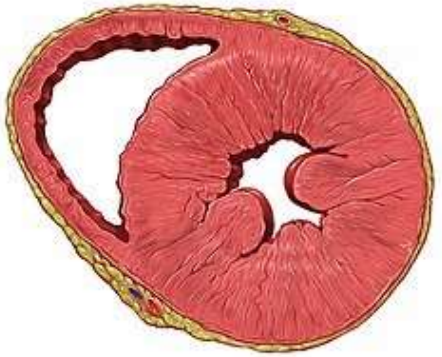
Inside the Hypertrophic Heart: Contemporary Insights Into HCM Pathophysiology



Professor Marc Dweck

Professor of Clinical Cardiology & Consultant Cardiologist

Vice-President EACVI

PACE-CME

Disclosures

- Professor Dweck is supported by the British Heart Foundation (FS/SCRF/21/32010) and is the recipient of the Sir Jules Thorn Award for Biomedical Research 2015 (15/JTA). He is Director of Image Analysis Core Lab within the Edinburgh Clinical Research Facility, University of Edinburgh.
- Professor Dweck has received speaker fees from Pfizer, Radcliffe Cardiology, Amarin, Bristol Myers Squibb, Edwards and Novartis. He has received consultancy fees from Novartis, Jupiter Bioventures, Astra-Zeneca, Novo Nordisk, UCB Biopharma, Beren and Silence therapeutics.

Most commonly inherited cardiovascular disease[1]

Estimated prevalence is **1:500**[2]
~86% of people are undiagnosed[3]
M = F

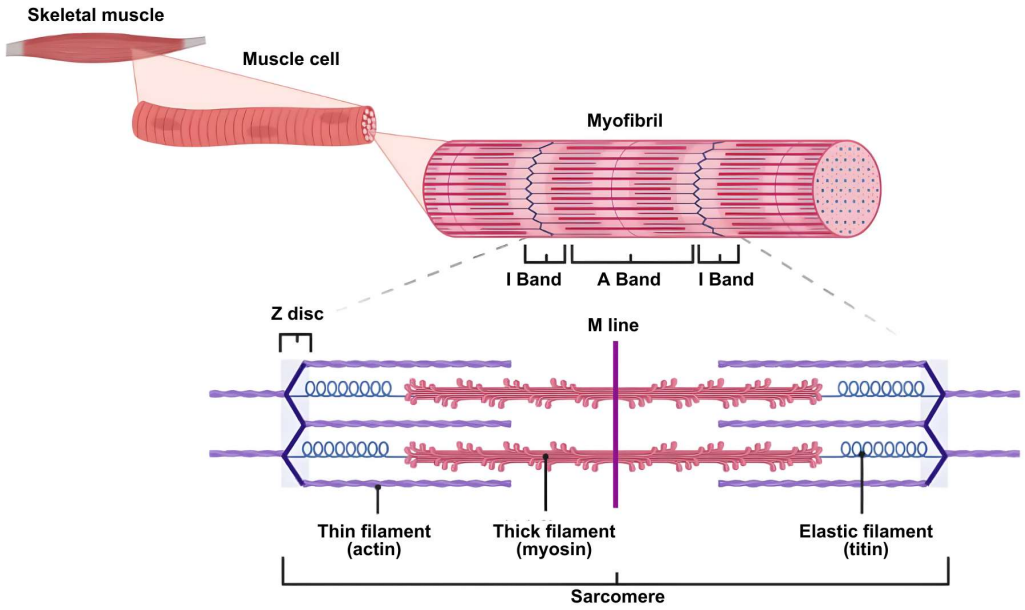
Unexplained LV wall thickness >15 mm (>13 mm in patients with a gene or first-degree relatives of patients with HCM)

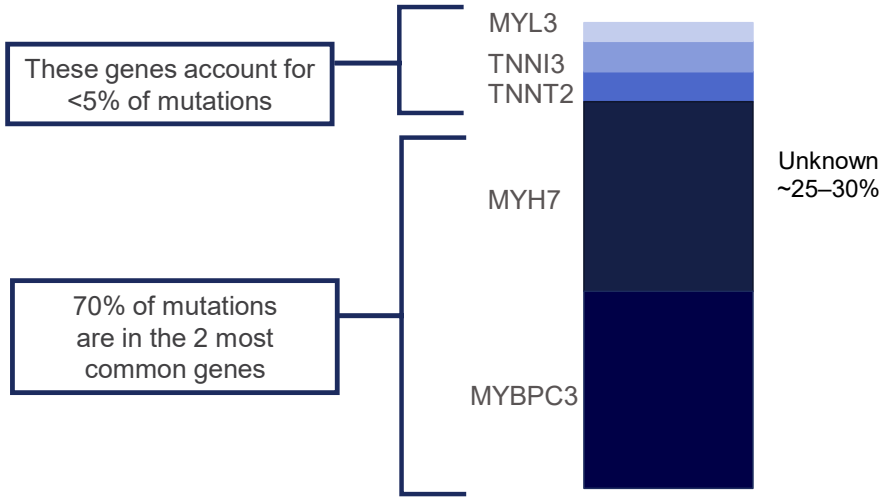
50% of cases familial
MYBCP3, MYH7
(thick filament)

oHCM, obstructive hypertrophic cardiomyopathy.

1. Maron BJ, et al. *JACC Heart Fail*. 2018;6(5):376-378. 2. Semsarian C, et al. *J Am Coll Cardiol*. 2015;65(12):1249-1254.
3. Maron MS, et al. *Am J Cardiol*. 2016;117(10):1651-1654.

A Disease of the Cardiac Sarcomere





Gene	Protein
MYL3	Myosin light chain 3
TPM1	Tropomyosin alpha-1 chain
TNNI3	Cardiac troponin I
TNNT2	Cardiac troponin T
MYH7	Beta-myosin heavy chain
MYBPC3	Cardiac myosin-binding protein C

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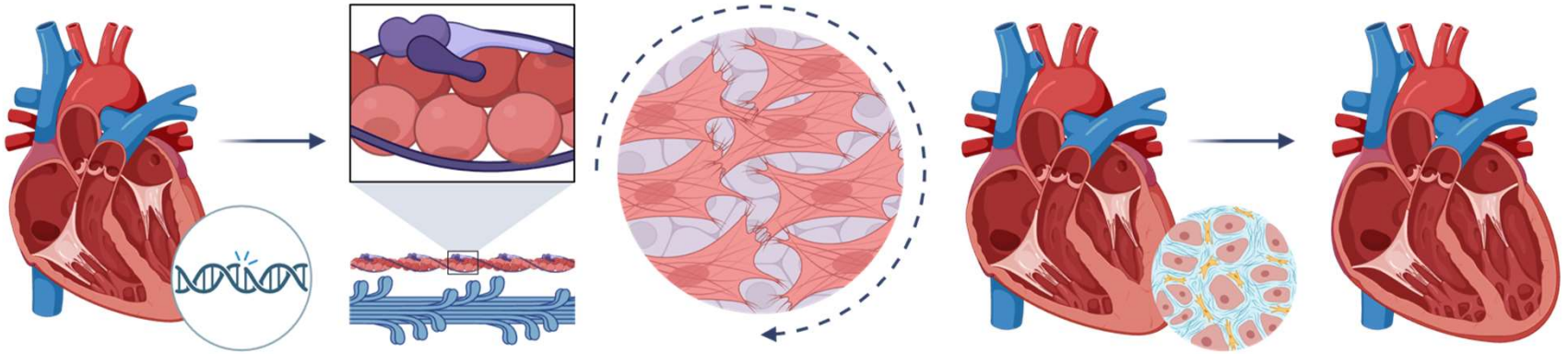
Elliott PM, et al. *Eur Heart J*. 2014;35(39):2733-2779. Maron BJ, et al. *J Am Coll Cardiol*. 2014;64(1):83-99.

Pathogenesis



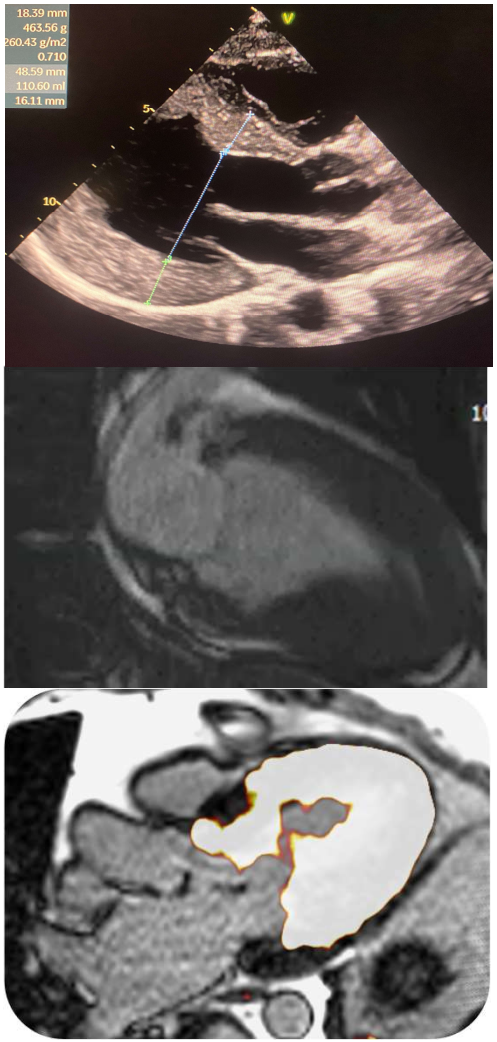
Sarcomeric and non-sarcomeric HCM

Increased actin-myosin cross-linking, resulting myocardial hypercontractility

Myocardial fibroblast activation

Left ventricular hypertrophy
Microvascular ischaemia
Myocardial disarray
Cardiac fibrosis

Adverse remodelling
Arrhythmias
Heart failure



1. Diagnosis
2. Risk stratification
3. Monitoring response to therapy

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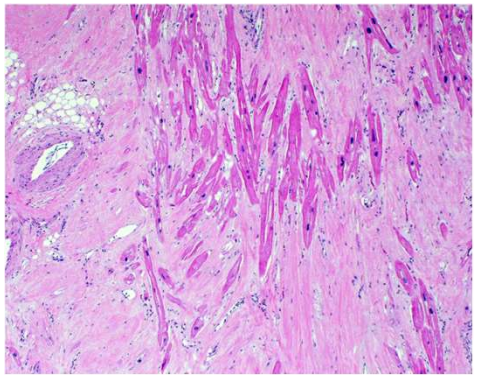
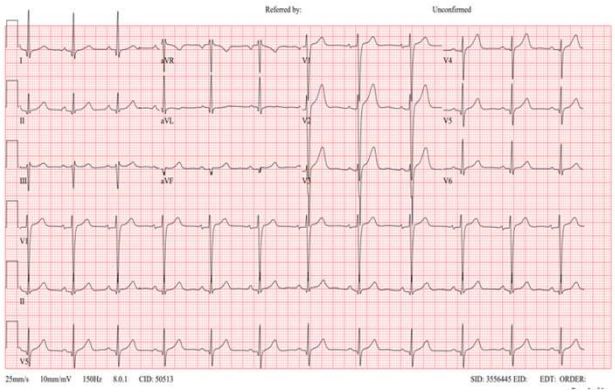
Diagnostic Tools

BIOPSY



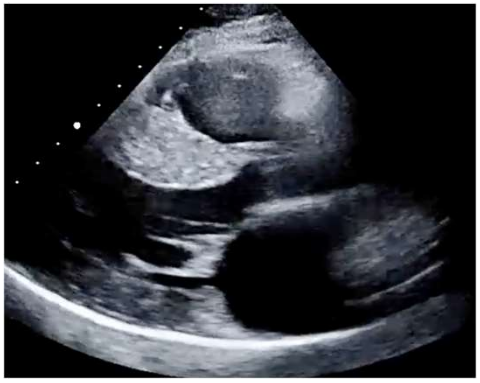
Myocyte hypertrophy,
fibrosis & disarray

ECG



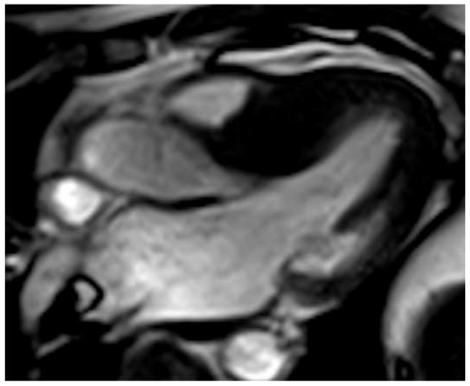
LVH, TWI, repolarisation
abnormality

ECHO



Left ventricular hypertrophy,
SAM & LVOT obstruction,
diastolic dysfunction,
LA dilatation

CMR



Left ventricular hypertrophy,
SAM & LVOT obstruction,
myocardial fibrosis (LGE, T1),
LA dilatation

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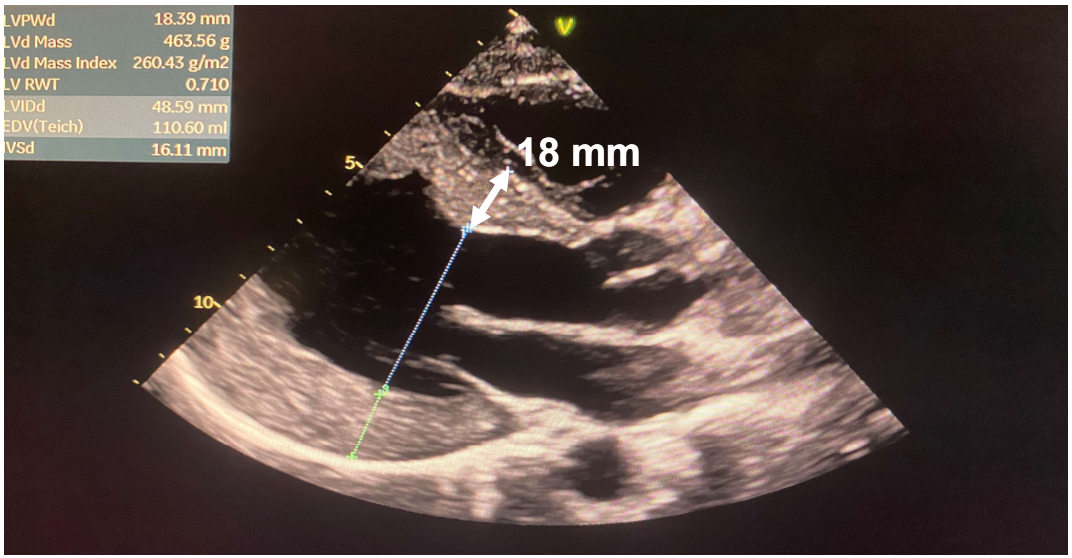
Diagnosis: Wall Thickness

- **Wall thickness >15 mm** anywhere in the left ventricle in the absence of another cause of hypertrophy
- **Wall thickness 13-15 mm** in family members of a patient with HCM or in conjunction with a positive genetic test

Nearly any pattern and distribution of wall thickening observed



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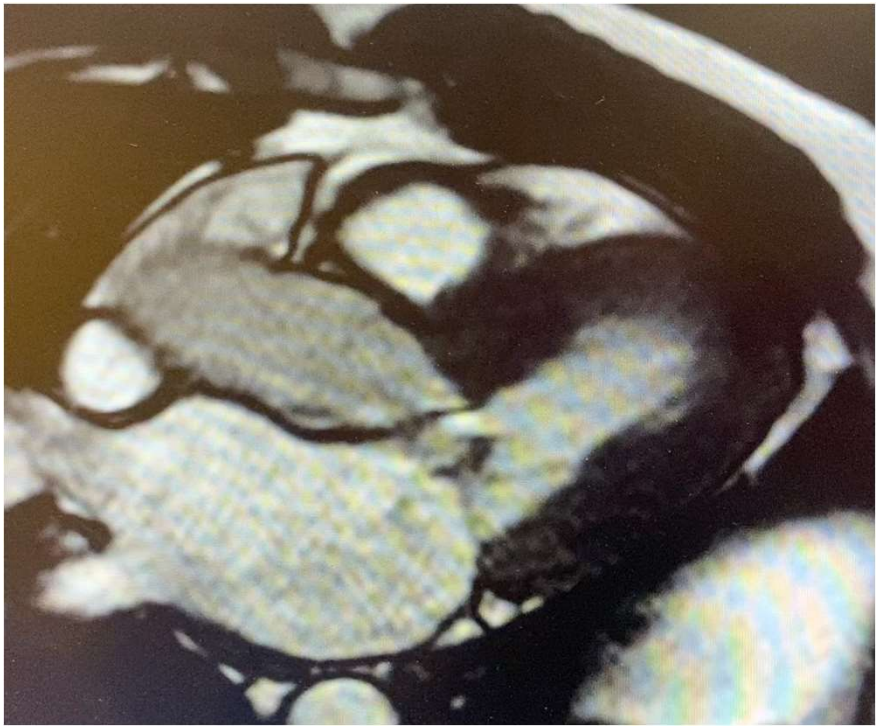
Dominguez F, et al. *Heart*. 2018;104(3):261-273.

