, the left ventricle has to generate higher pressures to overcome the left ventricular outflow track obstruction (LVOTO) and premature closure of the aortic valve. This may also cause a decline in pressure distal to the LVOTO.

Anterior septum or posterior wall thickness of 12 mm, posterior septum or free wall thickness of 14 mm, moderate

on ECG.) Prominent Q waves may be located in the inferolateral leads II, III, and AVF on the ECG in an estimated 12% to 14% of cases and less frequently in other lead locations. However, it is also important for the NP to recognize that such ECG findings may not be seen at all.⁸

■ Cardiac catheterization

Cardiac catheterization is helpful in assessing and quantifying the degree of obstruction of blood flow within the heart. This procedure typically includes a coronary angiogram and measures the pressures, gradients, and partial pressures of oxygen throughout the heart. In addition, tissue samples gathered during the procedure can be analyzed under a

microscope to confirm the genetic basis of HCM. During cardiac catheterization, catheters can measure the pressure difference between the left ventricle and ascending aorta. Patients with HCM who have a LVOTO can vary in the degree of severity of their underlying gradient and outflow tract obstruction

within the heart. LVOTO occurs usually when a thickening of the proximal portion of the interventricular septum occurs, resulting in narrowing or obstruction of the outflow track. Consequently, the left ventricle has to generate higher pressures to overcome the LVOTO, and premature closure of the aortic valve may occur. This may result in a decline in the pressure distal to the LVOTO. A mild LVOTO gradient may have a mean pressure gradient of less than 25 mm Hg, whereas those with severe LVOTO may have gradients greater than 40 mm Hg.

■ Electrophysiologic testing

This is an invasive diagnostic procedure that provides detailed information regarding the heart's electrical conduction system using catheters placed within the heart's chambers (atrium and ventricle). This diagnostic test can be used in conjunction with other medical information,



The ECG may show abnormal findings, including prominent Q waves and ST wave abnormalities.

SAM, and redundant MV leaflets contribute to the minor diagnostic criteria for HCM. Mitral regurgitation is another common finding in HCM patients with LVOTO and may result in symptoms, such as dyspnea. The mitral regurgitation is usually caused by distortion of the mitral valve, which can be seen on ECHO.¹¹

■ HCM findings on ECG

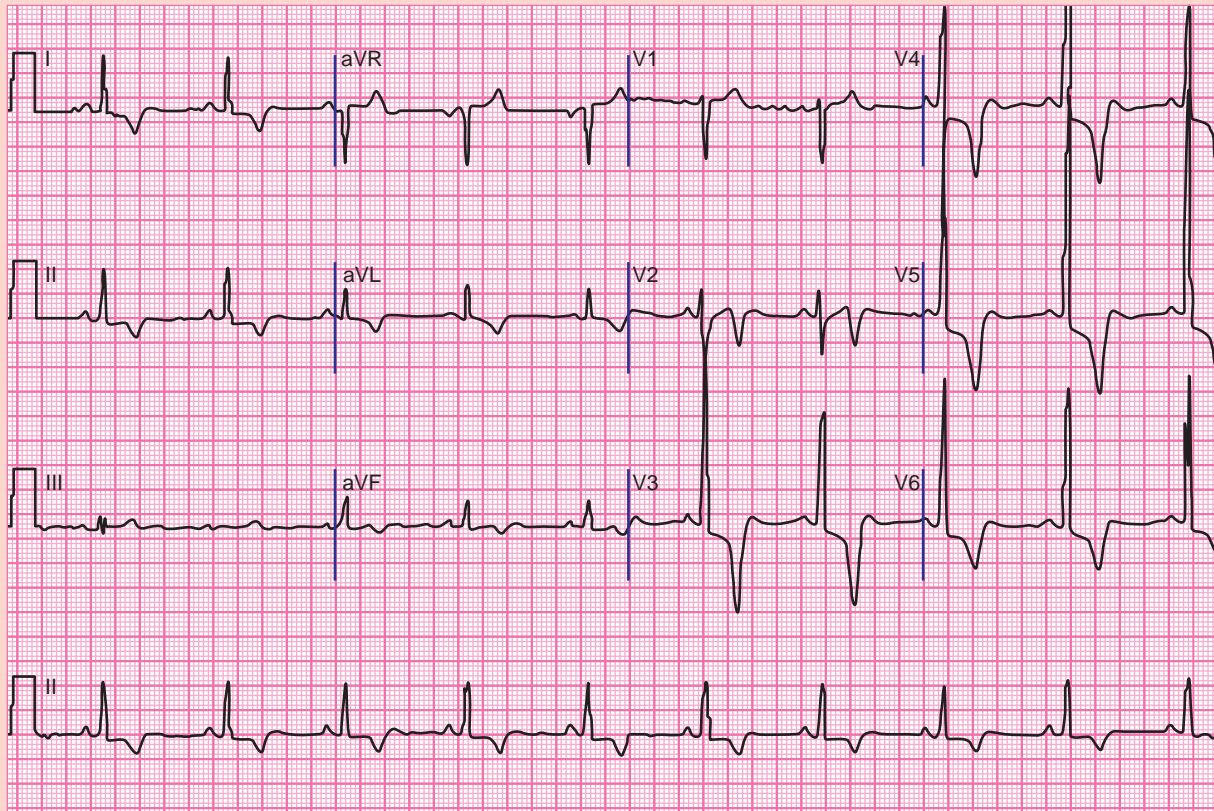
The ECG is often used to evaluate the presence of HCM. The procedure can inform the clinician of the heart rhythm abnormalities, such as premature ventricular beats or underlying hypertrophy. Because of its quick, noninvasive nature and widespread use in clinical practice, the ECG may be the first step to a possible diagnosis of HCM. The ECG may show abnormal findings, including prominent Q waves and ST wave abnormalities, which may be the first indication of potential ventricular hypertrophy.^{8,12} (See *HCM findings*

