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CLINICAL PRACTICE GUIDELINE: EXECUTIVE SUMMARY

2020 AHA/ACC Guideline for the Diagnosis and Treatment of Patients With Hypertrophic Cardiomyopathy: Executive Summary

A Report of the American College of Cardiology/American Heart Association Joint Committee on Clinical Practice Guidelines

Developed in collaboration with and endorsed by the American Association for Thoracic Surgery, American Society of Echocardiography, Heart Failure Society of America, Heart Rhythm Society, Society for Cardiovascular Angiography and Interventions, and Society for Cardiovascular Magnetic Resonance

Endorsed by The Pediatric & Congenital Electrophysiology Society

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ABSTRACT

AIM This executive summary of the hypertrophic cardiomyopathy clinical practice guideline provides recommendations and algorithms for clinicians to diagnose and manage hypertrophic cardiomyopathy in adult and pediatric patients as well as supporting documentation to encourage their use.

METHODS A comprehensive literature search was conducted from January 1, 2010, to April 30, 2020, encompassing studies, reviews, and other evidence conducted on human subjects that were published in English from PubMed, EMBASE, the Cochrane Collaboration, Agency for Healthcare Research and Quality reports, and other relevant databases.

STRUCTURE Many recommendations from the earlier hypertrophic cardiomyopathy guidelines have been updated with new evidence or a better understanding of earlier evidence. This summary operationalizes the recommendations from the full guideline and presents a combination of diagnostic work-up, genetic and family screening, risk stratification approaches, lifestyle modifications, surgical and catheter interventions, and medications that constitute components of guideline directed medical therapy. For both guideline-directed medical therapy and other recommended drug treatment regimens, the reader is advised to follow dosing, contraindications and drug-drug interactions based on product insert materials.

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TOP 10 TAKE-HOME MESSAGES - 2020 AHA/ACC GUIDELINE FOR THE DIAGNOSIS AND TREATMENT OF PATIENTS WITH HYPERTROPHIC CARDIOMYOPATHY

1. Shared decision-making, a dialogue between patients and their care team that includes full disclosure of all testing and treatment options, discussion of the risks and benefits of those options and, importantly, engagement of the patient to express their own goals, is particularly relevant in the management of conditions such as hypertrophic cardiomyopathy (HCM).
2. Although the primary cardiology team can initiate evaluation, treatment, and longitudinal care, referral to multidisciplinary HCM centers with graduated levels of expertise can be important to optimizing care for patients with HCM. Challenging treatment decisions—where reasonable alternatives exist, where the strength of recommendation is weak (e.g., any Class 2b decision) or is particularly nuanced, and for invasive procedures that are specific to patients with HCM—represent crucial opportunities to refer patients to these HCM centers.
3. Counseling patients with HCM regarding the potential for genetic transmission of HCM is one of the cornerstones of care. Screening first-degree family members of patients with HCM, using either genetic testing or an imaging/electrocardiographic surveillance protocol, can begin at any age and can be influenced by specifics of the patient/family history and family preference. As screening recommendations for family members hinge on the pathogenicity of any detected variants, the reported pathogenicity should be reconfirmed every 2 to 3 years.
4. Optimal care for patients with HCM requires cardiac imaging to confirm the diagnosis, characterize the pathophysiology for the individual, and identify risk markers that may inform decisions regarding interventions for left ventricular outflow tract obstruction and sudden cardiac death (SCD) prevention. Echocardiography continues to be the foundational imaging modality for patients with HCM. Cardiovascular magnetic resonance imaging will also be helpful in many patients, especially those in whom there is diagnostic uncertainty, poor echocardiographic imaging windows, or where uncertainty persists regarding decisions around implantable cardioverter-defibrillator (ICD) placement.
5. Assessment of an individual patient's risk for SCD continues to evolve as new markers emerge (e.g., apical aneurysm, decreased left ventricular systolic function, and extensive gadolinium enhancement). In addition to a full accounting of an individual's risk markers, communication with patients regarding not just the presence of risk markers but also the magnitude of their individualized risk is key. This enables the informed patient to fully participate in the decision-making regarding ICD placement, which incorporates their own level of risk tolerance and treatment goals.
6. The risk factors for SCD in children with HCM carry different weights than those observed in adult patients; they vary with age and must account for different body sizes. Coupled with the complexity of placing ICDs in young patients with anticipated growth and a higher risk of device complications, the threshold for ICD implantation in children often differs from adults. These differences are best addressed at primary or comprehensive HCM centers with expertise in children with HCM.
7. Septal reduction therapies (surgical septal myectomy and alcohol septal ablation), when performed by experienced HCM teams at dedicated centers, continue to improve in safety and efficacy such that earlier intervention may be possible in select patients

with drug-refractory or severe outflow tract obstruction causing signs of cardiac decompensation. Given the data on the significantly improved outcomes at comprehensive HCM centers, these decisions represent an optimal referral opportunity.

8. Patients with HCM and persistent or paroxysmal atrial fibrillation have a sufficiently increased risk of stroke such that oral anticoagulation with direct oral anticoagulants (or alternatively warfarin) should be considered the default treatment option independent of the CHA_2DS_2VASc score. As rapid atrial fibrillation is often poorly tolerated in patients with HCM, maintenance of sinus rhythm and rate control are key pursuits in successful treatment.
9. Heart failure symptoms in patients with HCM, in the absence of left ventricular outflow tract obstruction, should be treated similarly to other patients with heart failure symptoms, including consideration of advanced treatment options (e.g., cardiac resynchronization therapy, left ventricular assist device, transplantation). In patients with HCM, an ejection fraction <50% connotes significantly impaired systolic function and identifies individuals with poor prognosis and who are at increased risk for SCD.
10. Increasingly, data affirm that the beneficial effects of exercise on general health can be extended to patients with HCM. Healthy recreational exercise (moderate intensity) has not been associated with increased risk of ventricular arrhythmia events in recent studies. Whether an individual patient with HCM wishes to pursue more rigorous exercise/training is dependent on a comprehensive shared discussion between that patient and their expert HCM care team regarding the potential risks of that level of training/participation but with the understanding that exercise-related risk cannot be individualized for a given patient.

PURPOSE OF THE EXECUTIVE SUMMARY

This executive summary of the American Heart Association (AHA)/American College of Cardiology (ACC) hypertrophic cardiomyopathy (HCM) clinical practice guideline (1) provides a synopsis with algorithms to guide clinicians in the screening, diagnosis, and management of HCM in pediatric and adult patients.

The full guideline (1) recommends a combination of lifestyle modifications, medications, and surgical/catheter interventions that constitute components of guideline-directed medical therapy. For both guideline-directed medical therapy and other recommended drug treatment regimens, the reader is advised to follow dosing, contraindications and drug-drug interactions based on product insert materials.

The full guideline (1) replaces the 2011 guideline (2). Some recommendations from the earlier HCM guidelines have been updated by new evidence or a better understanding of earlier evidence, whereas others that were outdated, irrelevant, or overlapping were deleted or modified. The overall goal was to provide the clinician with concise, evidence-based, contemporary recommendations with supporting data to encourage their use. Sections were divided into the following: 1) diagnosis and follow-up (including genetic and family screening), 2) sudden cardiac death risk assessment and prevention, 3) medical, surgical, and catheter interventions in the management of HCM (obstructive HCM, nonobstructive HCM, atrial fibrillation, ventricular arrhythmias, advanced heart failure), and 4) lifestyle considerations (sports/activity, occupation, pregnancy, comorbidities). There was a strong emphasis on shared decision-making that accounts for patient choices, and the importance of skilled operators and experienced centers that can guide complex decision-making and perform complex procedures with superior outcomes.

The full guideline (1) contains Table 1 and Table 8, which are not cited in this executive summary. The 5 figures included in this executive summary are also included in the full guideline (1).

DOCUMENT REVIEW AND APPROVAL

The guideline was reviewed by 2 official reviewers each nominated by the ACC and AHA, 1 reviewer each from the American Association for Thoracic Surgery, American Society of Echocardiography, Heart Rhythm Society, Society for Cardiovascular Angiography and Interventions, the Society for Cardiovascular Magnetic Resonance, and 26 individual content reviewers. Information about the authors' relevant relationships with industry and other entities is available as Appendix 1 in the full guideline (1). Information about the reviewers' comprehensive relationships with industry and other entities was distributed to the writing committee and is published as Appendix 2 in the full guideline (1).

CLASS OF RECOMMENDATION AND LEVEL OF EVIDENCE

The Class of Recommendation (COR) indicates the strength of recommendation, encompassing the estimated magnitude and certainty of benefit in proportion to risk. The Level of Evidence (LOE) rates the quality of scientific evidence supporting the intervention on the basis of the type, quantity, and consistency of data from clinical trials and other sources (Table 2) (3).

