Advanced imaging for risk stratification of sudden death in hypertrophic cardiomyopathy

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ABSTRACT

Hypertrophic cardiomyopathy (HCM) is a common inherited cardiac condition, which typically manifests as left ventricular hypertrophy. A small subset of patients with HCM have an increased risk of sudden cardiac death (SCD) from ventricular arrhythmias. Risk of SCD can be effectively reduced following implantation of implantable cardiac defibrillators (ICD), although this treatment carries a risk of complications such as inappropriate shocks. With this in mind, we turn to advances in cardiac imaging to guide risk stratification for SCD and to select the appropriate individual who may benefit from ICD implantation. In this review, we have taken the opportunity to briefly summarise the role of imaging in the diagnosis of HCM before focusing on how specific imaging features influence risk of SCD in patients with HCM.

INTRODUCTION

Assessing for risk of sudden cardiac death (SCD) is an essential part of the evaluation of patients with hypertrophic cardiomyopathy (HCM). Although SCD is not frequent (~0.31–0.39 per HCM person-years), it is the most devastating manifestation of HCM independent of the presence of obstruction. A major challenge for the managing physician is to identify the individual at risk of SCD and for this, we turn to advances in cardiac imaging to guide stratification.

Implantable cardioverter defibrillator (ICD) insertion for the secondary prevention of SCD is given a class I indication in the American College of Cardiology (ACC)/American Heart Association (AHA) 2 and the European Society of Cardiology (ESC) 3 guidelines for the management of HCM. Risk stratification for primary prevention of SCD to decide on ICD implantation is more nuanced and is actively being refined, with some variability in recommendations from different society guidelines. 2–4 The general principle, however, is to identify high-risk patients based on the presence of clinical and imaging risk markers. 2–4

The 2011 ACC/AHA guidelines recommend an ICD in those with major SCD risk markers: SCD events in first-degree family members, maximal wall thickness ≥30mm, unexplained syncope, documented non-sustained ventricular tachycardia (VT) and abnormal blood pressure response with exercise. 2 The ESC guidelines recommend using a risk score (HCM Risk-SCD score) in those ≥16 years of age, which incorporates a few additional risk factors to that of the ACC/AHA guidelines, including left ventricular outflow tract (LVOT) gradient, two-dimensional left atrial size and age at the time of evaluation. 3,4 The risk score provides an individualised estimate of 5-year SCD risk, classified as low <4% (ICD not indicated), intermediate 4%–6% (ICD may be considered) or high >6% (ICD should be considered). 3 Its prognostic value was subsequently validated in a retrospective analysis of 3703 individuals with HCM. 6 Patients with a predicted 5-year risk of <4% (n=1524; 71%) had an observed 5-year SCD incidence of 1.4%; patients with a predicted risk of ≥6% had an observed SCD incidence of 8.9% at 5 years. However, some of the continuous variables used such as left atrial size and LVOT gradient have a high degree of variability, which may lead to measurement heterogeneity and thus misclassification of risk. The left atrium is a three-dimensional structure, and its dimensions can be measured inconsistently on two-dimensional echocardiography. Similarly, LVOT gradient is dynamic and influenced by loading conditions and medications.

