HCM: Multimodality Imaging

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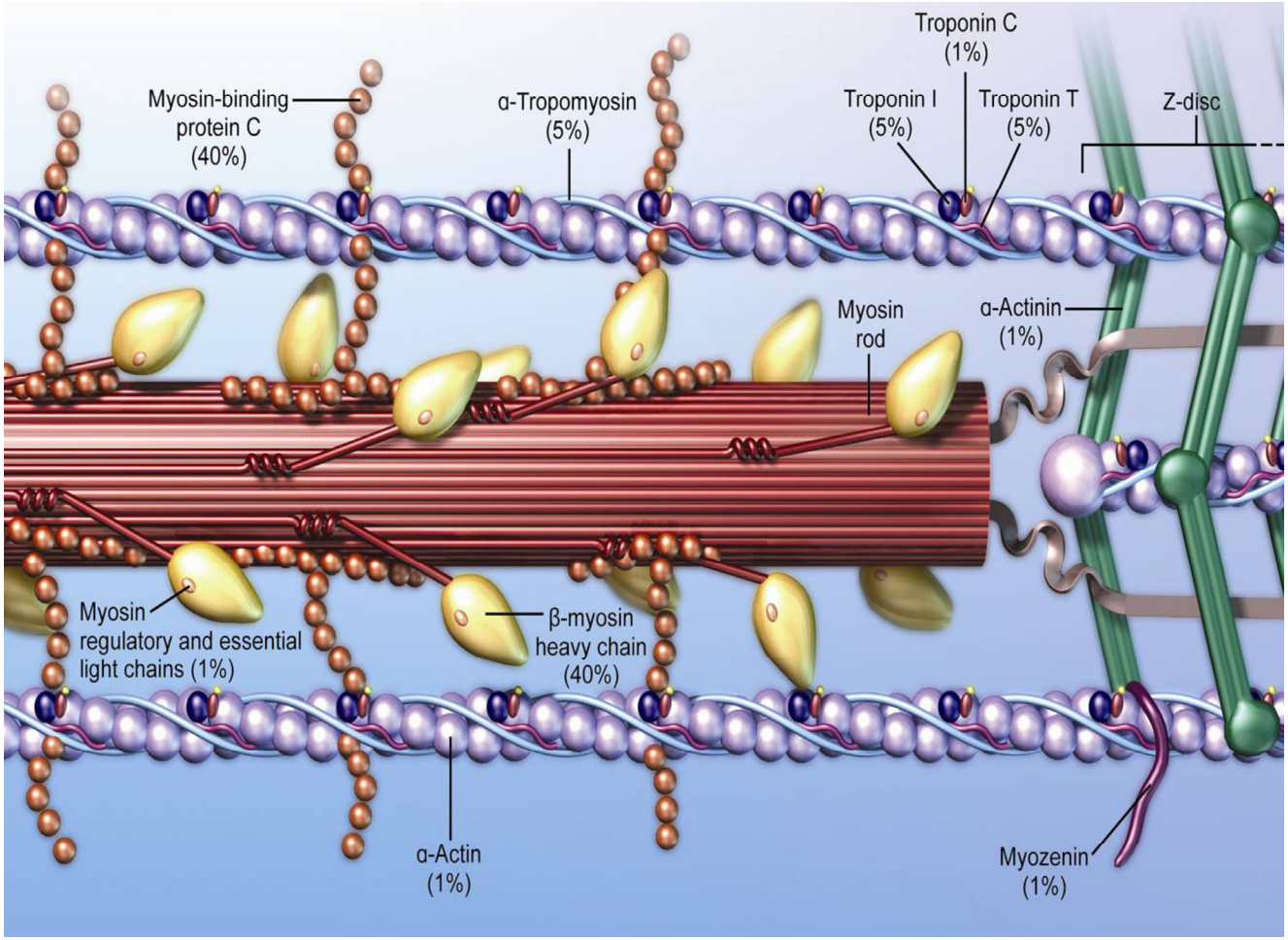
HCM: Commonest inherited heart muscle disease

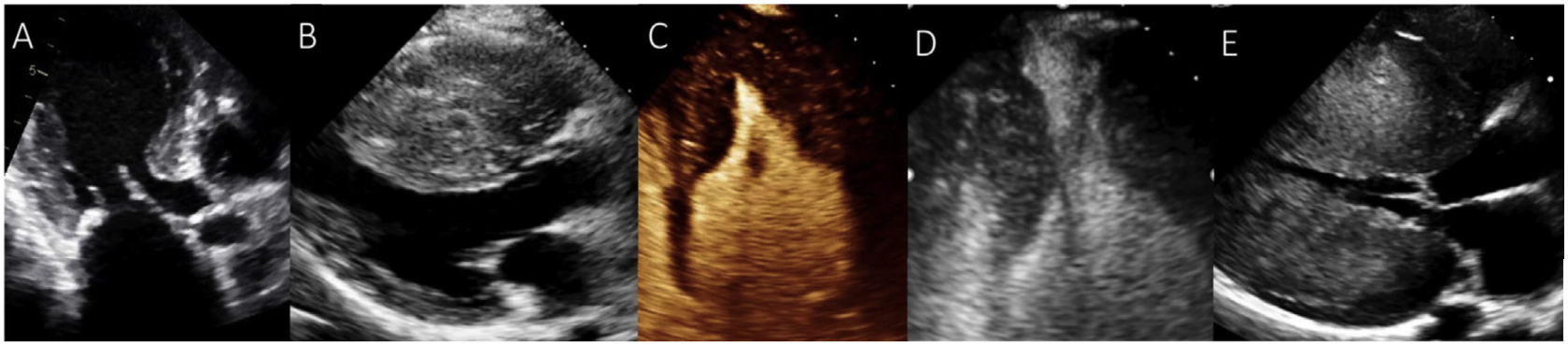
- Most common inherited cardiac dz
- 1 in 250
- 11 genes; most are AD
- Sarcomeric proteins
- Penetrance ~ 50-80%; age dependent
- Genetic testing yield: 60%
- Low risk of SCD in most

Diagnosing HCM – entirely dependent on IMAGING

- Unexplained LVH ≥15 mm (end-diastolic wall thickness) in absence of other cause of magnitude of this LVH (if 1st degree relative of proband, >13 mm)
- Septal/post ratio >1.3 in normotensive pts (>1.5 if HTive)
- Screening of probands requires imaging (especially if genotype +, phenotype --)



Phenotypic diversity



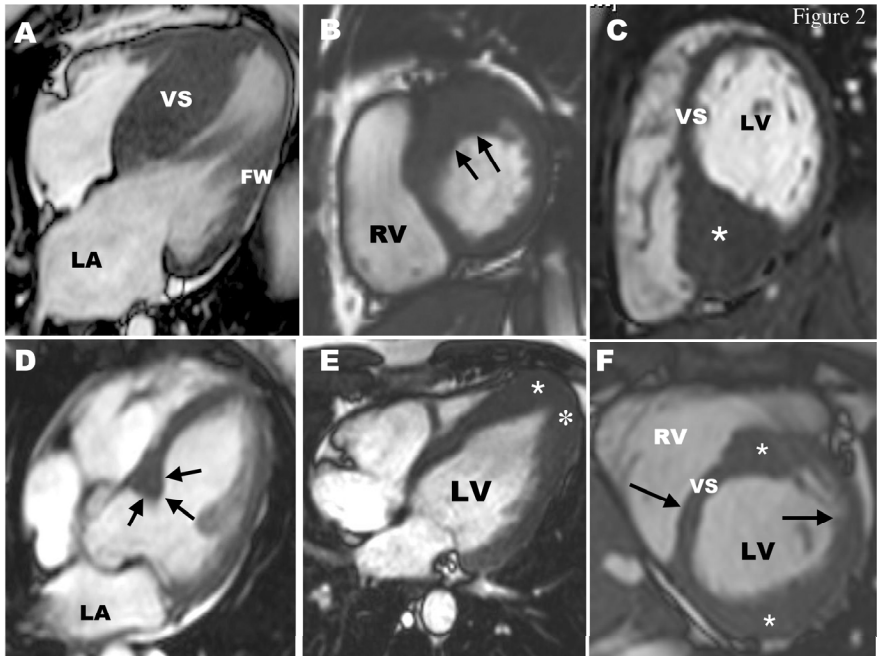
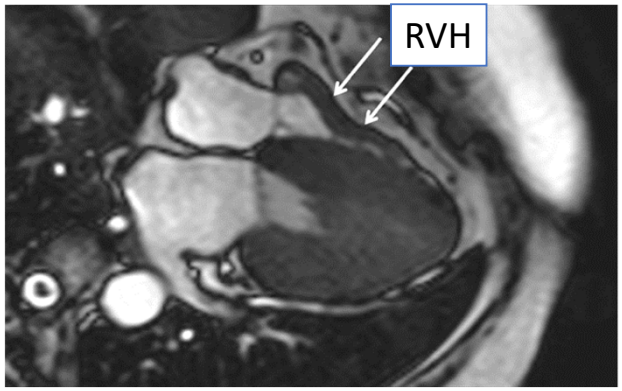
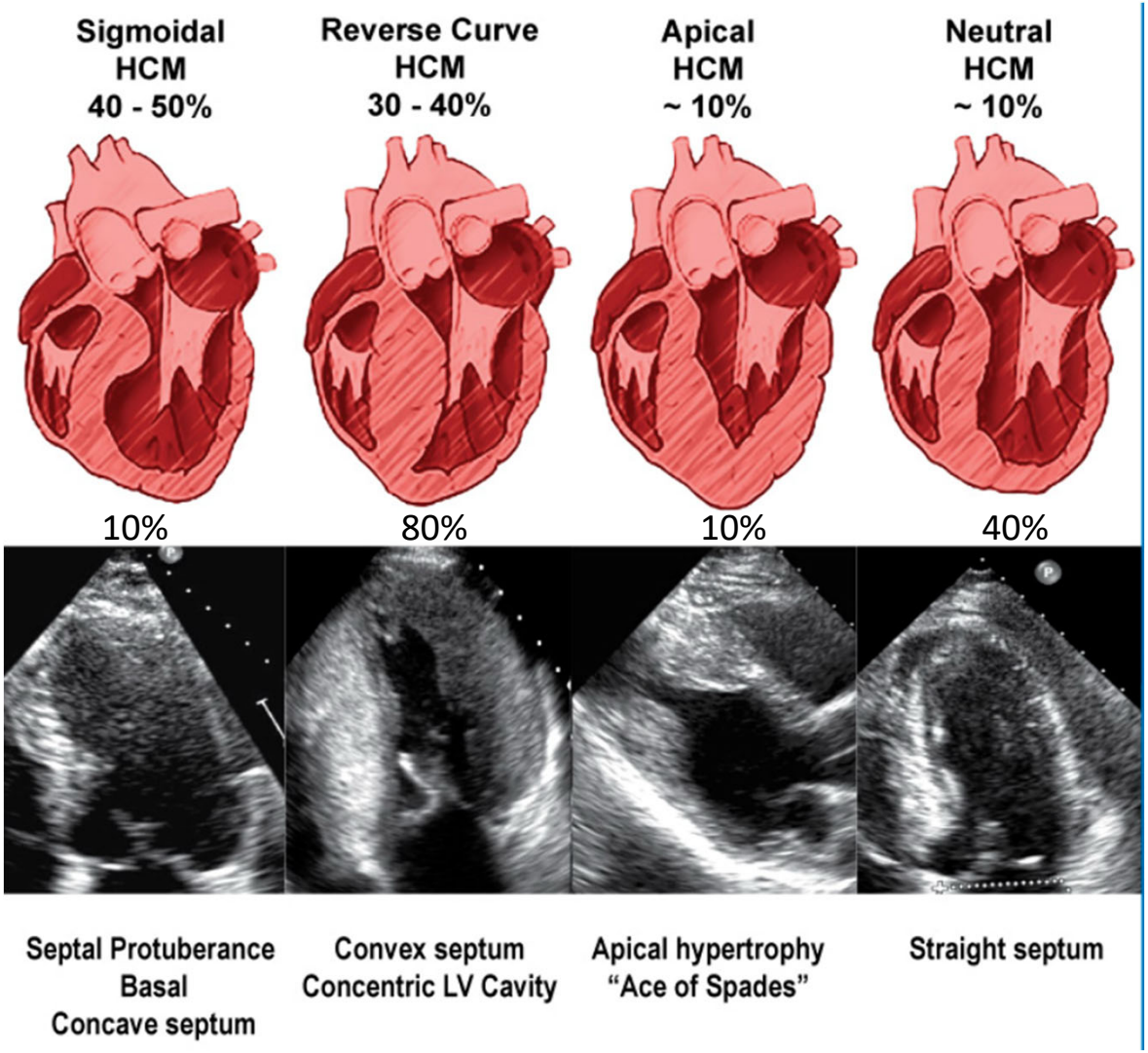


