AMERICAN COLLEGE of CARDIOLOGY FOUNDATION

American Heart Association®

ACCF/AHA Pocket Guideline

Adapted from the 2011 ACCF/AHA

Guideline for the Diagnosis and Treatment of Hypertrophic Cardiomyopathy

November 2011

Developed in Collaboration With the American Association for Thoracic Surgery, American Society of Echocardiography, American Society of Nuclear Cardiology, Heart Failure Society of America, Heart Rhythm Society, Society for Cardiovascular Angiography and Interventions, and Society of Thoracic Surgeons

The following material was adapted from the 2011 ACCF/AHA Guidelines for the Diagnosis and Treatment of Hypertrophic Cardiomyopathy (J Am Coll Cardiol 2011;XX:XX–XX). This pocket guideline is available on the World Wide Web sites of the American College of Cardiology (www.cardiosource.org) and the American Heart Association (my.americanheart.org).

Contents

1. Introduction

The impetus for the guidelines is based on an appreciation of the frequency of this clinical entity and a realization that many aspects of clinical management, including the use of diagnostic modalities, genetic testing, utilization of implantable cardioverter-defibrillators (ICDs), and therapies for refractory symptoms lack consensus. The discussion and recommendations about the various diagnostic modalities apply to patients with established HCM and to a variable extent to patients with a high index of suspicion of the disease.

Classification of Recommendations

The ACCF/AHA classifications of recommendations and levels of evidence are utilized, and described in more detail in *Table 1*.

Table 1. Applying Classification of Recommendations and Level of Evidence

ESTIMATE OF CERTAINTY (PRECISION) OF TREATMENT EFFECT	SIZE OF TREATMENT EFFECT	
	CLASS I ***Benefit >>> Risk*** **Procedure/Treatment SHOULD be performed/ administered**	**CLASS IIa** ***Benefit >> Risk*** ***Additional studies with focused objectives needed*** **IT IS REASONABLE to perform procedure/administer treatment**
LEVEL A **Multiple populations evaluated*** **Data derived from multiple randomized clinical trials or meta-analyses**	■ Recommendation that procedure or treatment is useful/effective ■ Sufficient evidence from multiple randomized trials or meta-analyses	■ Recommendation in favor of treatment or procedure being useful/effective ■ Some conflicting evidence from multiple randomized trials or meta-analyses
LEVEL B **Limited populations evaluated*** **Data derived from a single randomized trial or nonrandomized studies**	■ Recommendation that procedure or treatment is useful/effective ■ Evidence from single randomized trial or nonrandomized studies	■ Recommendation in favor of treatment or procedure being useful/effective ■ Some conflicting evidence from single randomized trial or nonrandomized studies
LEVEL C **Very limited populations evaluated*** **Only consensus opinion of experts, case studies, or standard of care**	■ Recommendation that procedure or treatment is useful/effective ■ Only expert opinion, case studies, or standard of care	■ Recommendation in favor of treatment or procedure being useful/effective ■ Only diverging expert opinion, case studies, or standard of care
Suggested phrases for writing recommendations	should is recommended is indicated is useful/effective/beneficial	is reasonable can be useful/effective/beneficial is probably recommended or indicated
Comparative effectiveness phrases†	treatment/strategy A is recommended/indicated in preference to treatment B treatment A should be chosen over treatment B	treatment/strategy A is probably recommended/indicated in preference to treatment B it is reasonable to choose treatment A over treatment B

CLASS IIb	CLASS III *No Benefit* or CLASS III *Harm*
Benefit ≥ Risk ***Additional studies with broad objectives needed; additional registry data would be helpful*** **Procedure/Treatment MAY BE CONSIDERED**	**Procedure/Test** / **Treatment** **COR III: No benefit** — **Not Helpful** / **No Proven Benefit** **COR III: Harm** — **Excess Cost w/o Benefit or Harmful** / **Harmful to Patients**
■ **Recommendation's usefulness/efficacy less well established** ■ **Greater conflicting evidence from multiple randomized trials or meta-analyses**	■ **Recommendation that procedure or treatment is not useful/effective and may be harmful** ■ **Sufficient evidence from multiple randomized trials or meta-analyses**
■ **Recommendation's usefulness/efficacy less well established** ■ **Greater conflicting evidence from single randomized trial or nonrandomized studies**	■ **Recommendation that procedure or treatment is not useful/effective and may be harmful** ■ **Evidence from single randomized trial or nonrandomized studies**
■ **Recommendation's usefulness/efficacy less well established** ■ **Only diverging expert opinion, case studies, or standard of care**	■ **Recommendation that procedure or treatment is not useful/effective and may be harmful** ■ **Only expert opinion, case studies, or standard of care**

	COR III: No Benefit	COR III: Harm
may/might be considered	is not recommended	potentially harmful
may/might be reasonable	is not indicated	causes harm
usefulness/effectiveness is unknown/unclear/uncertain or not well established	should not be performed/ administered/ other	associated with excess morbidity/mortality
	is not useful/ beneficial/ effective	should not be performed/ administered/ other

A recommendation with Level of Evidence B or C does not imply that the recommendation is weak. Many important clinical questions addressed in the guidelines do not lend themselves to clinical trials. Although randomized trials are unavailable, there may be a very clear clinical consensus that a particular test or therapy is useful or effective.

