What's all the Hype with HCM?

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Disclosure

- No disclosures

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Definition of HCM

2D echo showing a maximal end diastolic wall thickness of 15mm or greater anywhere within the left ventricle, in the absence of another cause of hypertrophy in adults

More limited hypertrophy (13-14mm) can be diagnostic when present in family members of a patient with HCM or in conjunction with a positive genetic test

JACC: 2020 Dec 22;76(25):3022-3055

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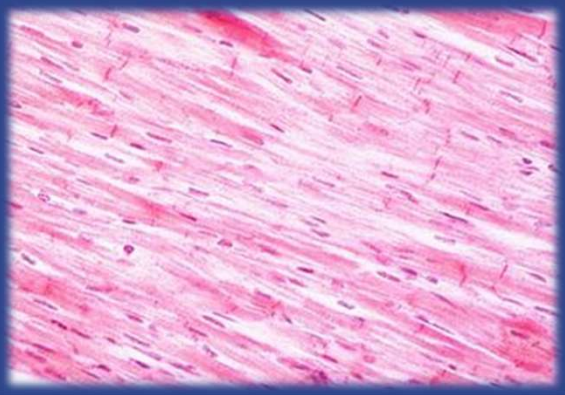
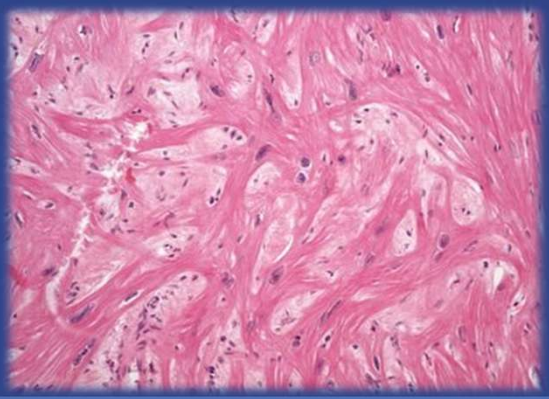
HCM: Histology

Normal Cardiac Myocyte



Note the disarray in the arrangement of the myocytes



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Hypertrophic Cardiomyopathies

ASH without Resting Murmur

LVOTO with classic Murmur

Mid-Ventricular HCM with associated Apical Aneurysm

Apical HCM

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Associations of HCM

- Common - SAM & hyperdynamic LV function
- Other morphologic abnormalities are also not diagnostic of HCM, but can be a part of it:
  - Hypertrophied and apically displaced papillary muscles,
  - Myocardial crypts
  - Anomalous insertion of the papillary muscle directly in the anterior leaflet of the mitral valve (absence of chordae tendinae)
  - Elongated mitral valve leaflets
  - Myocardial bridging-CA segment runs through myocardium & is compressed during systole
  - Right ventricular (RV) hypertrophy

Circulation: Cardiovascular Imaging. 2012;5:441–447

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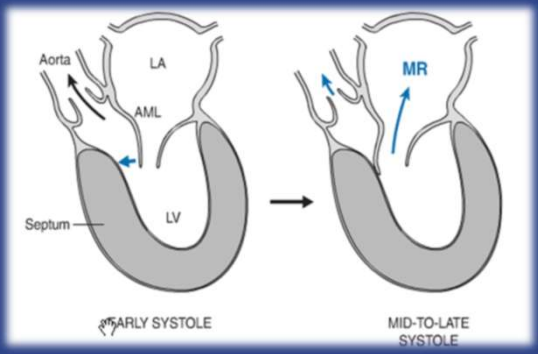
Why HCM Can Develop LVOT Obstruction



- Very thick septum and elongated MVL can lead to abnormal motion of the MV during systole or SAM
- MV gets sucked toward the septum (Venturi Effect), obstructs flow into the aorta

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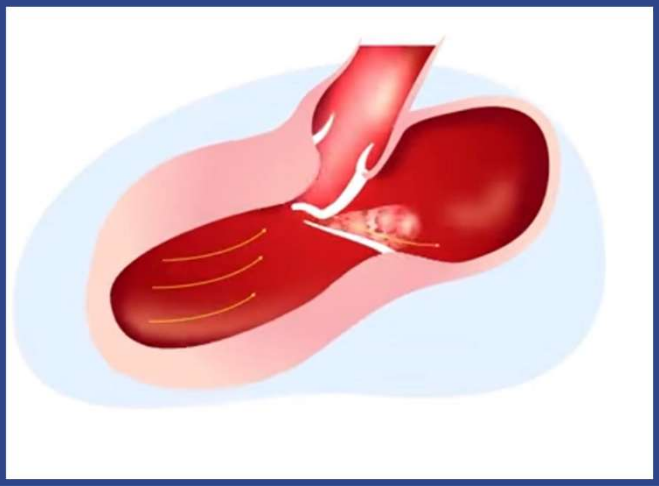
Dynamic LVOT Obstruction



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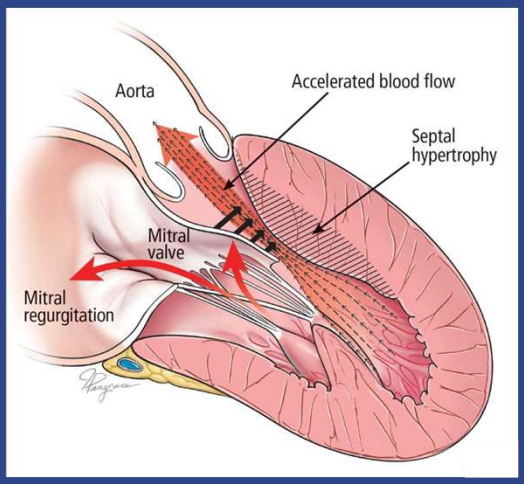
Significant LVOT Obstruction in HCM



- The presence of a peak LVOT gradient of 30 mmHg is considered to be indicative of obstruction
- Resting or provoked gradients >50 mmHg generally considered to be the threshold for therapy

