

AHA Clinical Update

ADAPTED FROM:

**2024 AHA/ACC/AMSSM/HRS/PACES/SCMR
Guideline for the Management of
Hypertrophic Cardiomyopathy**



American
Heart
Association.

Table 1. Applying Class of Recommendation and Level of Evidence to Clinical Strategies, Interventions, Treatments, or Diagnostic Testing in Patient Care

| CLASS (STRENGTH) OF RECOMMENDATION | LEVEL (QUALITY) OF EVIDENCE‡ |
|---|---|
| CLASS 1 (STRONG) Benefit >>> Risk Suggested phrases for writing recommendations: <ul style="list-style-type: none"> • Is recommended • Is indicated/useful/effective/beneficial • Should be performed/administered/other • Comparative-Effectiveness Phrases†: <ul style="list-style-type: none"> – Treatment/strategy A is recommended/indicated in preference to treatment B – Treatment A should be chosen over treatment B | LEVEL A <ul style="list-style-type: none"> • High-quality evidence‡ from more than 1 RCT • Meta-analyses of high-quality RCTs • One or more RCTs corroborated by high-quality registry studies |
| CLASS 2a (MODERATE) Benefit >> Risk Suggested phrases for writing recommendations: <ul style="list-style-type: none"> • Is reasonable • Can be useful/effective/beneficial • Comparative-Effectiveness Phrases†: <ul style="list-style-type: none"> – Treatment/strategy A is probably recommended/indicated in preference to treatment B – It is reasonable to choose treatment A over treatment B | LEVEL B-R (Randomized) <ul style="list-style-type: none"> • Moderate-quality evidence‡ from 1 or more RCTs • Meta-analyses of moderate-quality RCTs |
| CLASS 2b (Weak) Benefit ≥ Risk Suggested phrases for writing recommendations: <ul style="list-style-type: none"> • May/might be reasonable • May/might be considered • Usefulness/effectiveness is unknown/unclear/uncertain or not well-established | LEVEL B-NR (Nonrandomized) <ul style="list-style-type: none"> • Moderate-quality evidence‡ from 1 or more well-designed, well-executed nonrandomized studies, observational studies, or registry studies • Meta-analyses of such studies |
| CLASS 3: No Benefit (MODERATE) Benefit = Risk Suggested phrases for writing recommendations: <ul style="list-style-type: none"> • Is not recommended • Is not indicated/useful/effective/beneficial • Should not be performed/administered/other | LEVEL C-LD (Limited Data) <ul style="list-style-type: none"> • 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 |
| CLASS 3: Harm (STRONG) Risk > Benefit Suggested phrases for writing recommendations: <ul style="list-style-type: none"> • Potentially harmful • Causes harm • Associated with excess morbidity/mortality • Should not be performed/administered/other | LEVEL C-EO (Expert Opinion) <ul style="list-style-type: none"> • 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 recommendation (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.

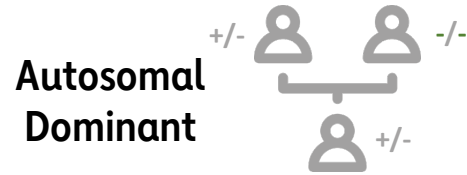
•‡The method of assessing quality is evolving, including the application of standardized, widely-used, and preferably validated evidence grading tools; and for systematic reviews, the incorporation of an Evidence Review Committee.

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



Hypertrophic Cardiomyopathy Prevalence and Characteristics

Inheritance Pattern



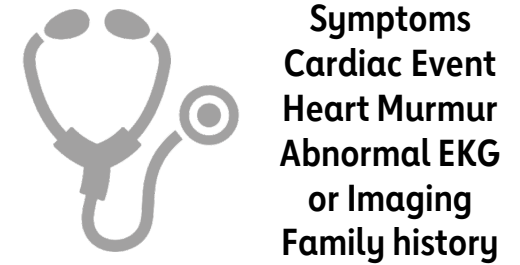
Sex Distribution



Disease Prevalence



Triggers for Evaluation



Underlying SDOH likely drive differences in prevalence, genetic testing, and cardiovascular outcomes by race and ethnicity

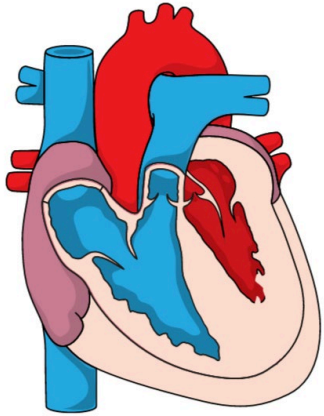
Differential Diagnosis: Non-HCM Causes of LV Hypertrophy

Metabolic & Multi-organ Syndromes
RASopathies (Noonan Syndrome)
Glycogen / Lysosomal storage diseases
Cardiac Amyloidosis
Sarcoidosis
Danon disease

Secondary Causes
Athlete's heart
Uncontrolled Hypertension
Valvular & subvalvular aortic stenosis

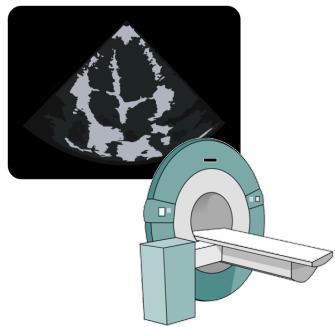
Abbreviations: EKG indicates electrocardiogram; SDOH, social determinants of health; and RAS, reticular activating system.

Defining Hypertrophic Cardiomyopathy in 2024



- Characterized by left ventricular hypertrophy
Asymmetric septal hypertrophy is most characteristic
- No other cardiac, systemic or metabolic disease capable of producing the magnitude of increased LV wall thickness present
- Disease-causing variant in a sarcomere gene identified or genetic etiology unresolved

Diagnostic Criteria in Adults



2D echocardiography or cardiac MRI

Maximal end-diastolic LV wall thickness > 15 mm

or

Maximal end-diastolic LV wall thickness 13-14 mm if there is a family history of HCM or a pathogenic sarcomere gene is present

Diagnostic Criteria in Children

2D echocardiography or cardiac MRI

LV wall thickness z-score > 2.5

or

LV wall thickness z-score >2 if there is a family history of HCM or a pathogenic sarcomere gene is present

Abbreviations: 2D indicates two dimensional; MRI, magnetic resonance imaging; mm, millimeter

Adverse Events Associated Hypertrophic Cardiomyopathy

Although some patients with HCM have a normal life expectancy without limiting symptoms, many will have important consequences