In a more recent study of 2094 patients with HCM, the HCM Risk-SCD score was shown to have good specificity (92%, 95% CI 91% to 94%) but poor sensitivity (34%, 95% CI 24% to 44%) in predicting SCD events in those with risk scores >6%. Similarly, in our recent experience with a large proportion of patients having myomectomy for obstructive HCM, the observed SCD-related event rates were similar across the three ESC SCD risk categories, with myomectomy mitigating SCD risk. 7 The findings were similar when ACC/AHA risk factors were used suggesting that neither recommendations fully encapsulate risk prediction following myomectomy. Given the inherent limitations of the above aforementioned approaches, we recommend using the recently proposed enhancement of the 2011 ACC/AHA guidelines (shown in figure 1), which was highly sensitive (95%, 95% CI 89% to 99%, in the intention-to-treat analysis) and specific (78%, 95% CI 76% to 80%) for predicting SCD events. 8 The number of patients needed to treat with ICD to prevent one SCD using the enhanced ACC/AHA guidelines was 6.6 vs 10.3 when using the ESC model (intermediate-risk/high-risk groups). This approach differs from the 2017 AHA/ACC/Heart Rythm Society guidelines for management of patients with ventricular arrhythmias and the prevention of SCD. 4 In the enhanced 2011 ACC/AHA guidelines, extensive late gadolinium enhancement (LGE), end-stage HCM with LV systolic dysfunction and the presence of an LV apical aneurysm all constituted major risk markers considered sufficient individually to justify ICD implantation. To support this approach, in the recent study by Maron et al of 2094 patients with HCM studied longitudinally, these additional major imaging markers were associated with appropriate ICD interventions in 21 patients (26%), including...
**Major Clinical Risk Markers**
1. Family history of SCD related to HCM
2. Unexplained syncope
3. Non-sustained VT (≥ 3 beats, ≥ 130 bpm)

**Major Imaging Risk Markers**
1. Severe LVH ≥ 30mm in any segment *
2. Extensive late gadolinium enhancement
3. End-stage HCM (LVEF < 50%)
4. LV apical aneurysm of any size

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**Risk Modifiers**
1. Abnormal blood pressure response to exercise
2. Resting LVOT obstruction (>50mmHg)
3. Moderate late gadolinium enhancement
4. Age ≥ 60 years (lowers risk)

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**Figure 1** Risk stratification for primary prevention of SCD using an enhancement of the 2011 AHA/ACC guidelines. In the enhanced AHA/ACC guidelines developed by Maron et al., presence of one of the major clinical or imaging risk markers is considered sufficient evidence of increased SCD risk to justify recommendation of prophylactic implantable cardiac defibrillator implantation. A positive family history is defined by sudden death in a first-degree or other close relative aged 50 years or younger definitively or likely caused by HCM. LGE with contrast CMR is considered extensive either by quantification (≥15% LV mass) or visual inspection. Age older than 60 years in itself is considered to confer lower risk of SCD. Other risk modifiers are used to support ICD decisions when associated with one or more major risk markers including abnormal blood pressure response to exercise, significant resting LVOT obstruction and moderate degree of LGE. AHA, American Heart Association; ACC, American College of Cardiology; CMR, cardiac magnetic resonance; HCM, hypertrophic cardiomyopathy; ICD, implantable cardiac defibrillators; LGE, late gadolinium enhancement; LVEF, left ventricular ejection fraction; LVH, left ventricular hypertrophy; LVOT, left ventricular outflow tract; SCD, sudden cardiac death; VT, ventricular tachycardia.

LV apical aneurysms (n=12), end-stage heart failure (n=6) and extensive LGE (n=3).8

With particular relevance to the cardiac imager, we address the established as well as potential risk modifiers of SCD in the sections below. Key articles are summarised in table 1.

**Left ventricular wall thickness**

The association between wall thickness and SCD risk in HCM suggests a relationship between the severity of disease expression and increased adverse events.9 10 Moreover, there is an independent relationship when LV hypertrophy is severe (≥30 mm, figure 2). Hence, left ventricular hypertrophy (LVH) ≥30 mm is a risk factor considered sufficient, even in the absence of other risk markers to consider ICD implantation.2 3 However, the negative predictive value appears to be limited as most SCD occurs in those with <30 mm of hypertrophy. It is also critical to recognise that the severity of LVH should not be simply considered in a binary manner. There is in fact, a continuous, linear relationship between wall thickness and increased SCD risk, with the association becoming particularly prognostic in young adults.9 The implication is that the SCD risk may be equal or greater in a young adult with maximal wall thickness ~30 mm versus older individuals. Recent data from the ShaRe registry (Sarcomeric Human Cardiomyopathy Registry) on 4591 patients indicate that those with a likely pathogenic/pathogenic sarcomere mutation had higher maximal wall thickness compared with mutation negative individuals, which may at least in part explain the higher adverse event rates in this cohort.11