* Data available from clinical trials or registries about the usefulness/efficacy in different subpopulations, such as sex, age, history of diabetes, history of prior myocardial infarction, history of heart failure, and prior aspirin use.

† For comparative effectiveness recommendations (Class I and IIa; Level of Evidence A and B only), studies that support the use of comparator verbs should involve direct comparisons of the treatments or strategies being evaluated.

2. Clinical Definition

The generally accepted definition of hypertrophic cardiomyopathy (HCM), is a disease state characterized by unexplained left ventricular (LV) hypertrophy associated with nondilated ventricular chambers in the absence of another cardiac or systemic disease that itself would be capable of producing the magnitude of hypertrophy evident in a given patient. Clinically, HCM is usually recognized by maximal LV wall thickness ≥15 mm, with wall thickness of 13 to 14 mm considered borderline, particularly in the presence of other compelling information (e.g., family history of HCM), based on echocardiography. In terms of LV wall-thickness measurements, the literature has been largely focused on echocardiography, although cardiovascular magnetic resonance (CMR) is now used with increasing frequency in HCM. In the case of children, increased LV wall thickness is defined as wall thickness ≥2 standard deviations above the mean (z score ≥2) for age, sex, or body size. However, it should be underscored that in principle, any degree of wall thickness is compatible with the presence of the HCM genetic substrate and that an emerging subgroup within the broad clinical spectrum is composed of family members with disease-causing sarcomere mutations but without evidence of the disease phenotype (i.e., LV hypertrophy).

3. Genetic Testing Strategies/Family Screening

Class I

1. Evaluation of familial inheritance and genetic counseling is recommended as part of the assessment of patients with HCM. (Level of Evidence: B)

2. Patients who undergo genetic testing should also undergo counseling by someone knowledgeable in the genetics of cardiovascular disease so that results and their clinical significance can be appropriately reviewed with the patient. (Level of Evidence: B)

3. Screening (clinical, with or without genetic testing) is recommended in first-degree relatives of patients with HCM. (Level of Evidence: B)

4. Genetic testing for HCM and other genetic causes of unexplained cardiac hypertrophy is recommended in patients with an atypical clinical presentation of HCM or when another genetic condition is suspected to be the cause. (Level of Evidence: B)

Class IIa	**1.** Genetic testing is reasonable in the index patient to facilitate the identification of first-degree family members at risk for developing HCM. (Level of Evidence: B)
Class IIb	**1.** The usefulness of genetic testing in the assessment of risk of sudden cardiac death (SCD) in HCM is uncertain. (Level of Evidence: B)
Class III: No Benefit	**1.** Genetic testing is not indicated in relatives when the index patient does not have a definitive pathogenic mutation. (Level of Evidence: B) **2.** Ongoing clinical screening is not indicated in genotype-negative relatives in families with HCM. (Level of Evidence: B)

4. Genotype-Positive/Phenotype-Negative Patients

Class I

1. In individuals with pathogenic mutations who do not express the HCM phenotype, it is recommended to perform serial electrocardiogram, transthoracic echocardiogram (TTE), and clinical assessment at periodic intervals (12 to 18 months in children and adolescents and about every 5 years in adults), based on the patient's age and change in clinical status. (Level of Evidence: B)

5. Echocardiography

Class I

1. A TTE is recommended in the initial evaluation of all patients with suspected HCM. (Level of Evidence: B)

2. A TTE is recommended as a component of the screening algorithm for family members of patients with HCM unless the family member is genotype negative in a family with known definitive mutations. (Level of Evidence: B)

3. Periodic (12 to 18 months) TTE screening is recommended for children of patients with HCM, starting by age 12 or earlier if a growth spurt or signs of puberty are evident and/or when there are plans for engaging in intense competitive sports or there is a family history of SCD. (Level of Evidence: C)

4. Repeat TTE is recommended for the evaluation of patients with HCM with a change in clinical status or new cardiovascular event. (Level of Evidence: B)

5. A transesophageal echocardiogram (TEE) is recommended for the intraoperative guidance of surgical myectomy. (Level of Evidence: B)

6. TTE or TEE with intracoronary contrast injection of the candidate's septal perforator(s) is recommended for the intraprocedural guidance of alcohol septal ablation. (Level of Evidence: B)

7. TTE should be used to evaluate the effects of surgical myectomy or alcohol septal ablation for obstructive HCM. (Level of Evidence: C)

Class IIa

1. TTE studies performed every 1 to 2 years can be useful in the serial evaluation of symptomatically stable patients with HCM to assess the degree of myocardial hypertrophy, dynamic obstruction, and myocardial function. (Level of Evidence: C)