**Sudden
Death**



**Progressive
Functional
Limitation**



Atrial Fibrillation

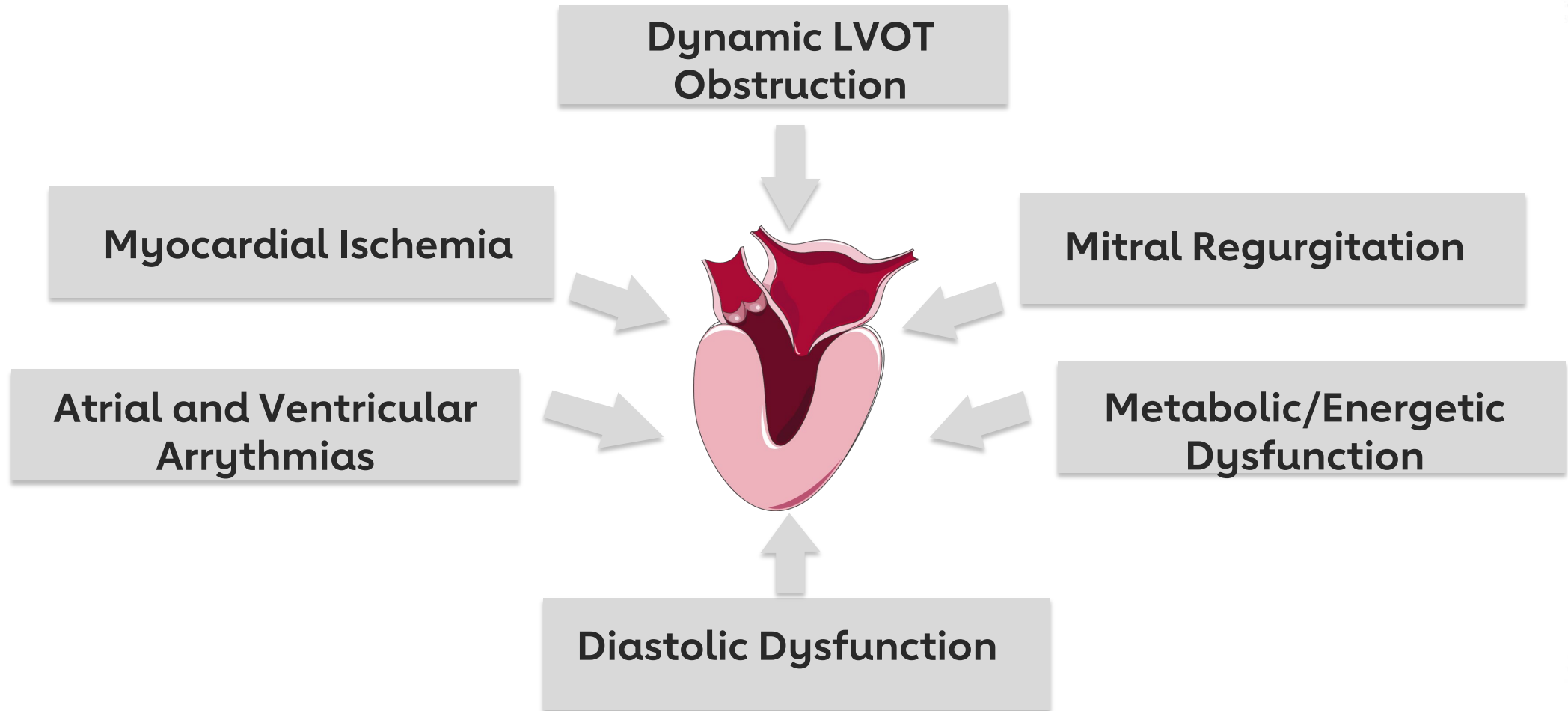


**Heart
Failure**



Thromboembolism

Pathophysiology of Hypertrophic Cardiomyopathy



Abbreviations: LVOT indicates left ventricular outflow tract.

Pathophysiology of HCM: LV Outflow Tract Obstruction



LVOTO, either at rest or with provocation, is present in many patients with HCM and primarily caused by systolic anterior motion of the mitral valve.

Peak gradient of ≥ 30 mm Hg is considered indicative of obstruction

Resting or provoked gradients ≥ 50 mmHg generally considered to be the threshold for advanced pharmacologic or septal reduction therapy in those patients with symptoms refractory to standard management.



Identify Site and Characteristics of Obstruction

Use invasive assessment for LVOTO if clinical and echo findings are discordant

Management Dependent on Site and Characteristics of Obstruction



LVOTO in HCM is primarily caused by basal septal hypertrophy and systolic anterior motion of the mitral valve

LVOTO in HCM is dynamic and sensitive to ventricular preload, afterload, and contractility

Abbreviations: HCM indicates hypertrophic cardiomyopathy; and LVOTO, left ventricular outflow tract.

Ommen, S.R. et al, 2024 AHA/ACC/AMSSM/HRS/PACES/SCMR Guideline for the Management of Hypertrophic Cardiomyopathy. *Circulation*.

Pathophysiology of HCM: Diastolic Dysfunction

Features of HCM that contribute to Diastolic Dysfunction



- Abnormal intracellular Ca reuptake
- Excess actin-myosin binding
- Microvascular ischemia
- Altered energetics



- Altered Ventricular Load with High Intracavitary Pressures
- Impaired LV compliance



Diastolic Dysfunction



Diastolic Dysfunction can contribute to:

Decreased Exercise Capacity

Heart Failure

Dyspnea

Adverse prognosis independent of LVOTO

Pathophysiology of HCM: Mitral Valve Abnormalities

Common abnormalities of the Mitral Valve in HCM

- Excessive leaflet length
- Anomalous papillary muscle insertion
- Anteriorly displaced papillary muscles



Mitral Regurgitation occurs

Primarily from leaflet abnormalities

Secondarily from Systolic Anterior Motion



Factors that affect the severity of LVOTO also may affect the degree of MR. Thus, imaging should be performed at rest and with provocation.

Abbreviations: HCM indicates hypertrophic cardiomyopathy; LVOTO, left ventricular outflow tract; MR, mitral regurgitation

Ommen, S.R. et al, 2024 AHA/ACC/AMSSM/HRS/PACES/SCMR Guideline for the Management of Hypertrophic Cardiomyopathy. *Circulation*.

Shared Decision-Making in HCM



Discussions should involve:

- Disclosure of risk and benefits of all screenings and therapies
- Anticipated outcomes of all options
- Goals, concerns and preferences of the patient (and family if the patient is a minor) (Class 1)

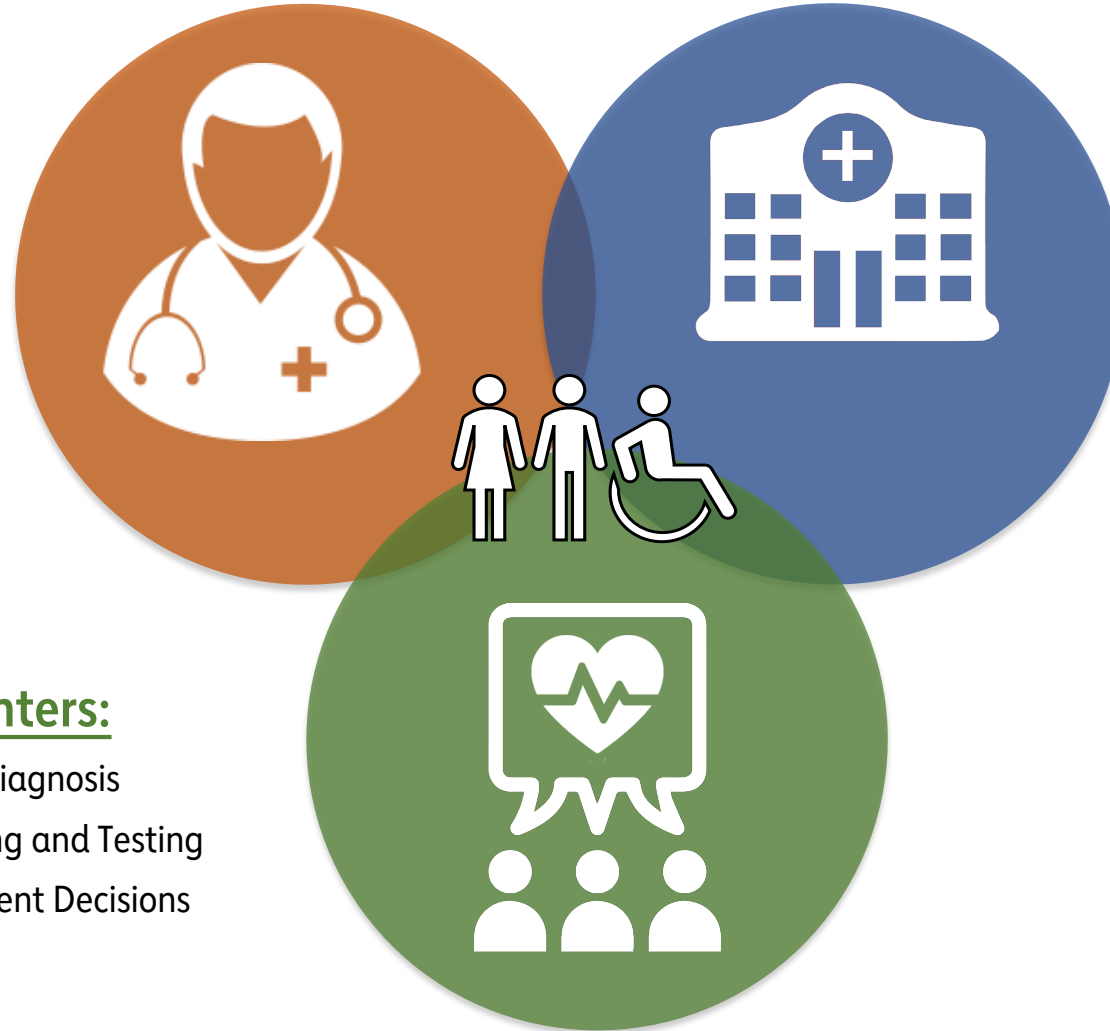
Shared decision discussions should be applied to:

- Genetic testing
- Medical and invasive therapies for LVOT obstruction
- Sudden death screening and ICD Implantation
- Participation in high-intensity exercise and competitive sports
- Pregnancy

Patient Centered Team-Based Care

Cardiologists Outside of HCM Centers:

- Initial and Surveillance Testing
- Initial Treatment Recommendations
- Rapid Assessment for Change in Disease Course



HCM Centers:

- Confirmation of Diagnosis
- Genetic Counseling and Testing
- Advanced Treatment Decisions

Comprehensive HCM Centers:

- HCM Center Activities, Plus
- Invasive Septal Reduction Therapies
- Catheter Ablation for Ventricular and Complex Atrial Tachyarrhythmias
- Advanced Heart Failure Therapies
- Management during Pregnancy

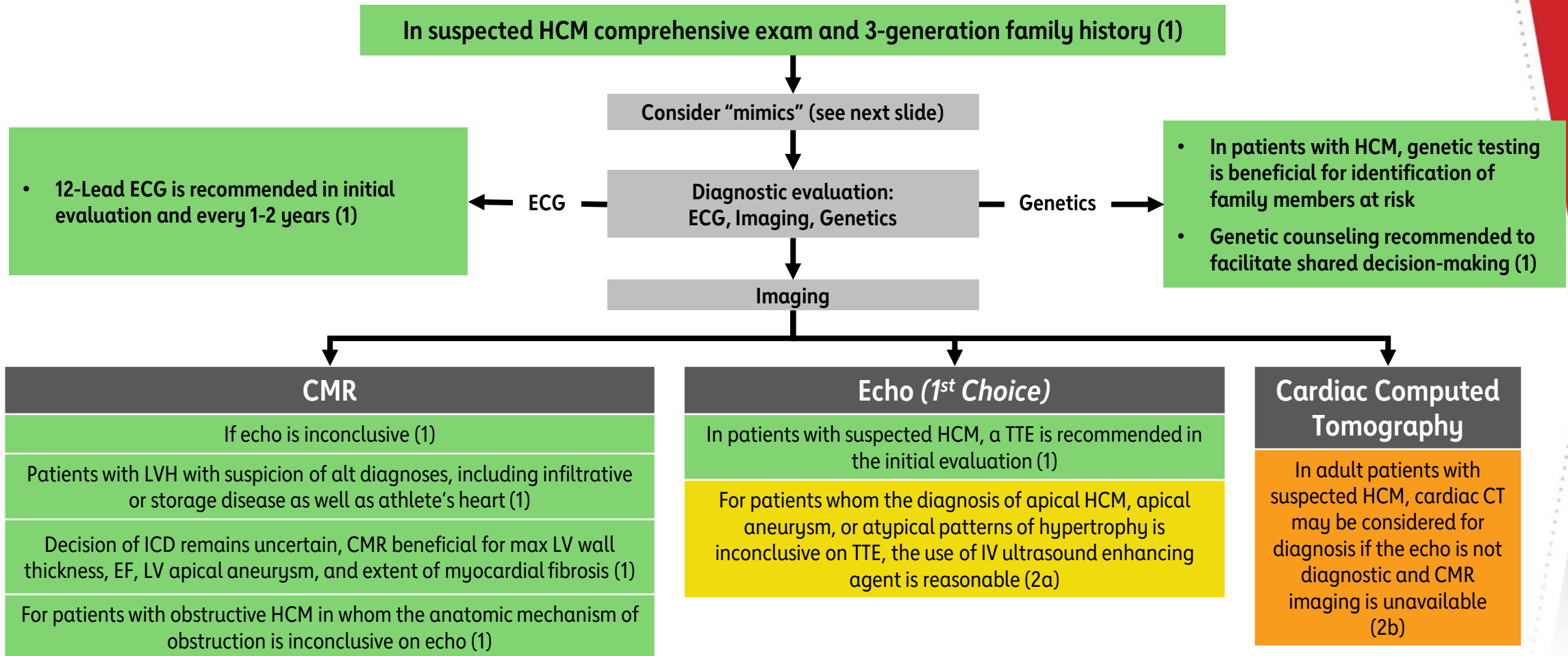
Abbreviations: HCM indicates hypertrophic cardiomyopathy.

Septal Reduction Therapy



- Referral to a comprehensive HCM Center with expertise in invasive septal reduction therapy to ensure optimal outcomes
- Invasive septal reduction therapy performed at centers with lower volumes and less expertise may be associated with worse outcomes