Types of aortic stenosis and their associated murmur locations²¹

Types of aortic stenosis	Most prominent murmur site
Valvular aortic stenosis	Harsh murmur over the right second intercostal space that radiates through the two carotid arteries into the neck.
Supravalvular aortic stenosis	Loudest at a point higher than it would be heard in valvular aortic stenosis, and also in some cases, further toward the right carotid artery.
Subvalvular aortic stenosis	The murmur that is most prominent above the left sternal border or the apex of the heart.

HCM findings on ECG

An ECG of an individual with HCM can show LVH and abnormal repolarization associated commonly with a large QRS amplitude (taller R waves in LV leads, deeper S-waves in RV leads)



Courtesy of Columbia University Department of Surgery, www.columbiasurgery.net.

such as a loss of consciousness to deduce the presence or absence of underlying dysrhythmias and for SCD risk stratification.^{2,10} An electrophysiology study will also provide specific details on the presence or absence of inducible dysrhythmias within the heart (atrial or ventricular in origin) and the associated rate, duration, and morphology of any inducible dysrhythmias. This information can help guide prescribed therapies (such as beta-blocker therapy or placement of an ICD).²

■ Magnetic resonance imaging

Magnetic resonance imaging (MRI) uses magnetic fields and radio waves to allow clinicians to see both a three-dimensional image of the heart and two-dimensional cross sections.^{2,10,13} A clinician can use an MRI to identify abnormal anatomical hypertrophy and structural changes within a patient's heart, which may aid in the diagnosis of HCM. MRI is often used to confirm a finding on an ECHO.^{2,10,13}

■ Cardiac exercise stress test

A cardiac stress test can show an individual's response to exercise, including the response of their heart rate and BP to the increasing blood flow and oxygenation demands of exercise. The exercise test is also helpful in the identification of underlying myocardial ischemia caused by a supply/demand mismatch. This provides a noninvasive measure of the heart's degree of functionality under conditions of exertion.^{2,10} Exercise-induced ECG changes can be recorded at rest with exercise and recovery and can be due to subtle ischemic changes secondary to LVOTO or diastolic dysfunction.¹⁴ Additionally, the inability of the ventricles to relax can result from the increased blood flow caused by the outflow tract obstruction and nonuniformity of ventricular contraction.¹⁴ This can also lead to delayed inactivation and an increase in chamber stiffness, known as diastolic dysfunction.^{2,10} In a case of diastolic dysfunction, the chamber stiffness triggered by HCM may cause blood to back up into the lungs, resulting in shortness of breath (usually during physical activity).

