In children, increased LV wall thickness is defined as wall thickness ≥2 standard deviations above the mean (z score ≥2) for age, sex, or body size.

Any wall thickness is compatible with HCM genetic substrate; an HCM subgroup is composed of family members with disease-causing sarcomere mutations without phenotype (no LVH).

### Genetic Testing Strategies/Family Screening

**Class I (Indicated)**
1. Evaluation of familial inheritance and genetic counseling is recommended. (LOE: B)
2. Screening (with or without genetic testing) is recommended in first-degree relatives. (LOE: B)
3. Genetic testing for HCM when other genetic causes of LVH are suspected. (LOE: B)

**Class IIa (Reasonable)**
1. Genetic testing in the index patient to help identify first-degree relatives at risk. (LOE: B)

**Class IIb (May be considered)**
1. Genetic testing in the assessment of risk of sudden cardiac death (SCD) in HCM. (LOE: B)

**Class III: No Benefit**
1. Genetic testing in relatives, if the index patient does not have a pathogenic mutation. (LOE: B)
2. Ongoing clinical screening in genotype-negative relatives in families with HCM. (LOE: B)

### Genotype-Positive/Phenotype-Negative Patients

**Class I (Indicated)**
1. ECG, TTE, clinical assessment: Q 12-18 mo in children/adolescents, Q 5 yrs in adults. (LOE: B)

**Echocardiography**

**Class I (Indicated)**
1. TTE in initial evaluation of all suspected patients or with a change in clinical status. (LOE: B)
2. TTE for screening family members unless family member is genotype negative. (LOE: B)
3. Periodic (12-18 mo) TTE screening for children of patients with HCM, starting age 12 or earlier if a growth spurt or signs of puberty are evident and/or when there are plans for engaging in intense competitive sports or there is a family history of SCD. (LOE: C)
4. TEE for the intraoperative guidance of surgical myectomy. (LOE: B)
5. TTE or TEE with IC contrast for procedural guidance of alcohol septal ablation. (LOE: B)
6. TTE to evaluate the effects of surgical myectomy or alcohol septal ablation for HOCM. (LOE: C)

**Class IIa (Reasonable)**
1. TTE every 1-2 yrs in stable patients to assess LVH, obstruction, and LV function. (LOE: C)
2. Exercise TTE to provoke LVOT obstruction in absence of resting gradient. (LOE: B)
3. TEE, if TTE is inconclusive, for clinical decision making about medical therapy and in planning for myectomy, exclusion of subaortic membrane or MR secondary to structural abnormalities of the mitral valve apparatus, or in assessment for the feasibility of alcohol septal ablation. (LOE: C)
4. TTE with contrast if apical HCM or apical infarction or severity of LVH is in doubt. (LOE: C)
5. Serial TTEs for unaffected patients with first-degree relative with HCM when genetic status is unknown (Q 12-18 months for children/adolescents; Q 5 yrs for adults). (LOE: C)

**Class III: No Benefit**
1. TTE more frequently than every 12 months in stable patients with HCM. (LOE: C)
2. TEE or contrast TTE, if TTE is diagnostic & no obstruction or MV disease is suspected. (LOE: C)

### Cardiac Magnetic Resonance (CMR)

**Class I (Indicated)**
1. In suspected HCM when echo is inconclusive for diagnosis. (LOE: B)
2. When magnitude and distribution of hypertrophy or anatomy of the mitral valve apparatus or papillary muscles is not adequately defined with echo, to plan invasive management. (LOE: B)

**Class IIa (Reasonable)**
1. To define apical hypertrophy and/or aneurysm if echo is inconclusive. (LOE: B)

**Class IIb (May be considered)**
1. When SCD risk stratification is inconclusive by conventional risk factors, CMR with assessment of late gadolinium enhancement may be considered in resolving clinical decision making. (LOE: C)
2. In LVH when there is suspicion of alternative diagnoses to HCM, including cardiac amyloidosis, Fabry disease, and genetic phenocopies such as LAMP2 cardiomyopathy. (LOE: C)
Stress Testing (GXT)
Class I (Indicated)
1. To determine functional capacity and response to therapy. (LOE: C)
2. GXT with monitoring ECG & BP for SCD risk stratification. (LOE: B)
3. When resting gradient is < 50 mmHg, to detect exercise-induced LVOT obstruction. (LOE: B)

Detection of Concomitant Coronary Disease

Asymptomatic Patients

Pharmacologic Management
Class I (Indicated)
1. Low hemodynamic: Drug therapy for symptoms and LVOT obstruction. (LOE: C)
2. Vasodilators/high doses of beta-blockers as first line therapy. (LOE: C)
3. Digitalis for dyspnea. (May be considered)
4. Diuretics with caution if dyspnea persists despite therapy. (LOE: C)

Invasive Therapies
Class I (Indicated)
1. Septal reduction should be performed by experienced operators in eligible patients. (LOE: C)
   * Operators with ≥ 20 procedures or work in a dedicated program with ≥ 50 cumulative total.
2. Surgical septal myectomy is first consideration for eligible patients. (LOE: B)
3. Surgical septal myectomy in eligible children who failed standard medical therapy. (LOE: C)
4. Alcohol septal ablation in eligible patients when surgery is contraindicated. (LOE: B)

