Risk Stratification and Role of Implantable Defibrillators for Prevention of Sudden Death in Patients With Hypertrophic Cardiomyopathy

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Hypertrophic cardiomyopathy (HCM) is the most common cause of sudden cardiac death (SCD) in young people, including trained athletes. It is now 30 years since the introduction of implantable cardioverter-defibrillators (ICDs) to clinical cardiovascular practice and coronary artery disease, and now device therapy represents the most significant therapeutic innovation and the only definitive strategy for prolonging the life of HCM patients. ICDs have proved effective in preventing SCD in young HCM patients with appropriate intervention rates of 11% for secondary and 4% for primary prevention, despite massive left ventricular (LV) hypertrophy, LV outflow obstruction, diastolic dysfunction or microvascular ischemia. Targeting candidates for prophylactic ICD therapy can be complex, compounded by the unpredictability of the arrhythmogenic substrate, the absence of a dominant risk factor, and difficulty in assembling randomized trials. However, a single major risk factor is often sufficient to justify an ICD, although additional markers and other disease features can resolve ambiguous decision-making. Nevertheless, the absence of all risk factors does not convey absolute immunity to SCD. The current risk factor algorithm, when combined with a measure of individual physician judgment (and patient autonomy considerations), is an effective guide to identifying high-risk HCM patients. ICDs have altered the natural history of HCM for many patients and provided an opportunity to achieve many decades of productive life, and the potential for normal or near-normal longevity. Indeed, prevention of SCD has now become a new paradigm in the management of HCM.

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Historical Perspectives of ICD Therapy

General
The ICD was developed by Drs Michel Mirowski and Morton Mower in the early 1980s for patients with coronary artery disease (CAD). Since then, the ICD has become a standard of care for patients at high risk of SCD, particularly those with CAD and advanced heart failure. The ICD has revolutionized the treatment of heart rhythm disorders and has become an essential tool for managing patients with ventricular arrhythmias. Over the past few decades, the ICD has undergone significant improvements, including advances in device technology, programming algorithms, and implantation techniques. These advancements have allowed for better patient outcomes and improved survival rates. The ICD has also become increasingly accessible and affordable, making it a viable option for a broader range of patients.

ICD in HCM
In their seminal 1980 paper, Mirowski et al reported 3 patients at exceptionally high risk for SCD after surviving 2 or more cardiac arrests. In the laboratory, the implanted defibrillator operated spontaneously and automatically to abort ventricular fibrillation (VF). Notably, 2 of the 3 original patients had HCM, but for the ensuing 2 decades patients with HCM were largely ignored with regard to ICD therapy. Pharmacologic therapy has failed to provide HCM patients with reliable protection against SCD. Historically, such medications have included β-blockers and verapamil, for which there are no prospective data, type IA antiarrhythmic agents such as quinidine and procainamide (abandoned because of the potential for proarrhythmia), and amiodarone, which is impractical for long-term administration to young patients given its potential for important side-effects. For example, in a large retrospective study, almost 30% of patients taking

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amiodarone (as an adjunct to ICD therapy) experienced appropriate ICD interventions. Furthermore, in an Italian HCM cohort largely devoid of ICD implants, cardioactive and antiarrhythmic drugs were ineffective in protecting patients from lethal ventricular tachyarrhythmias (VT), and SCD occurred in 20% of those taking amiodarone.

The ICD era in HCM treatment essentially began in earnest with the highly visible publication in 2000 of the first sizeable series of patients. This report triggered enthusiasm for device therapy in HCM patients and subsequently greater numbers of ICDs were implanted prophylactically. Interest in defibrillator therapy for HCM patients has stimulated more widespread application to other genetic heart diseases (ie, ion channelopathies (long QT and Brugada syndromes) and arrhythmogenic right ventricular cardiomyopathy).

Profile of SCD in HCM

Estimates of the SCD rate in HCM unavoidably emanate from hospital-based cohorts, and in the older literature were as high as 6%/year, which we now recognize as overestimates based on tertiary center data contaminated by the preferential referral of high-risk patients. However, reports over the past 10 years from less selected regional or community-based cohorts placed annual HCM mortality rates at a much more realistic ≤1%. Nevertheless, the traditional profile of SCD in HCM remains unchanged; that is, it usually occurs without warning in asymptomatic or mildly symptomatic young patients (predominantly <25 years of age). In this respect, HCM is the most common cause of sudden death in competitive athletes in the U.S. (Figure 1).

