2011 ACCF/AHA Guideline for the Diagnosis and Treatment of Hypertrophic Cardiomyopathy

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

Helping Cardiovascular Professionals Learn. Advance. Heal.

Citation

This slide set was adapted from the 2011 ACCF/AHA Guideline for the Diagnosis and Treatment of Hypertrophic Cardiomyopathy (*Journal of the American College of Cardiology).* Published on November 8, 2011, available at: http://content.onlinejacc.org/cgi/content/full/j.jacc.2011.06.011

The full-text guidelines are also available on the following Web sites:
ACC (www.cardiosource.org) and AHA (my.americanheart.org)

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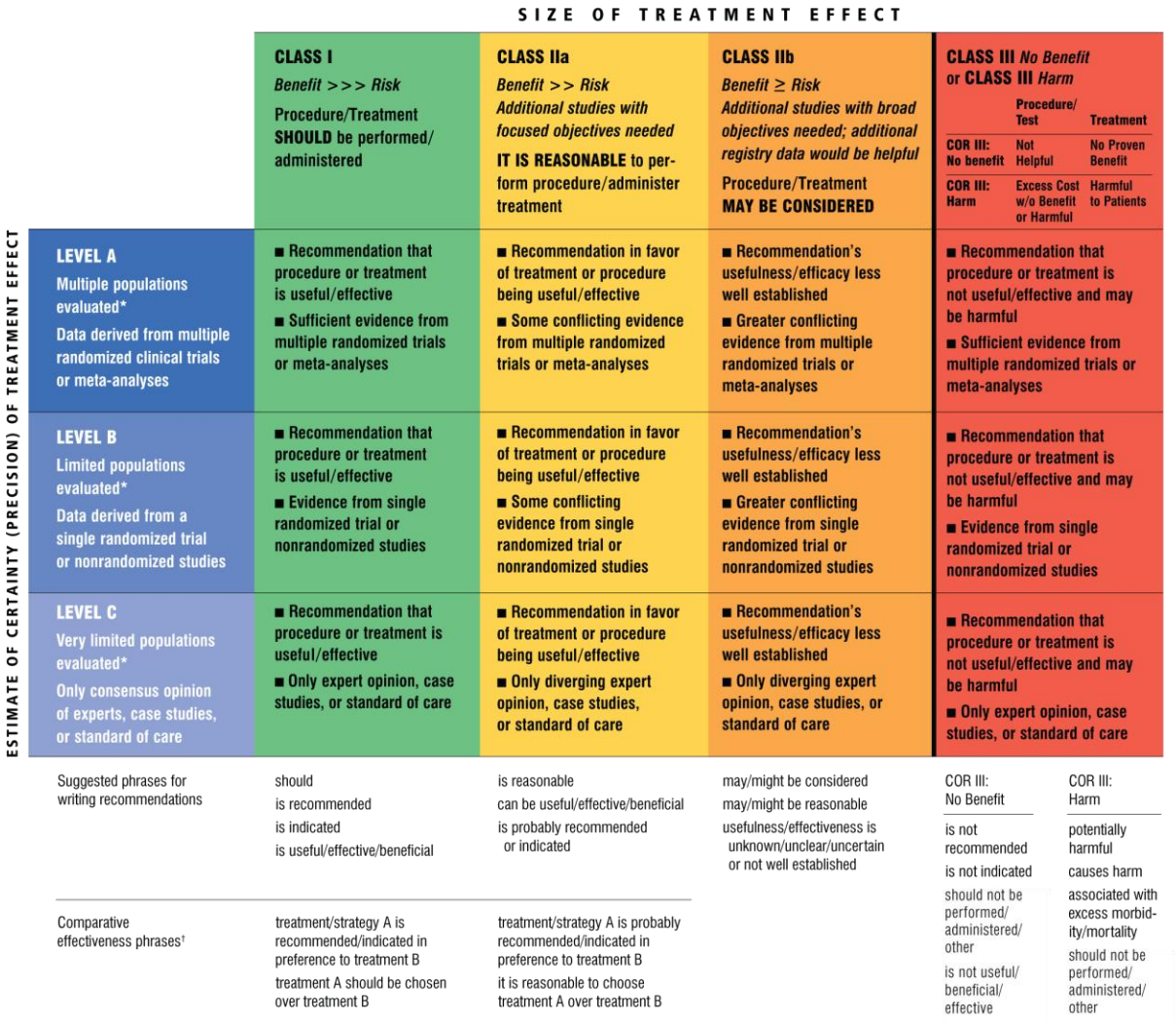
Classification of Recommendations and Levels of Evidence