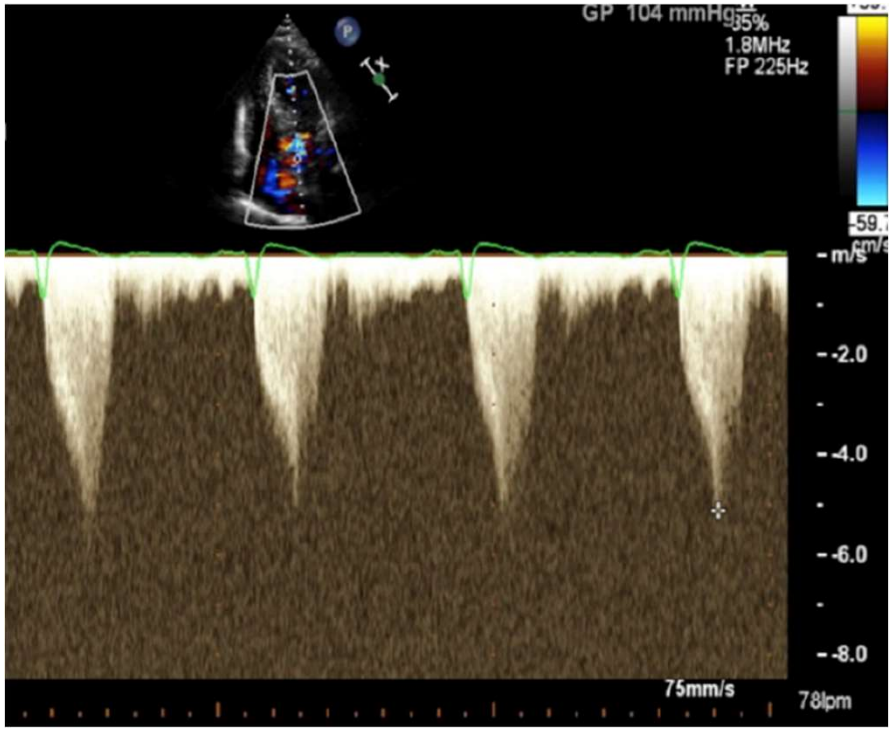
2 Main Subtypes of HCM

Normal	Non-obstructive HCM	Obstructive HCM
		

Up to 70% are obstructive.
HCM, hypertrophic cardiomyopathy.
Images from: Hypertrophekardiomyopathie. HCM im Überblick. https://www.hypertrophekardiomyopathie.de/hcmhocm-im-uberblick

Obstructive HCM





Continuous-Wave Doppler
Dagger-shaped Doppler profile requires distinction from more curved appearance of MR

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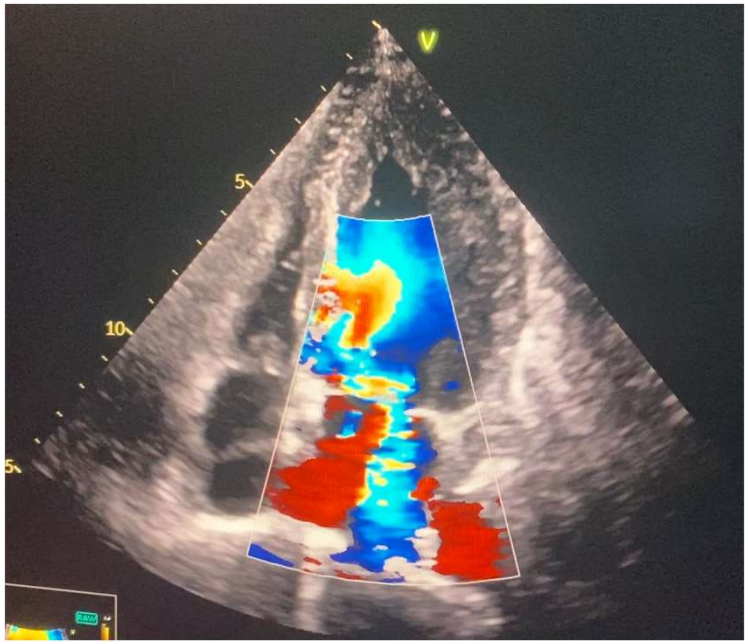
Assessment of Haemodynamics LVOTO / SAM

- 70% of patients with HCM often transient
- **Peak LVOT gradient >30 mmHg** consistent with obstruction
- **Peak LVOT gradient >50 mmHg** in conjunction with drug refractory symptoms considered the threshold for septal reduction therapies
- Provocative maneuvers (Valsalva, exercise, standing) should be performed in patients without a gradient at rest, particularly those with symptoms



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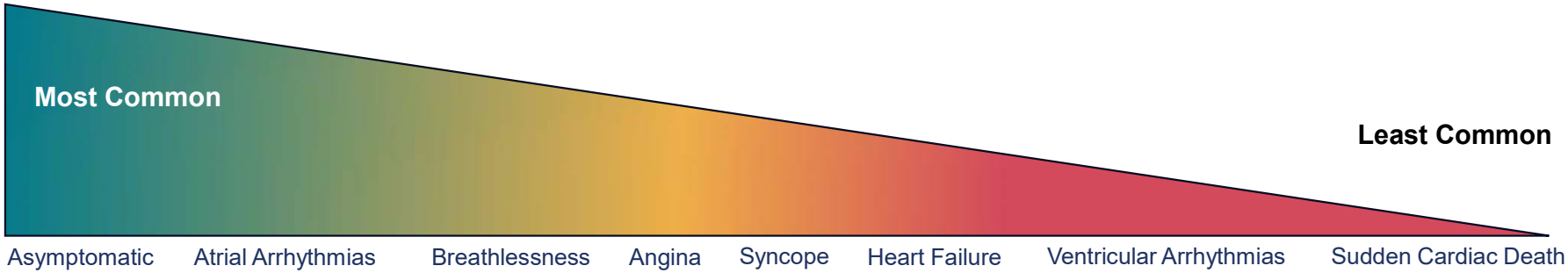
Diversity in Clinical Symptoms in Hypertrophic Cardiomyopathy



Different Mechanisms to Explain Symptoms

- Outflow tract obstruction
- Diastolic dysfunction
- Chronotropic dysfunction
- Microvascular ischaemia

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Cardiopulmonary Exercise Testing (CPEX) and pVO2

- Can provide an objective quantitative assessment of exercise status and symptom status
- Standard exercise tolerance testing limiting by baseline ECG changes
- Can be combined with echocardiography
- Diagnostic information (eg, HCM vs athlete heart) and prognostic information

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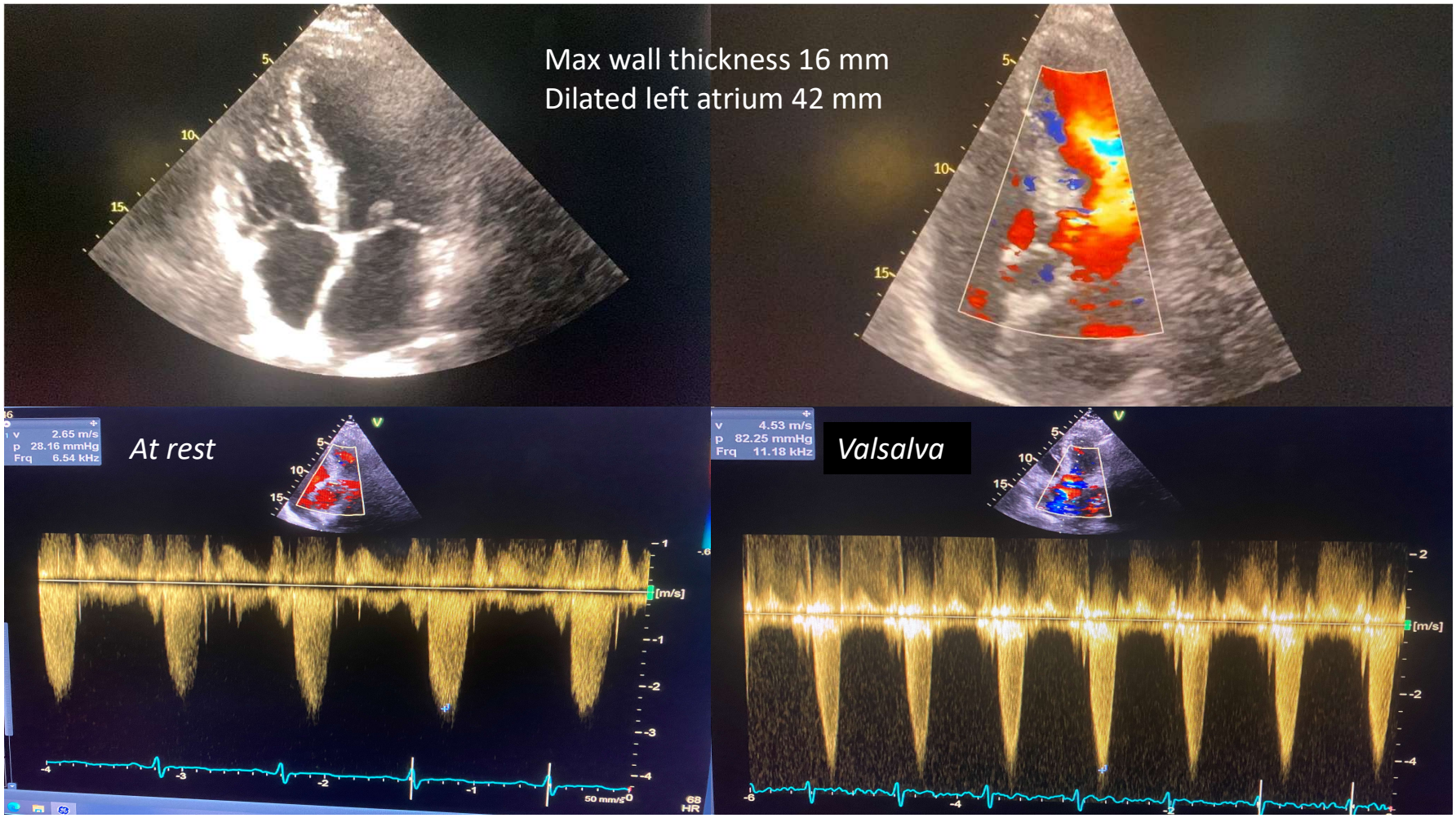
Challenges to Diagnosis

- Long asymptomatic phase
- Identification of outflow obstruction often requires provocation maneuver
- Genetic testing often not performed
- Phenocopies
- Apex often not well seen on echocardiography (standard wall thickness measurements may not apply)

Case Study

48-YEAR-OLD PATIENT

- SOBOE
- Murmur
- ECG – demonstrated LVH with subtle ST / T wave changes
- Noted to have a markedly abnormal ECG during an admission with glandular fever
- No family history of cardiomyopathy or SCD
- Normal ETT and Holter monitor


Max wall thickness 16 mm
Dilated left atrium 42 mm
At rest
v 2.65 m/s
p 28.16 mmHg
Frq 6.54 kHz
[m/s]
50 mm/s
68 HR
Valsalva
v 4.53 m/s
p 82.25 mmHg
Frq 11.18 kHz
[m/s]

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Case Study

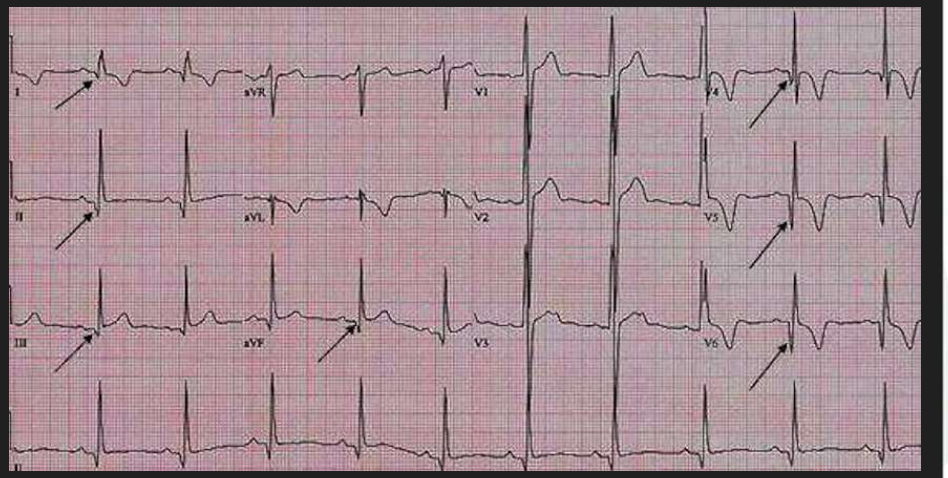
28-YEAR-OLD MAN

- Patient describes palpitations and syncope
- Previously fit and well
- Brother died suddenly playing football

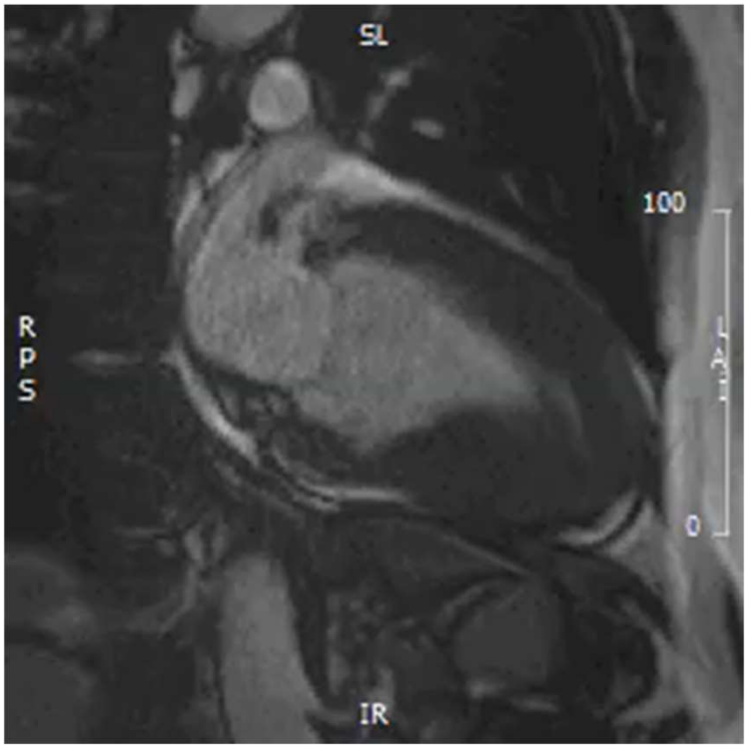
OE

- Pulse 60 regular
- HS normal
- Euvolaemic

- Echo reported as normal, difficult echo subject



Apical HCM



Apex often not well seen on echocardiography

Wall thickness cutoffs may be different compared to other regions of the LV

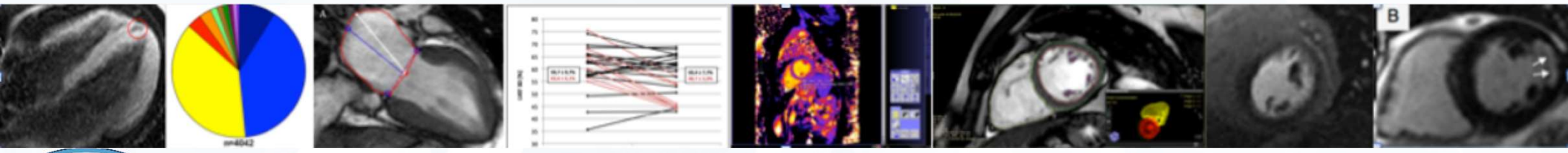
Conclusion

- Hypertrophic cardiomyopathy is common
- It is a disease of the sarcomere associated with hypercontractility and wall thickening
- It is often asymptomatic but can present with a wide range of symptoms due to different mechanisms