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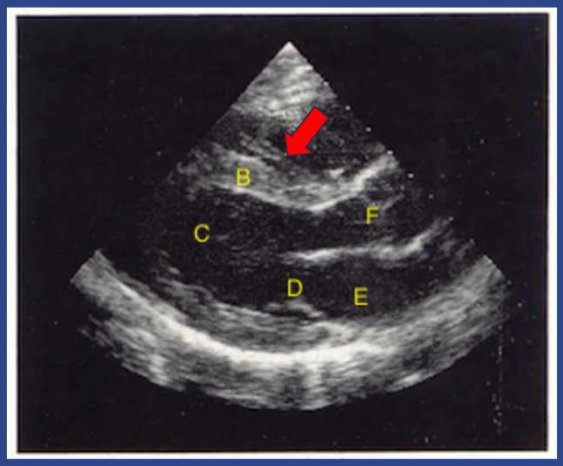
Measuring the Septal Thickness



- Be sure that you are measuring the LV and Not the insertion of the RV band
- Walk the RV band out and exclude it from measurement

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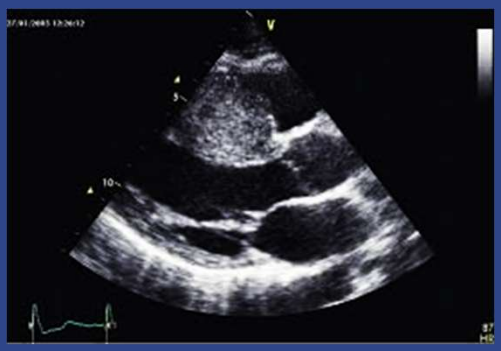
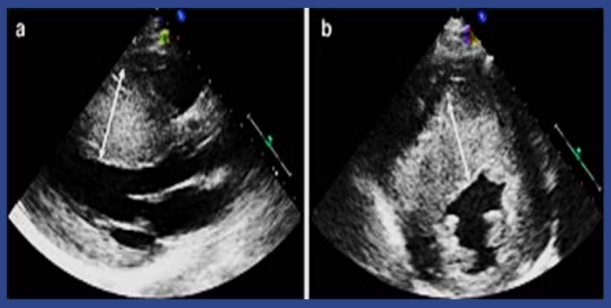
Pitfalls of LV Measurements

- Off axis imaging can lead to an overestimation of the LV septal thickness in a PLAX view





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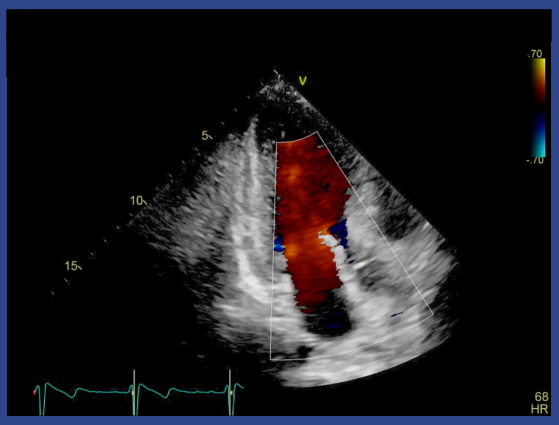
HCM: Asymmetrical Septal Hypertrophy



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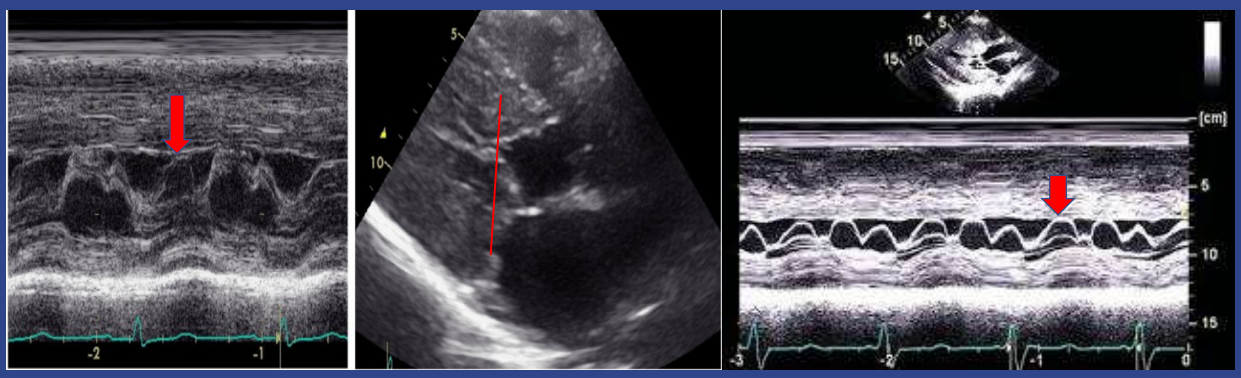
M-Mode: MV SAM



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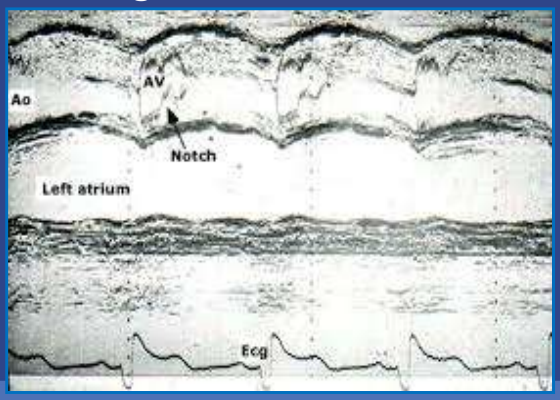
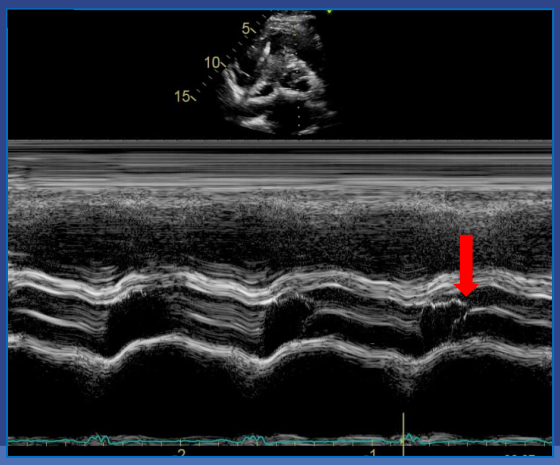
M-Mode Aortic Valve: Notching or Pre-closure

