DOI: 10.15825/1995-1191-2026-1-85-97

TREATMENT STRATEGIES FOR HYPERTROPHIC CARDIOMYOPATHY BASED ON ANATOMICAL VARIANT

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Hypertrophic cardiomyopathy (HCM) is the most common inherited heart disease and is characterized by a variety of manifestations: from morphological features and disease progression to clinical presentation and hemodynamic parameters. Management of HCM should be strictly individualized, considering not only hemodynamic parameters but also anatomical features. Classification based on the presence or absence of left ventricular outflow tract obstruction, as well as the location of interventricular septal hypertrophy (basal, midventricular, and apical), largely determines the optimal management strategy. Drug therapy, surgical myectomy, and alcohol septal ablation are the main treatment options for obstructive HCM, whereas management of the non-obstructive form focuses on symptom control and prevention of complications. Given the risk of sudden cardiac death, timely implantation of an implantable cardioverter-defibrillator in high-risk patients is of paramount importance.

Keywords: hypertrophic cardiomyopathy, left ventricular outflow tract obstruction, surgical myectomy, alcohol septal ablation, heart transplantation.

INTRODUCTION

HCM is the most common inherited heart disease, with an estimated prevalence of approximately 1 in 500 individuals [1].

It is defined by increased left ventricular (LV) wall thickness (≥15 mm in at least one myocardial segment in adults) that cannot be explained by abnormal loading conditions such as systemic hypertension [2]. HCM exhibits marked heterogeneity, encompassing a wide spectrum of morphological features, disease progression patterns, clinical manifestations, and hemodynamic profiles [3, 4].

Management strategies for HCM include both conservative and surgical approaches. Surgical options comprise septal myectomy, alcohol septal ablation, and, in advanced cases, heart transplantation (HT). HCM accounts for approximately 1–5% of all heart transplant indications, with post-transplant survival comparable to that observed in other forms of cardiomyopathy [5–7].

The selection of the most appropriate treatment strategy is largely determined by the specific anatomical variant of the disease. This article aims to discuss which treatment is preferable for various forms of this disease.

HCM CLASSIFICATION

Yifan Wang et al. conducted a comprehensive literature review and, drawing on multiple classification systems, systematically outlined the current criteria and distinguishing features of the various HCM subtypes (Table 1) [8].

NON-OBSTRUCTIVE AND OBSTRUCTIVE HCM

A key determinant in the diagnosis and risk stratification of HCM patients is the presence or absence of left ventricular outflow tract (LVOT) obstruction. Understanding the pathophysiological differences between obstructive and non-obstructive forms is critical for selecting the optimal treatment strategy.

Non-obstructive HCM

Historically, HCM was primarily viewed as a condition characterized by dynamic LVOT obstruction. It was this feature that defined its early names: idiopathic hypertrophic subaortic stenosis, hypertrophic obstructive cardiomyopathy, and muscular subaortic stenosis [15, 16].

It is noteworthy that, in recent years, comparatively less attention has been devoted to investigating the natural course of HCM in patients who do not develop LVOT obstruction under any circumstances, including during physical exertion [17–20].

Evidence from a large cohort study (n = 573) demonstrated that patients with non-obstructive HCM (unlike those with obstruction) generally exhibit a stable clinical course, often remaining asymptomatic or experiencing only mild manifestations over extended periods. Favorable outcomes in this group were frequently achieved with medical therapy [17].

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Medical Management of Patients with Non-obstructive HCM

The management of heart failure in patients with non-obstructive HCM largely aligns with established heart failure treatment guidelines. The primary objective of drug therapy is to reduce LV diastolic pressure and improve ventricular filling. This is typically accomplished through heart rate control using beta blockers, verapamil and diltiazem, alongside the judicious use of loop diuretics. In the absence of LVOT obstruction, cautious administration of nitrates and ranolazine may also be considered to alleviate symptoms in patients complaining of chest pain [3].

Targeted modulation of the renin-angiotensin-aldosterone system (RAAS) using antifibrotic and antihypertrophic agents represents a promising adjunctive strategy in the management of HCM. Evidence from a small randomized, double-blind, placebo-controlled trial suggests that losartan may slow the progression of myocardial hypertrophy and fibrosis in patients with non-obstructive HCM [21].