SIZE OF TREATMENT EFFECT

ESTIMATE OF CERTAINTY (PRECISION) 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	CLASS IIb *Benefit ≥ Risk* *Additional studies with broad objectives needed; additional registry data would be helpful* Procedure/Treatment **MAY BE CONSIDERED**	CLASS III *No Benefit* or CLASS III *Harm* 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
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	■ 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
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	■ 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
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	■ 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
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	may/might be considered may/might be reasonable usefulness/effectiveness is unknown/unclear/uncertain or not well established	COR III: No Benefit: is not recommended; is not indicated; should not be performed/ administered/ other; is not useful/ beneficial/ effective COR III: Harm: potentially harmful; causes harm; associated with excess morbidity/mortality; should not be performed/ administered/ other
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		

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.

Diagnosis

Genetic Testing Strategies/Family Screening

Genetic Testing Strategies/Family Screening

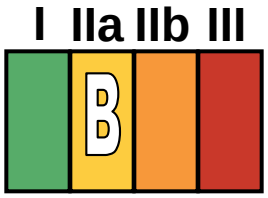
Evaluation of familial inheritance and genetic counseling is recommended as part of the assessment of patients with HCM.

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.

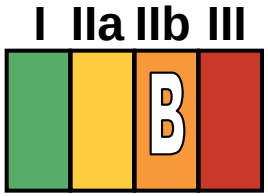
Screening (clinical, with or without genetic testing) is recommended in first-degree relatives of patients with HCM.

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.

Genetic Testing Strategies/Family Screening



Genetic testing is reasonable in the index patient to facilitate the identification of first-degree family members at risk for developing HCM.



The usefulness of genetic testing in the assessment of risk of SCD in HCM is uncertain.

Genetic Testing Strategies/Family Screening

No Benefit

Genetic testing is not indicated in relatives when the index patient does not have a definitive pathogenic mutation.

No Benefit

Ongoing clinical screening is not indicated in genotypenegative relatives in families with HCM.

Genotype-Positive/Phenotype-Negative Patients

Genotype-Positive/Phenotype-Negative Patients

In individuals with pathogenic mutations who do not express the HCM phenotype, it is recommended to perform serial ECG, 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.

Electrocardiography

Electrocardiography

A 12-lead ECG is recommended in the initial evaluation of patients with HCM.

Twenty-four–hour ambulatory (Holter) electrocardiographic monitoring is recommended in the initial evaluation of patients with HCM to detect VT and identify patients who may be candidates for ICD therapy.

Twenty-four–hour ambulatory (Holter) electrocardiographic monitoring or event recording is recommended in patients with HCM who develop palpitations or lightheadedness.

Electrocardiography

A repeat ECG is recommended for patients with HCM when there is worsening of symptoms.

A 12-lead ECG is recommended every 12 to 18 months as a component of the screening algorithm for adolescent first degree relatives of patients with HCM who have no evidence of hypertrophy on echocardiography.

A 12-lead ECG is recommended as a component of the screening algorithm for first-degree relatives of patients with HCM.

Electrocardiography

Twenty-four–hour ambulatory (Holter) electrocardiographic monitoring, repeated every 1 to 2 years, is reasonable in patients with HCM who have no previous evidence of VT to identify patients who may be candidates for ICD therapy.

Annual 12-lead ECGs are reasonable in patients with known HCM who are clinically stable to evaluate for asymptomatic changes in conduction or rhythm (i.e., AF).