- **Usually more severe when LVOT gradients are elevated**





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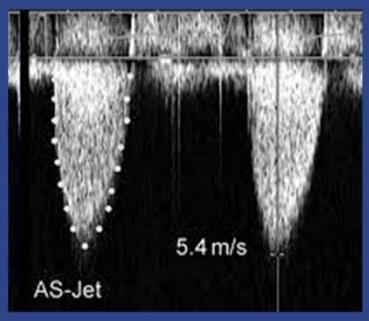
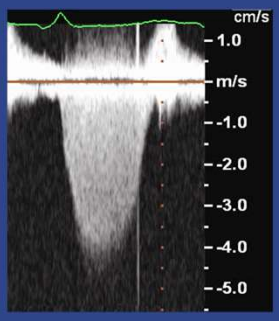
Differentiating Doppler Patterns: AS & MR



Aortic Stenosis



Mitral Regurgitation

St. Louis Arch

- U or V shaped
- Peaks in mid-systole

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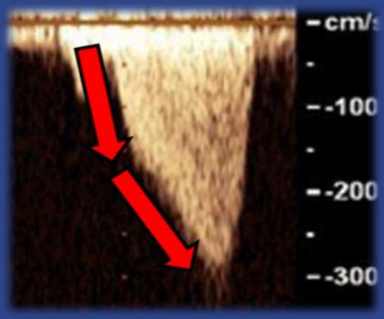
Differentiating Doppler Patterns: LVOT Obstruction



- Often described as dagger-shaped
- Note starts steep, then becomes less steep
- Very late peaking

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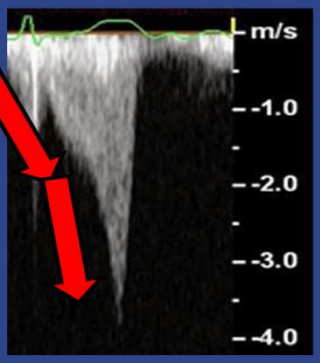
Differentiating Doppler Patterns: Mid-Cavity Obliteration



- Seen in hyperdynamic ventricles
- Starts slower, then accelerates
- Peaks in late-systole

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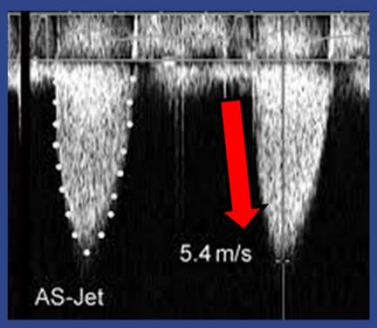
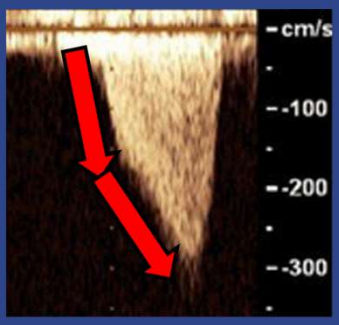
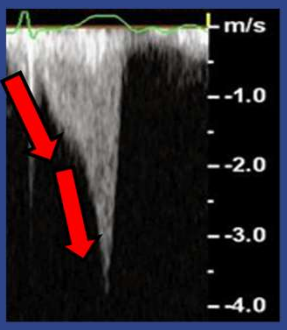
Differentiating Doppler Patterns



Aortic Stenosis



LVOT Obstruction



Mid-Cavity Obliteration

Can be significant variability!

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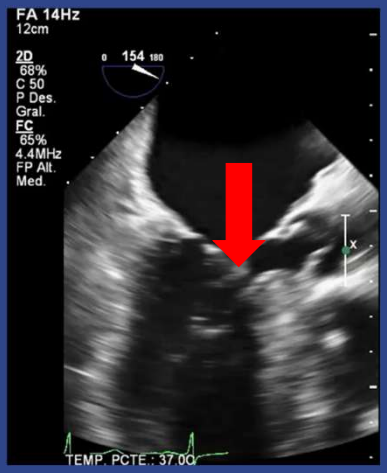
The Source of the LVOT Obstruction



- The resting/dynamic LVOTO comes from the anterior motion of the AMVL
- Any maneuver which decreases LV size will increase the obstruction because the MV and septum are closer together
- Any maneuver which increases LV size will decrease the obstruction because it is separating the MV from the septum

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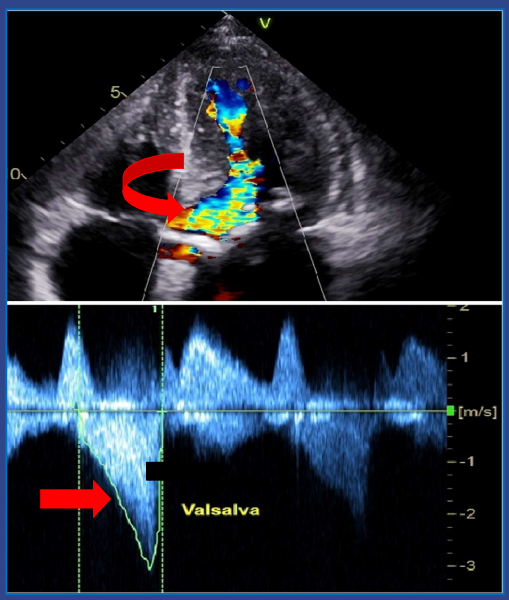
LVOT HCM

Tips:

- Color Doppler helps to localize obstruction
- Place sample volume within turbulent area
- Daggered shaped waveform
- Shape of waveform helps to distinguish between obstruction and MR or AS



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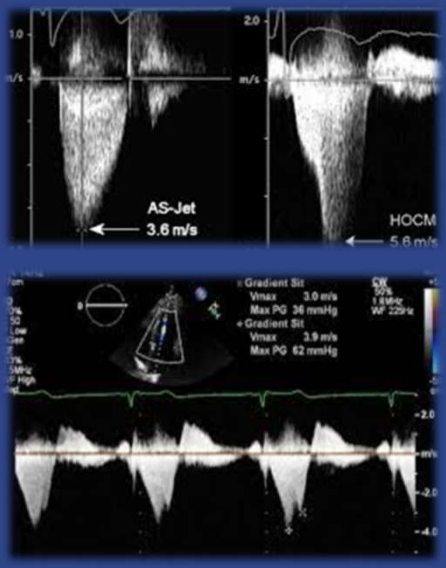
LVOTO with Aortic Stenosis