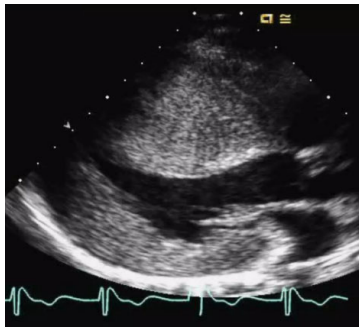
Figure 2



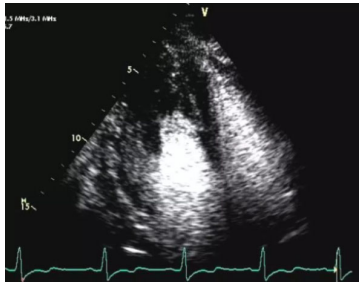
Maron MS et al, JCMR 2012

Patterns of LVH

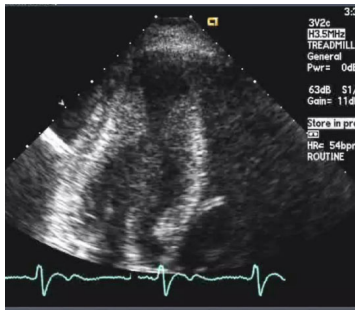




Reverse curve HCM



Apical HCM



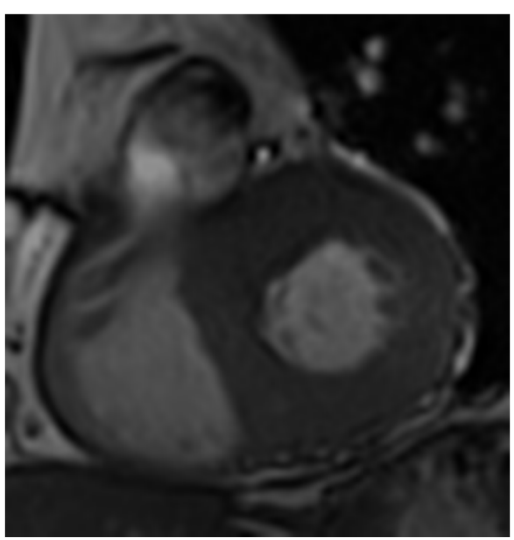
Apical HCM w aneurysm

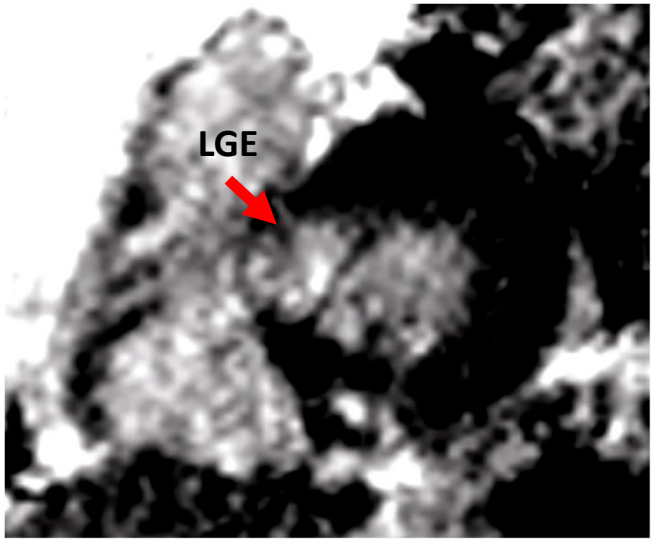
Basal Asymmetrical Septal Hypertrophy (ASH)

- Most common phenotype accounting for 60–70% of HCM cases
- Diagnostic criteria
  - Basal anterior septal thickness is ≥ 15 mm at end-diastole

AND

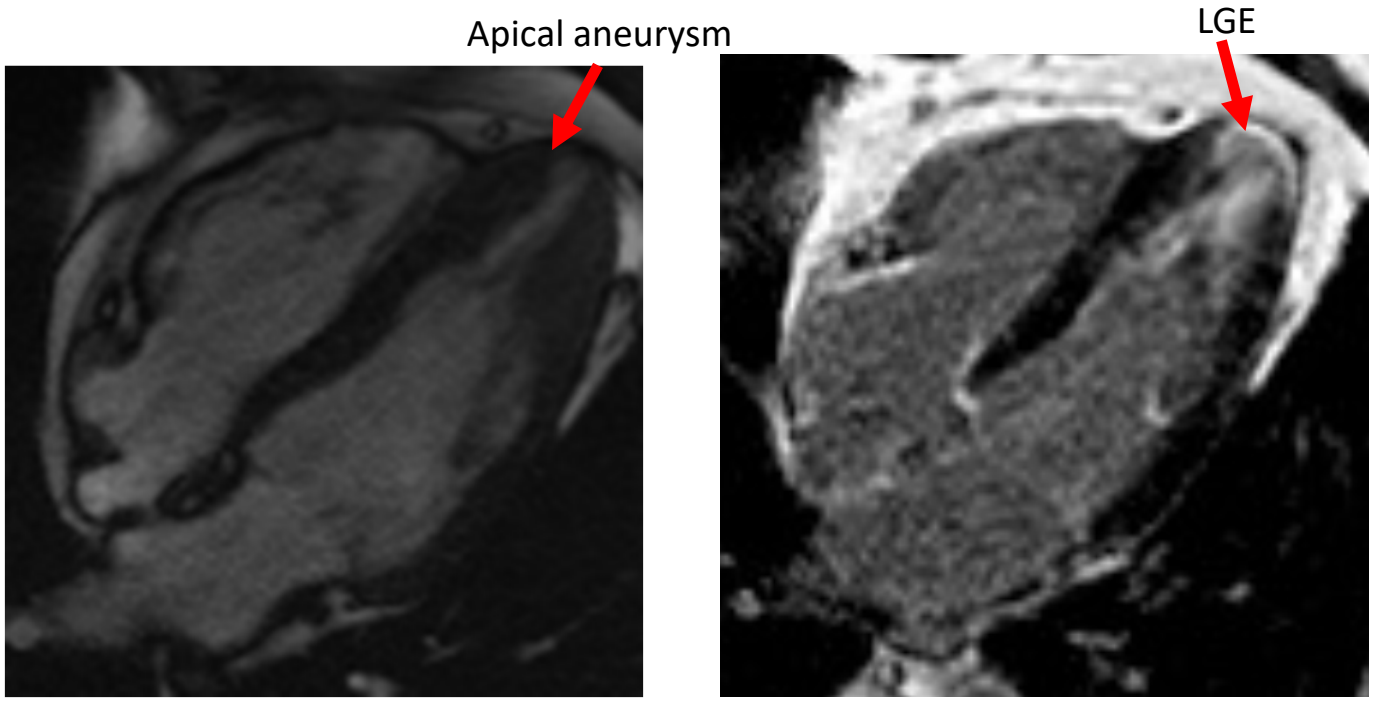
  - Ratio of septal to inferolateral wall thickness is ≥ 1.3





Midventricular obstruction

- Massive hypertrophy of mid-ventricular myocardium
- Can result in apical aneurysm formation
- Associated with thrombus formation
- Associated with a poorer prognosis

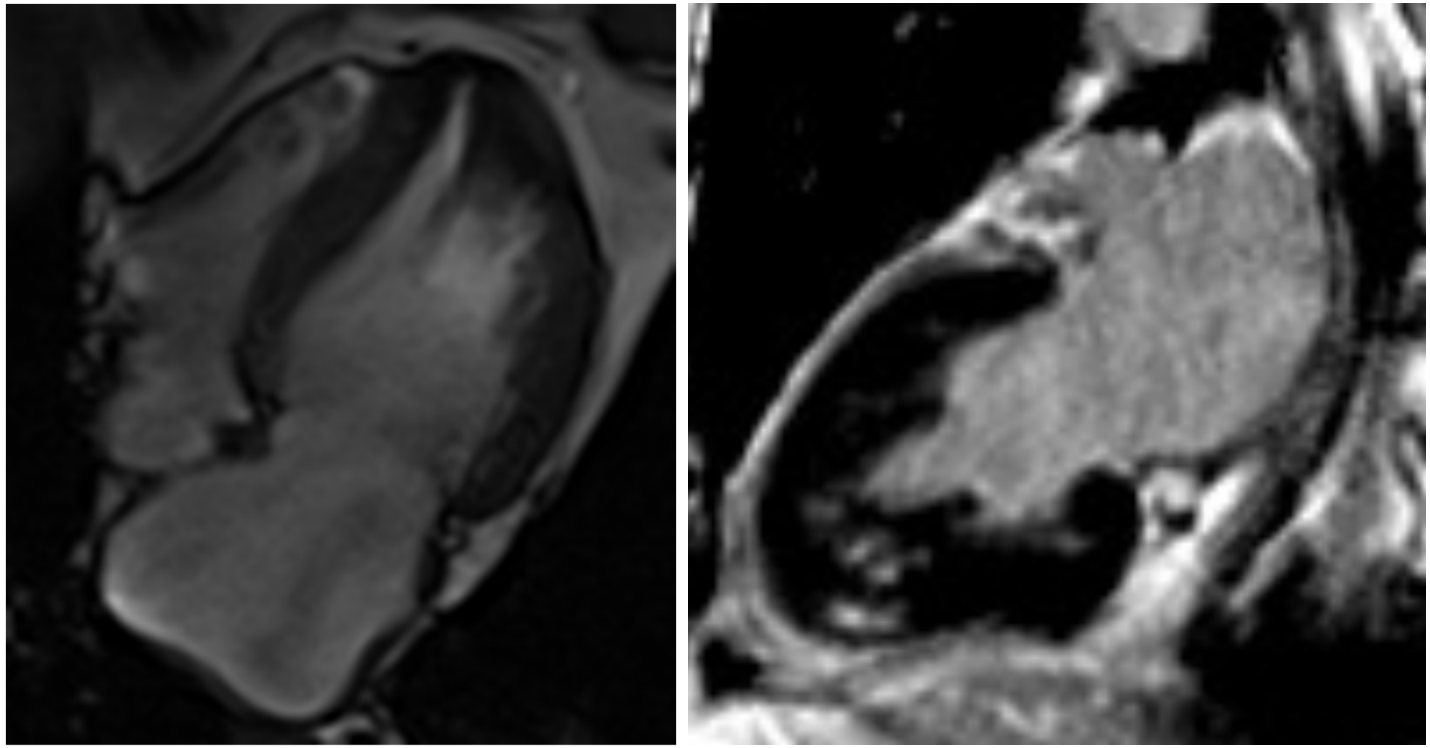


Apical HCM

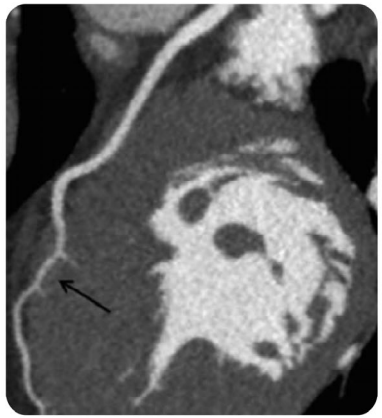
- 2-25% of HCM patients depending on ethnicity (Japanese) - Yamaguchi deeply inverted T waves
- Spade-like configuration
- Diagnostic criteria
  - Apical wall thickness > 15 mm

AND

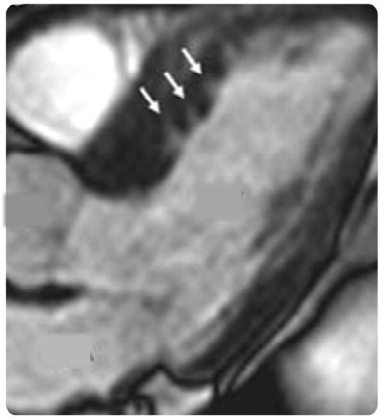
  - A ratio of apical to basal LV wall thicknesses ≥ 1.5