Surgical Treatment of Patients with Non-obstructive HCM

However, there is an important exception to this relatively favorable course of non-obstructive HCM. A small group of patients (estimated at less than 10%) develops progressive heart failure that is refractory to optimal medical therapy and is associated with severe symptoms (New York Heart Association Class III/IV). This occurs in so-called burned-out HCM, which is characterized by impaired LV contractile function, although in some cases systolic function may initially remain preserved. In such cases, heart transplantation (HT) remains the only definitive treatment option [6, 22, 23].

HT in patients with HCM is associated with favorable outcomes, with survival rates reported to be comparable to – and in some studies exceeding – those observed in other cardiac conditions. Reported post-transplant survival rates for HCM are approximately 85% at 1 year, 75% at 5 years, and 61% at 10 years [6].

Patients with non-obstructive HCM remain at risk for disease-related complications, such as sudden arrhythmic death, thromboembolic stroke, and atrial fibrillation. However, these complications are relatively rare in this patient group. It is emphasized that disease-related mortality in patients with non-obstructive HCM remains low, estimated at approximately 0.5% per year, with excellent long-term survival rates of 99% at 5 years and 97% at 10 years [17, 19, 24–26].

Obstructive HCM

Systolic anterior motion (SAM) of the mitral valve (MV) leaflet is a common echocardiographic finding in HCM, occurring in up to 95% of cases. Its clinical mani-

Table 1

HCM classification (adapted from [8])

Sign	Types
Presence of LVOT obstruction	– Obstructive HCM – Non-obstructive [9]
Types of non-obstructive HCM	– Standard/most common variant – With dilational transformation – Restrictive type – With reduced LV contractile function [10]
Morphological features of the heart and type of LVH	– Basal septal hypertrophy – Septal hypertrophy – Septal hypertrophy with anterior and anteroseptal walls – Apical LVH [11]
	– Septum curved into the LV cavity, forming a crescent-shaped LV cavity – S-shaped septum – Septal hypertrophy with a smooth contour – Apical LVH – Mid-septal hypertrophy [12]
	– Isolated septal hypertrophy – Hypertrophy of the septum and adjacent sections (except the apex) – Hypertrophy of the apex in combination with hypertrophy of other walls – Apical hypertrophy [13]
	– Apical LVH – Mid-septal hypertrophy – Basal septal LVH – Diffuse LVH [14]

Abbreviations: LVOT, left ventricular outflow tract; HCM, hypertrophic cardiomyopathy; LVH, left ventricular hypertrophy; LV, left ventricular.

festations varies widely, ranging from an incidental imaging observation to severe dynamic LVOT obstruction associated with significant hemodynamic compromise. Approximately 60–70% of patients with HCM exhibit either resting or provocable LVOT obstruction [27].

The severity of SAM is graded as follows:
– Mild: transient SAM without contact with the MV;
– Moderate: SAM with leaflet–septal contact lasting less than one-third of systole;
– Severe: SAM with persistent leaflet–septal contact exceeding one-third of systole [28].

Factors predisposing patients to dynamic LVOT obstruction and the SAM effect in HCM have been identified (Table 2) [27].

Interventricular Septum

It has been established that S-shaped deformation of the thickened interventricular septum (IVS) is an independent risk factor for the development of the SAM effect. This effect is attributed to vortex-induced redirection of early, low-velocity systolic blood flow beneath the MV, resulting in anterior displacement of the valve leaflets toward the IVS [27].

Mitral Valve

Normal MV leaflet lengths, as assessed by echocardiography (ECHO), are generally reported as 18–24 mm for the anterior mitral valve leaflet (AMVL) and 11–14 mm for the posterior mitral valve leaflet (PMVL) [29]. In HCM patients, transesophageal ECHO has demonstrated significant elongation of the MV leaflets compared with control subjects. Reported measurements show leaflet lengths of 31 ± 4 mm versus 22 ± 3 mm for the anterior leaflet, and 20 ± 2 mm versus 15 ± 3 mm for the posterior leaflet ($p < 0.00001$ for both cases) [30].

Similarly, a magnetic resonance imaging (MRI) study involving 172 HCM patients and 15 HCM gene carriers without disease manifestations revealed consistent MV leaflet elongation. The mean anterior leaflet length was 26 ± 5 mm, significantly greater than that observed in controls (19 ± 5 mm, $p < 0.001$). The posterior leaflet was also significantly longer in patients with HCM (14 ± 4 mm versus 10 ± 3 mm, $p < 0.001$) [31].

