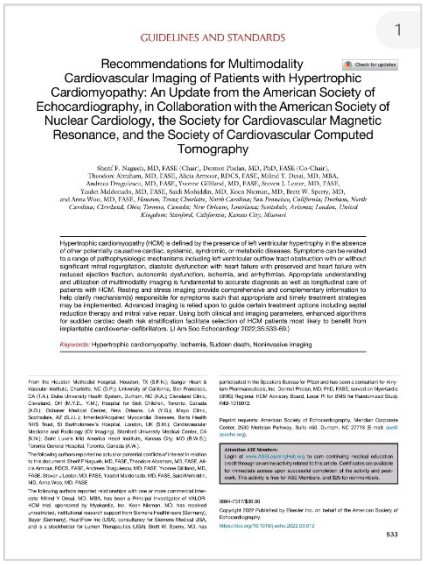
#ASEchoJC Twitter Chat
Tuesday, September 29, 2022 – 8 PM ET


GUIDELINES AND STANDARDS

Recommendations for Multimodality Cardiovascular Imaging of Patients with Hypertrophic Cardiomyopathy: An Update from the American Society of Echocardiography, in Collaboration with the American Society of Nuclear Cardiology, the Society for Cardiovascular Magnetic Resonance, and the Society of Cardiovascular Computed Tomography

- **Recommendations for Multimodality Cardiovascular Imaging of Patients with Hypertrophic Cardiomyopathy: An Update from the American Society of Echocardiography, in Collaboration with the American Society of Nuclear Cardiology, the Society for Cardiovascular Magnetic Resonance, and the Society of Cardiovascular Computed Tomography** (JASE, June 2022)

Authors

- Dermot Phelan, MD, FASE
- Sherif Nagueh, MD, FASE

Moderators:

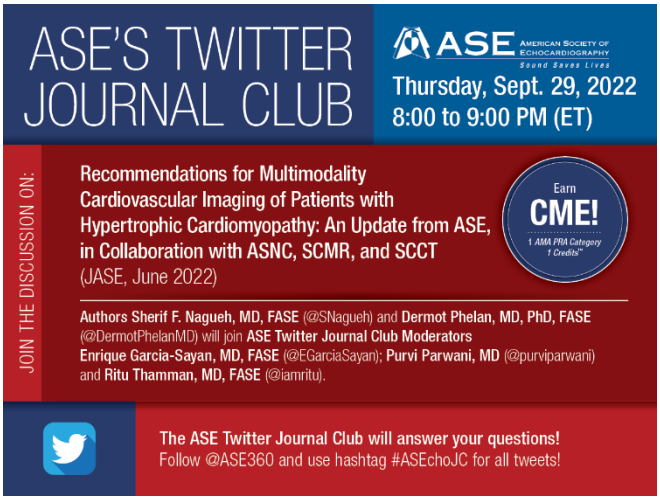
- Enrique Garcia-Sayan, MD, FASE
- Purvi Parwani, MD, FASE
- Ritu Thamman, MD, FASE

Tweetorial: 9-28-2022
https://twitter.com/EGarciaSayan/status/1575090095383715840

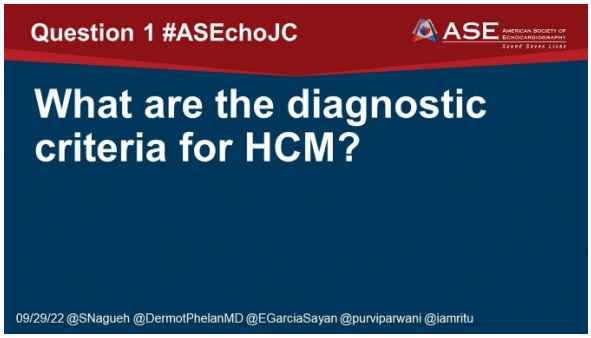
Introduction and Welcome: Welcome to #ASEchoJC! Tonight we'll be discussing the recommendations for multimodality #CVimaging in #HCM with guest authors @SNagueh & @DermotPhelanMD and my co-moderators @iamritu & @purviparwani. 10 questions for discussion follow.

Article:

👉 https://bit.ly/3K4CEVx



Q1:



A1 Notable Responses:

@iamritu: clinical diagnosis of HCM with #echofirst maximal end-diastolic wall thickness of ≥15 mm anywhere in LV, w/o another cause of hypertrophy

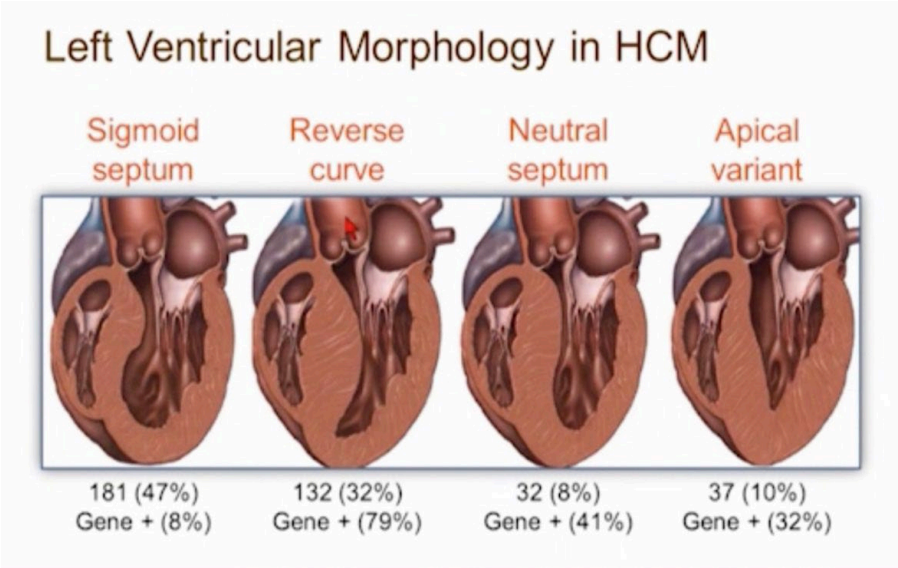
LVH (13–14 mm) can be diagnostic when seen in family members of HCM pt or with a positive genetic test. https://bit.ly/3BM3loV

@iamritu:

🔑Establish diagnosis & determine pattern of hypertrophy

🔑LV morphology in HCM: evaluate in long axis view

Sigmoid septum most common morphology #echofirst in HCM but reverse septal curve most number of genetic mutations https://bit.ly/3ztfRxO



@rajdoc2005: The lower threshold of 13-14 mm for family members/ +genetic test might help not to miss them - since they are at higher risk of disease progression to overt HCM!

@iamritu: This is 🔑! Phenotype +ve HCM patients have ↑risk for SCD even at lesser wall thicknesses. Need to recognize phenotype +ve status (typically wall thickness >15 mm) which allows for proper risk stratification (Holter monitoring, exercise stress testing, etc.)

@DermotPhelanMD:

📌LV end-diastolic wall thickness ≥ 15 mm in absence of other causes of LVH.
📌≥ 13 mm if there is a family Hx or known mutation
📌z-score >2 in children

@DermotPhelanMD:

🔶Important points:
🔺Make sure you only measure the compacted myocardium
🔺Avoid tangential cuts through the LV by confirming on SAX views
🔺Can assess by #EchoFirst # WhyCMR or #YesCCT

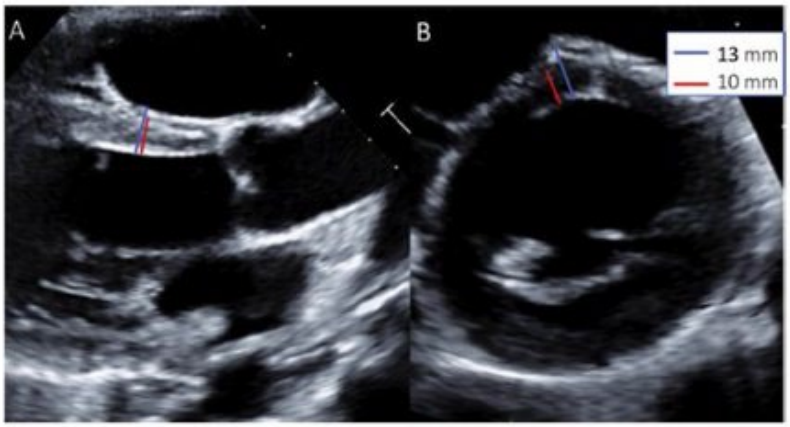


Figure 2 Challenges in measuring septal wall thickness with echocardiography. Female endurance athlete referred for evaluation of possible hypertrophic cardiomyopathy based on interventricular septal wall (IVS) measurement of 13 mm. Panels A and B are representative parasternal long-axis (PLAX) and short-axis (PSAX) images, respectively, which demonstrate inaccurate IVS measurement (*blue line*) that included right ventricular (RV) trabeculation. Comparison of the PLAX and PSAX views can help differentiate the true contractile IVS (*red line*) from RV trabeculation.

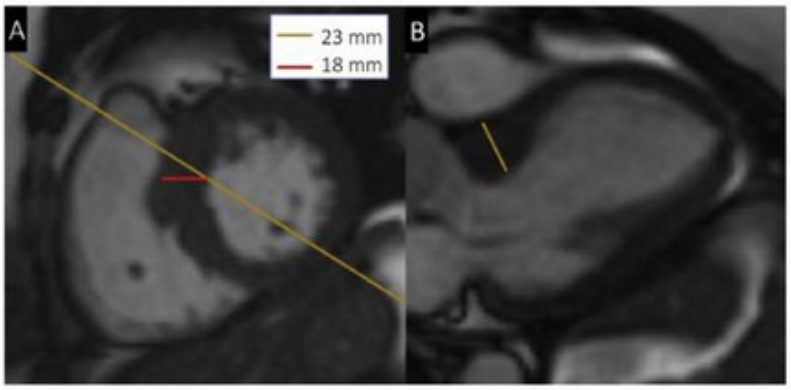


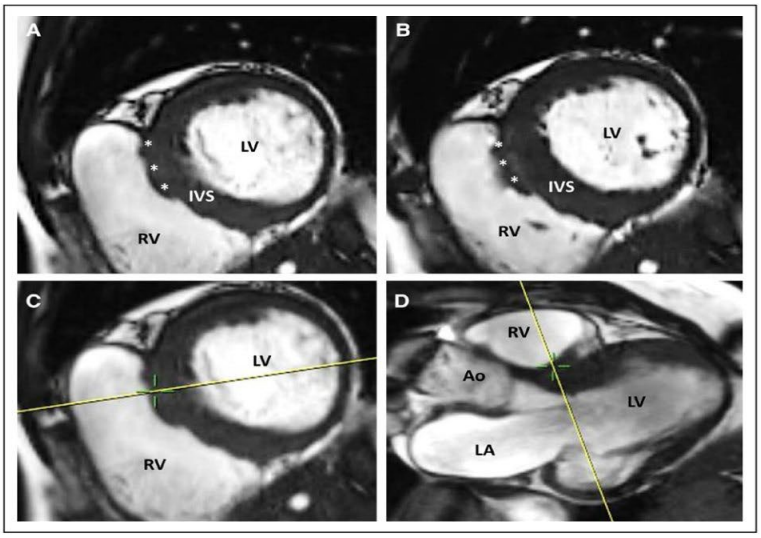
Figure 3 Challenges in measuring wall thickness with CMR. Cardiac magnetic resonance imaging demonstrating how tangential cuts through septum can overestimate interventricular septal (IVS) thickness on long-axis views. In the short-axis view **(A)**, the *yellow line* represents the plane used to obtain the long-axis view **(B)**. This line clearly cuts the muscle tangentially, which will overestimate the septal thickness, while the *red line* represents a more accurate nontangential plane through the compacted IVS thickness.