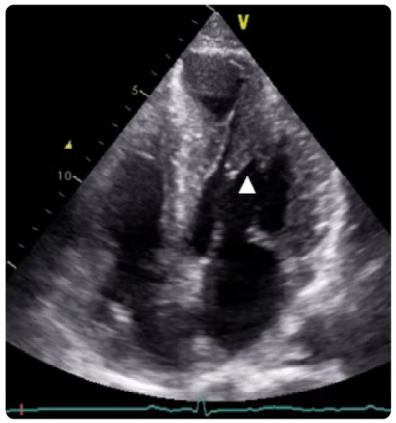
Accompanying features

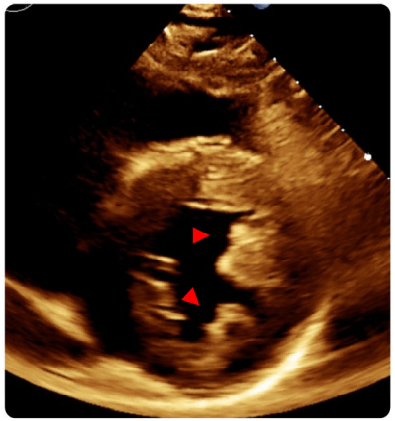


Myocardial Bridging



Myocardial crypts

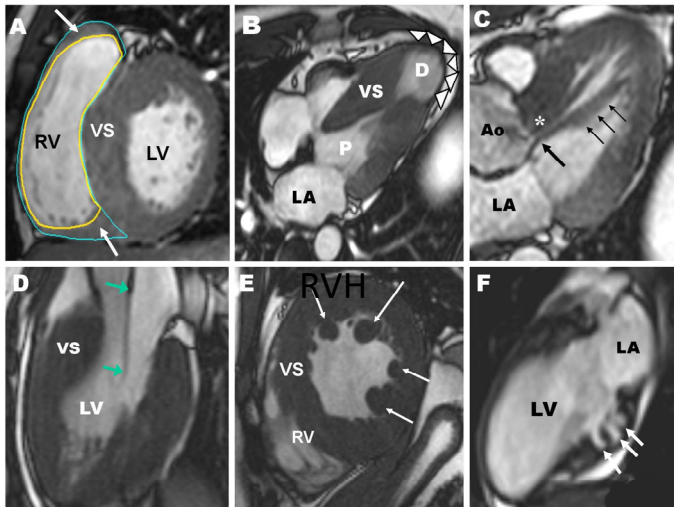




Papillary muscle abnormalities (apical displacement; bifid)

RVH

AML elongation



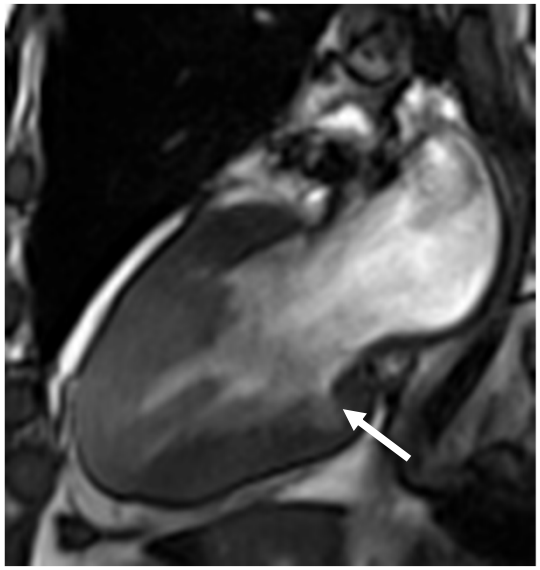
Abnormal chordae, paps

Crypts

Papillary muscle abn

Basal crypts

- Basal crypts are morphological signs of HCM
- Typically appreciated in the basal inferior/ inferolateral walls.
- Patients without phenotypic HCM, but known to carry disease-causing mutations (‘carriers’) small studies have identified a high prevalence of myocardial crypts , suggesting a potential role of crypts to identify patients who should proceed to genetic testing.

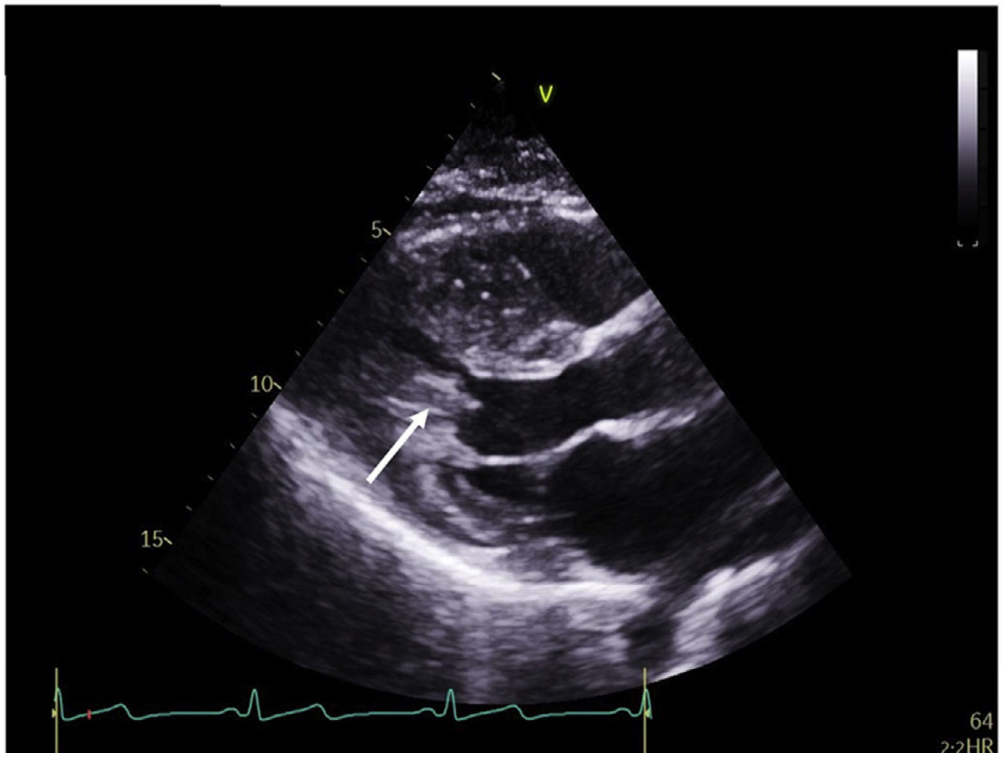


*Arrow pointing to basal crypt in the basal inferior wall in end-diastole (arrow), which is obliterated during systole.

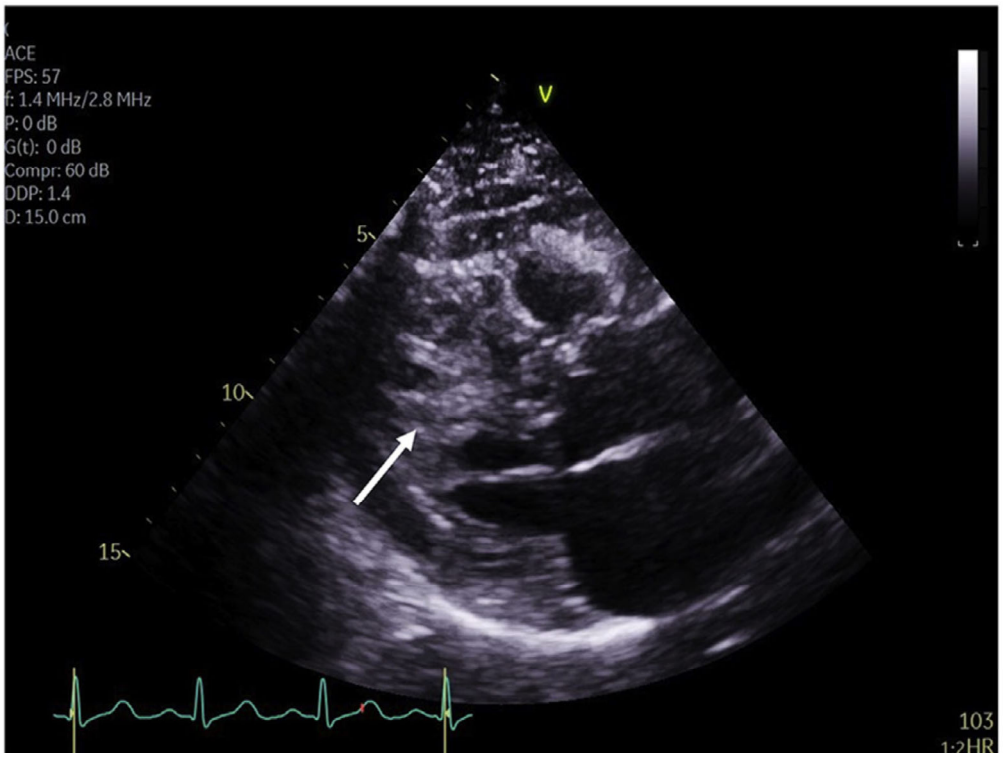
Child N et al. J Cardiovasc Magn Reson 2014;16:66.

Papillary muscle abnormalities

Anterior displacement of the papillary muscle

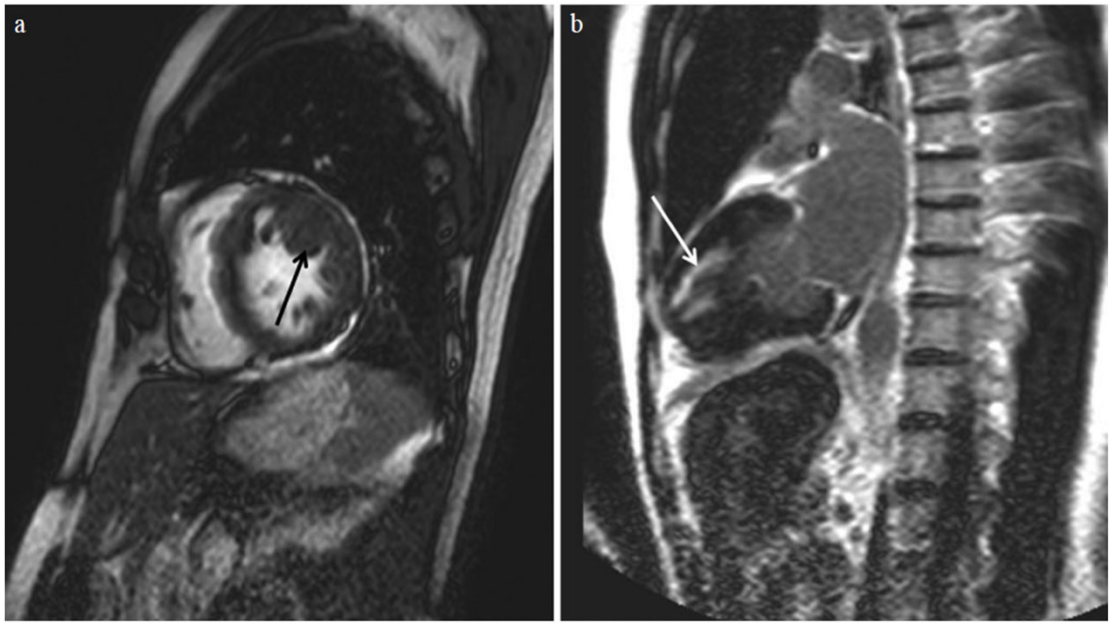


Bifid papillary muscle



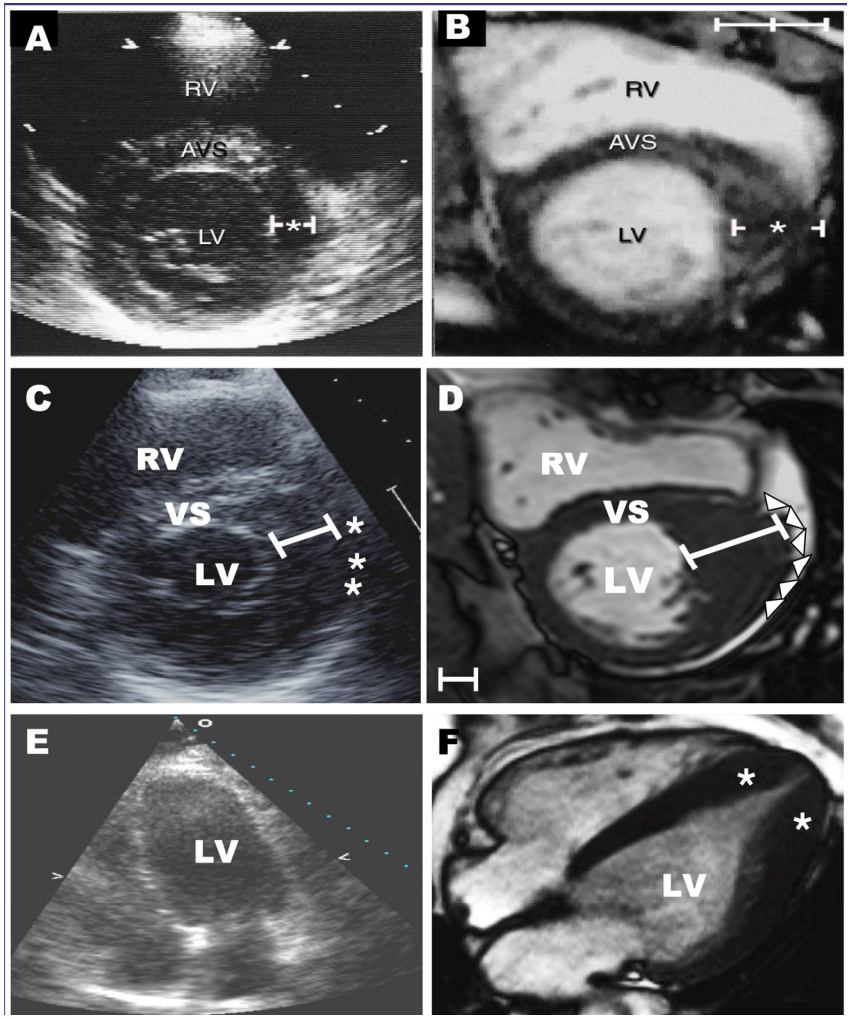
Papillary muscular abnormalities

- Hypertrophy and malposition of papillary muscles can be seen
- 1/3rd present with significant elongated anterior or posterior MVL
- When hypertrophied papillary muscles cause LVOT obstruction with clinical symptoms, myectomy combined with papillary muscle reorientation, should be performed