Given the emphasis on the use of LV wall thickness in making a diagnosis, determining SCD risk and candidacy for ICD implantation, it is critical to provide reliable measures of wall thickness.12 In a study of 195 individuals with HCM who underwent echocardiography and cardiac magnetic resonance (CMR), nearly half of the cohort had intermodal wall thickness measurement discrepancies ≥10% and in 16%, this discrepancy occurred at diagnostic (15 mm) or prognostic (30 mm) cut-offs.13 Thus at these clinically relevant thresholds, assessment of wall thickness should ideally be performed using CMR.

Using echocardiography, maximal wall thickness measurements should be obtained perpendicular to the ventricular septum in either the parasternal long-axis or short-axis imaging planes, with the maximal measurement derived from the thickest LV segment. Care needs to be taken to prevent overestimation (figure 3), which can occur if the right ventricular (RV) muscle structures are included.14 Use of an intravenous contrast agent should be considered2 if imaging is suboptimal or LV wall thickness measurements are borderline.5 Alternatively, in cases where echocardiography remains suboptimal despite use of contrast, CMR should be performed. CMR is particularly useful in identifying maximal hypertrophy in locations other than the basal anteroseptal wall segment due to the use of user-defined imaging planes. CMR is particularly useful in uncertain situations as it can provide additional information impacting management. More reliable wall measurements to guide decisions with regard to ICD implantation is possible due to high spatial resolution imaging allowing sharp contrast between bright blood and dark myocardium.15 CMR is also valuable to assess areas not well imaged using echocardiography, for example, the apex (figure 4).

In exceptional cases, where reliable left wall thickness measurements cannot be made using echocardiography and...
and is associated with a low risk of adverse events (figure 5C). Recent findings from the Hypertrophic Cardiomyopathy Registry comprising 2755 prospectively recruited patients with HCM demonstrate that individuals with LGE had thicker walls, more baseline arrhythmias and were more likely to be sarcomere mutation positive.\(^{20}\)

A number of cross-sectional studies have demonstrated the association between LGE and VT,\(^{21,22}\) suggesting that LGE may represent a structural nidus.\(^{15}\) There is accumulating evidence that presence of LGE may identify individuals at higher risk of SCD, who may benefit from primary prevention ICD. A recent meta-analysis of 2993 patients demonstrated that the presence of LGE was associated with increased SCD (OR 3.41).\(^{23}\) Chan et al conducted a study of 1293 individuals with HCM demonstrating that LGE extent >15% of LV mass portended a twofold increase in SCD risk in patients considered to be at lower risk.\(^{24}\) Chan et al reported that the presence of LGE was associated with increased SCD (OR 3.41).\(^{23}\) The authors found extent of LGE also predicted the development of end-stage HCM with systolic dysfunction (adjusted HR, 1.80/10% increase in LGE).\(^{23}\) LGE as a percentage of LV mass demonstrated a twofold increase in SCD risk in patients undergoing myomectomy.

CMR, contrast-enhanced, gated multidetector CT can be used to measure LV wall thickness.

Late gadolinium enhancement on CMR

CMR allows myocardial tissue characterisation through the use of gadolinium-based contrast agents. Following intravenous administration, gadolinium accumulates in areas of expanded extracellular space, which may represent an increase in the collagen volume fraction or fibrosis within the myocardium. CMR with delayed enhancement imaging is the non-invasive gold standard for quantification of focal myocardial fibrosis.\(^{16}\)

It uses an inversion pulse to suppress normal myocardium, followed by a standard gated T1-weighted gradient echo acquisition performed 10–15 min after gadolinium administration. In regions of fibrosis, T1 time is shorter than in adjacent normal myocardium, with a higher signal intensity, termed LGE. The physiological basis of LGE of fibrotic myocardial tissue relates to a combination of an increased volume of distribution for gadolinium and a prolonged washout related to the decreased capillary density within the myocardial fibrotic tissue.\(^{17}\)