TABLE 2 ACC/AHA Applying Class of Recommendation and Level of Evidence to Clinical Strategies, Interventions, Treatments, or Diagnostic Testing in Patient Care (Updated May 2019)*

Applying Class of Recommendation and Level of Evidence to Clinical Strategies, Interventions, Treatments, or Diagnostic Testing in Patient Care (Updated May 2019)*

CLASS (STRENGTH) OF RECOMMENDATION	
CLASS 1 (STRONG)	Benefit >>> Risk
Suggested phrases for writing recommendations: • Is recommended • Is indicated/useful/effective/beneficial • Should 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	
CLASS 2a (MODERATE)	Benefit >> Risk
Suggested phrases for writing recommendations: • Is reasonable • Can be useful/effective/beneficial • Comparative-Effectiveness Phrases†: – Treatment/strategy A is probably recommended/indicated in preference to treatment B – It is reasonable to choose treatment A over treatment B	
CLASS 2b (WEAK)	Benefit ≥ Risk
Suggested phrases for writing recommendations: • May/might be reasonable • May/might be considered • Usefulness/effectiveness is unknown/unclear/uncertain or not well-established	
CLASS 3: No Benefit (MODERATE) (Generally, LOE A or B use only)	Benefit = Risk
Suggested phrases for writing recommendations: • Is not recommended • Is not indicated/useful/effective/beneficial • Should not be performed/administered/other	
Class 3: Harm (STRONG)	Risk > Benefit
Suggested phrases for writing recommendations: • Potentially harmful • Causes harm • Associated with excess morbidity/mortality • Should not be performed/administered/other	

LEVEL (QUALITY) OF EVIDENCE‡	
LEVEL A	
• High-quality evidence‡ from more than 1 RCT • Meta-analyses of high-quality RCTs • One or more RCTs corroborated by high-quality registry studies	
LEVEL B-R	(Randomized)
• Moderate-quality evidence‡ from 1 or more RCTs • Meta-analyses of moderate-quality RCTs	
LEVEL B-NR	(Nonrandomized)
• Moderate-quality evidence‡ from 1 or more well-designed, well-executed nonrandomized studies, observational studies, or registry studies • Meta-analyses of such studies	
LEVEL C-LD	(Limited Data)
• Randomized or nonrandomized observational or registry studies with limitations of design or execution • Meta-analyses of such studies • Physiological or mechanistic studies in human subjects	
LEVEL C-EO	(Expert Opinion)
• Consensus of expert opinion based on clinical experience	

COR and LOE are determined independently (any COR may be paired with any LOE).

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

* The outcome or result of the intervention should be specified (an improved clinical outcome or increased diagnostic accuracy or incremental prognostic information).

† For comparative-effectiveness recommendations (COR 1 and 2a; LOE A and B only), studies that support the use of comparator verbs should involve direct comparisons of the treatments or strategies being evaluated.

‡ The method of assessing quality is evolving, including the application of standardized, widely-used, and preferably validated evidence grading tools; and for systematic reviews, the incorporation of an Evidence Review Committee.

COR indicates Class of Recommendation; EO, expert opinion; LD, limited data; LOE, Level of Evidence; NR, nonrandomized; R, randomized; and RCT, randomized controlled trial.

1. SHARED DECISION-MAKING

Recommendation for Shared Decision-Making
Referenced studies that support the recommendation are summarized in Online Data Supplement 1.

COR	LOE	RECOMMENDATION
1	B-NR	1. For patients with HCM or at risk for HCM, shared decision-making is recommended in developing a plan of care (including but not limited to decisions regarding genetic evaluation, activity, lifestyle, and therapy choices) that includes a full disclosure of the risks, benefits, and anticipated outcomes of all options, as well the opportunity for the patient to express their goals and concerns (4–9).

2. MULTIDISCIPLINARY HCM CENTERS

Tables in this section are located in the full guideline (1).

Recommendations for Multidisciplinary HCM Centers

COR	LOE	RECOMMENDATIONS
1	C-LD	1. In patients with HCM in whom septal reduction therapy (SRT) is indicated, the procedure should be performed at experienced centers (comprehensive or primary HCM centers) with demonstrated excellence in clinical outcomes for these procedures (10–12) (Table 3 and Table 4).
2a	C-LD	2. In patients with HCM, consultation with or referral to a comprehensive or primary HCM center is reasonable to aid in complex disease-related management decisions (13–22) (Table 3).

3. DIAGNOSIS, INITIAL EVALUATION, AND FOLLOW-UP

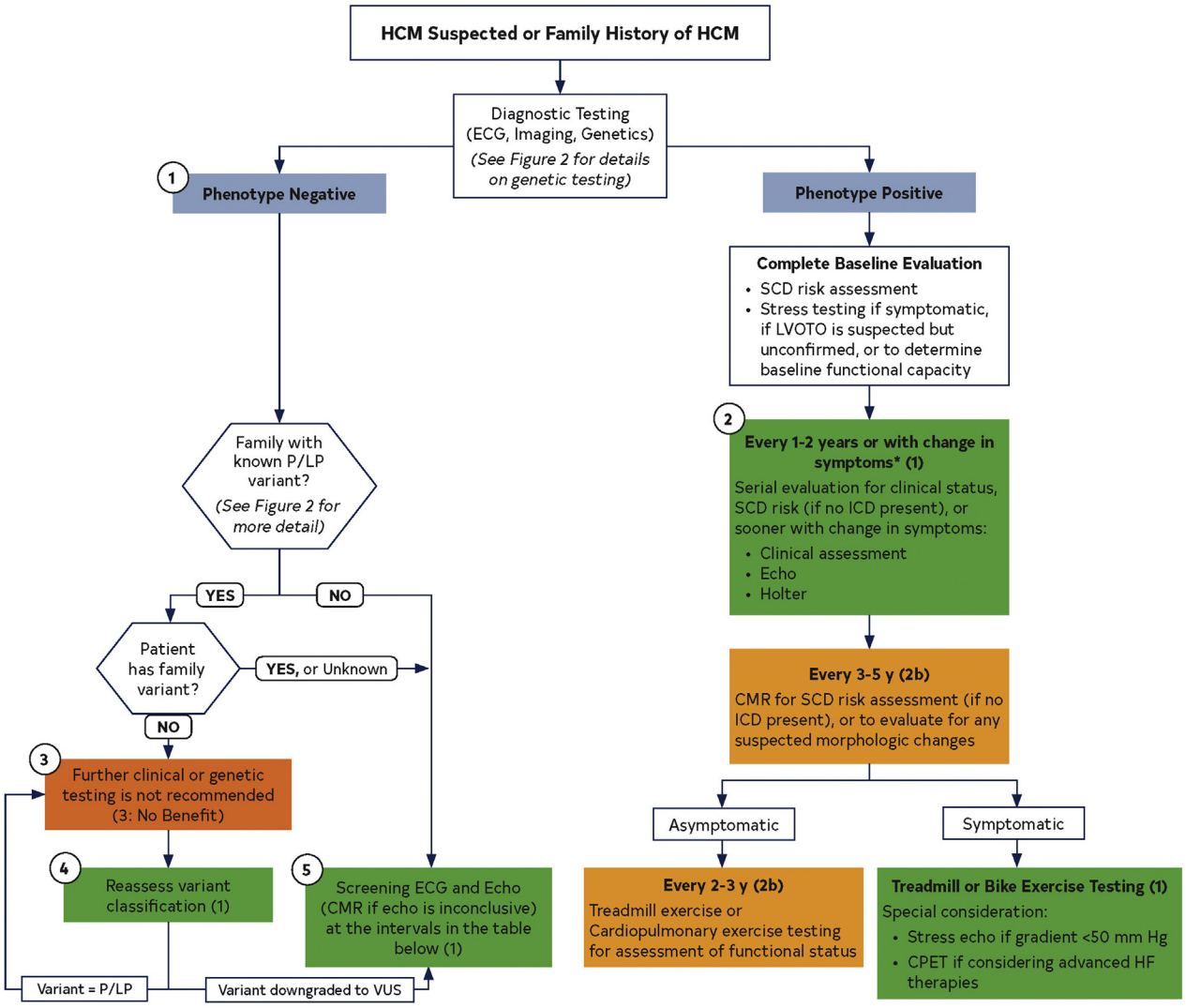
3.1. Clinical Diagnosis

Tables in this section are located in the full guideline (1). Figure 1 presents a recommended evaluation and testing for HCM.

Recommendation for Diagnosis, Initial Evaluation, and Follow-up
Referenced studies that support the recommendation are summarized in Online Data Supplement 2.

COR	LOE	RECOMMENDATION
1	B-NR	1. In patients with suspected HCM, comprehensive physical examination and complete medical and 3-generation family history is recommended as part of the initial diagnostic assessment (23–28) (Table 5 and Table 6).

FIGURE 1 Recommended Evaluation and Testing for HCM



Screening Asymptomatic First-Degree Relatives of Patients With HCM		
Age of First-Degree Relative	**Initiation of Screening**	**Surveillance Interval**
Children and adolescents from genotype-positive family and/or family with early onset HCM	At the time of diagnosis in another family member	Every 1-2 y
All other children and adolescents	At any time after the diagnosis in the family, but no later than puberty	Every 2-3 y
Adults	At the time of diagnosis in another family member	Every 3-5 y

Colors correspond to the Class of Recommendation in Table 2. *The interval may be extended, particularly in adult patients who remain stable after multiple evaluations. CMR indicates cardiovascular magnetic resonance; CPET, cardiopulmonary exercise test; ECG, electrocardiography/electrocardiogram; HCM, hypertrophic cardiomyopathy; HF, heart failure; ICD, implantable cardioverter-defibrillator; LVOTO, left ventricular outflow tract obstruction; P/LP, pathogenic or likely pathogenic variant; SCD, sudden cardiac death; and VUS, variant of unknown significance.