Intracoronary Stenting or Angioplasty
Class I (Indicated)
1. Intracoronary stenting or angioplasty in the setting of symptomatic CAD. (LOE: C)
2. Intracoronary stenting or angioplasty in the setting of asymptomatic CAD. (LOE: C)

Septal Reduction in Asymptomatic Patients

Invasive Therapies
Class I (Indicated)
1. Alcohol septal ablation as an alternative to surgery if this is the patient’s preference. (LOE: B)
2. Alcohol septal ablation is discouraged in marked (>30 mm) septal hypertrophy. (LOE: C)

Asymptomatic Patients

Pharmacologic Management
Class I (Indicated)
1. Low hemodynamic: Drug therapy for symptoms and LVOT obstruction. (LOE: C)
2. Vasodilators/high doses of beta-blockers as first line therapy. (LOE: C)
3. Digitalis for dyspnea. (May be considered)
4. Diuretics with caution if dyspnea persists despite therapy. (LOE: C)

Invasive Therapies
Class I (Indicated)
1. Intracoronary stenting or angioplasty in the setting of symptomatic CAD. (LOE: C)
2. Intracoronary stenting or angioplasty in the setting of asymptomatic CAD. (LOE: C)

Intracoronary Stenting or Angioplasty
Class I (Indicated)
1. Intracoronary stenting or angioplasty in the setting of symptomatic CAD. (LOE: C)
2. Intracoronary stenting or angioplasty in the setting of asymptomatic CAD. (LOE: C)

Septal Reduction in Asymptomatic Patients

Invasive Therapies
Class I (Indicated)
1. Alcohol septal ablation as an alternative to surgery if this is the patient’s preference. (LOE: B)
2. Alcohol septal ablation is discouraged in marked (>30 mm) septal hypertrophy. (LOE: C)

Asymptomatic Patients

Pharmacologic Management
Class I (Indicated)
1. Low hemodynamic: Drug therapy for symptoms and LVOT obstruction. (LOE: C)
2. Vasodilators/high doses of beta-blockers as first line therapy. (LOE: C)
3. Digitalis for dyspnea. (May be considered)
4. Diuretics with caution if dyspnea persists despite therapy. (LOE: C)

Invasive Therapies
Class I (Indicated)
1. Intracoronary stenting or angioplasty in the setting of symptomatic CAD. (LOE: C)
2. Intracoronary stenting or angioplasty in the setting of asymptomatic CAD. (LOE: C)

Intracoronary Stenting or Angioplasty
Class I (Indicated)
1. Intracoronary stenting or angioplasty in the setting of symptomatic CAD. (LOE: C)
2. Intracoronary stenting or angioplasty in the setting of asymptomatic CAD. (LOE: C)

Septal Reduction in Asymptomatic Patients

Invasive Therapies
Class I (Indicated)
1. Alcohol septal ablation as an alternative to surgery if this is the patient’s preference. (LOE: B)
2. Alcohol septal ablation is discouraged in marked (>30 mm) septal hypertrophy. (LOE: C)
Pacing

Class IIa (Reasonable)
1. RV apical pacer (not implanted for HOCM) may be used to relieve HOCM symptoms. (LOE: B)

Class IIb (May be considered)
1. In patients who are asymptomatic or whose symptoms are medically controlled. (LOE: C)
2. As a first-line therapy in refractory patients eligible for septal reduction. (LOE: B)

Sudden Cardiac Death Risk Stratification

Class I (Indicated)
1. Comprehensive SCD risk stratification at initial evaluation to determine presence of: (LOE: B)
   a. Personal history for VF, sustained VT, or SCD events, including appropriate ICD therapy.*
   b. Family history for SCD events, including appropriate ICD therapy.*
   c. Unexplained syncope.
   d. Documented NSVT ≥ 3 beats at ≥ 120 bpm on ambulatory (Holter) electrocardiogram.
   e. Maximal LV wall thickness greater than or equal to 30 mm.
   *Appropriate ICD therapy is triggered by VT or VF, in conjunction with symptoms.

Class IIa (Reasonable)
1. Assessment of BP response during exercise. (LOE: B)
2. SCD risk stratification every 12–24 months for patients without ICD. (LOE: C)

Class IIb (May be considered)
1. Usefulness of the following SCD risk modifiers is unclear but might be considered adjunctive:
   a. CMR imaging with late gadolinium enhancement. (LOE: C)
   b. Double and compound mutations (i.e., >1). (LOE: C)
   c. Marked LVOT obstruction. (LOE: B)

Class III: Harm
1. Invasive electrophysiology testing as routine SCD risk stratification strategy. (LOE: C)

Selection of Patients for Implantable Cardioverter-Defibrillators

Class I (Indicated)
1. ICD consideration should allow informed patient’s participation in decision making. (LOE: C)
2. For prior documented cardiac arrest, VF, or hemodynamically significant VT. (LOE: B)