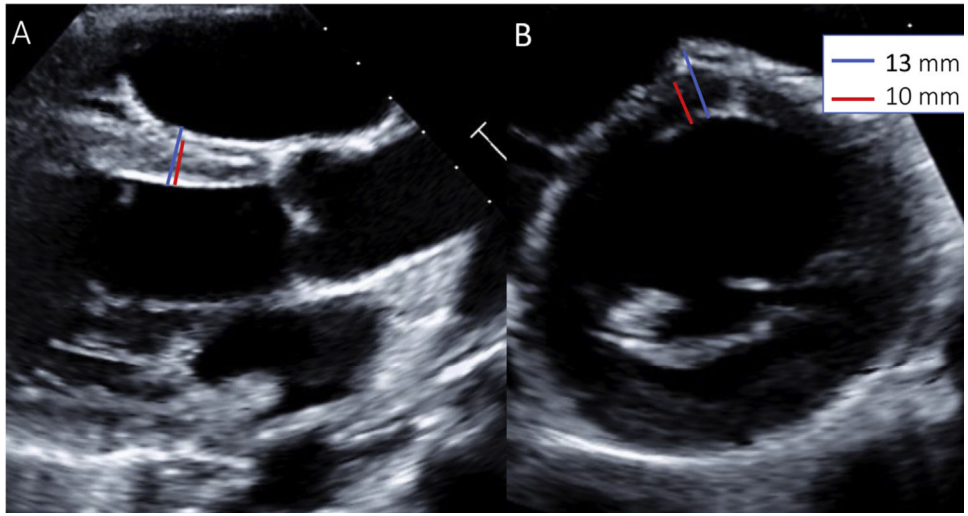


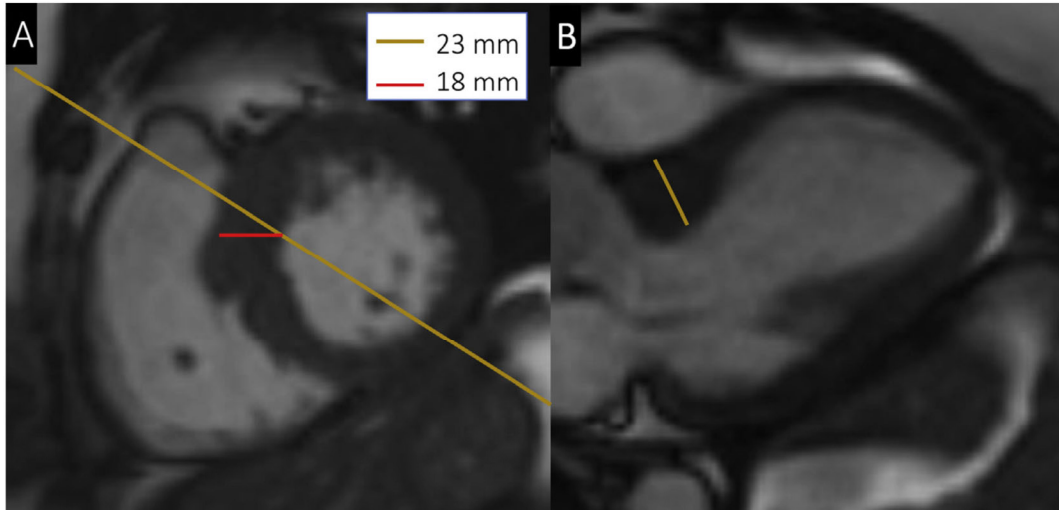
Amano Y, et al. Magn Reson Med Sci. 2018;17(2):120-31.

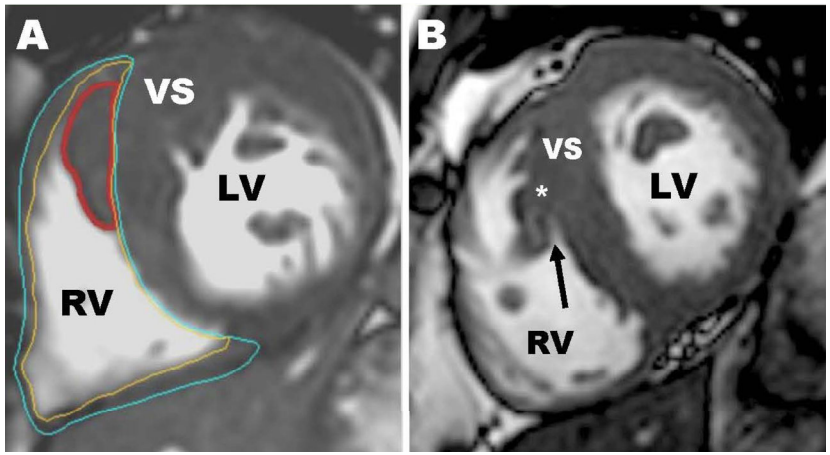
CMR “sees” LVH better than TTE



Measuring correctly

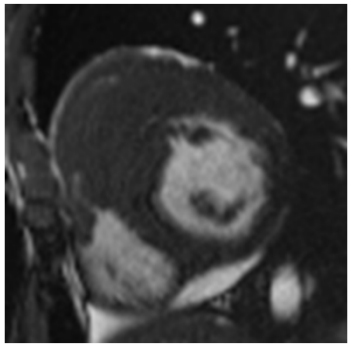


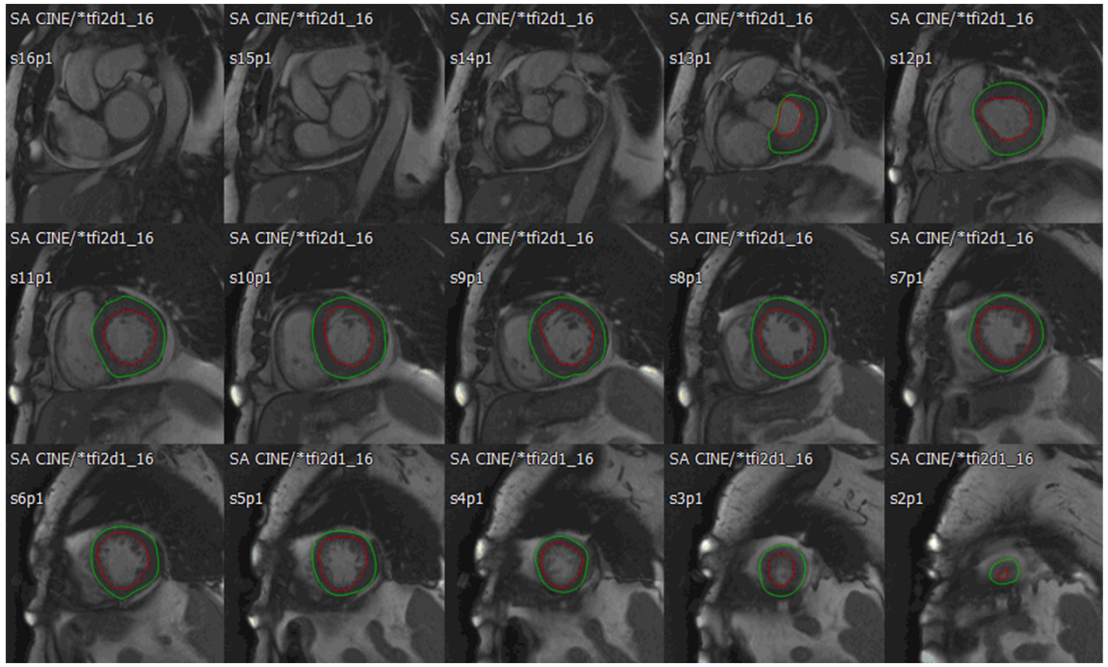




Nagueh et al, JASE 2022

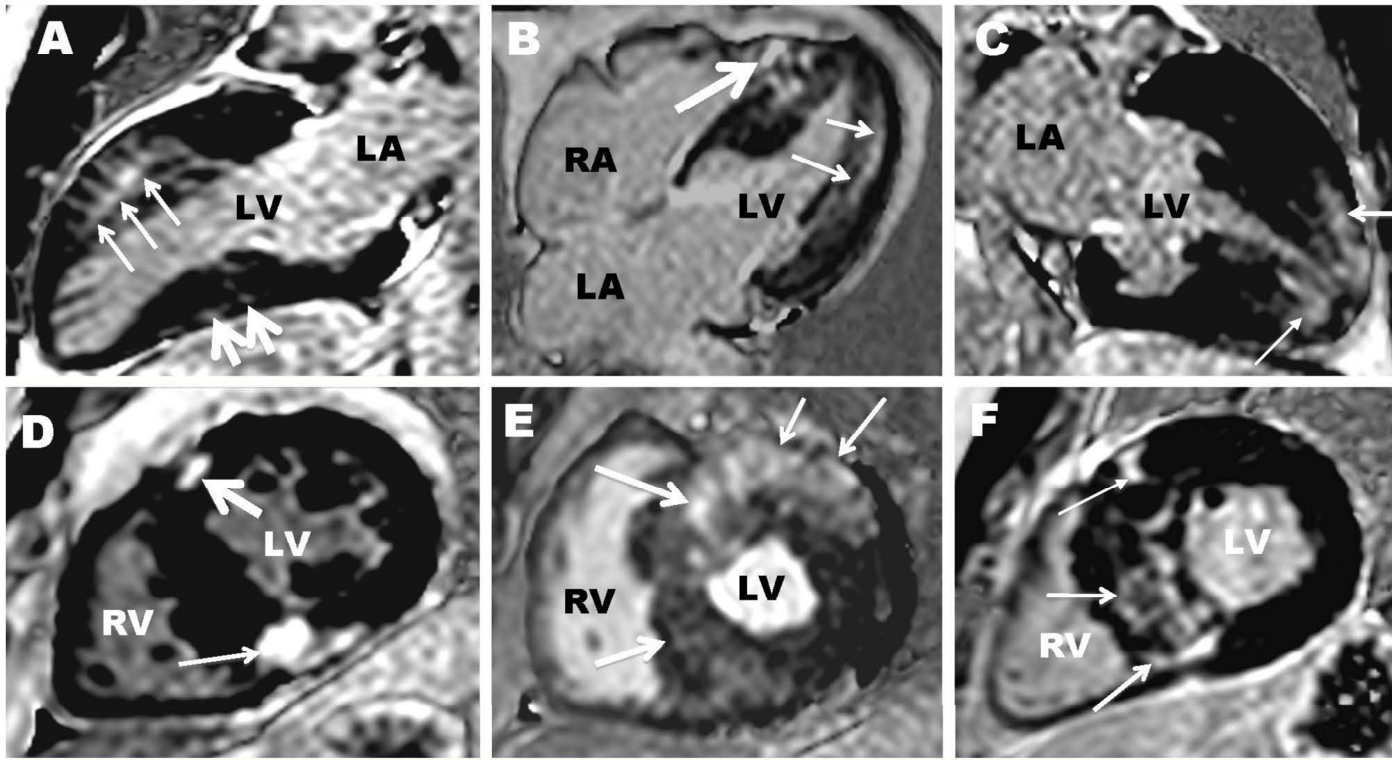
CMR is gold standard for volumes, mass





ED mass	**166.59 g**	(74-146)
EDV	**159.14 ml**	(76-160)
ESV	83.85 ml	(17-55)
SV	75.29 ml	(53-109)
EF	**47.31 %**	(59-79)
CO	5.36 l/min	
ED Mass/BSA	**103.10 g/m^2**	(48-78)
EDV/BSA	**98.43 ml/m^2**	(49-85)
ESV/BSA	**51.86 ml/m^2**	(11-31)
SV/BSA	46.57 ml/m^2	(34-60)
CO/BSA	3.32 l/(min*m^2)	

LGE patterns



- LGE identifies myocardial replacement fibrosis or scarring that contributes to risk stratification for HCM
- >15% LGE is significantly related to ventricular tachyarrhythmia and 2-fold increase in SCD event risk i.e. 6% risk at 5 years.
- LGE typically occurs in the segments with the greatest hypertrophy

Green JJ, Salerno M, et al. JACC CVI. 2012;5(4):370-7
Chan et al. *Circulation*.2014;130:484-495.