- Mid peaking vs late peaking
- Difficult to separate with CW Doppler
- Super imposed CW Doppler signal



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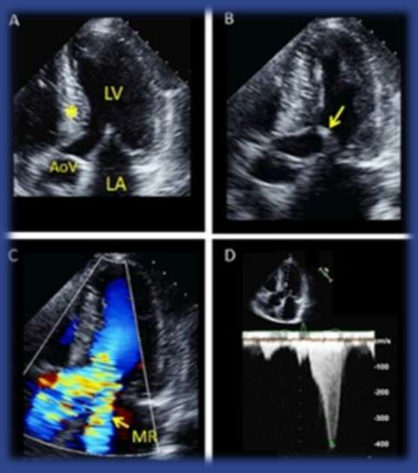
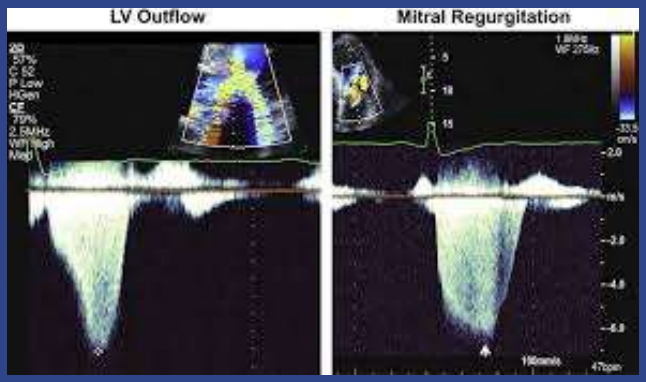
LVOT HCM with MR





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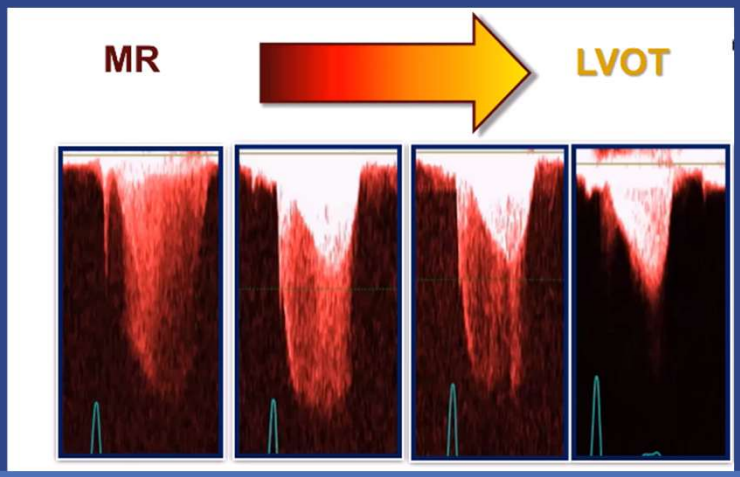
CW Doppler Sweep



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Provocative Testing

- Recommended:
  - In patients with HCM & Resting LVOT gradient <50mmHg TTE provocative maneuvers are recommended
  - Symptomatic patients with HCM without resting or provocable LVOT gradient ≥50 mmHg on TTE, exercise TTE recommended for detection & quantification of dynamic LVOT obstruction

Nagueh SF, Bierig SM, Budoff MJ, et al. The Role of Echocardiography in Hypertrophic Cardiomyopathy. J Am Soc Echocardiogr. 2011 May; 24(5): 473-98.

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Performing a Provocative Maneuver

- Align CW Doppler from an apical 5 or 3 chamber view through the obstructive area in the LVOT
- Use color Doppler for guidance
- Rehearse valsalva maneuver
- Practice staying in gradient with CW Doppler while Valsalva is performed by avoiding taking in a breath
- Text on screen “Valsalva”
- While collecting CW Doppler ask patient to Valsalva and then release

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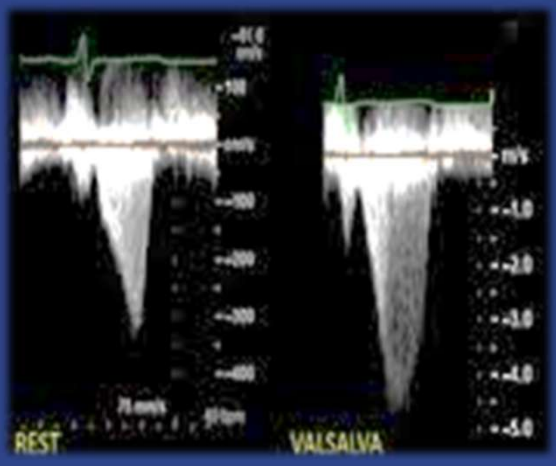
Provocative Maneuver: Valsalva



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Pitfalls of Performing a Valsalva

- Patient inhales/exhales, which moves Doppler out of the plane of obstruction = gradient loss
- Locate obstruction with color Doppler
  - Better localization
  - Helps to avoid MR
- Practice your approach for a Valsalva and coach patient

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Goal Directed Approach

- A goal directed approach to the Valsalva maneuver can be achieved using a blood pressure machine
- Connect a disposable 10-ml syringe or suction tubing to the BP machine
- Instruct the patient to blow into the syringe/tubing and try to maintain a pressure of >40 mmHg for >10-15 seconds
- LVOT gradients should be attempted while holding at 40mmHg

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Procedure:

1. Remove the coiled tubing from the sphygmo (the tubing that it came with).
2. Cut the designated oxygen/suction tubing in half.
3. Attach the non-cut end to the valve on the sphygmo
4. Place the sphygmo in a position so the patient can see the clock face.
5. Give the patient the other end of the oxygen/suction tubing and ask the patient to blow into the tubing until the dial reaches 40 mmHg.

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Procedure cont.

- Practice with the patient, managing inspiration/ expiration so when they prepare to blow and release the image is not lost.
- When ready, have the patient blow, obtaining 40 mmHg, or best effort if can't reach 40 mmHg. The patient must hold for 10 sec for an effective Valsalva.
- If the patient does not reach 40 mmHg, this can be documented, see reporting section.