Excess MV leaflet tissue often extends beyond the zone of coaptation and is referred to as the "residual leaflet", which contributes to increased leaflet length and surface area. This redundant tissue is typically the first to make contact with the IVS. In HCM, altered intraventricular hemodynamics resulting from abnormal cardiac geometry facilitate the displacement of this residual MV leaflet, thereby promoting LVOT obstruction [32].

Comparative analyses of MV and ventricular dimensions in patients with HCM and the SAM effect with data from the control group showed that in HCM patients, the distance from the MV leaflet closure site to the IVS (C-sept distance) was significantly reduced (12 ± 4 mm *vs.* 21 ± 3 mm, $p < 0.001$) [33].

Mitral regurgitation in HCM most commonly arises from incomplete closure of MV leaflets. This typically occurs when the PMVL fails to move anteriorly in coordination with the AMVL, either due to reduced length or limited mobility [34]. Accurate differentiation between SAM-related mitral regurgitation and primary MV regurgitation is essential for selecting an appropriate treatment strategy. Assessment of the regurgitant jet direction can aid in this distinction: a posteriorly directed jet is generally indicative of SAM-related regurgitation, whereas a centrally or anteriorly directed jet more often suggests intrinsic MV disease [35, 36].

Earlier theories attributed SAM to the Venturi effect, whereby IVS hypertrophy narrows the LVOT, reducing the cross-sectional increasing and thereby increasing blood flow velocity. The accelerated blood flow displaces

Table 2

Causes of systolic anterior motion (SAM) in HCM (adapted from [27])

Area of pathology	Features
Interventricular septum (IVS)	S-shaped septum
Mitral valve	Elongation of the Mitral valve leaflets (both anterior and posterior)
	Increased distance from the leaflet coaptation point to the apex of the anterior mitral leaflet (the area referred to as the "residual leaflet")
	Reduced distance between the leaflet coaptation point and the mitral annulus (C–sept distance)
	Venturi effect
	Anterior diastolic motion of the mitral leaflets
Papillary muscles	Papillary hypertrophy
	Increased number of papillary muscles
	Displacement and abnormal attachment of the papillary muscles
	Reduced distance between the papillary muscles
Chordal apparatus	Shortening and chordal fibrosis

the MV leaflets toward the IVS and LVOT, creating a "suction" effect [37].

Currently, a phenomenon known as "diastolic anterior motion" of the MV has been described and considered a precursor to SAM. During late diastole, mitral inflow generates a posteriorly directed vortex that propels the MV leaflets forward even before the onset of systole [38].

Papillary Muscles

Echocardiographic studies have shown that papillary muscle hypertrophy – defined as a short-axis thickness greater than 11 mm at end-diastole – occurs in more than 50% of patients. Cardiac MRI further demonstrates an increased number (2.5 *vs.* 2.1, $p < 0.001$) and total mass of papillary muscles in individuals with HCM [27, 39].

Displacement of the papillary muscles – whether anterior displacement, basal shift, or apical relocation – can disrupt LV hemodynamics. These alterations may lead to overlap between the LV inflow and outflow tracts [40].

An abnormal attachment of a papillary muscle directly to the AMVL poses a particular risk, as this causes a SAM effect and LVOT obstruction, which worsens the prognosis in HCM [41].

Papillary muscle anomalies have been shown to correlate with higher resting LVOT pressure gradients, independent of IVS thickness or baseline heart rate. Experimental *in vitro* models further suggest that such abnormalities can induce SAM in HCM even in the absence of septal hypertrophy. These variants are estimated to be present in up to 20% of patients with HCM [39, 40, 42].

MORPHOLOGICAL VARIANTS OF INTERVENTRICULAR SEPTAL HYPERTROPHY IN HCM AND TREATMENT STRATEGIES

The clinical course and choice of treatment for HCM are determined not only by the presence or absence of LVOT obstruction but also by underlying morphological features. In particular, the pattern and distribution of IVS hypertrophy is an important factor that must be considered in clinical decision-making. Based on the predominant location of maximal septal thickening, several morphological variants of HCM can be identified, with the basal, mid-ventricular, and apical forms being the most common. These anatomical subtypes carry distinct prognostic implications and play an important role in guiding treatment strategies [43].