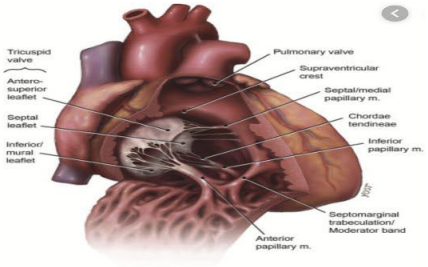
@EGarciaSayan: tips & tricks for LV wall measurement by #EchoFirst

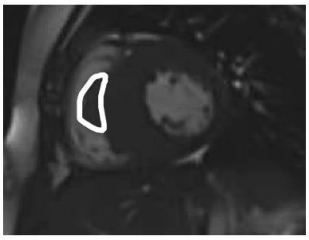
🔺Only measure compacted tissue, avoid LV & RV trabeculations
🔺Measurements performed with UEA & 3D more reproducible and closer to #WhyCMR
🔺Integrate LAX and SAX views

@purviparwani: Important point #ASEchoJC

When measuring LV septum important to exclude ->RV myocardium,

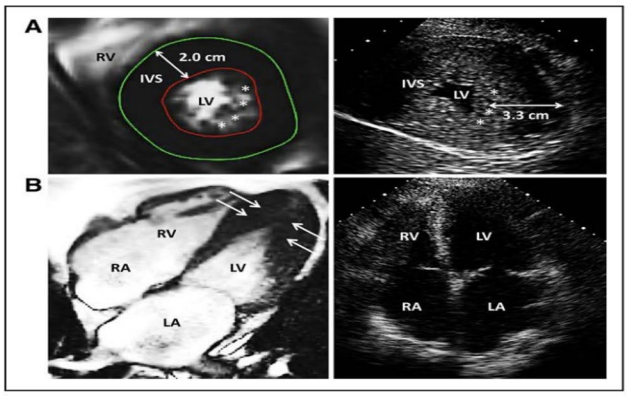






@purviparwani:

->When measuring LV septum important to also exclude LV trabeculation, Papillary Muscle, Apical Septal band -
-> Underestimation of the septal thickness can be due to Focal LV hypertrophy or poor #Echofirst windows, Use contrast

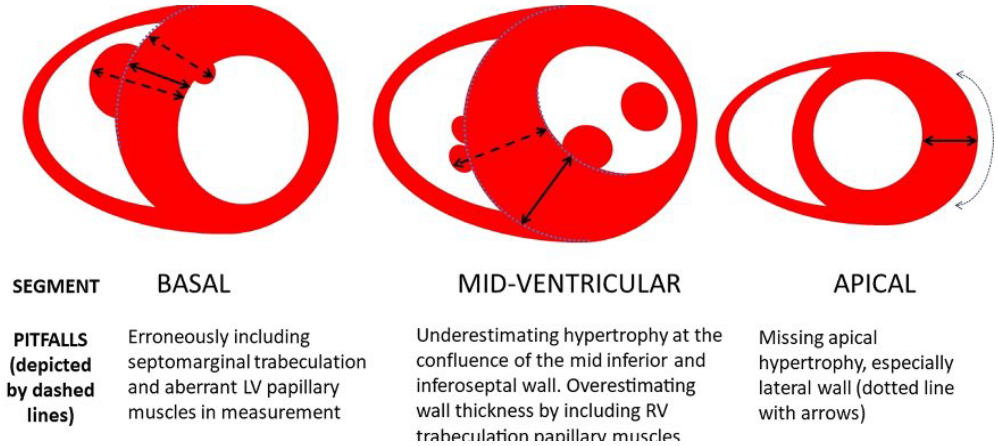


A. Inclusion of LV trabeculation

B. Poor Acoustic windows

@iamritu: This is a 🔑 point!

Measure in short-axis view orthogonal to circumference of endocardium & epicardium
exclude
septomarginal trabeculations
aberrant LV papillary
muscles attached to but not part of septum which could overestimate wall thickness & misdiagnose HCM



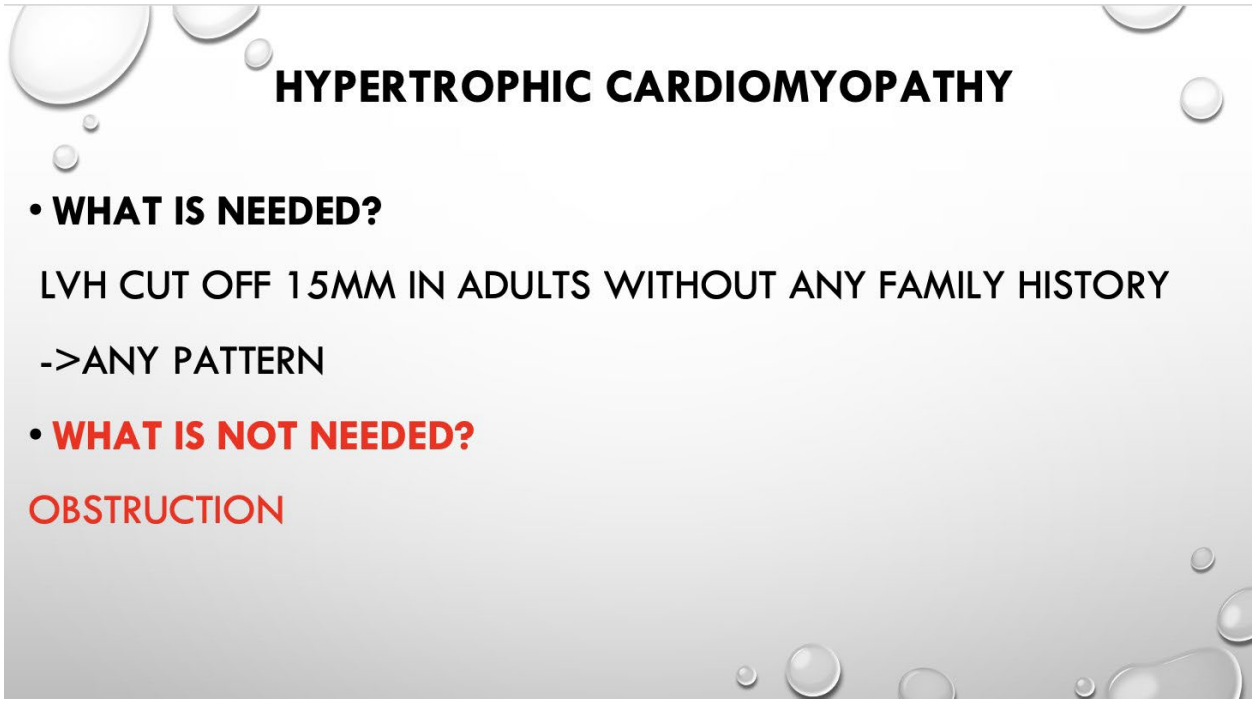
SEGMENT	BASAL	MID-VENTRICULAR	APICAL
PITFALLS (depicted by dashed lines)	Erroneously including septomarginal trabeculation and aberrant LV papillary muscles in measurement	Underestimating hypertrophy at the confluence of the mid inferior and inferoseptal wall. Overestimating wall thickness by including RV trabeculation papillary muscles	Missing apical hypertrophy, especially lateral wall (dotted line with arrows)

@evolutsapien: What is the proper location to measure septum in an elderly patient with sigmoid septum?

@boegel_kelly: I usually measure just distal to the sigmoid area for my IVSd and then make a notation in report detailing sigmoid septum that is ___ cm at its largest diameter, with comment whether it causes any obstruction to flow in LVOT

@GWhalleyPhD: Me too

@purviparwani: cutoff of 15mm without any family history. Important to remember for the diagnosis what is needed is LVH with cut off in any pattern. What's not needed for diagnosis is obstruction.

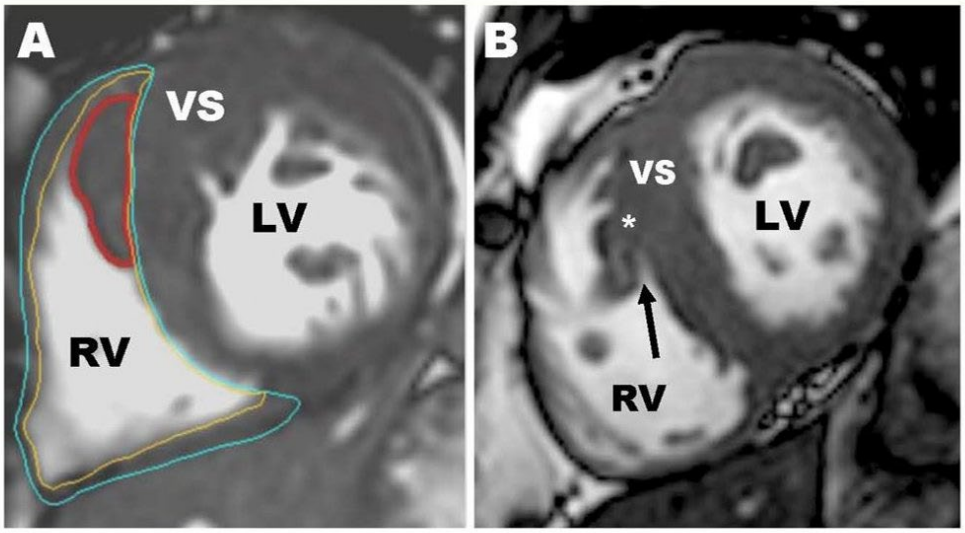


@rajdoc2005: 100% agree. HCM need not be H"O"CM!

@DavidWienerMD: Important point: HCM, not HOCM. Names matter!

@DermotPhelanMD: Beware RV trabeculations/crista supraventricularis!!

Crista Supraventricularis in HCM
Source of Wall Thickness Overestimation by MRI

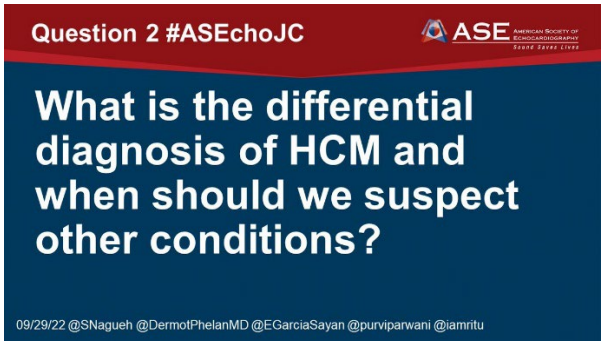


Maron MS. *Journal of CV Magnetic Resonance* 2012, 14:13.

@LP_DavidMD: Totally agree with everything that has been said. Would only add that for apical HCM, sticking with the cutoff of 15 mm will potentially lead to missed diagnosis. Lack of decreased LV wall thickness from base to apex should raise concerns between 10-15 mm

@danilorenzatti: Agree with @LP_DavidMD. Sometimes like in Apycal HCM you can't rely in the 15 mm cut-off. We should be aware the the myocardial thickness always get thinner towards the apex so having a septal apical segment of 11 mm is NOT normal. Apical relative hypertrophy

Question 2:



A2 Notable Responses:

@EGarciaSayan: #HCM differential outlined in Table 1:

⚠Athlete's heart, cardiac amyloidosis, HTN heart disease, Friedrich's, Fabry, PRKAG2, Danon may all mimic HCM.
⚠Need good history & ECG!
⚠How can #EchoFirst (with GLS) and multi-modality imaging (#WhyCMR #YesCCT) help?

Table 1 Typical Features and Findings in Phenocopies of HCM

Most Common Phenocopies	Clinical Features	Typical ECG Findings	Typical Echo Findings	Typical CMR Findings	Genetics and Additional Features
Children/Adolescents					
Danon Disease	Mild skeletal myopathy, ophthalmic abnormalities, intellectual disability.	May reveal pre-excitation syndrome.	Massive concentric LVH, occasionally dilated cardiomyopathy.	LVH is often severe. LGE can be extensive, but often conspicuously sparing of the mid septum	X-linked dominant disorder, although isolated cardiac form can present in older females. Diagnosis based on elevated CK, muscle biopsy, genetic testing (LAMP2 gene mutation)
Adults <40 years					
PRKAG2	Proximal myopathy, myalgia, epilepsy, early-onset hypertension	Pre-excitation syndrome, bundle branch block, high voltages. Atrial fibrillation, atrial flutter. Advanced atrioventricular blocks, marked sinus bradycardia, or sinus block	Variable degree of increased LV wall thickness. Diastolic and systolic dysfunction	Highly variable findings from minimal asymmetric hypertrophy without LGE in early stages to severe hypertrophy with extensive LGE in advanced stages	Autosomal dominant, PRKAG2 gene mutation
Friedrichs Ataxia	Progressive ataxia, loss of deep tendon reflexes, motor weakness, cerebral dysarthria, diabetes mellitus	Lateral T wave flattening or inversion. Supraventricular and ventricular arrhythmia	Mild concentric remodeling, followed by hypertrophy, less often eccentric, hypertrophy. Impaired relaxation. Ultimately dilatation with systolic dysfunction. Sparkling texture.	In early and intermediate disease: concentric remodeling or hypertrophy. In late disease: replacement fibrosis.	Autosomal recessive, serum alpha-tocopherol level, brain MRI
Anderson- Fabry Disease	Multi-system disease: peripheral neuropathy, cutaneous lesions, progressive renal insufficiency with proteinuria, coronary small vessel disease.	LVH with repolarization abnormalities, conduction abnormalities, preexcitation, atrial and ventricular arrhythmia.	Concentric, asymmetric, and eccentric hypertrophy. Impaired relaxation. Normal ejection fraction. Thinned basal inferolateral LV wall in advanced disease. RV hypertrophy. Prominent papillary muscle. Aortic dilatation.	LGE typically involves the mid segments of the lateral wall with subendocardial sparing. Involvement of the basal third of other LV walls in severe cases. Short T1 relaxation time may be present in the septum.	X-linked recessive, deficiency of alpha-galactosidase A activity. Males present at a younger age.
Athlete's Heart	Asymptomatic	Sinus bradycardia, LVH, early repolarization, first degree heart bock, Wenckebach, ectopic atrial or junctional rhythm.	End-diastolic wall thickness typically below 15 mm. Balanced four chambers dilation. Normal/low-normal biventricular ejection fraction with normal/ supranormal diastolic function.	LGE absent except occasionally at the RV insertion points. Normal ECV.	Supranormal functional capacity.