LGE

- LGE is present in 50% of cases
- A high proportion of patients with **reverse septal curvature hypertrophy and apical aneurysm** patterns have LGE
- Isolated basal septal hypertrophy demonstrates LGE less frequently
- The reverse septal curvature pattern is associated with the majority (79%) of cases with >10% LGE
- In patients with LGE present, ESC risk score is higher than those without LGE

TABLE 4 LGE Amount by HCM Morphology

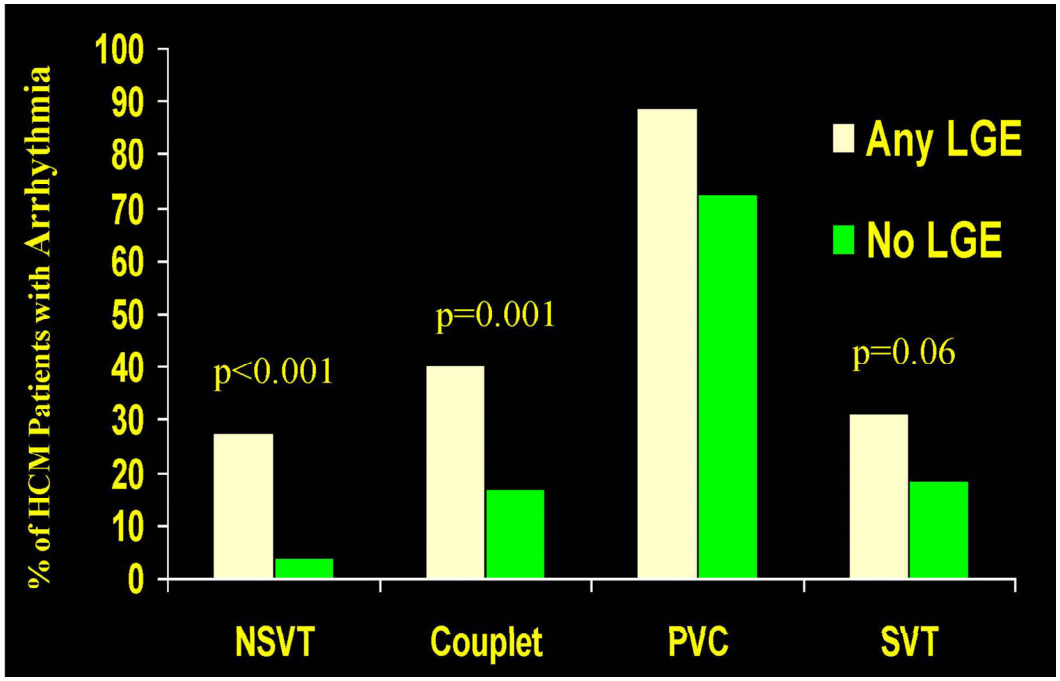
	No LGE (n = 1,265)	<5% (n = 990)	5%-10% (n = 182)	10%-15% (n = 54)	>15% (n = 46)
Isolated basal septal	767 (66.5)	353 (30.6)	25 (2.2)	8 (0.7)	0 (0.0)
Reverse curvature septal	322 (31.4)	498 (48.5)	127 (12.4)	36 (3.5)	43 (4.2)
Apical	116 (54.2)	81 (37.8)	15 (7.0)	1 (0.5)	1 (0.5)
Concentric	19 (57.6)	11 (33.3)	0 (0.0)	3 (9.1)	0 (0.0)
Apical aneurysm	25 (32.1)	35 (44.9)	13 (16.7)	5 (6.4)	0 (0.0)
Other	16 (48.5)	12 (36.4)	2 (6.1)	1 (3.0)	2 (6.1)

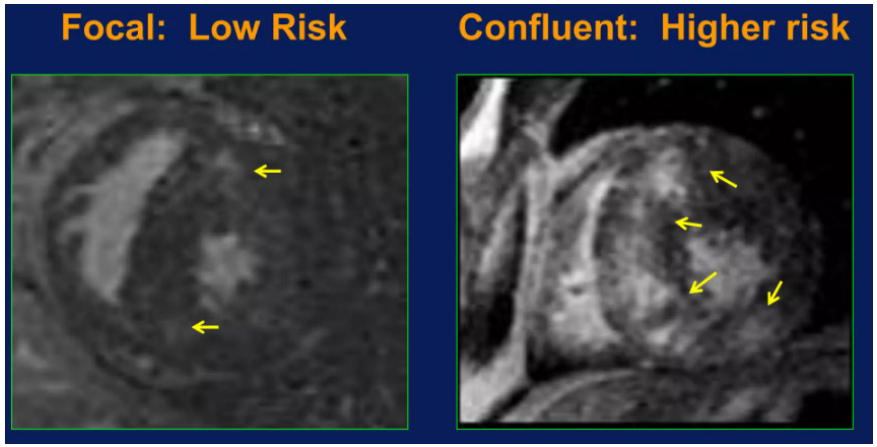
Values are n (%).

HCM = hypertrophic cardiomyopathy; LGE = late gadolinium enhancement.

Neubauer, S, Kramer CM, et al. JACC, 2019. 74(19): p. 2333-2345

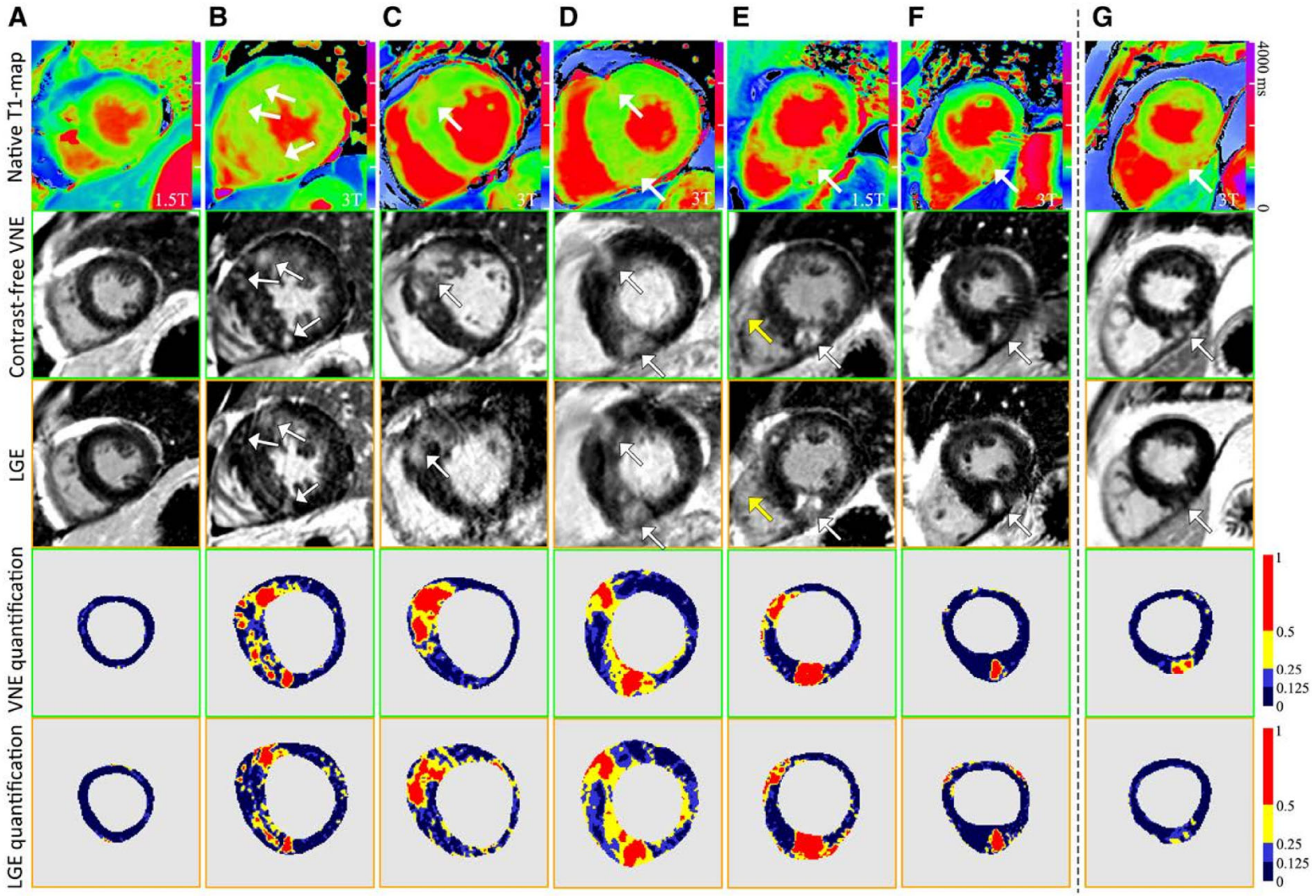
LGE & arrhythmias





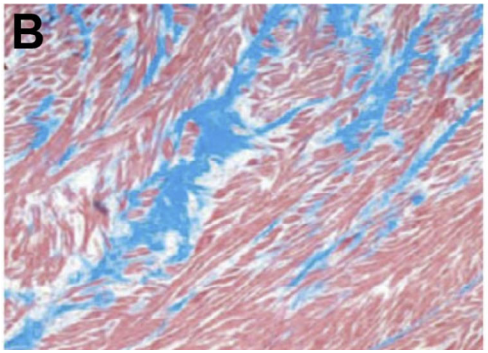
Adabag et al, JACC 2008

Noncontrast virtual LGE using AI



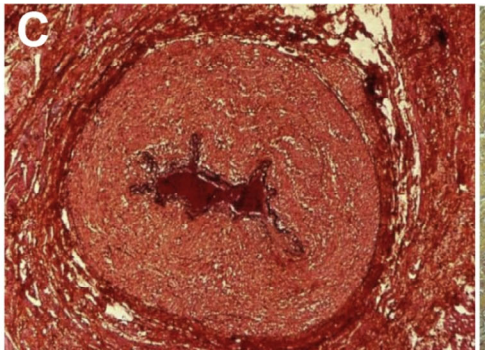
Zhang et al, Circ 2021

Linking imaging to see pathological abnormalities



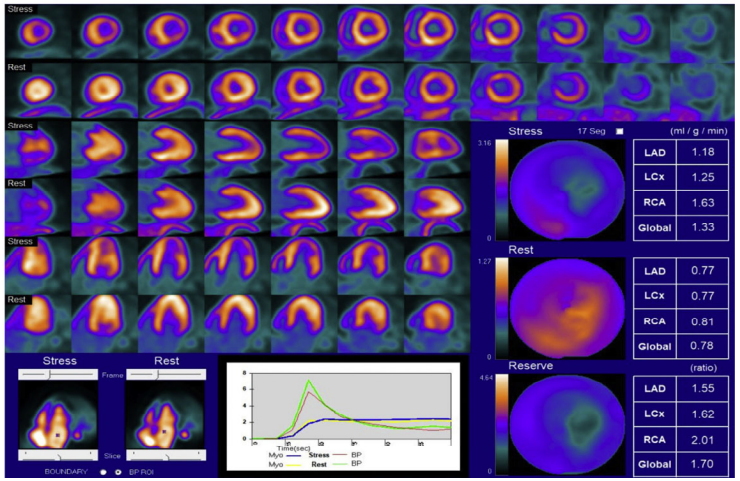
Interstitial fibrosis

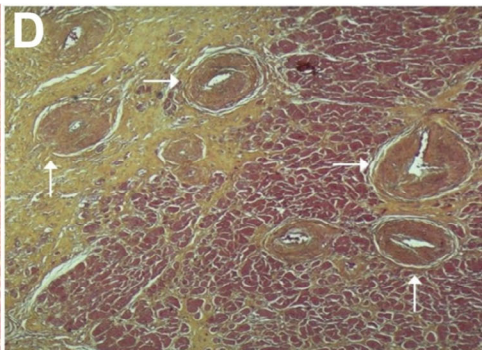
↑ T1



Smooth muscle hypertrophy, intimal hyperplasia, CMD (coronary microvascular disease)