HCM with Basal Hypertrophy

Basal IVS hypertrophy is confined to the subaortic region and extends directly to the point where the chords attach to the apex of the AMVL [43].

Focal thickening of the IVS in the subaortic region, which causes the SAM effect and LVOT obstruction, makes this anatomical subtype the most promising candidate for various treatment methods aimed at reducing IVS thickness [43].

Pharmacological Treatment of Patients with HCM and Basal Hypertrophy

A meta-analysis of 37 studies involving 1,898 patients showed that, in obstructive HCM, various medications reduce the LVOT pressure gradient to varying degrees [44].

The most pronounced reductions were observed with disopyramide (–43.5 mmHg [95% CI, –51.6 to –35.3]) and cardiac myosin inhibitors (CMIs) (–34.8 mmHg [95% CI, –40.6 to –29.0]). In contrast, beta blockers (–20.7 mmHg [95% CI, –29.4 to –12.0]) and calcium channel blockers (–14.7 mmHg [95% CI, –23.3 to –6.1]) were associated with more modest reductions. An interaction p-value <0.01 indicates a statistically significant difference between groups.

Within the class of CMIs, mavacamten demonstrated a greater reduction in LVOT gradient compared with aficamten. Among beta blockers, metoprolol was associated with the largest gradient reduction, while verapamil showed the greatest efficacy among calcium channel blockers (p for interaction <0.01). These findings are consistent with results observed for provocable LVOT gradients [44].

Surgical Treatment of Patients with HCM and Basal Hypertrophy

According to the generally accepted definition, LVOT obstruction in HCM is defined as a peak LVOT pressure gradient of ≥30 mmHg, as measured by Doppler echocardiography. However, a higher threshold of ≥50 mmHg is generally used to guide consideration of invasive treatment [3].

According to European clinical guidelines [3], invasive treatment is indicated to reduce LVOT obstruction in the presence of the following criteria:

– A resting gradient or maximum induced gradient ≥50 mmHg (e.g., during exercise or Valsalva maneuver);
– Moderate to severe mitral regurgitation associated with SAM;
– Presence of symptoms, typically New York Heart Association (NYHA) functional class II–IV;
– Syncope during physical exertion or unexplained recurrent syncope despite optimal medical therapy;
– Atrial fibrillation or moderate to severe left atrial dilatation.

In patients with obstructive HCM who remain resistant to medical therapy, septal myectomy (Morrow procedure) is an effective surgical option. This procedure involves resection of the basal portion of the IVS (transaortic [45] or transmucosal [46] approach), resulting in enlargement of the LVOT lumen and elimination of the

cause of SAM [47]. In cases of severe septal hypertrophy, a more extensive resection may be required, extending the myectomy into the mid-ventricular region beyond the point of mitral leaflet–septal contact (modified Morrow procedure) [48].

For patients at elevated surgical risk or those who are not suitable candidates for surgery, percutaneous alcohol septal ablation may be performed [49, 50]. This procedure involves injecting a small amount of 96% ethanol into the first septal branch of the left anterior descending artery. This induces a controlled localized infarction of the basal IVS, which subsequently leads to LVOT remodeling [51].

In pediatric patients, LVOT obstruction often requires a repeat septal myectomy. This may be due to various reasons, including inadequate myocardial resection during the initial procedure, the presence of a mid-ventricular component of obstruction, papillary muscle anomalies, and postoperative LV remodeling, including regrowth of septal myocardium. Notably, Osiev et al. reported a case of successful alcohol septal ablation in a 6-year-old patient [52].

Table 3 provides a comparative overview of surgical and endovascular approaches for reducing IVS thickness [3, 53].

HCM with Midventricular Hypertrophic Cardiomyopathy

Midventricular hypertrophic cardiomyopathy (MVHCM) is defined as a pattern of IVS thickening that extends from the MV chordae to the septal region opposite the papillary muscle heads [43].

Isolated MVHCM occurs in less than 10% of patients with obstructive HCM, but it may present with clinical manifestations similar to those observed in subaortic LVOT obstruction [54–56].

About 25% of these patients have an LV apical aneurysm, and in some of them, a thrombus forms within this aneurysm [3, 57].

