

# Clinical Update

---

Adapted from: 2020 ACC/AHA Guideline for  
the Diagnosis and Treatment of Patients  
with Hypertrophic Cardiomyopathy



American  
Heart  
Association.



# ACC/AHA 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	
<b>CLASS 1 (STRONG)</b>	<b>Benefit &gt;&gt;&gt; Risk</b>
<b>Suggested phrases for writing recommendations:</b> <ul style="list-style-type: none"> <li>• Is recommended</li> <li>• Is indicated/useful/effective/beneficial</li> <li>• Should be performed/administered/other</li> <li>• Comparative-Effectiveness Phrases†:               <ul style="list-style-type: none"> <li>– Treatment/strategy A is recommended/indicated in preference to treatment B</li> <li>– Treatment A should be chosen over treatment B</li> </ul> </li> </ul>	
<b>CLASS 2a (MODERATE)</b>	<b>Benefit &gt;&gt; Risk</b>
<b>Suggested phrases for writing recommendations:</b> <ul style="list-style-type: none"> <li>• Is reasonable</li> <li>• Can be useful/effective/beneficial</li> <li>• Comparative-Effectiveness Phrases†:               <ul style="list-style-type: none"> <li>– Treatment/strategy A is probably recommended/indicated in preference to treatment B</li> <li>– It is reasonable to choose treatment A over treatment B</li> </ul> </li> </ul>	
<b>CLASS 2b (WEAK)</b>	<b>Benefit ≥ Risk</b>
<b>Suggested phrases for writing recommendations:</b> <ul style="list-style-type: none"> <li>• May/might be reasonable</li> <li>• May/might be considered</li> <li>• Usefulness/effectiveness is unknown/unclear/uncertain or not well-established</li> </ul>	
<b>CLASS 3: No Benefit (MODERATE)</b> (Generally, LOE A or B use only)	<b>Benefit = Risk</b>
<b>Suggested phrases for writing recommendations:</b> <ul style="list-style-type: none"> <li>• Is not recommended</li> <li>• Is not indicated/useful/effective/beneficial</li> <li>• Should not be performed/administered/other</li> </ul>	
<b>Class 3: Harm (STRONG)</b>	<b>Risk &gt; Benefit</b>
<b>Suggested phrases for writing recommendations:</b> <ul style="list-style-type: none"> <li>• Potentially harmful</li> <li>• Causes harm</li> <li>• Associated with excess morbidity/mortality</li> <li>• Should not be performed/administered/other</li> </ul>	

LEVEL (QUALITY) OF EVIDENCE‡	
<b>LEVEL A</b>	<ul style="list-style-type: none"> <li>• High-quality evidence‡ from more than 1 RCT</li> <li>• Meta-analyses of high-quality RCTs</li> <li>• One or more RCTs corroborated by high-quality registry studies</li> </ul>
<b>LEVEL B-R</b>	<b>(Randomized)</b>
<ul style="list-style-type: none"> <li>• Moderate-quality evidence‡ from 1 or more RCTs</li> <li>• Meta-analyses of moderate-quality RCTs</li> </ul>	
<b>LEVEL B-NR</b>	<b>(Nonrandomized)</b>
<ul style="list-style-type: none"> <li>• Moderate-quality evidence‡ from 1 or more well-designed, well-executed nonrandomized studies, observational studies, or registry studies</li> <li>• Meta-analyses of such studies</li> </ul>	
<b>LEVEL C-LD</b>	<b>(Limited Data)</b>
<ul style="list-style-type: none"> <li>• Randomized or nonrandomized observational or registry studies with limitations of design or execution</li> <li>• Meta-analyses of such studies</li> <li>• Physiological or mechanistic studies in human subjects</li> </ul>	
<b>LEVEL C-EO</b>	<b>(Expert Opinion)</b>
<ul style="list-style-type: none"> <li>• Consensus of expert opinion based on clinical experience</li> </ul>	

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).

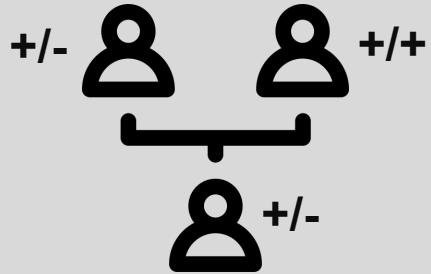
† For comparative-effectiveness recommendations (COR 1 and 2a; LOE A and B only), studies that support the use of comparator verbs should involve direct comparisons of the treatments or strategies being evaluated.

‡ 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.

# Hypertrophic Cardiomyopathy (HCM) is a Globally Prevalent & Common Genetic Heart Disease

## Inheritance Pattern



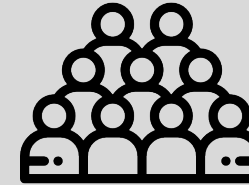
**Autosomal Dominant**

## Sex Distribution



**Women diagnosed less commonly**

## Disease Prevalence

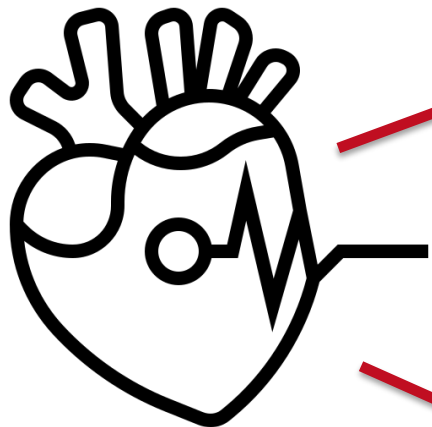


**Estimated  
1:200 – 1:500**

## Triggers for Evaluation



**Symptoms**  
Cardiac Event  
Heart Murmur  
Abnormal EKG  
Cardiac Imaging  
Family Studies



**HCM**

**$\frac{2}{3}$  have LVOTO**

**LV Outflow Tract  
Obstruction  
(LVOTO)**

**$\frac{1}{3}$  do not have LVOTO**

## Other non-HCM Causes of LV Hypertrophy

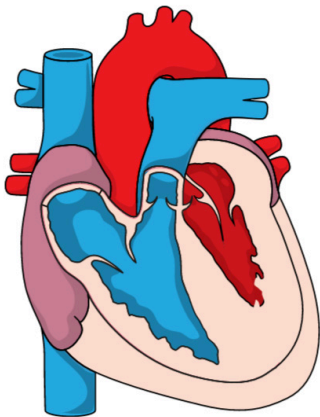
### Metabolic & Multi-organ Syndromes

RASopathies  
Mitochondrial myopathies  
Glycogen / Lysosomal storage diseases  
Amyloidosis  
Sarcoidosis  
Hemochromatosis  
Danon disease

### Secondary Causes

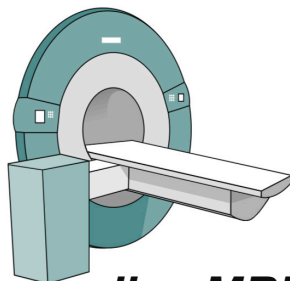
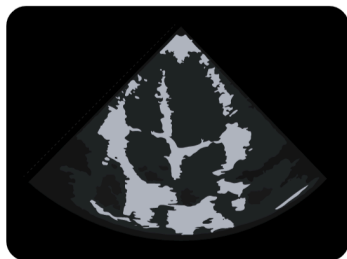
Athlete's heart  
Hypertension  
Valvular & subvalvular stenosis

## Defining Hypertrophic Cardiomyopathy in 2020



- Morphologic expression confined solely to the heart
- Characterized by left ventricular (LV) hypertrophy  
*Basal anterior septum in continuity with the anterior free wall = most common*
- No other cardiac, systemic or metabolic disease capable of producing the magnitude of hypertrophy present
- Disease-causing sarcomere (or sarcomere-related) variant identified or genetic etiology unresolved

### Diagnostic Criteria in Adults



#### 2D echocardiography or cardiac MRI

**Maximal end-diastolic LV wall thickness  $\geq 15$  mm**

**Maximal end-diastolic LV wall thickness 13-14 mm in family member of HCM pt. or in conjunction with positive genetic test**

### Other Nondiagnostic Morphologic Abnormalities Associated with HCM

Systolic anterior motion (SAM) of the mitral valve

Hyperdynamic LV function

Hypertrophied & apically displaced papillary muscles

Myocardial crypts

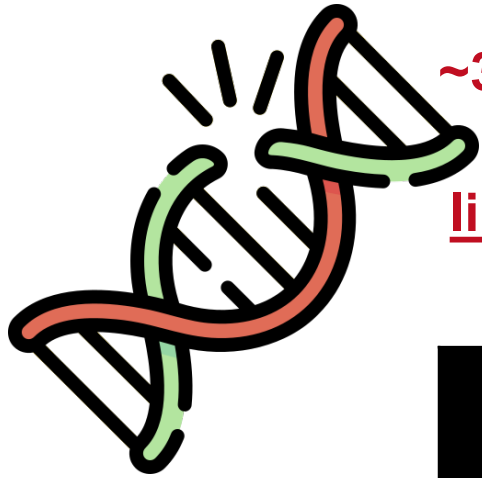
Anomalous papillary muscle insertion in anterior MV leaflet

Elongated mitral valve leaflets

Myocardial bridging

Right ventricular hypertrophy

# Genetic Etiology of Hypertrophic Cardiomyopathy (HCM)



~30-60% of HCM patients have an identifiable pathogenic or likely-pathogenic genetic variant



Many others have no genetic evidence of disease and / or no other affected family members