3.2. Echocardiography

Recommendations for Echocardiography
Referenced studies that support the recommendations are summarized in Online Data Supplement 3.

COR	LOE	RECOMMENDATIONS
1	B-NR	1. In patients with suspected HCM, a transthoracic echocardiogram (TTE) is recommended in the initial evaluation (29–34).
1	B-NR children C-LD adults	2. In patients with HCM with no change in clinical status or events, repeat TTE is recommended every 1 to 2 years to assess the degree of myocardial hypertrophy, dynamic left ventricular outflow tract obstruction (LVOTO), mitral regurgitation, and myocardial function (35–42) (Figure 1).
1	B-NR	3. For patients with HCM who experience a change in clinical status or a new clinical event, repeat TTE is recommended (35,38,43–46).
1	B-NR	4. For patients with HCM and resting left ventricular outflow tract gradient <50 mm Hg, a TTE with provocative maneuvers is recommended (47–50).
1	B-NR	5. For symptomatic patients with HCM who do not have a resting or provocable outflow tract gradient ≥50 mm Hg on TTE, exercise TTE is recommended for the detection and quantification of dynamic LVOTO (49–54).
1	B-NR	6. For patients with HCM undergoing surgical septal myectomy, intraoperative transesophageal echocardiogram (TEE) is recommended to assess mitral valve anatomy and function and adequacy of septal myectomy (55–58).
1	B-NR	7. For patients with HCM undergoing alcohol septal ablation, TTE or intraoperative TEE with intracoronary ultrasound-enhancing contrast injection of the candidate's septal perforator(s) is recommended (31,59–63).
1	B-NR	8. For patients with HCM who have undergone SRT, TTE within 3 to 6 months after the procedure is recommended to evaluate the procedural results (17,64–66).
1	B-NR	9. Screening: In first-degree relatives of patients with HCM, a TTE is recommended as part of initial family screening and periodic follow-up (31–33,35,36,61) (Figure 1, Table 6).
1	B-NR	10. Screening: In individuals who are genotype-positive or phenotype-negative, serial echocardiography is recommended at periodic intervals depending on age (1 to 2 years in children and adolescents, 3 to 5 years in adults) and change in clinical status (Table 6; Figure 1) (28,67,68–71) .
2a	C-LD	11. For patients with HCM, TEE can be useful if TTE is inconclusive in clinical decision-making regarding 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 the assessment of the feasibility of alcohol septal ablation (55–58).
2a	B-NR	12. For patients with HCM in whom the diagnoses of apical HCM, apical aneurysm, or atypical patterns of hypertrophy is inconclusive on TTE, the use of an intravenous ultrasound-enhancing agent is reasonable particularly if other imaging modalities such as cardiovascular magnetic resonance (CMR) are not readily available or contraindicated (72,73).
2a	C-LD	13. For asymptomatic patients with HCM who do not have a resting or provocable outflow tract gradient ≥50 mm Hg on standard TTE, exercise TTE is reasonable for the detection and quantification of dynamic LVOTO (43,48,49,51–54).

3.3. Cardiovascular Magnetic Resonance Imaging

Recommendations for CMR Imaging
Referenced studies that support the recommendations are summarized in Online Data Supplement 4.

COR	LOE	RECOMMENDATIONS
1	B-NR	1. For patients suspected to have HCM in whom echocardiography is inconclusive, CMR imaging is indicated for diagnostic clarification (74–80).
1	B-NR	2. For patients with left ventricular hypertrophy in whom there is a suspicion of alternative diagnoses including infiltrative or storage disease as well as athlete's heart, CMR imaging is useful (74–80) (Figure 1).
1	B-NR	3. For patients with HCM who are not otherwise identified as high risk for sudden cardiac death (SCD), or in whom a decision to proceed with implantable cardioverter-defibrillator (ICD) remains uncertain after clinical assessment that includes personal/family history, echocardiography, and ambulatory electrocardiographic monitoring, CMR imaging is beneficial to assess for maximum left ventricular (LV) wall thickness, ejection fraction (EF), LV apical aneurysm, and extent of myocardial fibrosis with late gadolinium enhancement (38,74–87).
1	B-NR	4. For patients with obstructive HCM in whom the anatomic mechanism of obstruction is inconclusive on echocardiography, CMR imaging is indicated to inform the selection and planning of SRT (88–92).
2b	C-EO	5. For patients with HCM, repeat contrast-enhanced CMR imaging on a periodic basis (every 3 to 5 years) for the purpose of SCD risk stratification may be considered to evaluate changes in late gadolinium enhancement and other morphologic changes, including EF, development of apical aneurysm, or LV wall thickness (Figure 1, Table 7).

3.4. Cardiac Computed Tomography

Recommendation for Cardiac Computed Tomography (CT)

COR	LOE	RECOMMENDATION
2b	C-LD	1. In adult patients with suspected HCM, cardiac CT may be considered for diagnosis if the echocardiogram is not diagnostic and CMR imaging is unavailable (34,93,94).

3.5. Heart Rhythm Assessment

Recommendations for Heart Rhythm Assessment
Referenced studies that support the recommendations are summarized in Online Data Supplement 5.

COR	LOE	RECOMMENDATIONS
1	B-NR	1. In patients with HCM, a 12-lead ECG is recommended in the initial evaluation and as part of periodic follow-up (every 1 to 2 years) (95–97) (Figure 1, Table 6).
1	B-NR	2. In patients with HCM, 24- to 48-hour ambulatory electrocardiographic monitoring is recommended in the initial evaluation and as part of periodic follow-up (every 1 to 2 years) to identify patients who are at risk for SCD and guide management of arrhythmias (Figure 1) (98–100).
1	B-NR	3. In patients with HCM who develop palpitations or lightheadedness, extended (>24 hours) electrocardiographic monitoring or event recording is recommended, which should not be considered diagnostic unless patients have had symptoms while being monitored (101).
1	B-NR	4. In first-degree relatives of patients with HCM, a 12-lead ECG is recommended as a component of the screening algorithm (95–97) (Figure 1, Table 6).

(Continued)

COR	LOE	RECOMMENDATIONS
2a	B-NR	5. In patients with HCM who have additional risk factors for atrial fibrillation (AF), such as left atrial dilatation, advanced age, and New York Heart Association (NYHA) class III to class IV heart failure (HF), and who are eligible for anticoagulation, extended ambulatory monitoring is reasonable to screen for AF as part of initial evaluation and periodic follow-up (every 1 to 2 years) (102–106) (Figure 1).
2b	B-NR	6. In adult patients with HCM without risk factors for AF and who are eligible for anticoagulation, extended ambulatory monitoring may be considered to assess for asymptomatic paroxysmal AF as part of initial evaluation and periodic follow-up (every 1 to 2 years) (102–106).

3.6. Angiography and Invasive Hemodynamic Assessment

Recommendations for Angiography and Invasive Hemodynamic Assessment
Referenced studies that support the recommendations are summarized in Online Data Supplement 6.

COR	LOE	RECOMMENDATIONS
1	B-NR	1. For patients with HCM who are candidates for SRT and for whom there is uncertainty regarding the presence or severity of LVOTO on noninvasive imaging studies, invasive hemodynamic assessment with cardiac catheterization is recommended (45,107–109).
1	B-NR	2. In patients with HCM with symptoms or evidence of myocardial ischemia, coronary angiography (CT or invasive) is recommended (110).
1	B-NR	3. In patients with HCM who are at risk of coronary atherosclerosis, coronary angiography (CT or invasive) is recommended before surgical myectomy (111).

3.7. Exercise Stress Testing

Recommendations for Exercise Stress Testing
Referenced studies that support the recommendations are summarized in Online Data Supplement 7.

COR	LOE	RECOMMENDATIONS
1	B-NR	1. For symptomatic patients with HCM who do not have resting or provocable outflow tract gradient ≥50 mm Hg on TTE, exercise TTE is recommended for the detection and quantification of dynamic LVOTO (50,112).
1	B-NR	2. In patients with nonobstructive HCM and advanced HF (NYHA functional class III to class IV despite guideline-directed management and therapy), cardiopulmonary exercise stress testing should be performed to quantify the degree of functional limitation and aid in selection of patients for heart transplantation or mechanical circulatory support (113,114).
2a	B-NR	3. In patients with HCM, exercise stress testing is reasonable to determine functional capacity and to provide prognostic information as part of initial evaluation (113,114).
2a	C-LD	4. For asymptomatic patients with HCM who do not have a resting or provocable outflow tract gradient ≥50 mm Hg on standard TTE, exercise TTE is reasonable for the detection and quantification of dynamic LVOTO (49,51–54,115).
2b	C-EO	5. In patients with obstructive HCM, who are being considered for SRT, and in whom functional capacity or symptom status is uncertain, exercise stress testing may be reasonable (Figure 1).
2b	C-EO	6. In patients with HCM in whom functional capacity or symptom status is uncertain, exercise stress testing may be considered every 2 to 3 years (Figure 1).