Diagnosis and Initial Evaluation

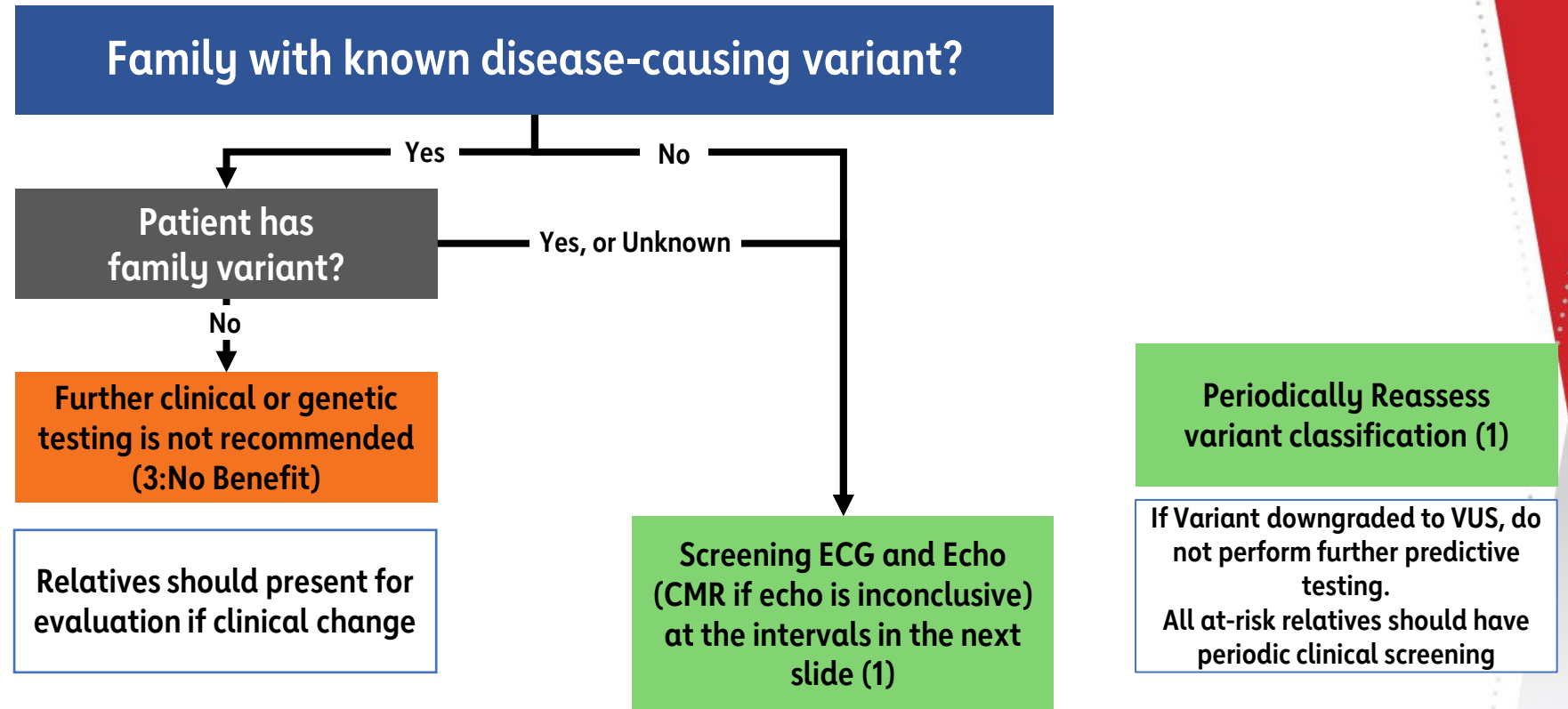


Abbreviations: CMR indicates cardiac magnetic resonance; CT, computed tomography; ECG, electrocardiography; echo, echocardiography; EF, ejection fraction; HCM, hypertrophic cardiomyopathy; ICD, implantable cardiac defibrillator; IV, intravenous; LVH, left ventricular hypertrophy; SCD, sudden cardiac death; and TTE, transthoracic echo

Clinical Features in Patients with “HCM Mimics”

| LIFE STAGE | SYSTEMIC FEATURES | POSSIBLE ETIOLOGY | DIAGNOSTIC APPROACH |
|---|---|--|--|
| Infants (0-12 months) and toddlers | Dysmorphic features, failure to thrive, metabolic acidosis | <ul style="list-style-type: none"> • RASopathies (e.g. Noonan Syndrome) • Glycogen storage diseases, other metabolic or mitochondrial diseases • Infant of a mother with diabetes | <ul style="list-style-type: none"> • Geneticist assessment • Newborn metabolic screening • Specific metabolic assays • Genetic testing |
| Early childhood | Delayed or abnormal cognitive development, visual or hearing impairment | <ul style="list-style-type: none"> • RASopathies (e.g. Noonan Syndrome) • Mitochondrial diseases | <ul style="list-style-type: none"> • Biochemical screening • Genetic testing |
| School age and adolescence | Skeletal muscle weakness or movement disorder | <ul style="list-style-type: none"> • Friedrich ataxia, Danon disease • Mitochondrial disease | <ul style="list-style-type: none"> • Biochemical screening • Neuromuscular assessment • Genetic testing |
| Adulthood | Movement disorder, peripheral neuropathy, renal dysfunction | Anderson-Fabry disease, Friedrich ataxia, infiltrative disorders (e.g., amyloidosis), glycogen storage diseases | <ul style="list-style-type: none"> • Biochemical screening, • Neuromuscular assessment • Genetic testing |

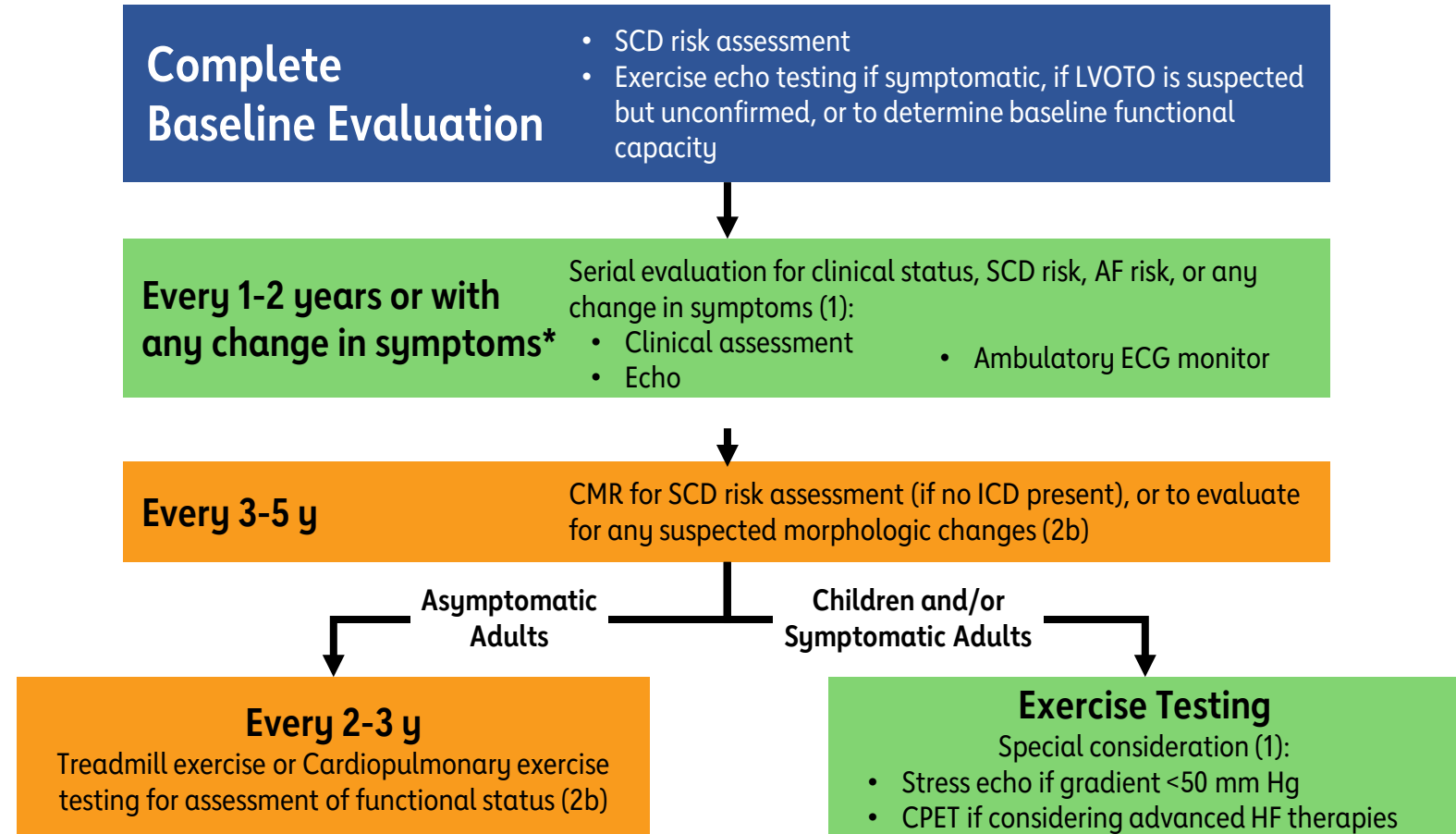
Guidance for Family Management



Abbreviations: 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.