In HCM, LGE (figure 5A) is present in approximately 50% of individuals,\(^{18-20}\) typically in mid-wall of the hypertrophied myocardium. Small, focal areas of LGE confined to the RV insertion point do not reflect replacement fibrosis or scarring and is associated with a low risk of adverse events (figure 5C). Recent findings from the Hypertrophic Cardiomyopathy Registry comprising 2755 prospectively recruited patients with HCM demonstrate that individuals with LGE had thicker walls, more baseline arrhythmias and were more likely to be sarcomere mutation positive.\(^{20}\)

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Taken together, the findings from these two, large independent cohorts, suggest that the extent of LGE may be more important than the presence of LGE per se.

Currently, there are a number of practical challenges preventing the routine adoption of LGE quantification in daily clinical use.\textsuperscript{15} There is current lack of uniformity in quantification technique due to the heterogeneity in CMR scanning hardware, LGE protocols, contrast dose/type, optimisation of inversion times.\textsuperscript{35} Several lines of evidence suggest that LGE quantification with an intensity of \( >6 \text{SD} \) above normal myocardium is an optimal threshold for detection in HCM (figure 5B), although other methods including full width at half maximum (ie, pixels that are \( \geq 50\% \) the signal intensity of a hyper enhanced area) also have high reproducibility.\textsuperscript{24}

Currently, qualitative assessment of LGE is to be considered as an arbitrator in resolving clinical decision making in patients where uncertainty remains about ICD implantation. Extensive LGE should sway the decision towards ICD implantation and conversely those with no or minimal LGE, away from device therapy.\textsuperscript{15, 18, 19}

**LVOT obstruction**

A number of studies report a significant association between LVOT obstruction, with peak basal (resting) gradients \( >30 \text{ mm Hg} \) and SCD, although this observation has not been consistently reproducible.\textsuperscript{25, 26} SCD risk appears to be positively correlated with the degree of LVOT obstruction. Nearly one-third of
patients will have obstruction under resting conditions (gradi- 

te >30 mm Hg). Obstruction causes a complex interplay of 

abnormalities stemming from the abrupt increase in LV systolic 

pressure leading to prolongation of ventricular relaxation, elevation of LV diastolic pressure, mitral regurgitation, myocardial ischemia, and a decrease in forward cardiac output.

Initial evaluation by echocardiography should incorporate 

assessment for the mechanism of LVOT obstruction, and in 

cases where echocardiography is not definitive, evaluation with 

CMR is needed (figure 6). Although septal reduction therapy is 

primarily used to address symptoms of progressive heart failure, 

survival of patients after septal myectomy has been reported to 

be superior to survival of unoperated patients, and this improved 

outcome was related, in part, to greater freedom from SCD.27

Continuous wave Doppler is used to assess the presence and severity of LVOT obstruction. It is a late-peakin systolic 

velocity that reflects the occurrence of subaortic obstruction 

late in systole, and the peak gradient influences treatment deci- 

sions.3, 4 Importantly, care needs to be taken to distinguish LVOT 

obstruction from mitral regurgitation. In the setting of resting 

outflow gradient <30 mm Hg, provocative measures may be 

used to ascertain if higher gradients can be elicited, ideally with 

physiological exercise (stress echocardiography) but alternatively 

with Valsalva manoeuvre or amyl nitrite.5 The role of stress 

echocardiography including exercise-related LVOT obstruction 

was assessed in a study of 426 asymptomatic or minimally symp- 
tomatic patients with HCM.28 After a mean follow-up of 8.7 

years, per cent of age-sex predicted metabolic equivalents (HR: 0.76), heart rate recovery at 1 min (HR: 0.89) and atrial fibril-

lation (HR: 2.7) but not post-stress LVOT gradient (HR: 1.01) 

predicted were associated with adverse outcomes.

