Advanced Cardiac Imaging

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Disclosures

- Nothing to disclose

Case

27 yo Egyptian man with history of syncope and mitral regurgitation presents to the ED after syncopal episode while walking up stairs to his apartment.

He reports >10 episodes of syncope in last 4-6 months with first episode 2 years ago. He is also having daily exertional dyspnea, chest tightness, and lightheadedness. Denies palpitations, orthopnea, or edema.

Case –History

- Per patient echocardiogram results in Egypt “mitral regurgitation and narrowing of another valve”

- Family history –
  - Both mother and father with unknown cardiac “valvular” issues. Both alive and well
  - No family history of sudden cardiac death or syncope

Exam

Vitals
BP 119/59 HR 92 bpm RR 16 Ht 5’8 Wt 218 lbs BMI 33

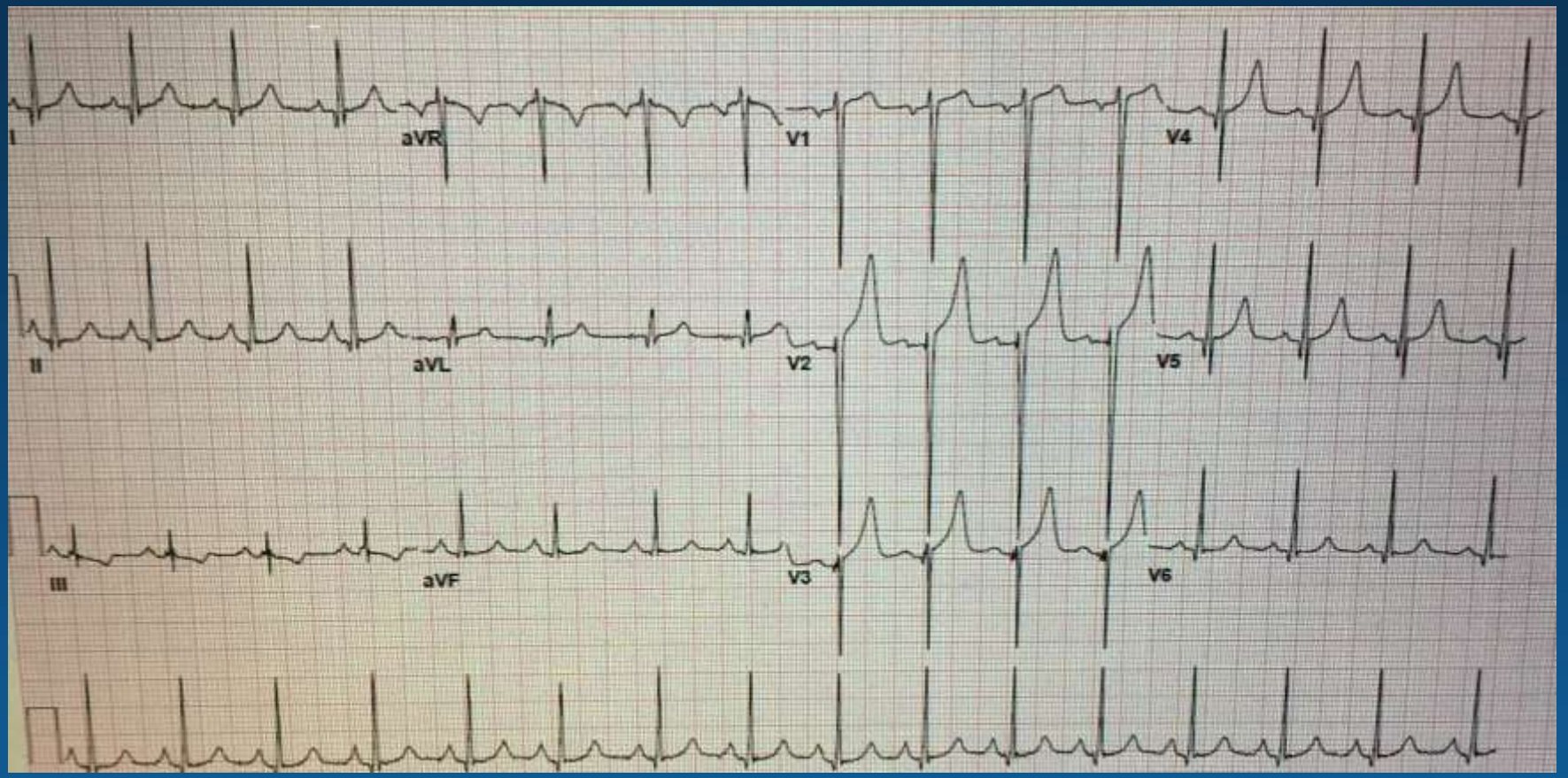
Gen
-obese, young gentleman, no distress, no craniofacial abnormalities