Guidance for Individuals Diagnosed with Clinical HCM

Phenotype Positive



Abbreviations: 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.

Screening with Electrocardiography and 2D Echocardiography Recommendations in Asymptomatic Family Members*

| AGE OF FIRST-DEGREE RELATIVE | INITIATION OF SCREENING | REPEAT ECG, ECHO |
|--|---|------------------|
| Pediatric | | |
| Children and adolescents from families with a disease-causing sarcomere variant, and families with early onset disease | At the time HCM is diagnosed in a family member | Every 1-2 y |
| All other children | At any time after HCM is diagnosed in a family member but no later than puberty | Every 2-3 y |
| Adults | At the time HCM is diagnosed in another family member | Every 3-5 y |

* Includes all asymptomatic, phenotype-negative first-degree relatives deemed to be at-risk for developing HCM based on family history or genotype status and may sometimes include more distant relatives based on clinical judgment. Screening interval may be modified (e.g., at onset of new symptoms or in families with a malignant clinical course or late-onset HCM).

Abbreviations: ECG indicates electrocardiogram; Echo, echocardiogram; and HCM, hypertrophic cardiomyopathy.

Genetic Testing

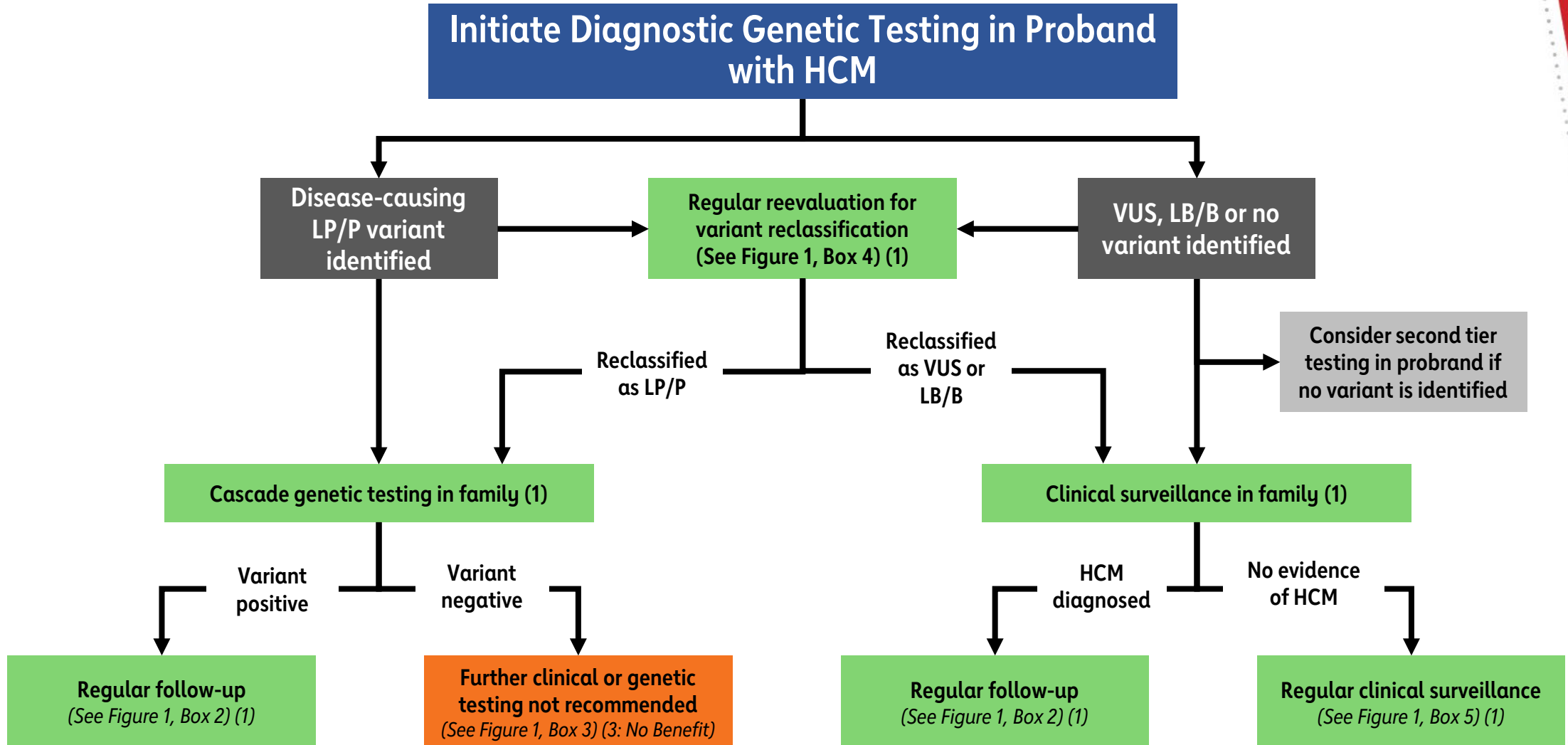






Figure 1 refers to figure on slide 18 (two slides prior).

Abbreviations: HCM indicates hypertrophic cardiomyopathy; LB/B, likely benign/benign; LP/P, likely pathogenic or pathogenic; and VUS: variant of unknown significance

Heart Rhythm Assessment in HCM

| COR | RECOMMENDATIONS |
|-----|--|
| 1 |  24- to 48-hour ambulatory ECG monitoring is recommended in the initial evaluation and as part of periodic follow up (every 1-2 years) to identify patients at risk for SCD and guide management of arrhythmias (Class 1) |
| 1 |  In patients with HCM who develop palpitations or lightheadedness, extended (>24h) ECG monitoring or event recording is recommended (Class 1) |
| 1 |  In patients with HCM who are deemed high risk for AF based on risk factors or risk score, and who are eligible for anticoagulation, extended ambulatory monitoring is recommended to screen for AF as part of initial evaluation and annual follow-up. (Class 1) |
| 2b |  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 1-2 years) (Class 2b) |

Abbreviations: AF indicates atrial fibrillation; ECG:, electrocardiography; HCM, hypertrophic cardiomyopathy; and SCD, sudden cardiac death.

Risk Assessment of Sudden Cardiac Death (SCD) in HCM



At initial evaluation and every 1-2 years (Class 1)



Assess the following (Class 1):

- Personal history of cardiac arrest, sustained ventricular arrhythmia, OR unexplained syncope suspected to be arrhythmic
- Family history of premature SCD in a close relative
- Maximal LV wall thickness ≥ 30 mm, EF $\leq 50\%$, LV apical aneurysm
- NSVT or VT episodes on continuous ambulatory electrocardiographic monitoring



CMR imaging to help decision regarding ICD if risk remains “unresolved” or if the patient is unsure about ICD placement (Class 1)