2. Exercise TTE can be useful in the detection and quantification of dynamic left ventricular outflow tract (LVOT) obstruction in the absence of resting outflow tract obstruction in patients with HCM. (Level of Evidence: B)

3. TEE can be useful if TTE is inconclusive for clinical decision making about medical therapy and in situations such as planning for myectomy, exclusion of subaortic membrane or mitral regurgitation secondary to structural abnormalities of the mitral valve apparatus, or in assessment for the feasibility of alcohol septal ablation. (Level of Evidence: C)

4. TTE combined with the injection of an intravenous contrast agent is reasonable if the diagnosis of apical HCM or apical infarction or severity of hypertrophy is in doubt, particularly when other imaging modalities such as CMR are not readily available, not diagnostic, or contraindicated. (Level of Evidence: C)

5. Serial TTE studies are reasonable for clinically unaffected patients who have a first-degree relative with HCM when genetic status is unknown. Such follow-up may be considered every 12 to 18 months for children or adolescents from high-risk families and every 5 years for adult family members. (Level of Evidence: C)

Class III: No Benefit

1. TTE studies should not be performed more frequently than every 12 months in patients with HCM when it is unlikely that any changes have occurred that would have an impact on clinical decision making. (Level of Evidence: C)

2. Routine TEE and/or contrast echocardiography is not recommended when TTE images are diagnostic of HCM and/or there is no suspicion of fixed obstruction or intrinsic mitral valve pathology. (Level of Evidence: C)

6. Stress Testing

Class IIa

1. Treadmill exercise testing is reasonable to determine functional capacity and response to therapy in patients with HCM. (Level of Evidence: C)

2. Treadmill testing with monitoring of an electrocardiogram and blood pressure is reasonable for SCD risk stratification in patients with HCM. (Level of Evidence: B)

3. In patients with HCM who do not have a resting peak instantaneous gradient of greater than or equal to 50 mm Hg, exercise echocardiography is reasonable for the detection and quantification of exercise-induced dynamic LVOT obstruction. (Level of Evidence: B)

7. Cardiac Magnetic Resonance

Class I

1. CMR imaging is indicated in patients with suspected HCM when echocardiography is inconclusive for diagnosis. (Level of Evidence: B)

2. CMR imaging is indicated in patients with known HCM when additional information that may have an impact on management or decision making regarding invasive management, such as magnitude and distribution of hypertrophy or anatomy of the mitral valve apparatus or papillary muscles, is not adequately defined with echocardiography. (Level of Evidence: B)

Class IIa

1. CMR imaging is reasonable in patients with HCM to define apical hypertrophy and/or aneurysm if echocardiography is inconclusive. (Level of Evidence: B)

Class IIb

1. In selected patients with known HCM, when SCD risk stratification is inconclusive after documentation of the conventional risk factors, CMR imaging with assessment of late gadolinium enhancement may be considered in resolving clinical decision making. (Level of Evidence: C)

2. CMR imaging may be considered in patients with LV hypertrophy and the suspicion of alternative diagnoses to HCM, including cardiac amyloidosis, Fabry disease, and genetic phenocopies such as *LAMP2* cardiomyopathy. (Level of Evidence: C)

8. Detection of Concomitant Coronary Disease

Class I	**1.** Coronary arteriography (invasive or computed tomographic imaging) is indicated in patients with HCM with chest discomfort who have an intermediate to high likelihood of coronary artery disease (CAD) when the identification of concomitant CAD will change management strategies. (Level of Evidence: C)
Class IIa	**1.** Assessment of coronary anatomy with computed tomographic angiography is reasonable for patients with HCM with chest discomfort and a low likelihood of CAD to assess for possible concomitant CAD. (Level of Evidence: C) **2.** Assessment of ischemia or perfusion abnormalities suggestive of CAD with single-photon emission computed tomography or positron emission tomography myocardial perfusion imaging (because of excellent negative predictive value) is reasonable in patients with HCM with chest discomfort and a low likelihood of CAD to rule out possible concomitant CAD. (Level of Evidence: C)

Class III: No Benefit

1. Routine single-photon emission computed tomography myocardial perfusion imaging or stress echocardiography is not indicated for detection of "silent" CAD-related ischemia in patients with HCM who are asymptomatic. (Level of Evidence: C)