Twenty-four–hour ambulatory (Holter) electrocardiographic monitoring might be considered in adults with HCM to assess for asymptomatic paroxysmal AF/atrial flutter.

Diagnosis

Imaging

Echocardiography

A TTE is recommended in the initial evaluation of all patients with suspected HCM.

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.

Periodic (12 to 18 months) TTE screening is recommended for children of patients with HCM, starting by age 12 years 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.

Echocardiography

Repeat TTE is recommended for the evaluation of patients with HCM with a change in clinical status or new cardiovascular event.

A TEE is recommended for the intraoperative guidance of surgical myectomy.

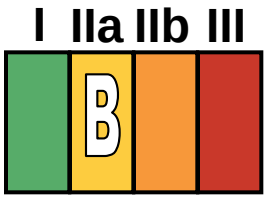
TTE or TEE with intracoronary contrast injection of the candidate's septal perforator(s) is recommended for the intraprocedural guidance of alcohol septal ablation.

Echocardiography

TTE should be used to evaluate the effects of surgical myectomy or alcohol septal ablation for obstructive HCM.

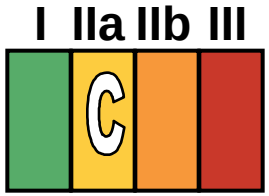
TTE studies performed every 1 to 2 years can be useful in the serial evaluation of symptomatically stable patients with HCM to assess the degree of myocardial hypertrophy, dynamic obstruction, and myocardial function.

Echocardiography



Exercise TTE can be useful in the detection and quantification of dynamic LVOT obstruction in the absence of resting outflow tract obstruction in patients with HCM.

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.



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 are contraindicated.

Echocardiography

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.

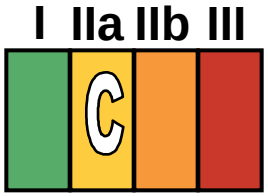
No Benefit

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.

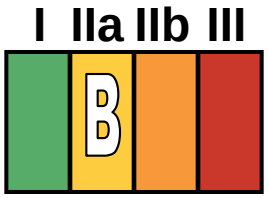
No Benefit

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.

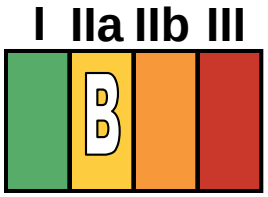
Stress Testing



Treadmill exercise testing is reasonable to determine functional capacity and response to therapy in patients with HCM.



Treadmill testing with monitoring of an ECG and blood pressure is reasonable for SCD risk stratification in patients with HCM.



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.

Cardiac Magnetic Resonance

Cardiac Magnetic Resonance

CMR imaging is indicated in patients with suspected HCM when echocardiography is inconclusive for diagnosis.

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.

CMR imaging is reasonable in patients with HCM to define apical hypertrophy and/or aneurysm if echocardiography is inconclusive.

Cardiac Magnetic Resonance

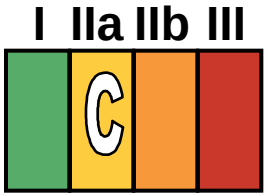
In selected patients with known HCM, when SCD risk stratification is inconclusive after documentation of the conventional risk factors (Section 6.3.1), CMR imaging with assessment of LGE may be considered in resolving clinical decision making.

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.

Detection of Concomitant Coronary Disease

Detection of Concomitant Coronary Disease

Coronary arteriography (invasive or computed tomographic imaging) is indicated in patients with HCM with chest discomfort who have an intermediate to high likelihood of CAD when the identification of concomitant CAD will change management strategies.



Assessment of coronary anatomy with CTA is reasonable for patients with HCM with chest discomfort and a low likelihood of CAD to assess for possible concomitant CAD.

Assessment of ischemia or perfusion abnormalities suggestive of CAD with SPECT or PET MPI (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.

Detection of Concomitant Coronary Disease

No Benefit

Routine SPECT MPI or stress echocardiography is not indicated for detection of “silent” CAD-related ischemia in patients with HCM who are asymptomatic.