Although SCD risk is lower in midlife and beyond, achieving this measure of longevity does not confer immunity to SCD. No relationship between SCD risk and gender has been identified, and although no differences in risk according to race are known, SCD is common in African-American competitive athletes with HCM.

Selection of HCM Patients for ICDs

Conventional Risk Markers

There has been a substantial investigative effort in ICD decision-making toward reliably identifying the relatively small subset of patients within the broad HCM disease spectrum.
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who are at unacceptably high risk for SCD.\textsuperscript{3,4,7–10,13,23–27} There is universal agreement concerning the prudence of secondary prevention with ICDs for patients who have survived a cardiac arrest with documented VF or an episode of sustained VT, because of the relatively high recurrence rates in this subgroup.\textsuperscript{4}

However, selection of patients most likely to benefit from ICD therapy for primary prevention is less resolved, with guidelines an evolving process. Such stratification of risk in HCM is predicated on generally accepted noninvasive markers, usually in clinically stable patients, that have emerged from observational studies.\textsuperscript{13,15,22,23–27} Of note, this strategy differs conceptually from that in patients with CAD, in whom primary prevention is based largely on a single predominant risk marker emanating from randomized trials, in which a major clinical event (myocardial infarction) led to LV remodeling and systolic dysfunction (ejection fraction $\leq 30–35\%$), usually associated with disease progression.\textsuperscript{19}

The conventional primary prevention risk factors in HCM, which assume greater weight in younger patients $<50$ years of age, are: (1) family history of $\geq 1$ HCM-related SCDs; (2) $\geq 1$ episode of unexplained, recent syncope; (3) massive LV hypertrophy (thickness $\geq 30$ mm); (4) nonsustained VT on serial ambulatory 24-h (Holter) ECGs; and (5) hypotensive or attenuated blood pressure response to exercise (Figure 2). However, the exercise blood pressure response is tested less commonly than other risk factors\textsuperscript{13} and it rarely represents the sole indicator for a prophylactic implant in clinical practice,\textsuperscript{3,13} although it is often used as an arbitrator when the risk assessment is otherwise ambiguous. Also, as a matter of practice, isolated brief runs of nonsustained VT on single, random 24-h ambulatory (Holter) ECGs have not usually triggered decisions for a prophylactic ICD, although frequent and/or prolonged ($>10$ beats) bursts of nonsustained VT, or repetitive runs over extended serial monitoring periods, intuitively carry greater weight.

Potential Arbitrators

Certain features of HCM can contribute to resolution of ICD decisions when there is uncertainty regarding assignment of risk level, on a case-by-case basis (Figures 2, 3).

LV Apical Aneurysms. This recently reported subset of

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Figure 2. Risk stratification for sudden cardiac death (SCD). (Top) Pyramid profile currently used to identify patients at highest risk for SCD who are potential candidates for an implantable cardioverter-defibrillator (ICD). BP, blood pressure; LV, left ventricular; LVH, left ventricular hypertrophy; NSVT, nonsustained ventricular tachycardia; VT, ventricular tachycardia. “Following alcohol septal ablation, sustained VT has been reported in a significant minority of patients ($\approx 10\%$) over the short term.”\textsuperscript{36} (Reproduced with permission.) (Bottom) Direct relation between magnitude of LV hypertrophy (maximum [max] wall thickness by echocardiography) and SCD risk. Mild hypertrophy generally conveys lower risk and extreme hypertrophy (wall thickness $\geq 30$ mm) is associated with the highest risk.\textsuperscript{23} (Reproduced with permission.)
patients has been associated with a substantial (10%) annual event rate, largely because of the arrhythmogenic substrate created by the fibrotic thin-walled aneurysm, as well as the myocardial scarring identifiable by contrast cardiac magnetic resonance (CMR) in the contiguous distal LV wall.(27,28) Successful mapping and ablation of the VT focus associated with the aneurysm has been reported.(29)

**End-Stage Phase**  
In a small minority of HCM patients, approximately 3%, widespread (often transmural) LV scarring evolves to irreversible systolic dysfunction, often associated with wall thinning and cavity dilatation.(30) (Figure 3D). Inevitably, an adverse clinical course with progressive heart failure and often atrial and ventricular tachyarrhythmias ensues. ICDs are used in the end-stage as a bridge to heart transplantation.