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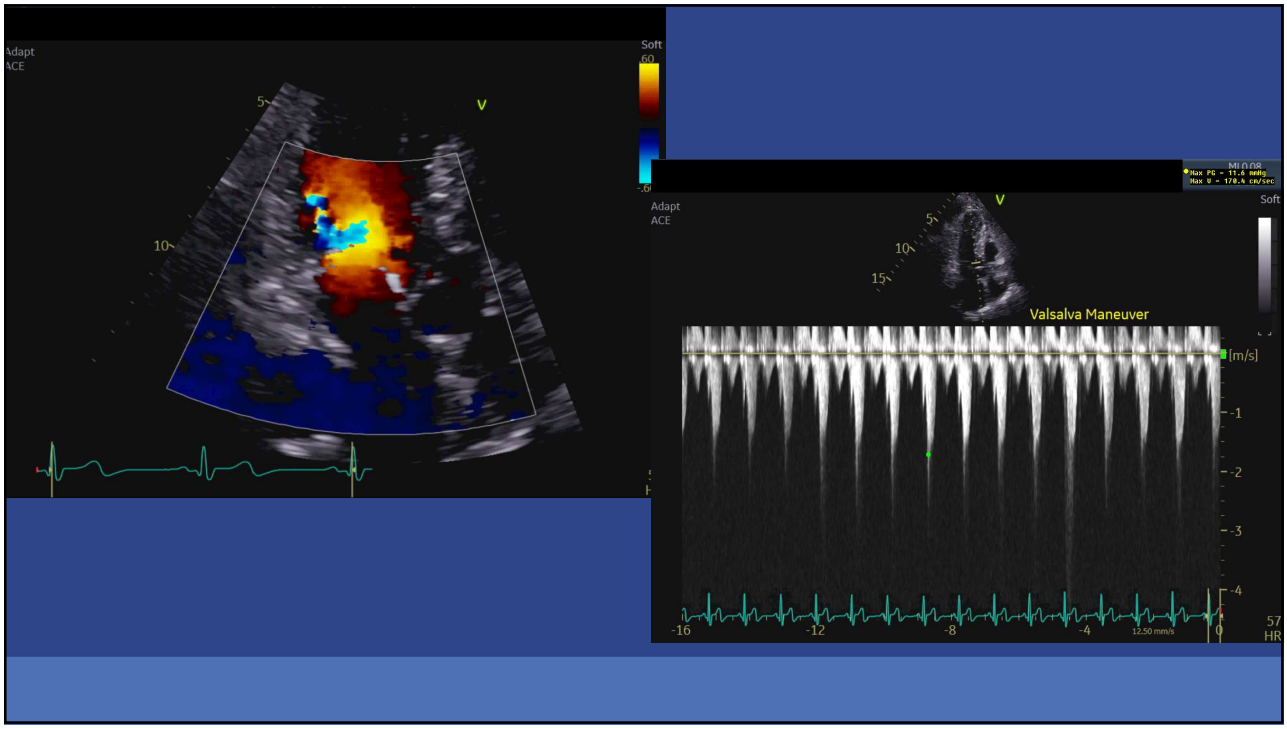

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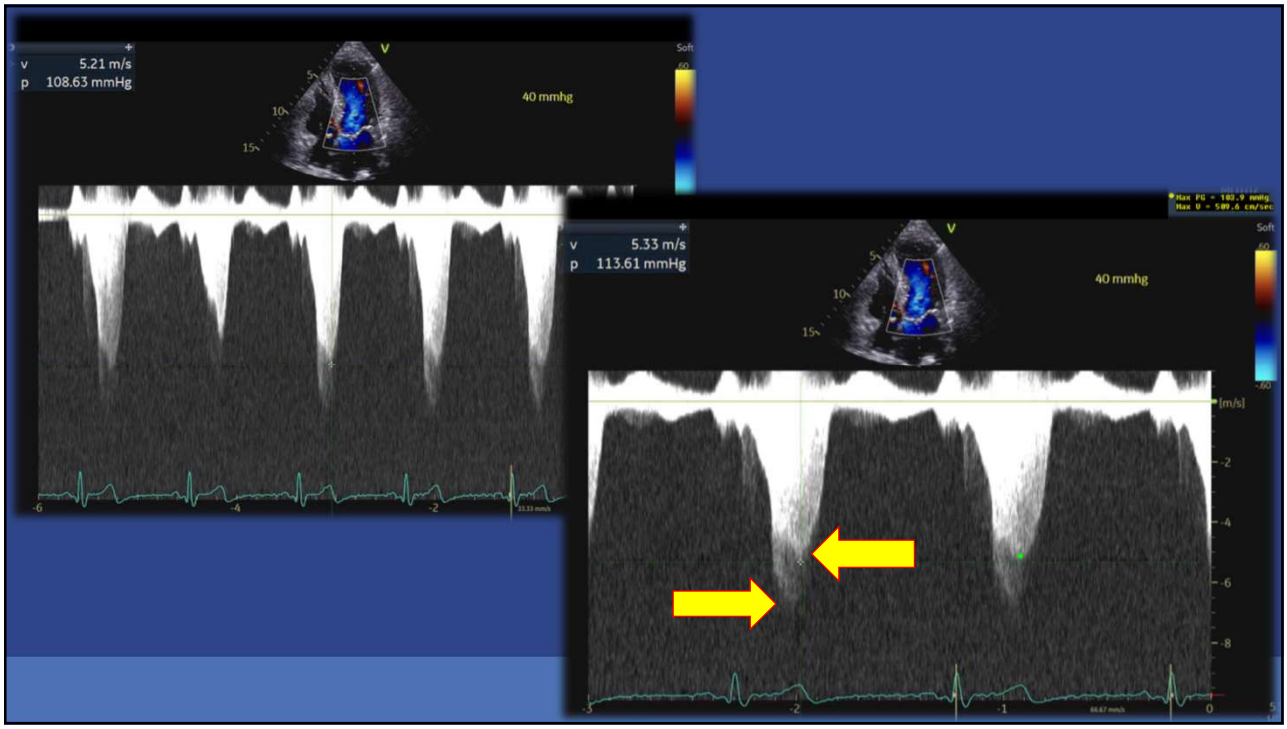