3.8. Genetics and Family Screening

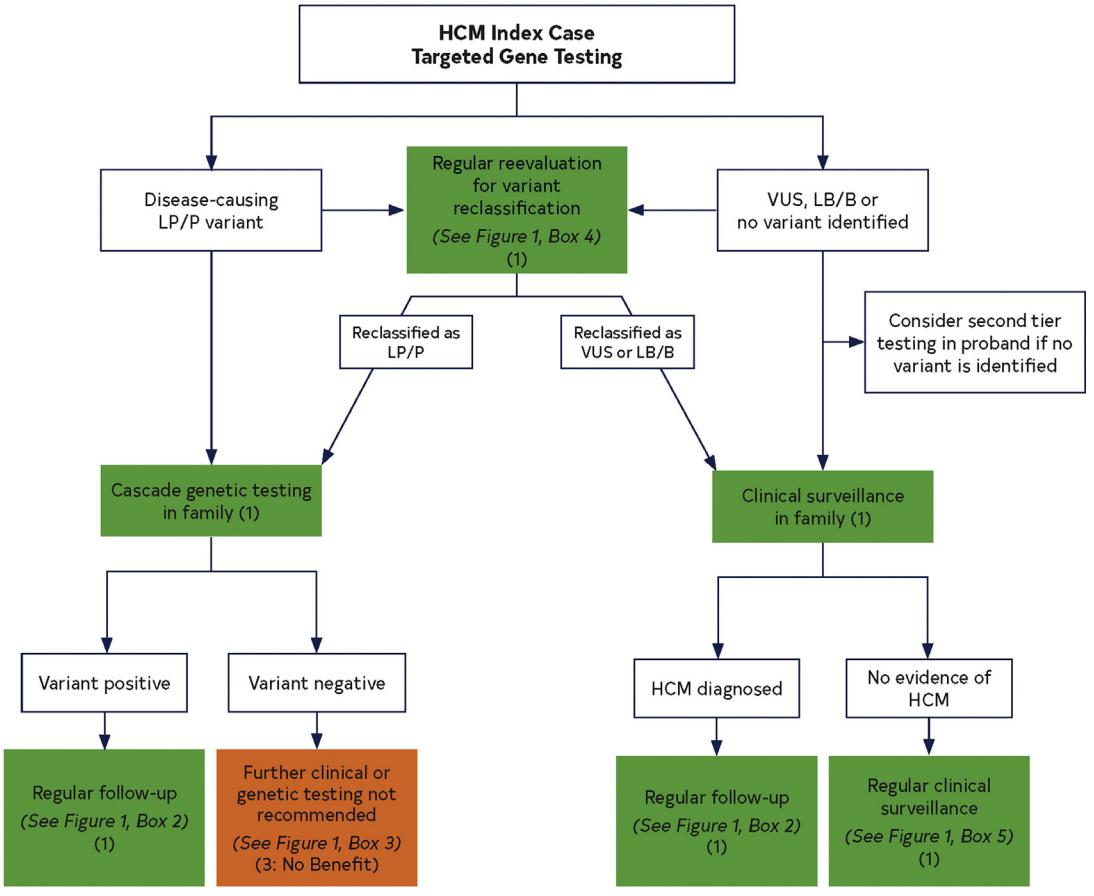
Figure 2 presents a genetic testing process for HCM.

Recommendations for Genetics and Family Screening
Referenced studies that support the recommendations are summarized in Online Data Supplements 8 and 9.

COR	LOE	RECOMMENDATIONS
1	B-NR	1. In patients with HCM, evaluation of familial inheritance, including a 3-generation family history, is recommended as part of the initial assessment (23,25–28,117,118).
1	B-NR	2. In patients with HCM, genetic testing is beneficial to elucidate the genetic basis to facilitate the identification of family members at risk for developing HCM (cascade testing) (119–122).
1	B-NR	3. In patients with an atypical clinical presentation of HCM or when another genetic condition is suspected to be the cause, a work-up including genetic testing for HCM and other genetic causes of unexplained cardiac hypertrophy ("HCM phenocopies") is recommended (123–125).
1	B-NR	4. In patients with HCM who choose to undergo genetic testing, pre- and posttest genetic counseling by an expert in the genetics of cardiovascular disease is recommended so that risks, benefits, results, and their clinical significance can be reviewed and discussed with the patient in a shared decision-making process (23–25,117).
1	B-NR	5. When performing genetic testing in an HCM proband, the initial tier of genes tested should include genes with strong evidence to be disease-causing in HCM* (119,122,126,127,128,129).
1	B-NR	6. In first-degree relatives of patients with HCM, both clinical screening (ECG and 2D echocardiogram) and cascade genetic testing (when a pathogenic/likely pathogenic variant has been identified in the proband) should be offered (23,67,123,130,131,132).
1	B-NR	7. In families where a sudden unexplained death has occurred with a postmortem diagnosis of HCM, postmortem genetic testing is beneficial to facilitate cascade genetic testing and clinical screening in first-degree relatives (133,134).
1	B-NR	8. In patients with HCM who have undergone genetic testing, serial reevaluation of the variant(s) identified is recommended to assess for variant reclassification, which may impact diagnosis and cascade genetic testing in family members (135–137). (Figure 1 and Figure 2)
1	B-NR	9. In affected families with HCM, preconception and prenatal reproductive and genetic counseling should be offered (23–25,116,117).
2b	B-NR	10. In patients with HCM, the usefulness of genetic testing in the assessment of risk of SCD is uncertain (121,137–139).
2b	B-NR	11. In patients with HCM who harbor a variant of uncertain significance, the usefulness of clinical genetic testing of phenotype-negative relatives for the purpose of variant reclassification is uncertain (28,70,118,119).
3: No benefit	B-NR	12. For patients with HCM who have undergone genetic testing and were found to have no pathogenic variants (i.e., harbor only benign/likely benign variants), cascade genetic testing of the family is not useful (118–121).
3: No benefit	B-NR	13. Ongoing clinical screening is not indicated in genotype-negative relatives in families with genotype-positive HCM, unless the disease-causing variant is downgraded to variant of uncertain significance, likely benign, or benign variant during follow-up (15,135,140,141,142,143,144).

*Strong evidence HCM genes include, at the time of this publication, *MYH7, MYBPC3, TNNI3, TNNT2, TPM1, MYL2, MYL3*, and *ACTC1*.

FIGURE 2 Genetic Testing Process in HCM



Colors correspond to the Class of Recommendation in Table 2. HCM indicates hypertrophic cardiomyopathy; LB/B, likely benign/benign; LP/P, likely pathogenic or pathogenic; and VUS, variant of unknown significance.

3.9. Genotype-Positive, Phenotype-Negative

Recommendations for Individuals Who Are Genotype-Positive, Phenotype-Negative
Referenced studies that support the recommendations are summarized in Online Data Supplement 10.

COR	LOE	RECOMMENDATIONS
1	B-NR	1. In individuals who are genotype-positive, phenotype-negative for HCM, serial clinical assessment, electrocardiography, and cardiac imaging are recommended at periodic intervals depending on age (every 1 to 2 years in children and adolescents, and every 3 to 5 years in adults) and change in clinical status (28,67,69-71). (Figure 1 and Figure 2, Table 6)
2a	C-LD	2. In individuals who are genotype-positive, phenotype-negative for HCM, participation in competitive athletics of any intensity is reasonable (132).
3: No benefit	B-NR	3. In individuals who are genotype-positive, phenotype-negative for HCM, ICD is not recommended for primary prevention (28,69-71,132,145).

4. SCD RISK ASSESSMENT AND PREVENTION

4.1. SCD Risk Assessment

Recommendations for SCD Risk Assessment
Referenced studies that support the recommendations are summarized in Online Data Supplement 11.

COR	LOE	RECOMMENDATIONS
1	B-NR	1. In patients with HCM, a comprehensive, systematic noninvasive SCD risk assessment at initial evaluation and every 1 to 2 years thereafter is recommended and should include evaluation of these risk factors (38,78,79,81,83-87,99,146-160) (Figure 1 and Figure 3, Table 7): a. Personal history of cardiac arrest or sustained ventricular arrhythmias b. Personal history of syncope suspected by clinical history to be arrhythmic c. Family history in close relative of premature HCM-related sudden death, cardiac arrest, or sustained ventricular arrhythmias d. Maximal LV wall thickness, EF, LV apical aneurysm e. Nonsustained ventricular tachycardia episodes on continuous ambulatory electrocardiographic monitoring
1	B-NR	2. For patients with HCM who are not otherwise identified as high risk for SCD, or in whom a decision to proceed with ICD placement remains uncertain after clinical assessment that includes personal/family history, echocardiography, and ambulatory electrocardiographic monitoring, CMR imaging is beneficial to assess for maximum LV wall thickness, EF, LV apical aneurysm, and extent of myocardial fibrosis with late gadolinium enhancement (78,79,81,83-86,146,155) (Table 7).
2a	B-NR	3. For patients who are ≥16 years of age with HCM, it is reasonable to obtain echocardiography-derived left atrial diameter and maximal left ventricular outflow tract gradient to aid in calculating an estimated 5-year sudden death risk that may be useful during shared decision-making for ICD placement (147,157) (Table 7).