No Benefit

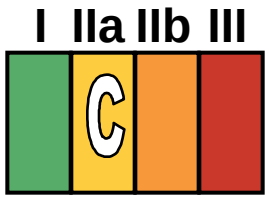
Assessment for the presence of blunted flow reserve (microvascular ischemia) using quantitative myocardial blood flow measurements by PET is not indicated for the assessment of prognosis in patients with HCM.

Management of HCM

Asymptomatic Patients

Asymptomatic Patients

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.



Low-intensity aerobic exercise is reasonable as part of a healthy lifestyle for patients with HCM.

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.

Asymptomatic Patients

Harm

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.

Harm

In patients with HCM with resting or provocable outflow tract obstruction, regardless of symptom status, pure vasodilators and high-dose diuretics are potentially harmful.

Management of HCM

Symptomatic Patients

Pharmacologic Management

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.

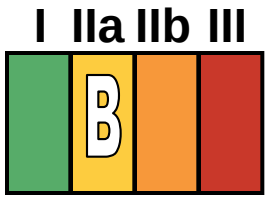
If low doses of beta-blocking drugs are ineffective for controlling symptoms (angina or dyspnea) in patients with HCM, it is useful to titrate the dose to a resting heart rate of <60 to 65 bpm (up to generally accepted and recommended maximum doses of these drugs).

Pharmacologic Management

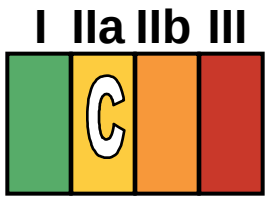
Verapamil therapy (starting in low doses and titrating up to 480 mg/d) is recommended for the treatment of symptoms (angina or dyspnea) in patients with obstructive or nonobstructive HCM who do not respond to beta-blocking drugs or who have side effects or contraindications to beta-blocking drugs. However,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

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.

Pharmacologic Management



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.



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.

Pharmacologic Management

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.

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.

Pharmacologic Management

The usefulness of ACE inhibitors or ARBs 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.

In patients with HCM who do not tolerate verapamil or in whom verapamil is contraindicated, diltiazem may be considered.

Pharmacologic Management

Harm

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.

Harm

Verapamil is potentially harmful in patients with obstructive HCM in the setting of systemic hypotension or severe dyspnea at rest.

Harm

Digitalis is potentially harmful in the treatment of dyspnea in patients with HCM and in the absence of AF.

Pharmacologic Management

Harm

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.

Harm

Dopamine, dobutamine, norepinephrine, and other intravenous positive inotropic drugs are potentially harmful for the treatment of acute hypotension in patients with obstructive HCM.

Invasive Therapies

Invasive Therapies

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.†

*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 (Section 6.2.2.3)

†Eligible patients are defined by all of the following:

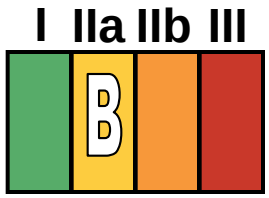
a. Clinical: Severe dyspnea or chest pain (usually NYHA functional classes III or IV) or occasionally other exertional symptoms (such as syncope or near syncope) that interfere with everyday activity or quality of life despite optimal medical therapy.

b. Hemodynamic: Dynamic LVOT gradient at rest or with physiologic provocation 50 mm Hg associated with septal hypertrophy and SAM 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.

Invasive Therapies

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.

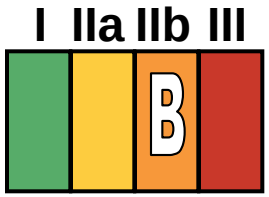


Surgical septal myectomy, when performed in experienced centers, can be beneficial and is the first consideration for the majority of eligible patients with HCM with severe drug-refractory symptoms and LVOT obstruction.

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.

Invasive Therapies

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 NYHA functional classes III or IV).



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.

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.

Invasive Therapies

Harm

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.

Harm

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.

Invasive Therapies

Harm

Mitral valve replacement for relief of LVOT obstruction should not be performed in patients with HCM in whom septal reduction therapy is an option.