Cardiac Exam
- regular, normal rate, hyperdynamic LV on palpation

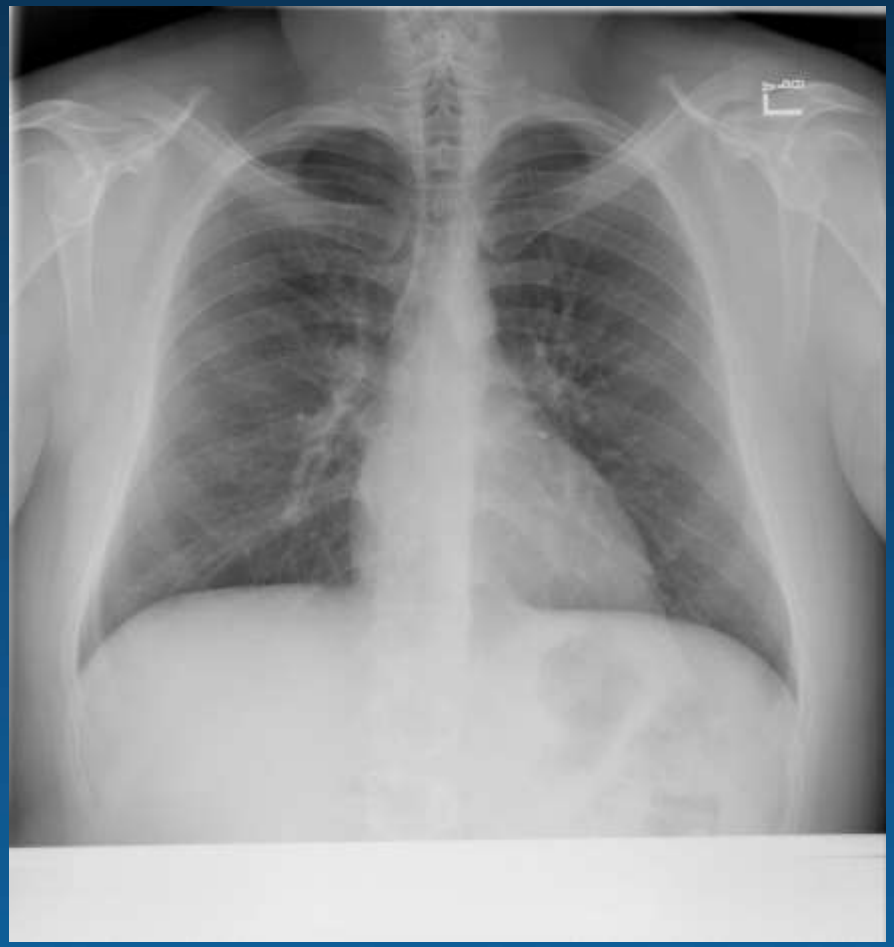
- 3/6 early mid peaking systolic murmur best heard left sternal border increases with valsalva