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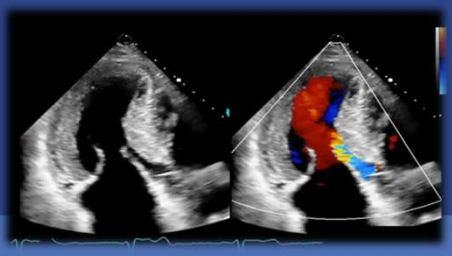
Tips

- Practice the maneuver with the patient prior and determine best probe position to obtain optimal imaging
- Record a loop while the maneuver is being performed with 2D and CFD to determine whether SAM occurs/worsens
- Make sure not in mid-cavity area of obstruction



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Reporting:

- Document pressure achieved and length of hold.
- The patient achieved ___ mmHg of intraoral pressure using the sphygmomanometer and held pressure for ____ seconds.

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How does Exercise Induce LVOTO?

- Exercise decreases afterload, which means LV is Smaller
- Increased cardiac contractility means the LV size is Smaller
- Even though venous return increases with exercise, the decreased afterload and increased contractility result in LVOTO

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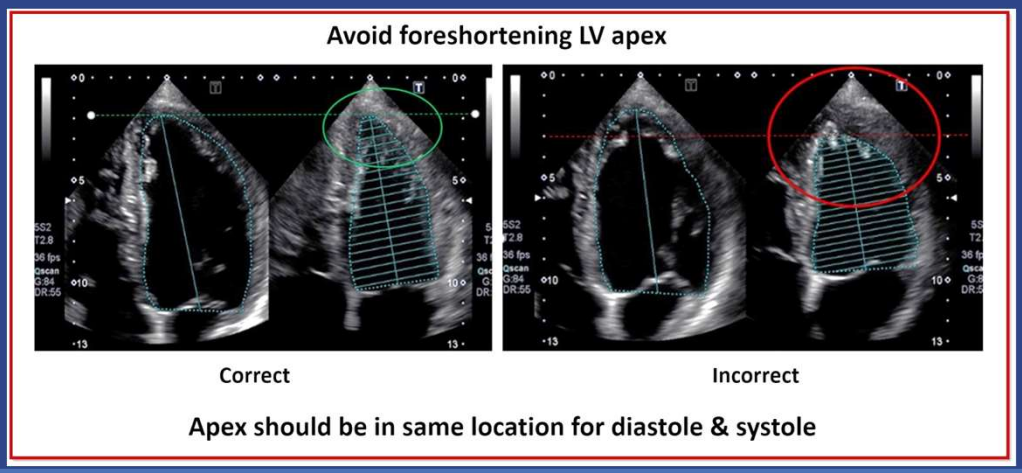
Challenges with the apex



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HCM: Apical

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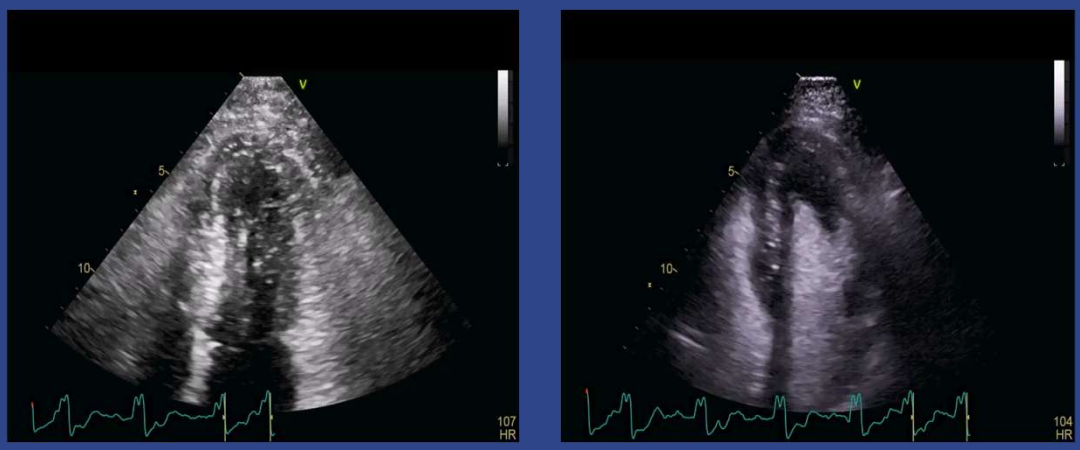
Apical 4 HCM



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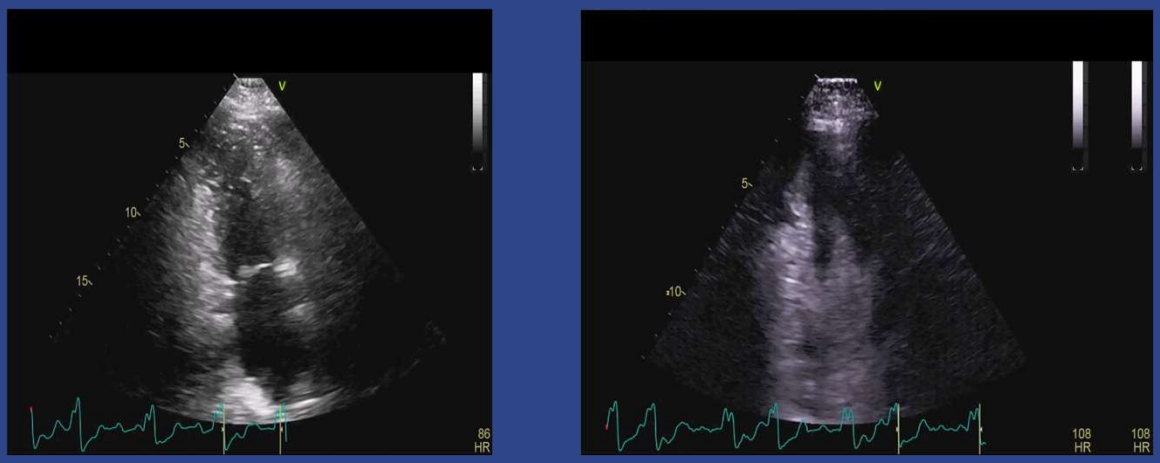
Apical 2 HCM