One of the limitations of using LVOT obstruction for risk 

stratification is its variation according to loading conditions 

day-to-day activities. Thus, its presence should not be 

used in isolation, but rather as a modifying factor to guide ICD 

implantation.

LV apical aneurysm

LV mid-cavity obstruction can occur in a subset of patients with 

HCM with ~25% demonstrating an apical aneurysm.39 The 

incidence of apical aneurysm is between 1.3% and 4.8% within 

a general HCM population.30, 31 In a subset of 21 patients with 

apical HCM and apical aneurysm, a Japanese group reported 

an event rate of 10.1%/year (ICD following ventricular tachycar- 

dia or fibrillation, appropriate ICD discharge or stroke).32

Figure 4  Illustrative case: HCM with mid-cavity obliteration and apical aneurysm. An individual was evaluated after presenting with palpitations. 

12-lead ECG demonstrated voltage criteria for left ventricular hypertrophy and ambulatory ECG monitoring was significant for four consecutive ventricular beats at a rate of 148 beats/min. Transthoracic echocardiography (A) apical four-chamber, (B) apical long-axis and C) contrast-enhanced apical long-axis views demonstrating mid to distal left ventricular wall thickening suggestive of apical variant HCM with possible small apical aneurysm. Use of the HCM Risk-SCD score5 predicted an intermediate risk of sudden cardiac death of 4.3% at 5 years which placed the patients in an ambiguous grey zone of SCD risk stratification. Further assessment with CMR (D) CMR SSFP horizontal long-axis sequence, (E) SSFP short-

axis sequence of the distal third of the left ventricle and (F) short-axis PSIR sequence at the same level approximately 10 min following the 

administration of intravenous contrast. CMR images demonstrate apical HCM with mid-cavity obliteration and apical aneurysm with associated, 

extensive hyperenhancement conferring an increased risk of SCD. Following a shared decision-making process, an ICD was implanted. The patient 

was maintained on 50 mg of atenolol daily, with successful amelioration of symptoms. The patient remained virtually asymptomatic for 3 

years, after which they had a syncopal event followed by an ICD discharge for ventricular tachycardia (220 bpm), which was successfully aborted. CMR, cardiac magnetic resonance; ICD, implantable cardiac defibrillator; PSIR, phase-sensitive inversion recovery; SSFP, steady-state free precision.
In an independent cohort of apical HCM with apical aneurysm (n=93), Rowin et al reported a rate of HCM-related deaths and aborted SCD of 6.4%/year, threefold greater than the 2.0%/year event rate in 1847 patients with apical HCM without aneurysms. In a subsequent study (n=118 with LV apical aneurysm), Rowin et al reported that 36% of SCD events occurred in those ≥60 years. Though ageing in itself is considered protective against SCD, the aforementioned findings suggest the need to consider primary prevention ICD implantation milderly patients with apical aneurysms.

The 2011 ACC/AHA HCM guidelines recommend the inclusion of LV apical aneurysm as a risk modifier when assessing...
risk for SCD. Echocardiography, with care taken to avoid image foreshortening may allow adequate visualisation of the apical hypertrophy and aneurysms. Contrast-enhanced echocardiography help to distinguish apical hypertrophy against the presence of an apically displaced papillary muscle, in addition to excluding a thrombus (figure 4C). CMR has increased sensitivity to detect apical aneurysm and should be performed when apical aneurysm is suspected but not identified, for instance, in patients with mid cavity obstruction (figure 4).

**Left atrial size**

The left atrium (LA) is commonly enlarged in HCM and its size provides important prognostic information. Few observational studies have reported a positive correlation between LA size, measured using either LA volume (>34 mL/m²) or anteroposterior diameter (>48 mm), with adverse cardiovascular events including SCD.34 35

The HCM Risk-SCD score incorporates the left atrial diameter measured anteroposteriorly using M-mode or two-dimensional echocardiography in the parasternal long-axis plane. As LA is a three-dimensional structure, its dimensions can be measured incorrectly using a single two-dimensional measurement in isolation (online supplementary figure 1). Evaluation of LA size is best undertaken using LA volume indexed to body surface area to account for alterations in LA chamber size in all directions.