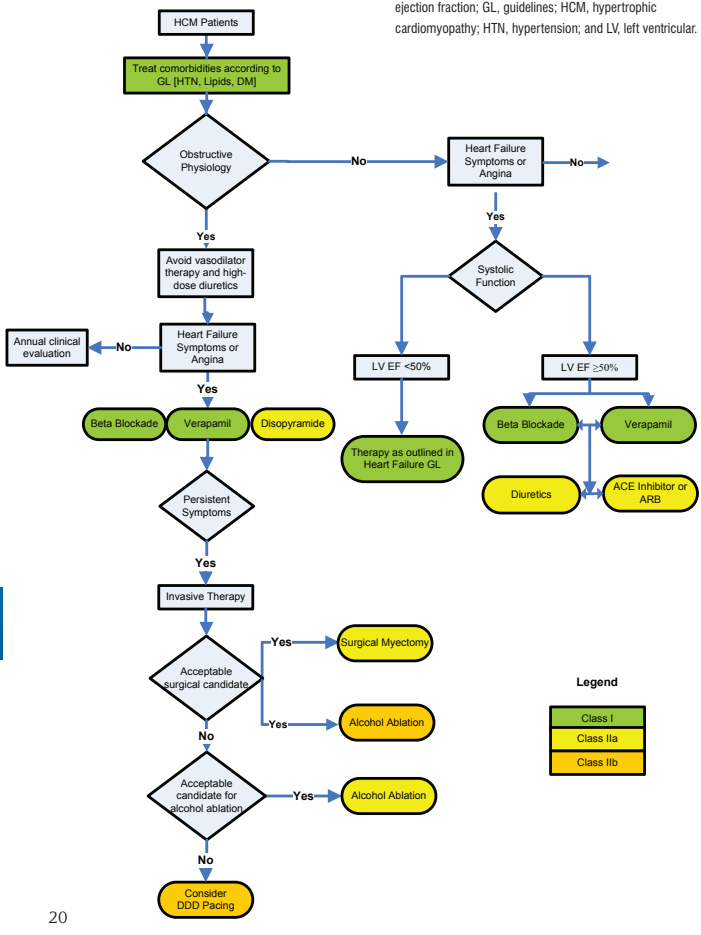
2. Assessment for the presence of blunted flow reserve (microvascular ischemia) using quantitative myocardial blood flow measurements by positron emission tomography is not indicated for the assessment of prognosis in patients with HCM. (Level of Evidence: C)

9. Asymptomatic Patients

Class I	**1.** For patients with HCM, it is recommended that comorbidities that may contribute to cardiovascular disease (e.g., hypertension, diabetes, hyperlipidemia, obesity) be treated in compliance with relevant existing guidelines. (Level of Evidence: C)
Class IIa	**1.** Low-intensity aerobic exercise is reasonable as part of a healthy lifestyle for patients with HCM. (Level of Evidence: C)
Class IIb	**1.** The usefulness of beta blockade and calcium channel blockers to alter clinical outcome is not well established for the management of asymptomatic patients with HCM with or without obstruction. (Level of Evidence: C)
Class III: Harm	**1.** Septal reduction therapy should not be performed for asymptomatic adult and pediatric patients with HCM with normal effort tolerance regardless of the severity of obstruction. (Level of Evidence: C) **2.** In patients with HCM with resting or provocable outflow tract obstruction, regardless of symptom status, pure vasodilators and high-dose diuretics are potentially harmful. (Level of Evidence: C)

Figure 1. Treatment Algorithm

ACE indicates angiotensin-converting enzyme; ARB, angiotensin receptor blocker; DM, diabetes mellitus; EF, ejection fraction; GL, guidelines; HCM, hypertrophic cardiomyopathy; HTN, hypertension; and LV, left ventricular.



10. Pharmacologic Management

Class I

1. Beta-blocking drugs are recommended for the treatment of symptoms (angina or dyspnea) in adult patients with obstructive or nonobstructive HCM but should be used with caution in patients with sinus bradycardia or severe conduction disease. (Level of Evidence: B)

2. If low doses of beta-blocking drugs are ineffective for controlling symptoms (angina or dyspnea) in patients with HCM, it is useful to titrate the dose to a resting heart rate of less than 60 to 65 bpm (up to generally accepted and recommended maximum doses of these drugs). (Level of Evidence: B)

3. Verapamil therapy (starting in low doses and titrating up to 480 mg/d) is recommended for the treatment of symptoms (angina or dyspnea) in patients with obstructive or nonobstructive HCM who do not respond to beta-blocking drugs or who have side effects or contraindications to beta-blocking drugs. However, verapamil should be used with caution in patients with high gradients, advanced heart failure, or sinus bradycardia. (Level of Evidence: B)

4. Intravenous phenylephrine (or another pure vasoconstricting agent) is recommended for the treatment of acute hypotension in patients with obstructive HCM who do not respond to fluid administration. (Level of Evidence: B)

Class IIa

1. It is reasonable to combine disopyramide with a beta-blocking drug or verapamil in the treatment of symptoms (angina or dyspnea) in patients with obstructive HCM who do not respond to beta-blocking drugs or verapamil alone. (Level of Evidence: B)

2. It is reasonable to add oral diuretics in patients with nonobstructive HCM when dyspnea persists despite the use of beta blockers or verapamil or their combination. (Level of Evidence: C)