4.2. Patient Selection for ICD Placement

Recommendations for ICD Placement in High-Risk Patients With HCM
Referenced studies that support the recommendations are summarized in Online Data Supplement 12.

COR	LOE	RECOMMENDATIONS
1	C-EO	1. In patients with HCM, application of individual clinical judgment is recommended when assessing the prognostic strength of conventional risk marker(s) within the clinical profile of the individual patient, as well as a thorough and balanced discussion of the evidence, benefits, and estimated risks to engage the fully informed patient's active participation in ICD decision-making (13,146,161-163).
1	B-NR	2. For patients with HCM, and previous documented cardiac arrest or sustained ventricular tachycardia, ICD placement is recommended (146,148,161-163) (Figure 3, Table 7).
2a	B-NR	3. For adult patients with HCM with ≥1 major risk factors for SCD, it is reasonable to offer an ICD. These major risk factors include (38,81,87,99,146,149-157,159-161) (Figure 3, Table 7): a. Sudden death judged definitively or likely attributable to HCM in ≥1 first-degree or close relatives who are ≤50 years of age; b. Massive left ventricular hypertrophy ≥30 mm in any left ventricular segment; c. ≥1 Recent episodes of syncope suspected by clinical history to be arrhythmic (i.e., unlikely to be of neurocardiogenic [vasovagal] etiology, or related to LVOTO); d. LV apical aneurysm, independent of size; e. LV systolic dysfunction (EF <50%).
2a	B-NR	4. For children with HCM who have ≥1 conventional risk factors, including unexplained syncope, massive left ventricular hypertrophy, nonsustained ventricular tachycardia, or family history of early HCM-related SCD, ICD placement is reasonable after considering the relatively high complication rates of long-term ICD placement in younger patients (164-171) (Figure 3, Table 7).

(Continued)

2a B-NR	5. For patients ≥16 years of age with HCM and with ≥1 major SCD risk factors, discussion of the estimated 5-year sudden death risk and mortality rates can be useful during the shared decision-making process for ICD placement (157,161) (Figure 3, Table 7).
2b B-NR	6. In select adult patients with HCM and without major SCD risk factors after clinical assessment, or in whom the decision to proceed with ICD placement remains otherwise uncertain, ICD may be considered in patients with extensive late gadolinium enhancement by contrast-enhanced CMR imaging or non-sustained ventricular tachycardia present on ambulatory monitoring (84–86,99,146,157,161,170) (Figure 3, Table 7).
2b C-LD	7. In select pediatric patients with HCM in whom risk stratification is otherwise less certain, it may be useful to consider additional factors such as extensive late gadolinium enhancement on contrast-enhanced CMR imaging and systolic dysfunction in risk stratification (172,173) (Figure 3, Table 7).
3: Harm B-NR	8. In patients with HCM without risk factors, ICD placement should not be performed (85,146).
3: Harm B-NR	9. In patients with HCM, ICD placement for the sole purpose of participation in competitive athletics should not be performed (174).

TABLE 7 Established Clinical Risk Factors for HCM Sudden Death Risk Stratification

Family history of sudden death from HCM	Sudden death judged definitively or likely attributable to HCM in ≥1 first-degree or close relatives who are ≤50 years of age. Close relatives would generally be second-degree relatives; however, multiple SCDs in tertiary relatives should also be considered relevant.
Massive LVH	Wall thickness ≥30 mm in any segment within the chamber by echocardiography or CMR imaging; consideration for this morphologic marker is also given to borderline values of ≥28 mm in individual patients at the discretion of the treating cardiologist. For pediatric patients with HCM, an absolute or z-score threshold for wall thickness has not been established; however, a maximal wall that corresponds to a z-score ≥20 (and >10 in conjunction with other risk factors) appears reasonable.
Unexplained syncope	≥1 Unexplained episodes involving acute transient loss of consciousness, judged by history unlikely to be of neurocardiogenic (vasovagal) etiology, nor attributable to LVOTO, and especially when occurring within 6 months of evaluation (events beyond 5 years in the past do not appear to have relevance).
HCM with LV systolic dysfunction	Systolic dysfunction with EF <50% by echocardiography or CMR imaging.
LV apical aneurysm	Apical aneurysm defined as a discrete thin-walled dyskinetic or akinetic segment of the most distal portion of the LV chamber; independent of size.
Extensive LGE on CMR imaging	Diffuse and extensive LGE, representing fibrosis, either quantified or estimated by visual inspection, comprising ≥15% of LV mass (extent of LGE conferring risk has not been established in children).
NSVT on ambulatory monitor	It would seem most appropriate to place greater weight on NSVT as a risk marker when runs are frequent (≥3), longer (≥10 beats), and faster (≥200 bpm) occurring usually over 24 to 48 hours of monitoring. For pediatric patients, a VT rate that exceeds the baseline sinus rate by >20% is considered significant.

CMR indicates cardiovascular magnetic resonance; ICD, implantable cardioverter-defibrillator; LGE, late gadolinium enhancement; LV, left ventricular; LVH, left ventricular hypertrophy; LVOTO, left ventricular outflow tract obstruction; NSVT, nonsustained ventricular tachycardia; and SCD, sudden cardiac death.

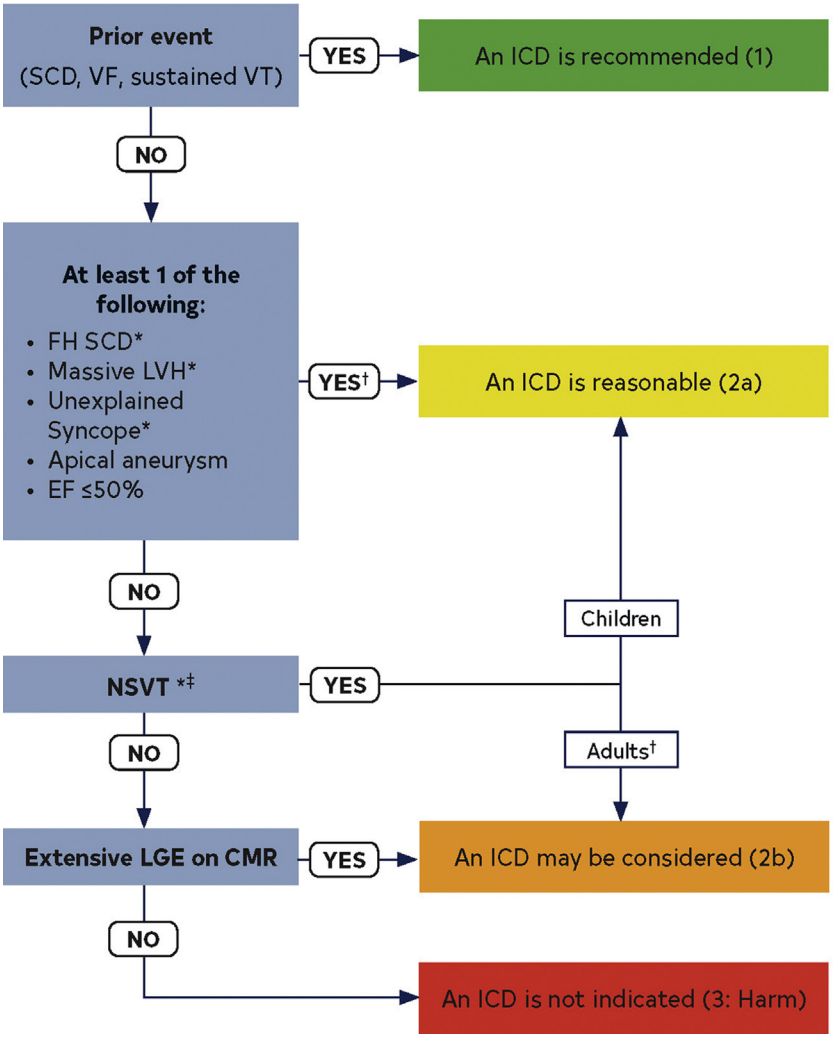
4.3. Device Selection Considerations

Figure 3 presents ICD patient selection.

Recommendations for Selection of ICD Device Type
Referenced studies that support the recommendations are summarized in Online Data Supplement 13.

COR	LOE	RECOMMENDATIONS
1	B-NR	1. In patients with HCM who are receiving an ICD, either a single chamber transvenous ICD or a subcutaneous ICD is recommended after a shared decision-making discussion that takes into consideration patient preferences, lifestyle, and expected potential need for pacing for bradycardia or ventricular tachycardia termination (175–190).
1	B-NR	2. In patients with HCM who are receiving an ICD, single-coil ICD leads are recommended in preference to dual coil leads (187).
2a	B-NR	3. In patients with HCM who are receiving an ICD, dual-chamber ICDs are reasonable for patients with a need for atrial or atrioventricular sequential pacing for bradycardia/conduction abnormalities, or as an attempt to relieve symptoms of obstructive HCM (most commonly in patients >65 years of age) (191–198).
2a	C-LD	4. In selected adult patients with nonobstructive HCM receiving an ICD who have NYHA class II to ambulatory class IV HF, left bundle branch block (LBBB), and LV ejection fraction (LVEF) <50%, cardiac resynchronization therapy for symptom reduction is reasonable (199–203).
2b	C-LD	5. In patients with HCM in whom a decision has been made for ICD implantation and who have paroxysmal atrial tachycardias or AF, dual-chamber ICDs may be reasonable, but this decision must be balanced against higher complication rates of dual chamber devices (191–198).