Importantly, left atrial size in itself should not be used to dictate management decisions for SCD prevention.15

**LV systolic dysfunction**

The 2011 ACC/AHA HCM guidelines recommend (class IIb) consideration of ICD implantation in individuals with non-obstructive HCM with left ventricular ejection fraction (LVEF) <50% and no ICD indication. 2 Although prophylactic ICD implantation is an accepted therapy for primary prevention in patients with HCM with systolic dysfunction, these patients were not included in most primary prevention ICD trials.

It is imperative to recognise the challenges in reliably measuring LVEF using echocardiography in individuals with significantly increased wall thickness, particularly involving the mid-LV to distal LV. Assessment of LVEF using CMR should be considered in this context (figure 4).

### Assessment of SCD risk in athletes

Recent ESC recommendations for athletes with HCM suggest it is reasonable to maintain participation in competitive sports for asymptomatic adults with mild clinical expression, low ESC risk score in the absence of high-risk features such as history of aborted SCD, exercise-induced ventricular tachycardia, significant LVOT obstruction or abnormal blood pressure response to exercise. In a study of 35 athletes with HCM, traditional SCD criteria have poor sensitivity (~0%) in identifying SCD risk (incidence 0.3% per year), highlighting the limitations of applying traditional SCD risk methods and imaging criteria in this unique patient population. Further evidence is needed to more precisely guide athletes with HCM towards participation in competitive sports.

### Other techniques

**Nuclear cardiology**

Autopsy findings from individuals with HCM-related SCD found a high prevalence of histological changes consistent with acute/subacute myocardial ischaemia, possibly forming the substrate for arrhythmogenesis. In a study of 133 individuals with HCM who underwent position emission tomography (PET), the presence of significant flow heterogeneity (defined as the ratio of the highest to lowest regional stress myocardial blood flow >1.85) was significantly associated with ventricular arrhythmias (n=23). This suggests the potential role of PET in SCD risk assessment; although until further larger studies are performed, we cannot recommend its routine adoption.

**Myocardial strain**

Recently, myocardial strain assessment has garnered interest as a novel imaging technique to more sensitively assess myocardial contractile performance. Myocardial strain refers to the degree of deformation of a fixed material point within the myocardium from its initial length throughout the cardiac cycle. Currently, left ventricular global longitudinal strain (GLS) using speckle tracking echocardiography is the most ubiquitously used strain parameter in HCM (online supplementary figure 2). In a recent systemic review of 14 observational studies in patients with HCM (n=3154), we found a clear association between abnormal LV GLS with adverse cardiac outcomes including

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CMR, cardiac magnetic resonance; LGE, late gadolinium enhancement; LV, left ventricular; LVEF, left ventricular ejection fraction; LVH, left ventricular hypertrophy; LVOT, left ventricular outflow tract; SCD, sudden cardiac death.
mortality, appropriate ICD discharge and ventricular arrhythmias. A barrier to the routine use of LV GLS for clinical decision making is the lack of prospective evaluations to confirm its utility in HCM. Furthermore, there are variations in the normal reference range in part contributed by the lack of standardisation between various echocardiography platforms. Uniformity in strain assessment is needed particularly given differences in strain values between high-risk and low-risk groups are small, relative to technique variability.

Illustrative case and summative approach to imaging-guided SCD risk stratification

An illustrative case (figure 4) highlights the incremental utility of multimodality cardiac imaging techniques (table 2) for SCD risk stratification in HCM.

CONCLUSIONS

To summarise, we recommend initial evaluation with a thorough and methodological echocardiogram, looking for features of heightened SCD risk. This includes assessment for maximal wall thickness, LVOT obstruction, left atrial enlargement and apical aneurysm. CMR should be considered for identification of myocardial fibrosis, which can act as an arbitrator in patients where uncertainty remains about ICD implantation.

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