<u>Sarcomere Genes Implicated in HCM</u>
<i>MYH7</i>
<i>MYBPC3</i>
<i>TNNI3</i>
<i>TNNT2</i>
<i>TPM1</i>
<i>MYL2</i>
<i>MYL3</i>
<i>ACTC1</i>



**Two most common genes that harbor pathogenic variants in HCM (70%)**

# Adverse Event Associated Hypertrophic Cardiomyopathy

Majority of patients with HCM have a normal life expectancy without limiting symptoms or the need for major treatments



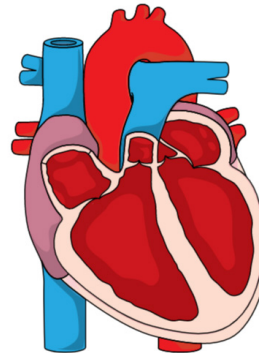
Sudden Death



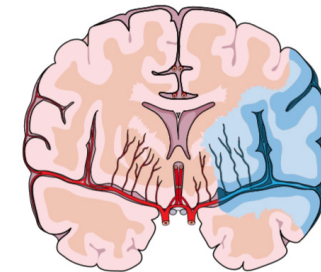
Progressive Functional Limitation



Atrial Fibrillation



Heart Failure

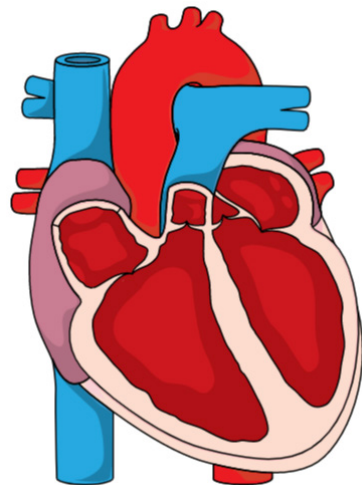


Thromboembolism

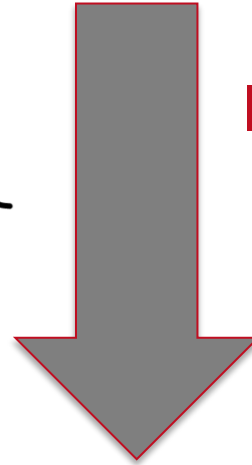
# Hypertrophic Cardiomyopathy Mortality Rates Now < 1% per Year



**Sudden Death**

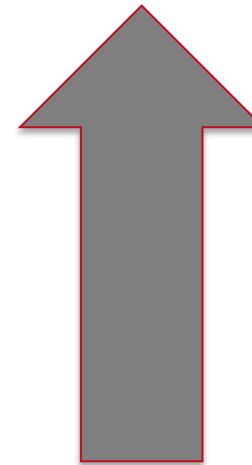


**Heart Failure**



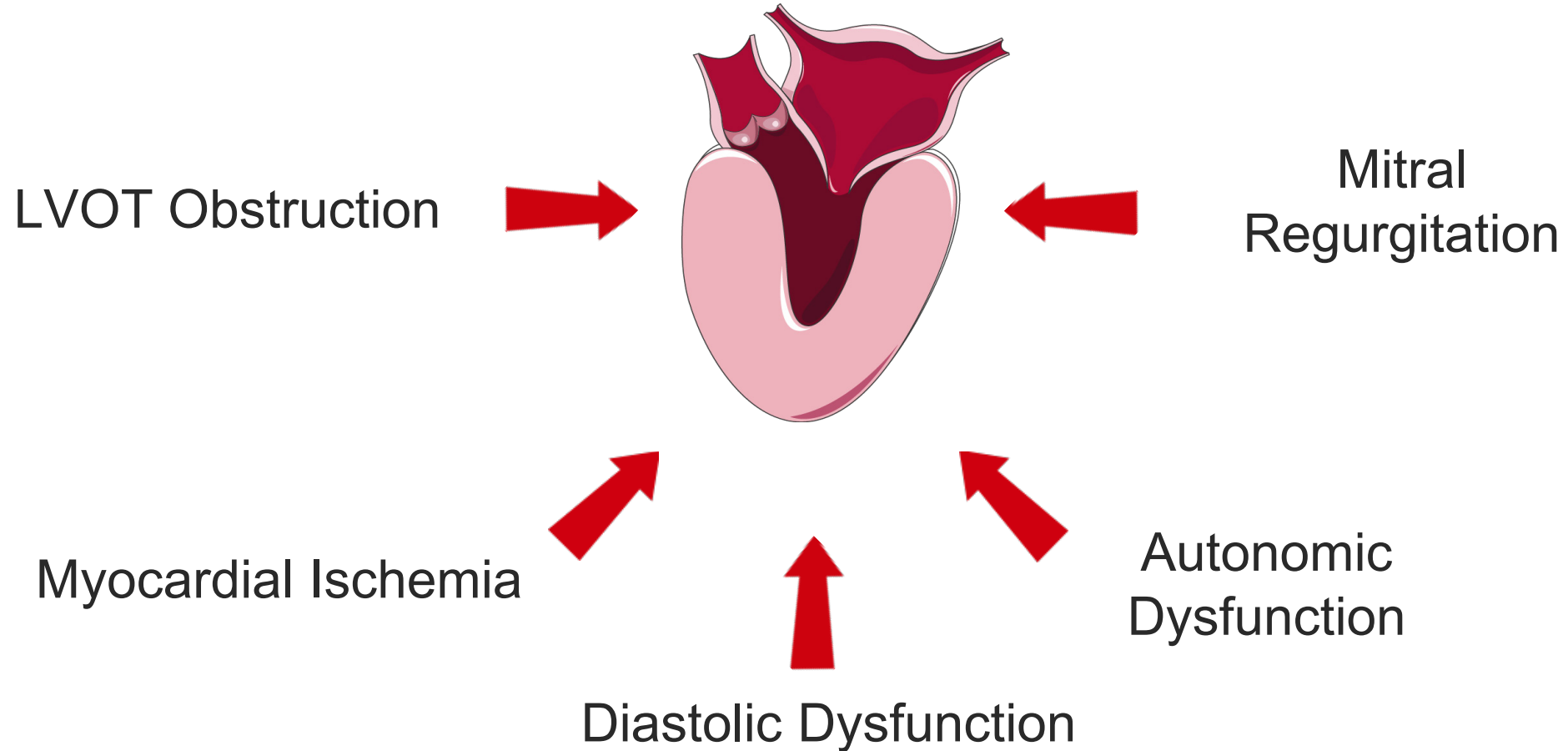
**Improvements in Risk Stratification**

**ICD Implantation**



***Now the greatest unmet treatment need in adults***

# Pathophysiologic Myocardial Changes in Hypertrophic Cardiomyopathy



Abbreviations: LVOT indicates left ventricular outflow tract.

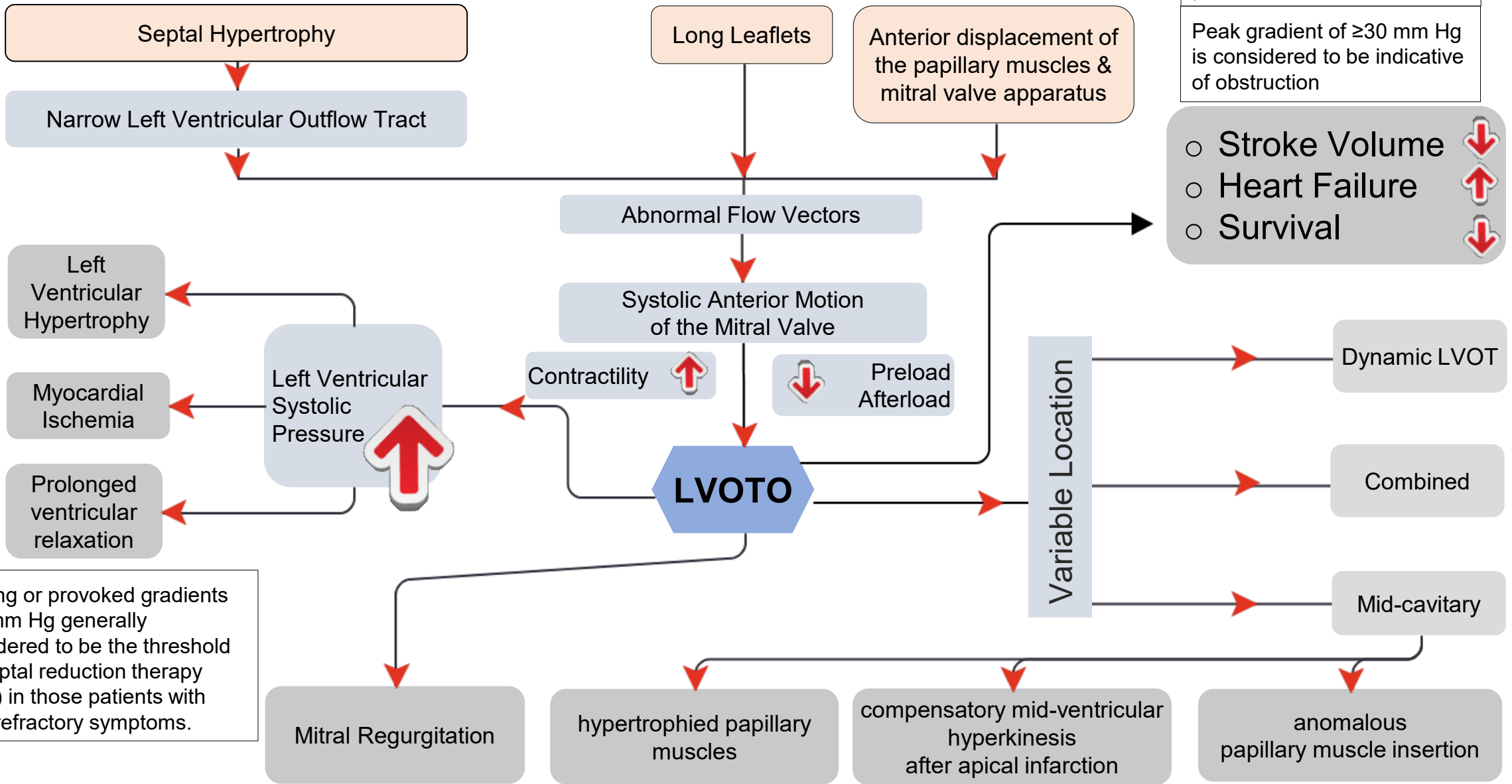
Ommen, SR et al. 2020 ACC/AHA Guideline for the Diagnosis and Treatment of Patients with Hypertrophic Cardiomyopathy. *Circulation*. XXX:XX-XX.