PET

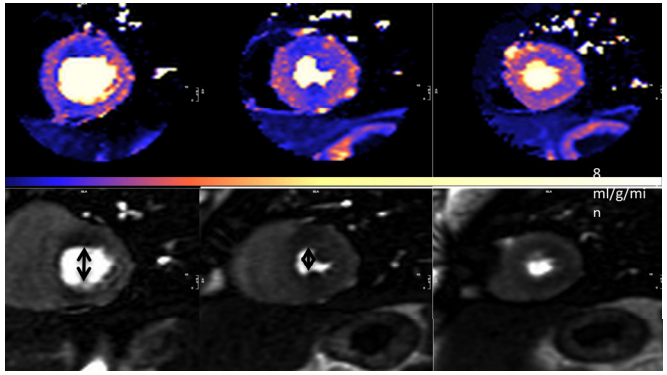




Replacement fibrosis

Focal LGE

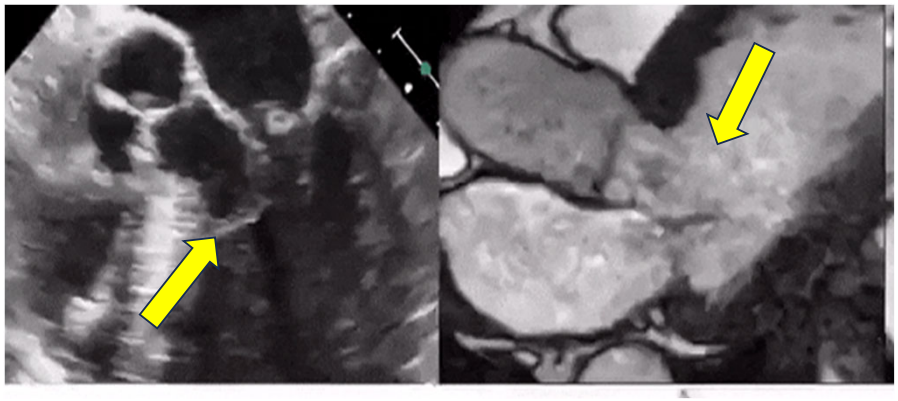
Stress perfusion CMR

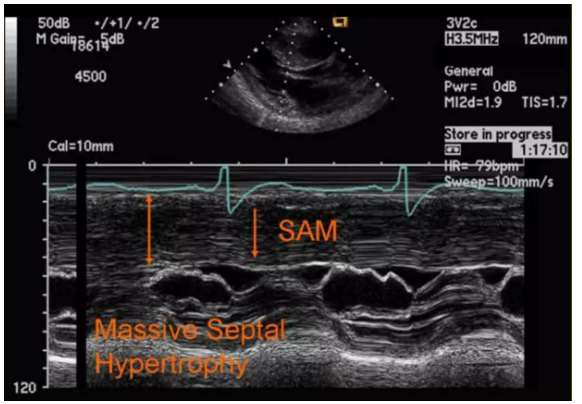


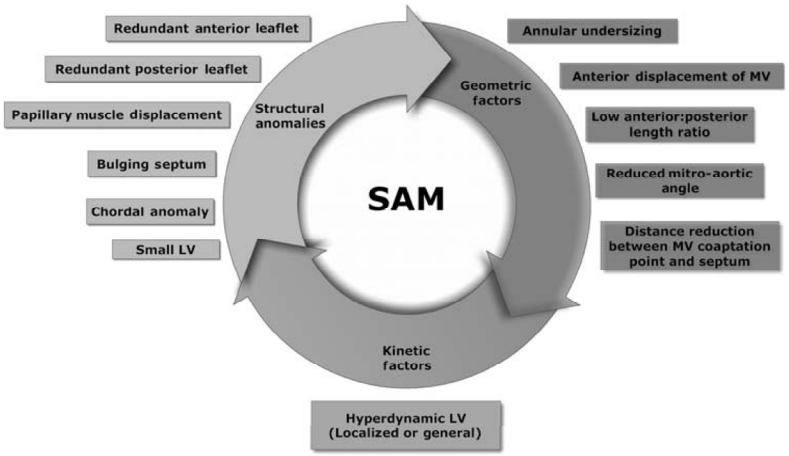
Apical ischemia common in HCM

Hughes et al, Circ cvi 2023

SAM and LVOTO



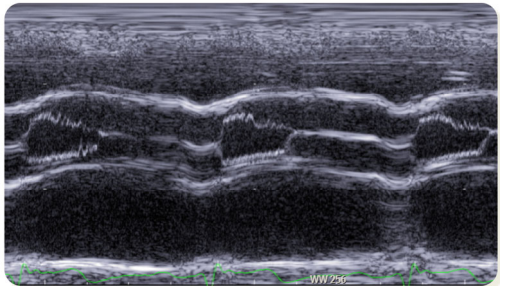




1/3rd: LVOTO at rest >30 mm Hg
1/3rd: LVOTO with provocation (exercise, dobutamine, Valsalva) >30 mm Hg
1/3rd: no LVOTO

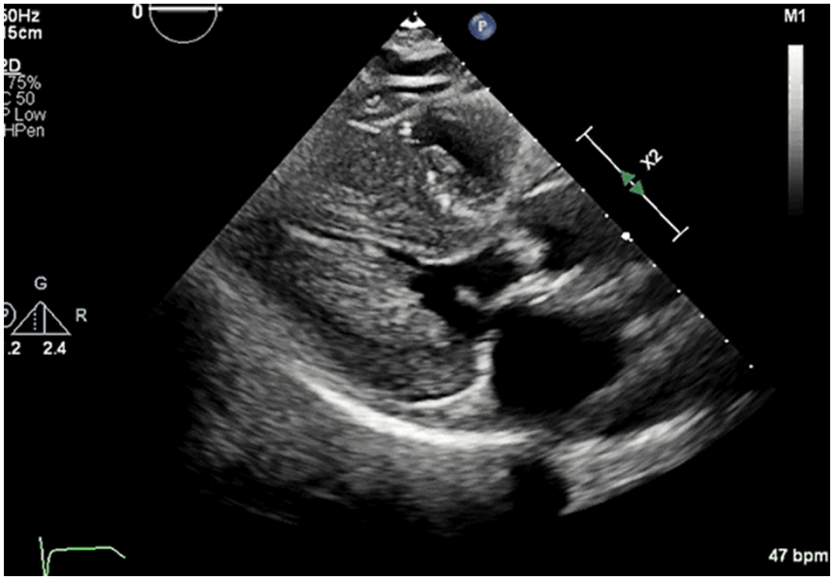
Dynamic, depends on load

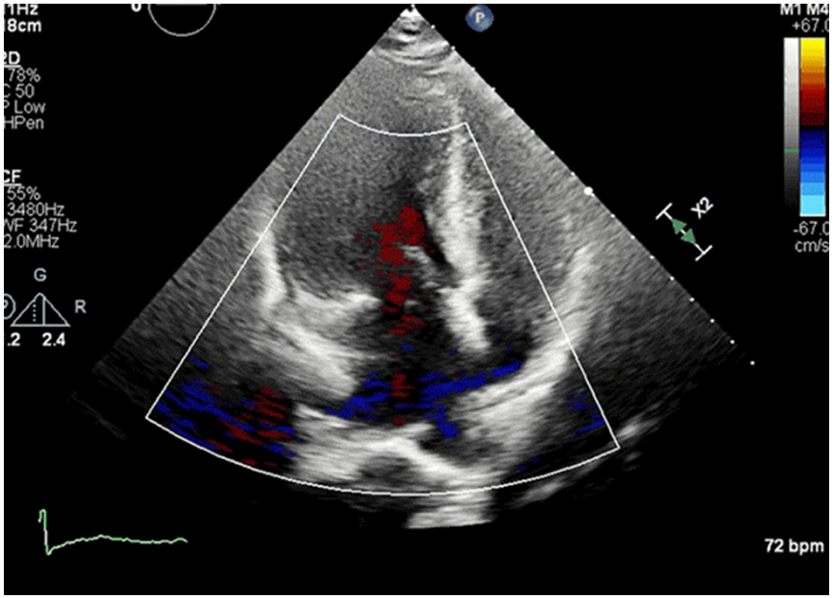
Must confirm SAM and exclude subaortic membrane, mitral leaflet abn and midcavity obstruction



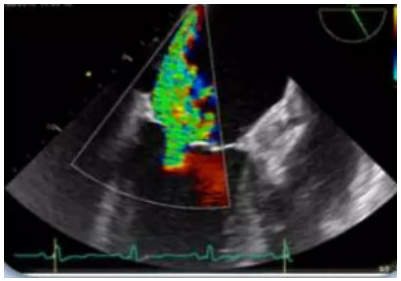
Aortic valve: mid systolic fluttering & closure

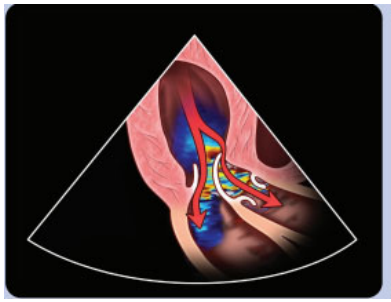
SAM

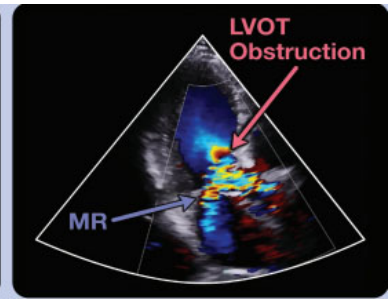




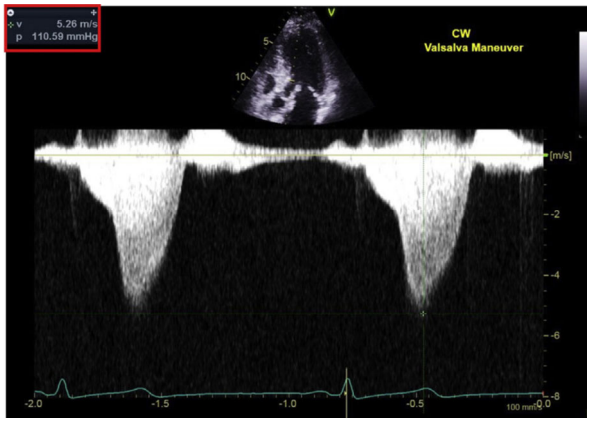
- Mid systolic closure, fluttering, fibrotic changes at contact point of AML-septum
- MR – mid to late systolic
- LAI > 34 ml/m2 associated w more LVH and DD, and predicts MACE



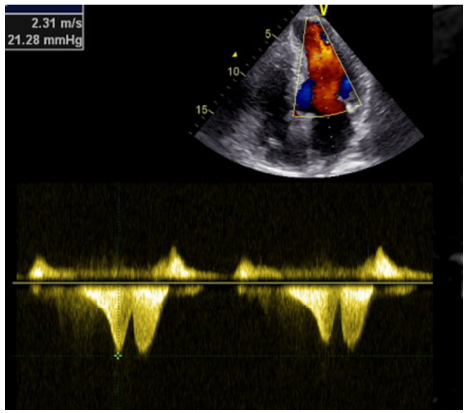




SAM is a dynamic process with variation

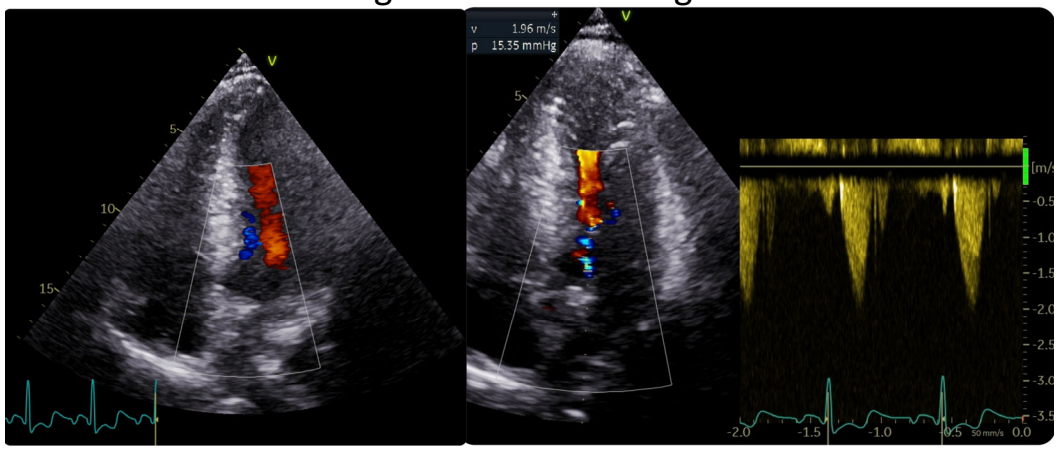


LVOTO>50 mm Hg – significant, rest or exercise or dobutamine or Valsalva >30 mm Hg in trials to start Rx