**LV Outflow Obstruction**  
A gradient ≥30 mmHg at rest is a quantitative measure of elevated intraventricular pressure and wall stress. In 2 studies, obstruction had a very modest, though statistically significant, relationship to SCD risk (positive predictive value, only 5–10%).(31,32) but another investigation showed no such relationship.(33) Other obstacles to outflow obstruction as a primary risk factor include its dynamic nature, and the high frequency of gradients encountered in the HCM patient population (ie, 70%, at rest or with physiologic exercise).(34) Reduction of obstruction by myectomy (or alcohol ablation) is not considered a primary strategy for mitigating SCD risk.

**Delayed Enhancement (DE)**  
Because current risk stratification cannot guide SCD prevention in precise terms for each HCM patient, and SCD is known to occur in occasional patients without any conventional risk factors, there is the ongoing aspiration to identify more sensitive and specific clinical markers. Ideally, this would lead to a single, non-invasive, repeatable and accurate quantitative test (without adding to patient risk).

There is considerable interest surrounding in vivo detection of LV myocardial fibrosis on contrast-CMR as DE, and its relationship to determinants of SCD and disease progression.(35–39) DE has been linked to the underlying and unstable electrical substrate in HCM by the observation that non-

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**Figure 3.** Morphology of HCM patient subgroups associated with possible risk for sustained ventricular tachyarrhythmias. **(A)** Massive hypertrophy with ventricular septal (VS) thickness of 55 mm. **(B)** Akinetic thin-walled LV apical aneurysm with mid-cavity muscular apposition. D, distal (cavity); LA, left atrium; P, proximal (cavity); **(B1)** Contrast-cardiovascular magnetic resonance shows delayed enhancement (ie, scar) involving the thin aneurysm rim (arrowheads) and contiguous myocardium (large arrow); small apical thrombus is evident (small arrow). **(C)** Large transmural ventricular septal scar (arrow) resulting from alcohol ablation(45) (arrow) (reproduced with permission). **(D)** “End-stage” heart showing extensive and transmural septal scarring, extending into the anterior wall (arrowheads)(13) (Panels A, B, B1 and D) (reproduced with permission).
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sustained VT on ambulatory Holter ECG is more common in patients with DE than in patients without DE. Whether DE can be regarded as a bona fide risk marker in HCM will ultimately require adequately powered studies in large populations with sufficient numbers of events accrued over many years. However, solely the presence of DE in a given HCM patient is unlikely to become a reliable risk marker, given that fully 50% of all patients with this disease show some DE.

Alcohol Septal Ablation Percutaneous alcohol septal ablation is an alternative therapeutic strategy for selected HCM patients to the preferred option of surgical myectomy, with the capability of reducing the LV outflow gradient and heart failure symptoms, by creating a sizeable transmural scar occupying on average 30% of the ventricular septum and 10% of the overall LV chamber. There is accumulating evidence and concern that the potential arrhythmogenicity of the scar created by alcohol septal ablation can augment risk in the HCM population. Several studies have documented the occurrence of sustained ventricular arrhythmias and SCD following septal ablation in approximately 10% of patients with or without SCD risk factors. In a recent single center experience, tenCate et al from the Thoraxcenter reported that long-term outcome and survival was 4-fold less favorable with alcohol ablation compared to myectomy; more than 20% of ablation patients experienced sudden or other cardiac death, aborted sudden death, and/or appropriate ICD discharge, resulting in an annual event rate of 4.4% following ablation. This experience led the Rotterdam investigators to recommend myectomy as the preferred intervention for obstructive HCM. These findings are similar to those at Massachusetts General Hospital, where an event rate of 5%/year (for VT, VF, and/or appropriate ICD discharges) was reported. Furthermore, high-risk HCM patients implanted with ICDs after alcohol ablation have appropriate interventions for VT/VF of 3–10%/year, which is substantially higher than the extremely low incidence of sustained VT reported after myectomy (0.2–0.9%/year). Indeed, the multicenter HCM-ICD registry demonstrated the rate of appropriate ICD therapy among alcohol ablation patients with primary prevention ICDs to be 3-fold more frequent than in other patients (10.3%/year vs 3.6%/year). It is not entirely certain how commonly such arrhythmias are caused by the alcohol ablation procedure itself or alternatively to the underlying disease.