- faint holosystolic murmur heard at apex

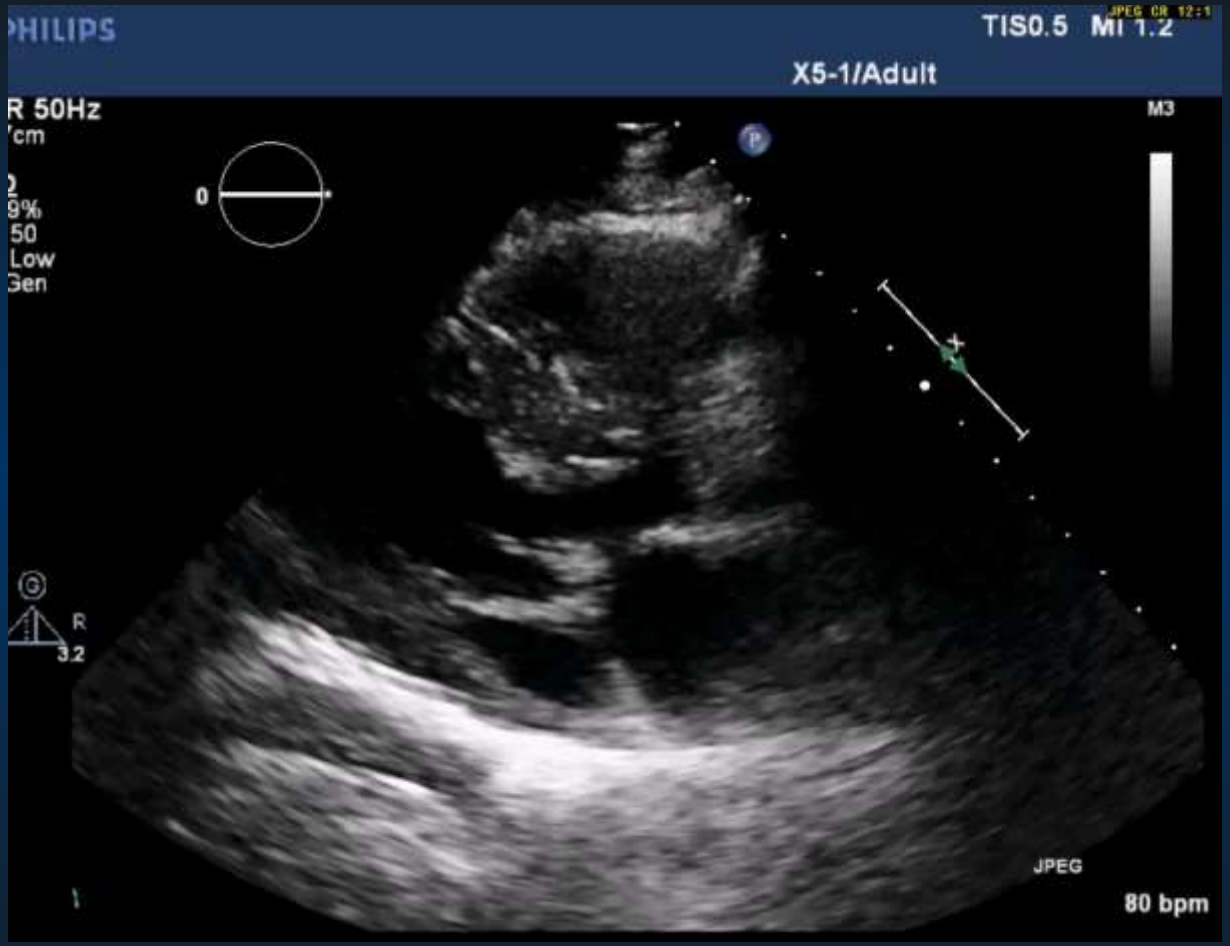
ECG



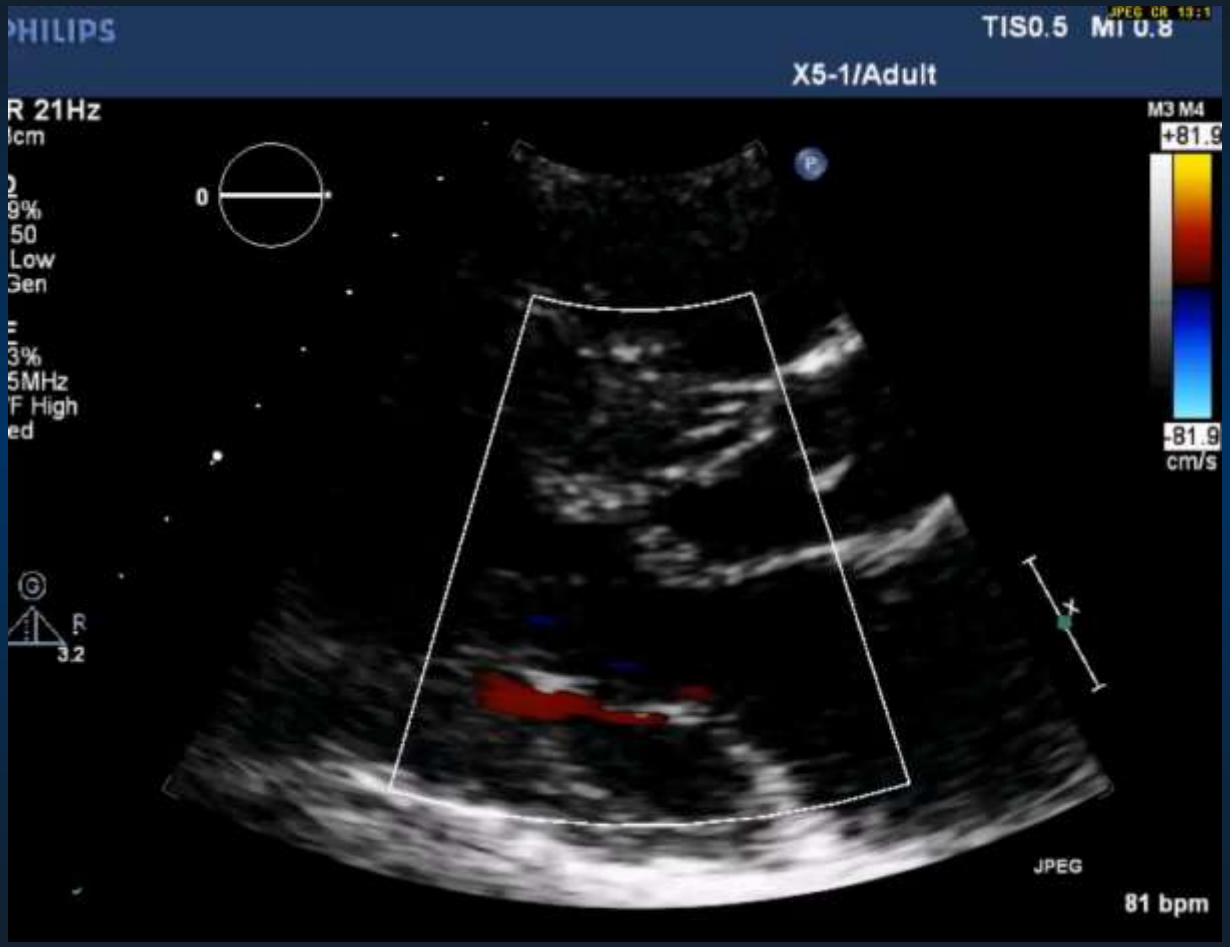
Chest X-ray PA film



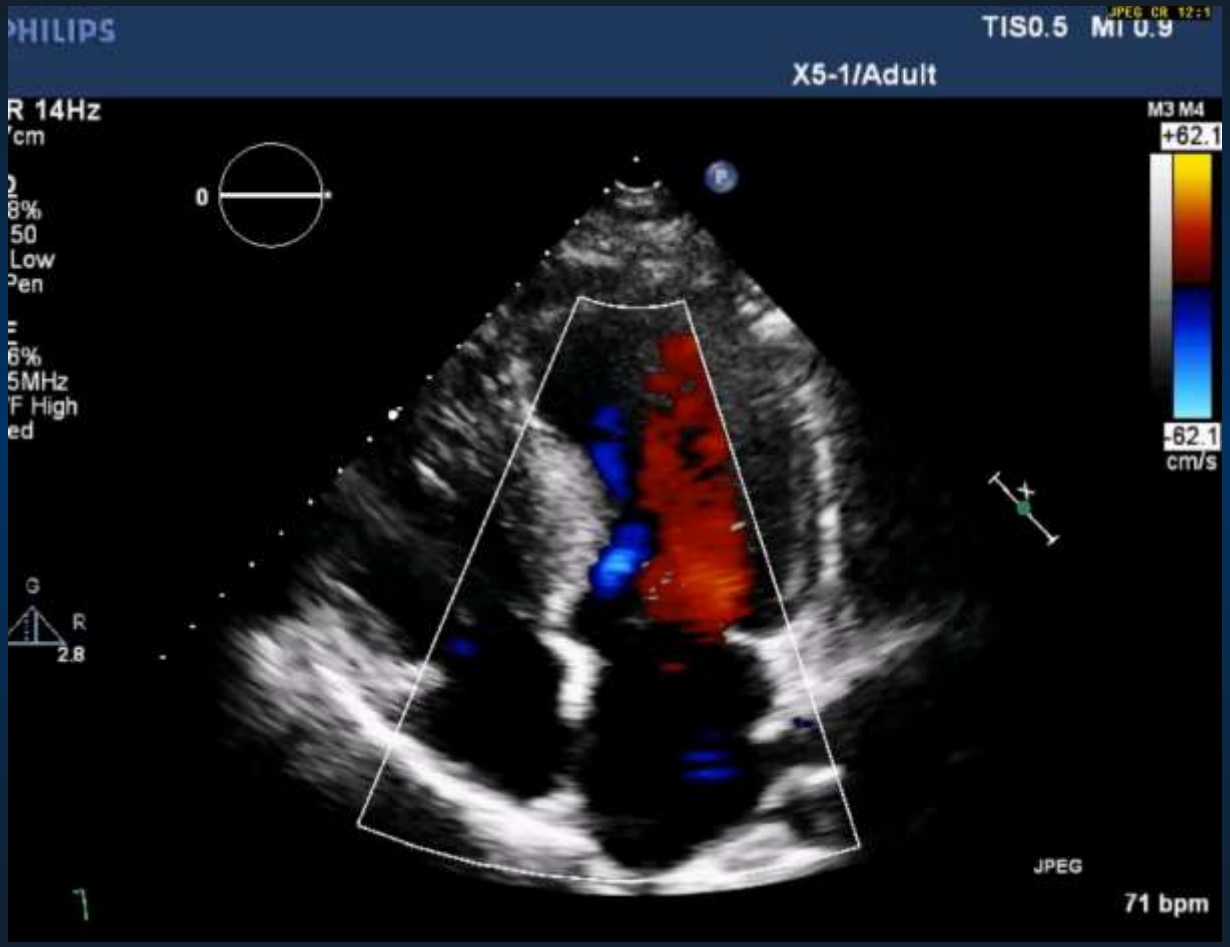
2D Echo: Parasternal Long Axis