Class IIb

1. Beta-blocking drugs might be useful in the treatment of symptoms (angina or dyspnea) in children or adolescents with HCM, but patients treated with these drugs should be monitored for side effects, including depression, fatigue, or impaired scholastic performance. (Level of Evidence: C)

2. It may be reasonable to add oral diuretics with caution to patients with obstructive HCM when congestive symptoms persist despite the use of beta blockers or verapamil or their combination. (Level of Evidence: C)

3. The usefulness of angiotensin-converting enzyme inhibitors or angiotensin receptor blockers in the treatment of symptoms (angina or dyspnea) in patients with HCM with preserved systolic function is not well established, and these drugs should be used cautiously (if at all) in patients with resting or provocable LVOT obstruction. (Level of Evidence: C)

4. In patients with HCM who do not tolerate verapamil or in whom verapamil is contraindicated, diltiazem may be considered. (Level of Evidence: C)

Class III: Harm

1. Nifedipine or other dihydropyridine calcium channel-blocking drugs are potentially harmful for treatment of symptoms (angina or dyspnea) in patients with HCM who have resting or provocable LVOT obstruction. (Level of Evidence: C)

2. Verapamil is potentially harmful in patients with obstructive HCM in the setting of systemic hypotension or severe dyspnea at rest. (Level of Evidence: C)

3. Digitalis is potentially harmful in the treatment of dyspnea in patients with HCM and in the absence of atrial fibrillation (AF). (Level of Evidence: B)

4. The use of disopyramide alone without beta blockers or verapamil is potentially harmful in the treatment of symptoms (angina or dyspnea) in patients with HCM with AF because disopyramide may enhance atrioventricular conduction and increase the ventricular rate during episodes of AF. (Level of Evidence: B)

5. Dopamine, dobutamine, norepinephrine, and other intravenous positive inotropic drugs are potentially harmful for the treatment of acute hypotension in patients with obstructive HCM. (Level of Evidence: B)

11. Invasive Therapies

Class I

1. Septal reduction therapy should be performed only by experienced operators* in the context of a comprehensive HCM clinical program and only for the treatment of eligible patients with severe drug-refractory symptoms and LVOT obstruction†. (Level of Evidence: C)

*Experienced operators are defined as an individual operator with a cumulative case volume of at least 20 procedures or an individual operator who is working in a dedicated HCM program with a cumulative total of at least 50 procedures.

†Eligible patients are defined by all of the following:

a. Clinical: Severe dyspnea or chest pain (usually New York Heart Association functional classes III or IV) or occasionally other exertional symptoms (such as syncope or near syncope) that interfere with everyday activity or quality of life despite optimal medical therapy.
b. Hemodynamic: Dynamic LVOT gradient at rest or with physiologic provocation greater than or equal to 50 mm Hg associated with septal hypertrophy and systolic anterior motion of the mitral valve.
c. Anatomic: Targeted anterior septal thickness sufficient to perform the procedure safely and effectively in the judgment of the individual operator.

Class IIa

1. Consultation with centers experienced in performing both surgical septal myectomy and alcohol septal ablation is reasonable when discussing treatment options for eligible patients with HCM with severe drug-refractory symptoms and LVOT obstruction. (Level of Evidence: C)

2. Surgical septal myectomy, when performed in experienced centers, can be beneficial and is the first consideration for the majority of eligible patients with HCM with severe drug-refractory symptoms and LVOT obstruction. (Level of Evidence: B)

3. Surgical septal myectomy, when performed at experienced centers, can be beneficial in symptomatic children with HCM and severe resting obstruction (>50 mm Hg) for whom standard medical therapy has failed. (Level of Evidence: C)

4. When surgery is contraindicated or the risk is considered unacceptable because of serious comorbidities or advanced age, alcohol septal ablation, when performed in experienced centers, can be beneficial in eligible adult patients with HCM with LVOT obstruction and severe drug-refractory symptoms (usually New York Heart Association functional classes III or IV). (Level of Evidence: B)

Class IIb	**1.** Alcohol septal ablation, when performed in experienced centers, may be considered as an alternative to surgical myectomy for eligible adult patients with HCM with severe drug-refractory symptoms and LVOT obstruction when, after a balanced and thorough discussion, the patient expresses a preference for septal ablation. (Level of Evidence: B)
	2. The effectiveness of alcohol septal ablation is uncertain in patients with HCM with marked (i.e., >30 mm) septal hypertrophy, and therefore the procedure is generally discouraged in such patients. (Level of Evidence: C)
Class III: Harm	**1.** Septal reduction therapy should not be done for adult patients with HCM who are asymptomatic with normal exercise tolerance or whose symptoms are controlled or minimized on optimal medical therapy. (Level of Evidence: C)
	2. Septal reduction therapy should not be done unless performed as part of a program dedicated to the longitudinal and multidisciplinary care of patients with HCM. (Level of Evidence: C)

3. Mitral valve replacement for relief of LVOT obstruction should not be performed in patients with HCM in whom septal reduction therapy is an option. (Level of Evidence: C)