- As we will hear, multimodality imaging plays a key role in diagnosis, risk stratification and monitoring of therapy

Echocardiography and Beyond: Comprehensive Imaging Strategies for HCM Evaluation

Prof. Jeanette Schulz-Menger
ECRC, Charité Campus Buch, University Medicine Berlin
HELIOS-Clinics Berlin,
Department for Cardiology and Nephrology
jeanette.schulz-menger@charite.de
www.cmr-berlin.org



CHARITÉ UNIVERSITÄTSMEDIZIN BERLIN
MDC MAX-DELBRÜCK-CENTRUM FÜR MOLEKULARE MEDIZIN IN DER HELMHOLTZ-GEMEINSCHAFT
BIH Berlin Institute of Health Charité & MDC
DZHK DEUTSCHES ZENTRUM FÜR HERZ-KREISLAUF-FORSCHUNG E.V.
Helios

Disclosure

- Nothing relevant to this topic

ESC European Society of Cardiology
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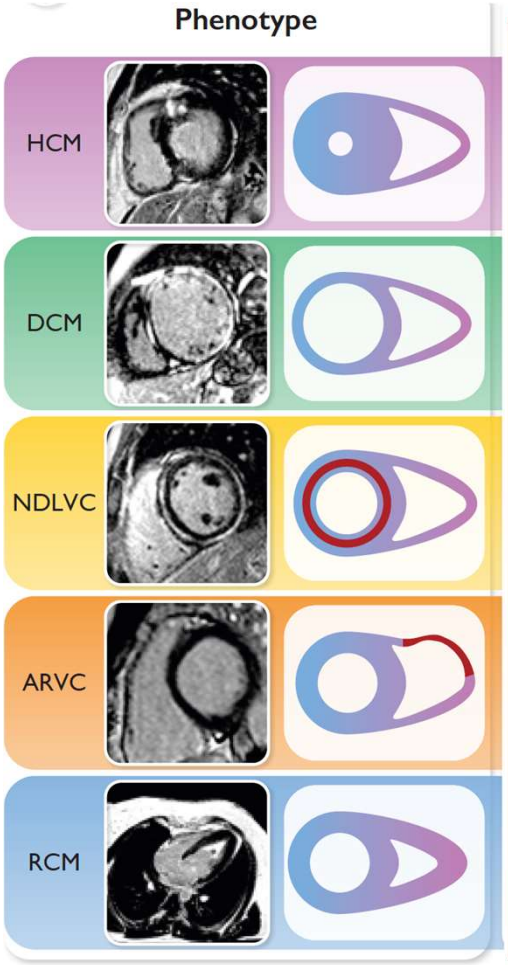
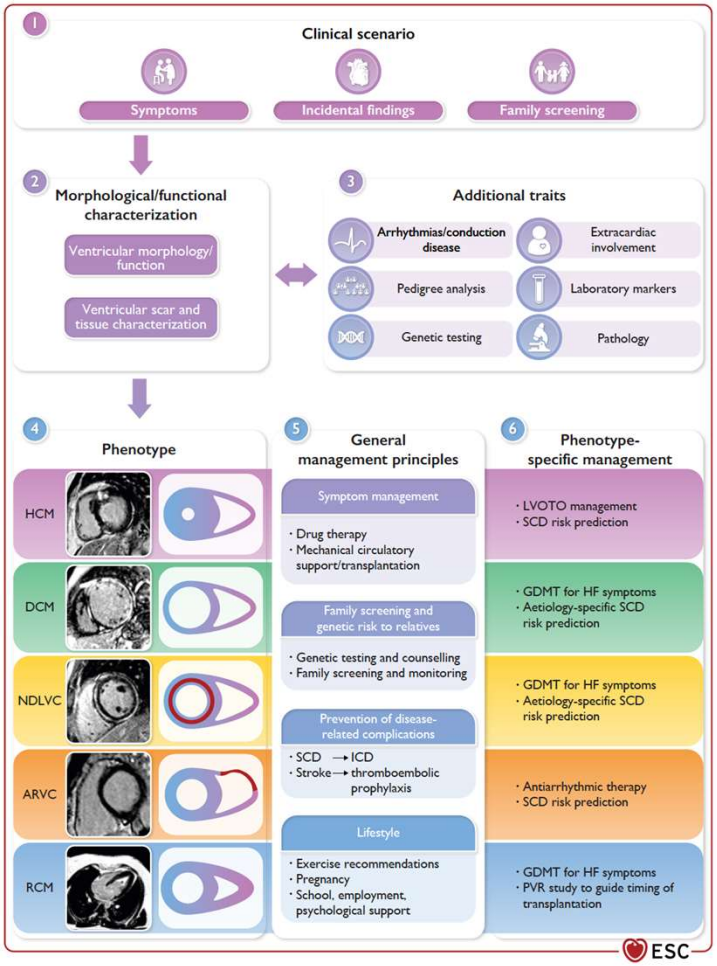
ESC GUIDELINES

2023 ESC Guidelines for the management of cardiomyopathies

Developed by the task force on the management of cardiomyopathies of the European Society of Cardiology (ESC)

Authors/Task Force Members: Elena Arbelo [*†], (Chairperson) (Spain), Alexandros Protonotarios [‡], (Task Force Co-ordinator) (United Kingdom), Juan R. Gimeno [‡], (Task Force Co-ordinator) (Spain), Eloisa Arbustini (Italy), Roberto Barriales-Villa (Spain), Cristina Basso (Italy), Connie R. Bezzina (Netherlands), Elena Biagini (Italy), Nico A. Blom[1] (Netherlands), Rudolf A. de Boer (Netherlands), Tim De Winter (Belgium), Perry M. Elliott (United Kingdom), Marcus Flather (United Kingdom), Pablo Garcia-Pavia (Spain), Kristina H. Haugaa (Sweden), Jodie Ingles (Australia), Ruxandra Oana Jurcut (Romania), Sabine Klaassen (Germany), Giuseppe Limongelli (Italy), Bart Loeys[2] (Belgium), Jens Mogensen (Denmark), Iacopo Olivotto (Italy), Antonis Pantazis (United Kingdom), Sanjay Sharma (United Kingdom), J. Peter Van Tintelen (Netherlands), James S. Ware (United Kingdom), Juan Pablo Kaski [*†], (Chairperson) (United Kingdom), and ESC Scientific Document Group





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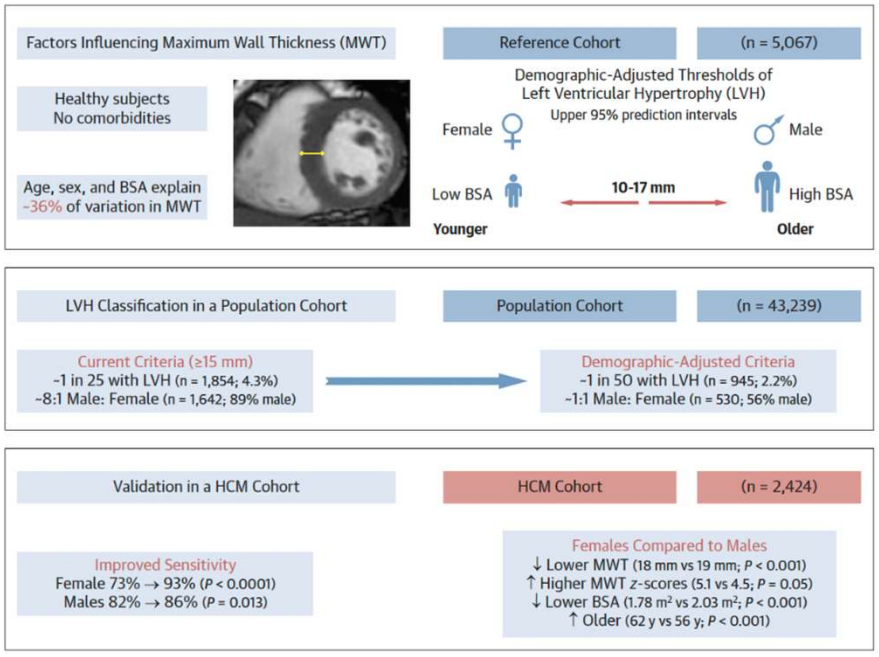
ORIGINAL RESEARCH

Demographic-Based Personalized Left Ventricular Hypertrophy Thresholds for Hypertrophic Cardiomyopathy Diagnosis

Hunain Shiwani, MD,[a,b] Rhodri H. Davies, MD, PhD,[a,b] Constantin-Cristian Topriceanu, MD,[b] Raffaello Ditaranto, MD, PhD,[a,b,c] Anjali Owens, MD,[d] Betty Raman, MD, DPhil,[e] João Augusto, MD,[f] Rebecca K. Hughes, MD, PhD,[a,b] Camilla Torlasco, MD, PhD,[g] Ben Dowsing, MD,[a,b] Jessica Artico, MD,[a,b] George Joy, MD, PhD,[a,b] Inês Miranda, MD,[f] Walter Witschey, PhD,[h] Jose F. Rodriguez-Palomares, MD, PhD,[i] Clara Badia-Molins, MD,[i] Lia Crotti, MD, PhD,[g,j] Mario Cortina-Borja, PhD,[k] Michael L. Chuang, MD,[l,m] Raymond Y. Kwong, MD, MPH,[n] Christopher M. Kramer, MD,[o] Warren Manning, MD,[m,p] Carolyn Y. Ho, MD,[n] Peter Kellman, PhD,[q] Alun D. Hughes, MD, PhD,[b] Elena Biagini, MD, PhD,[c] Saidi Mohiddin, MD,[a,r] Luis Lopes, MD, PhD,[a,b] Harold Litt, MD, PhD,[h] Victor A. Ferrari, MD,[d] Gabriella Captur, MD, PhD,[b] James C. Moon, MD,[a,b] the PRECISION-HCM Collaborative

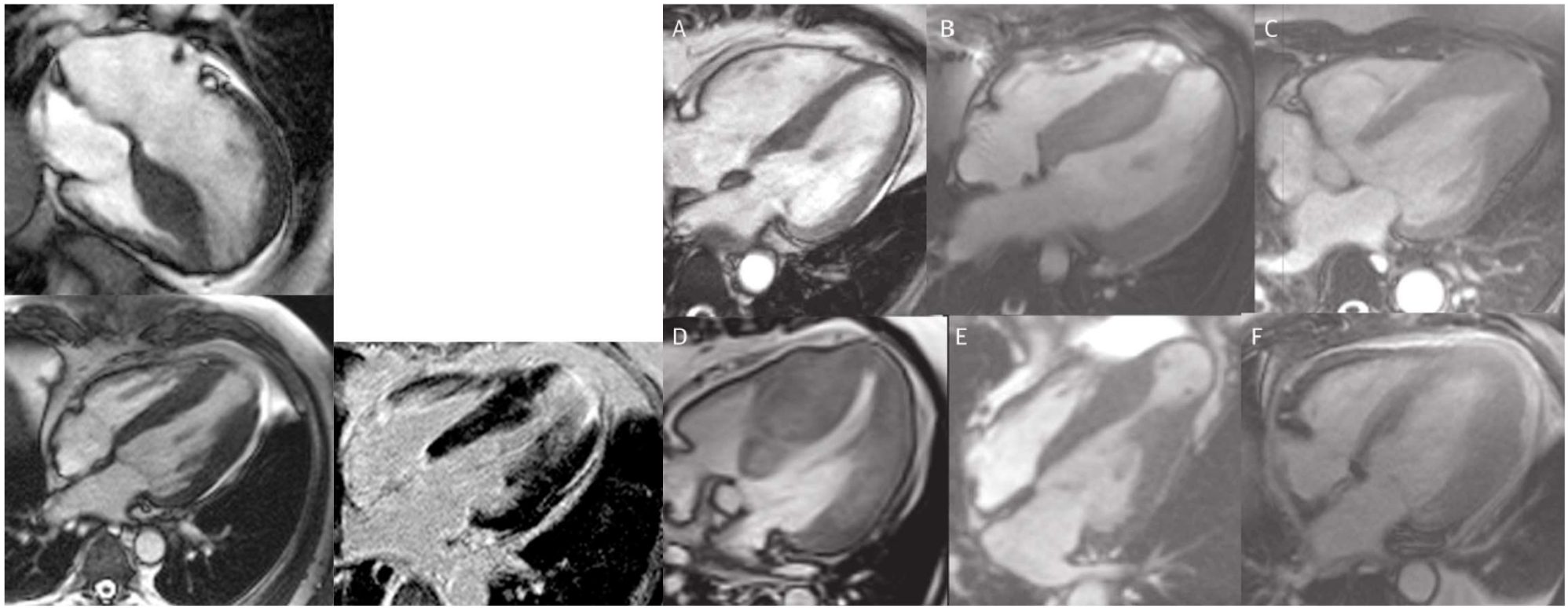
CENTRAL ILLUSTRATION Demographic-Adjusted Left Ventricular Hypertrophy Thresholds for Hypertrophic Cardiomyopathy Diagnosis



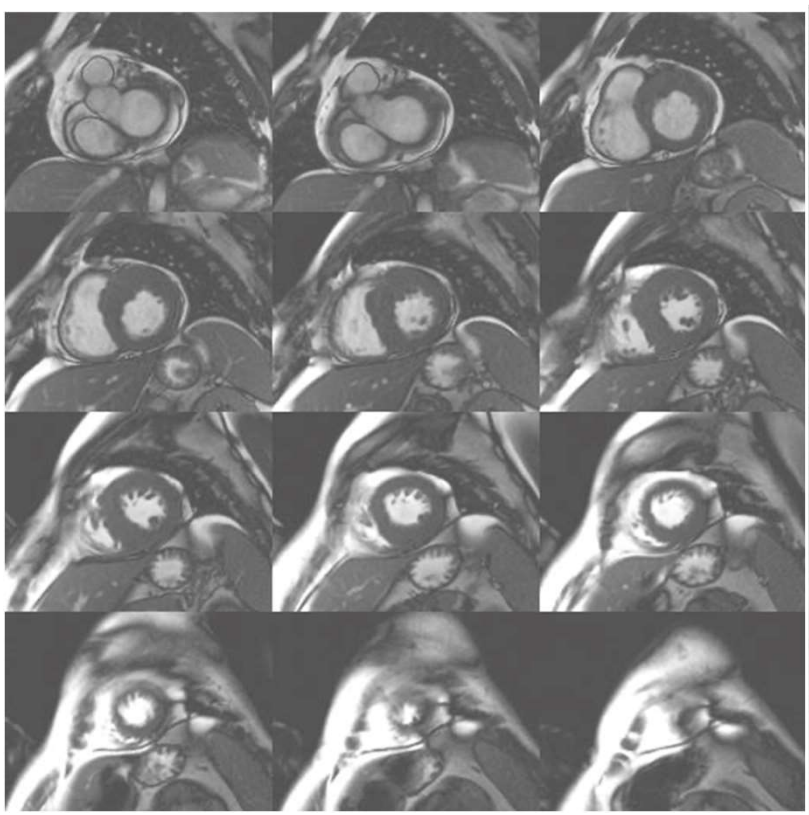
Shiwani H, et al. JACC. 2025;85(7):685-695.