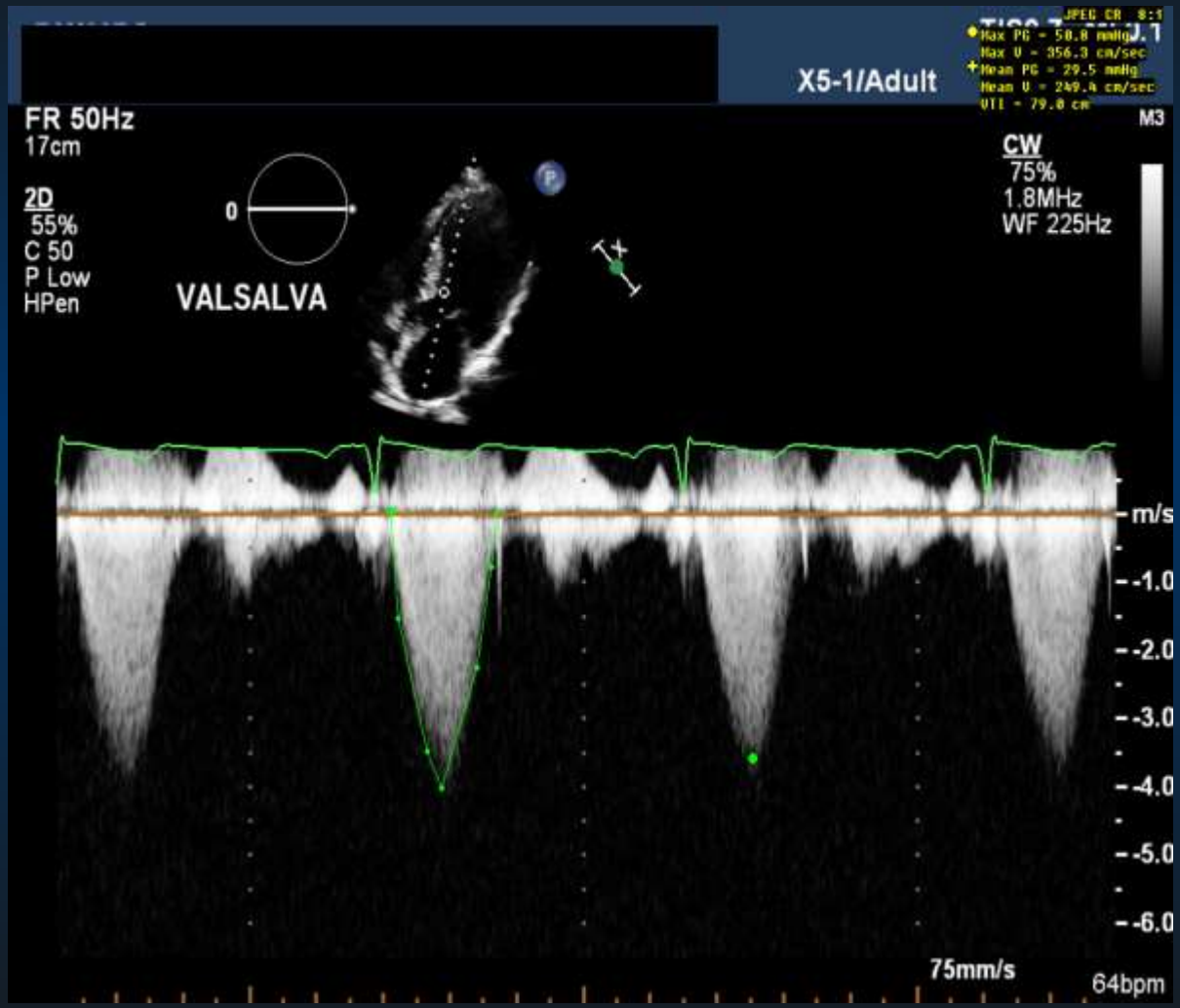
Echo: Parasternal with color doppler



Transthoracic Echo: 4C w/color doppler



Peak LVOT gradient



LVOT peak gradient
Valsalva- 51 mmHg

Echo Findings

- Asymmetric left ventricular hypertrophy with septum measuring 1.6 cm on parasternal long axis
- Presence of systolic anterior motion of mitral leaflet
- LV outflow tract peak gradient of 23 mmHg at rest and 51 mmHg with Valsalva
- Moderate mitral regurgitation
- Left atrial enlargement

Diagnosis?