Whether HCM patients should have routine ICD implantation following alcohol septal ablation is unresolved. However, based on these considerations and concern that alcohol-induced infarcts could compound preexisting and underlying myocardial electrical instability, some practitio-
bers have regarded alcohol ablation as a risk arbitrator\textsuperscript{13,44,47} and prudently implanted ICDs in selected patients with other risk markers post-procedure (Figure 2).\textsuperscript{42}

**Uncertain Contributors to Risk**

**Atrial Fibrillation (AF)**
AF is the most common sustained arrhythmia in HCM (20–25\% of patients)\textsuperscript{49,50} associated with progressive heart failure and embolic stroke.\textsuperscript{16} However, there is no compelling evidence that paroxysmal AF is a predictor specifically of SCD in cohort analyses, although reported as an occasional trigger for VT.\textsuperscript{49}

**Genotyping**
It was 20 years ago that there was initial recognition that mutations in genes encoding proteins of the cardiac sarcomere cause HCM, which created substantial enthusiasm for identifying malignant or benign genetic substrates in order to facilitate assignment of SCD risk level.\textsuperscript{51} Although now commercially available, genetic testing has not achieved the initial expectation that it would become a reliable strategy for predicting prognosis or selecting patients for primary prevention ICDs.\textsuperscript{13,52} The gene-based hypothesis for risk stratification is clinically untenable, because of the heterogeneity of HCM, now with >1,000 mutations in ≥11 genes, including many mutations for which the pathologic significance remains unresolved.\textsuperscript{44} Selected clinical situations in which molecular diagnosis may predict prognosis have recently emerged, including LAMP2 cardiomyopathy,\textsuperscript{53} an X-linked lysosomal storage disease, and possibly double sarcomere mutations\textsuperscript{54} (Figure 5).

**Programmed Stimulation**
Laboratory electrophysiologic testing, which directly probes the electrical properties of the heart, has proved an impractical prognostic strategy now abandoned in HCM clinical practice as nonspecific, expensive, irrelevant with respect to clinical arrhythmias, and without advantage over current noninvasive risk stratification.\textsuperscript{13} Also,
there is insufficient evidence to regard specific 12-lead ECG patterns, T-wave alternans, or coronary arterial bridging as risk markers in HCM.\textsuperscript{13}

\textbf{Modifiable Risk Markers}

Participation in intense competitive sports can represent a potential risk factor in athletes with HCM, even when conventional markers are absent.\textsuperscript{6} The generally accepted recommendation of Bethesda Conference #36 for trained athletes with HCM to reduce SCD risk is the withdrawal from most competitive sports.\textsuperscript{55} In older HCM patients, coexistent obstructive CAD may increase the overall SCD risk, potentially modifiable by coronary arterial intervention.

The risk stratification algorithm in HCM (Figure 2) has proved to be a useful guide to selecting patients for ICD therapy, but a few SCDs have been reported in young HCM patients judged to be at low risk without any of the acknowledged risk markers, underscoring that the risk stratification strategy in HCM remains imprecise.\textsuperscript{13,56} Specifically, in preliminary data from the Minneapolis Heart Institute over the past 15 years, SCD events occurred in approximately 2\% (0.5%/year) of young apparently low-risk patients, without primary prevention ICDs.

\textbf{The Risk Factor Debate}

There has been a measure of controversy concerning the precise selection of HCM patients for prophylactic ICD strategies. Clinical practice in the European cardiovascular community has ascribed to a virtual mandatory minimum of 2 risk factors before recommending a prophylactic defibrillator.\textsuperscript{5,12} Certainly, the presence of multiple risk factors creates a clinical environment in which ICD decision-making is more intuitive and easier. Nevertheless, there is also considerable evidence that a single strong marker of increased risk within the clinical profile of an individual patient is sufficient to establish SCD risk as unacceptably increased, sufficient to raise the option of a primary prevention ICD.\textsuperscript{8-10} Indeed, an important proportion of appropriate ICD interventions for VT/VF occur in patients implanted because of only 1 risk factor (ie, 35\%) and device therapy interventions do not differ significantly between patients with 1, 2 or \geq 3 risk markers (Figure 6). Rigid adherence to a minimum of 2 risk markers before recommending a primary prevention ICD raises the possibility that some deserving 1-risk factor patients will be relegated to a lower level of consideration for ICD therapy, or left essentially unprotected.