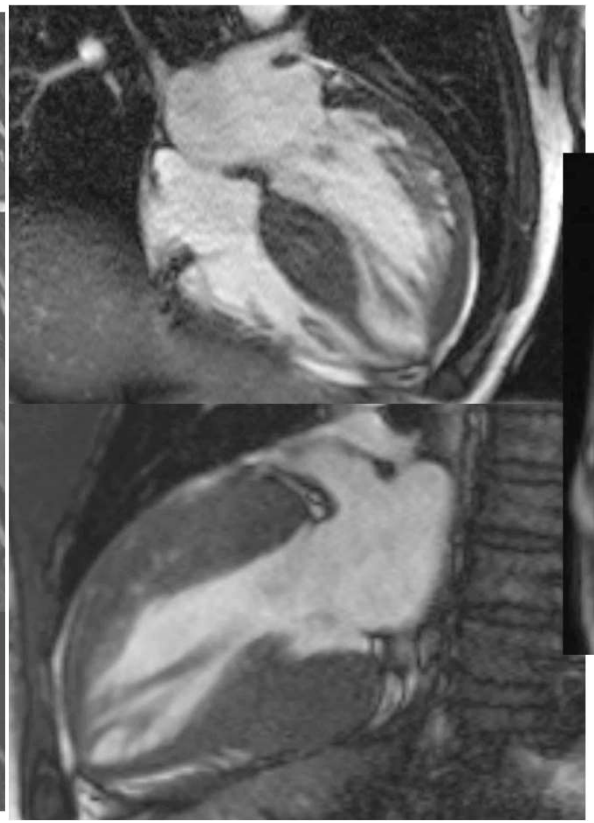
In healthy subjects, age, sex, and BSA collectively explain ~36% of the variation in MWT. Demographic-adjusted LVH thresholds show considerable range, varying with sex, BSA, and age. In a population cohort, demographic-adjusted criteria reduce LVH ascertainment, balance the male-to-female ratio, and attenuate demographic skews, compared with the current fixed threshold. Validation in an HCM cohort demonstrated improved diagnostic sensitivity for HCM, especially for female individuals. BSA = body surface area; HCM = hypertrophic cardiomyopathy.

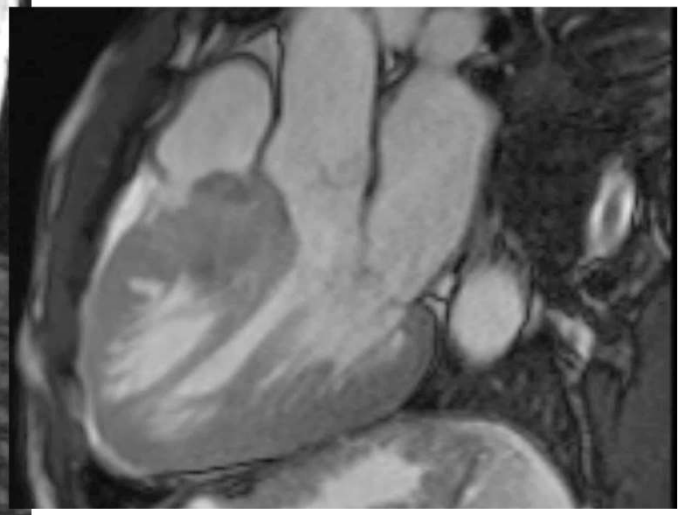
Phenotypes HCM



Neubauer S, et al. *J Am Coll Cardiol*. 2019;74(19):2333-2345.







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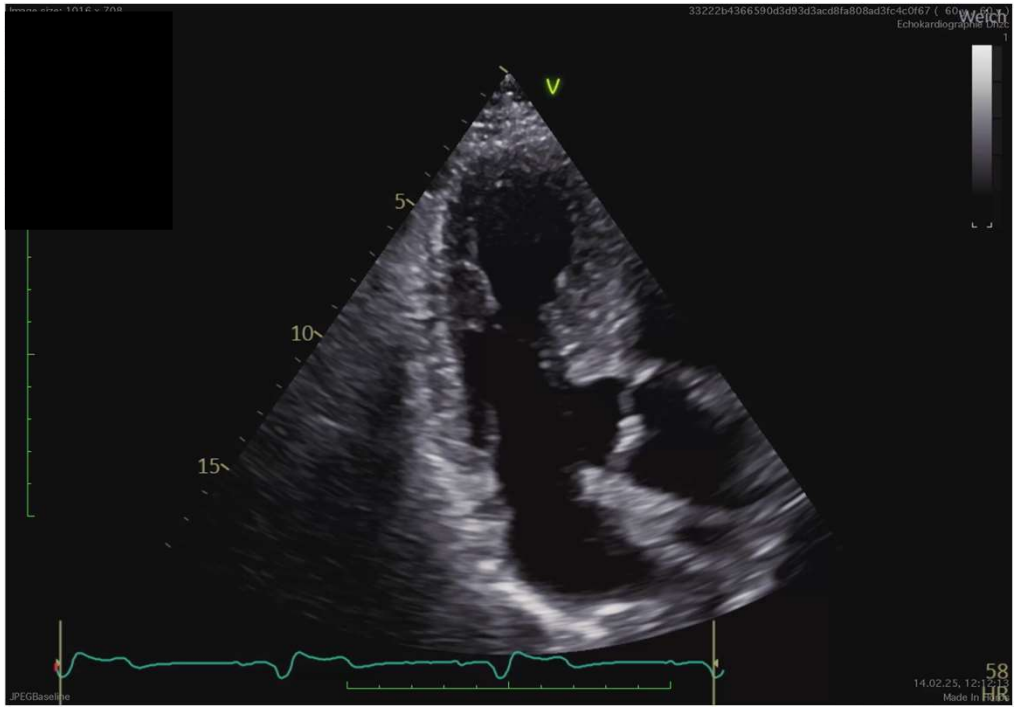
Recommendation Table 16 — Recommendation for evaluation of left ventricular outflow tract obstruction

Recommendations	Class[a]	Level[b]
In all patients with HCM, at initial evaluation, transthoracic 2D and Doppler echocardiography are recommended, at rest and during Valsalva manoeuvre in the sitting and semi-supine positions—and then on standing if no gradient is provoked—to detect LVOTO.[84,86,365,525,584,587,589–594]	I	B
In symptomatic patients with HCM and inconclusive non-invasive cardiac imaging, left and right heart catheterization may be considered to assess the severity of LVOTO and to measure LV filling pressures.[603]	IIb	C
Transoesophageal echocardiography should be considered in patients with HCM and LVOTO if the mechanism of obstruction is unclear or when assessing the mitral valve apparatus before a septal reduction procedure, or when severe mitral regurgitation caused by intrinsic valve abnormalities is suspected.[599–602]	IIa	C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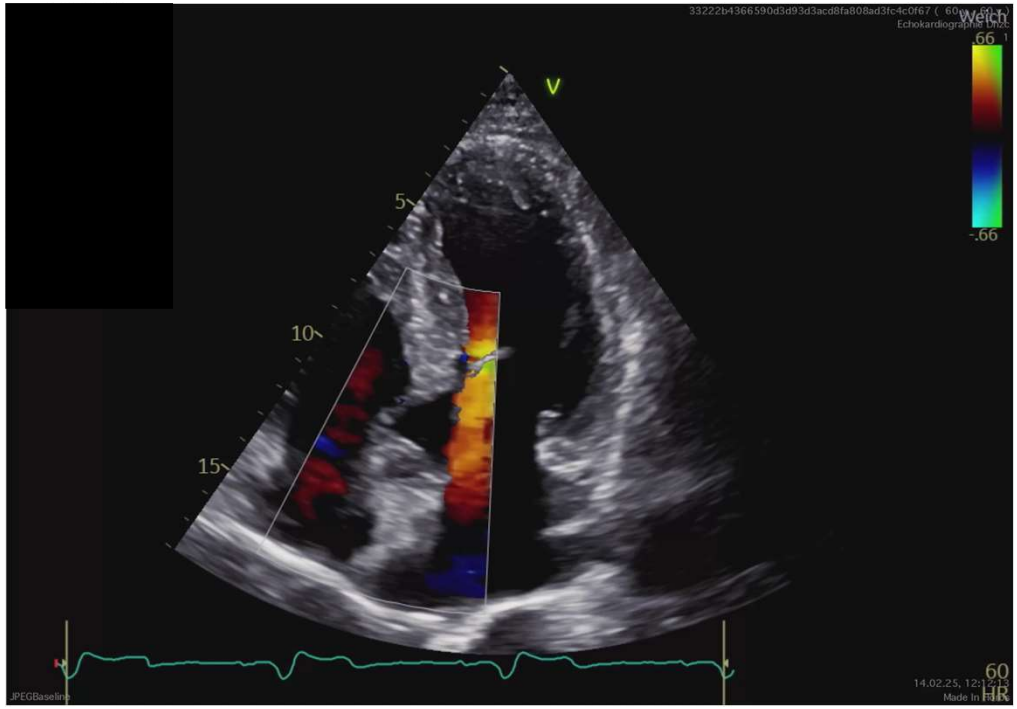
JPEGBaseline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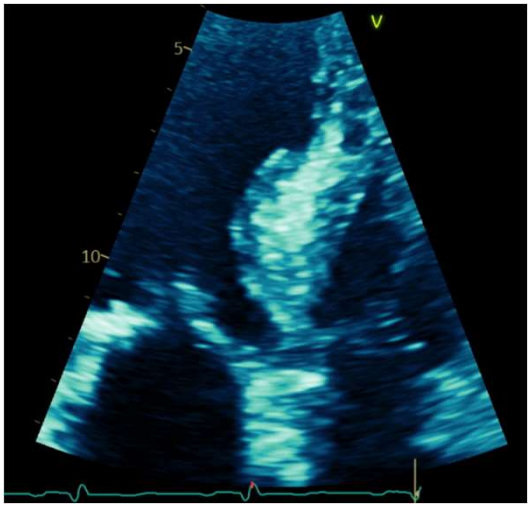

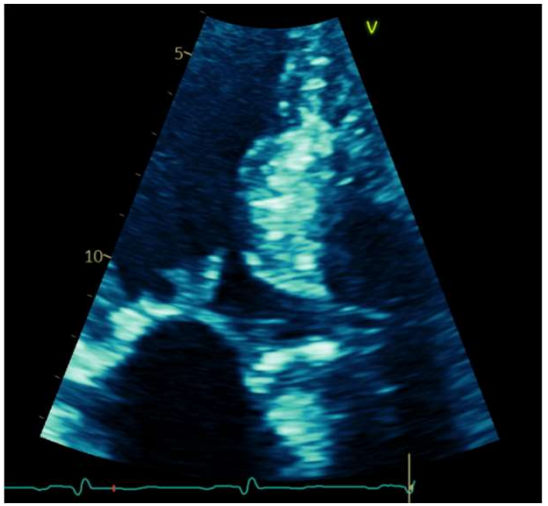

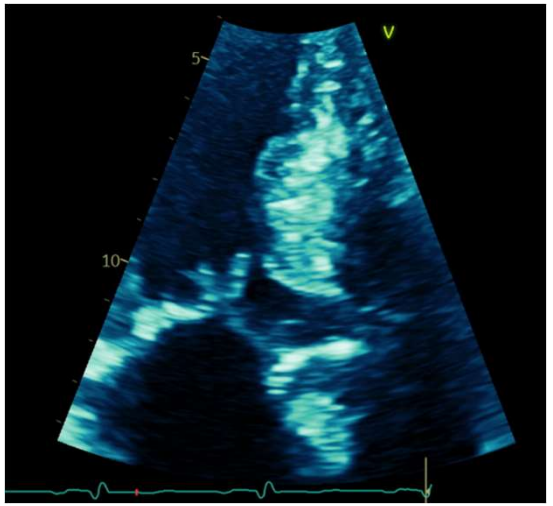

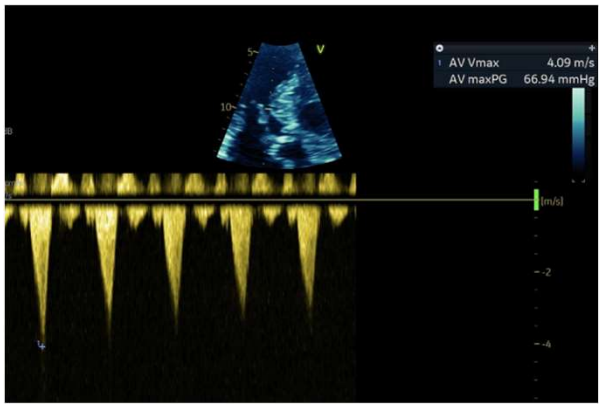

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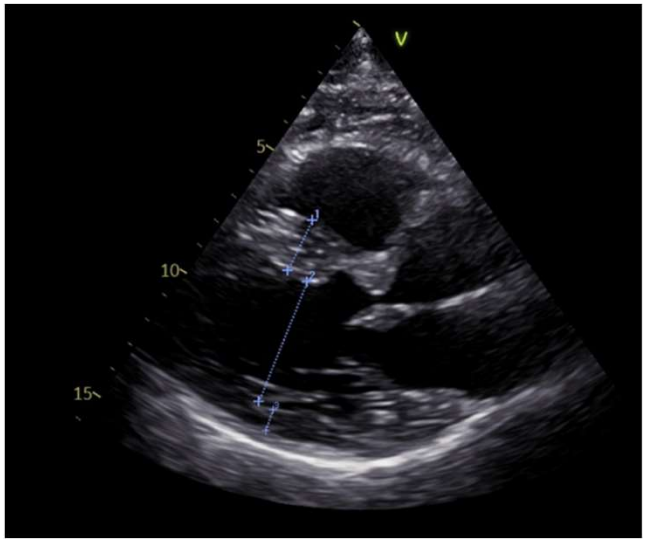
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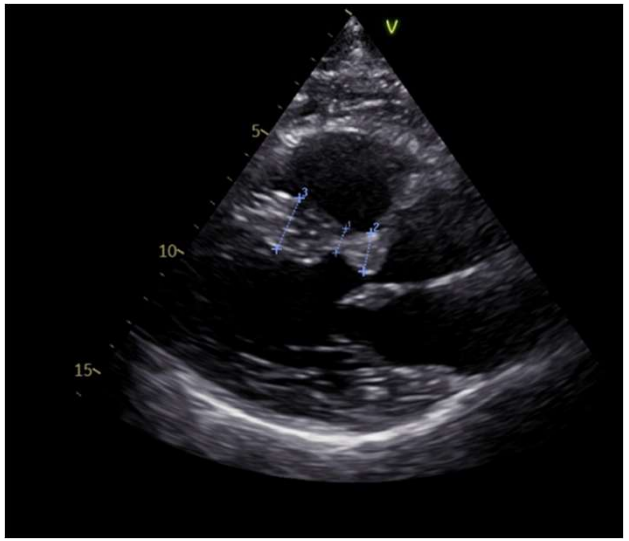
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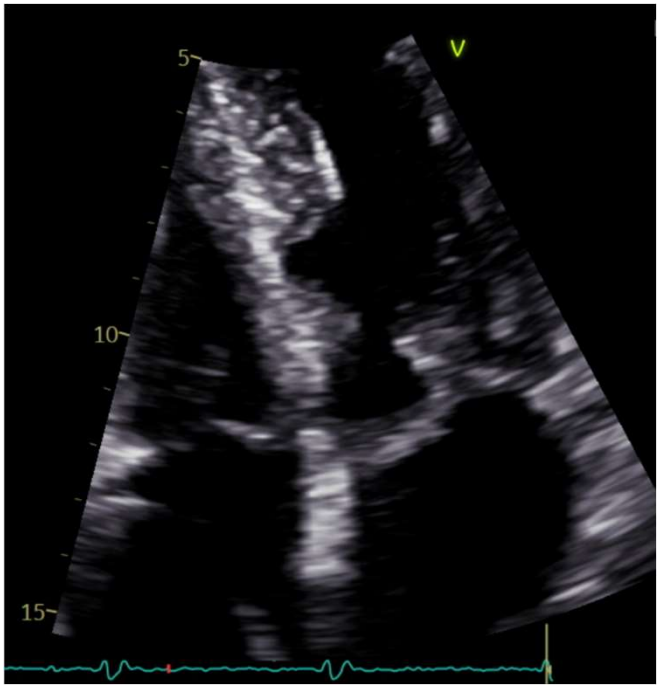
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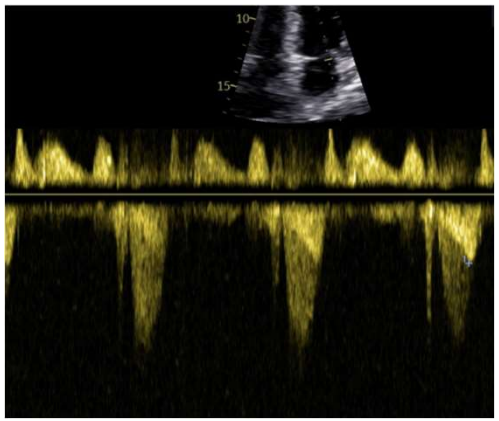
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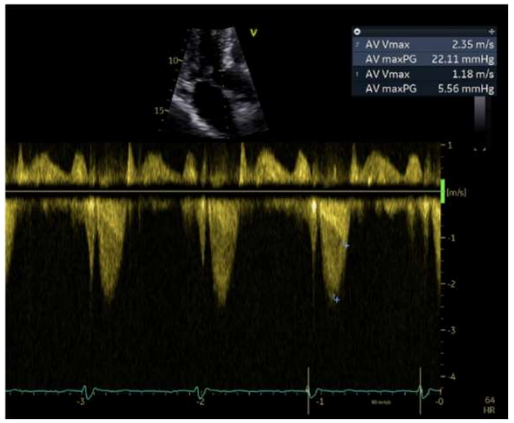
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AV maxPG 22.11 mmHg
AV Vmax 1.18 m/s
AV maxPG 5.56 mmHg
[m/s]