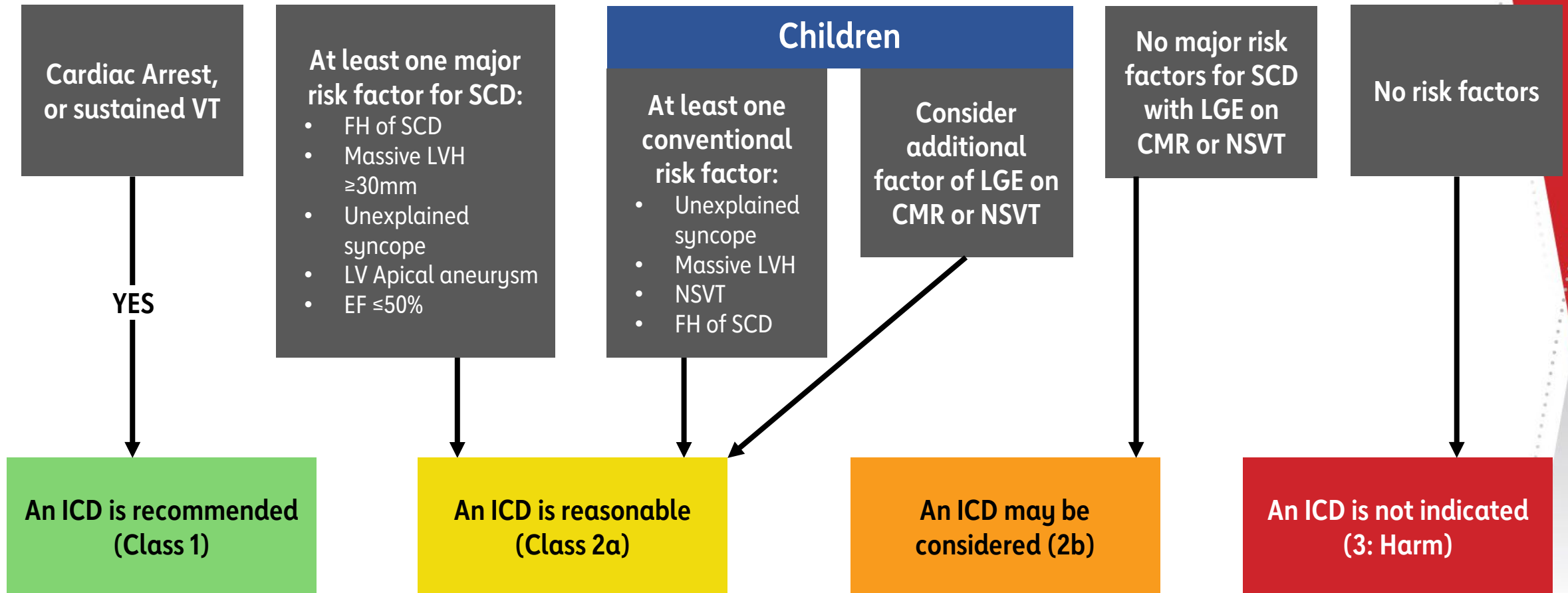
≥ 16 years old, reasonable to obtain echocardiographic LA diameter and maximal LVOT gradient to assist in shared decision making for ICD placement (Class 2a)



< 16 years of age it is reasonable to calculate an estimated 5-year sudden death risk that includes echocardiographic parameters and genotype that may be useful during shared decision-making for ICD placement (Class 2a)

Abbreviations: EF indicates ejection fraction; NSVT, non-sustained ventricular tachycardia; CMR, cardiovascular magnetic resonance; ICD, implantable cardioverter-defibrillator; LA, left atrium; LVOT, left ventricular outflow tract.

ICD Placement in High-Risk Patients with HCM



Abbreviations: ICD indicates implantable cardioverter defibrillator; SCD, sudden cardiac death; VF, ventricular fibrillation; VT, ventricular tachycardia; LVH, left ventricular hypertrophy; FH, family history; EF, ejection fraction; NSVT, non-sustained ventricular tachycardia; LGE, late gadolinium enhancement; and CMR, cardiac magnetic resonance imaging.

Pharmacologic Management of Obstructive and Non-Obstructive HCM

Obstructive

Symptoms r/t LVOTO

Step-Wise Approach:

1. **Non-Vasodilating β -Blocker**
2. If not effective or not tolerated, switch to **Non-Dihydropyridine CCBs**
3. If persistent severe symptoms,
 - Add **Myosin Inhibitor** (ie Mavacamten)
 - OR-
 - Add **Disopyramide**
 - OR-
 - **Septal Reduction Therapy** performed at experienced centers (Class 1)

Consider Discontinuing:

- Vasodilators
- Digoxin
- High Dose Diuretics (Class 2b)

If persistent dyspnea with volume overload, consider **Low Dose Diuretics** (Class 2b)

Acute Hypotension

1. **Intravenous Fluids**
2. **Phenylephrine \pm β -Blocker** (without inotropic activity) (Class 1)

Verapamil potentially HARMFUL in:

- Severe Dyspnea at Rest
- Very High Gradients (>100 mmHg)
- Children < 6 Weeks (Class 3: Harm)

Non-Obstructive HCM Preserved LVEF

Symptomatic:

β -Blocker or Non-Dihydropyridine CCBs (Class 1)

Oral Diuretic if evidence of congestion (Class 2a)

Usefulness of **ACEi / ARBs** in symptomatic patients with LVEF>50% is not well established (Class 2b)

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, apical myectomy by experienced surgeons at comprehensive centers may be considered to reduce symptoms (Class 2b)

Asymptomatic:

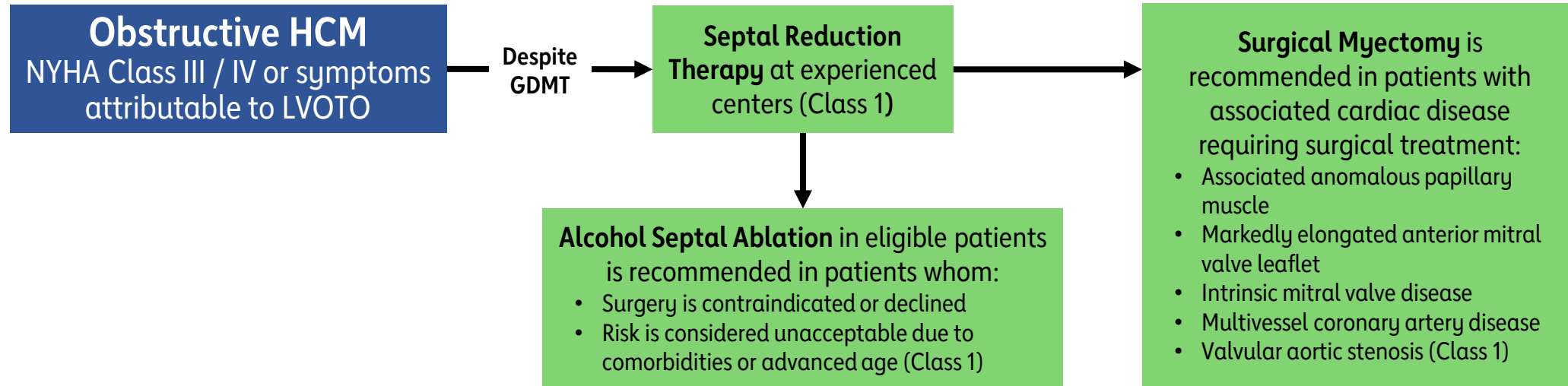
Benefits of **β -Blocker or CCBs** is not well established (Class 2b)

- ≤ 45 years
- Asymptomatic
- Pathogenic sarcomere variant carrier
- Mild phenotype:

Valsartan may be considered to potentially slow adverse cardiac remodeling (Class 2b)

Abbreviations: ACEi indicates angiotensin-converting enzyme inhibitors; ARB, angiotensin receptor blockers; CCBs, calcium channel blockers; GDMT, guideline-directed medical therapy; HCM, hypertrophic cardiomyopathy; LVOTO, left ventricular outflow tract obstruction; and r/t, related to

Invasive Management of Obstructive HCM



Mitral Valve Replacement **should not** be performed for sole purpose of relieving LVOTO (Class 3:Harm)

Septal Reduction Therapy **should not** be performed in asymptomatic patients with normal exercise capacity (Class 3:Harm)