Harm

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

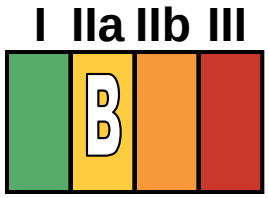
Harm

Alcohol septal ablation should not be done in patients with HCM who are <21 years of age and is discouraged in adults <40 years of age if myectomy is a viable option.

Management of HCM

Pacing

Pacing



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.

Permanent pacing may be considered in medically refractory symptomatic patients with obstructive HCM who are suboptimal candidates for septal reduction therapy.

Pacing

No Benefit

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.

No Benefit

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 who are candidates for septal reduction.

Patients With LV Systolic Dysfunction

Patients With LV Systolic Dysfunction

Patients with nonobstructive HCM who develop systolic dysfunction with an EF ≤50% should be treated according to evidence-based medical therapy for adults with other forms of heart failure with reduced EF, including ACE inhibitors, ARBs, beta blockers, and other indicated drugs.

Other concomitant causes of systolic dysfunction (such as CAD) should be considered as potential contributors to systolic dysfunction in patients with HCM.

Patients With LV Systolic Dysfunction

ICD therapy may be considered in adult patients with advanced (as defined by NYHA functional class III or IV heart failure) nonobstructive HCM, on maximal medical therapy, and EF ≤50%, who do not otherwise have an indication for an ICD.

For patients with HCM who develop systolic dysfunction, it may be reasonable to reassess the use of negative inotropic agents previously indicated, for example, verapamil, diltiazem, or disopyramide, and to consider discontinuing those therapies.

Selection of Patients for Heart Transplantation

Selection of Patients for Heart Transplantation

Patients with advanced heart failure (end stage) and nonobstructive HCM not otherwise amenable to other treatment interventions, with EF ≤50% (or occasionally with preserved EF), should be considered for heart transplantation.

Symptomatic children with HCM with restrictive physiology who are not responsive to or appropriate candidates for other therapeutic interventions should be considered for heart transplantation.

Selection of Patients for Heart Transplantation

Harm

Heart transplantation should not be performed in mildly symptomatic patients of any age with HCM.

Prevention of SCD

SCD Risk Stratification

All patients with HCM should undergo comprehensive SCD risk stratification at initial evaluation to determine the presence of the following:

a. A personal history for ventricular fibrillation, sustained VT, or SCD events, including appropriate ICD therapy for ventricular tachyarrhythmias.†

b. A family history for SCD events, including appropriate ICD therapy for ventricular tachyarrhythmias.†

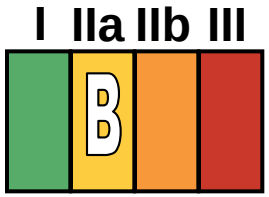
c. Unexplained syncope.

d. Documented NSVT defined as ≥3 beats at ≥120 bpm on ambulatory (Holter) ECG.

e. Maximal LV wall thickness ≥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.

SCD Risk Stratification



It is reasonable to assess blood pressure response during exercise as part of SCD risk stratification in patients with HCM.

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).

SCD Risk Stratification

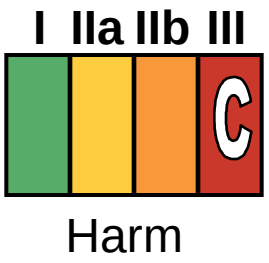
The usefulness of the following potential SCD risk modifiers is unclear but might be considered in selected patients with HCM for whom risk remains borderline after documentation of conventional risk factors:

a. CMR imaging with LGE

b. Double and compound mutations (i.e., >1)

c. Marked LVOT obstruction

SCD Risk Stratification



Invasive electrophysiologic testing as routine SCD risk stratification for patients with HCM should not be performed.

Selection of Patients for ICDs

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 (Figure 4).

ICD placement is recommended for patients with HCM with prior documented cardiac arrest, ventricular fibrillation, or hemodynamically significant VT.

Selection of Patients for ICDs

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

a. Sudden death presumably caused by HCM in ≥first-degree relatives.

b. A maximum LV wall thickness ≥30 mm.

c. One or more recent, unexplained syncopal episodes

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‡.