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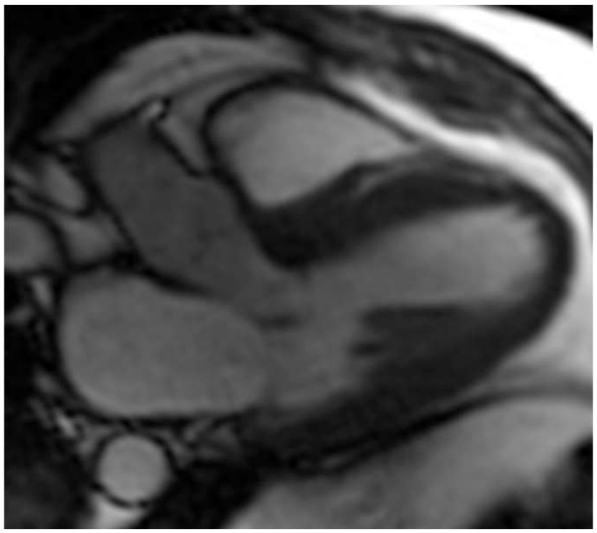
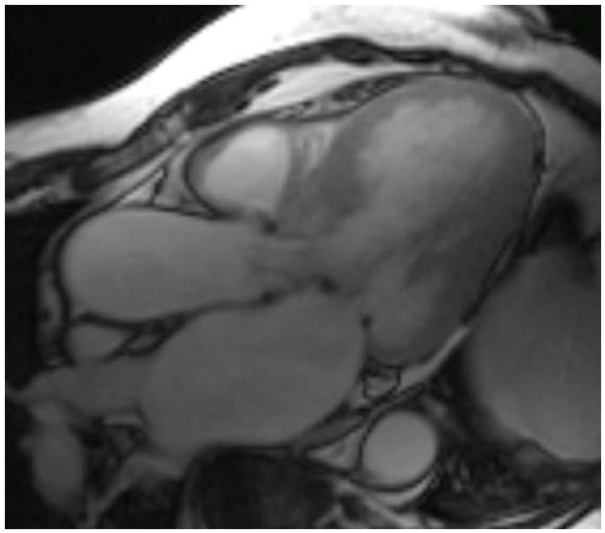
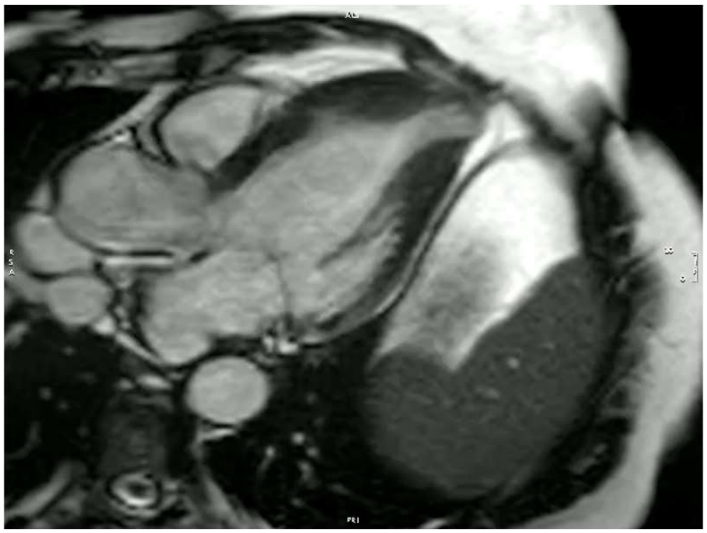
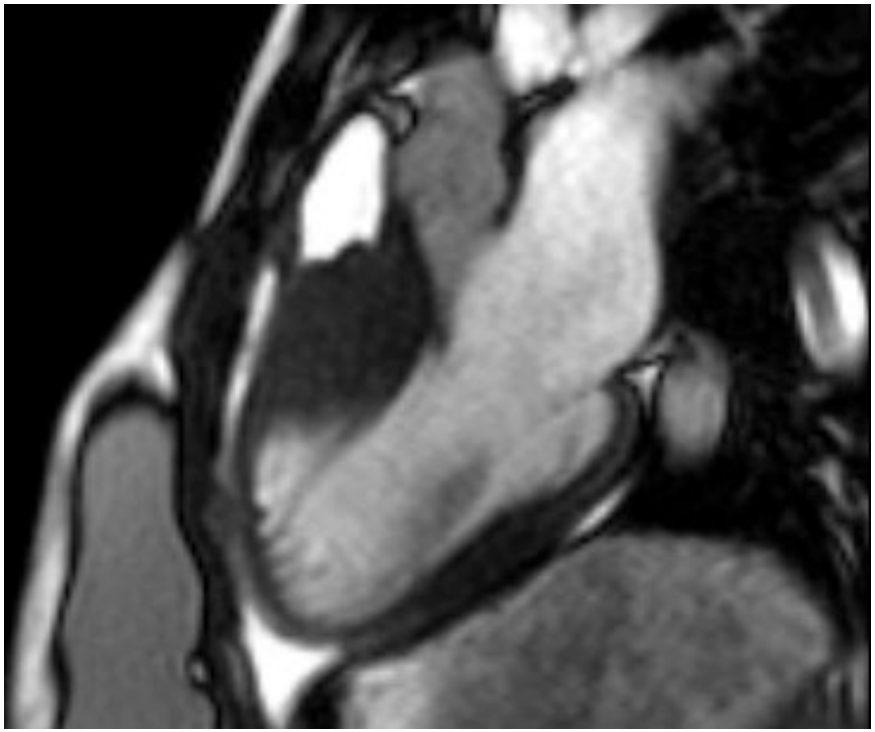
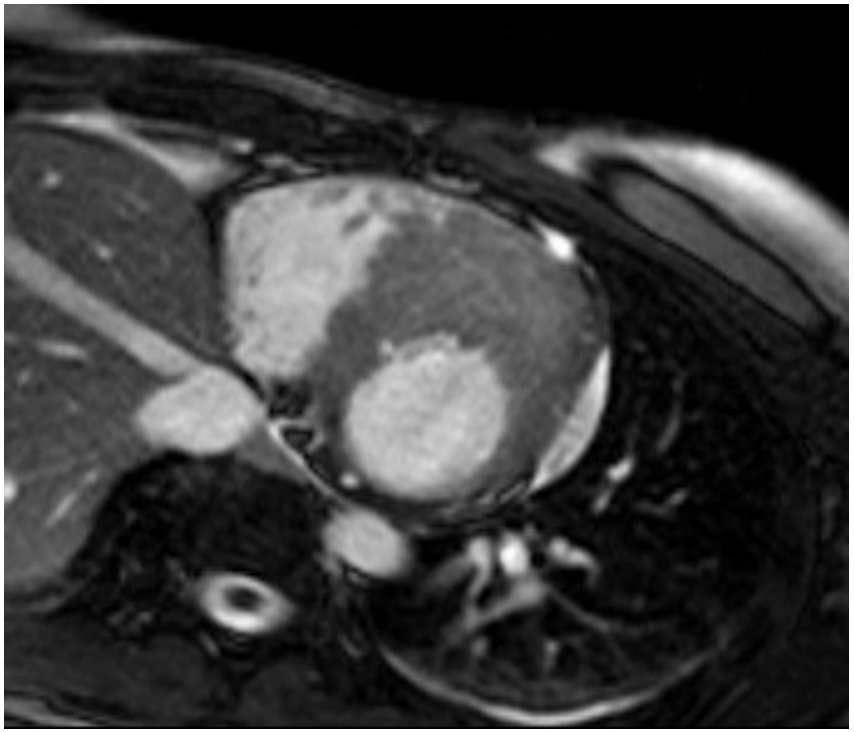
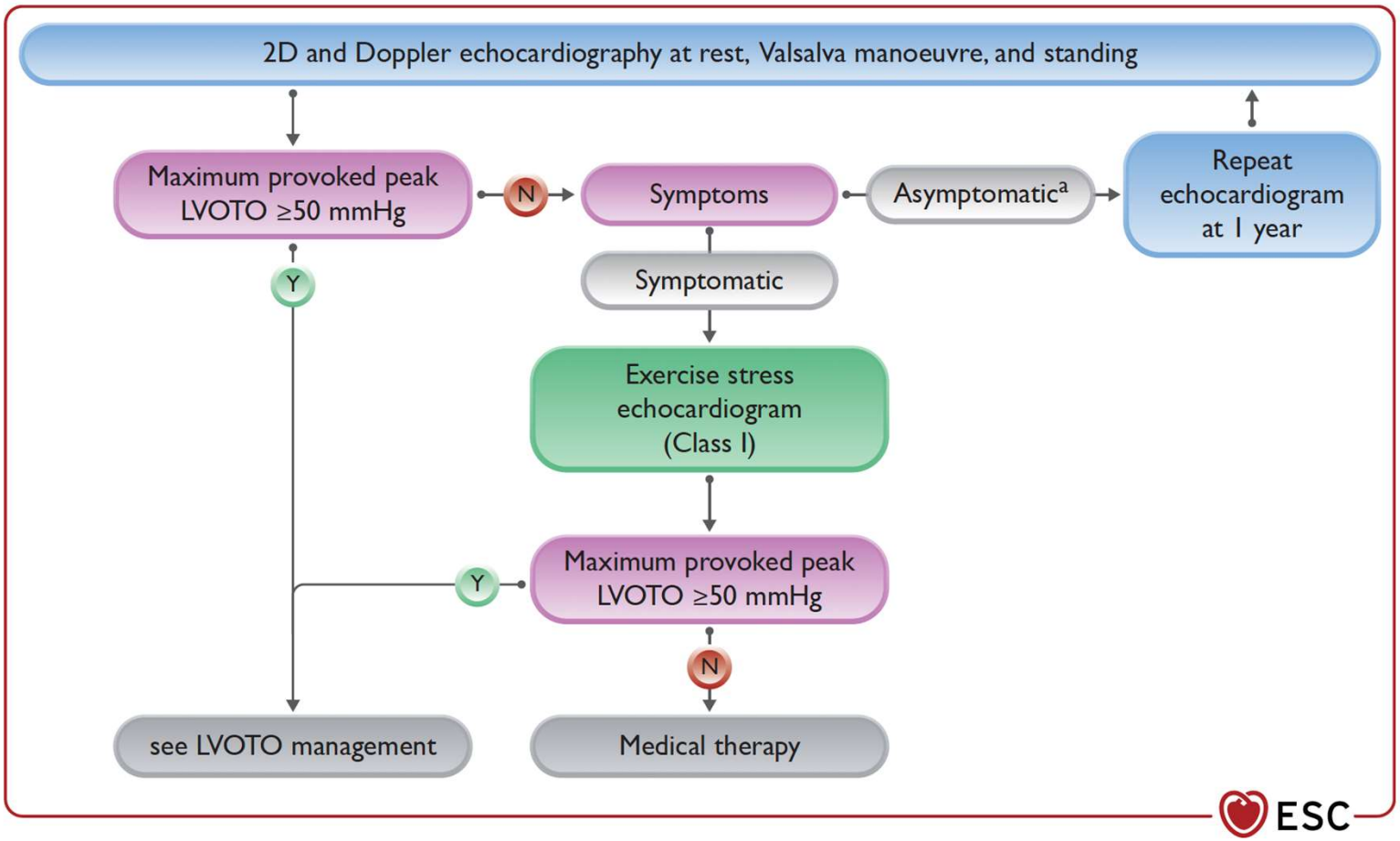
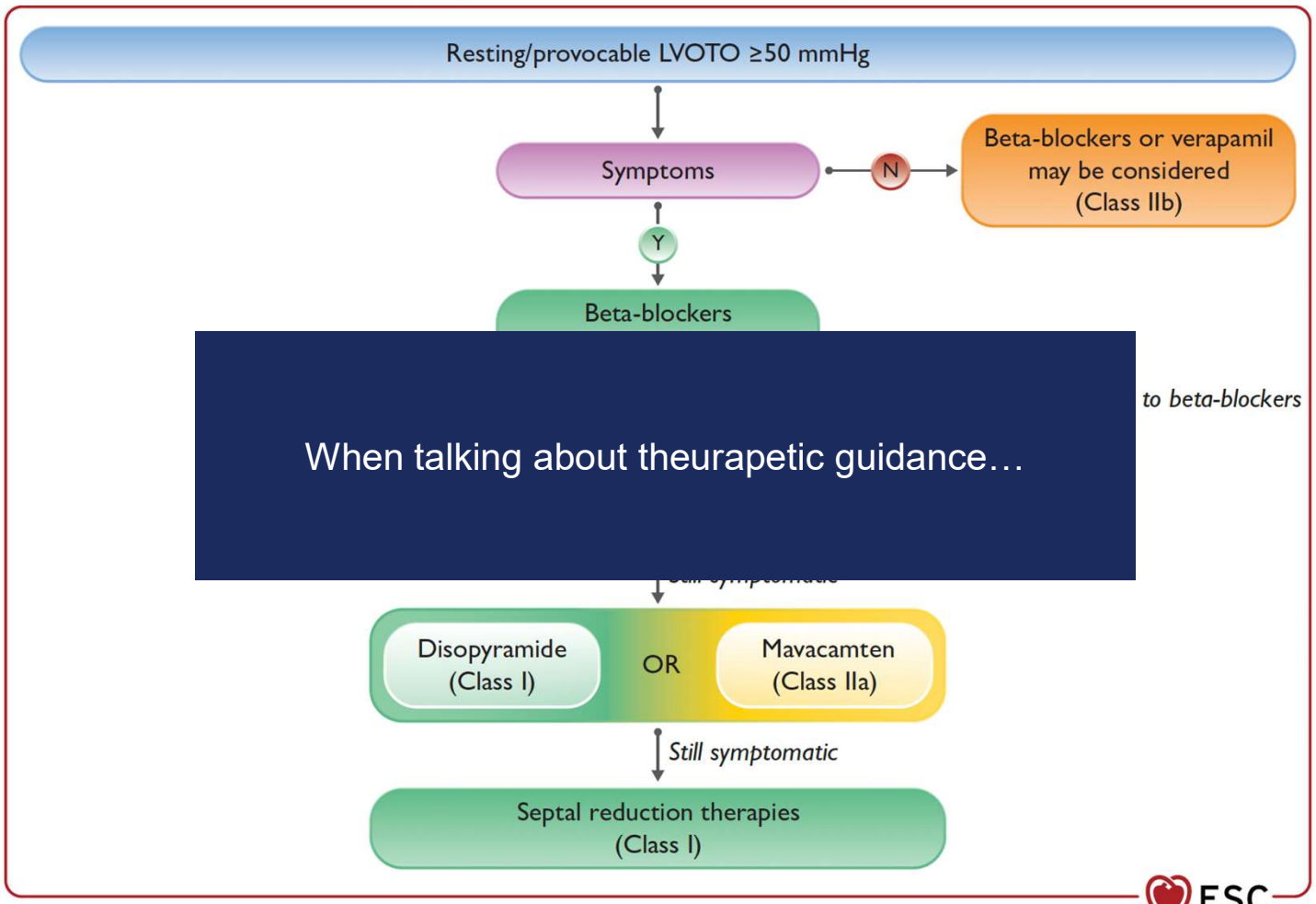