@LilyLeiZhang1: Also tumors ie lipoma, sarcomas, metastatic disease, melanoma, lymphoma can invade the LV

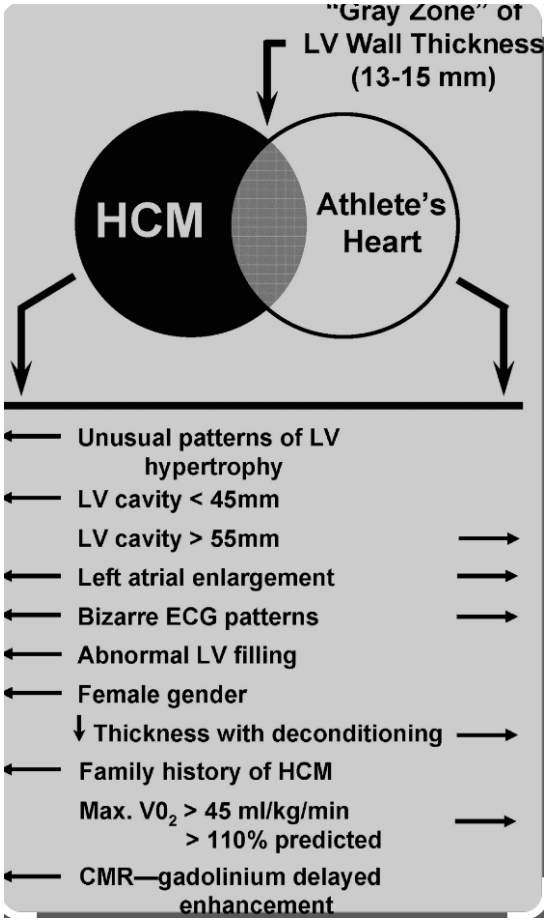
@purviparwani: #HCM and Athletes Heart

IMPORTANT to keep in mind

🍩 Unusual pattern of LVH
🍩LV Cavity <45 in HCM
🍩 LAE
🍩 Abnormal EKG
🍩 Abnormal LGE
Can be tricky, in the example left is athlete, right is HCM

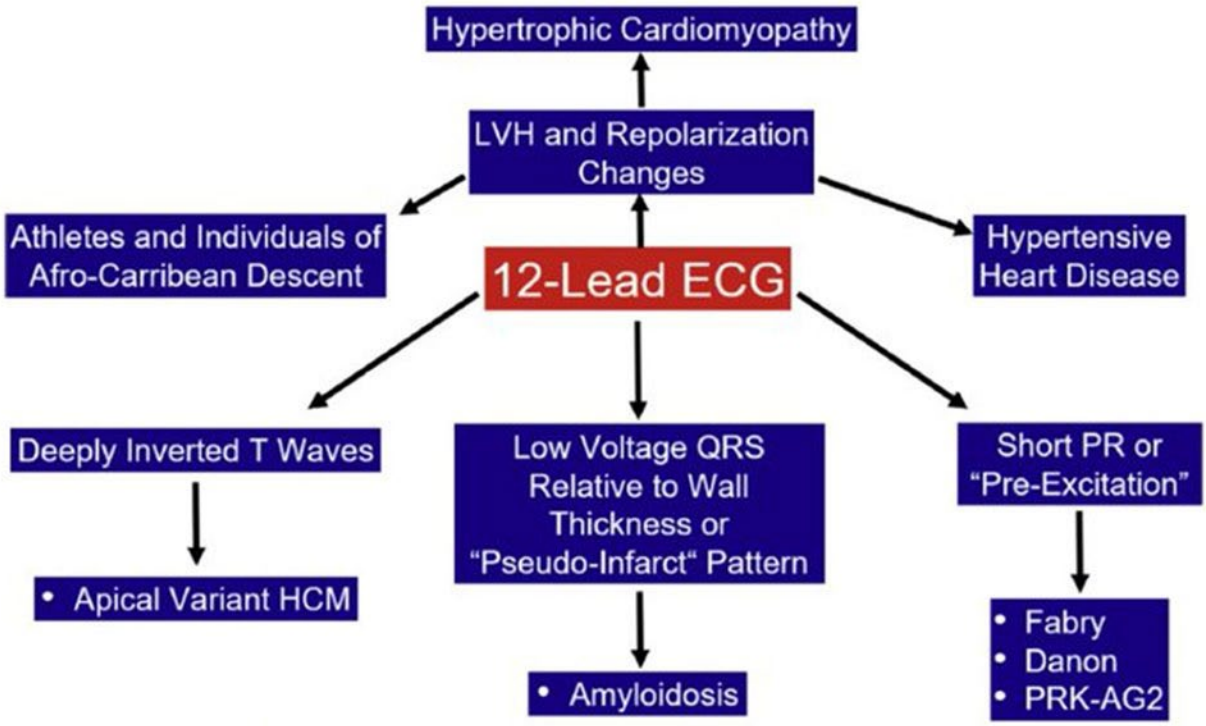
use #WhyCMR

Link: https://ahajournals.org/doi/full/10.1161/01.cir.91.5.1596



@DermotPhelanMD:

🤔Think – what causes increased wall thickness?
🔶Afterload: HTN Heart disease, AS, Athlete's heart
🔶Storage/Infiltration: Amyloid, AFD, Danon, PR KAG2
🆘ECG can help!



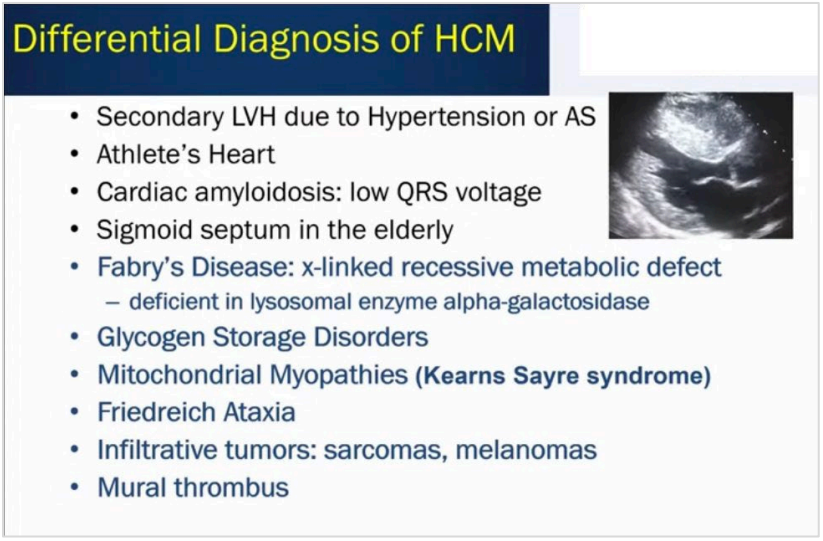
@iamritu: Infiltrative storage dx

Glycogen storage dz:ie Danon in adults -LVH

Fabry:HCM/SAM

Common BSH, a portion do have sarcomeric mutations, but most do not. https://ncbi.nlm.nih.gov/pubmed/31699273

Rare: marked ASH, mimicking HCM from pheochromocytoma



@LilyLeiZhang1: One of favorite board's questions is deposition disorders such as amyloid, sarcoidosis and Fabry's … also for ? Apical HCM, rule out Loeffler

@purviparwani:

#whyCMR can help with differentiation

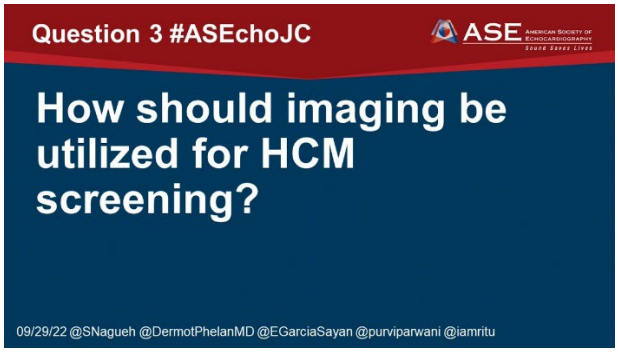
-> Think Fabry's or Dannon when massive LVH without LGE or non-septal LGE
-> think of Fabry when lateral LGE on #WHYCMR (NOT THE THICKEST PORTION) and Low mapping values
->https://jcmr-online.biomedcentral.com/articles/10.1186/s12968-016-0233-6
Case of Fabry disease on #whyCMR

https://twitter.com/i/status/1575641882079608832

@rajdoc2005: Yes. Good to remember the T1 values are low in Fabry's. Can help differentiate from HCM

@vidhu_anand: Very important point as all other reasons for hypertrophied LV lead to increased T1

Question 3:



A3 Notable Responses:

@DermotPhelanMD:

#EchoFirst is first line.
#WhyCMR is complementary if suspicion raised on Echo
@mmartinezheart

TABLE 6 Screening With Electrocardiography and 2D Echocardiography in Asymptomatic Family Members*

Age of First-Degree Relative	Initiation of Screening	Repeat ECG, Echo
Pediatric		
Children and adolescents from genotype-positive families, and families with early onset disease	At the time HCM is diagnosed in another family member	Every 1-2 y
All other children and adolescents	At any time after HCM is diagnosed in a family member but no later than puberty	Every 2-3 y
Adults	At the time HCM is diagnosed in another family member	Every 3-5 y

*Includes all asymptomatic, phenotype-negative first-degree relatives deemed to be at-risk for developing HCM based on family history or genotype status and may sometimes include more distant relatives based on clinical judgment. Screening interval may be modified (e.g., at onset of new symptoms or in families with a malignant clinical course or late-onset HCM).

ECG indicates electrocardiogram; Echo, echocardiogram; and HCM, hypertrophic cardiomyopathy.

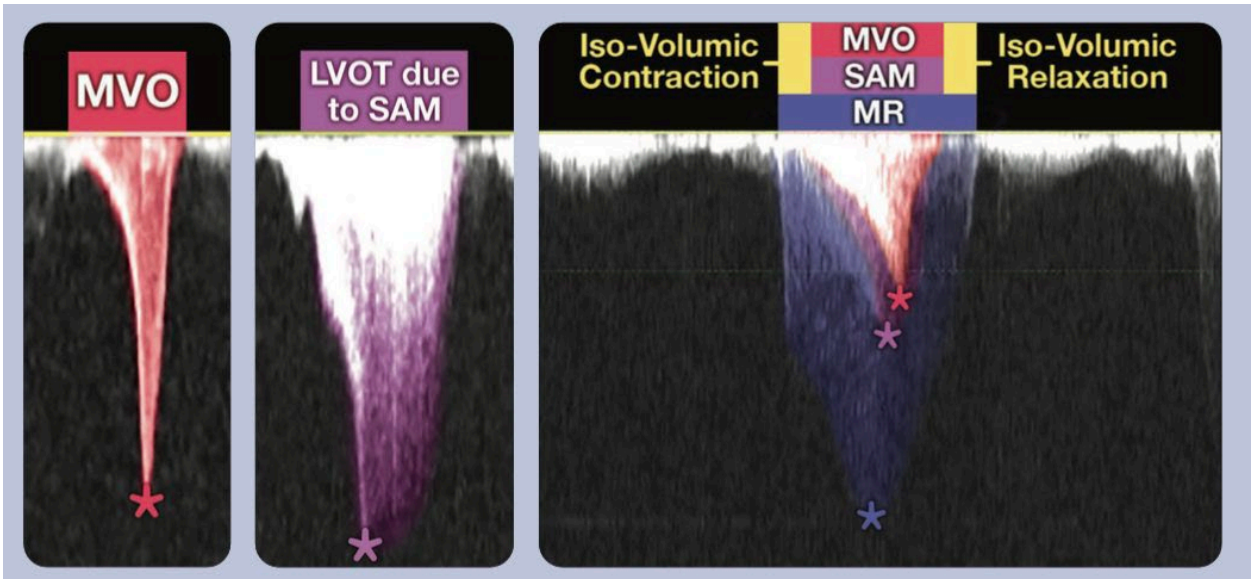
J Am Coll Cardiol. 2020 Dec 22;76(25):e159-e240.

> **@SunthankarMD:** This table is a go-to resource anytime I see a pediatric patient in clinic with family history of HCM

@rajdoc2005: Practices differ. We get a #WhyCMR in almost all our HCM patients as a routine standard practice from our @jct_ucb HCM clinic!! There is something new we learn from many cases on #WhyCMR

@iamritu:

On #echofirst Look for presence/severity of LVOT obstruction
May have contamination of LVOT signal with MR
MR velocity is higher & signal is of longer duration
(spans isovolumic contraction & relaxation) vs LVOT signal



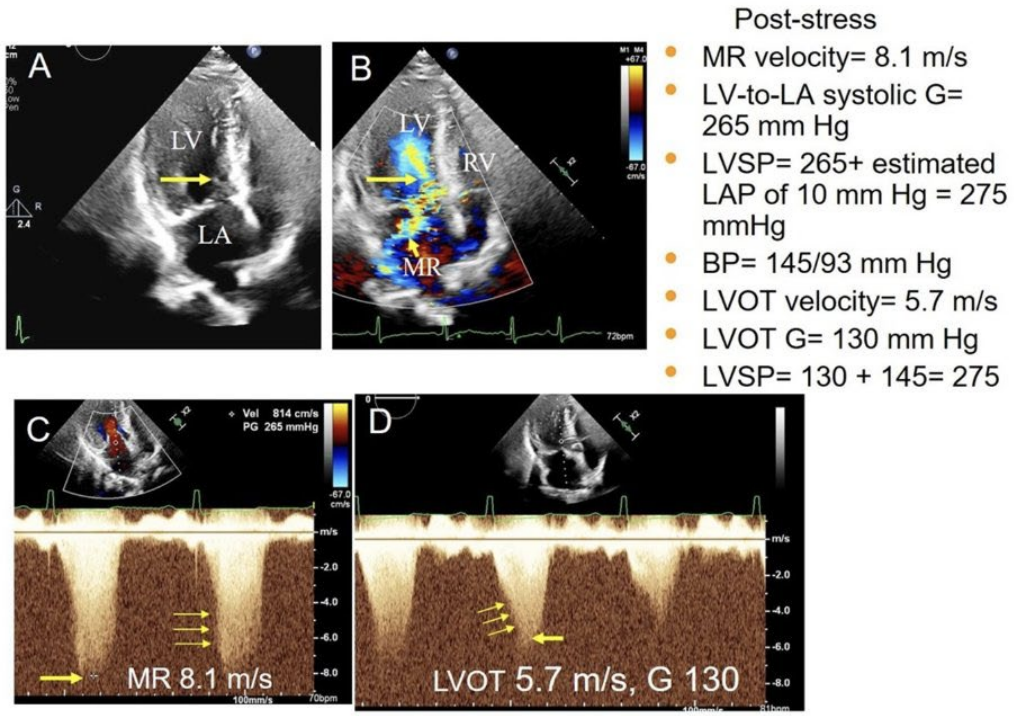
@purviparwani: VERY IMPORTANT point!