# Left Ventricular Outflow Tract Obstruction (LVOTO)

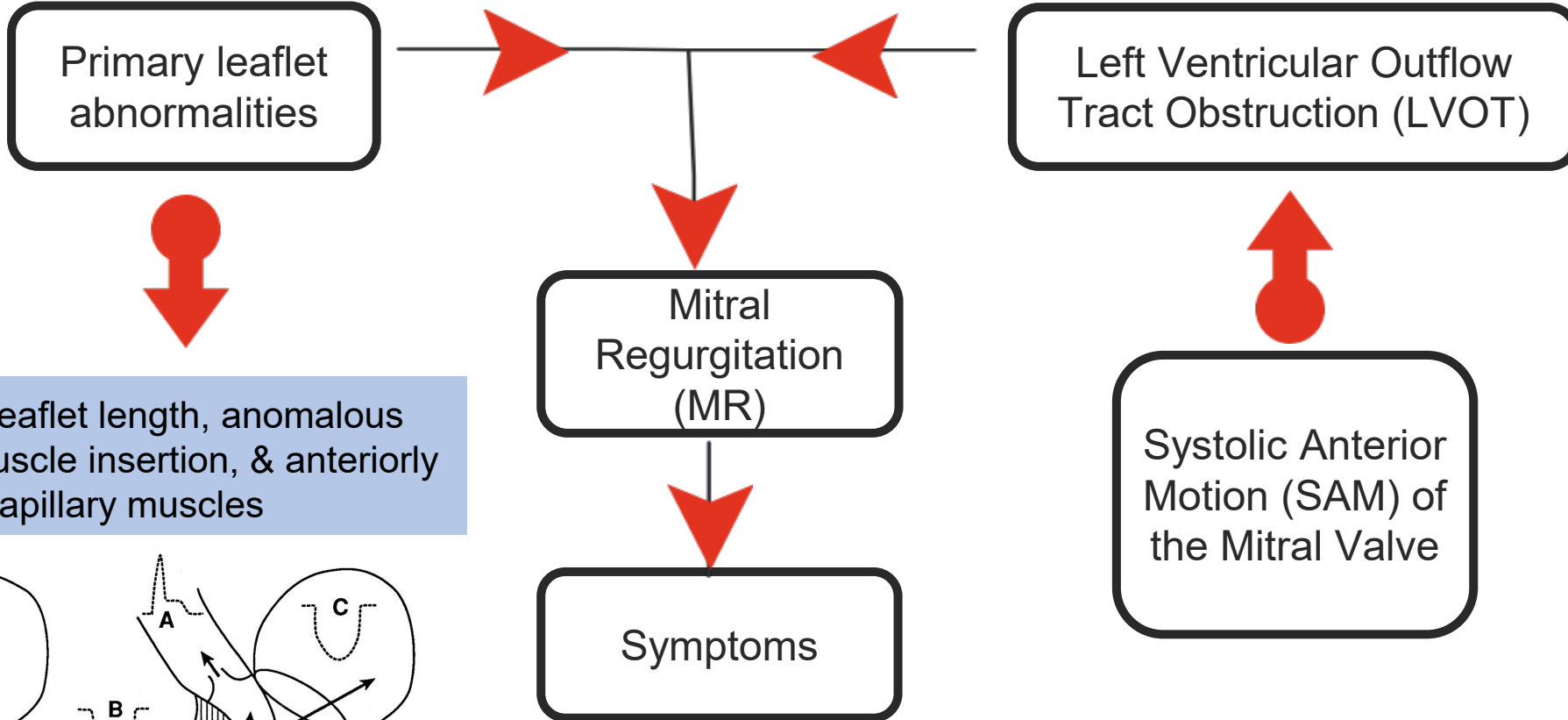
LVOTO, either at rest or with provocation, is present in approximately 75% (2/3) of patients with HCM

Peak gradient of  $\geq 30$  mm Hg is considered to be indicative of obstruction

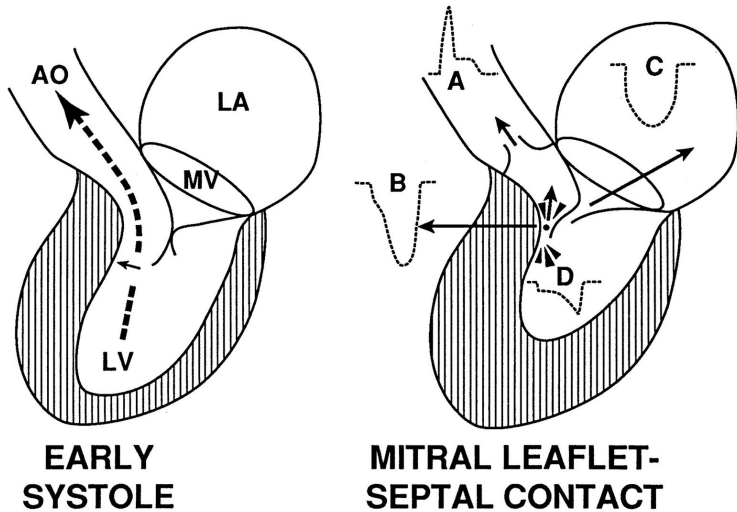


Resting or provoked gradients  $\geq 50$  mm Hg generally considered to be the threshold for septal reduction therapy (SRT) in those patients with drug refractory symptoms.

# A Closer Look at Left Ventricular Outflow Tract Obstruction (LVOTO)

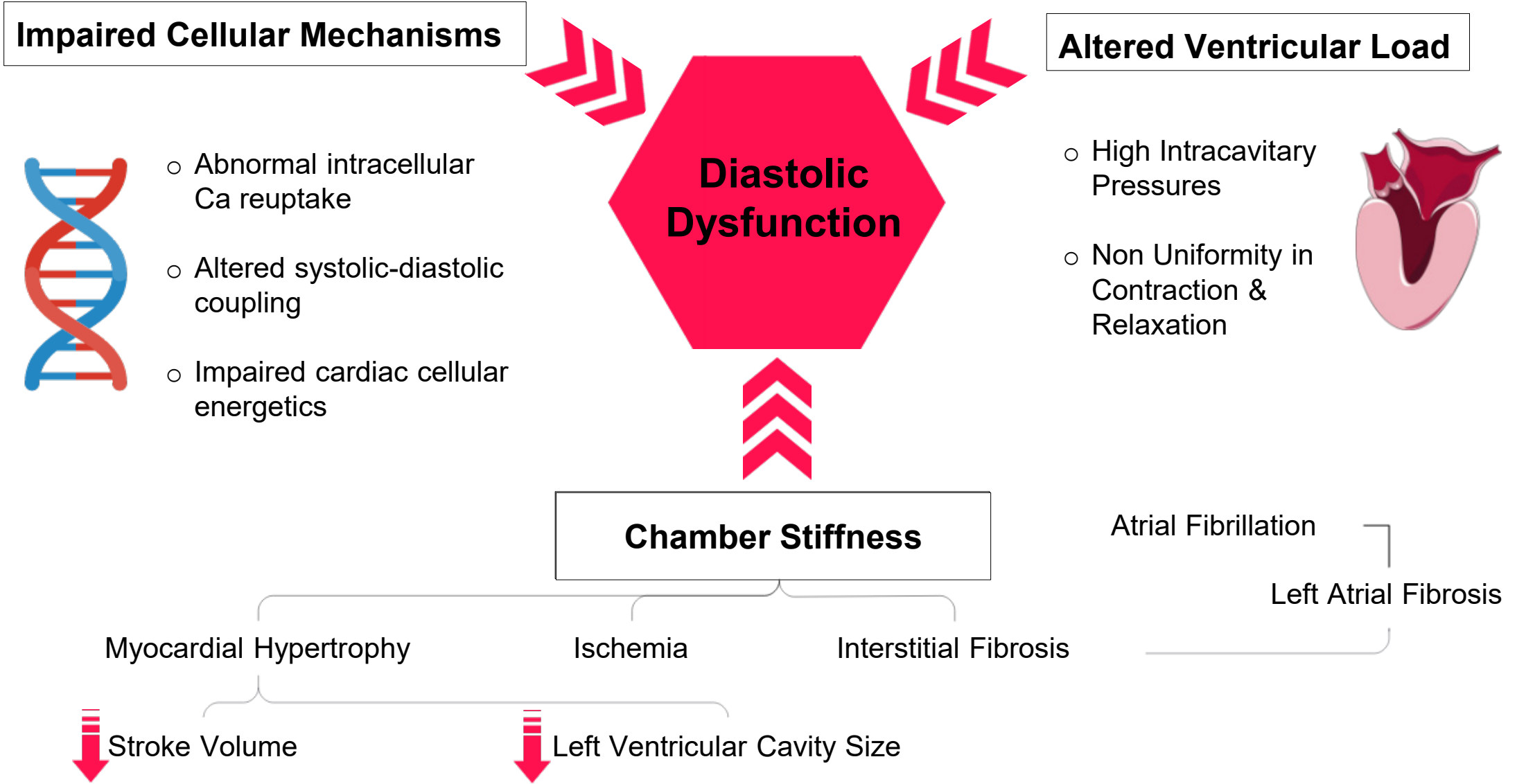


Excessive leaflet length, anomalous papillary muscle insertion, & anteriorly displaced papillary muscles