Exercise stress testing is important because approximately 25% of patients with HCM have an abnormal BP response with exercise.¹⁵ This is defined by either a failure of systolic BP to rise more than 20 mm Hg or a fall in the systolic BP.^{1,8,15}

The inability to augment and sustain systolic BP during exercise is caused by the LVOTO, or systemic vasodilatation during exercise, and may be seen on a cardiac exercise test.^{2,10} It is speculated that the fall in BP associated with bradycardia may be the result of an abnormal autonomic reflex response to the obstruction.^{2,10} The exercise test is helpful in unmasking many of these findings and guiding treatment.

■ Dysrhythmias and HCM

Patients with HCM may exhibit various dysrhythmias, including atrial fibrillation, atrial flutter, ventricular ectopy, VT, and ventricular fibrillation (VF). Atrial fibrillation is caused by a rapid, disorganized atrial electrical signal causing the heart's upper chambers (the atria) to contract very fast and irregularly (fibrillation).^{2,10} In fact, atrial fibrillation is the most encountered dysrhythmia in clinical practice and is very common in patients with



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HCM.² Dysrhythmia symptoms may include heart palpitations, shortness of breath, and weakness.^{2,10} While typically not life threatening, if sustained, unrecognized, and untreated, atrial fibrillation could potentially lead to complications, such as stroke.

Atrial flutter also involves a fast heartbeat (tachycardia in the right or left atrium of the heart) resulting from a rapid electrical circuit in the atrium. Palpitations, a fluttering feeling in the chest, shortness of breath, anxiety, and weakness are some common symptoms of atrial flutter that may be reported by a patient.^{2,10}

NSVT and VT are rapid ventricular rhythms typically characterized by a rapid pulse rate for several consecutive ventricular heartbeats. Symptoms characteristic of nonsustained and sustained VT include chest discomfort, palpitations, light-headedness, dizziness, near syncope or syncope, and result from impairment in circulation. Nonsustained ventricular dysrhythmias can quickly deteriorate to sustained VT leading to the absence of a pulse, loss of consciousness, hypotension, and ultimately, cardiac arrest if not terminated.^{2,8} While VT can occur

without the presence of heart disease in those with HCM, underlying causes such as worsening hypertrophy, heart failure, and valvular heart disease must be considered.^{2,16} The presence of a myocardial infarction/myocardial ischemia must also be ruled out. An NP must be aware of the potential presence of atrial and ventricular dysrhythmias in those with HCM.

■ Overview of pharmacologic management of HCM

Pharmacologic therapy has been the traditional primary therapeutic approach for relieving symptoms of HCM, blocking the effects of catecholamines that exacerbate the outflow tract obstruction, and to slow the heart rate so that diastolic filling is enhanced.^{2,10} The use of several classes of medications may be considered based on the individual profile. Beta-blockers have been traditionally administered to HCM patients to alleviate symptoms as well as treat atrial and ventricular dysrhythmias.^{8,10} Judgment regarding treatment with beta-blockers is often difficult because of the unpredictable day-to-day nature of symptoms.^{8,10} Patient responses to these drugs are highly variable. Propranolol was the first drug used to treat HCM, and derivatives of the drug have been developed and utilized.⁶