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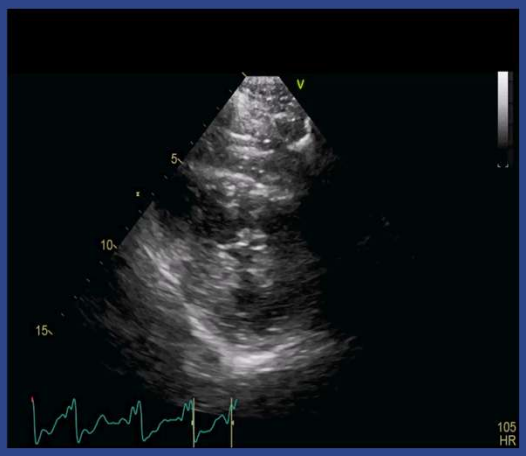
SAX & Apical 4 HCM



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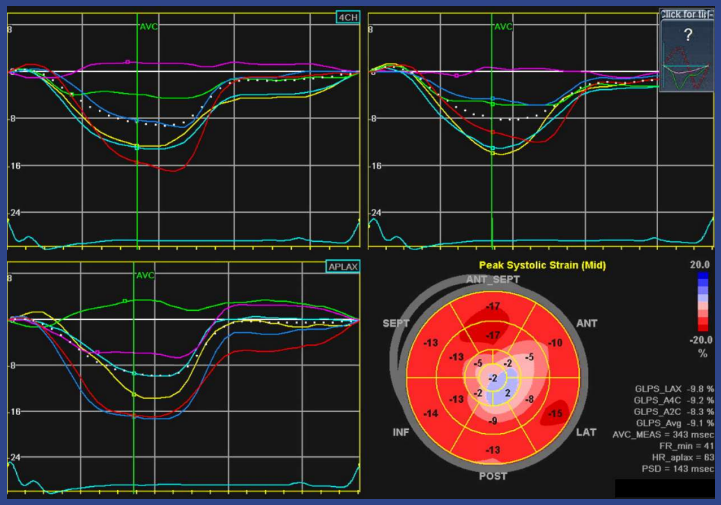
Apical HCM Strain



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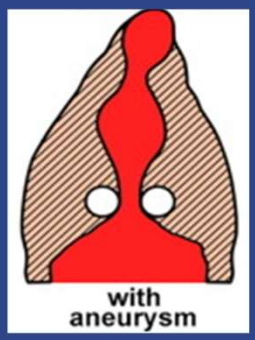
Spectrum Between Apical and Mid-Ventricular



As thickness moves towards base (mid segment), an apical aneurysm often develops

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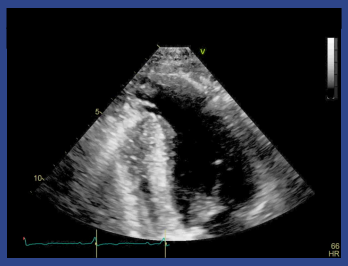
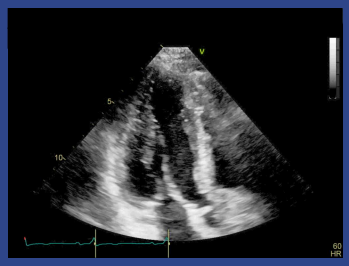
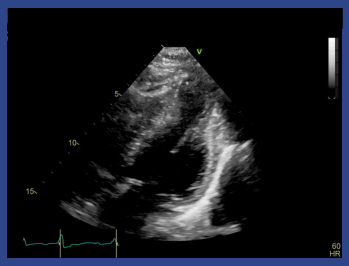
HCM: Mid Ventricular







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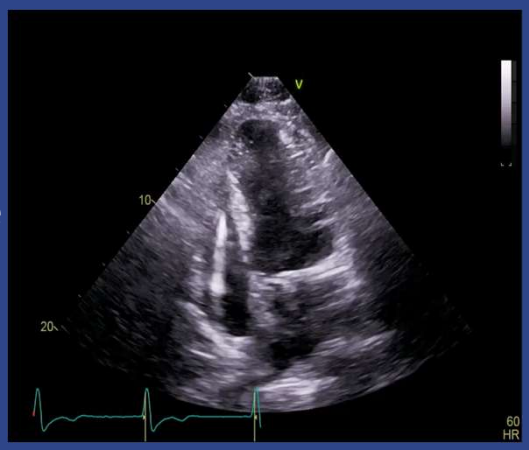
Mid Ventricular HCM

- 72 y/o female
- Chronic diastolic HF due to HCM
- Stage 3 Chronic Kidney disease
- Echo to evaluate LVF



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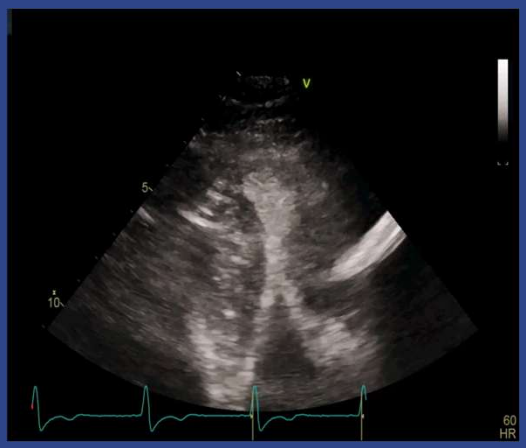
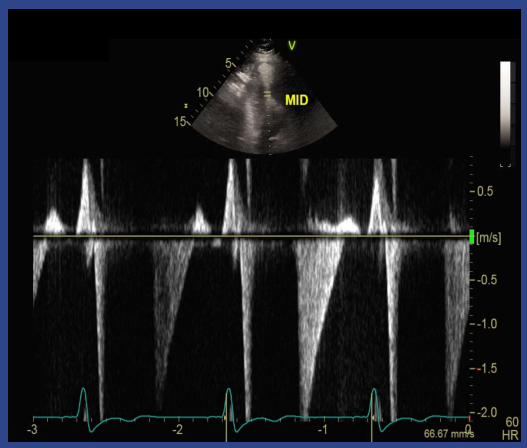
Mid Ventricular HCM





