Class I Recommendations for Echocardiography in Hypertrophic Cardiomyopathy (HCM)*

Suspected or established HCM
1. TTE is recommended in initial evaluation of all suspected HCM patients.
2. In patients with established HCM & no change in clinical status, repeat TTE recommended every 1 to 2 years.
3. In patients with established HCM & change in clinical status, prompt repeat TTE is recommended.
4. In patients who have undergone septal reduction therapy, TTE within 3-6 months of the procedure recommended to evaluate procedural results.

Screening & follow-up
1. In first-degree relatives of patients with HCM, TTE is recommended as part of initial family screening & periodic follow-up.
2. In individuals who are genotype-positive/phenotype-negative, serial TTE is recommended at periodic intervals depending on age & clinical status.

Provocative testing
1. In patients with HCM & resting LVOT gradient <50 mmHg, TTE with provocative maneuvers recommended.
2. In symptomatic patients with HCM without resting or provocative LVOT gradient ≥50 mmHg on TTE, exercise TTE recommended for detection & quantification of dynamic LVOT obstruction.

Septal reduction therapies
1. For patients with HCM undergoing surgical septal myectomy, intraoperative TEE recommended to assess mitral valve anatomy & function & adequacy of septal myectomy.
2. For patients with HCM undergoing alcohol septal ablation, TTE or intraoperative TEE with intracoronary ultrasound-enhancing contrast injection of candidate’s septal perforator(s) is recommended.

Goals of Echocardiographic Assessment in Hypertrophic Cardiomyopathy (HCM)

Establish diagnosis & determine pattern of hypertrophy
Clinical diagnosis should be suspected with evidence of a large end-diastolic wall thickness of >15 mm anywhere in the left ventricle, absent another cause of hypertrophy in adults.
Differentiate sigmoid septum (with ovoid cavity) versus reverse curve (with crescent cavity) versus apical hypertrophic phenotypes.
Massive left ventricular hypertrophy >30 mm in any left ventricular segment is a risk factor for sudden cardiac death (SCD).

Evaluate global myocardial function
Systolic dysfunction defined as LVEF <50%.
Strain abnormalities correlate with increased wall thickness & delayed gadolinium enhancement by MRI.

Establish presence & severity of LVOT obstruction
Peak LVOT gradient of ≥50 mmHg at rest or with provocation or exercise indicates obstruction.
Differentiate SAM-mediated LVOT obstruction from mid-ventricular obstruction (MVO; ”dagger” shaped).
Caution with contamination of LVOT signal with MR. MR velocity is higher & signal is of longer duration (spreading isovolumic contraction & relaxation) vs LVOT signal.
MR contour may be incomplete if Doppler signal not optimally aligned.
Estimated LVOT gradient from MR signal calculated as: LV Pressure - Systolic BP, where

LV Pressure = 4 x ( Peak MR Velocity )^2 + LA Pressure (assume 10 - 15 mmHg)

Evaluate degree & direction of mitral regurgitation, & intrinsic structure of mitral valve & papillary muscles
MR caused by LVOT obstruction results from SAM & results in a jet direction that is posterior & lateral in orientation & predominantly mid-to-late systolic.
Central or anterior jets should prompt further evaluation for intrinsic valvular or papillary abnormalities.

Evaluate for LV apical aneurysm
TTE with ultrasound-enhancing agent should be performed in patients with HCM with suspected apical or mid-ventricular hypertrophy to evaluate for apical aneurysms.
Presence of an LV apical aneurysm is an established clinical risk factor for SCD.

Plan & guide septal reduction therapies & subsequently assess their efficacy

Preoperative measurements include:
A) IVS maximum thickness
B) Anterior leaflet length
C) Apical extent of septal bulge
D) Distance from aortic annulus to mitral-septal contact

Pre-myectomy measurements

Postoperative measurements include:
A) IVS maximum thickness
B) Anterior leaflet length
C) Apical extent of septal bulge
D) Distance from aortic annulus to mitral-septal contact

Identify appropriate septal perforator that supplies SAM-septal contact (by TTE or TEE).
Inappropriate targets:
- Distal Septum
- LV Papillary Muscles
- RV Papillary Muscles

Alcohol septal ablation

Abbreviations
HCM Hypertrophic cardiomyopathy
LVOT Left ventricular outflow tract
LVSP Left ventricular systolic pressure
MR Mitral regurgitation
MVO Mid-ventricular obstruction
SAM Systolic anterior motion
SCD Sudden cardiac death

* ACC/AHA Guideline Document (See full citation on back)

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**Recommended Evaluation & Testing for Hypertrophic Cardiomyopathy**

**Family History of Hypertrophic Cardiomyopathy (HCM) or Suspected HCM**

Perform Diagnostic Evaluation (ECG, Imaging)

**Phenotype Negative**

Family with a known pathogenic or likely pathogenic variant?

- Yes
  - Patient has family variant?
    - Yes, or unknown
      - **Class I**
        - Screening ECG & Echo at the intervals in the table below
    - No
      - **Class III: No Benefit**
        - Further clinical or genetic testing is not recommended

- No
  - **Class III: No Benefit**
    - Further clinical or genetic testing is not recommended

**Phenotype Positive**

**Risk factors for SCD**
- Unexplained Syncope
- Massive LVH
- Family history SCD
- Apical aneurysm
- EF <50%
- Extensive LGE on CMR

Perform Comprehensive Baseline Evaluation with SCD risk assessment (consider ESC guidelines)

- **Stress testing**
  - to determine baseline functional status
  - to clarify symptom status
  - to confirm presence and quantify severity of LVOTO

**Class I**
- Every 1-2 years or sooner if change in symptoms
  - Perform serial evaluation of clinical status & SCD risk with
    - Clinical assessment
    - Echo
    - Holter

**Class IIb**
- Every 3-5 years
  - Perform CMR for SCD risk assessment or to evaluate for any suspected morphologic changes

- **If Symptomatic**
  - Treadmill or Bike Exercise Testing
    - Special considerations:
      - Stress echo if gradient <50 mmHg
      - CPET if considering advanced HF therapies

- **If Asymptomatic**
  - Every 2-3 years
    - Treadmill exercise or CPET testing for assessment of functional status

**Screening Asymptomatic First-Degree Relatives of Patients with HCM**

<table>
<thead>
<tr>
<th>Age of First-Degree Relative</th>
<th>Initiation of Screening</th>
<th>Surveillance Interval</th>
</tr>
</thead>
<tbody>
<tr>
<td>Children &amp; adolescents from genotype-positive family and/or family with early onset HCM</td>
<td>At the time of diagnosis in another family member</td>
<td>Every 1-2 years</td>
</tr>
<tr>
<td>All other children &amp; adolescents</td>
<td>At any time after the diagnosis in the family (no later than puberty)</td>
<td>Every 2-3 years</td>
</tr>
<tr>
<td>Adults</td>
<td>At the time of diagnosis in another family member</td>
<td>Every 3-5 years</td>
</tr>
</tbody>
</table>

**Class (Strength) of Recommendation**

- **Class I (STRONG)** Benefit >>> Risk
- **Class IIb (WEAK)** Benefit ≥ Risk
- **Class III: No Benefit** Benefit = Risk