THE MASQUERADER | Could It Be Hypertrophic Cardiomyopathy (HCM)?

1. Investigate for HCM and conduct further diagnostic workup based on the presence of symptoms, abnormal ECG result indicating LV hypertrophy, and/or systolic ejection murmur[1-3]

2. Identify and characterize LV hypertrophy using 2D echocardiography (echo) or M-mode imaging[4,5]

3. Assess for systolic anterior motion (SAM) of the mitral valve and left ventricular outflow tract (LVOT) obstruction using 2D and Doppler echo[3,5]

4. Evaluate LVOT obstruction both at rest AND with provocation (stress echo preferred over Valsalva maneuver in certain instances)[5,6]

5. Consider CMR imaging when echo results are inconclusive or for prognostic purposes[3,5,7]

Any Signs and Symptoms Suggestive of HCM?

Common Symptoms[8]

- Dyspnea
- Fatigue
- Chest pain
- Palpitations
- Syncope

ECG Abnormalities[1-3]

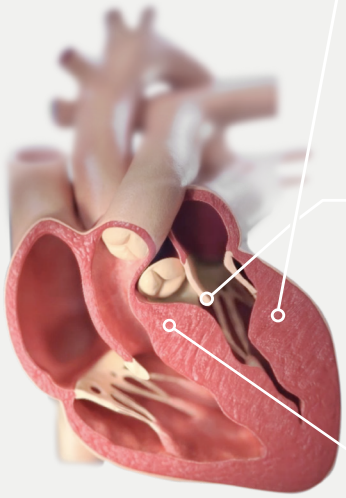
- ST-T wave changes
- Pathological Q waves
- Increased QRS voltages
- Deep T-wave inversion (apical HCM)

Note: Although no specific ECG pattern is pathognomonic, these ECG anomalies are consistent with HCM

Systolic Ejection Murmur[3,6]

- Best heard over the left sternal border
- Becomes louder with maneuvers that decrease preload or afterload (eg, Valsava maneuver)

Consider starting with echo



A Identification and Characterization of LV Hypertrophy

HCM is defined by an unexplained maximal end-diastolic wall thickness of ≥15 mm anywhere in the left ventricle in patients without family history of HCM and 13-14 mm in those with family history of HCM or with a positive genetic test[3,6]

Suggested echo techniques:

- **2D echo (measures all LV segments, from base to apex)[4]**
- **M-mode (measures only the basal segments of the interventricular septum and the posterior wall)[4]**
- **All imaging views are important for characterizing and defining LV hypertrophy[5]**

NOTE: LV hypertrophy may also be isolated in the LV free wall or apex.[5] During these instances, consider LV opacification with contrast, TTE with contrast, or CMR imaging[5]

B SAM

SAM of the mitral valve or chordae occurs when abnormalities of the mitral valve apparatus cause the leaflets to be swept into the LVOT by drag forces in the presence of hyperdynamic ejection fraction, making mitral valve SAM one of the causes of LVOT obstruction[5,6]

Suggested echo techniques:

- **2D echo[3]**
- **M-mode[9]**
- **Color Doppler (LVOT turbulence)[5]**

NOTE: Because SAM can result in the distortion of mitral leaflet coaptation and subsequently MR, particular attention should be paid to avoid mixing LVOT gradient with MR jet[5]

C LVOT Obstruction

Per current data, the majority of diagnosed patients had obstructive HCM.[10] LVOT obstruction can result from septal hypertrophy that leads to narrowing of the LVOT or anatomic alterations in the mitral valve and apparatus[6]

Suggested echo techniques[5]:

- **Continuous-wave Doppler (peak gradient)**
- **Pulsed-wave Doppler (site of obstruction)**

NOTE: LVOT gradient should be measured in all patients under resting AND provocative conditions (stress echo preferred over Valsalva maneuver in certain instances)[5,6]; otherwise obstruction, which portends a worse prognosis and affects management, might be missed[6,11]

Need More Information? Consider CMR Imaging

Use if echo findings are inconclusive, if ECG and echo results do not match, or if risk stratification is needed[3,5,7]

NOTE: Even though CMR imaging is helpful in identifying HCM in instances when echo may be lacking, it is more helpful in some instances (eg, detection of LV apical and anterolateral hypertrophy) than others (eg, Doppler echo is preferred over CMR imaging for the quantification of LVOT obstruction)[3]

HCM Is Difficult to Monitor – Do So Objectively

As patients subtly adapt their lifestyles, subjective assessment of worsening symptoms and function may be complicated.[12] Therefore, objective monitoring over a patient's lifetime is crucial.

Test[6]	Frequency[6]
12-Lead ECG or TTE	Every 1-2 years
24- to 48-hour ambulatory ECG monitoring	Every 1-2 years
Cardiopulmonary exercise testing	Every 2-3 years for patients whose functional capacity or symptom status is uncertain
CMR imaging	Every 3-5 years

2D, 2-dimensional; CMR, cardiac magnetic resonance; ECG, electrocardiogram; LV, left ventricular; MR, mitral regurgitation.

1. Maron MS et al. *Circ Cardiovasc Imaging.* 2017;10(7):e005372. **2.** Finocchiaro G et al. *Heart Rhythm.* 2020;17(1):142-151. **3.** Elliott PM et al. *Eur Heart J.* 2014;35(39):2733-2779. **4.** Pantazis A et al. *Echo Res Pract.* 2015;2(1):R45-R53. **5.** Nagueh SF et al. *J Am Soc Echocardiogr.* 2011;24(5):473-498. **6.** Ommen SR et al. *J Am Coll Cardiol.* 2020. doi:10.1016/j.jacc.2020.08.045. **7.** Amano Y et al. *Magn Reson Med Sci.* 2018;17(2):120-131. **8.** Cleveland Clinic. Heart failure: understanding heart failure. https://my.clevelandclinic.org/health/diseases/17069-heart-failure-understanding-heart-failure. Accessed September 3, 2020. **9.** Losi MA et al. *Cardiovasc Ultrasound.* 2010;8:7. **10.** Maron MS et al. *Circulation.* 2006;114(21):2232-2239. **11.** O'Mahony C et al. *Circ Arrhythm Electrophysiol.* 2013;6(2):443-451. **12.** Jacoby DL et al. *CMAJ.* 2013;185(2):127-134.

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