4. Alcohol septal ablation should not be done in patients with HCM with concomitant disease that independently warrants surgical correction (e.g., coronary artery bypass grafting for CAD, mitral valve repair for ruptured chordae) in whom surgical myectomy can be performed as part of the operation. (Level of Evidence: C)

5. Alcohol septal ablation should not be done in patients with HCM who are less than 21 years of age and is discouraged in adults less than 40 years of age if myectomy is a viable option. (Level of Evidence: C)

12. Pacing

Class IIa	**1.** In patients with HCM who have had a dual-chamber device implanted for non-HCM indications, it is reasonable to consider a trial of dual-chamber atrial-ventricular pacing (from the right ventricular apex) for the relief of symptoms attributable to LVOT obstruction. (Level of Evidence: B)
Class IIb	**1.** Permanent pacing may be considered in medically refractory symptomatic patients with obstructive HCM who are suboptimal candidates for septal reduction therapy. (Level of Evidence: B)
Class III: No Benefit	**1.** Permanent pacemaker implantation for the purpose of reducing gradient should not be performed in patients with HCM who are asymptomatic or whose symptoms are medically controlled. (Level of Evidence: C) **2.** Permanent pacemaker implantation should not be performed as a first-line therapy to relieve symptoms in medically refractory symptomatic patients with HCM and LVOT obstruction in patients who are candidates for septal reduction. (Level of Evidence: B)

13. Sudden Cardiac Death Risk Stratification

Class I

1. All patients with HCM should undergo comprehensive SCD risk stratification at initial evaluation to determine the presence of: (Level of Evidence: B)

a. A personal history for ventricular fibrillation, sustained ventricular tachycardia, or SCD events, including appropriate ICD therapy for ventricular tachyarrhythmias.*
b. A family history for SCD events, including appropriate ICD therapy for ventricular tachyarrhythmias.*
c. Unexplained syncope.
d. Documented nonsustained ventricular tachycardia (NSVT) defined as 3 or more beats at greater than or equal to120 bpm on ambulatory (Holter) electrocardiogram.
e. Maximal LV wall thickness greater than or equal to 30 mm.

*Appropriate ICD discharge is defined as ICD therapy triggered by VT or ventricular fibrillation, documented by stored intracardiac electrogram or cycle-length data, in conjunction with the patient's symptoms immediately before and after device discharge.

Class IIa

1. It is reasonable to assess blood pressure response during exercise as part of SCD risk stratification in patients with HCM. (Level of Evidence: B)

2. SCD risk stratification is reasonable on a periodic basis (every 12 to 24 months) for patients with HCM who have not undergone ICD implantation but would otherwise be eligible in the event that risk factors are identified (12 to 24 months). (Level of Evidence: C)

Class IIb

1. The usefulness of the following potential SCD risk modifiers is unclear but might be considered in selected patients with HCM for whom risk remains borderline after documentation of conventional risk factors:

a. CMR imaging with late gadolinium enhacement. (Level of Evidence: C)

b. Double and compound mutations (i.e., >1). (Level of Evidence: C)

c. Marked LVOT obstruction. (Level of Evidence: B)

Class III: Harm

1. Invasive electrophysiologic testing as routine SCD risk stratification in patients with HCM should not be performed. (Level of Evidence: C)

14. Selection of Patients for Implantable Cardioverter-Defibrillators

Class I

1. The decision to place an ICD in patients with HCM should include application of individual clinical judgment, as well as a thorough discussion of the strength of evidence, benefits, and risks to allow the informed patient's active participation in decision making. (Level of Evidence: C)

2. ICD placement is recommended for patients with HCM with prior documented cardiac arrest, ventricular fibrillation, or hemodynamically significant ventricular tachycardia. (Level of Evidence: B)

Class IIa

1. It is reasonable to recommend an ICD for patients with HCM with:

a. Sudden death presumably caused by HCM in 1 or more first-degree relatives. (Level of Evidence: C)
b. A maximum LV wall thickness greater than or equal to 30 mm. (Level of Evidence: C)
c. One or more recent, unexplained syncopal episodes. (Level of Evidence: C)

2. An ICD can be useful in select patients with NSVT (particularly those <30 years of age) in the presence of other SCD risk factors or modifiers*. (Level of Evidence: C)

*See Section 6.3.1.2 of the full-text guideline for SCD risk factors or modifiers.

3. An ICD can be useful in select patients with HCM with an abnormal blood pressure response with exercise in the presence of other SCD risk factors or modifiers.* (Level of Evidence: C)

4. It is reasonable to recommend an ICD for high-risk children with HCM, based on unexplained syncope, massive LV hypertrophy, or family history of SCD, after taking into account the relatively high complication rate of long-term ICD implantation. (Level of Evidence: C)

Class IIb

1. The usefulness of an ICD is uncertain in patients with HCM with isolated bursts of nonsustained ventricular tachycardia when in the absence of any other SCD risk factors or modifiers.* (Level of Evidence: C)

2. The usefulness of an ICD is uncertain in patients with HCM with an abnormal blood pressure response with exercise when in the absence of any other SCD risk factors or modifiers*, particularly in the presence of significant outflow obstruction. (Level of Evidence: C)

*See Section 6.3.1.2 of the full-text guideline for SCD risk factors or modifiers.