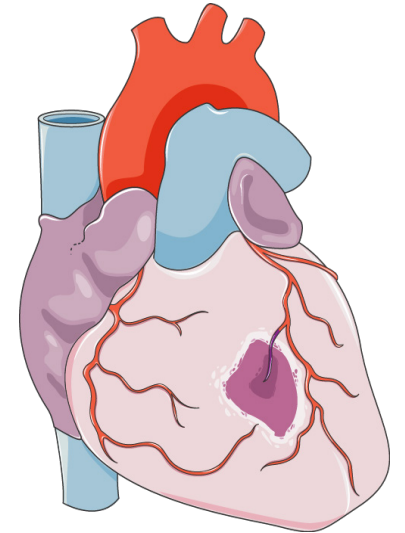
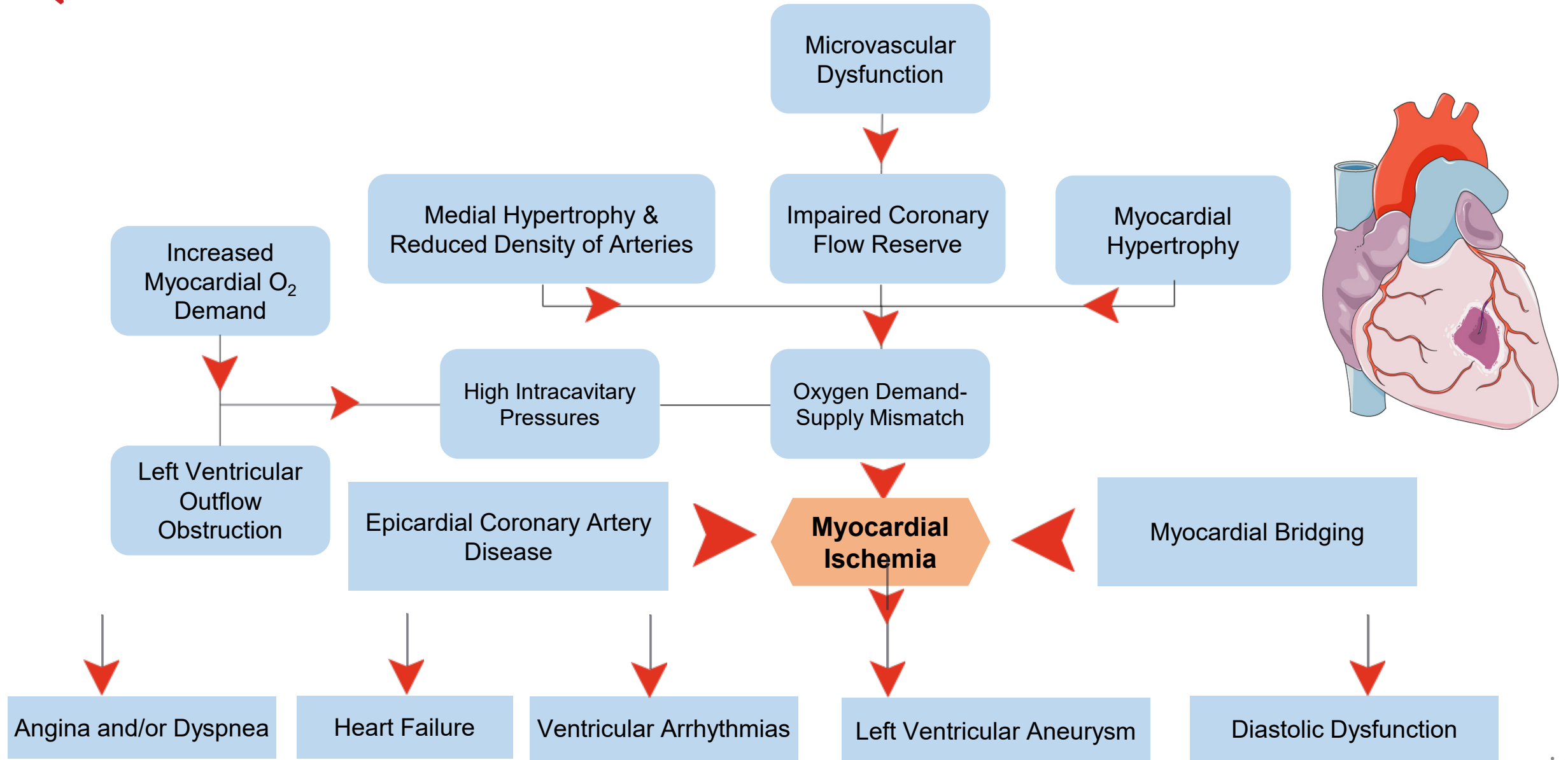


- In MR caused by LVOTO, SAM of the mitral valve leads to loss of leaflet coaptation, and the jet is predominantly mid-to-late systolic and posterior or lateral in orientation. However, central and anterior jets may also result from SAM of the mitral valve
- Factors that affect the severity of LVOTO also may affect the degree of MR. Thus, significant MR may not be evident without provocation for LVOTO and SAM of the mitral valve.

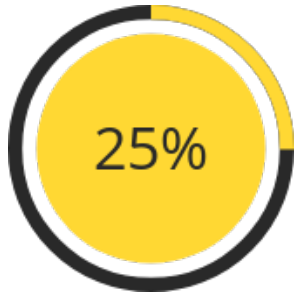
# Pathophysiology of Diastolic Dysfunction in HCM



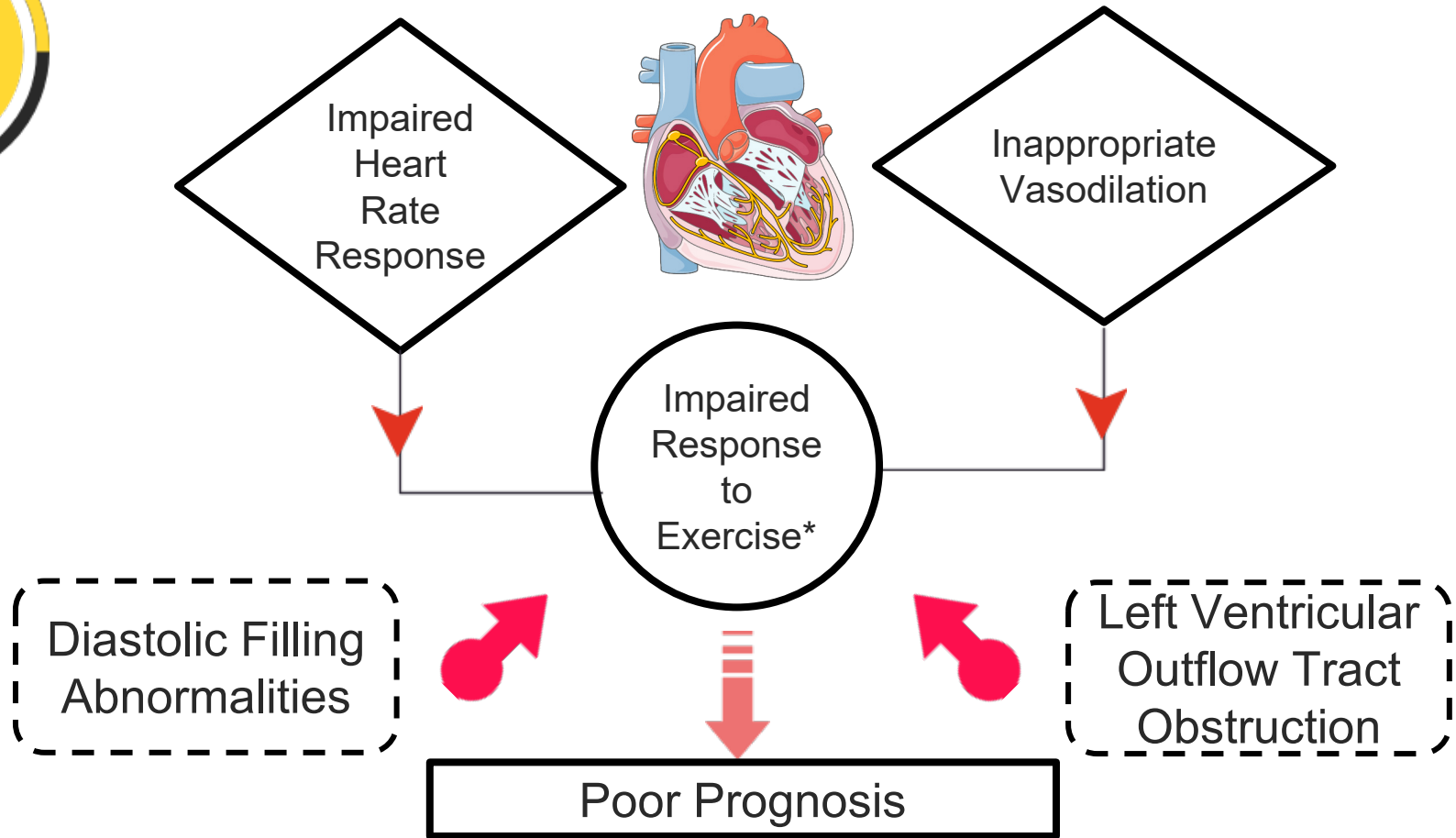
# Pathophysiologic Mechanisms of Myocardial Ischemia in HCM