What Next?

Morphologic Diagnosis of Hypertrophic Cardiomyopathy

- Hypertrophic & non dilated left ventricle
- Typically at least > 15 mm LV hypertrophy
- Absence of another cardiac or systemic diagnosis capable of causing this degree of hypertrophy
- May or may not have LV outflow tract obstruction

Gersh B et al. J Am Coll Cardiol. 2011;58(25):e212-e260

Cardiac MRI?

CMR in HCM Evaluation

- High spatial resolution and tomographic capability
- Better visualization of anterolateral wall, apex and right ventricular involvement
- More accurate assessment of LV mass and in some cases LV wall thickness
- Detection and quantification of myocardial fibrosis via late gadolinium enhancement

To AC JACC Imag. 4 (10) 1123-1131

ACC/AHA Guidelines for Cardiac MR in HCM

- Class I

  - CMR indicated if diagnostic uncertainty after echocardiogram
  - CMR indicated in patients when further anatomic definition may impact management or decision making regarding invasive therapy

- Class IIa

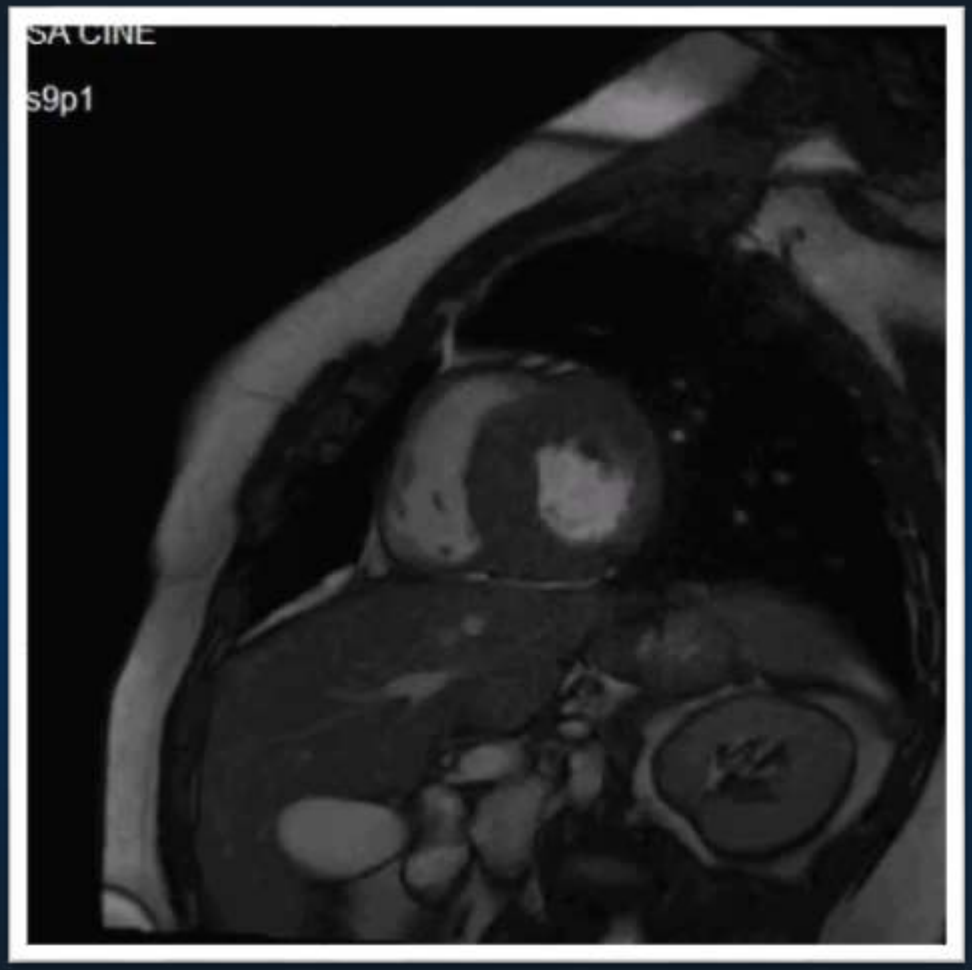
  - CMR reasonable to define LV apical hypertrophy or aneurysm if echo inconclusive