Medical Management of Patients with HCM and MVHCM

The main medications used in such cases (beta blockers, calcium channel blockers, and disopyramide) are aimed at reducing LV contractile function and increasing its filling time [58]. These drugs help reduce the pressure gradient in the mid-ventricular region and improve diastolic function in patients with HCM and mid-ventricular obstruction [57, 59].

Two recently published studies, based on analyses of three clinical cases, reported favorable clinical and morphological effects of mavacamten administered at doses of 5–10 mg in patients with HCM and midventricular obstruction. Over a follow-up period of 16–24 weeks, patients demonstrated a reduction in symptom severity, accompanied by a marked decrease in peak mid-ventricular pressure gradient – from 50–77 mmHg to 8–11 mmHg – as well as a 5- to 7-fold reduction in N-terminal pro–B-type natriuretic peptide (NT-proBNP) levels [60, 61]. Additionally, evidence of reverse LV remodeling was observed, including expansion of the LV cavity, reduction in IVS thickness and left atrial size, and improvement in LV diastolic function, while preserving systolic performance [60].

In rare cases, thrombi are found within the aneurysm, which should be treated with long-term oral anticoagulant therapy [57].

Table 3

Comparison of surgical and endovascular treatment methods in terms of reducing septal thickness [3, 53]

Myectomy	Alcohol septal ablation
Perioperative mortality	
Higher	Lower
Surgical complications	
Higher risk of VSD	Higher risk of atrioventricular block (7–20% of patients), requiring implantation of a permanent pacemaker
Higher incidence of postoperative bleeding (OR 0.18; 95% CI 0.11–0.32; $p < 0.0001$)	Higher residual LVOT gradient
Higher risk of the need for intra-aortic balloon counterpulsation	Higher likelihood of repeat intervention (7–20% of patients)
Surgical limitations	
Although the overall risk of VSD after septal myectomy is low, it may be increased in patients with moderate left ventricular hypertrophy (wall thickness ≤16 mm)	May be less effective in patients with severe LVH (≥30 mm)
Total adjusted treatment cost and hospital length of stay	
Higher	Lower

Abbreviations: VSD, ventricular septal defect; LVOT, left ventricular outflow tract; LVH, left ventricular hypertrophy; LV, left ventricular.

Surgical Treatment of Patients with HCM and MVHCM

In patients with HCM and MVHCM whose symptoms remain refractory to medical therapy, treatment options are more limited than in cases of HCM with basal hypertrophy.

Recent Russian study suggests that extended septal myectomy, combined with interventions on subvalvular structures, can effectively relieve obstruction in both the mid- and basal segments of the left ventricle, while also increasing LV cavity size [62]. When performing myectomy in patients with obstructive HCM in whom subaortic hypertrophy extends into the midventricular region, careful control of the volume of resected IVS tissue is crucial. Transesophageal echocardiography is routinely used to visualize anatomical landmarks within the left ventricle.

In cases with severe basal IVS hypertrophy, septal myectomy via a subaortic (transaortic) approach can be technically demanding. When obstruction involves the midventricular segment, the procedure is typically initiated through a transaortic myectomy, removing the IVS muscle as close as possible to the apex of the heart [43].

In cases of isolated MVHCM, septal myectomy via a transapical approach is generally considered the preferred surgical strategy. Transapical access provides a wide field of view and access to the (IVS and papillary muscles [54].

It is generally accepted that, in the absence of LVOT obstruction, alcohol septal ablation is not indicated for HCM. However, some clinical studies demonstrate that it may be useful for symptom relief and reduction of intraventricular gradients in patients with symptomatic midventricular obstruction [63, 64].

Some studies suggest that cardiac pacing may reduce obstruction and alleviate symptoms in patients with midventricular obstruction [65, 66]. A randomized, placebo-controlled trial involving 17 patients with midventricular obstruction and a mean NYHA functional class III despite optimal medical therapy demonstrated that individualized optimization of pacing parameters can reduce intraventricular obstruction and improve exercise tolerance in patients with severemid-ventricular obstruction symptoms.

In this study, the mean pressure gradient decreased from 80 ± 29 mmHg prior to pacing to 31 ± 21 mmHg at the optimal pacing configuration, representing an approximate 60% reduction ($p < 0.0001$). Personalized pacemaker programming was associated with an improvement in exercise tolerance, as evidenced by an increase in the distance covered during the 6-minute walk test (328.5 ± 99.9 m *vs.* 285.8 ± 105.5 m; $p = 0.018$) [58].