Avoid contamination with MR #Echofirst #ASEchoJC

Use BP to your advantage:

See an example: https://acc.org/Education-and-Meetings/Patient-Case-Quizzes/2022/04/12/17/17/Measuring-Left-Ventricular-Outflow-Tract-Signal-Gradient-in-HCM

Figure 7



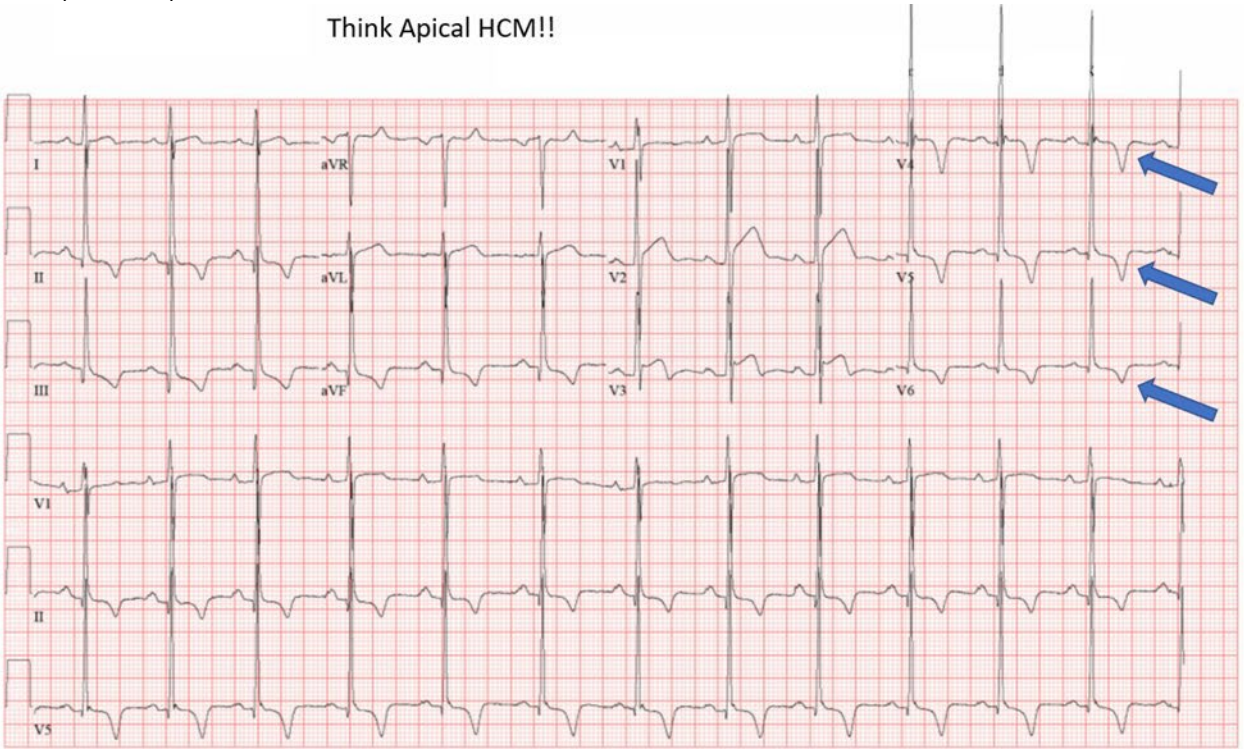
(Panel A) Apical four-chamber view (arrow pointing toward SAM). (Panel B) Color Doppler showing severe LVOT turbulence and moderate to severe posterolaterally directed MR. (Panel C) MR signal with peak velocity and gradient. (Panel D) LVOT signal with peak velocity and gradient. The middle signal shows the accurate LVOT signal; the first signal is likely mixed with the MR and the third signal appears incomplete (thin yellow arrows define the characteristic envelope shapes of both the MR and LVOT signal).

@vidhu_anand: Also very important to different LVOT from mid cavitary obstruction. The color Doppler can be very helpful. Also imp to remember there can be multiple levels of obstruction

It can be really challenging to differentiate the levels of obstruction but it has therapeutic implications - response to medication vs myectomy/ septal ablation

@DermotPhelanMD:

⏰If screening athletes with ECG - be wary of ECG with deep lateral TWI.
🆘Liberal use of UEA to make sure we don't miss apical HCM.
#WhyCMR is your friend



@rajdoc2005: Its a good idea to think of Apical HCM in the presence of these EKG changes - even in non-athletes! We see tons of patients with Apical HCM in their 60s @jct_ucb - diagnosed for the first time on #EchoFirst with #UEA. #WhyCMR helps as well!

@jct_ucb: And don't forget to give the patient a copy of their ECG with their apical variant HCM diagnosis documented on it in case they end up in an ED

@DavidWienerMD: I suggest they take a photo and keep it on their cell phone

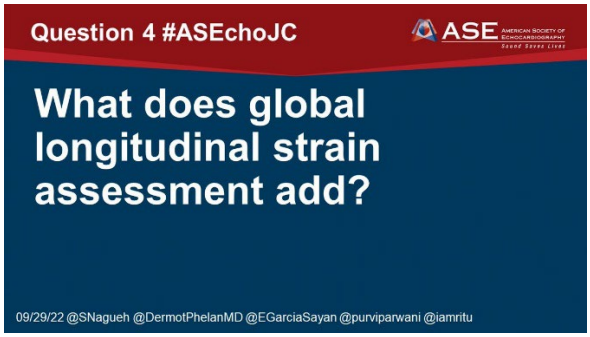
@kgzimmerman: Any tips for strain with thick hearts?

@DermotPhelanMD: Hypertrophy causes reduction in strain. Important to avoid apical foreshortening. ROI can be challenging if very asymmetric - follow the endocardium as much as you can.

@purviparwani: Few points to remember #ECHOFIRST #cvImaging

- Careful with OVER or UNDERESTIMATE the thickness. Use SAX measurements
- Rare to have massive LVH without any LGE
- Risk is incremental with LVH thickness
- Look for Crypts, thick papillary muscle
- Careful with OffAxis

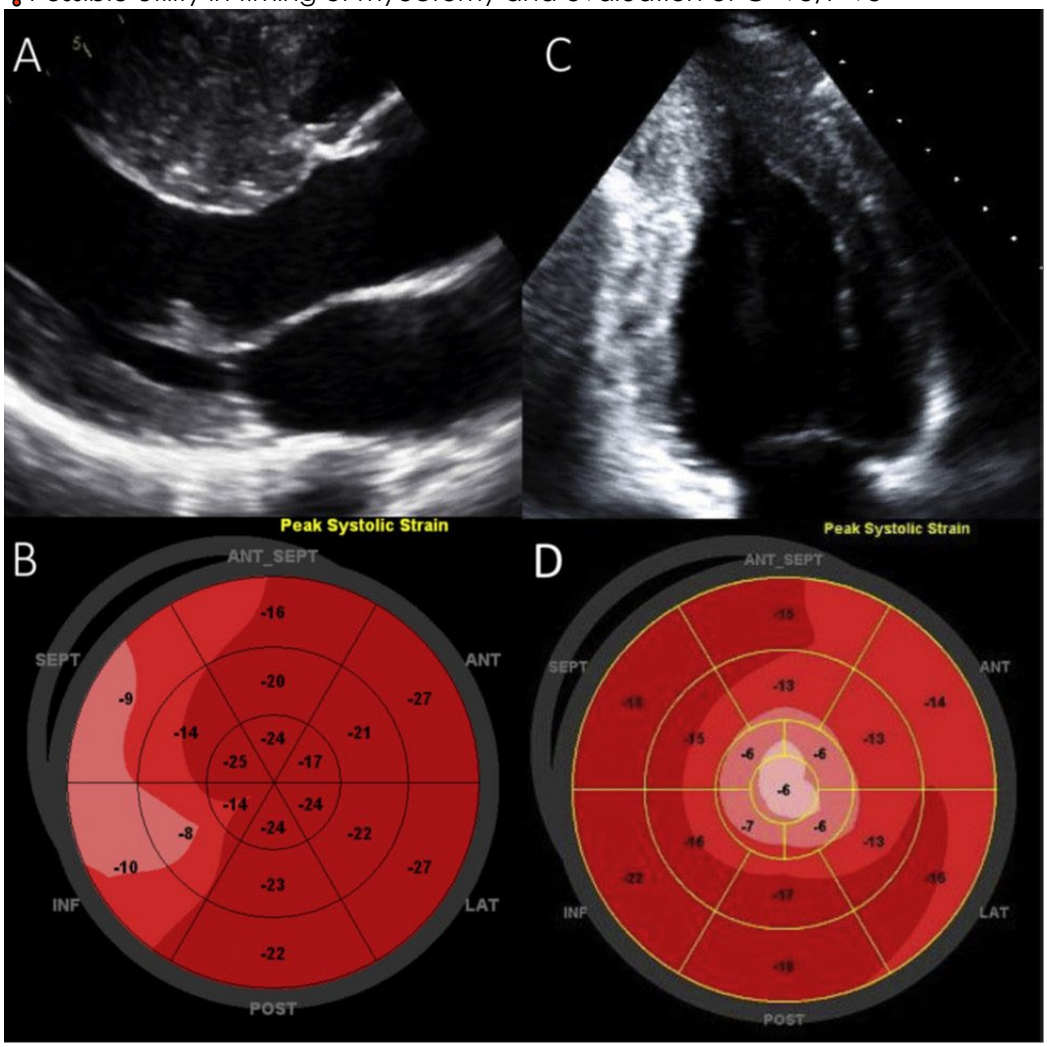
Question 4:



A4 Notable responses

@DermotPhelanMD:

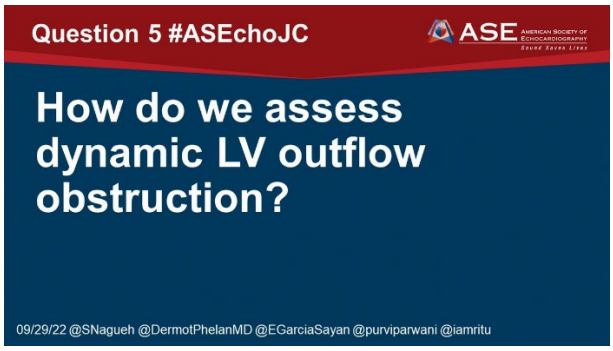
🔶Diagnosis (site of greatest hypertrophy and fibrosis has reduced strain)
🔶Prognosis (lower stain associated with Vent Arrhythmias, ICD discharge & death)
❓Possible utility in timing of myectomy and evaluation of G+ve/P-ve



@iamritu: Those pts with HCM & ventricular arrhythmias showed worse GLS than those without them
Abnormal GLS is an independent predictor of outcomes in HCM
Strain patterns may vary based on type of HCM

@vidhu_anand: Strain pattern can also help rule out infiltrative CMP such as amyloid

Question 5:

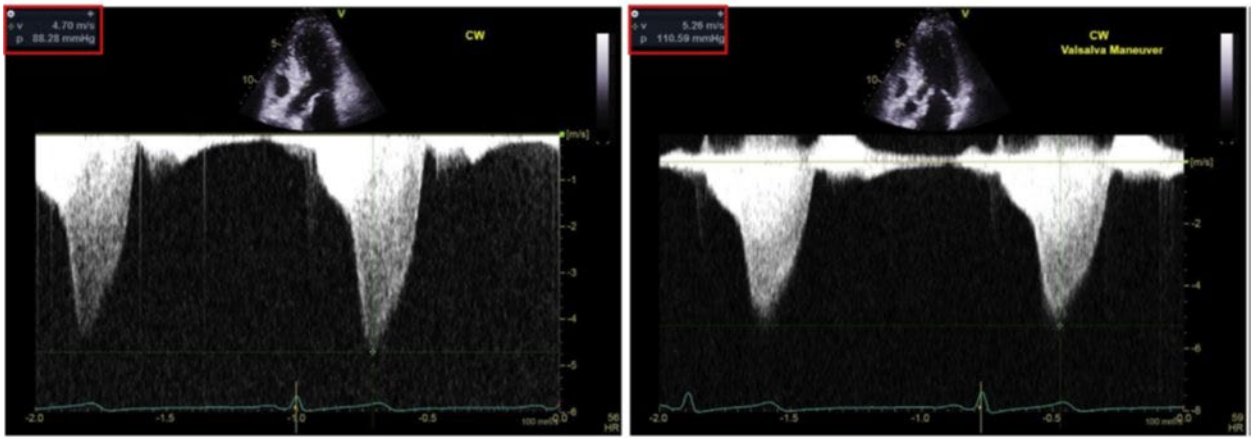


A5 Notable responses

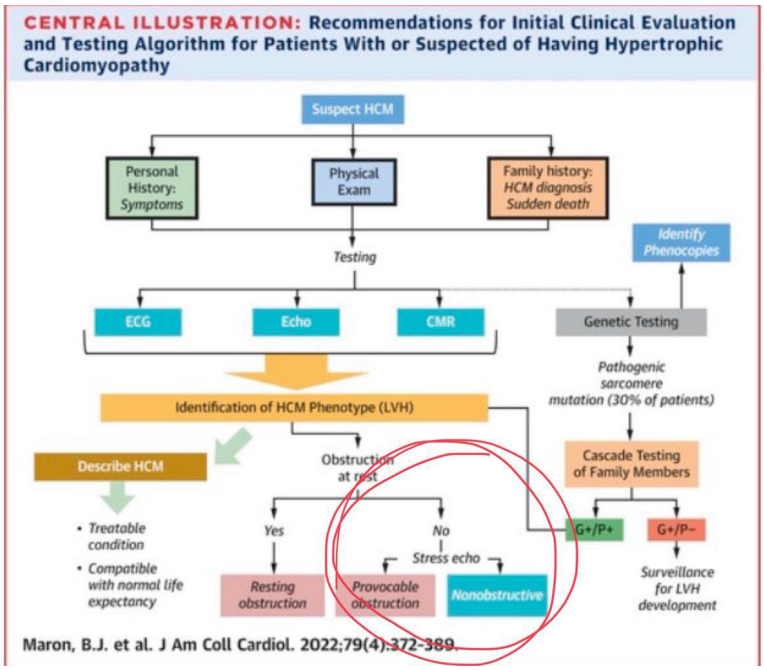
@DermotPhelanMD:

- Visualize: B-Mode PLAX/PSAX/A3C/A5C
- Localize: Color Doppler, PW Doppler
- Quantify: CW Doppler
- Provoke: Valsalva, Exercise, amyl nitrate etc
- Report: Location of obstruction, resting and stress gradients

Late-peaking, dagger-shaped LVOT velocity waveform at rest and with Valsalva

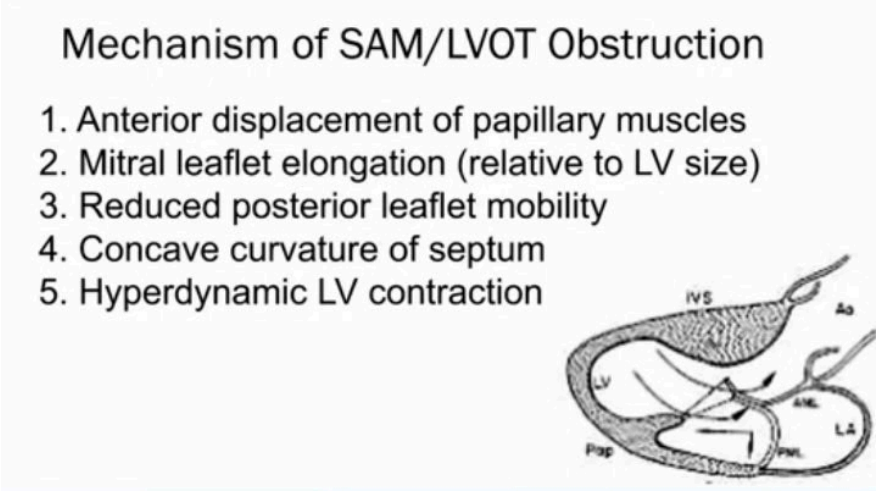


@iamritu:
Resting signs assoc w latent LVOT obstruction in HCM #ASEchoJC
When both MV coaptation length ≥10 mm (long)&LV Outflow D<20 mm short,severe LVOT obstruction likely
when neither seen, severe obstruction unlikely
if only 1 seen, need further test https://bit.ly/2V6oYnH



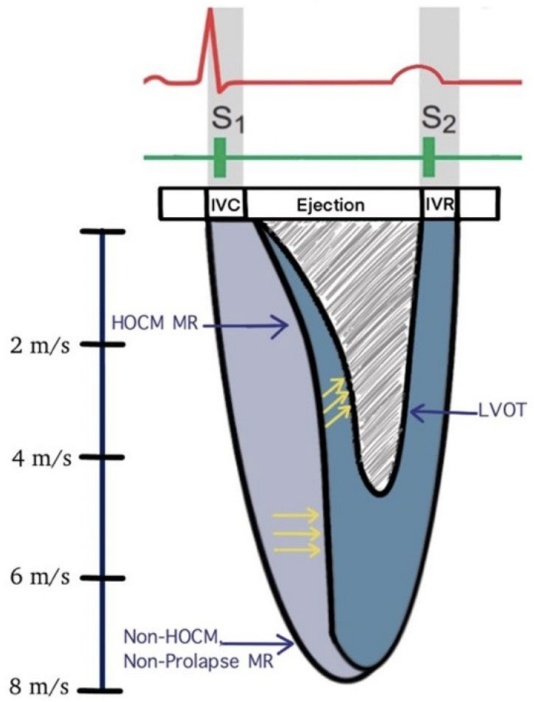
@iamritu

Another key point is that the amount of LVH does not correlate with LVOT obstruction #ASEchoJC Look for other anatomic variants that can cause LVOT



@purviparwani: Important to remember the contamination with MR signal.
-> LVOT signal velocity is lower
->LVOT signal is narrower and spans the ejection time
-> Characteristic late peaking, dagger-shaped, and continues to show rightward curvature in mid-late systole

Figure 8



@rajdoc2005: Great reminder. This concept is heavily tested in Echo boards and can be really challenging in clinical practice. It's important to educate the sonographers how to carefully try to tease out the LVOT velocity with little/ no contamination from the MR jet.

@boegel_kelly: This can be difficult to master and can require diligence and some patience to define the different signals. Important to pay close attention to signal shape, velocity and duration

@purviparwani:

Differentiating LVOT signal from MR signal

-> See the velocity
-> See the envelope
-> See the shape of the envelope

Differentiating the HOCM MR Signal From the LVOT Signal

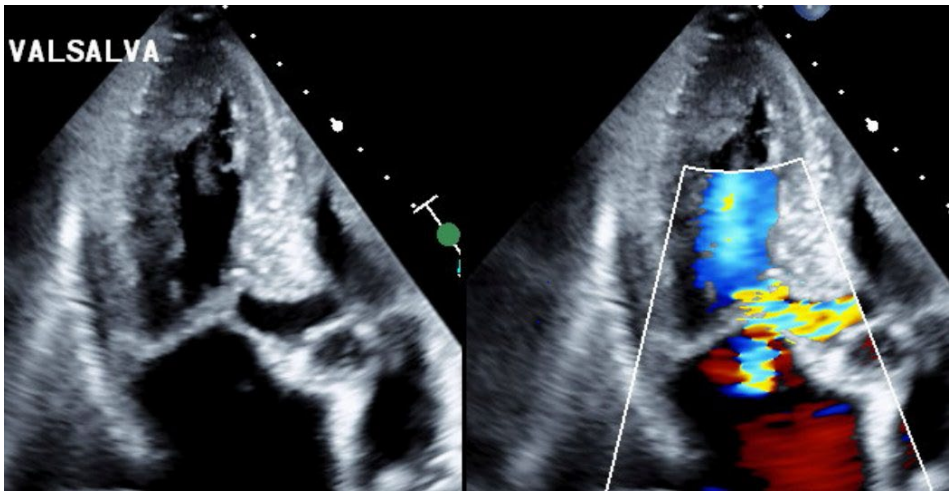
The appearance of the HOCM MR and LVOT signals can be confusing and the differentiation can be challenging. If the MR signal is mistaken as an LVOT signal, the LVOT gradient can be overestimated. The features listed in Table 1 help differentiate these signals:

Table 1

HOCM MR Signal	LVOT Signal
• MR signal velocity is always higher because it gives the LV-to-LA gradient	• LVOT signal velocity is lower
• HOCM MR envelope is wider, begins after the onset of ejection, and terminates at the end of IVR	• LVOT signal is narrower and spans the ejection time
• Curved initially but then straightens out (perpendicular to baseline) by midsystole	• Characteristic late peaking, dagger shaped, and continues to show rightward curvature in mid-late systole

HOCM = hypertrophic obstructive cardiomyopathy; IVR = isovolumetric relaxation; LA = left atrial; LV = left ventricular; LVOT = left ventricular outflow tract; MR = mitral regurgitation.

@DermotPhelanMD: I love color compare when localizing obstruction. Patient below had no obstruction at rest - see what a good Valsalva can do.



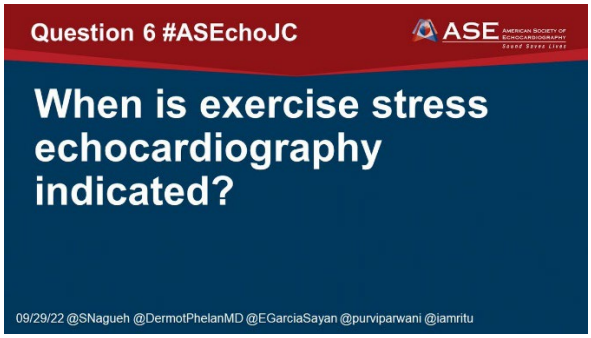
@boegel_kelly: I like this too although it's important that good 2D imaging is obtained as it's own clip as well as adding the color Doppler decreases your frame rate significantly.

@purviparwani: Use the strain phase of the Valsalva maneuver to precipitate LVOT obstruction, forced exhalation against a closed airway, results in less in venous return

A limitation to the Valsalva maneuver is the subjective nature of the effort and thus the variable response.

> **@rajdoc2005:** Some echo labs in the community do not use Valsalva at all! 😕😕😕 Important to try to educate their teams on how its done and what we are looking for!
>
> **@purviparwani:** I would probably say most don't in community. HCM remains under diagnosed!

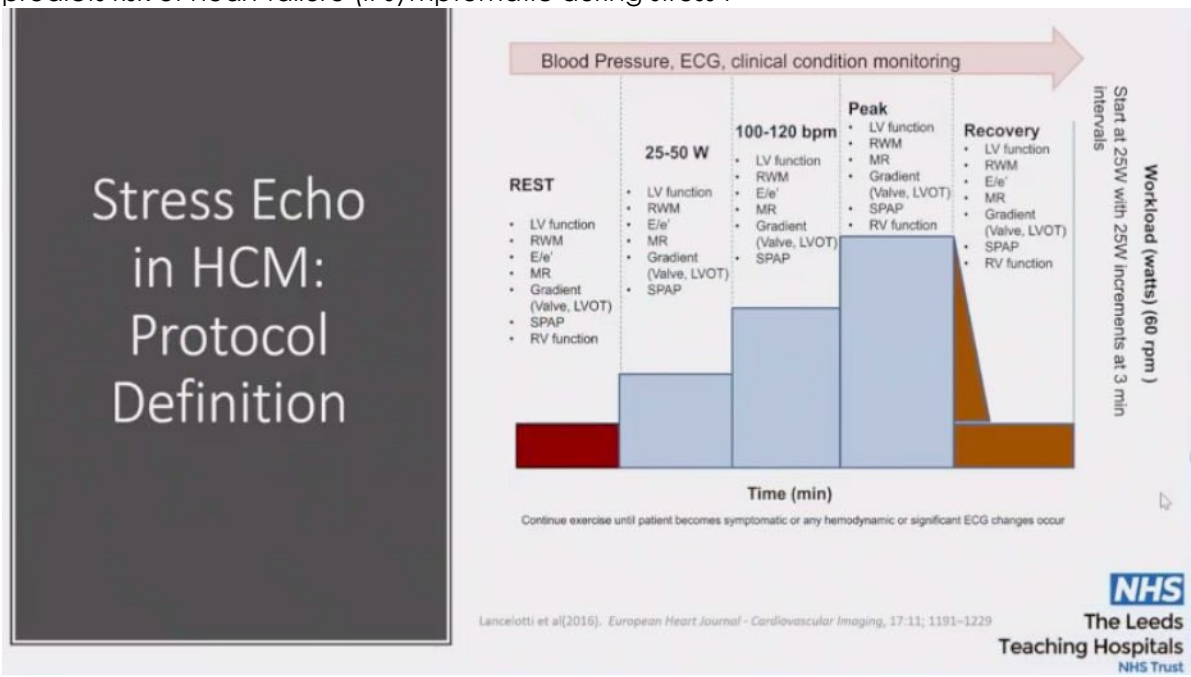
Question 6:



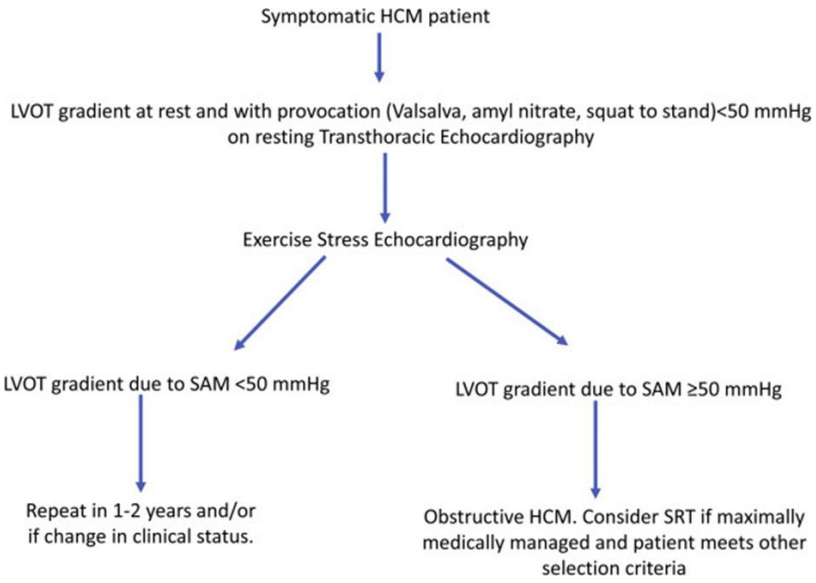
A6 Notable responses

@SNagueh: In symptomatic patients without significant dynamic obstruction at rest or with Valsalva. Goal is to identify exercise induced LVOT dynamic obstruction with SAM at ≥ 50 mmHg. Medical therapy should not be withheld before test.