2D and Doppler echocardiography at rest, Valsalva manoeuvre, and standing
Maximum provoked peak LVOTO ≥50 mmHg
N
Symptoms
Asymptomatic[a]
Repeat echocardiogram at 1 year
Y
Symptomatic
Exercise stress echocardiogram (Class I)
Y
Maximum provoked peak LVOTO ≥50 mmHg
N
see LVOTO management
Medical therapy
ESC


Resting/provocable LVOTO ≥50 mmHg
Symptoms
N
Beta-blockers or verapamil may be considered (Class IIb)
Y
Beta-blockers
to beta-blockers
When talking about theurapetic guidance…
Disopyramide (Class I)
OR
Mavacamten (Class IIa)
Still symptomatic
Septal reduction therapies (Class I)
ESC

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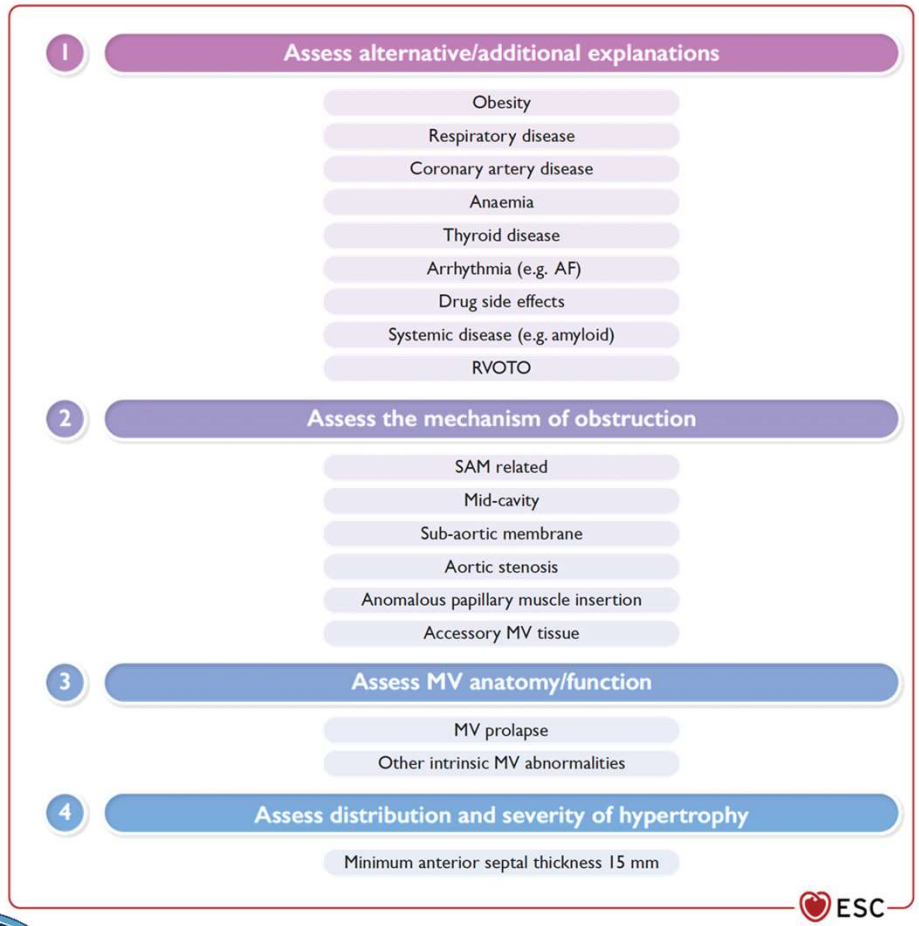
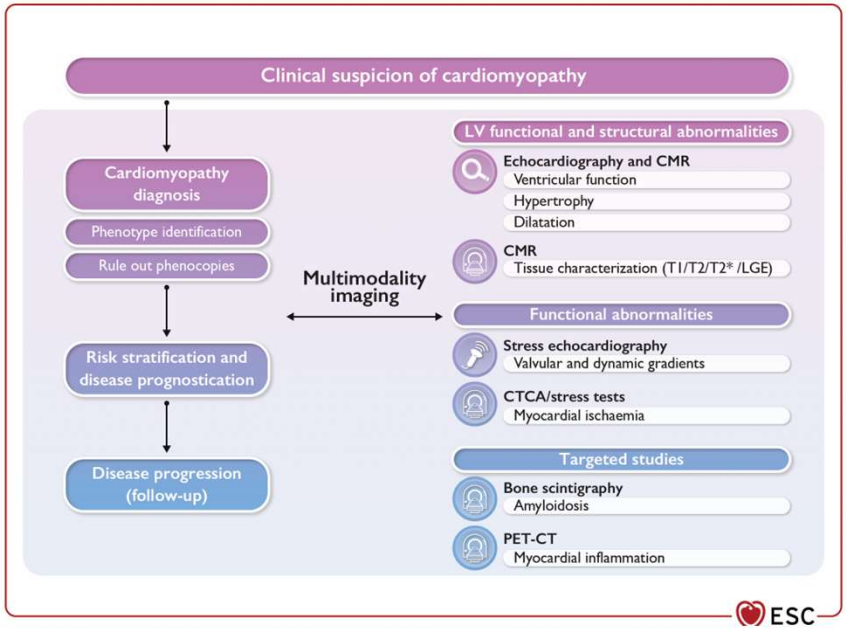

1
Assess alternative/additional explanations
Obesity
Respiratory disease
Coronary artery disease
Anaemia
Thyroid disease
Arrhythmia (e.g. AF)
Drug side effects
Systemic disease (e.g. amyloid)
RVOTO
2
Assess the mechanism of obstruction
SAM related
Mid-cavity
Sub-aortic membrane
Aortic stenosis
Anomalous papillary muscle insertion
Accessory MV tissue
3
Assess MV anatomy/function
MV prolapse
Other intrinsic MV abnormalities
4
Assess distribution and severity of hypertrophy
Minimum anterior septal thickness 15 mm
ESC


Clinical suspicion of cardiomyopathy
Cardiomyopathy diagnosis
Phenotype identification
Rule out phenocopies
Risk stratification and disease prognostication
Disease progression (follow-up)
Multimodality imaging
LV functional and structural abnormalities
Echocardiography and CMR
Ventricular function
Hypertrophy
Dilatation
CMR
Tissue characterization (T1/T2/T2* /LGE)
Functional abnormalities
Stress echocardiography
Valvular and dynamic gradients
CTCA/stress tests
Myocardial ischaemia
Targeted studies
Bone scintigraphy
Amyloidosis
PET-CT
Myocardial inflammation
ESC

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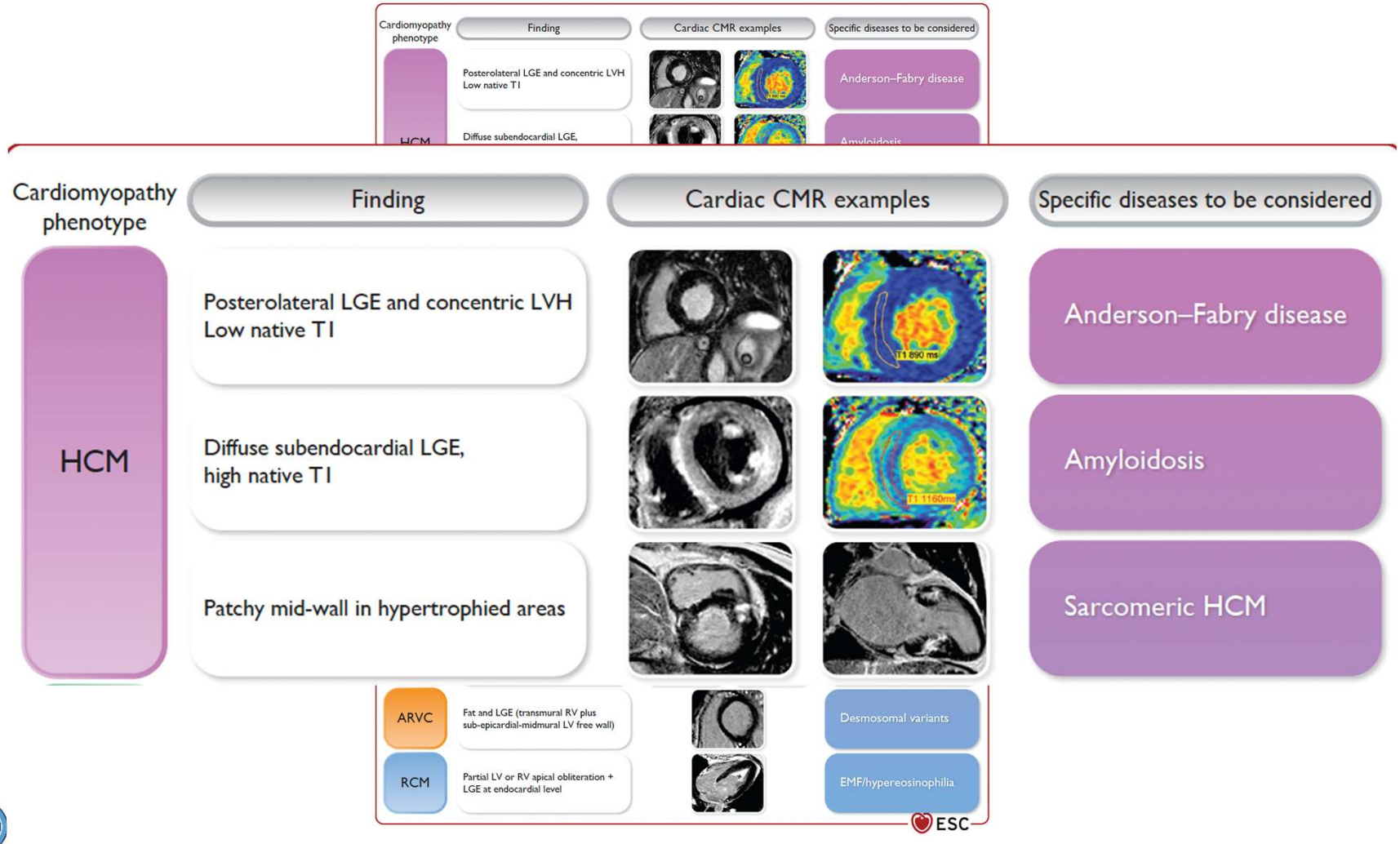
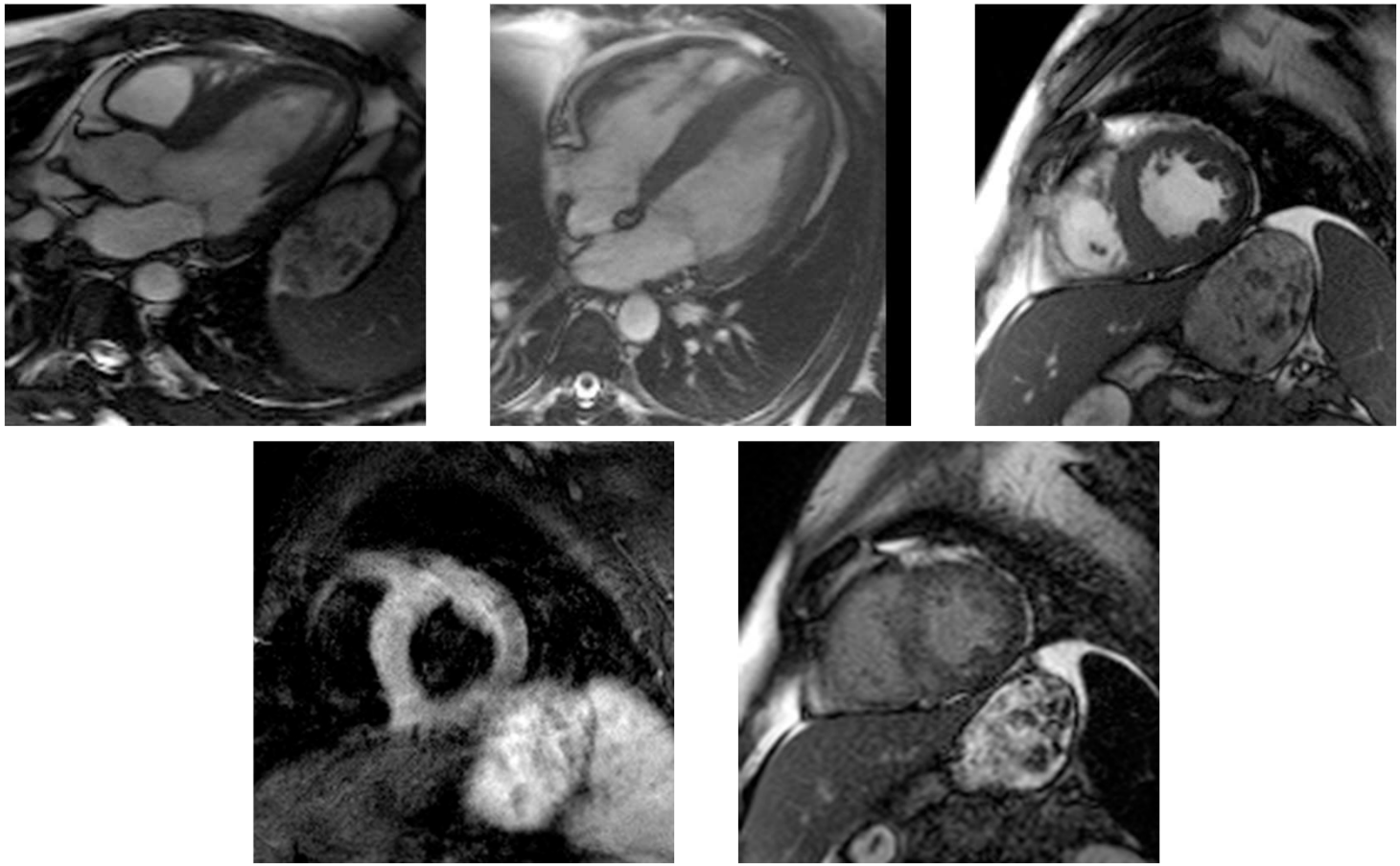

Cardiomyopathy phenotype
Finding
Cardiac CMR examples
Specific diseases to be considered
HCM
Posterolateral LGE and concentric LVH
Low native T1
Anderson–Fabry disease
Diffuse subendocardial LGE,
high native T1
Amyloidosis
Patchy mid-wall in hypertrophied areas
Sarcomeric HCM
T1 890 ms
T1 1160ms
ARVC
Fat and LGE (transmural RV plus sub-epicardial-midmural LV free wall)
Desmosomal variants
RCM
Partial LV or RV apical obliteration + LGE at endocardial level
EMF/hypereosinophilia
ESC

Recommendation Table 5 — Recommendations for cardiac magnetic resonance indication in patients with cardiomyopathy

Recommendations	Class[a]	Level[b]
Contrast-enhanced CMR is recommended in patients with cardiomyopathy at initial evaluation.[10,90,116,119–143]	I	B

Common Pitfall



PACE-CME

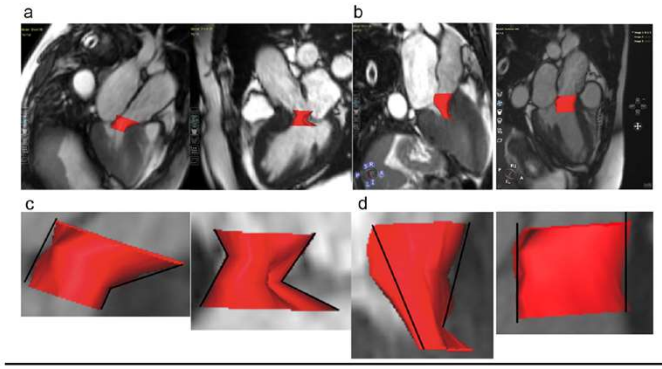
LVOT Obstruction: Fully Understood?