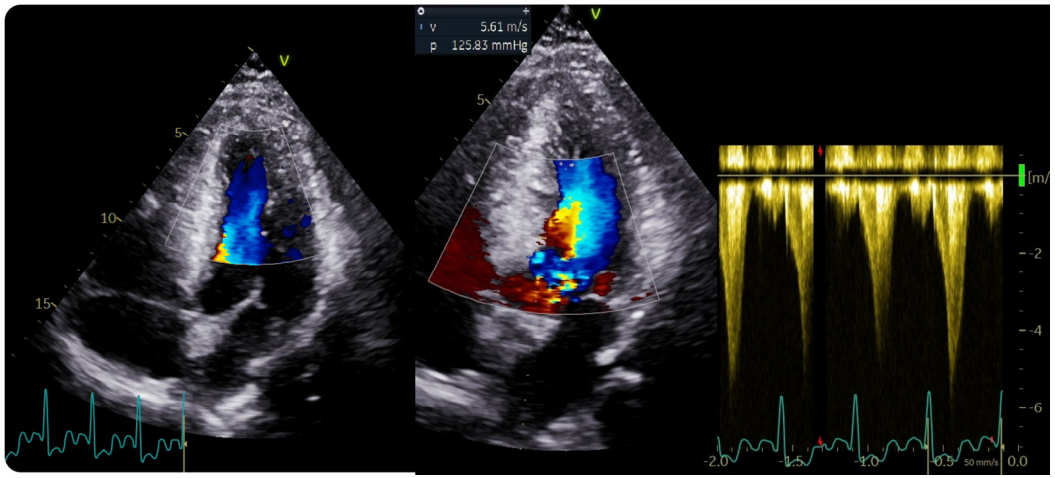


Mid cavitary obstruction (not due to SAM): lobster claw sign

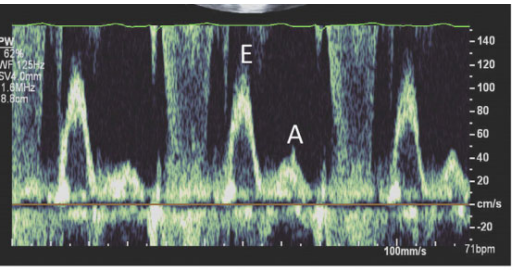
Rest gradient: 15 mm Hg

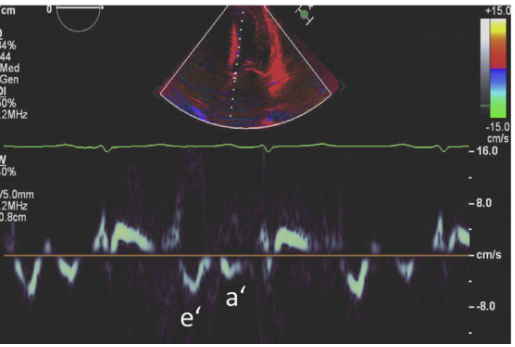


Post treadmill exercise gradient: 15 mm Hg



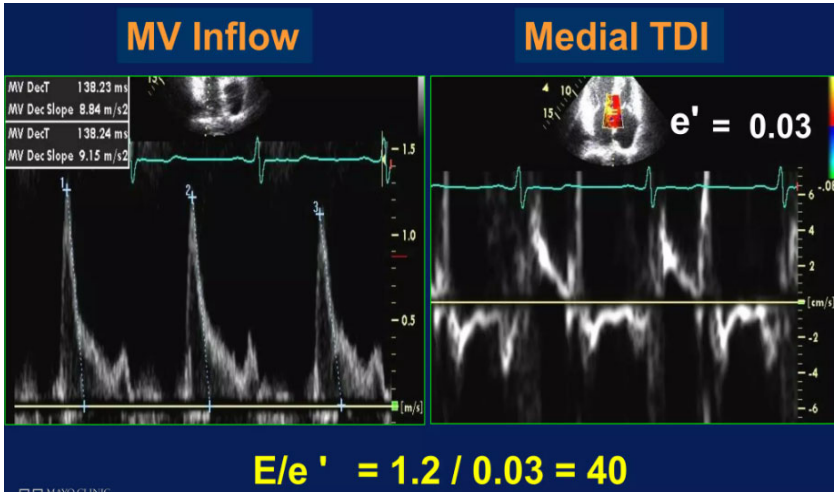
Diastolic dysfunction





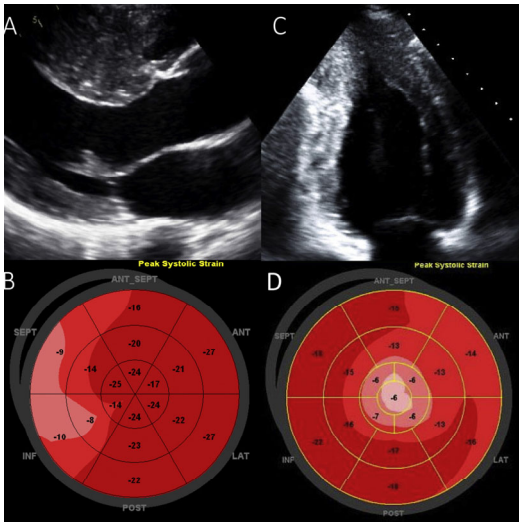
Diastolic dyfn:

- Universal
- Present even in asymptomatic pts with + phenotype and even in early pre-phenotypic stage
- Grade 2 or 3 in symptomatic pts
- ↑E
- ↓e'
- ↑E/e' (>14), ↑LAP



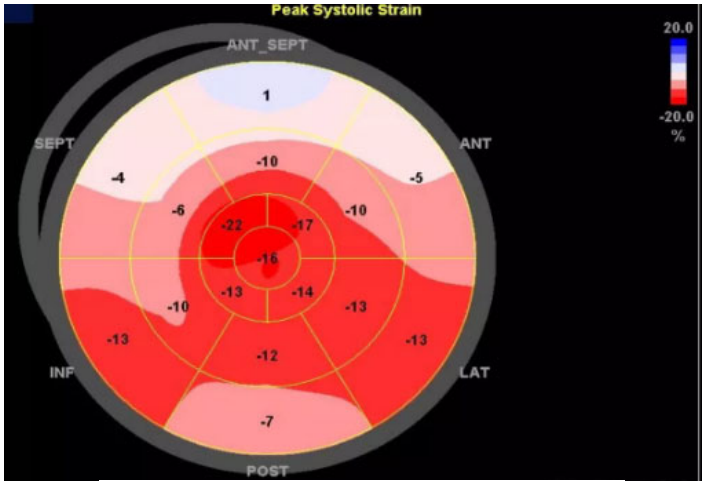
Nagueh et al, JASE 2022

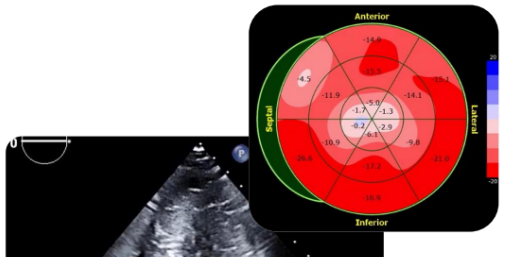
Echo Strain abnormalities

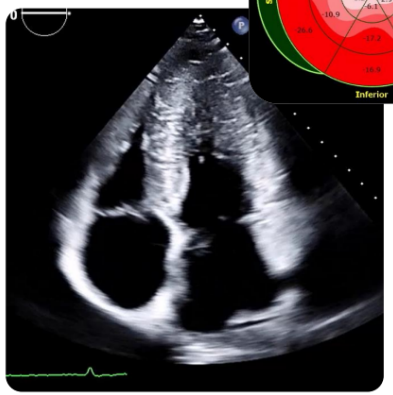


Longitudinal strain:

↓ Regional strain in areas of LVH
↓ Global (<-17%)
↓ LS with base to apex gradient
↑ CS
↓ untwisting (diastole)
Normal twist/torsion (systole)







Apical variant of HCM: apical LVH with cavity obliteration

TEE during myectomy

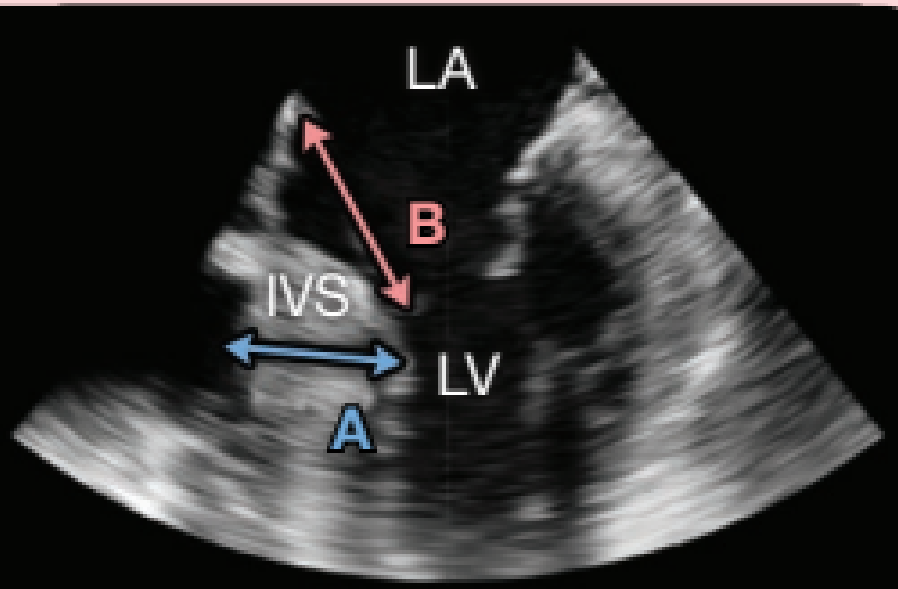
Preoperative measurements include:

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

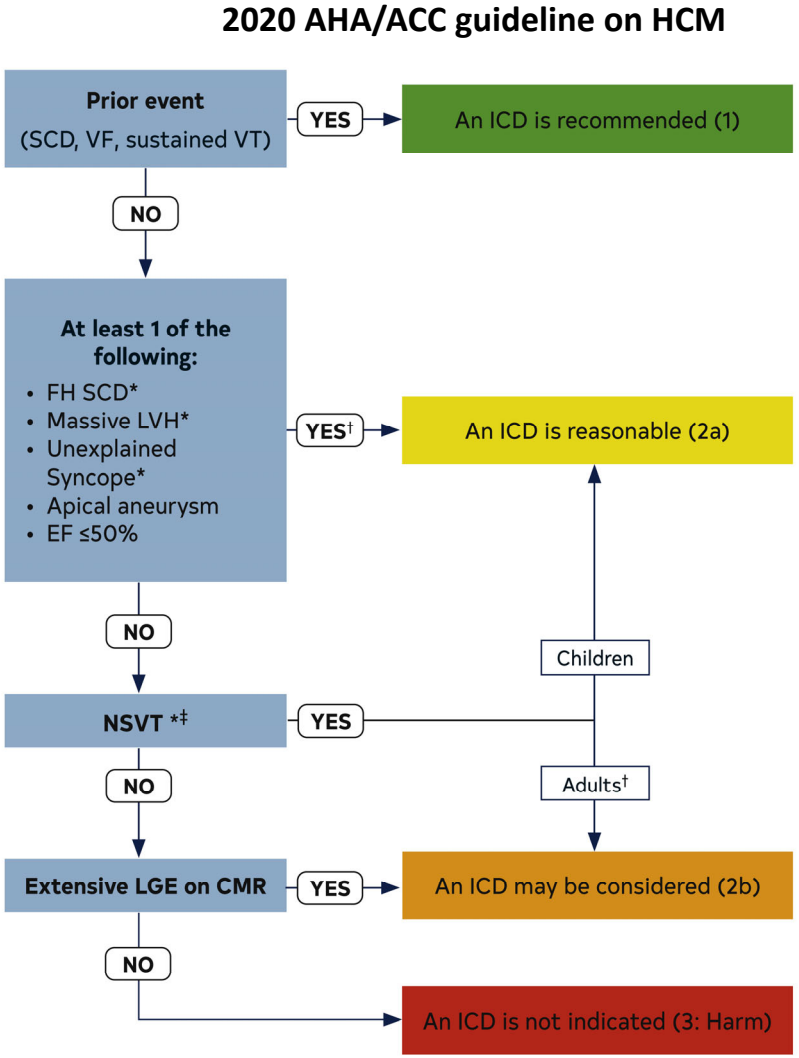
SCD in HCM: Imaging plays a key role

Predictors of SCD

1. Massive LVH (> 30 mm)
2. LVEF <50%
3. Apical aneurysm
4. LGE>15%

Imaging

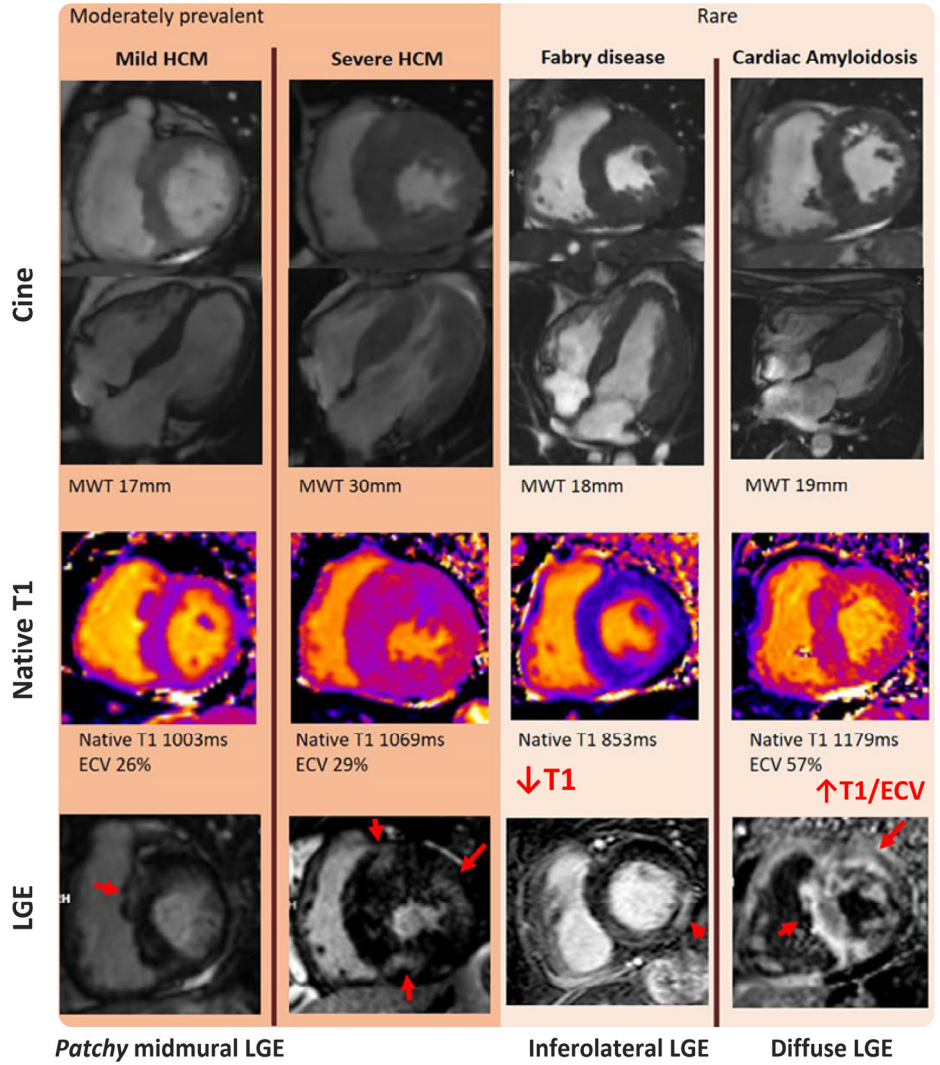
5. SCA (personal/FHx)
6. Syncope
7. Exercise hypotension
8. NSVT



Ommen et al, 2022

Not all “LVH” is HCM

- Amyloid
  - Symmetric left ventricular thickening
  - Rapid wash-out of gadolinium from blood and myocardium or endomyocardial LGE
  - **Very elevated native T1 and ECV**
- Athlete’s Heart
  - HCM is most common cause of SCD in athletes
  - Symmetric wall thickness <1.6cm
  - Utilize the diastolic wall thickness and left ventricular end-diastolic volume ratio (DWT/LVEDV) <0.15mm/m^2/mL
  - **Normal ECV**
- Fabry’s Disease
  - Concentric but can have asymmetric septal thickening
  - Delayed gadolinium-enhanced imaging typically mid wall and basal inferolateral segments
  - **Low native T1**
- Hypertensive CM
  - **Symmetric LV wall thickening with no SAM**
  - Linear or patchy LGE at the septal or inferior wall
- Thickened LV apex
  - Differential includes: LV thrombus, hypertrabeculation, noncompaction, hypereosinophilic CM
  - SSFP and delayed gadolinium enhanced imaging



Perry et al, EHJ 2014

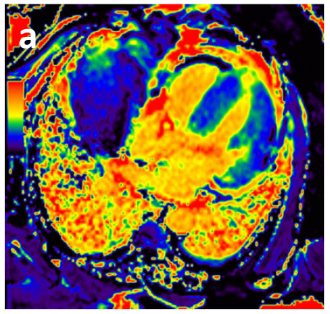
Athlete's heart/hypertrophy

- HCM - both cellular hypertrophy and interstitial fibrosis resulting in an increase in ECV
- In athletic hypertrophy, there is predominately myocyte hypertrophy without significant fibrosis resulting in a reduction in the ECV
- Normal ECV values = 25.3 ± 3.5%

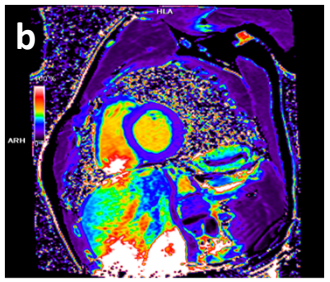
HCM vs athlete's heart

- Asymmetric
- Mitral S'< 9 cm/s, abnormal LS
- WT>20 mm (especially non-Black)
- Diastolic dysfunction
- LVH doesn't regress (after exercise holiday)
- Small LV cavity (EDD<45 mm)
- SAM and LVOTO
- Intraventricular dyssynchrony > 45 ms

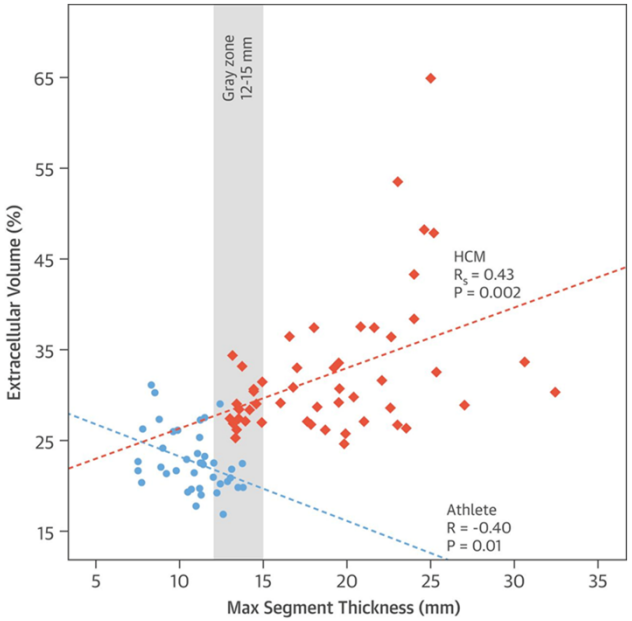
ECV maps



HCM (ECV = 42%)



Athlete's heart (ECV = 23%)



50 HCM, 40 athletes, 35 controls.
AUC RUC for ECV 0.94

Swoboda PP, et al. J Am Coll Cardiol. 2016;67(18):2189-90.

Gottbrecht M, Kramer CM, Salerno M. Radiology 2019;290:317-26.

Echocardiography Recommendations in Hypertrophic Cardiomyopathy

COR	LOE	Recommendations
1	B-NR	1. In patients with suspected HCM, a TTE is recommended in the initial evaluation.
1	B-NR children	2. In patients with HCM with no change in clinical status or events, repeat TTE is recommended every 1 to 2 years to assess the degree of myocardial hypertrophy, dynamic LVOTO, MR, and myocardial function.
1	C-LD adults	2. In patients with HCM with no change in clinical status or events, repeat TTE is recommended every 1 to 2 years to assess the degree of myocardial hypertrophy, dynamic LVOTO, MR, and myocardial function.
1	B-NR	3. For patients with HCM who experience a change in clinical status or a new clinical event, repeat TTE is recommended.
1	B-NR	4. For patients with HCM and resting LVOT gradient <50 mm Hg, a TTE with provocative maneuvers is recommended.

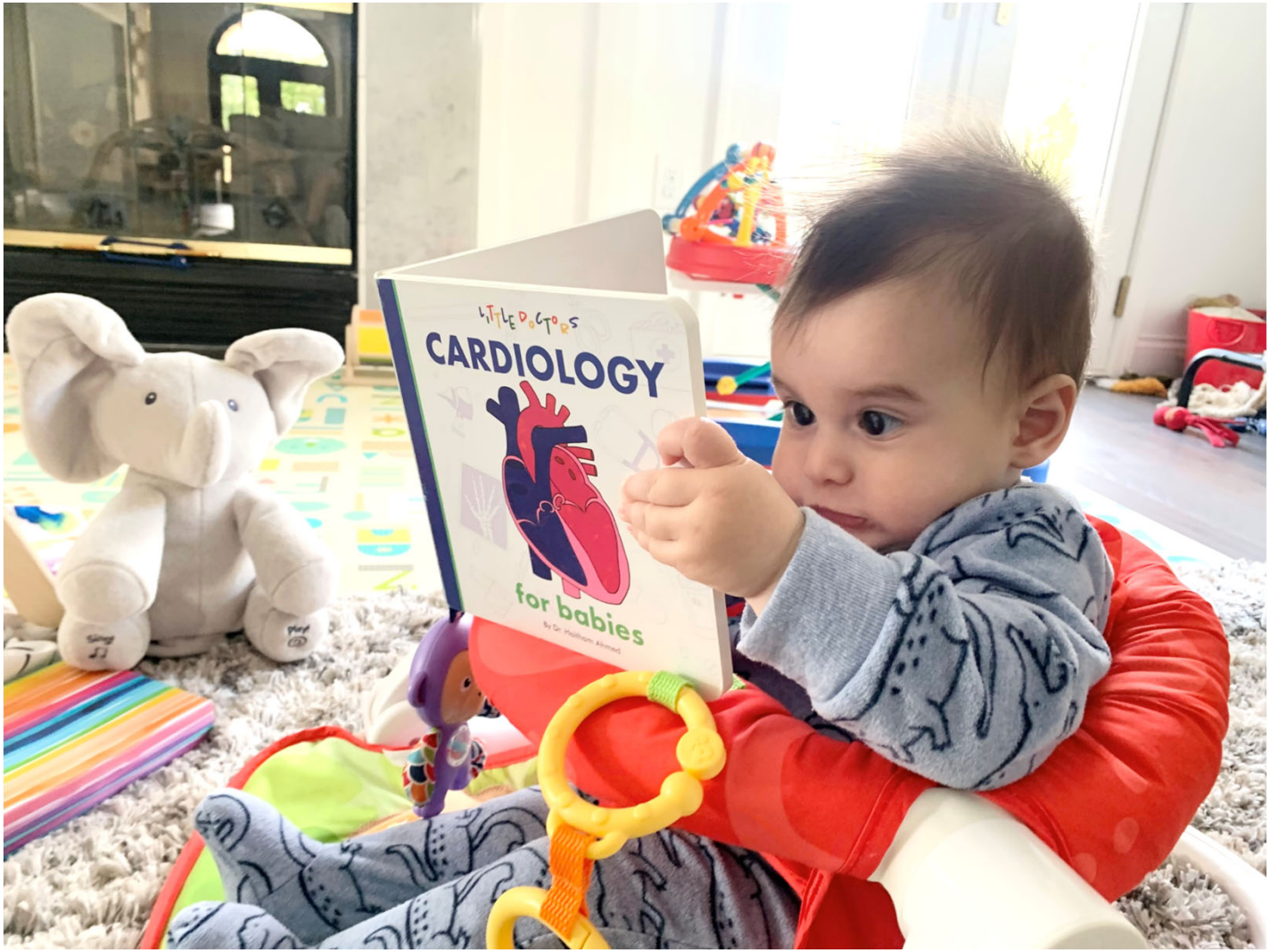
Ommen et al, Circ 2020

Cardiovascular Magnetic Resonance (CMR) Imaging Recommendations in HCM

COR	LOE	Recommendations
1	B-NR	1. For patients suspected to have HCM in whom echocardiography is inconclusive, CMR imaging is indicated for diagnostic clarification
1	B-NR	2. For patients with LVH in whom there is a suspicion of alternative diagnoses, including infiltrative or storage disease as well as athlete's heart, CMR imaging is useful.
1	B-NR	3. For patients with HCM who are not otherwise identified as high risk for SCD, or in whom a decision to proceed with ICD remains uncertain after clinical assessment that includes personal/family history, echocardiography, and ambulatory electrocardiographic monitoring, CMR imaging is beneficial to assess for maximum LV wall thickness, ejection fraction (EF), LV apical aneurysm, and extent of myocardial fibrosis with LGE.
1	B-NR	4. For patients with obstructive HCM in whom the anatomic mechanism of obstruction is inconclusive on echocardiography, CMR imaging is indicated to inform the selection and planning of SRT.

Ommen et al, Circ 2020

Thank You

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Thank You