# Autonomic Dysfunction in Hypertrophic Cardiomyopathy



The prevalence of autonomic dysfunction in HCM is uncertain, although studies have described an abnormal blood pressure response to exercise in 25% of patients (1-3)



\* Failure to increase systolic blood pressure by at least 20 mm Hg, or a drop in systolic blood pressure during exercise of >20 mm Hg from the peak value obtained

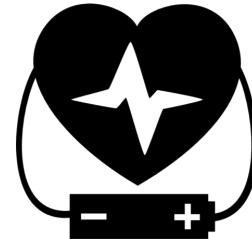
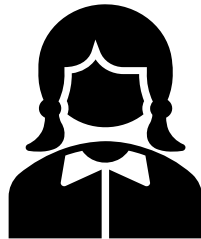
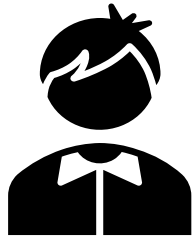
1. Maron BJ, Wolfson JK, Epstein SE, et al. Intramural ("small vessel") coronary artery disease in hypertrophic cardiomyopathy. *J Am Coll Cardiol.* 1986;8:545– 57.
2. Karamitsos TD, Dass S, Suttie J, et al. Blunted myocardial oxygenation response during vasodilator stress in patients with hypertrophic cardiomyopathy. *J Am Coll Cardiol.* 2013;61:1169–76.
3. Raphael CE, Cooper R, Parker KH, et al. Mechanisms of myocardial ischemia in hypertrophic cardiomyopathy: insights from wave intensity analysis and magnetic resonance. *J Am Coll Cardiol.* 2016 Oct 11;68(15):1651-1660.

# Recommendations for Shared Decision-Making in HCM



## Discussions should involve:

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



## Shared decision discussions should be applied to:

- Genetic testing
- Sudden death risk assessment and ICD implantation
- Participation in high-intensity exercise and competitive sports
- Medical and invasive therapies for LVOT obstruction



# Teams Based Approach to Hypertrophic Cardiomyopathy Care



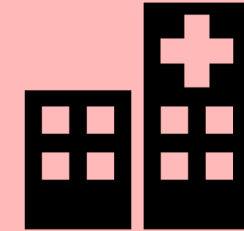
## Cardiologists Working Outside of HCM Centers:

- 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 and Procedures



## Comprehensive HCM Centers:

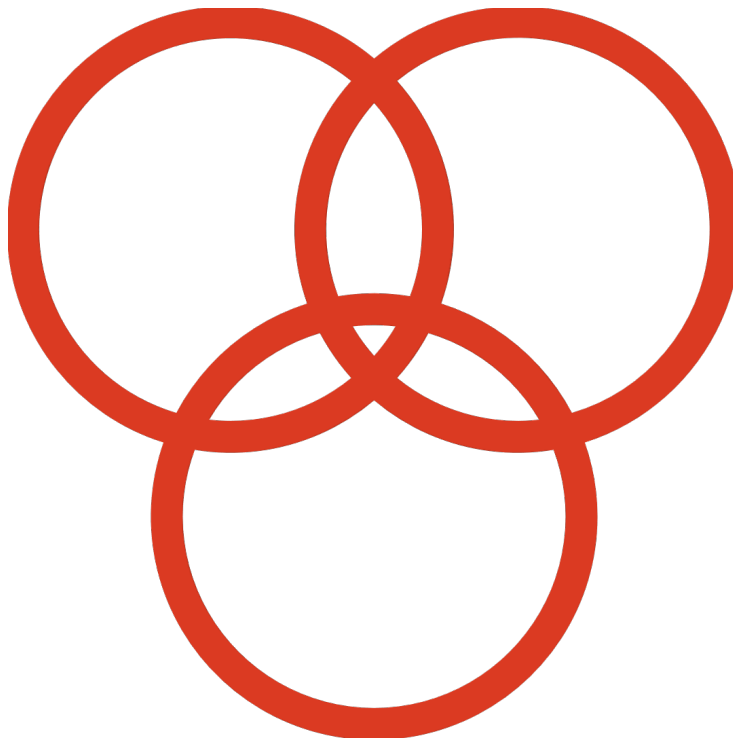
- Complex Invasive Septal Reduction Therapies
- Catheter Ablation for Ventricular and Complex Atrial Tachyarrhythmias
- Advanced Heart Failure Therapies

# Role of the Cardiologist



## Cardiologists Outside of HCM Centers:

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



## Comprehensive HCM Centers:

- Complex Invasive Septal Reduction Therapies
- Catheter Ablation for Ventricular and Complex Atrial Tachyarrhythmias
- Advanced Heart Failure Therapies

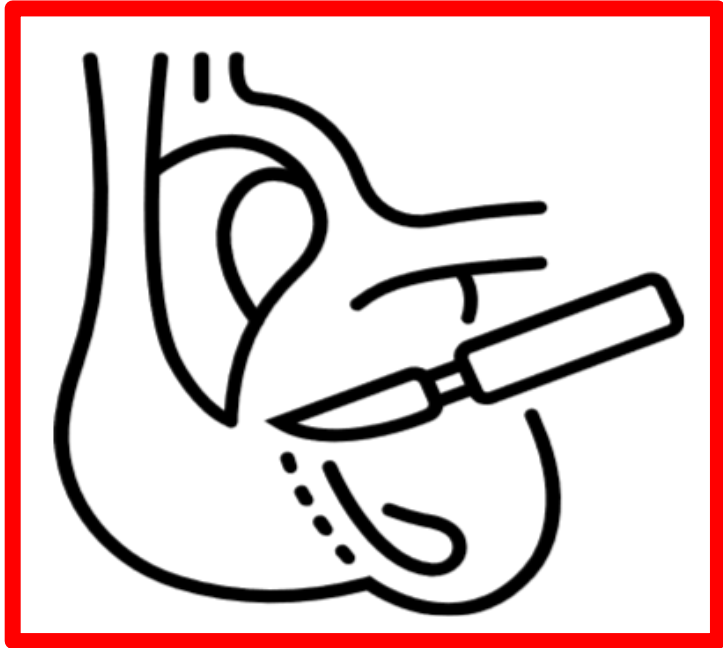


## HCM Centers:

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



## Recommendations for Septal Reduction Therapy



- Invasive septal reduction therapy is associated with increased morbidity and mortality at low volume centers defined as centers with the lowest tertiles of hospital volumes
- Referral to a high volume HCM Center should be strongly considered for invasive septal reduction therapy
- Centers performing invasive septal reduction therapies should aim for outcomes similar to comprehensive HCM Centers

# Clinical Features in Patients with “HCM Mimics”

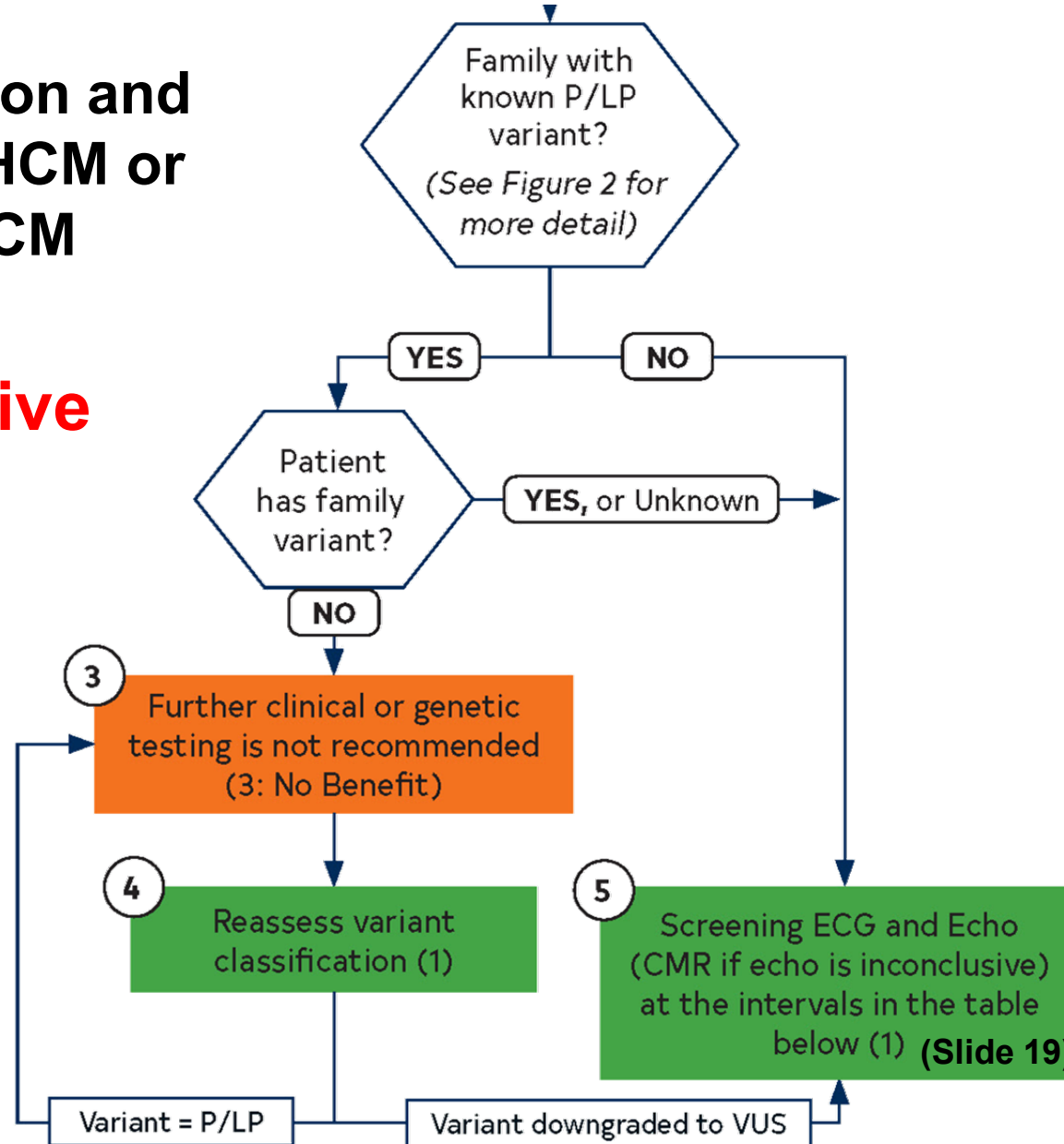
Life Stage	Systemic Features	Possible Etiology	Diagnostic Approach
Infants (0-12 months) and toddlers	Dysmorphic features, failure to thrive, metabolic acidosis	<ul style="list-style-type: none"> <li>• RASopathies</li> <li>• Glycogen storage diseases, other metabolic or mitochondrial diseases</li> <li>•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