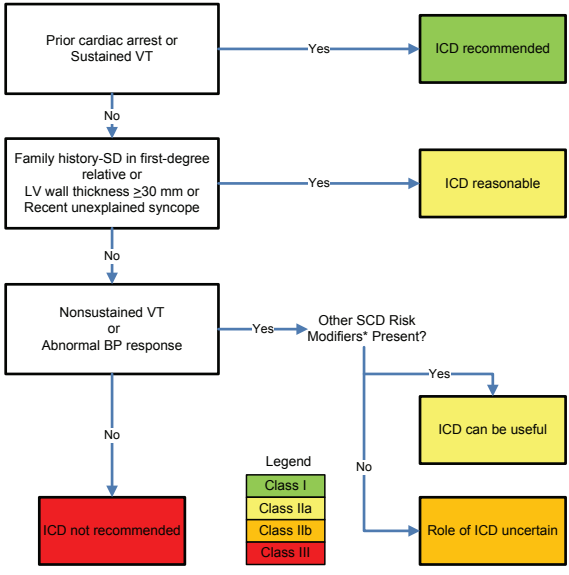
Class III: Harm

1. ICD placement as a routine strategy in patients with HCM without an indication of increased risk is potentially harmful. (Level of Evidence: C)

2. ICD placement as a strategy to permit patients with HCM to participate in competitive athletics is potentially harmful. (Level of Evidence: C)

3. ICD placement in patients who have an identified HCM genotype in the absence of clinical manifestations of HCM is potentially harmful. (Level of Evidence: C)

Figure 2. Indications for ICDs in HCM



Regardless of the level of recommendation put forth in these guidelines, the decision for placement of an ICD must involve prudent application of individual clinical judgment, thorough discussions of the strength of evidence, the benefits, and the risks (including but not limited to inappropriate discharges, lead and procedural complications) to allow active participation of the fully informed patient in ultimate decision making.

BP indicates blood pressure; ICD, implantable cardioverter-defibrillator; LV, left ventricular; SCD, sudden cardiac death; SD, sudden death; and VT, ventricular tachycardia.

15. Participation in Competitive or Recreational Sports and Physical Activity

Class IIa	**1.** It is reasonable for patients with HCM to participate in low-intensity competitive sports (e.g., golf and bowling). (Level of Evidence: C)
	2. It is reasonable for patients with HCM to participate in a range of recreational sporting activities as outlined in Table 2. (Level of Evidence: C)
Class III: Harm	**1.** Patients with HCM should not participate in intense competitive sports regardless of age, sex, race, presence or absence of LVOT obstruction, prior septal reduction therapy, or implantation of a cardioverter-defibrillator for high-risk status. (Level of Evidence: C)

Table 2. Recommendations for the Acceptability of Recreational (Noncompetitive) Sports Activities and Exercise in Patients With HCM*

Intensity Level	Eligibility Scale for HCM†
High	
Basketball (full court)	0
Basketball (half court)	0
Body building‡	1
Gymnastics	2
Ice hockey‡	0
Racquetball/squash	0
Rock climbing‡	1
Running (sprinting)	0
Skiing (downhill)‡	2
Skiing (cross-country)	2
Soccer	0
Tennis (singles)	0
Touch (flag) football	1
Windsurfing§	1
Moderate	
Baseball/softball	2
Biking	4
Modest hiking	4
Motorcycling‡	3
Jogging	3
Sailing§	3
Surfing§	2
Swimming (laps)§	5
Tennis (doubles)	4
Treadmill/stationary bicycle	5
Weightlifting (free weights)‡‖	1
Hiking	3
Low	
Bowling	5
Golf	5
Horseback riding‡	3
Scuba diving§	0
Skating¶	5
Snorkeling§	5
Weights (nonfree weights)	4
Brisk walking	5

*Recreational sports are categorized according to high, moderate, and low levels of exercise and graded on a relative scale (from 0 to 5) for eligibility, with 0 to 1 indicating generally not advised or strongly discouraged; 4 to 5, probably permitted; and 2 to 3, intermediate and to be assessed clinically on an individual basis. The designations of high, moderate, and low levels of exercise are equivalent to an estimated >6, 4 to 6, and <4 metabolic equivalents, respectively.

†Assumes absence of laboratory DNA genotyping data; therefore, limited to clinical diagnosis.

‡These sports involve the potential for traumatic injury, which should be taken into consideration for individuals with a risk for impaired consciousness.