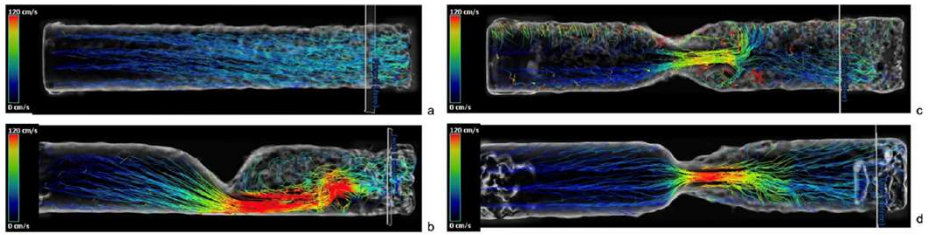
The International Journal of Cardiovascular Imaging
https://doi.org/10.1007/s10554-024-03242-4

ORIGINAL PAPER

Hypertrophic obstructive cardiomyopathy-left ventricular outflow tract shapes and their hemodynamic influences applying CMR

T. Mayr[1,2] · L. Riazy[1,2,3] · R. F. Trauzeddel[1,2,6] · J. P. Bassenge[1,4] · S. Wiesemann[1,2,3] · E. Blaszczyk[1,2,3] · M. Prothmann[5] · T. Hadler[1,2] · S. Schmitter[4] · Jeanette Schulz-Menger[1,2,5]





Alexander Schulz[1], Tess E. Wallace[1,2], Kelvin Chow[3], Xiaoming Bi[4], Amine Amyar[1], Jennifer Rodriguez[1], Fahime Ghanbari[1], Martin S. Maron[5], Ethan J. Rowin[5], Peter Kellmann[6] Warren J. Manning[1,7], Reza Nezafat[1]

Quantitative Myocardial Blood Flow and Perfusion Reserve with Exercise CMR **JCMR 2025 – in press, currently proofs**

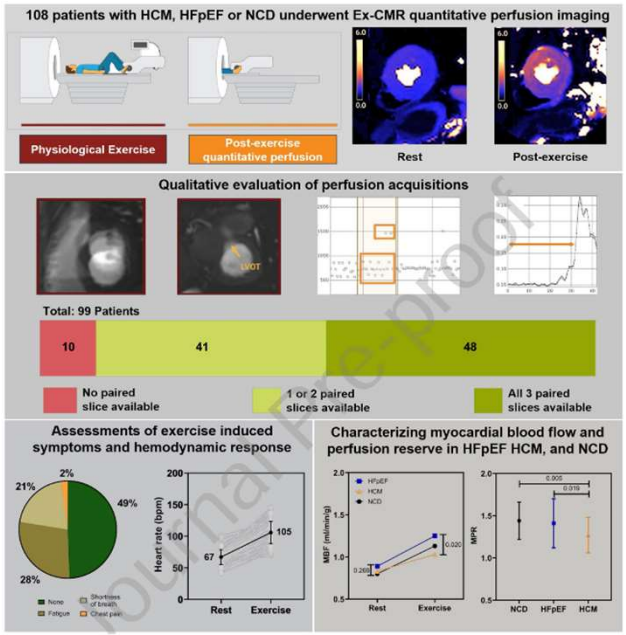


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Table 17 **Imaging evaluation in hypertrophic cardiomyopathy**

Item to assess	Primary imaging modality	Comments
LV wall thickness	ECHO/CMR	• All LV segments from base to apex examined in end-diastole, preferably in the 2D short-axis view, ensuring that the wall thickness is recorded at mitral, mid-LV, and apical levels. • CMR is superior in the detection of LV apical and anterolateral hypertrophy, aneurysms,[580] and thrombi,[581] and is more sensitive in the detection of subtle markers of disease in patients with sarcomeric protein gene variants (e.g. myocardial crypts, papillary muscle abnormalities).[159,582,583]
Systolic function (global and regional)	ECHO/CMR	• Ejection fraction is a suboptimal measure of LV systolic performance when hypertrophy is present. • Doppler myocardial velocities and deformation parameters (strain and strain rate) are typically reduced at the site of hypertrophy despite a normal EF and may be abnormal before the development of increased wall thickness in genetically affected patients.
Diastolic function	ECHO	• Routine examination should include mitral inflow assessment, tissue Doppler imaging, pulmonary vein flow velocities, pulmonary artery systolic pressure, and LA size/volume.
Mitral valve	ECHO	• Assess presence and degree of SAM and mitral regurgitation. The presence of a central- or anteriorly directed jet of mitral regurgitation should raise suspicion of an intrinsic/primary mitral valve abnormality and prompt further assessment.
LVOT	ECHO	• See *Figure 12*.
LA dimensions	ECHO/CMR	• Provides important prognostic information.[365,525,584] • Most common mechanisms of LA enlargement are SAM-related mitral regurgitation and elevated LV filling pressures.
Myocardial fibrosis/LGE	CMR	• The distribution and severity of interstitial expansion can suggest specific diagnoses. Anderson–Fabry disease is characterized by a reduction in non-contrast T1 signal and the presence of posterolateral LGE.[134,155] In cardiac amyloidosis, there is often global, subendocardial or segmental LGE and a highly specific pattern of myocardial and blood-pool gadolinium kinetics caused by similar myocardial and blood T1 signals.[585,586]

From Therapy to Outcomes: Imaging-Guided Assessment in Obstructive HCM

Tomaž Podlesnikar

UKC MARIBOR Univerzitetni klinični center Maribor

univerzitetni klinični center ljubljana
University Medical Centre Ljubljana

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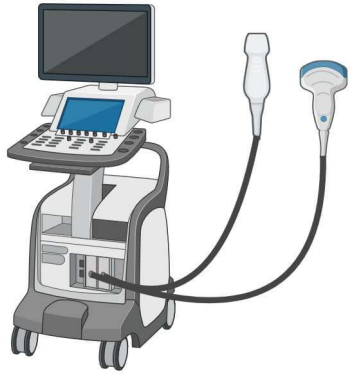
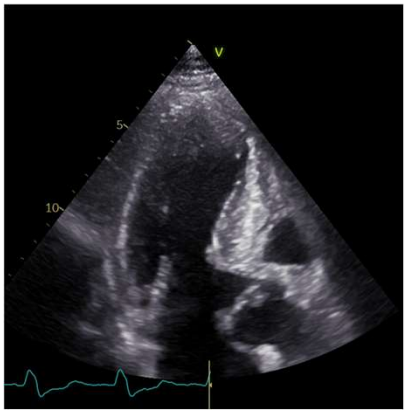
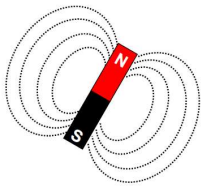
Disclosure

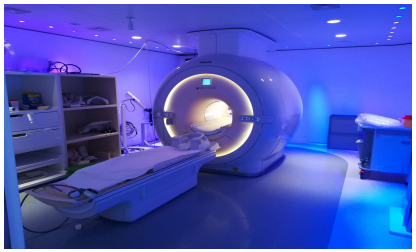
- Dr. Podlesnikar has no relevant relationships to disclose.

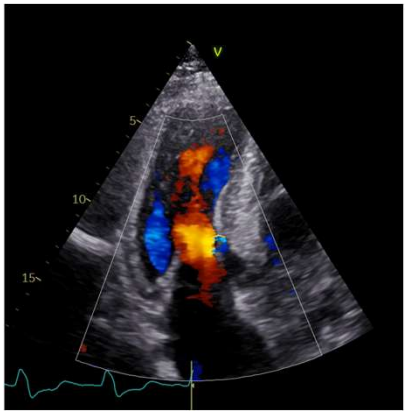
Imaging to Assess LVOTO

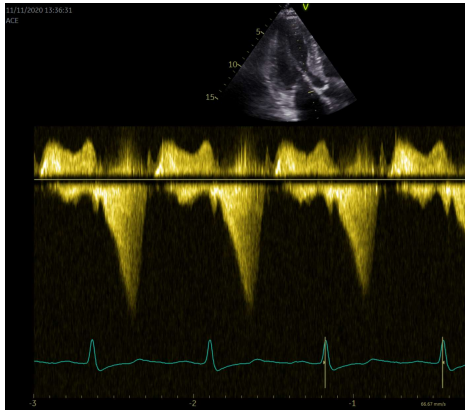


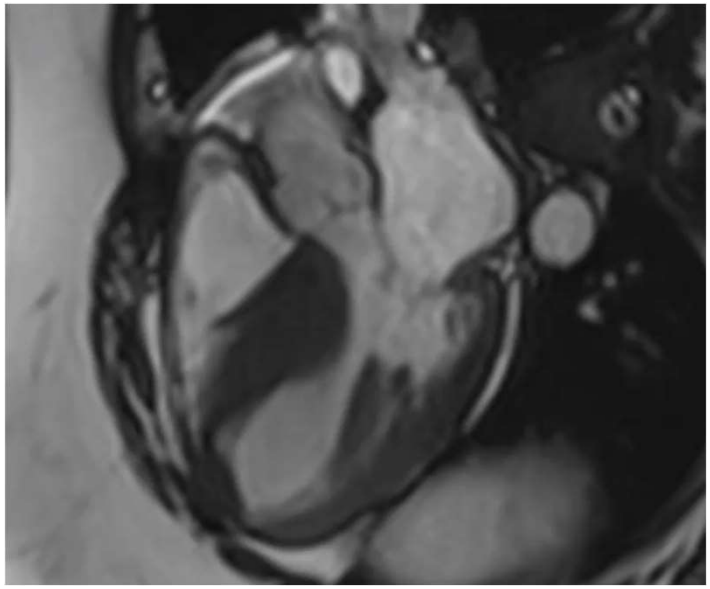












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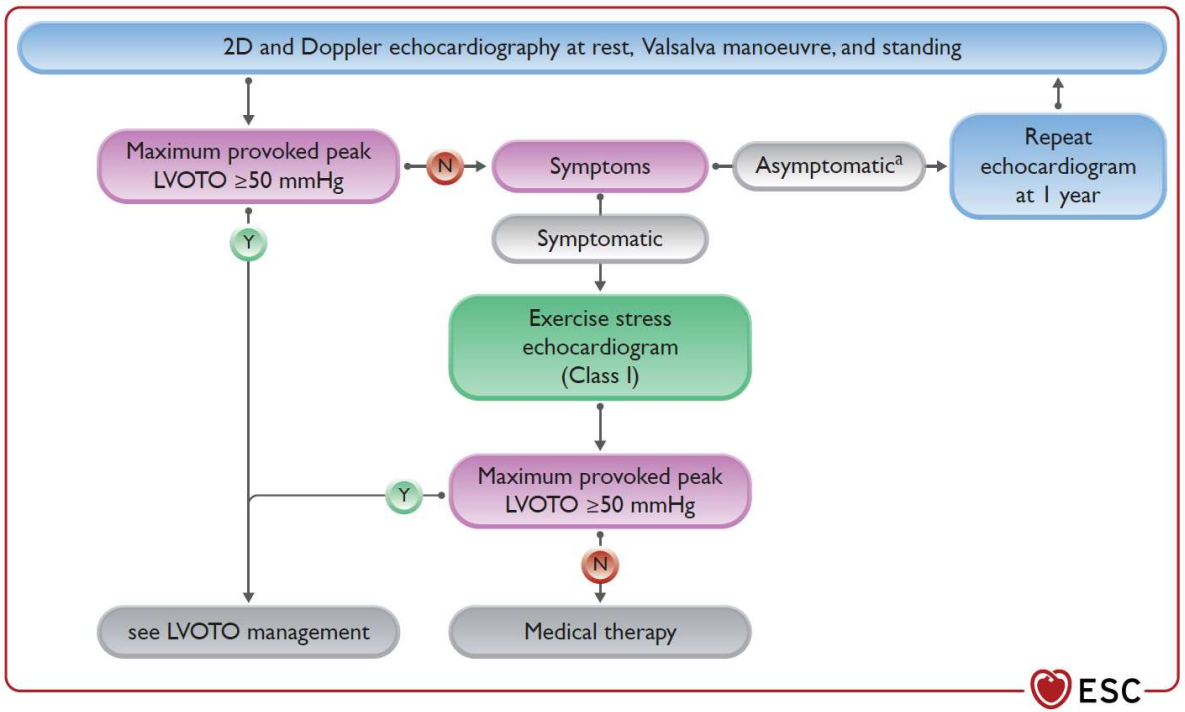
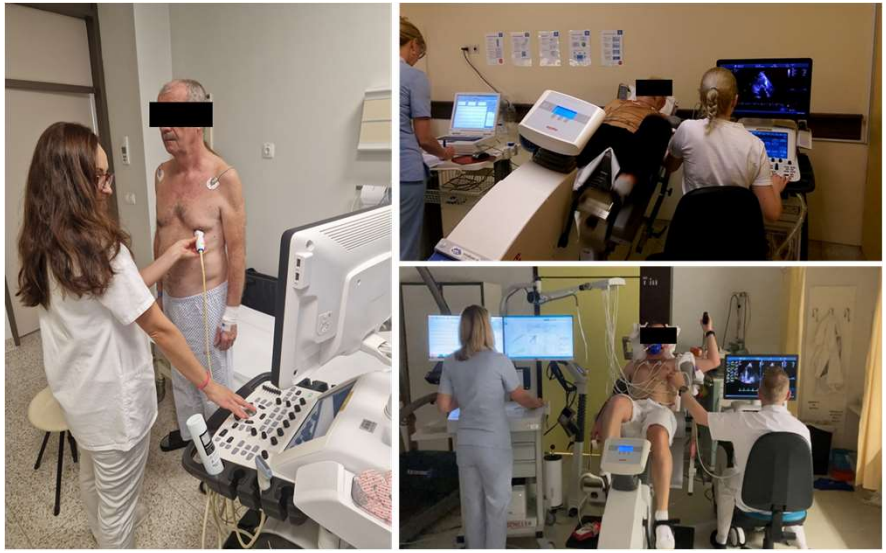
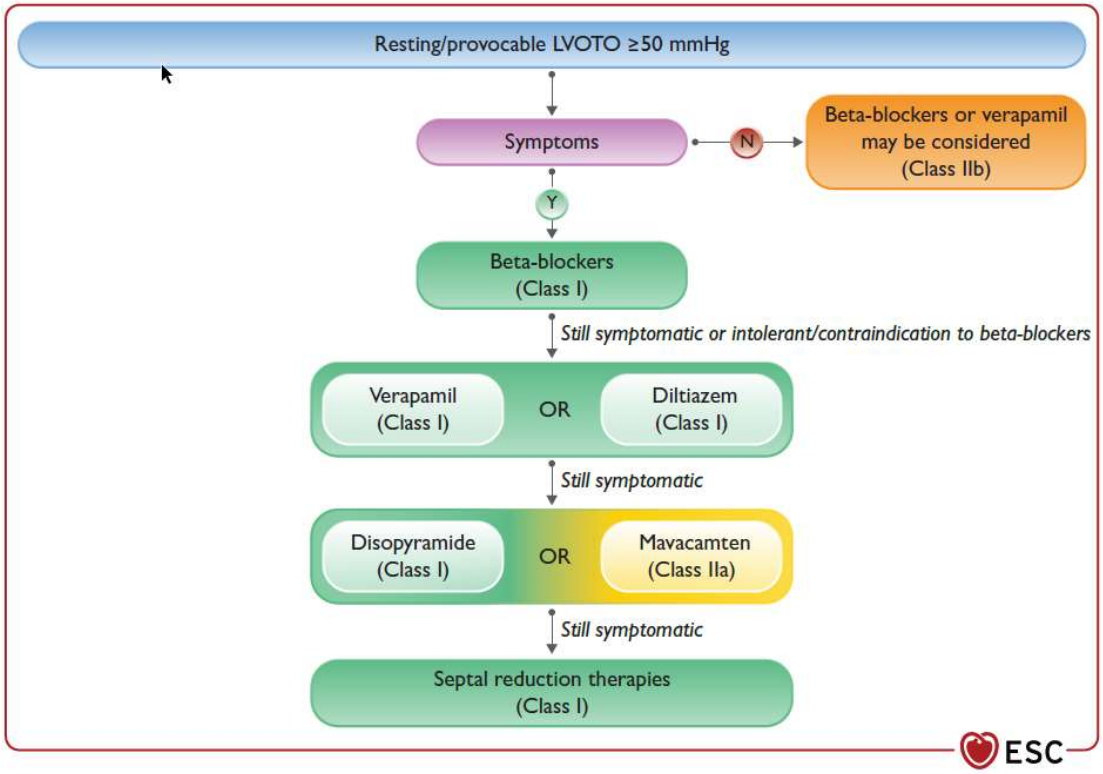
Protocol for the Assessment of LVOTO





