The Role of Echocardiography in Hypertrophic Cardiomyopathy

ASE American Society of Echocardiography — Sound Saves Lives

ASE Foundation

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

Suspected or established HCM

1. TTE is recommended in initial evaluation of all suspected HCM
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 provocable 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

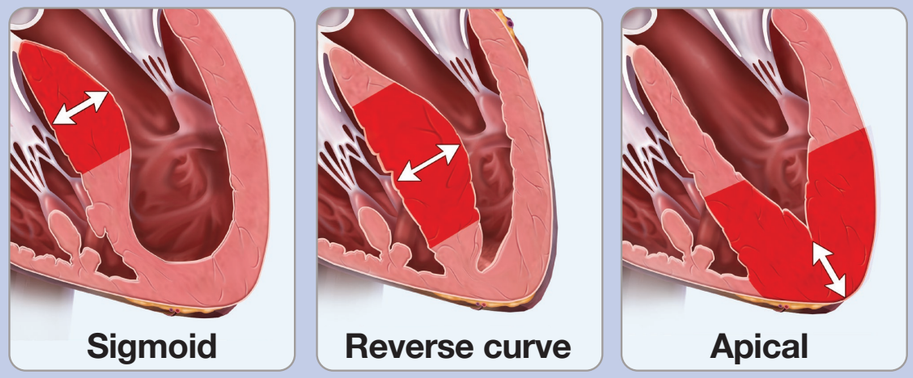
Goals of Echocardiographic Assessment in Hypertrophic Cardiomyopathy (HCM)

Establish diagnosis & determine pattern of hypertrophy

Clinical diagnosis should be suspected with imaging evidence of a maximal 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

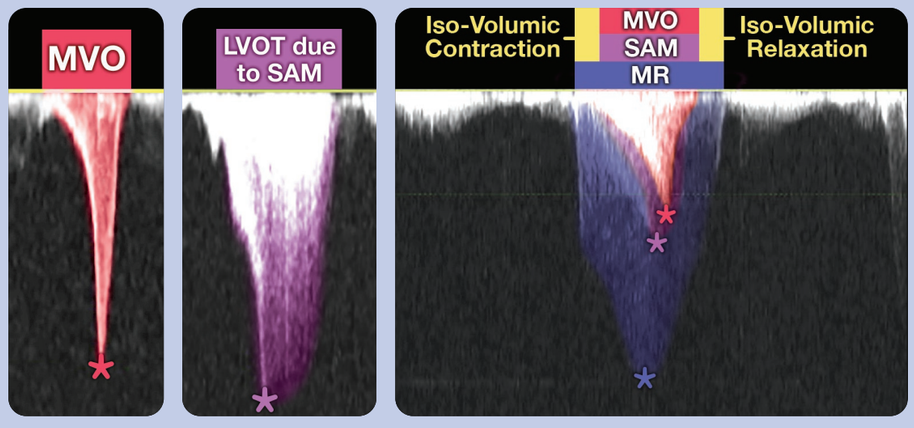
Massive left ventricular hypertrophy >30 mm in any left ventricular segment is a risk factor for sudden cardiac death (SCD)



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

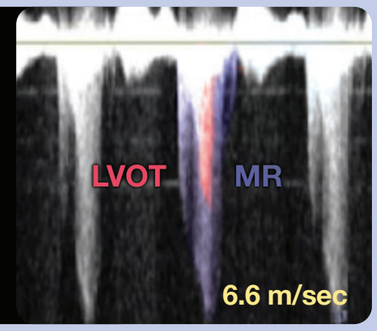
Differentiate SAM-mediated LVOT obstruction from mid-ventricular obstruction (MVO; "dagger" shaped)

Caution with contamination of LVOT signal with MR. MR velocity is higher & signal is of longer duration (spanning 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

$$\text{LV Pressure} = 4 \times (\text{Peak MR}_{\text{velocity}})^2 + \text{LA Pressure}$$
(assume 10 - 15 mmHg)