Abbreviations: RAS indicates reticular activating system.

Ommen, SR et al. 2020 ACC/AHA Guideline for the Diagnosis and Treatment of Patients with Hypertrophic Cardiomyopathy. *Circulation*. XXX:XX-XX.

# Recommended Evaluation and Testing for Suspected HCM or Family History of HCM

## Phenotype Negative



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.

## (1) Slide 19 - Screening with Electrocardiography and 2D Echocardiography Recommendations in Asymptomatic Family Members\*

Age of First-Degree Relative	Initiation of Screening	Repeat ECG, Echo
<b>Pediatric</b>		
Children and adolescents from genotype-positive families, and families with early onset disease	At the time HCM is diagnosed in another family member	Every 1-2 y
All other pediatric	At any time after HCM is diagnosed in a family member but no later than puberty	Every 2-3 y
<b>Adults</b>	At the time HCM is diagnosed in another family member	Every 3-5 y

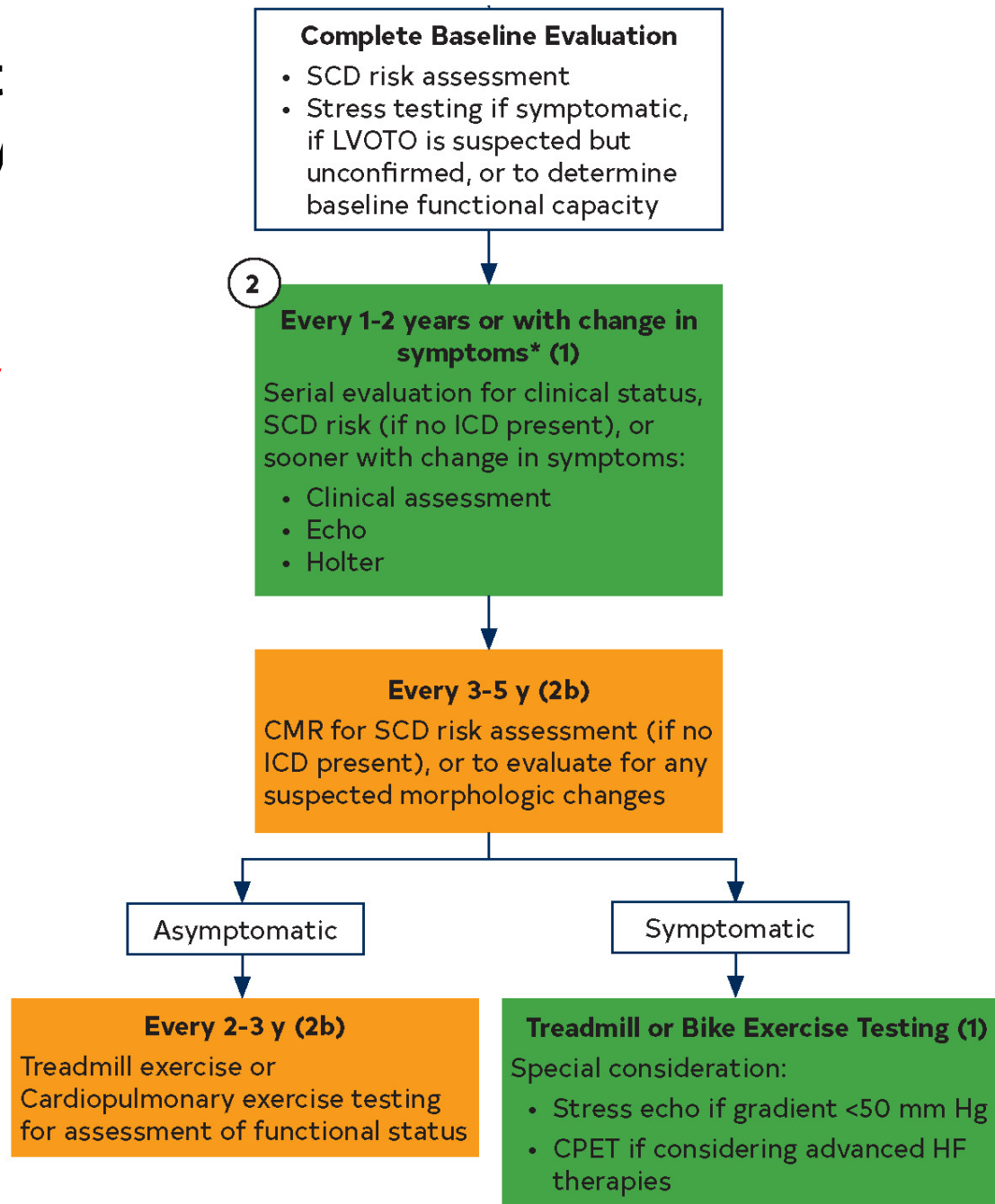
\*Includes all asymptomatic, phenotype-negative first-degree relatives deemed to be at-risk for developing HCM based on family history or genotype status and may sometimes include more distant relatives based on clinical judgment. Screening interval may be modified (e.g., at onset of new symptoms or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

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

Ommen, SR et al. 2020 ACC/AHA Guideline for the Diagnosis and Treatment of Patients with Hypertrophic Cardiomyopathy. *Circulation*. XXX:XX-XX.

# Recommended Evaluation and Testing of Phenotype Positive 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.

# Echocardiography Recommendations in Hypertrophic Cardiomyopathy

COR	LOE	Recommendations
1	B-NR	1. In patients with suspected HCM, a TTE is recommended in the initial evaluation.
1	B-NR children	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 LVOTO, MR, and myocardial function.
1	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 LVOTO, MR, and myocardial function.
1	B-NR	3. For patients with HCM who experience a change in clinical status or a new clinical event, repeat TTE is recommended.
1	B-NR	4. For patients with HCM and resting LVOT gradient <50 mm Hg, a TTE with provocative maneuvers is recommended.

Abbreviations: COR indicates classification of recommendation; LOE, level of evidence; B-NR, Level B nonrandomized; C-LD, Level C, limited data; TTE, transthoracic echocardiogram; LVOTO, left ventricular outflow tract obstruction; MR, mitral regurgitation; LVOT, left ventricular outflow tract.

Ommen, SR et al. 2020 ACC/AHA Guideline for the Diagnosis and Treatment of Patients with Hypertrophic Cardiomyopathy. *Circulation*. XXX:XX-XX.

## Cardiovascular Magnetic Resonance (CMR) Imaging Recommendations in HCM

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
1	B-NR	2. For patients with LVH in whom there is a suspicion of alternative diagnoses, including infiltrative or storage disease as well as athlete's heart, CMR imaging is useful.
1	B-NR	3. For patients with HCM who are not otherwise identified as high risk for SCD, or in whom a decision to proceed with ICD 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, ejection fraction (EF), LV apical aneurysm, and extent of myocardial fibrosis with LGE.
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.

Abbreviations: SCD indicates sudden cardiac death; ICD, implanted cardioverter-defibrillator; LV, left ventricular; LGE, late gadolinium enhancement; SRT, septal reduction therapy; COR, classification of recommendation; LOE, level of evidence; B-NR, Level B nonrandomized.

# Risk Assessment of Sudden Cardiac Death (SCD) in HCM



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



Assess the following (Class I):

- ✓ 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,  $EF \leq 50\%$ , apical aneurysm
- ✓ NSVT episodes on continuous ambulatory electrocardiographic monitoring; In select adult patients without major SCD risk factors, ICD may be considered in NSVT present on ambulatory monitoring (Class IIb).