FIGURE 3 **ICD Patient Selection**



Colors correspond to the Class of Recommendation in Table 2. *ICD decisions in pediatric patients with HCM are based on ≥1 of these major risk factors: family history of HCM SCD, NSVT on ambulatory monitor, massive LVH, and unexplained syncope. †In patients >16 years of age, 5-year risk estimates can be considered to fully inform patients during shared decision-making discussions. ‡It would seem most appropriate to place greater weight on frequent, longer, and faster runs of NSVT. CMR indicates cardiovascular magnetic resonance; EF, ejection fraction; FH, family history; HCM, hypertrophic cardiomyopathy; ICD, implantable cardioverter-defibrillator; LGE, late gadolinium enhancement; LVH, left ventricular hypertrophy; NSVT, nonsustained ventricular tachycardia; SCD, sudden cardiac death; VF, ventricular fibrillation; and VT, ventricular tachycardia.

5. MANAGEMENT OF HCM

5.1. Management of Symptomatic Patients With Obstructive HCM

5.1.1. Pharmacologic Management of Symptomatic Patients With Obstructive HCM

Tables in this section are located in the full guideline (1).

Recommendations for Pharmacologic Management of Patients With Obstructive HCM
Referenced studies that support the recommendations are summarized in Online Data Supplement 14.

COR	LOE	RECOMMENDATIONS
1	B-NR	1. In patients with obstructive HCM and symptoms* attributable to LVOTO, nonvasodilating beta blockers, titrated to effectiveness or maximally tolerated doses, are recommended (204–206).
1	Verapamil B-NR Diltiazem C-LD	2. In patients with obstructive HCM and symptoms* attributable to LVOTO, for whom beta blockers are ineffective or not tolerated, substitution with non-dihydropyridine calcium channel blockers (e.g., verapamil, diltiazem) is recommended (207–209).
1	B-NR	3. For patients with obstructive HCM who have persistent severe symptoms* attributable to LVOTO despite beta blockers or non-dihydropyridine calcium channel blockers, either adding disopyramide in combination with 1 of the other drugs, or SRT performed at experienced centers,† is recommended (17,50,210–213).
1	C-LD	4. For patients with obstructive HCM and acute hypotension who do not respond to fluid administration, intravenous phenylephrine (or other vasoconstrictors without inotropic activity), alone or in combination with beta-blocking drugs, is recommended (214).
2b	C-EO	5. For patients with obstructive HCM and persistent dyspnea with clinical evidence of volume overload and high left-sided filling pressures despite other HCM guideline-directed management and therapy, cautious use of low-dose oral diuretics may be considered.
2b	C-EO	6. For patients with obstructive HCM, discontinuation of vasodilators (e.g., angiotensin-converting enzyme inhibitors, angiotensin receptor blockers, dihydropyridine calcium channel blockers) or digoxin may be reasonable because these agents can worsen symptoms caused by dynamic outflow tract obstruction.
3: Harm	C-LD	7. For patients with obstructive HCM and severe dyspnea at rest, hypotension, very high resting gradients (e.g., >100 mm Hg), as well as all children <6 weeks of age, verapamil is potentially harmful (207,215).

*Symptoms include effort-related dyspnea or chest pain; and occasionally other exertional symptoms (e.g., syncope, near syncope) that are attributed to LVOTO and interfere with everyday activity or quality of life. †Comprehensive or primary HCM centers with demonstrated excellence in clinical outcomes for these procedures (Table 3 and Table 4).

5.1.2. Invasive Treatment of Symptomatic Patients With Obstructive HCM

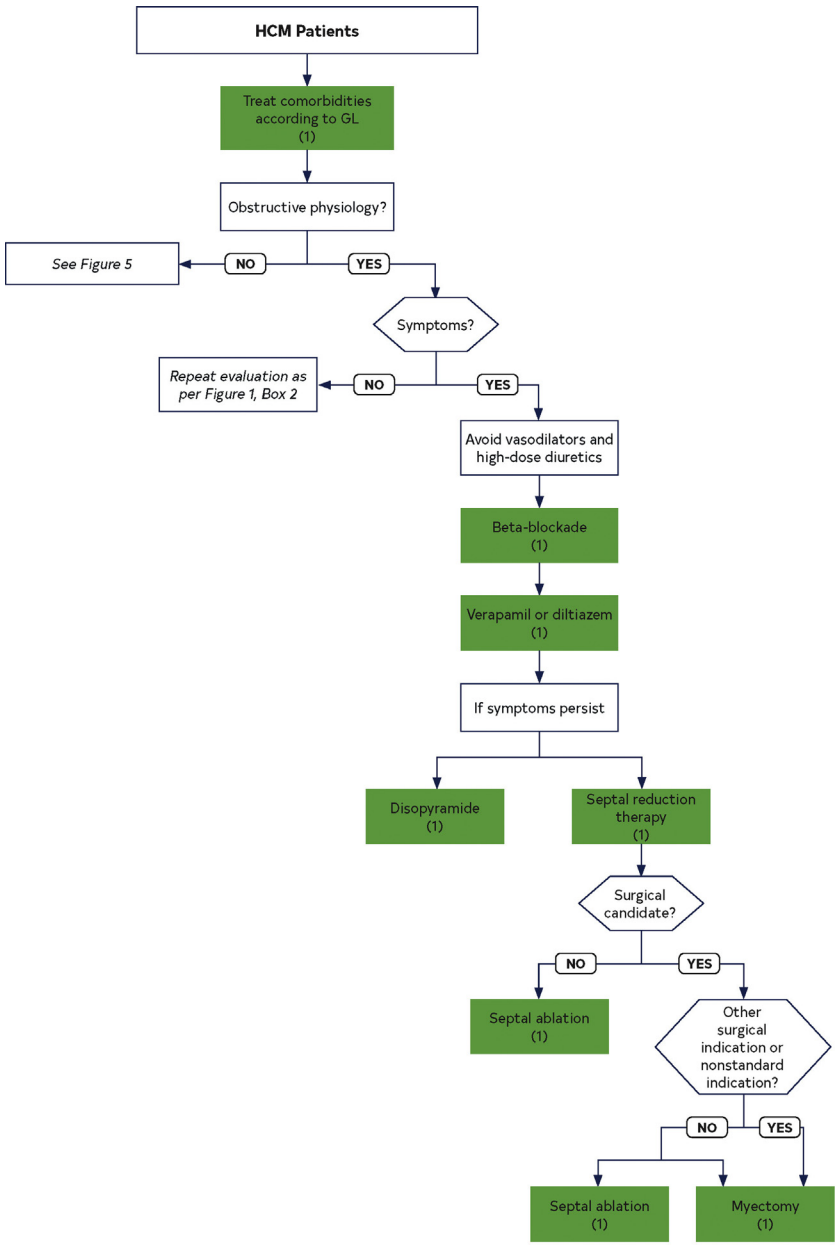
Tables in this section are located in the full guideline (1). **Figure 4** presents a management diagram of symptoms in patients with HCM.

Recommendations for Invasive Treatment of Symptomatic Patients With Obstructive HCM
Referenced studies that support the recommendations are summarized in Online Data Supplement 15.

COR	LOE	RECOMMENDATIONS
1	B-NR	1. In patients with obstructive HCM who remain severely symptomatic despite guideline-directed management and therapy, SRT in eligible patients,* performed at experienced centers,† is recommended for relieving LVOTO (17,50,213) (Table 3 and Table 4).
1	B-NR	2. In symptomatic patients with obstructive HCM who have associated cardiac disease requiring surgical treatment (e.g., associated anomalous papillary muscle, markedly elongated anterior mitral leaflet, intrinsic mitral valve disease, multivessel coronary artery disease, valvular aortic stenosis), surgical myectomy, performed at experienced centers,† is recommended (92,216–218) (Table 3 and Table 4).
1	C-LD	3. In adult patients with obstructive HCM who remain severely symptomatic, despite guideline-directed management and therapy and in whom surgery is contraindicated or the risk is considered unacceptable because of serious comorbidities or advanced age, alcohol septal ablation in eligible patients,* performed at experienced centers,† is recommended (219–221) (Table 3 and Table 4).
2b	B-NR	4. In patients with obstructive HCM, earlier (NYHA class II) surgical myectomy performed at comprehensive HCM centers (Table 3 and Table 4), may be reasonable in the presence of additional clinical factors, including (17,222–233): a. Severe and progressive pulmonary hypertension thought to be attributable to LVOTO or associated mitral regurgitation. b. Left atrial enlargement with ≥1 episodes of symptomatic AF. c. Poor functional capacity attributable to LVOTO as documented on treadmill exercise testing. d. Children and young adults with very high resting LVOT gradients (>100 mm Hg).
2b	C-LD	5. For severely symptomatic patients with obstructive HCM, SRT in eligible patients,* performed at experienced centers† (Table 3 and Table 4), may be considered as an alternative to escalation of medical therapy after shared decision-making including risks and benefits of all treatment options (10,213,221,234,235).
3: Harm	C-LD	6. For patients with HCM who are asymptomatic and have normal exercise capacity, SRT is not recommended (224,232).
3: Harm	B-NR	7. For symptomatic patients with obstructive HCM in whom SRT is an option, mitral valve replacement should not be performed for the sole purpose of relief of LVOTO (236,237).