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‡.

‡SCD risk modifies are discussed in Section 6.3.1.2 of the full text guideline.

Selection of Patients for ICDs

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.

The usefulness of an ICD is uncertain in patients with HCM with isolated bursts of NSVT when in the absence of any other SCD risk factors or modifiers‡.

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.

‡ SCD risk modifiers are discussed in Section 6.3.1.2.

Selection of Patients for ICDs

Harm

ICD placement as a routine strategy in patients with HCM without an indication of increased risk is potentially harmful.

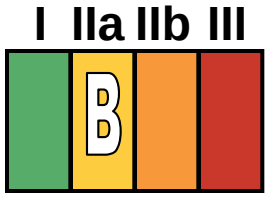
Harm

ICD placement as a strategy to permit patients with HCM to participate in competitive athletics is potentially harmful.

Harm

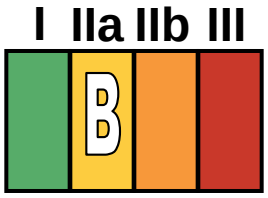
ICD placement in patients who have an identified HCM genotype in the absence of clinical manifestations of HCM is potentially harmful.

Selection of ICD Device Type



In patients with HCM who meet indications for ICD implantation, single-chamber devices are reasonable in younger patients without a need for atrial or ventricular pacing

In patients with HCM who meet indications for ICD implantation, dual-chamber ICDs are reasonable for patients with sinus bradycardia and/or paroxysmal AF.



In patients with HCM who meet indications for ICD implantation, dual-chamber ICDs are reasonable for patients with elevated resting outflow gradients >50 mm Hg and significant heart failure symptoms who may benefit from right ventricular pacing (most commonly, but not limited to, patients **>**65 years of age)

Participation in Competitive or Recreational Sports and Physical Activity

It is reasonable for patients with HCM to participate in low intensity competitive sports (e.g., golf and bowling)

It is reasonable for patients with HCM to participate in a range of recreational sporting activities as outlined in Table 4.

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.

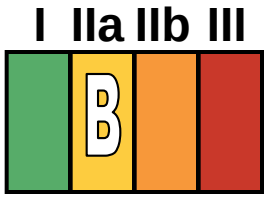
Management of AF

Management of AF

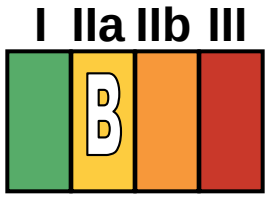
Anticoagulation with vitamin K antagonists (i.e., warfarin, to an INR 2.0 to 3.0) is indicated in patients with paroxysmal, persistent, or chronic AF and HCM. (Anticoagulation with direct thrombin inhibitors [i.e., dabigatran] may represent another option to reduce the risk of thromboembolic events,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

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.

Management of AF



Disopyramide (with ventricular rate–controlling agents) and amiodarone are reasonable antiarrhythmic agents for AF in patients with HCM.



Radiofrequency ablation for AF can be beneficial in patients with HCM who have refractory symptoms or who are unable to take antiarrhythmic drugs

Maze procedure with closure of LA appendage is reasonable in patients with HCM with a history of AF, either during septal myectomy or as an isolated procedure in selected patients.

Management of AF

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.

Other Issues

Management of HCM

Pregnancy/Delivery

Pregnancy/Delivery

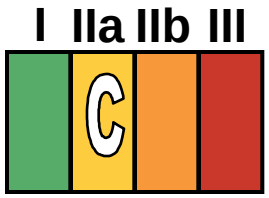
In women with HCM who are asymptomatic or whose symptoms are controlled with beta-blocking drugs, the drugs should be continued during pregnancy, but increased surveillance for fetal bradycardia or other complications is warranted.

For patients (mother or father) with HCM, genetic counseling is indicated before planned conception.

In women with HCM and resting or provocable LVOT obstruction ≥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.

Pregnancy/Delivery

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.



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.

Harm

For women with advanced heart failure symptoms and HCM, pregnancy is associated with excess morbidity/mortality.