On the other hand, not all 1-risk factor patients deserve consideration for an ICD. HCM is a heterogeneous genetic disease and inevitably there are numerous complex clinical scenarios involving ambiguities and gray areas with respect to the presence, strength or number of risk factors that ultimately affect ICD decision-making. One specific example is the elderly patient with syncope as a single risk factor. Such a patient may not be a candidate for primary prevention, given that HCM-related SCD is uncommon in this age range, survival to advanced age itself declares lower risk status in this disease, and syncope is relatively common in elderly individuals.

\textbf{ICD Experience in HCM}

\textbf{Efficacy}

There is now substantial evidence for the reliability of ICDs in preventing SCD in HCM patients.\textsuperscript{8-12,14-17,48,56-63} In selected subsets of high-risk patients, ICD interventions have proved to be both frequent and highly effective in aborting VT/VF. The largest and most recent of these studies is an international, multicenter retrospective/prospective registry assessment of 506 HCM patients from 42 centers in the U.S., Europe, and Australia\textsuperscript{9} (Figure 7).

Patients in this ICD registry were predominantly asymptomatic or with only mild limiting heart failure symptoms, and of relatively young age (average, 42 years old). Over a mean follow-up of only 3.7 years, appropriate device discharges (defibrillation shocks or antitachycardia pacing) for VF or rapid VT occurred in 20\% of patients (Figure 7), an implant-to-life-saved ratio of 5:1. The overall appropriate ICD discharge rate was 5.5%/year, at an average age of only 44 years. Cumulative probability of an appropriate ICD intervention 5 years after implant was almost 25\%, indicating the potential for ongoing SCD risk in the absence of an ICD.

The intervention rate for secondary prevention (implant after cardiac arrest) was highest (ie, 11%/year), and for primary prevention was 4%/year in those patients implanted based only on conventional risk factors (Figure 7). Consistent with the predilection of SCD in young patients with HCM, almost 30\% of those receiving ICDs were \leq 20 years of age, largely for primary prevention, and they experienced appropriate ICD interventions at an average of 18±4 years of age.
(ie, 7%/year). Intervention rates in the multicenter study are consistent with those reported individually from other countries, including Spain,16 Poland,17,58 Canada,12 Portugal,60 United Kingdom,51 Germany,59 Australia,11 Italy9 and other U.S. centers.48,62,63

It is notable that the ICD has performed so reliably in terminating VT/VF in patients with a disease such as HCM, which is associated with unique pathology and hemodynamic features such as a substantial increase in LV mass (wall thickening up to 3- to 5-fold greater than normal),23,64 dynamic obstruction to LV outflow with elevated intraventricular pressures,31,34 diastolic dysfunction and microvascular myocardial ischemia due to small vessel disease.65

In our large registry experience the ICD proved effective in preventing SCD in virtually every patient with the typical clinical and phenotypic expression of HCM (ie, in the absence of systolic dysfunction).8–10 However, a notable exception in the registry of 506 patients was a 21-year-old student with extreme LV hypertrophy, preserved systolic function, and syncope, who died suddenly when a mechanically defective ICD failed to deliver an appropriate shock because of massive electrical overstress resulting from short-circuiting,9,66 a design flaw known only to the manufacturer. Another exception is LAMP2 cardiomyopathy, a HCM phenocopy with massive LV hypertrophy, which appears largely refractory to ICD therapy (Figure 5).53

**Event Occurrence**

The timing of SCD events in HCM is largely unpredictable8–10,14,57,67 (Figures 7 and 8). The interval from ICD implant to first appropriate device intervention is quite variable, and often considerable in length (ie, as long as 10 years be-
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In addition, several patients have survived >10 years (and up to 30 years) after cardiac arrest with or without the aid of a defibrillator intervention.67

Therefore, initial clinical recognition of high-risk status and ICD implantation can precede by many years the occurrence of potentially lethal VT that the defibrillator is required to interrupt.5–10,13,57

Notably, development of disabling heart failure symptoms following major arrhythmic events appear to be uncommon in HCM,67 and at present there is no evidence that ICDs merely shift the mode of demise from SCD to progressive heart failure, as suggested in CAD.68 This observation raises the distinct possibility of achieving normal or near-normal longevity with (and because of) the ICD (Figure 9).