Apical HCM

Apical hypertrophy with obliteration of the LV apex is confined to the apex and the distal portion of the IVS [43].

Morphologically, apical HCM can be classified into three subtypes:

– Isolated type: hypertrophy confined to the apical region;
– Mixed type: combined apical and septal hypertrophy, with maximal wall thickness at the apex;
– Relative type: considered an early or less pronounced phenotypic expression of apical HCM [67].

In most cases, apical HCM is asymptomatic [68] and is generally associated with a favorable prognosis [69, 70].

However, extensive hypertrophy in this region can reduce LV end-diastolic volume, contributing to the development of diastolic heart failure [71].

Pharmacological Treatment of Patients with HCM and Apical Hypertrophy

In patients with apical HCM, treatment typically begins with beta blockers. These agents are commonly prescribed in part to reduce the frequency of unstable ventricular arrhythmias. The therapeutic benefits of beta blockers include:

– Reduction of heart rate, particularly during physical exertion;
– Improvement in diastolic heart function;
– Alleviation of symptoms such as chest pain and dyspnea through decreased myocardial oxygen demand;
– Reduction of the mid-ventricular pressure gradient in patients with concomitant mid-ventricular obstruction [68].

The beneficial effects of Verapamil and Diltiazem are attributed to their negative inotropic and chronotropic properties, which lead to a reduction in chest pain, an increase in LV diastolic filling time, and improved myocardial perfusion [72].

Low-dose loop and thiazide diuretics may be used to reduce dyspnea and manage volume overload in patients with apical HCM [68].

RAAS inhibitors are prescribed to slow the progression of myocardial hypertrophy and fibrosis, alleviate symptoms, and reduce mortality [21].

In patients with symptomatic non-obstructive HCM, treatment with Mavacamten has been associated with a significant, dose-dependent reduction in NT-proBNP levels. However, it has not demonstrated a significant effect on symptom relief or improvement in exercise capacity [73].

Surgical Treatment of Patients with HCM and Apical Hypertrophy

Transapical myectomy, aimed at enlarging the LV cavity, is effective in improving diastolic function, reducing symptoms, and potentially delaying or preventing the need for heart transplantation in patients with heart failure secondary to apical HCM [71, 74, 75].

HCM with Hypertrophy of the Entire IVS

Although this variant is rare, patients with obstructive HCM sometimes present with hypertrophy extending to all three zones of the IVS [43]. This phenotype is associated with a less favorable surgical prognosis.

A study published in 2018 evaluated 469 patients with obstructive HCM who underwent surgical myectomy. The cohort included patients with basal hypertrophy (n = 248), hypertrophy of the entire IVS (n = 141), and diffuse LV hypertrophy (n = 80). The mean follow-up period was 2.5 ± 1.4 years. Postoperative assessment showed that patients with basal septal hypertrophy experienced a thinner IVS, lower LV mass, and reduced gadolinium uptake on MRI. This group also demonstrated higher survival rates following myectomy. In contrast, patients with hypertrophy involving the entire IVS and those with diffuse LV hypertrophy exhibited poorer survival outcomes after surgical myectomy [14].

Preclinical HCM

A distinct subgroup among HCM patients consists of genotype-positive but phenotype-negative individuals, commonly referred to as having preclinical HCM [31, 76, 77].

In this group, LV structure and dimensions are typically within normal limits. However, they exhibit structural abnormalities of the MV compared with healthy individuals. These include a longer AMVL (17.1 ± 0.4 *vs.* 16.0 ± 0.4 mm/m^2, p = 0.006), a thicker PMVL (1.79 ± 0.008 *vs.* 1.62 ± 0.007 cm, p = 0.06), and a reduced distance between the papillary muscles (31.1 ± 0.7 *vs.* 34.2 ± 0.9 mm, p = 0.007).

The prevalence of SAM is also significantly higher in preclinical HCM compared with healthy controls (15.2% *vs.* 1.6%, p = 0.006). These changes are more pronounced in HCM with a distinct phenotype and the presence of genetic markers. Therefore, early diagnosis supported by comprehensive imaging is essential for timely identification, accurate risk stratification, and appropriate selection of management strategies [78].