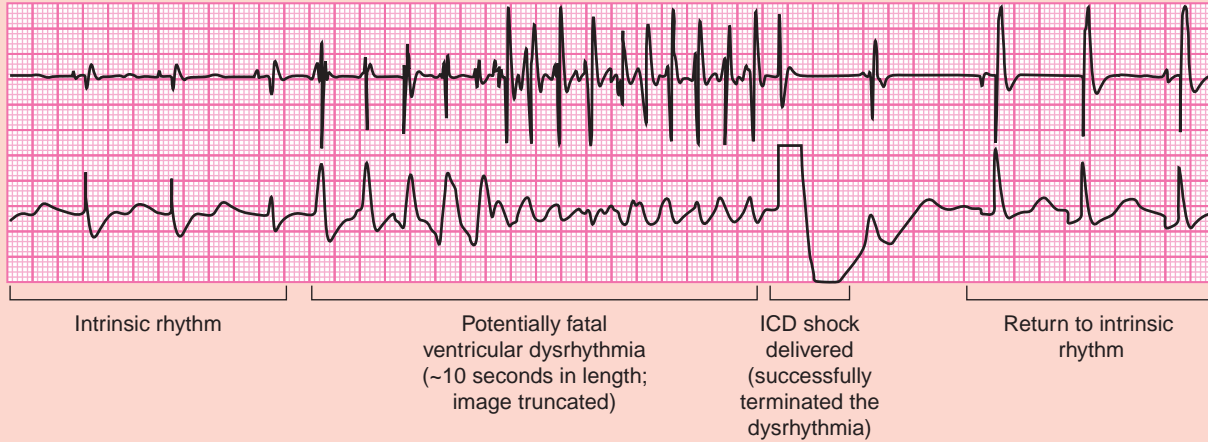
Research suggests that standard dosages of beta-blockers can mitigate disabling symptoms. There is little evidence that beta-blocking agents consistently reduce outflow obstruction. For this reason, beta-blockers are a preferred drug treatment strategy for symptomatic

patients with outflow gradients present only with exertion.^{2,8} This class of pharmaceuticals positively affects exertional dyspnea and exercise intolerance. The benefits of beta-blocking agents are a decrease in heart rate, consequent prolongation of diastole and relaxation, and an increase in passive ventricular filling.^{2,8} Common adverse reactions include fatigue, impotence, sleep disturbances, and chronic incompetence.^{2,8}

Calcium channel blockers are another class of drugs that can be used alone or in combination with beta-blockers. The calcium antagonist, verapamil, is one of several negative inotropic agents used for the treatment of HCM. Verapamil has a favorable effect on symptoms due to ventricular relaxation, relieving myocardial ischemia, and decreasing LV contractility. Verapamil maintains significant adverse consequences, including augmented outflow obstruction and pulmonary edema, as a result of the vasodilating properties predominating over negative inotropic effects. Because of these concerns, clinicians should be cautious about administering verapamil to patients with resting outflow obstruction and severe limiting symptoms.

ICD tracing

ICD tracing of VT to VF captured and transmitted by home monitoring. The patient with a known history of HCM experienced VF during sleep, which was successfully terminated by a single 34-J shock delivered from her ICD that restored her underlying intrinsic rhythm.



ICD tracing courtesy of the authors.

Genes associated with HCM

Most common proteins, genes, and the associated frequency seen in HCM on cardiogenetic testing

Protein	Gene	Frequency of HCM
Cardiac myosin-binding protein C	MYBPC3	~20%-45%
B-myosin heavy chain	MYH7	~15%-20%
Cardiac troponin	TNNT1 & TNNT3	~6%-7%

Adapted from Ho CY. Hypertrophic cardiomyopathy in 2012. *Circulation*. 2012;125(11):1432-1438.

■ **ICD therapy**

The ICD is an effective life-saving therapy against SCD. The ICD is surgically placed under the skin in the anterior pectoral chest wall, but specifically, it is the lead wires and defibrillator coils of the ICD that deliver the electrical shock to terminate ventricular dysrhythmias. The ICD monitors the underlying heart rhythm. Relying on a series of preprogrammed algorithms in the ICD, the ICD can detect the occurrence of VT or ventricular fibrillation. At this point, the ICD is capable of either pace terminating the ventricular rhythm or delivering an internal electric shock to terminate a ventricular dysrhythmia, restoring the intrinsic rhythm, usually normal sinus rhythm. (See *ICD tracing*.)