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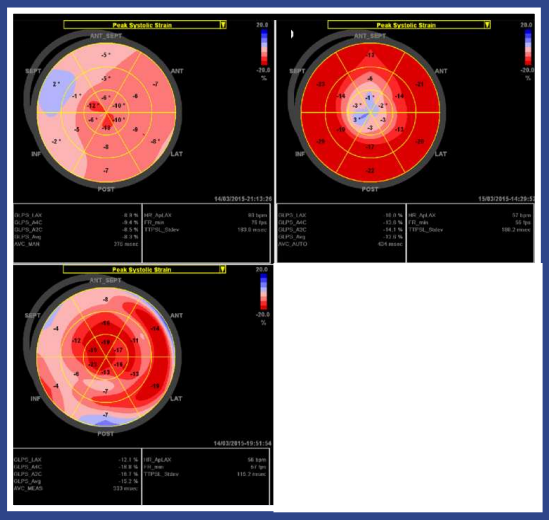
Strain with Hypertrophic Cardiomyopathy

Septal Hypertrophy
76 yo Female
IVSd -18mm
LVPWd 12mm

Concentric Hypertrophy
70 yo Male
LVPWd & IVSd 17mm
LVEF - 60%



Apical Hypertrophy
59 yo Male
IVSd & LVPWd NL
Wall thickness at apex 20mm EF – 60%

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HCM and Contrast

Indicated for endocardial border definition

- **Defines areas throughout the LV**
  - **Basal Septum**
  - **Lateral Wall**
  - **Apex**

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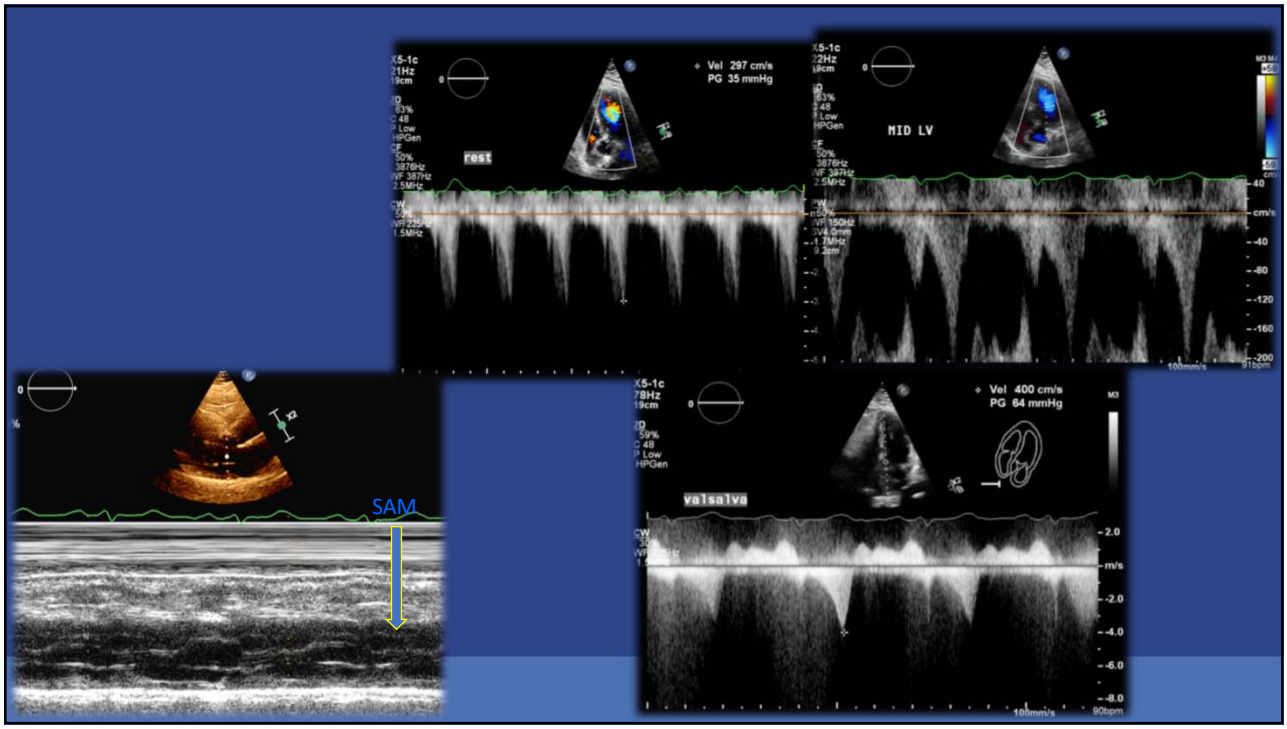

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Additional HCM Protocol

1. Confirm the presence of LVH
    1. Unexplained wall thickness of >15 mm in any segment
    2. M-mode across aortic valve – aortic pre-closure
    3. M-mode across mitral valve - SAM
2. Assessment of LVOT Obstruction
    1. Color Doppler across LVOT from multiple views
    2. Apical 5 or 3 chamber approach with Spectral Doppler (PD & CW)
    3. Addition of a valsalva maneuver in a sitting or standing position and if no gradient provoked attempt them on standing
    4. Addition of goal directed Sypg. at 40mmHg for 10-15 seconds
3. Evaluation for Mitral Regurgitation
    1. SAM results in failure of leaflet coaptation
        1. Mid to late systolic
        2. Inferolaterally oriented
4. Strain
    1. Significantly reduced longitudinal strain
    2. Longitudinal abnormalities often focal or segmental based on location of hypertrophy
5. Contrast
    1. Enhances endocardial borders and Doppler through LVOTO

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A Word of Caution…
Non-HCM Cause of LVOTO

- Hypertensive heart disease
- Post-Cardiac Surgery (Immediate)
- Sigmoid Septum/Basal Septal Hypertrophy
- Hyperdynamic Cardiac Function
- Takotsubo Cardiomyopathy

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Take home message…

- HCM is more common than we think
- Please have high index of suspicion
- Echo is the starting place for HCM diagnosis
- Obtain strain if IVSd > 1.5
- Low threshold for contrast utilization in apical HCM
- Differentiate between MR and LVOT CW jets
- Attempt provocative maneuvers if there is no resting gradient

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