*General eligibility criteria for septal reduction therapy: a) Clinical: Severe dyspnea or chest pain (usually NYHA functional class III or class IV), or occasionally other exertional symptoms (e.g., syncope, near syncope), when attributable to LVOTO, that interferes with everyday activity or quality of life despite optimal medical therapy. b) Hemodynamic: Dynamic LVOT gradient at rest or with physiologic provocation with approximate peak gradient of ≥50 mm Hg, associated with septal hypertrophy and SAM of mitral valve. c) Anatomic: Targeted anterior septal thickness sufficient to perform the procedure safely and effectively in the judgment of the individual operator. †Comprehensive or primary HCM centers with demonstrated excellence in clinical outcomes for these procedures (Table 3 and Table 4).

FIGURE 4 Management of Symptoms in Patients With HCM



Colors correspond to the Class of Recommendation in Table 2. GL indicates guideline; HCM, hypertrophic cardiomyopathy; HFpEF, heart failure with preserved ejection fraction; HFrEF, heart failure with reduced ejection fraction; and SRT, septal reduction therapy.

5.2. Management of Patients With Nonobstructive HCM With Preserved EF

Recommendations for Management of Patients With Nonobstructive HCM With Preserved EF
Referenced studies that support the recommendations are summarized in Online Data Supplement 15.

COR	LOE	RECOMMENDATIONS
1	C-LD	1. In patients with nonobstructive HCM with preserved EF and symptoms of exertional angina or dyspnea, beta blockers or non-dihydropyridine calcium channel blockers are recommended (209,238–246).
2a	C-EO	2. In patients with nonobstructive HCM with preserved EF, it is reasonable to add oral diuretics when exertional dyspnea persists despite the use of beta blockers or non-dihydropyridine calcium channel blockers.
2b	C-LD	3. In patients with nonobstructive HCM with preserved EF, the usefulness of angiotensin-converting enzyme inhibitors and angiotensin receptor blockers in the treatment of symptoms (angina and dyspnea) is not well established (247).
2b	C-LD	4. In highly selected patients with apical HCM with severe dyspnea or angina (NYHA class III or class IV) despite maximal medical therapy, and with preserved EF and small LV cavity size (LV end-diastolic volume $<$50 mL/m^2 and LV stroke volume $<$30 mL/m^2), apical myectomy by experienced surgeons at comprehensive centers may be considered to reduce symptoms (248).
2b	C-EO	5. In asymptomatic patients with non-obstructive HCM, the benefit of beta blockers or calcium channel blockers is not well established.

5.3. Management of Patients With HCM and Atrial Fibrillation

Recommendations for Management of Atrial Fibrillation
Referenced studies that support the recommendations are summarized in Online Data Supplement 16.

COR	LOE	RECOMMENDATIONS
1	B-NR	1. In patients with HCM and clinical AF, anticoagulation is recommended with direct-acting oral anticoagulants as first-line option and vitamin K antagonists as second-line option, independent of CHA_2DS_2-VASc score (249–253).
1	C-LD	2. In patients with HCM and subclinical AF detected by internal or external cardiac device or monitor of >24 hours' duration for a given episode, anticoagulation is recommended with direct-acting oral anticoagulants as first-line option and vitamin K antagonists as second-line option, independent of CHA_2DS_2-VASc score (102,103,249,254).
1	C-LD	3. In patients with AF in whom rate control strategy is planned, either beta blockers, verapamil, or diltiazem are recommended, with the choice of agents according to patient preferences and comorbid conditions (104,255).
2a	C-LD	4. In patients with HCM and subclinical AF detected by internal or external device or monitor, of >5 minutes' but <24 hours' duration for a given episode, anticoagulation with direct-acting oral anticoagulants as first-line option and vitamin K antagonists as second-line option can be beneficial, taking into consideration duration of AF episodes, total AF burden, underlying risk factors, and bleeding risk (102,103,249,254,256).
2a	B-NR	5. In patients with HCM and poorly tolerated AF, a rhythm control strategy with cardioversion or antiarrhythmic drugs can be beneficial with the choice of an agent according to AF symptom severity, patient preferences, and comorbid conditions (104,210,257–268).

(Continued)

2a	B-NR	6. In patients with HCM and symptomatic AF, as part of a AF rhythm control strategy, catheter ablation for AF can be effective when drug therapy is ineffective, contraindicated, or not the patient's preference (257,269,270).
2a	B-NR	7. In patients with HCM and AF who require surgical myectomy, concomitant surgical AF ablation procedure can be beneficial for AF rhythm control (104,258,271–273).

5.4. Management of Patients With HCM and Ventricular Arrhythmias

Recommendations for the Management of Patients With HCM and Ventricular Arrhythmias
Referenced studies that support the recommendations are summarized in Online Data Supplement 17.

COR	LOE	RECOMMENDATIONS
1	B-NR	1. In patients with HCM and recurrent poorly tolerated life-threatening ventricular tachyarrhythmias refractory to maximal antiarrhythmic drug therapy and ablation, heart transplantation assessment is indicated in accordance with current listing criteria (21,274).
1	Amiodarone, B-NR Dofetilide, C-LD Mexiletine, C-LD Sotalol, C-LD	2. In adults with HCM and symptomatic ventricular arrhythmias or recurrent ICD shocks despite beta-blocker use, antiarrhythmic drug therapy listed is recommended, with the choice of agent guided by age, underlying comorbidities, severity of disease, patient preferences, and balance between efficacy and safety (275–278).
1	C-LD	3. In children with HCM and recurrent ventricular arrhythmias despite beta-blocker use, antiarrhythmic drug therapy (amiodarone (275,276), mexiletine (278), sotalol (275,276)) is recommended, with the choice of agent guided by age, underlying comorbidities, severity of disease, patient preferences, and balance of efficacy and safety.
1	C-LD	4. In patients with HCM and pacing-capable ICDs, programming antitachycardia pacing is recommended to minimize risk of shocks (279,280).
2a	C-LD	5. In patients with HCM and recurrent symptomatic sustained monomorphic ventricular tachycardia, or recurrent ICD shocks despite optimal device programming, and in whom antiarrhythmic drug therapy is either ineffective, not tolerated, or not preferred, catheter ablation can be useful for reducing arrhythmia burden (20,281,282).

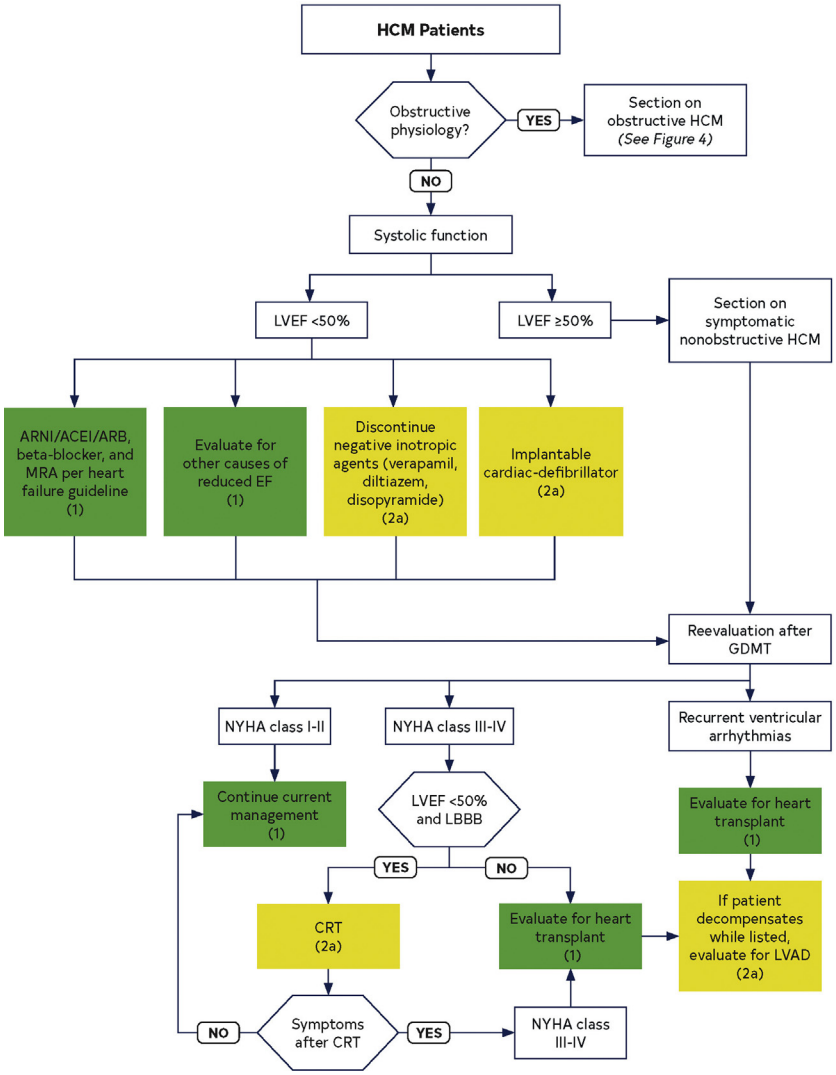
5.5. Management of Patients With HCM and Advanced HF

Figure 5 presents a heart failure algorithm.