**IF** none of the above:



CMR to help decision regarding ICD (Class I)

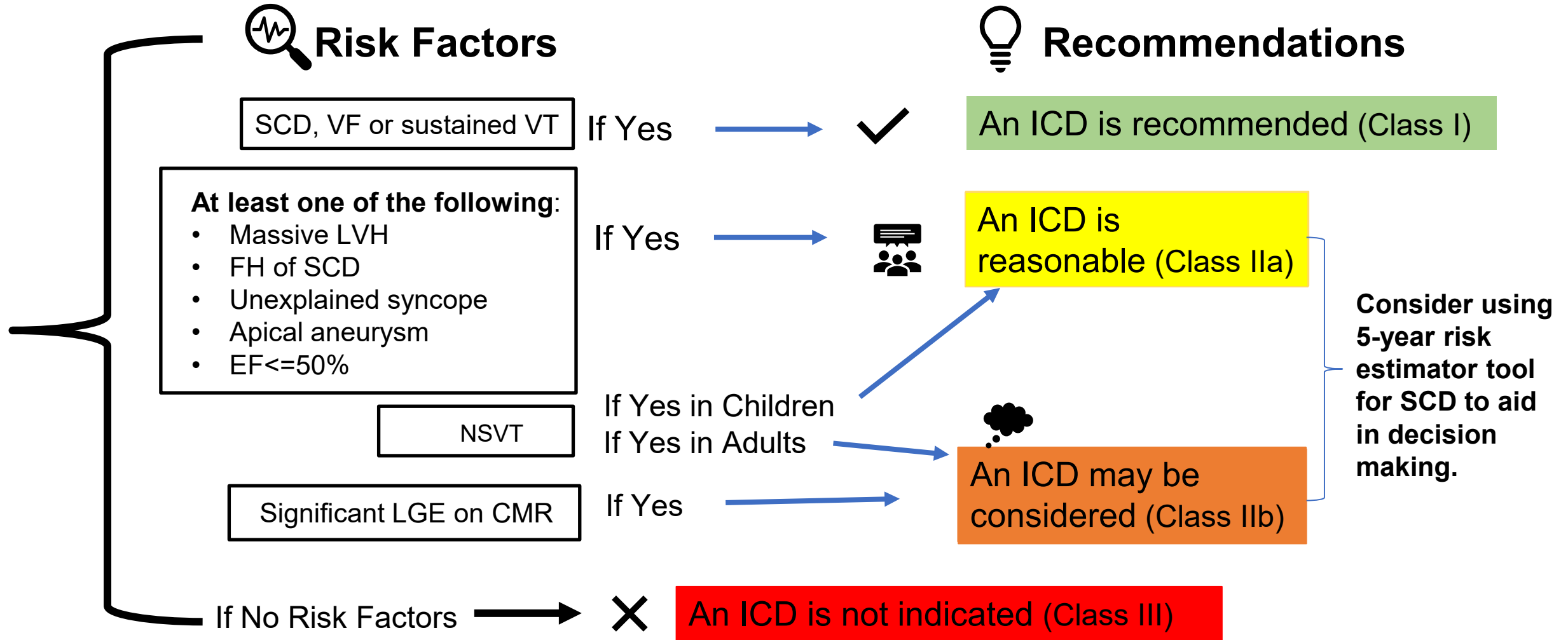


Reasonable to obtain echocardiographic LA diameter and LVOT gradient (Class IIa)

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.



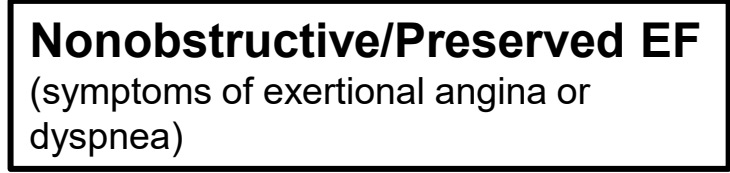
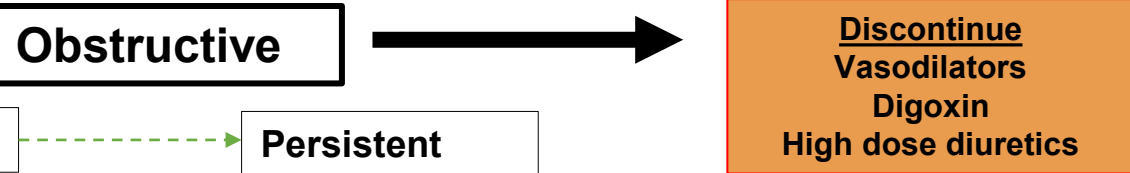
# Indications for ICD in HCM Patients



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; CMR, cardiac magnetic resonance imaging.

Ommen, SR et al. 2020 ACC/AHA Guideline for the Diagnosis and Treatment of Patients with Hypertrophic Cardiomyopathy. *Circulation*. XXX:XX-XX.

# Pharmacologic Management Based on Type of HCM



Symptoms r/t LVOTO

Persistent dyspnea with volume overload

Acute Hypotension

ARBs/ACEi in symptomatic patients is not well established.

non-vasodilating  $\beta$  blockers

Cautious use of low-dose oral diuretics

IV Fluids

Therefore treatment includes:  
 $\beta$  blockers or CCBs\*

Verapamil is potentially harmful in severe dyspnea at rest, very high gradients and in children < 6 weeks

If no response:  
Phenylephrine  $\pm$   $\beta$  blockers

If dyspnea continues:  
Diuresis

If not effective:  
CCBs \*

If persistent severe symptoms:

Add Disopyramide

OR

Septal reduction therapy in eligible patients

In persistent NYHA class III/VI + apical HCM  
Treat with GDMT for HFpEF  
Apical myectomy‡



The following is not well established:  
 $\beta$  blockers or CCBs if asymptomatic

\*non-dihydropyridine calcium channel blockers (CCBs)  
‡LV end-diastolic volume <50 mL/m<sup>2</sup> and LV stroke volume <30 mL/m<sup>2</sup>

Abbreviations: CCBs indicates calcium channel blockers; LVOTO, left ventricular outflow tract obstruction; IV, intravenous; EF, ejection fraction; ACEi, angiotensin-converting enzyme inhibitors; ARBs, angiotensin receptor blockers; GDMT: goal-directed medical therapy; HFpEF: heart failure with preserved ejection fraction

Ommen, SR et al. 2020 ACC/AHA Guideline for the Diagnosis and Treatment of Patients with Hypertrophic Cardiomyopathy. *Circulation*. XXX:XX-XX.

# Invasive Management of Obstructive HCM

**Obstructive HCM with NYHA class III/IV despite GDMT**

**SRT at an experienced center**

**Myectomy is recommended if associated conditions exist where surgical treatment is necessary (such as: associated anomalous papillary muscle, markedly elongated anterior mitral leaflet, intrinsic mitral valve disease, multivessel CAD, valvular aortic stenosis)**

**Alcohol septal ablation is recommended for eligible patients if surgery is contraindicated or the risk is considered unacceptable because of serious comorbidities or advanced age**

For severely symptomatic patients, SRT in eligible patients, performed at experienced centers, may be considered as an alternative to escalation of medical therapy after shared decision-making including risks and benefits of all treatment options

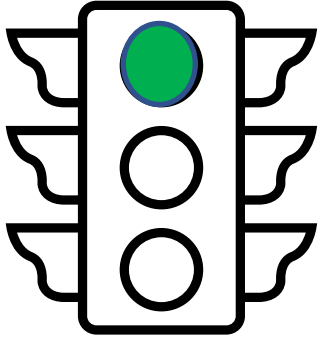
**For symptomatic patients in whom SRT is an option, mitral valve replacement should not be performed for the sole purpose of relief of LVOTO**

Myectomy is reasonable in patients with NYHA class II if:

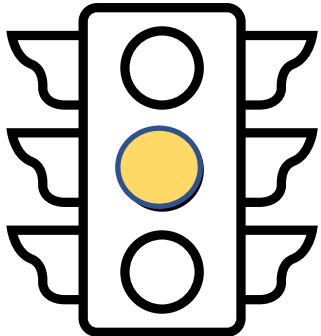
- a) Severe and progressive pulmonary hypertension thought to be attributable to LVOTO or associated MR
- b) Left atrial enlargement with  $\geq 1$  episodes of symptomatic AF
- c) Poor functional capacity attributable to LVOTO as documented on treadmill exercise testing
- d) Children and young adults with very high resting LVOT gradients ( $>100$  mm Hg)

Abbreviations: GDMT indicates guideline directed medical therapy; NYHA, New York Heart Association; SRT, septal reduction therapy; CAD, coronary artery disease; MR, mitral regurgitation; LVOTO, left ventricular outflow tract obstruction; AF, atrial fibrillation.