- Class IIb

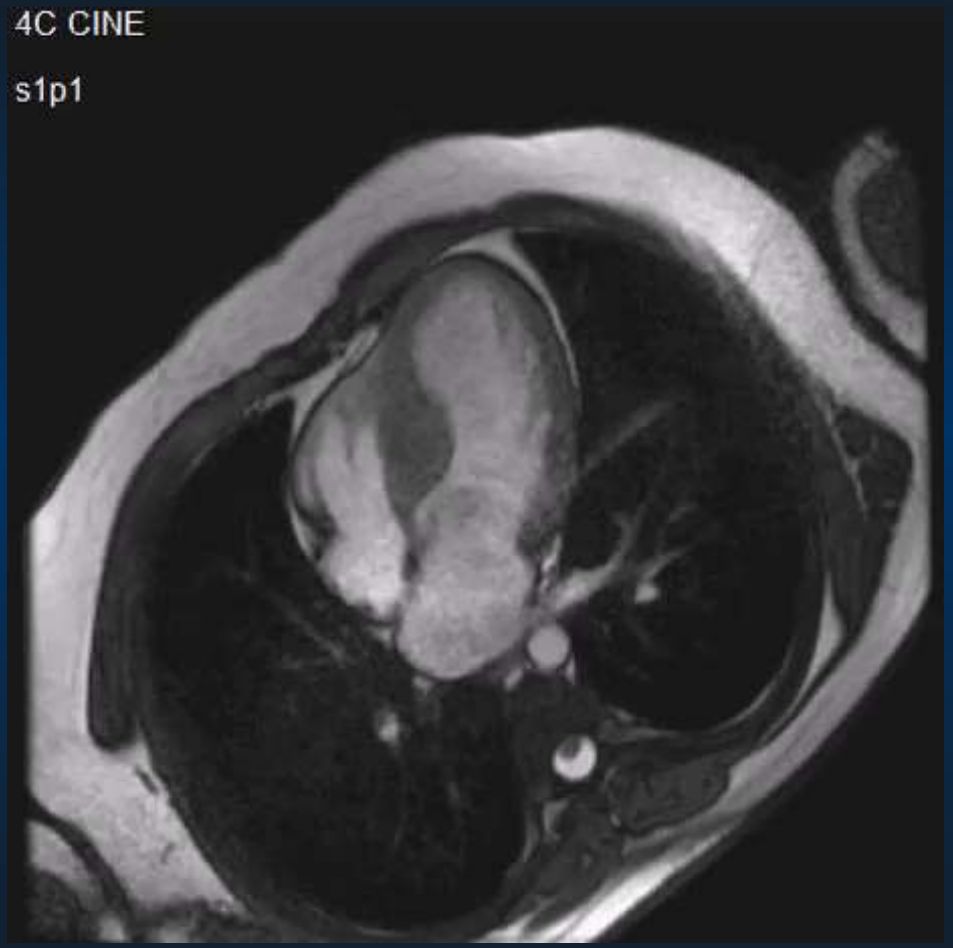
  - CMR with late gadolinium enhancement can be considered for risk stratification for sudden cardiac if risk is inconclusive after evaluating for traditional risk factors