Class IIa (Reasonable)
1. It is reasonable to recommend an ICD for patients with HCM with:
   a. Sudden death presumably caused by HCM in 1 or more first-degree relatives. (LOE: C)
   b. A maximum LV wall thickness greater than or equal to 30 mm. (LOE: C)
   c. One or more recent, unexplained syncopal episodes. (LOE: C)
2. For NSVT (particularly <30 years of age) & other SCD risk factors or modifiers*. (LOE: C)
3. For Abnormal exercise BP response & other SCD risk factors or modifiers.* (LOE: C)
4. For high-risk children with unexplained syncope, massive LVH, or FH of SCD. (LOE: C)

Class IIb (May be considered)
1. For isolated NSVT bursts in the absence of any other SCD risk factors or modifiers.* (LOE: C)
2. For abnormal GXT BP, without SCD risk factors/modifiers*, esp. no LVOT obstruction. (LOE: C)

Class III: Harm
1. As a routine strategy without an indication of increased risk. (LOE: C)
2. As a strategy to permit patients with HCM to participate in competitive athletics. (LOE: C)
3. For patient with HCM genotype without clinical manifestations of HCM. (LOE: C)

Management of Atrial Fibrillation

Class I (Indicated)
1. Warfarin (INR 2–3) for AF. (dabigatran* may be an option, but no data in HCM!). (LOE: C)
2. Rate control in AF with RVR; can require high ββ & CCB doses. (LOE: C)
   * Avoid dabigatran in prosthetic valves, sig. valve disease, liver failure, or CrCl<15 mL/min.

Class IIa (Reasonable)
1. Disopyramide (rate-controlling agents) & amio are reasonable antiarrhythmics. (LOE: B)
2. RF ablation for AF with refractory symptoms or inability to take antiarrhythmic drugs. (LOE: B)
3. Maze + LA appendage closure during septal myectomy or as an isolated procedure. (LOE: C)

Class IIb (May be considered)
1. Sotalol, dofetilide, & dronedarone, especially + ICD, but clinical experience is limited! (LOE: C)

Pregnancy/Delivery

Class I (Indicated)
1. ββ for symptoms should be continued (? fetal bradycardia or other complications ). (LOE: C)
2. For mother or father with HCM, genetic counseling before planned conception. (LOE: C)
3. If ≥ 50 mmHg LVOT obstruction &/or uncontrolled symptoms, referral to high risk OB. (LOE: C)
4. Asymptomatic HCM does not contraindicate pregnancy, but risk should be evaluated. (LOE: C)

Class IIa (Reasonable)
1. With mild to moderate symptoms. Expert maternal/fetal medical specialist is advised. (LOE: C)

Class IIb (Reasonable)
1. With advanced CHF symptoms, there is excess morbidity/mortality. (LOE: C)
Participation in Competitive or Recreational Sports and Physical Activity

Class IIa (Reasonable)
1. Low-intensity competitive sports (e.g., golf and bowling). (LOE: C)
2. A range of recreational sporting activities as outlined in Table 2. (LOE: C)

Class III: Harm
1. Intense competitive sports regardless of age, sex, race, presence or absence of LVOT obstruction, prior septal reduction therapy, or ICD implantation for high-risk status. (LOE: C)

*Recreational sports are categorized according to high (6 METS), moderate (4-6 METS), and low (4 METS) levels of exercise and graded on a relative scale (from 0 to 5) for eligibility:

0 to 1: generally not advised or strongly discouraged
2 to 3: intermediate and to be assessed clinically on an individual basis
4 to 5: probably permitted

†Assumes absence of laboratory DNA genotyping data; therefore, limited to clinical diagnosis.
‡These sports involve the potential for traumatic injury, which should be taken into consideration for individuals with a risk for impaired consciousness.
§The possibility of impaired consciousness occurring during water-related activities should be taken into account with respect to the individual patient’s clinical profile.
ǁRecommendations generally differ from those for weight-training machines (nonfree weights), based largely on the potential risks of traumatic injury associated with episodes of impaired consciousness during bench-press maneuvers; otherwise, the physiologic effects of all weight-training activities are regarded as similar with respect to the present recommendations.
¶Individual sporting activity not associated with the team sport of ice hockey.
Figure 1. Treatment Algorithm

1. **HCM Patients**
   - TreatConsidering according to GL HCM, Lopes, DMD

2. **Obstructive Physiology**
   - Annual clinical evaluation
     - AVOID vasodilator therapy and high-dose diuretics
     - Heart Failure Symptoms or Angina
       - LV EF ≤50%
       - LV EF <30%
         - Beta Blockade
         - Verapamil
         - Diuretics
         - ACE Inhibitors or ARB

3. **Heart Failure Symptoms or Angina**
   - Therapy as outlined in Heart Failure GL
     - Persistent Symptoms
       - Beta Blockade
       - Verapamil
       - Digoxin

4. **Invasive Therapy**
   - Acceptable surgical candidacy
     - Yes: Surgical Myectomy
   - Acceptable candidate for alcohol ablation
     - Yes: Alcohol Ablation
     - No: Consider TTOO Pacing

Legend:
- Class I
- Class Ia
- Class Ib