Recommendations for Patients With HCM and Advanced HF
Referenced studies that support the recommendations are summarized in Online Data Supplement 18.

COR	LOE	RECOMMENDATIONS
1	C-LD	1. In patients with HCM who develop systolic dysfunction with an LVEF <50%, guideline-directed therapy for HF with reduced EF is recommended (38,283,284).
1	C-LD	2. In patients with HCM and systolic dysfunction, diagnostic testing to assess for concomitant causes of systolic dysfunction (such as coronary artery disease) is recommended (22,36,285).
1	B-NR	3. In patients with nonobstructive HCM and advanced HF (NYHA functional class III to class IV despite guideline-directed therapy) cardiopulmonary exercise test should be performed to quantify the degree of functional limitation and aid in selection of patients for heart transplantation or mechanical circulatory support (113,114).
1	B-NR	4. In patients with nonobstructive HCM and advanced HF (NYHA class III to class IV despite guideline-directed therapy) or with life-threatening ventricular arrhythmias refractory to maximal guideline-directed therapy, assessment for heart transplantation in accordance with current listing criteria is recommended (21,274,286,287).
2a	C-EO	5. For patients with HCM who develop systolic dysfunction (LVEF <50%), it is reasonable to discontinue previously indicated negative inotropic agents (specifically, verapamil, diltiazem, or disopyramide).
2a	B-NR	6. In patients with nonobstructive HCM and advanced HF (NYHA functional class III to class IV despite GDMT) who are candidates for heart transplantation, continuous-flow LV assist device therapy is reasonable as a bridge to heart transplantation (288–291).
2a	C-LD	7. In patients with HCM and LVEF <50%, ICD placement can be beneficial (284).
2a	C-LD	8. In patients with HCM and LVEF <50%, NYHA functional class II to class IV symptoms despite guideline-direct therapy, and LBBB, CRT can be beneficial to improve symptoms (199–203).

FIGURE 5 Heart Failure Algorithm



Colors correspond to the Class of Recommendation in Table 2. ACE indicates angiotensin-converting enzyme; ARB, angiotensin receptor blocker; ARNI, angiotensin receptor-neprilysin inhibitors; CRT, cardiac resynchronization therapy; EF, ejection fraction; GDMT, guideline-directed management and therapy; HCM, hypertrophic cardiomyopathy; LBBB, left bundle branch block; LVAD, left ventricular assist device; LVEF, left ventricular ejection fraction; MRA, mineralocorticoid receptor antagonist; and NYHA, New York Heart Association.

6. LIFESTYLE CONSIDERATIONS FOR PATIENTS WITH HCM

6.1. Sports and Activity

Recommendations for Sports and Activity
Referenced studies that support the recommendations are summarized in Online Data Supplement 19.

COR	LOE	RECOMMENDATIONS
1	B-NR	1. For most patients with HCM, mild- to moderate-intensity recreational* exercise is beneficial to improve cardiorespiratory fitness, physical functioning, and quality of life, and for their overall health in keeping with physical activity guidelines for the general population (292–294).
1	C-EO	2. For athletes with HCM, a comprehensive evaluation and shared discussion of potential risks of sports participation by an expert provider is recommended (295).
2a	C-EO	3. For most patients with HCM, participation in low-intensity competitive sports is reasonable (2,297).
2a	C-LD	4. In individuals who are genotype-positive, phenotype-negative for HCM, participation in competitive athletics of any intensity is reasonable (2,174.296–301).
2b	C-LD	5. For patients with HCM, participation in high-intensity recreational activities or moderate- to high-intensity competitive sports activities may be considered after a comprehensive evaluation and shared discussion, repeated annually with an expert provider who conveys that the risk of sudden death and ICD shocks may be increased, and with the understanding that eligibility decisions for competitive sports participation often involve third parties (e.g., team physicians, consultants, and other institutional leadership) acting on behalf of the schools or teams (174,295,298–301).
3: Harm	B-NR	6. In patients with HCM, ICD placement for the sole purpose of participation in competitive athletics should not be performed (2,174,302).

*Recreational exercise is done for the purpose of leisure with no requirement for systematic training and without the purpose to excel or compete against others.

6.2. Occupation

Recommendations for Occupation in Patients With HCM

COR	LOE	RECOMMENDATIONS
2a	C-EO	1. For patients with HCM, it is reasonable to follow Federal Motor Carrier Safety Administration cardiovascular disease guidelines that permit driving commercial motor vehicles, if they do not have an ICD or any major risk factors for SCD and are following a guideline-directed management plan (303).
2a	C-EO	2. For pilot aircrew with a diagnosis of HCM, it is reasonable to follow Federal Aviation Administration guidelines that permit consideration of multicrew flying duties, provided they are asymptomatic, are deemed low risk for SCD, and can complete a maximal treadmill stress test at 85% peak heart rate (304).
2b	C-EO	3. Patients with HCM may consider occupations that require manual labor, heavy lifting, or a high level of physical performance after a comprehensive clinical evaluation, risk stratification for SCD, and implementation of guideline-directed management. Before a shared decision between a clinician and patient is reached, the clinician should convey that risks associated with the physical requirements of these occupations are uncertain.

6.3. Pregnancy

Recommendations for Pregnancy in Patients With HCM
Referenced studies that support the recommendations are summarized in Online Data Supplement 20.

COR	LOE	RECOMMENDATIONS
1	B-NR	1. For pregnant women with HCM and AF or other indications for anticoagulation, low-molecular-weight heparin or vitamin K antagonists (at maximum therapeutic dose of <5 mg daily) are recommended for stroke prevention (249,250,305).
1	C-LD	2. In pregnant women with HCM, selected beta blockers should be administered for symptoms related to outflow tract obstruction or arrhythmias, with monitoring of fetal growth (306,307).
1	C-LD	3. In most pregnant women with HCM, vaginal delivery is recommended as the first-choice delivery option (306,308).
1	B-NR	4. In affected families with HCM, preconceptional and prenatal reproductive and genetic counseling should be offered (302,306,307,308).
1	C-EO	5. For pregnant women with HCM, care should be coordinated between their cardiologist and an obstetrician. For patients with HCM who are deemed high risk, consultation is advised with an expert in maternal-fetal medicine.
2a	C-LD	6. For women with clinically stable HCM who wish to become pregnant, it is reasonable to advise that pregnancy is generally safe as part of a shared discussion regarding potential maternal and fetal risks, and initiation of guideline-directed therapy (309-312).
2a	C-LD	7. In pregnant women with HCM, cardioversion for new or recurrent AF, particularly if symptomatic, is reasonable (302,313).
2a	C-LD	8. In pregnant women with HCM, general or epidural anesthesia is reasonable, with precautions to avoid hypotension (310).
2a	C-EO	9. In pregnant women with HCM, it is reasonable to perform serial echocardiography, particularly during the second or third trimester when hemodynamic load is highest, or if clinical symptoms develop (309).
2b	C-EO	10. In pregnant women with HCM, fetal echocardiography may be considered for diagnosis of fetal HCM in the context of prenatal counseling.

6.4. Comorbidities

Table 9 addresses lifestyle considerations for patients with HCM.

Recommendations for Patients With Comorbidities
Referenced studies that support the recommendations are summarized in Online Data Supplement 21.

COR	LOE	RECOMMENDATIONS
1	C-EO	1. In patients with HCM, adherence to the guidelines on the prevention of atherosclerotic cardiovascular disease is recommended to reduce risk of cardiovascular events (294).
1	B-NR	2. In patients with HCM who are overweight or obese, counseling and comprehensive lifestyle interventions are recommended for achieving and maintaining weight loss (294) and possibly lowering the risk of developing LVOTO, HF, and AF (314–316).
1	C-LD	3. In patients with HCM and hypertension, lifestyle modifications and medical therapy for hypertension are recommended (294) with preference for beta blockers and non-dihydropyridine calcium channel blockers in patients with obstructive HCM (310,316–319).
1	C-LD	4. In patients with HCM, assessment for symptoms of sleep disordered breathing is recommended and, if present, referral to a sleep medicine specialist for evaluation and treatment (320–323).

TABLE 9 Lifestyle Considerations for Patients With HCM

Lifestyle Considerations*	
Sports/activity	For most patients with HCM, mild- to moderate-intensity recreational exercise is beneficial to improve cardiorespiratory fitness, physical functioning, and quality of life, and for their overall health in keeping with physical activity guidelines for the general population
Pregnancy	For women with clinically stable HCM who wish to become pregnant, it is reasonable to advise that pregnancy is generally safe as part of a shared discussion regarding potential maternal and fetal risks, and initiation of guideline-directed therapy.
Comorbidities	The clinician should monitor and counsel patients on prevention and treatment of comorbid conditions that can worsen severity of HCM (atherosclerotic cardiovascular disease, obesity, hypertension, sleep-disordered breathing)

*Shared decision-making is an important component of counseling and lifestyle modifications.

HCM indicates hypertrophic cardiomyopathy.

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