Gersch BJ et al. *Circulation* 2011; 124:2761-2796

Cardiac MR: Short axis cine



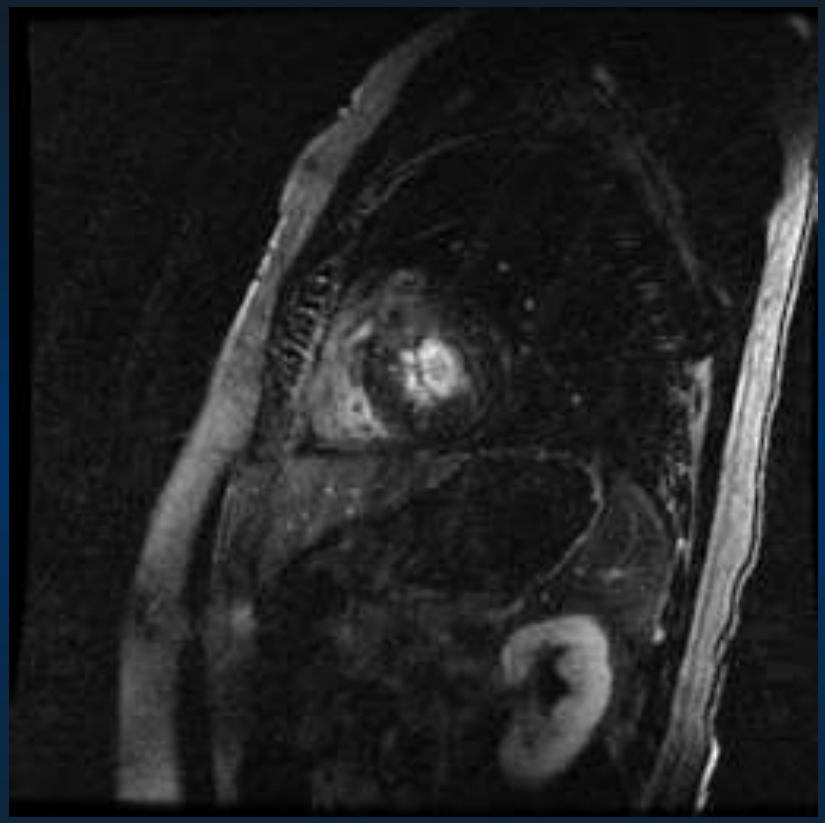
Cardiac MRI: Four Chamber Cine



Asymmetric thickening of septum

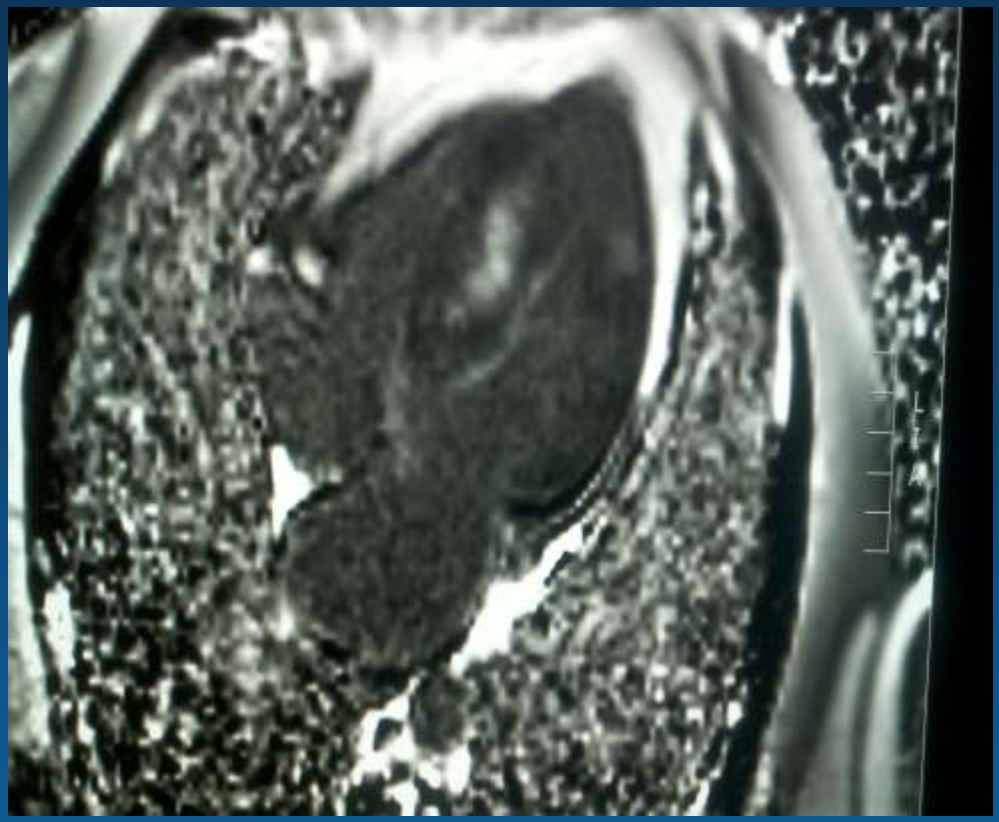
Septum 2.8 cm
Maximal Basal Thickening 3.3 cm

Cardiac MRI : Delayed Enhancement



LGE in mid interventricular wall

CMR: Delayed Enhancement



Case Management

- Started on Metoprolol XL and titrated up to 300 mg daily with resolution of syncope but still had mild symptoms
- Disopyramide was added and titrated up to 150 mg TID with resolution of symptoms
- Decision was made to place ICD
  - based on LV wall thickness just under 30 mmHg, age, significant myocardial fibrosis on MRI and syncope(although likely attributable to LVOT obstruction since resolved with medications)

Conclusions

Transthoracic echo is primary imaging modality for diagnosis of Hypertrophic Cardiomyopathy

Cardiac MRI has emerged as useful complementary tool for diagnosis and risk stratification with the ability to detect and measure extent of fibrosis and provide better assessment of LV wall thickness, mass, and anatomic variation.

In patients with medical refractory symptoms and inducible LVOT gradient >50mmHg. Septal reduction therapy with either surgical myomectomy or alcohol septal ablation can be considered.

ICD is indicated for primary prevention of sudden cardiac death in high risk individuals based on family history of sudden death, unexplained syncope, presence and extent of fibrosis, and wall thickness > 30mm.

Symptom Management