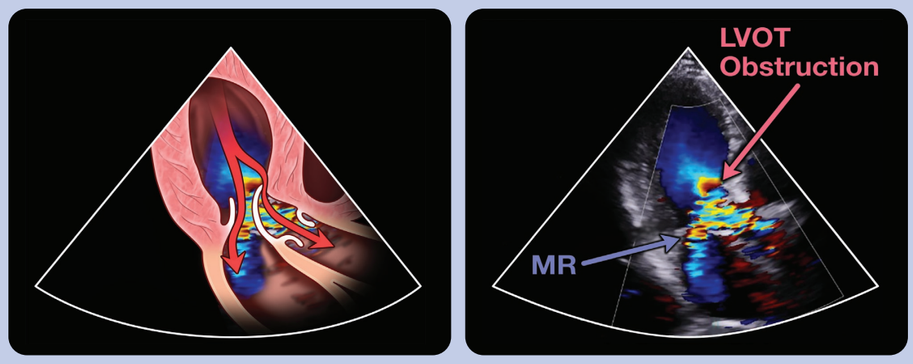
Peak $MR_{velocity}$ = 6.6 m/sec
Peak LVSP = $4(6.6^2)$ + LAP (10 mmHg)
SBP = 113 mmHg
LVSP - SBP = LVOT gradient
174 + 10 = 184 - 113 = 71 mmHg



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 or lateral in orientation & predominantly mid-to-late systolic

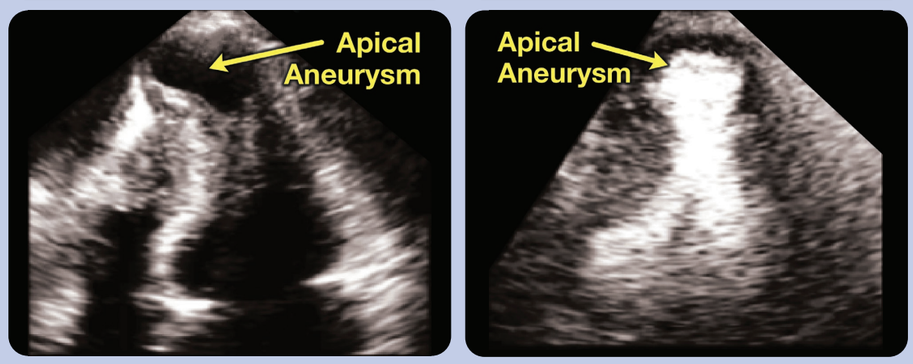
Central or anterior jets should prompt further evaluation for intrinsic valvular or papillary abnormalities



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

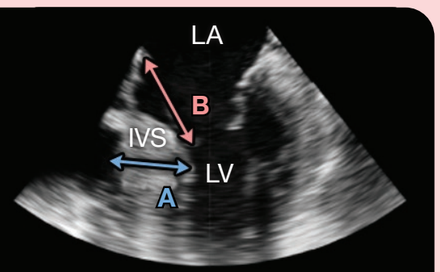
Preoperative measurements include:

A) IVS maximum thickness

B) Anterior leaflet length

C) Apical extent of septal bulge

D) Distance from aortic annulus to mitral-septal contact

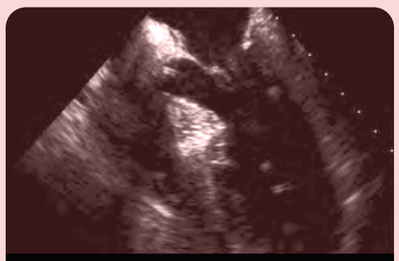


Pre-myectomy measurements

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	**LVOTO**	Left ventricular outflow tract obstruction	**MR**	Mitral regurgitation	**SAM**	Systolic anterior motion
LVOT	Left ventricular outflow tract	**LVSP**	Left ventricular systolic pressure	**MVO**	Mid-ventricular obstruction	**SCD**	Sudden cardiac death

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

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Poster Authors: Karan Kapoor, MD; Allison G Hays, MD, FASE. Design and illustration by medmovie.com

Recommended Evaluation & Testing for Hypertrophic Cardiomyopathy

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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

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

Class (Strength) of Recommendation

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

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