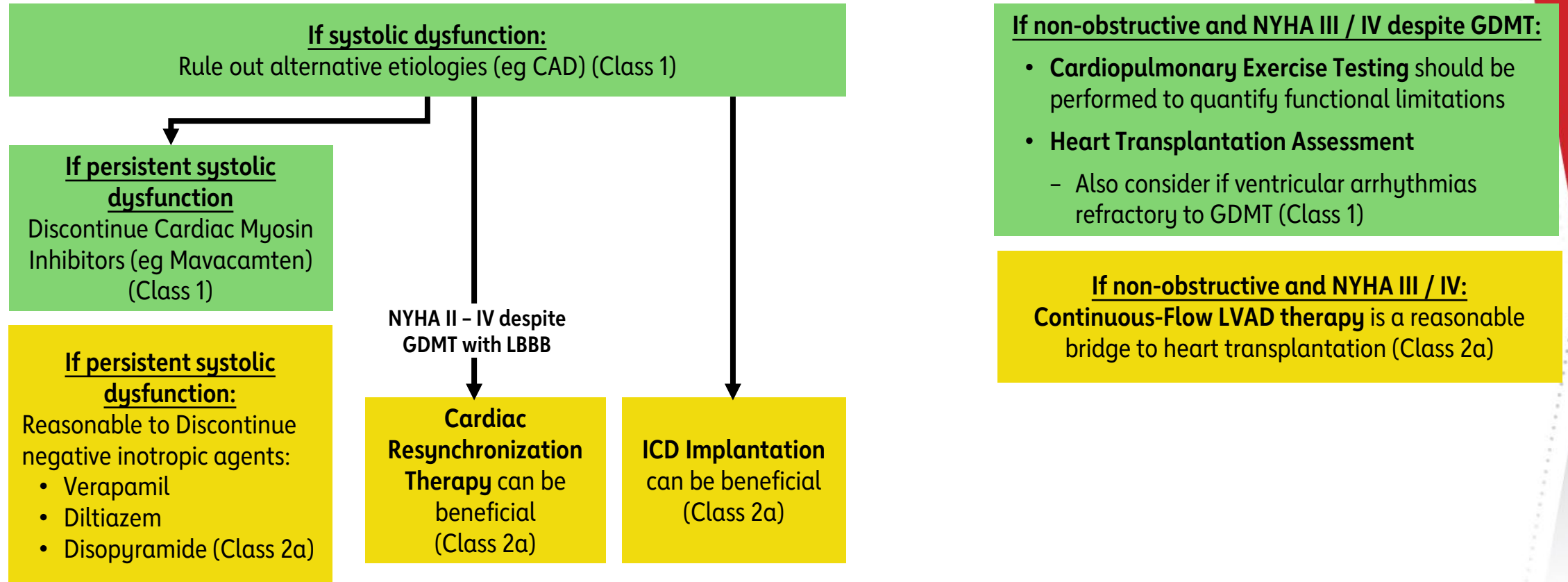
Septal Reduction Therapy **may be considered** as alternative to escalation of medical therapy after shared-decision making (Class 2b)

Surgical Myectomy is reasonable in NYHA Class II if:

- Severe PH attributable to LVOTO or MR
- LAE with ≥ 1 episodes of symptomatic AF
- Poor functional capacity attributable to LVOTO
- Children or young adults Class with very high LVOT gradients (> 100 mmHg) (Class 2b)

Abbreviations: AF indicates atrial fibrillation; GDMT, guideline-directed medical therapy; LVOTO, HCM, hypertrophic cardiomyopathy; LAE, left atrial enlargement; left ventricular outflow tract obstruction; NYHA, New York Heart Association; and MR, mitral regurgitation;

Hypertrophic Cardiomyopathy with Advanced Heart Failure



Abbreviations: CAD indicates coronary artery disease; GDMT, guideline-directed medical therapy; LBBB, left bundle branch block; LVAD, left ventricular assist device; LVEF, left ventricular ejection fraction; and NYHA, New York Heart Association

Management of Atrial Fibrillation in Patients with HCM

| COR | RECOMMENDATIONS |
|-----|--|
| 1 | In patients with clinical AF or subclinical AF (≥ 24 hours), anticoagulation with direct-acting oral anticoagulants (DOACs) is <u>first line</u> |
| 1 | Anticoagulation with Vitamin K Antagonists is <u>second line</u> |
| 1 | β-Blocker, Verapamil, or Diltiazem is recommended if pursuing rate control strategy |
| 2a | In patients with subclinical AF, lasting > 5 minutes but < 24 hours for a given episode, anticoagulation with DOAC as <u>first line</u> , and vitamin K Antagonist as <u>second line</u> can be beneficial |
| 2a | Patients with poorly tolerated AF, a rhythm control strategy with cardioversion or anti-arrhythmic drugs can be beneficial |
| 2a | AF catheter ablation can be effective when drug therapy is 1) ineffective, 2) contraindicated or 3) not patient's preference |
| 2a | In patients with AF undergoing myectomy, concomitant surgical AF ablation can be beneficial |

Abbreviations: AF indicates atrial fibrillation; and DOACs, direct-acting oral anticoagulants

HCM-AF Risk Calculator: Risk for Atrial Fibrillation in Hypertrophic Cardiomyopathy

HCM-AF Risk Calculator

Risk For Atrial Fibrillation in Hypertrophic Cardiomyopathy

This score provides patients who have an HCM diagnosis with individualized estimates of their risk for developing new-onset atrial fibrillation in the five-year period following their evaluation. These predictions are based on the previously published risk model from Carrick et al. (2021). *Circ Arrhythm Electrophysiol*, 14:e009796. DOI: 10.1161/CIRCEP.120.009796

Transverse Left Atrial Dimension, mm

Age at HCM Diagnosis, y

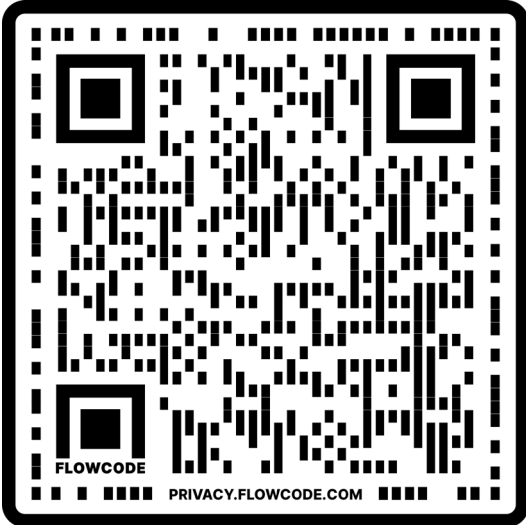
Age at Clinical Evaluation, y

NYHA Class II, III, or IV Heart Failure Symptoms No Yes

HCM-AF Score

2-Year Risk Estimate (%)

5-Year Risk Estimate (%)