Once the potential risk is recognized and presented to the patient, it is difficult and probably imprudent to temporize or delay potentially preventive treatment. When the decision to implant an ICD in a high-risk HCM patient is made, it is likely to represent a life-long preventative measure.

Data on ICD performance in HCM patients are assembled out of necessity in cohorts judged to be at high risk in clinical practice, and therefore the reported appropriate ICD discharge rates may not be entirely representative of a truly general HCM population with a more benign profile.3,4,21 Also, reliance on retrospective, observational studies is unavoidable, because it is virtually impossible to design randomized ICD trials in this disease given its unique features, including the relatively low prevalence of HCM in cardiovascular practice, its heterogeneous clinical expression, and infrequent events dispersed over decades. Also, there are ethical and other considerations related to randomizing patients in a study involving a life-saving device.

HCM vs CAD

High-risk HCM patients are much younger on average with extended periods of future SCD risk (average 42 years at implant and 44 at first ICD intervention).3–17,23–28,48 compared to patients with CAD who are of relatively advanced
The decision to implant an ICD prophylactically for SCD prevention in HCM patients always includes the consideration of the potential complications and inconvenience incurred by a permanent device vs the obvious life-saving benefit should the ICD effectively terminate a lethal arrhythmia. Nevertheless, these 2 scenarios are not of equal weight, given the power of ICDs for preserving life. However, a measure of hesitancy toward life-long prophylactic ICDs may arise in pediatric cardiology practice, confronted by the clinical paradox in which active and apparently healthy young HCM patients are exposed to the greatest SCD risk (by age), but also device complications.

ICD-related complications are well documented, including infection, pocket hematoma, pneumothorax and venous thrombosis in a small minority. However, 25% of HCM patients experience inappropriate shocks (5/ year), resulting from lead fracture or dislodgement, oversensing, double counting and programming malfunctions, or when triggered inadvertently by sinus tachycardia or AF. The highest rate of complications is in younger HCM patients, largely because their activity level and body growth places a continual strain on the leads, which are regarded as the weakest link in the system. Extended lead survival is par-
ticularly crucial for young patients because they will have their ICDs for decades (probably most of their lives).

Industry-related ICD problems have disproportionately affected HCM patients, in which prominent recalls have included defective generators resulting in several deaths, and small-diameter high voltage leads prone to fracture. The implant procedure has been largely free of significant risk, without reported deaths, although selected patients with extreme hypertrophy, or who have been administered amiodarone, may require high-energy output generators or epicardial leads.

ICDs Worldwide: Cultural and Other Factors

The rate of prophylactic defibrillator implants in HCM patients is related in part to the country of origin as determined by differences in healthcare systems and a number of cultural, societal, and economic factors. The considerable variability among nations and cultures with respect to ICD implants undoubtedly affects clinical decision-making and, in particular, the threshold for recommending primary prevention devices to HCM patients. ICDs are most common in the U.S., more than 2-fold that of Germany and 5–10-fold greater than in most European countries, including the UK, where the mandatory 2-risk factor model for primary prevention prevails.

In Japan, other Asian countries, the Middle East, and Eastern Europe, HCM implant rates have been generally quite low, probably related primarily to cultural factors. These observations raise the distinct possibility that HCM patients with the same level of risk in different countries may not have the same access to prophylactic ICDs and the opportunity for SCD prevention.

ICD Decision-Making

Clinical dilemmas inevitably arise concerning ICD recommendations, because many HCM patients fall into ambiguous gray zones in which the level of risk cannot be assessed with precision using conventional risk factors (or arbitrators), and a measure of individual clinical judgment and experience of the managing physician with direct knowledge of the patient’s clinical profile and desires is necessary (Figure 10). Indeed, transparency, full disclosure and informed consent in association with autonomous input concerning the desires of the fully informed patient and family is necessary for resolving otherwise ambiguous decisions for prophylactic ICDs in which there are gaps in knowledge or absence of sufficient evidence-based data.

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