■ **Brief overview of HCM surgical approaches**

Septal myectomy has been shown to significantly ease symptoms in more than 90% of patients, many of whom can live a normal lifestyle with minimal or no symptoms.^{2,3} This

surgical approach involves removal of a portion of heart muscle and has been shown to be an extremely effective treatment for HCM over the past 50 years.² Alcohol septal ablation is another approach used to treat HCM that involves the surgical injection of an alcohol solution into the heart, leading to localized necrosis of a small portion of cardiac muscle.² However, to date, no randomized clinical trial data exist comparing these two approaches. Heart transplantation is reserved for those with end-stage heart failure.²⁻⁴

■ **Genetics of HCM**

HCM is a genetically inherited disease that occurs in an autosomal dominant manner—meaning affected individuals have a 50% chance of passing the condition on to each of their children.^{3,6} Multiple genes are known to be associated with HCM (see *Genes associated with HCM*) with numerous new genetic mutations being identified each year.^{2-4,6} Familial HCM is genetically heterogeneous because it can be

caused by genetic alternations at more than one locus. The same genotype could yield a different phenotype in various individuals within the same family, with multiple degrees of clinical symptoms and expressed hypertrophy. In individuals with identified pathogenic mutations but without the

methods of genetic testing do not detect all types of mutations; since not all genes are known, not all can be analyzed.² In addition, the highest yield is in those with a positive family history of HCM and clinical feature, whereas the lowest yield is in those with no family history and those with very late onset of the disease. Genetic test results can be ambiguous in those with a variant of unknown significance because it may be unknown if the DNA variant is pathogenic (disease causing, disease modifying, or inconsequential).³ However, the results of genetic testing do not significantly change individual prognosis; they can guide individual treatment, management, and follow-up within families.¹⁷



The benefits of cardiogenetic testing include confirmation of the definitive diagnosis of HCM and guiding treatment.

HCM phenotype, the guidelines recommend serial ECG, echocardiography, and clinical assessments at periodic intervals depending on the age of the individual.² Screening of first-degree relatives is useful to identify additional affected family members to continue surveillance and minimize symptoms and to implement appropriate treatment and management.¹⁷ Genetic testing can be used to confirm a diagnosis of HCM. However, multiple factors must be considered prior to undergoing testing, including the clinical and family history, ECG, echo phenotypes, and benefits/risks of genetic testing.


■ Family counseling

The NPs, along with other health professionals who specialize in genetics, play a critical role in counseling patients¹⁸ regarding HCM. However, once a specific mutation has been associated with HCM in a family, mutation-specific genetic testing should be offered to additional family members. The benefits of cardiogenetic testing include confirmation of the definitive diagnosis of HCM and guiding treatment.¹⁹ If genetic testing in an individual identifies a pathogenic mutation, then additional family members can undergo genetic testing for that specific mutation. This will allow for appropriate follow-up and treatment in those with a pathogenic mutation identified and reassurance for those who test negative for the mutation. Options for reproductive planning can be discussed, including the 50% risk of transmission to each offspring, in those who test positive. Those who test negative and do not harbor a specific HCM mutation previously identified as running within their family can be reassured of their health status. In addition, because a given individual does not harbor a specific HCM mutation, they cannot pass it along to subsequent generations. For those who test positive, screening and assessment of each individual's risk for SCD can be undertaken and appropriate treatment, management, and follow-up implemented. However, the limitations of cardiogenetic testing must also be recognized, and families must be counseled that current

■ The role of the NP

Nurses trained in genetics are ideally suited to actively engage in clinical care, education, and research.²⁰ The NP may suspect HCM in a patient after completing a thorough history and physical exam. A three-generation family pedigree, gathering complete details on any reported symptoms and the associated onset and duration (whether at rest or during physical activity), sudden death details within a family, an ECG, and ECHO should be obtained. Referral to an experienced HCM center of excellence and team is recommended for individuals and families suspected of having HCM. This will allow further evaluation and diagnostic testing, the opportunity to discuss clinical findings, treatment and management of the disease, and provision of genetic counseling and testing.

■ Moving forward

As genetics (or the study of *single* genes and their effects) and genomics (or the study of *all* genetic material in the genome) become more integrated into clinical care and testing for inherited cardiomyopathies (such as HCM), it is imperative that NPs be well-equipped with knowledge in these areas. Ultimately, it is this awareness that will allow NPs to have the necessary skill set to provide personalized genetic and genomic care. Competencies have been developed around genetic and genomic content in academic programs to help NPs integrate these advances into their clinical care, research, and education. Furthermore, NPs will also be responsible for ensuring appropriate treatment, management, and surveillance of patients and families. NPs are ideally suited to actively engage in the care and treatment of individuals with or at risk for HCM. 

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