@iamritu: Getting baseline exercise stress echo is key in HCM
if Pt has HCM & SOBOE - is it exercise-induced SAM or new MR or exertional arrhythmia? #ASEchoJC
Another factor for SCD in HCM=⬇ BP during exercise stress test
predicts risk of heart failure (if symptomatic during stress t

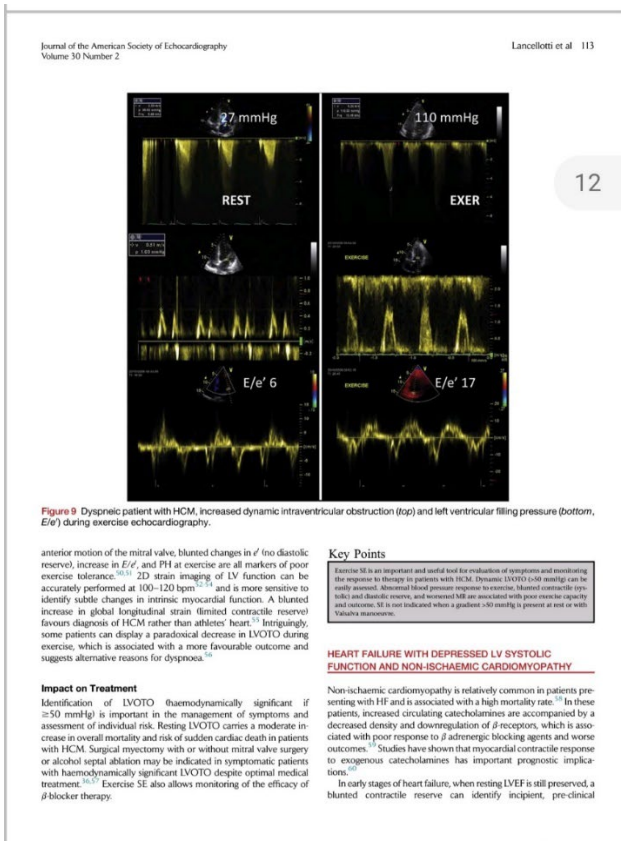


@SNagueh



@purviparwani: Remember Valsalva Dynamic response is lower than that of exercise stress test, when patient can, always exercise them

@DavidWienerMD: An example of exercise stress in #HCM, from @ASE360 guideline on stress testing in non ischemic diseases: increased gradient and worse filling pressures with exercise. Read it at https://asecho.org/guideline/the-clinical-use-of-stress-echocardiography-in-non-ischaemic-heart-disease/

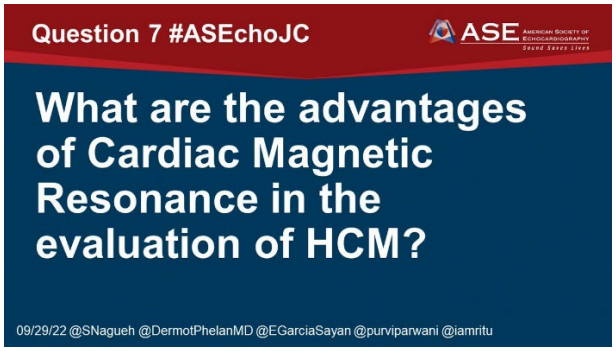


@rajdoc2005: Extremely helpful to unmask symptoms and hemodynamics. But these "exercise" stress echos have to be carefully done and ideally physician supervised - to get all the important Information we can glean from a stress echo well done!

@DavidWienerMD: We did 2 today, and I planned the sequence of image acquisition and what to acquire in advance, with the sonographer and fellow

@rajdoc2005: THAT is exactly what should be done. Just perfect!👍👍

Question 7:



A7 Notable responses

@SNagueh

1) -Identify increased wall thickness in regions that could be missed by echo, as anterolateral wall.

- Accurate measurement of LV mass, maximum wall thickness, apical hypertrophy

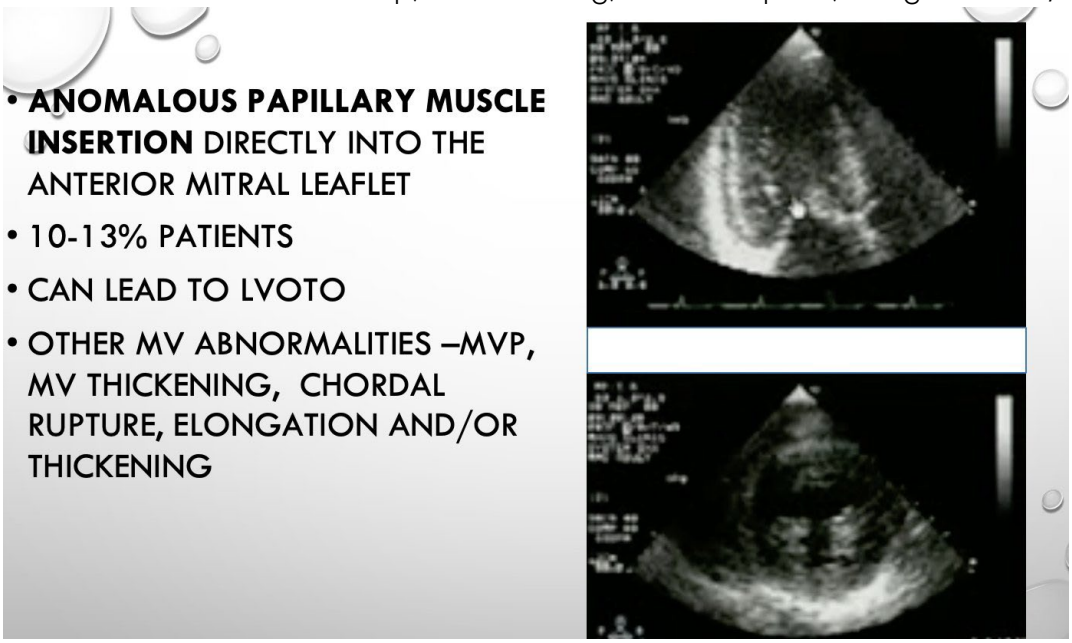
- Accurate measurement of LV and RV volumes and EF

2) -Identifies details of mitral valve apparatus and presence of muscle bands in LVOT

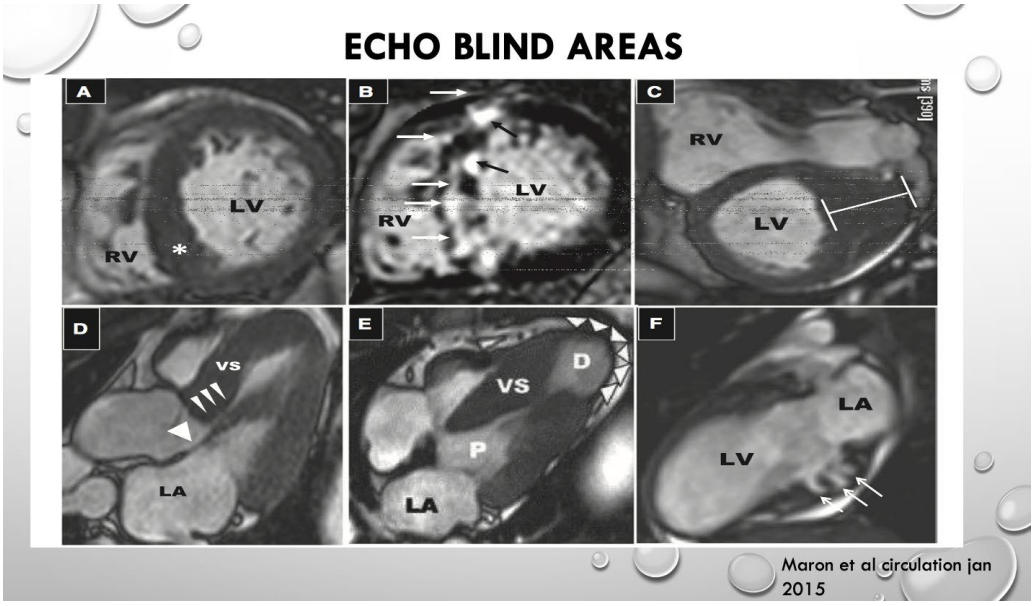
-Identification of regions with LGE, scar burden, apical aneurysm, and ECV

- Detects ischemia

@purviparwani: Anomalous papillary muscle insertion directly into the anterior mitral leaflet #ASEchoJC #whyCMR #echofirst
->10-13% of patients
->Can lead to LVOTO
->Other MV abnormalities –mvp, mv thickening, chordal rupture, elongation and/or thickening



@purviparwani: #whyCMR
-> To confirm the diagnosis
->To distinguish between inherited and non-inherited
->For initial or subsequent evaluation of left/ right ventricular function (LVEF/ RVEF)
->To assess the extent of LGE
continued..

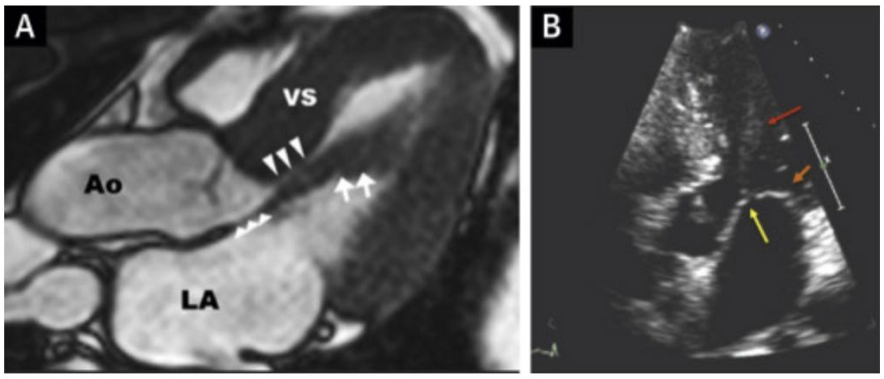


@DermotPhelanMD:
#WhyCMR - beautiful views of apical aneurysm with tissue characterization.
Major impact to patient care to identify this - needs anticoagulation and ICD
https://twitter.com/i/status/1575647151018823680

@MaheshAnandCh: Main pts with HCM have abnormal mitral valve apparatus that may effect residual MR post myectomy. Does MR have any role over echo for surgical planning?

@purviparwani: yes TEE #Echofirst or #whyCMR can be useful for better visualization of the mitral complex, elongated AML

Great article : https://sciencedirect.com/science/article/pii/S0735109716007518



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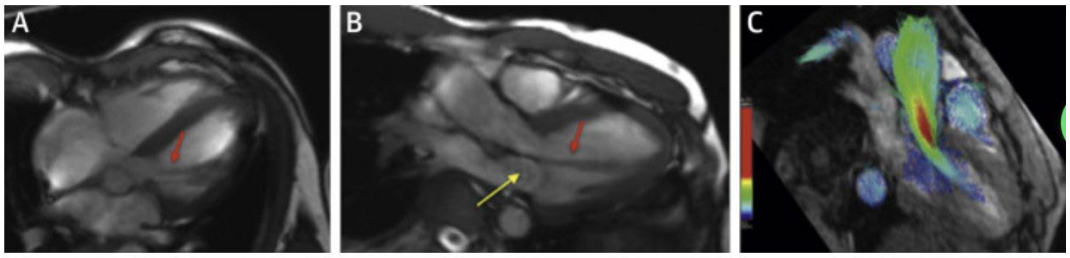

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Figure 8. CMR of Bifid Anomalous Anterolateral Papillary Muscle With Insertion Into the Midanterior Mitral Leaflet