# Atrial Fibrillation (AF) in Hypertrophic Cardiomyopathy

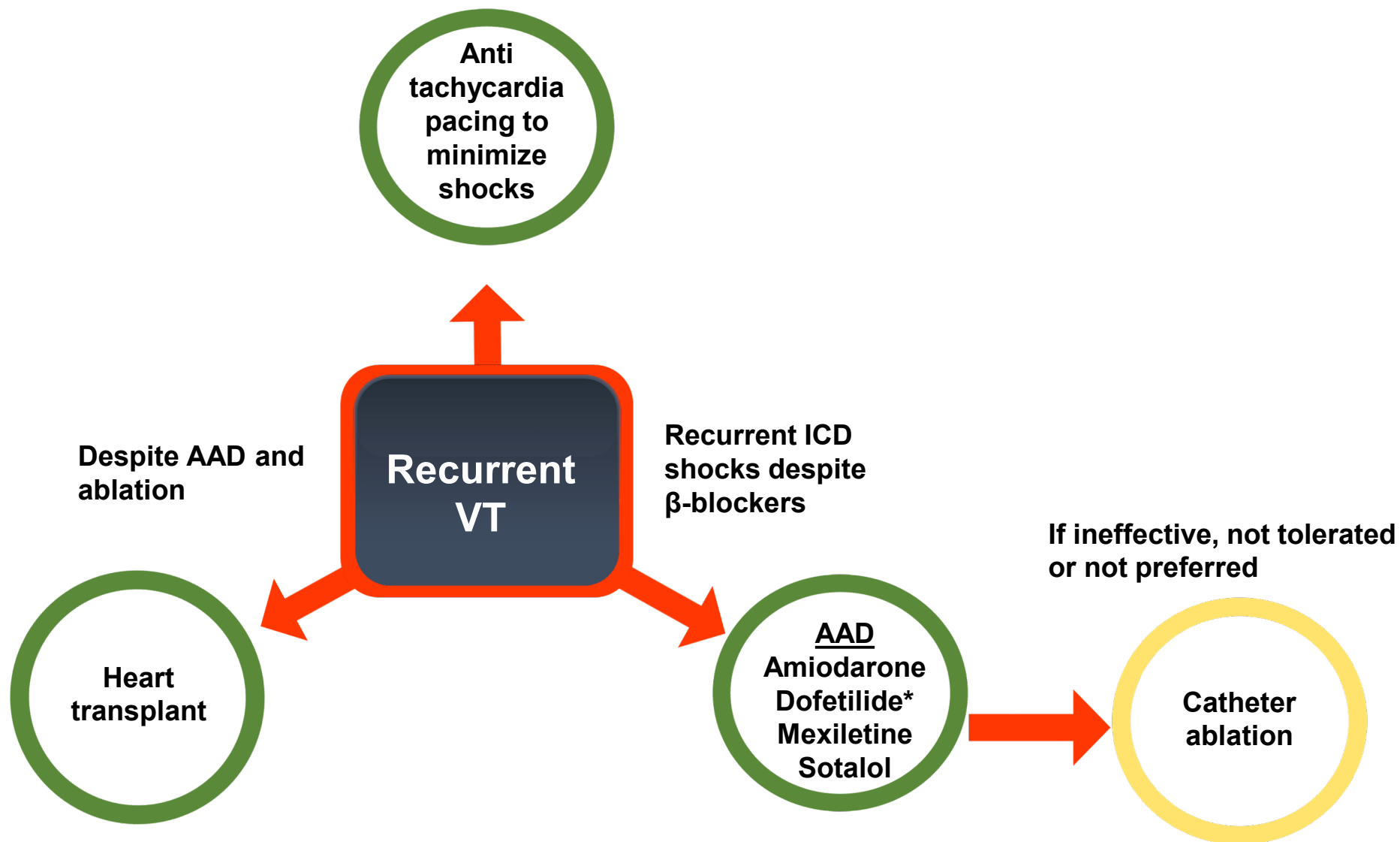


- ✓ In patients with clinical AF or subclinical AF (duration  $\geq$  24 hours) DOACs are first line
- ✓ Vitamin K antagonists are second line Independent of CHA<sub>2</sub>DS<sub>2</sub>-VASc score
- ✓ For rate control strategy use either beta blockers, verapamil or diltiazem.



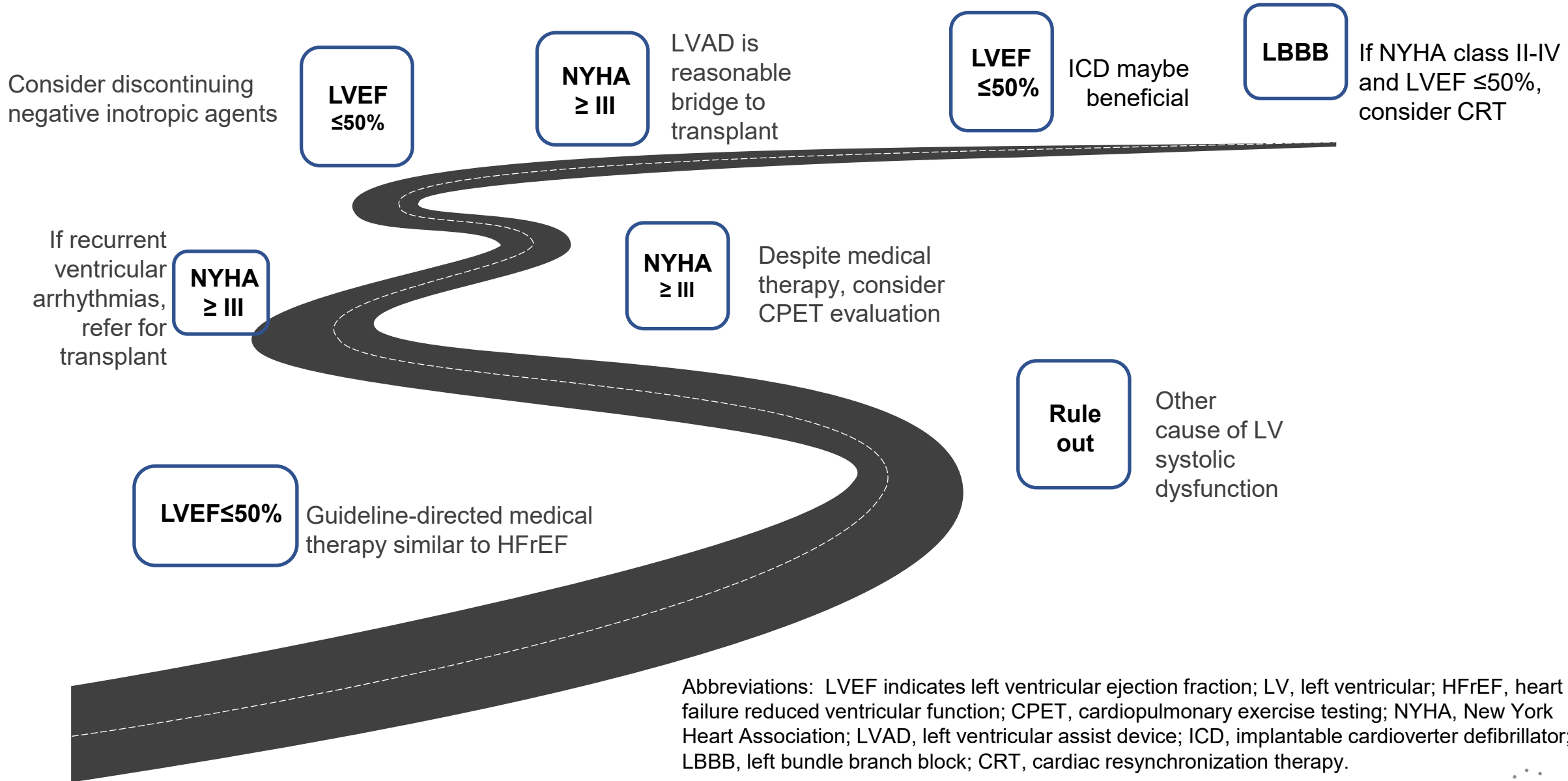
- ✓ In patients with poorly tolerated AF, a rhythm control strategy with cardioversion or anti-arrhythmic drugs can be beneficial
- ✓ AF catheter ablation can be effective when drug therapy is ineffective, contraindicated or not patient's preference
- ✓ In patients with AF undergoing myectomy , surgical AF ablation can be beneficial

# Management of HCM and Ventricular Arrhythmias



\* Not in children

# Hypertrophic Cardiomyopathy with Advanced Heart Failure



# Lifestyle Considerations in HCM



Mild to moderate intensity exercise if beneficial (Class I)

Comprehensive evaluation and shared discussion regarding sports participation are recommended (Class I)



Patients with other comorbidities, prevention and management of atherosclerotic cardiovascular disease are recommended (Class I)



It is reasonable to follow the Federal Motor Carrier Safety Guidelines that permit driving commercial vehicles for those who do not have ICD or any major risk for SCD (Class IIa)



For pilots with HCM, it is reasonable to permit multi-crew flying duties if they are asymptomatic, low risk for SCD and complete a treadmill stress test (Class IIa)



Moderate to high intensity exercise maybe considered after comprehensive evaluation and shared discussion (Class IIb)



ICD placement for the sole purpose of participation in competitive athletics should not be performed (Class III)



## Recommendations for HCM in Pregnancy



In high risk HCM, consultation with a maternal-fetal medicine expert is recommended (Class I).



In patients with HCM and atrial fibrillation or other indications for anti-coagulation, low-molecular weight heparin or low dose warfarin are recommended (Class I).



Selected beta-blocker should be administered for symptoms of LVOT obstruction or arrhythmia, with continued fetal monitoring (Class I).



Vaginal delivery is the first-choice delivery option in HCM (Class I).



In clinically stable HCM, it is reasonable to advise pregnancy is generally safe as part of shared discussion (Class IIa).



Reasonable to cardiovert new or recurrent atrial fibrillation, especially if symptomatic (Class IIa).



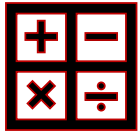
Reasonable to perform serial echocardiography in the second or third trimester, or if symptoms develop (Class IIa).



## Unmet Needs



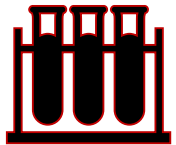
Randomized clinical trials are needed to prevent or attenuate disease progression, explore new therapies and gender-specific outcomes in HCM.



New risk factors to enhance the power of risk stratification algorithms, especially in children.



Pharmacological and catheter-based ablation for arrhythmia management, especially in young patients.



Greater access to genetic counseling and testing.



More data needed regarding potential risks of exercise and sports in patients with HCM.

Many thanks to our Guideline Ambassadors who were guided by Dr. Elliott Antman in developing this translational learning product adapted from the ACC/AHA 2020 Hypertrophic Cardiomyopathy Guideline.

Yuvraj Chowdhury, MD  
Marat Fudim, MD, MHS  
Ahmad Masri, MD, MS  
Nosheen Reza, MD  
Jainy Savla, MD  
Lina Ya'qoub, MD