PREVENTION OF SUDDEN CARDIAC DEATH (SCD) IN PATIENTS WITH HCM

Despite advances in medical and interventional therapies, SCD remains a major clinical concern in patients with HCM. The implantable cardioverter defibrillator (ICD) is an established and effective strategy for both primary and secondary prevention of SCD in this population [3, 4].

Secondary prevention involves ICD implantation in patients who have survived cardiac arrest due to ventricular tachycardia (VT) or ventricular fibrillation, or in those with a documented history of sustained, hemodynamically significant VT [3].

For primary prevention, ICD implantation is recommended in patients with HCM who present with at least one major risk factor for SCD [4] and a calculated 5-year SCD risk of ≥4% (Class IIb) to ≥6% (Class IIa) according to the HCM Risk-SCD model [3].

Guidelines from both American and European cardiology societies largely overlap in identifying SCD risk factors in HCM, although certain differences remain (Tables 4 and 5) [3, 4].

Table 4

Risk factors for sudden cardiac death (SCD) in patients with hypertrophic cardiomyopathy (HCM) according to the American Heart Association [4]

SCD risk factors	Description
Family history of SCD from HCM	Family history of SCD, judged definitively or likely attributable to HCM in ≥1 first-degree or close relatives who are ≤50 years of age.
Massive LVH	Maximum wall thickness ≥28–30 mm in any segment within the chamber according to echocardiography or cardiovascular magnetic resonance (CMR) imaging
Unexplained syncope	≥1 unexplained episodes involving acute transient loss of consciousness, judged by history unlikely to be of neurocardiogenic (vasovagal) etiology, especially when occurring within the last 6 months
Nonsustained ventricular tachycardia (NSVT)	A significant risk marker is frequent (≥3) and prolonged (≥10 beats) episodes of NSVT with a high ventricular rate (VR) (≥200 beats per minute), as determined by Holter monitoring.
LV systolic dysfunction	Ejection fraction <50% by echocardiography or CMR imaging.
LV apical aneurysm	Independent of the size of the aneurysm
Extensive late gadolinium enhancement	Diffuse and extensive late gadolinium enhancement on CMR imaging, representing myocardial fibrosis comprising ≥15% of LV mass.

Abbreviations: LVH, left ventricular hypertrophy; LV, left ventricular.

Table 5

Risk factors for sudden cardiac death (SCD) in patients with hypertrophic cardiomyopathy (HCM) according to the European Society of Cardiology [3]

SCD risk factors	Description
Family history of SCD from HCM	– SCD in a first-degree relative <40 years of age, irrespective of a confirmed diagnosis of HCM – SCD in a first-degree relative of any age with a confirmed diagnosis of HCM
Massive LVH	Maximum wall thickness ≥30–35 mm according to echocardiography
Syncope	Unexplained, non- neurocardiogenic syncope, particularly episodes, occurring within 6 months prior to evaluation
Nonsustained ventricular tachycardia (NSVT)	A consecutive series of ≥3 ventricular contractions with a frequency of ≥120 beats per minute, lasting <30 seconds, regardless of the number and duration of NSVT episodes per day, as well as ventricular rate (VR) during paroxysm. The occurrence of NSVT during or immediately after physical exertion may indicate an increased risk of SCD.
Age	SCD risk is higher in younger patients. In this population, major risk factors, such as NSVT, severe LVH, and unexplained syncope, carry greater adverse prognostic significance.
Left atrial dilation	However, current evidence is insufficient to establish a clear association between SCD and specific parameters such as left atrial area or volume.
LVOT obstruction	However, the prognostic significance of provoked LVOT obstruction and the effectiveness of different treatment strategies (conservative and invasive) in preventing SCD remain poorly understood.

Abbreviations: LVOT, left ventricular outflow tract; LVH, left ventricular hypertrophy; LV, left ventricular.

CONCLUSION

The treatment strategy for HCM should be strictly individualized, taking into account not only hemodynamic status but also detailed anatomical features. HCM classification based on the presence or absence of LVOT obstruction, as well as the localization of IVS hypertrophy, is essential for selecting the optimal management approach.

Medical therapy, surgical septal myectomy, and alcohol septal ablation are the main treatments for obstructive HCM, whereas management of the non-obstructive form primarily focuses on symptom control and prevention of complications.

Given the risk of SCD, timely implantation of an ICD in high-risk patients remain critical components of comprehensive care.

The authors declare no conflict of interest.

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The article was submitted to the journal on 6.09.2025