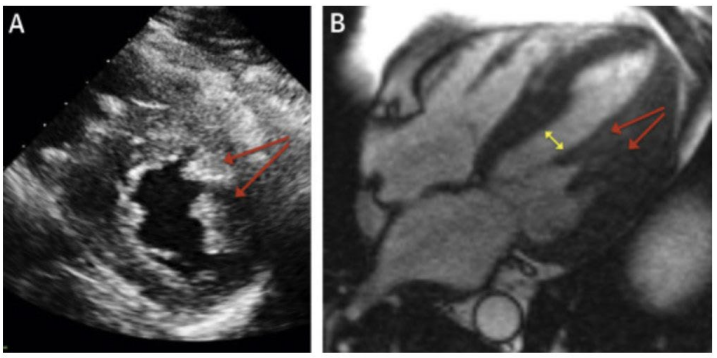

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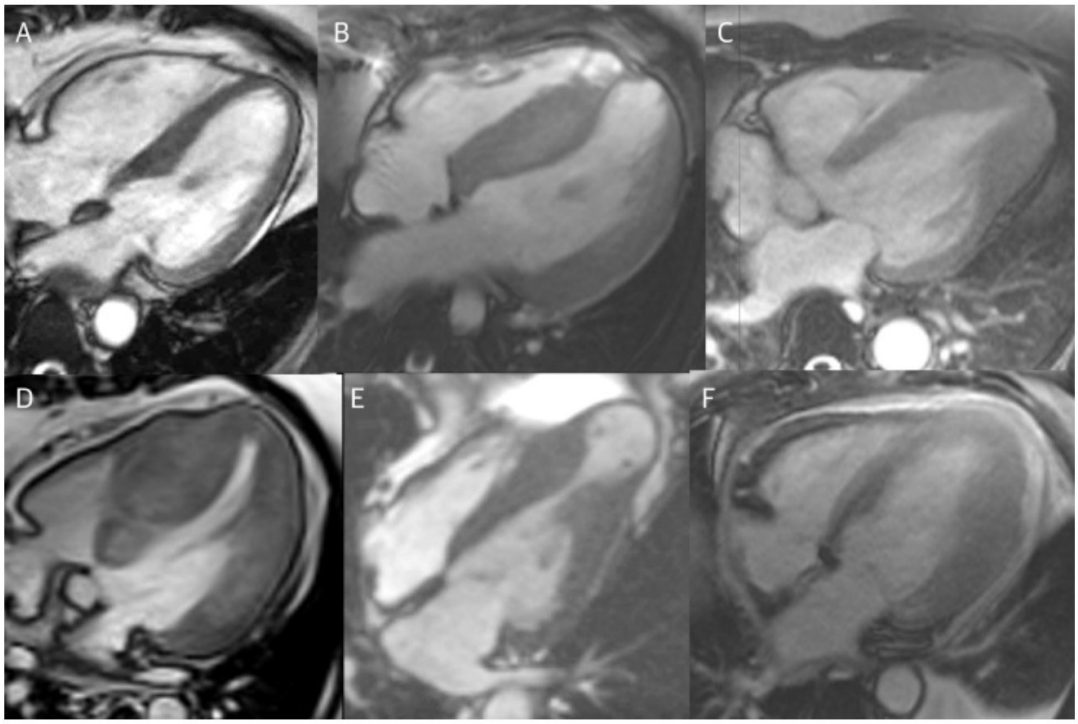
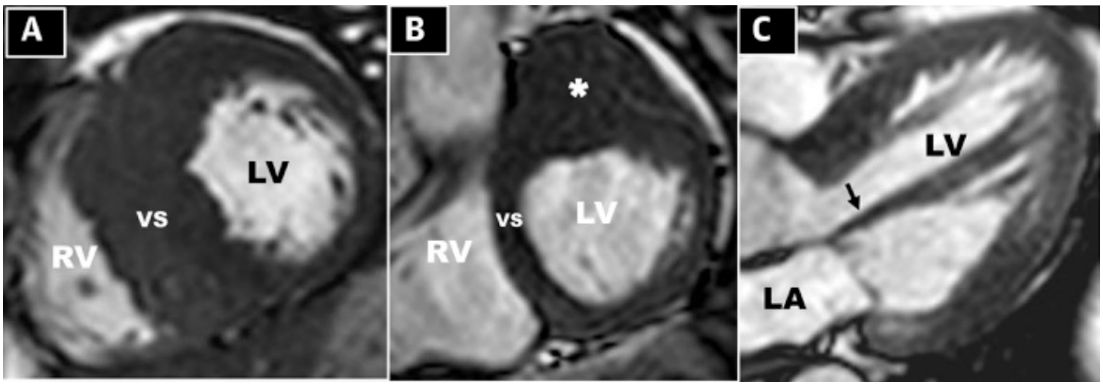
Figure 6. Anterior Displacement of Hypertrophied Anterolateral Papillary Muscle

@purviparwani

#whyCMR in HCM #ASEchoJC
->To measure maximum LV hypertrophy, as opposed to the contralateral wall and myocardial mass
-> To evaluate 'burned out" HCM and subsequent HF
-> For prognostic evaluation (maximal LVH, HF, LGE)
https://sciencedirect.com/science/article/pii/S0735109719376831
@ChrisKramerMD @salernomdphd

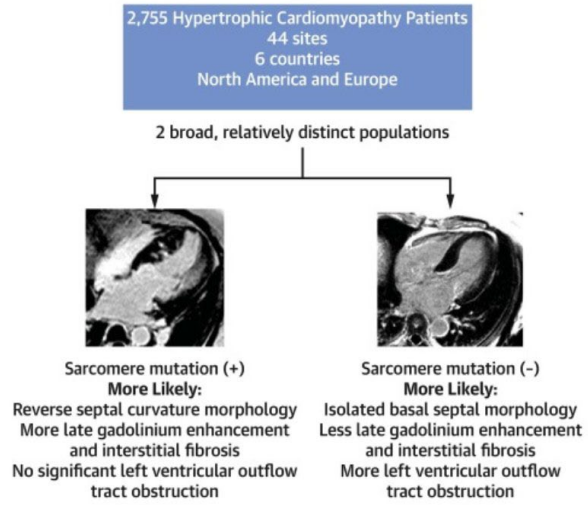


@iamritu: #WhyCMR 🔑 in HCM
-Massive septal hypertrophy
independent SCD risk -anterolateral free wall hypertrophy(area can be blind to echo beam)
-anomalous insertion of AL pap muscle directly into base of ant mitral leaflet ➡️ midventricular obstruction



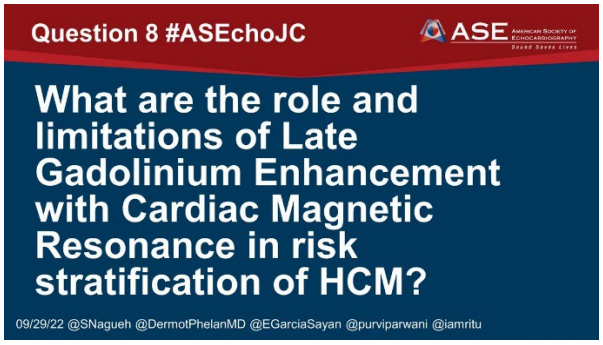
@purviparwani: #WhyCMR in #HCM #ASEchoJC
HCM registry data
-> sarcomere mutation positive pts-> ⬆️reverse septal curvature morphology, ⬆️fibrosis, but ⬇️resting obstruction,
Sarcomere negative pts -> ⬆️ isolated basal septal hypertrophy with obstruction, but ⬇️fibrosis.
https://sciencedirect.com/science/article/pii/S0735109719376831

CENTRAL ILLUSTRATION: Hypertrophic Cardiomyopathy: Overall Design and Findings



Neubauer, S. et al. J Am Coll Cardiol. 2019;74(19):2333–45.

Question 8:



A8 Notable responses

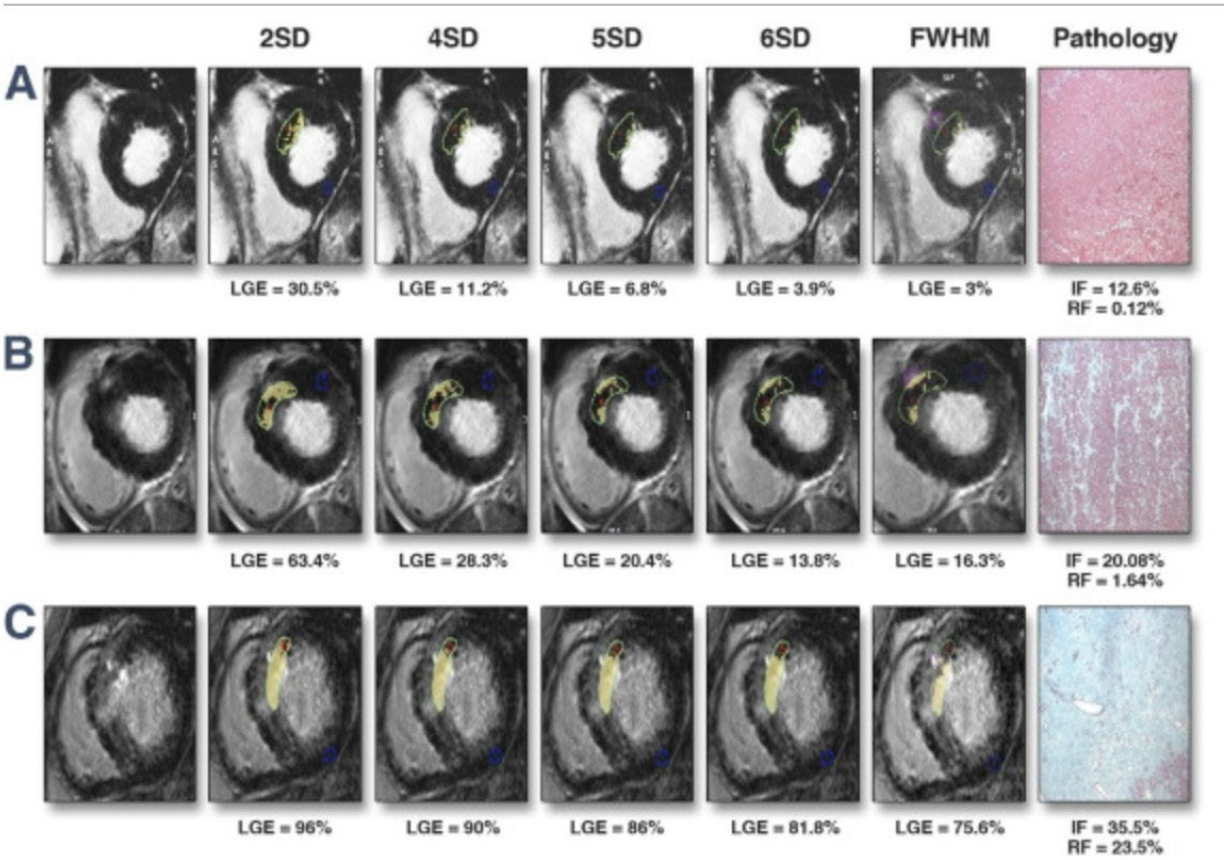
@SNagueh

1/2
LGE and SCD
-Common in >50%, patchy mid myocardial and in areas with hypertrophy
-Often seen in RV insertion points and if only here it is not associated with increased risk of SCD
-Scar burden related to ventricular arrhythmias, SCD, and HF diagnosis and admissions, low EF

2/2
LGE$SCD
-visual or measurement based on signal intensity of affected region versus normal segment
-LGE ≥15% associated with increased risk of SCD.
-Repeat CMR every 3-5 years for changes in wall thickness and LGE, and to identify new apical aneurysms or LV EF reduction

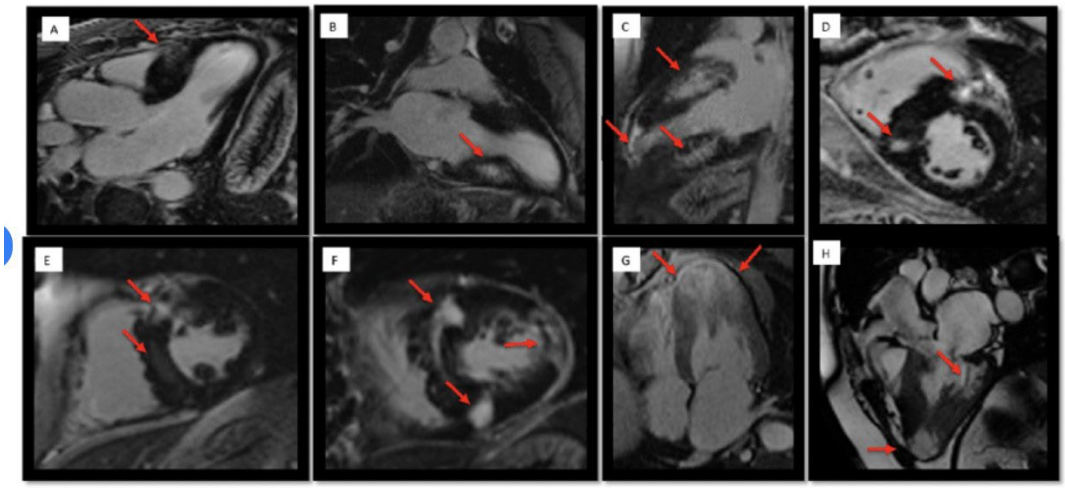
@vidhu_anand: Important to quantify LGE as much as possible given importance of the same in recent guidelines. What method do you use? FWHM? >2 or 3SD of normative?