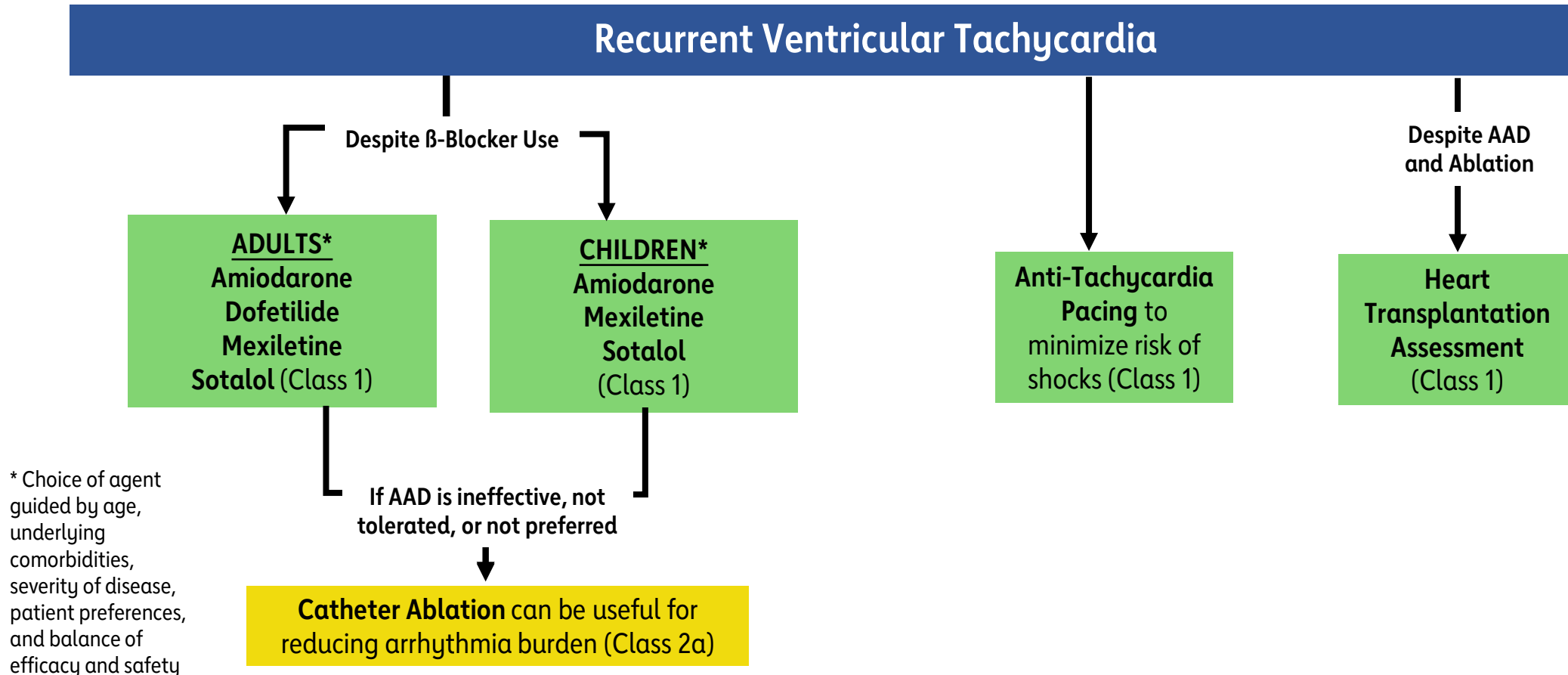


<https://professional.heart.org/en/guidelines-and-statements/hcm-af-risk-calculator>

Abbreviations: AF indicates atrial fibrillation; and HCM, hypertrophic cardiomyopathy










Management of Ventricular Tachycardia in Patients with HCM






Abbreviations: AADs indicates anti-arrhythmic drugs; and HCM, hypertrophic cardiomyopathy

Recreational Physical Activity and Competitive Sports in HCM

| COR | RECOMMENDATIONS |
|----------------------|---|
| 1 |  Mild- to moderate-intensity recreational exercise is encouraged for all patients with HCM. |
| 1 |  Elite athletes engaging in competition should undergo comprehensive evaluation with an expert provider. |
| 2a |  In individuals who are genotype-positive, phenotype-negative for HCM, participation in competitive sports is reasonable. |
| 2a |  Vigorous recreational activities are reasonable for patients with HCM accompanied by annual evaluations. |
| 2b |  Competitive sports may be considered after annual comprehensive evaluation and shared decision-making that includes an expert in HCM and sports cardiology. |
| 3: No Benefit |  Universal restriction from vigorous physical activity or competitive sports is not indicated |
| 3: Harm |  ICD placement for the sole purpose of participation in competitive athletics should not be performed. |

Abbreviations: HCM indicates hypertrophic cardiomyopathy; ICD, implantable cardioverter defibrillator; and SCD, sudden cardiac death.






Occupation Recommendations in HCM






| COR | RECOMMENDATIONS |
|-----|---|
| 2a |  Follow the Federal Motor Carrier Safety Guidelines for those without ICD or major risk factors for SCD and are using a GDMT plan. |
| 2a |  For pilots, follow Federal Aviation Administration guidelines for multicrew flying duties if they are asymptomatic, low risk for SCD and complete a treadmill stress test at 85% of peak heart rate. |
| 2b |  Occupations that require manual labor, heavy lifting, or a high level of physical performance may be reasonably considered after annual comprehensive evaluation, SCD risk assessment, and GDMT in the context of shared decision-making. |

Abbreviations: GDMT indicates guideline-directed management and therapy; HCM, hypertrophic cardiomyopathy; ICD, implantable cardioverter defibrillator; and SCD, sudden cardiac death.

Ommen, S.R. et al, 2024 AHA/ACC/AMSSM/HRS/PACES/SCMR Guideline for the Management of Hypertrophic Cardiomyopathy. *Circulation*.

Pregnancy in HCM

| COR | RECOMMENDATIONS |
|-----|--|
| 1 |  In high risk HCM, consultation with a maternal-fetal medicine expert is recommended. |
| 1 |  In families affected by HCM, preconception and prenatal reproductive and genetic counseling recommended. |
| 1 |  In patients with HCM and AF low molecular weight heparin or low dose warfarin are recommended. |
| 1 |  Beta-blocker for symptoms of LVOT obstruction or arrhythmia, with monitoring of fetal growth. |
| 1 |  Vaginal delivery is the first-choice in HCM. |

| COR | RECOMMENDATIONS |
|------------|---|
| 2a |  In clinically stable HCM, it is reasonable to advise pregnancy is generally safe as part of shared discussion and initiation of GDMT. |
| 2a |  Reasonable to cardiovert new or recurrent atrial fibrillation, especially if symptomatic. |
| 2a |  Reasonable to use general or epidural anesthesia, with precautions to avoid hypotension. |
| 2a |  Reasonable to perform serial echocardiography in the second or third trimester, or if symptoms develop. |
| 3: Harm |  Use of mavacamten is contraindicated due to potential teratogenic effects. |

Abbreviations: GDMT indicates guideline-directed management and therapy; HCM, hypertrophic cardiomyopathy; and LVOT, left ventricular outflow tract.

Unmet Needs and Future Directions

Expanding access to genetic counseling and testing, as well as improving interpretation of results (particularly pertaining to variants of unknown significance)



Refining criteria for HCM to improve diagnostic accuracy and facilitate potential targeted therapies



Improving management and risk stratification of arrhythmias in patients with HCM



Developing safe and effective therapies that attenuate and prevent disease progression



Incorporating new risk factors and tools to improve screening, risk stratification, and disease monitoring



Advancing care for patients with non-obstructive HCM



Abbreviations: HCM indicates hypertrophic cardiomyopathy

Acknowledgments

Many thanks to our Guideline Ambassadors who were guided by Dr. Elliott Antman in developing this guideline derivative product in support of the 2024 AHA/ACC/AMSSM/HRS/PACES Guideline for the Management of Hypertrophic Cardiomyopathy.

Dr. Ijeoma Eleazu
Dr. Ahmed Kazem
Dr. Dennis Narcisse

Dr. Jessica Regan
Dr. Raymond Yeow

The American Heart Association requests this electronic slide deck be cited as follows:

Eleazu, I., Kazem, A., Narcisse, D., Regan, J., Yeow, R., Reyna, G., Bezanson, J. L., & Antman, E. M. (2024). AHA Clinical Update; Adapted from: [PowerPoint slides]. Retrieved from the 2024 ACC/AHA/AMSSM/HRS/PACES Guideline for the Management of Hypertrophic Cardiomyopathy. <https://professional.heart.org/en/science-news>.