§The possibility of impaired consciousness occurring during water-related activities should be taken into account with respect to the individual patient's clinical profile.

‖Recommendations generally differ from those for weight-training machines (nonfree weights), based largely on the potential risk of traumatic injury associated with episodes of impaired consciousness during bench-press maneuvers; otherwise, the physiologic effects of all weight-training activities are regarded as similar with respect to the present recommendations.

¶Individual sporting activity not associated with the team sport of ice hockey.

16. Management of Atrial Fibrillation

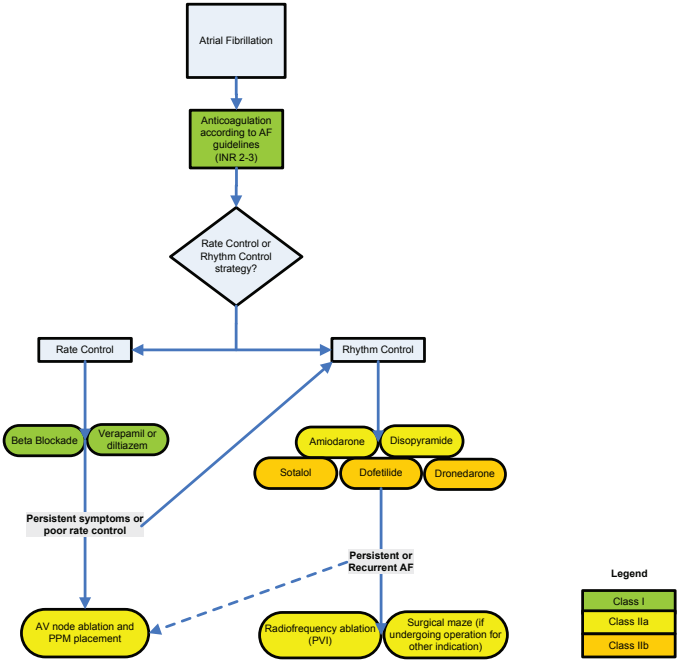
Class I

1. Anticoagulation with vitamin K antagonists (i.e., warfarin, to an international normalized ratio of 2.0 to 3.0) is indicated in patients with paroxysmal, persistent, or chronic AF and HCM. (Anticoagulation with direct thrombin inhibitors [i.e., dabigatran*] may represent another option to reduce the risk of thromboembolic events, but data for patients with HCM are not available). (Level of Evidence: C)

2. Ventricular rate control in patients with HCM with AF is indicated for rapid ventricular rates and can require high doses of beta antagonists and nondihydropyridine calcium channel blockers. (Level of Evidence: C)

* Dabigatran should not be used in patients with prosthetic valves, hemodynamically significant valve disease, advanced liver failure, or severe renal failure (creatinine clearance <15 mL/min).

Figure 3. Management of AF in HCM



AF indicates atrial fibrillation; AV, atrioventricular; INR, international normalized ratio; PPM, permanent pacemaker; and PVI, pulmonary vein isolation.

Class IIa

1. Disopyramide (with ventricular rate-controlling agents) and amiodarone are reasonable antiarrhythmic agents for AF in patients with HCM. (Level of Evidence: B)

2. Radiofrequency ablation for AF can be beneficial in patients with HCM who have refractory symptoms or who are unable to take antiarrhythmic drugs. (Level of Evidence: B)

3. Maze procedure with closure of left atrial appendage is reasonable in patients with HCM with a history of AF, either during septal myectomy or as an isolated procedure in selected patients. (Level of Evidence: C)

Class IIb

1. Sotalol, dofetilide, and dronedarone might be considered alternative antiarrhythmic agents in patients with HCM, especially in those with an ICD, but clinical experience is limited. (Level of Evidence: C)

17. Pregnancy/Delivery

Class I

1. In women with HCM who are asymptomatic or whose symptoms are controlled with beta-blocking drugs, the drugs should be continued during pregnancy, but increased surveillance for fetal bradycardia or other complications is warranted. (Level of Evidence: C)

2. For patients (mother or father) with HCM, genetic counseling is indicated before planned conception. (Level of Evidence: C)

3. In women with HCM and resting or provocable LVOT obstruction greater than or equal to 50 mm Hg and/or cardiac symptoms not controlled by medical therapy alone, pregnancy is associated with increased risk, and these patients should be referred to a high-risk obstetrician. (Level of Evidence: C)

4. The diagnosis of HCM among asymptomatic women is not considered a contraindication for pregnancy, but patients should be carefully evaluated in regard to the risk of pregnancy. (Level of Evidence: C)

Class IIa	**1.** For women with HCM whose symptoms are controlled (mild to moderate), pregnancy is reasonable, but expert maternal/fetal medical specialist care, including cardiovascular and prenatal monitoring, is advised. (Level of Evidence: C)
Class III: Harm	**1.** For women with advanced heart failure symptoms and HCM, pregnancy is associated with excess morbidity/mortality. (Level of Evidence: C)

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