@purviparwani: Great question! #whyCMR #Echofirst
-> Either 6 SD or FWHM are preferred in HCM.
-> Large studies used a threshold of 15% LGE of the LV mass using the 6-SD technique showed LGE >15% was associated with an increased risk of SCD
https://sciencedirect.com/science/article/pii/S1936878X13002283



@purviparwani:

LGE is noted in approximately half of the HCM pts
Most commonly patchy & mid myocardial within segments of maximal hypertrophy
Several studies have demonstrated ↑ ventricular arrhythmias, SCD, and all-cause mortality in patients who have LGE

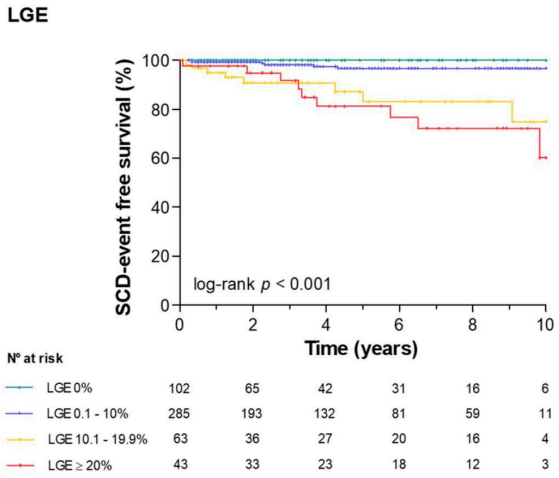


@purviparwani:

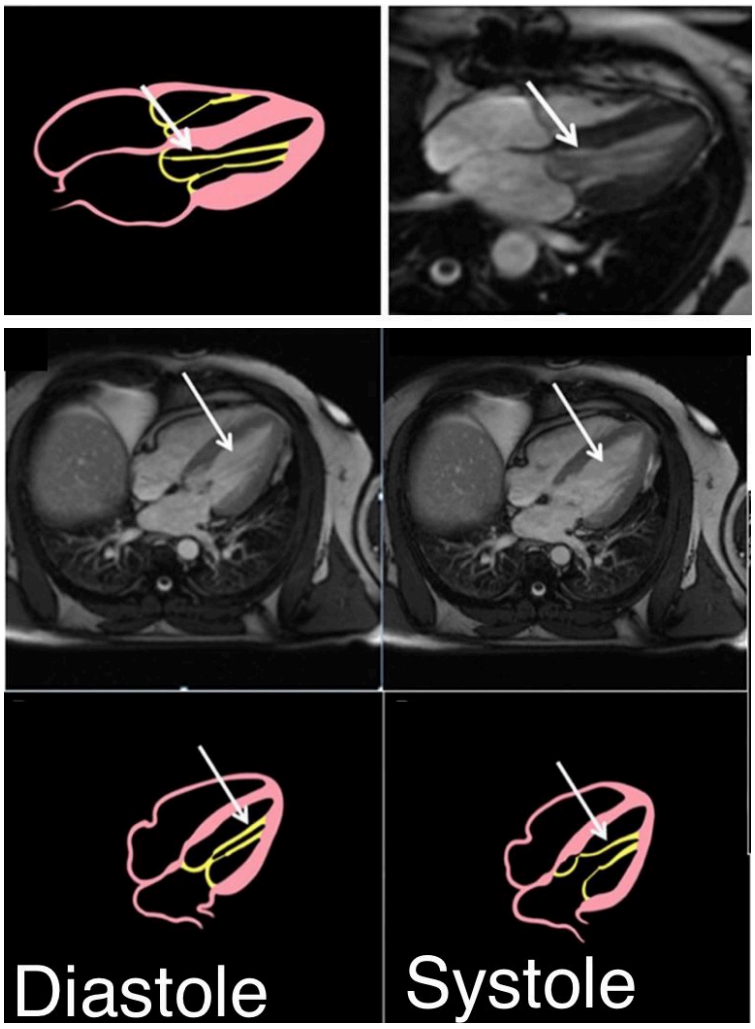
->Isolated LGE at the RV insertion points does not appear to be associated with increased risk. -> Although LGE is present in >50% of patients with HCM, the overall prevalence of SCD in these patients is far lower.

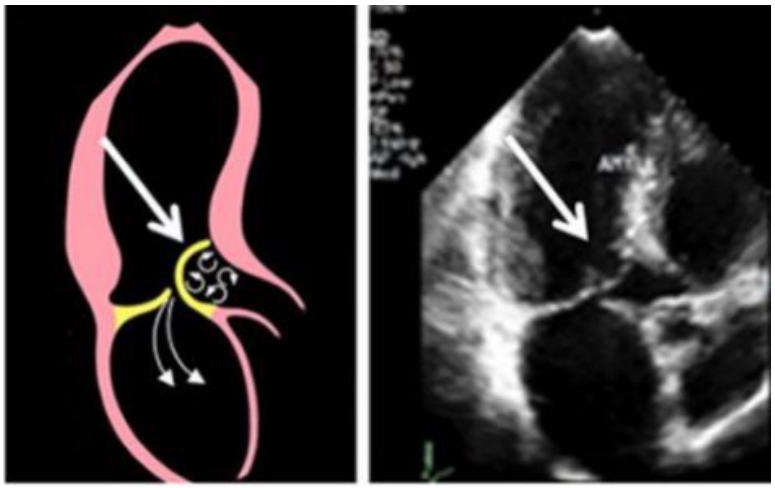
@iamritu

#WhyCMR 🔑 to see
extensive LGE of ≥15% of LV mass 2x ↑ in SCD risk HCM
https://bit.ly/364wgbN
elongated anterior mitral leaflet, abnl chordal attachment to base of anterior mitral leaflet,bifid anterolateral papillary muscle
https://bit.ly/3r8V13b



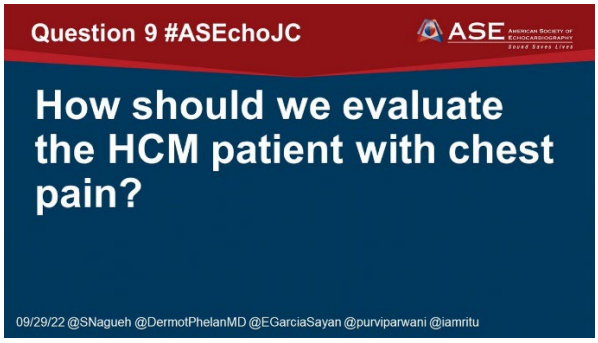
Survival analysis through Kaplan-Meier according to the ACCF/AHA, HCM Risk-SCD and LGE classifications





@purviparwani
#whyCMR #ASEchoJC #SAM LVOT obstruction on 4Dflow
https://twitter.com/i/status/1575654222380441600

Question 9:



A9 Notable responses

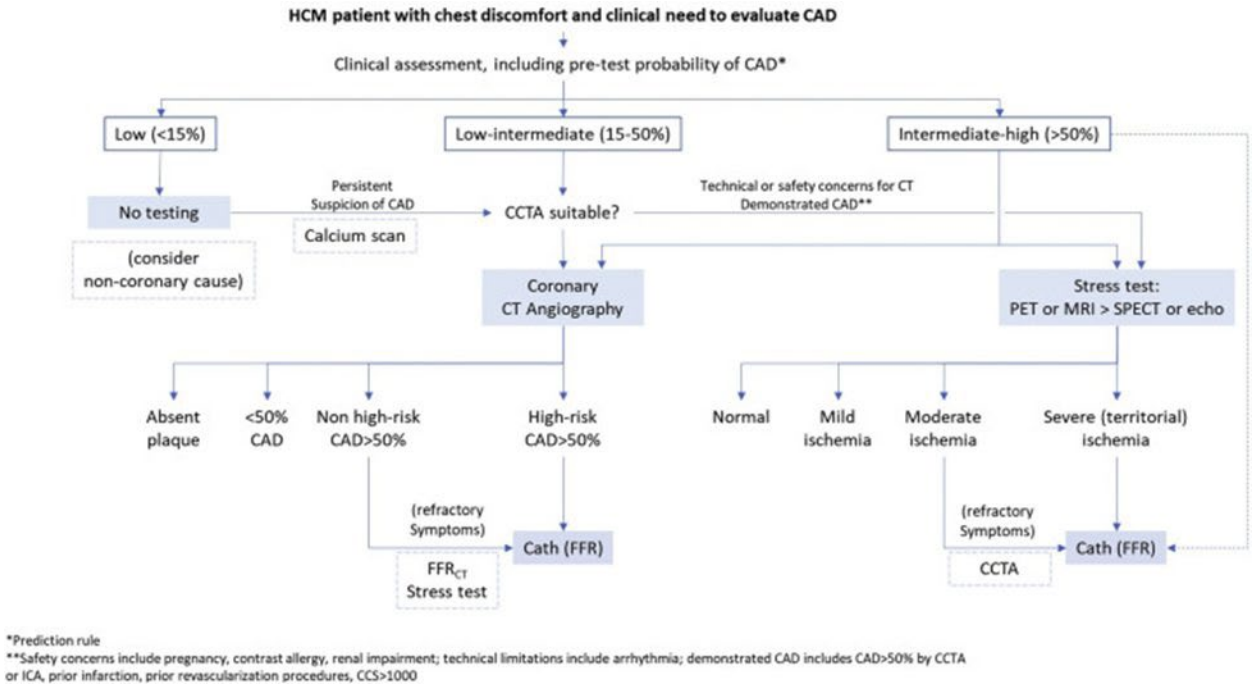
@SNagueh

1/3
-First step is clinical evaluation
-Low probability (<15%) CAD, no testing reasonable
-If symptoms persist, reasonable to obtain coronary calcium scan
-In patients with low to intermediate probability (15-50%), coronary CT for epicardial CAD and myocardial bridges

2/3

-Not good candidate for coronary CT or high probability or known CAD, stress testing by PET or CMR perfusion recommended.
-Invasive angiography in presence of significant disease (left main or 3 vessel disease) by CT or functional stress testing. FFR measurement if needed

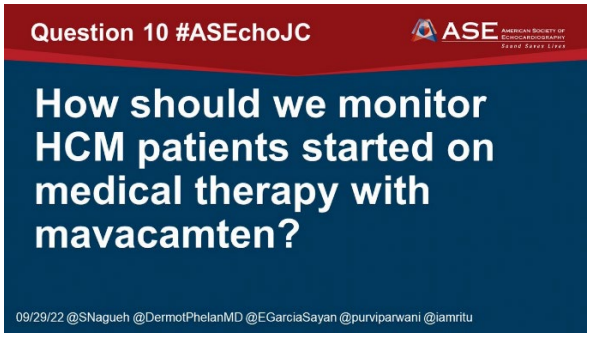
3/3



*Prediction rule
**Safety concerns include pregnancy, contrast allergy, renal impairment; technical limitations include arrhythmia; demonstrated CAD includes CAD>50% by CCTA or ICA, prior infarction, prior revascularization procedures, CCS>1000

@iamritu: HCM can have micro vascular obstruction & increased myocardial oxygen demand causing exertional CP or HCM w severe diastolic dysfunction & restrictive phenotype without LVOTO w CP/SOB with high LVEDP & worse prognosis

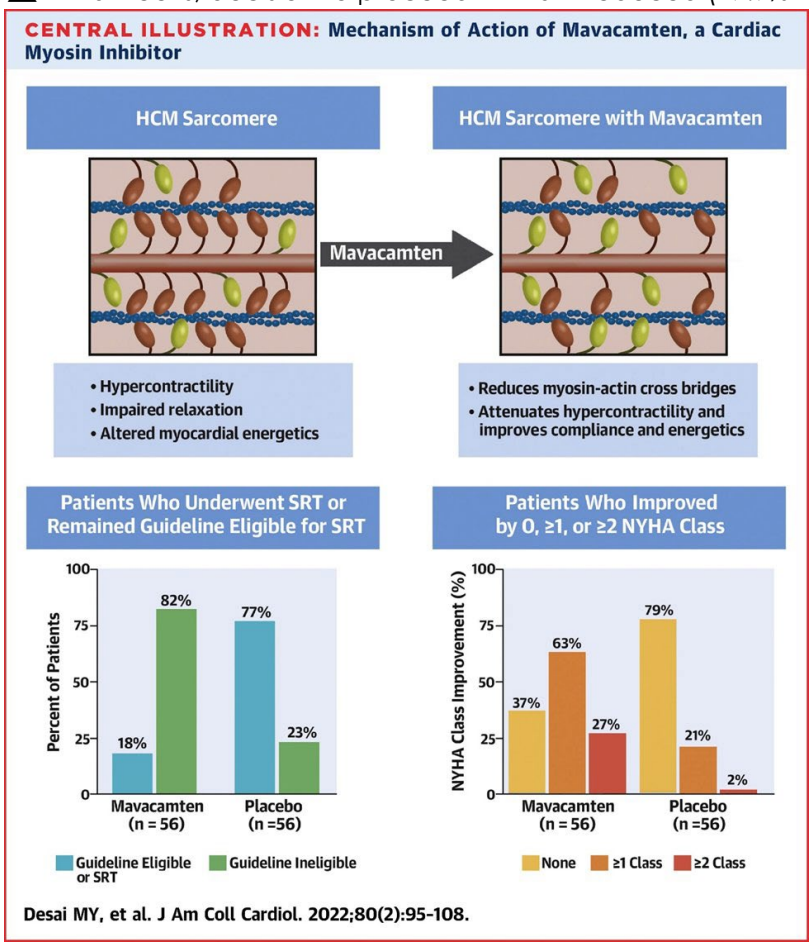
Question 10:



A10 Notable responses

@EGarciaSayan

Highlights from VALOR-HCM trial:
🔺Adults with severe symptoms despite OMT + peak grad >50 mmHg
🔺Referred <12 months for SRT & considering scheduling procedure
🔺Randomized to mavacamten or placebo
🔺At 16 weeks, decision to proceed with SRT reduced (17.9% vs 76.8%)



@SNagueh:

1/2

-Need echo at baseline for rest and Valsalva LVOT gradient and LV EF. Do not start if EF<55%
- Avoid concomitant use of mavacamten in patients on disopyramide, ranolazine, verapamil with a beta blocker, or diltiazem with a beta blocker
- REMS program, starting dose 5 mg/day

-Stop drug if heart failure symptoms develop, clinical status worsens or EF<50%
-Monthly echo with clinical evaluation each month for first 3 months and after 12 weeks, repeat evaluation every 12 weeks unless dose changes or EF <50% in which case repeat echo in 4 months.

> **@bobatr0n1:** Can you elaborate on the avoidance of these concomitant medications? I thought it was an add-on medication to those that remain symptomatic.
>
> **@MaheshAnandCh:** The medication was studied on a background of beta blockade or CCB but not both. In addition folks were not on Norpace. Unrelated, there are medications interactions to be aware of

@iamritu:
mavacamten/Camzyos initiation & treatment will be serially #EchoFirst-guided — Rx will be restricted through REMS program https://bit.ly/3rdNC2k
#Valor used core lab-measured LVEF, LVOT gradient at rest, & Valsalva provocation
will this be sufficient in real world?

@SNagueh:
Need carefully collected registry data to answer this most important question.

@MaheshAnandCh
There is only more myosin modulation coming down the pipeline- which will lead to a busy echo lab