Cardim N, et al. *Eur Heart J Cardiovasc Imaging*. Published online November 4, 2025. doi:10.1093/ehjci/jeaf282

Protocol for the Treatment of LVOTO



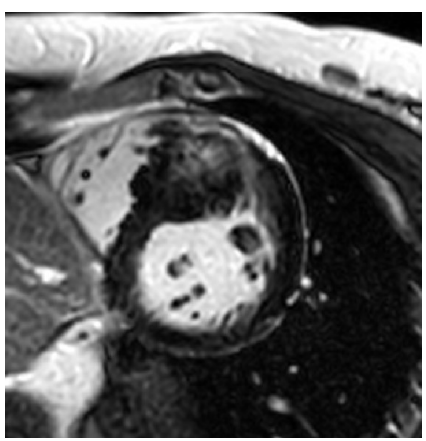
1	B-R	3. For patients with obstructive HCM who have persistent symptoms* attributable to LVOTO despite beta blockers or nondihydropyridine calcium channel blockers, adding a myosin inhibitor (adult patients only), or disopyramide (in combination with an atrioventricular nodal blocking agent), or SRT performed at experienced centers,§ is recommended.[7–14]

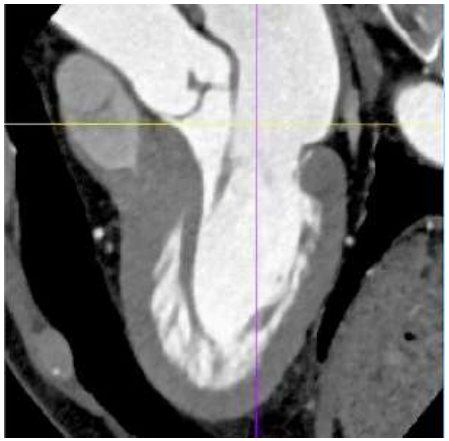
- SRT, when performed by experienced operators in comprehensive HCM centers, is very effective for relieving LVOTO and can be used instead of mavacamten or disopyramide

MMI in the Assessment, Periprocedural Monitoring, and Follow-Up After SRT

Surgical myectomy ± MV surgery

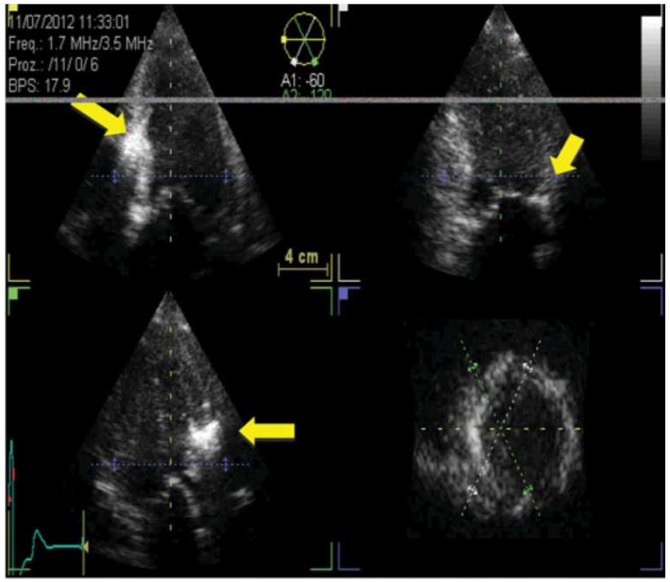
- Marked mitral leaflet/chordal elongation
- PM abnormalities – hypertrophy, bifidity, anterior/apical displacement, direct insertion into the anterior mitral valve leaflet





Alcohol septal ablation

- Myocardial contrast echocardiography is essential prior to alcohol injection



PACE-CME

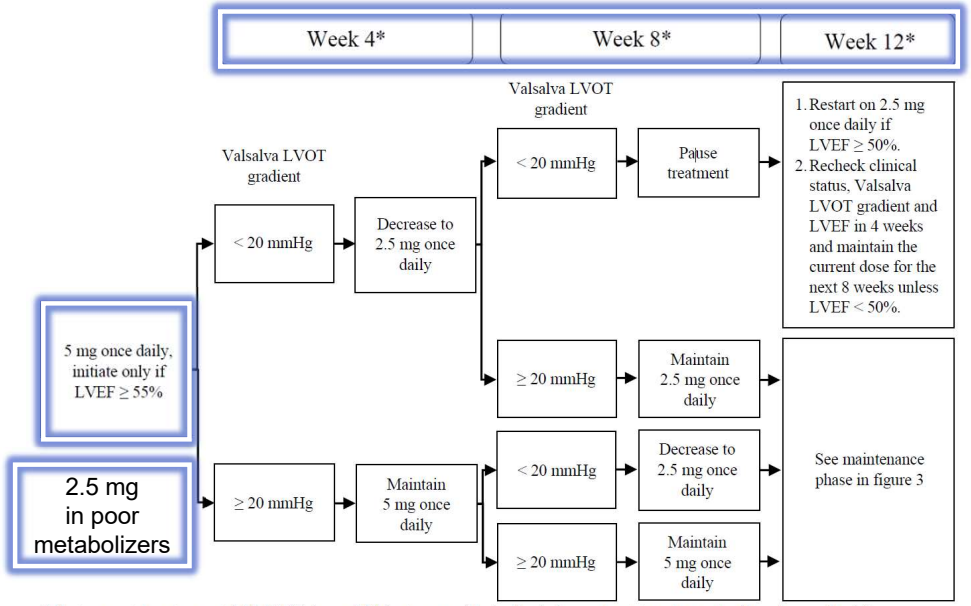
Cardim N, et al. *Eur Heart J Cardiovasc Imaging*. 2015;16(3):280.

Imaging in Monitoring Treatment With Cardiac Myosin Inhibitors

- CMIs act by reducing actin-myosin cross-bridge formation and LV contractility
- Close monitoring of LV systolic function is mandated during drug administration, dose titration, and maintenance treatment
- LVEF ≥55% and LVEF ≥60% are a prerequisite for initiating treatment with the CMIs mavacamten and aficamten, respectively

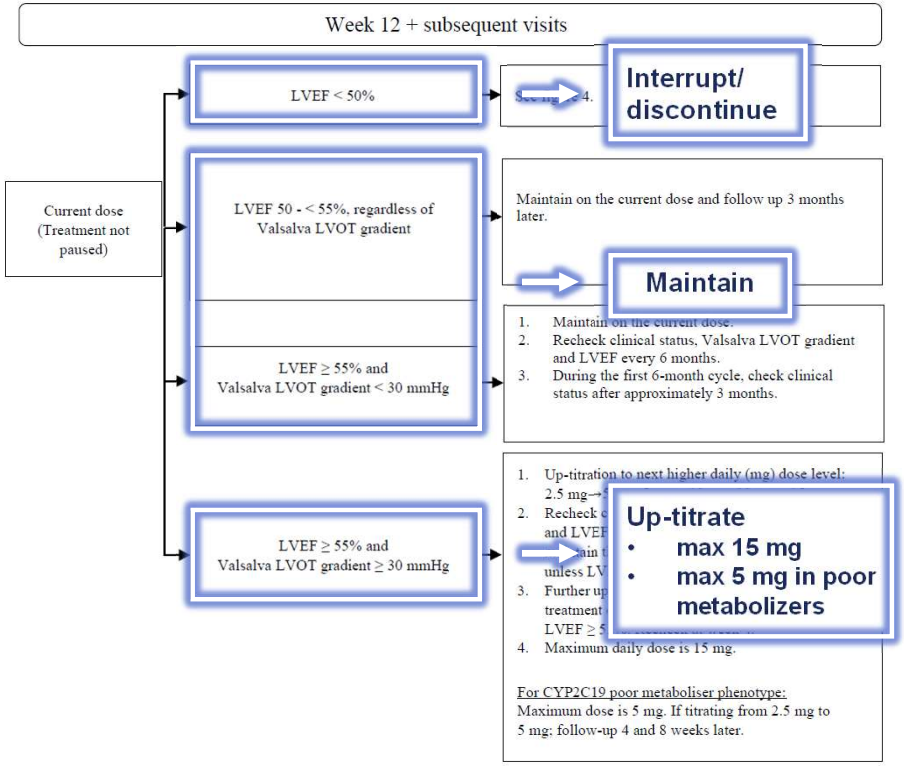
Mavacamten SmPC

Figure 2: Treatment initiation in CYP2C19 intermediate, normal, rapid and ultra-rapid metaboliser phenotype



* Interrupt treatment if LVEF is < 50% at any clinical visit; restart treatment after 4 weeks if LVEF ≥ 50% (see figure 4).
LVEF = left ventricular ejection fraction; LVOT = left ventricular outflow tract

Figure 3: Maintenance phase



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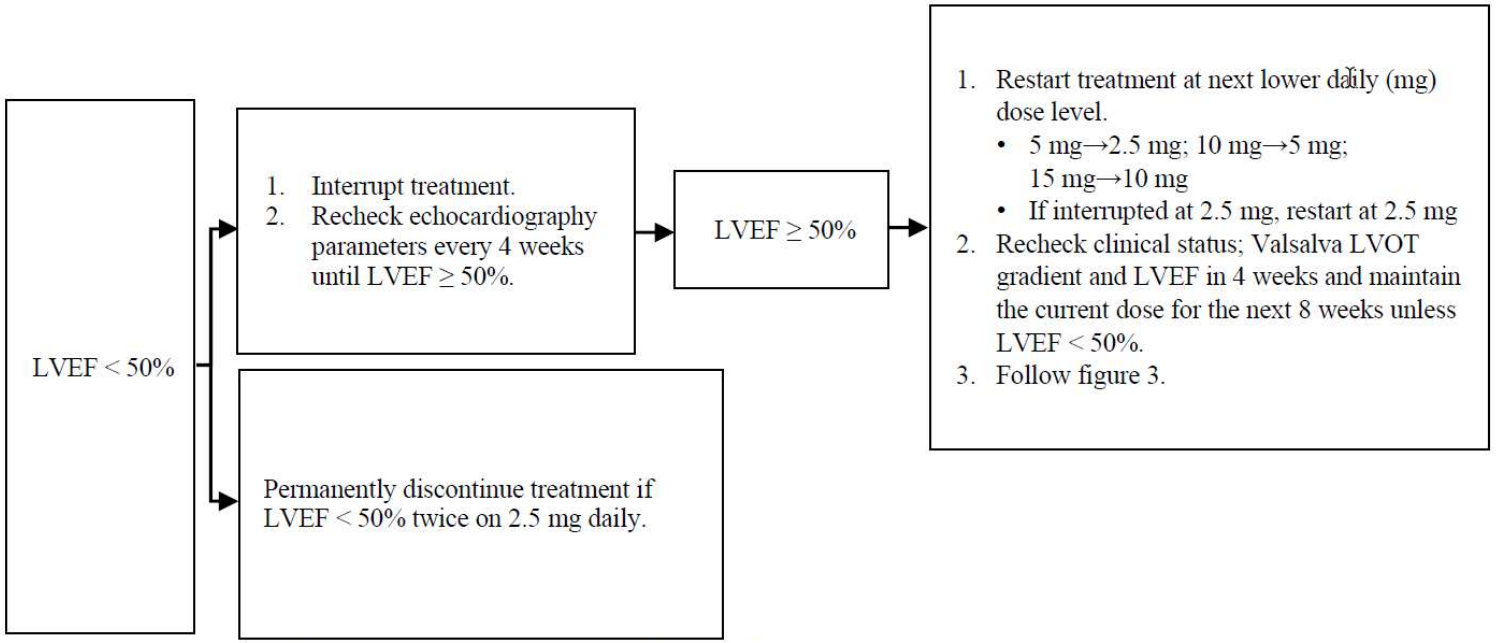
Mavacamten: Treatment Interruption

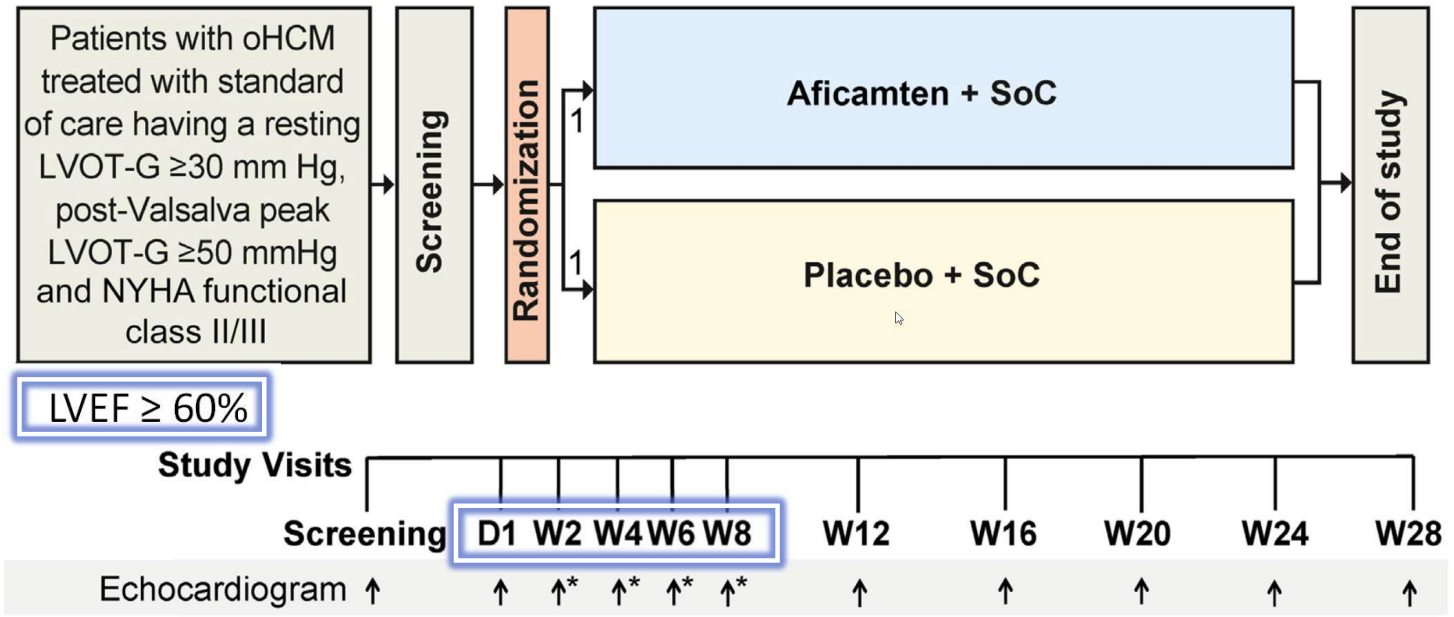
Figure 4: Treatment interruption at any clinic visit if LVEF < 50%



LVEF = left ventricular ejection fraction; LVOT = left ventricular outflow tract

Aficamten: Treatment Initiation and Monitoring

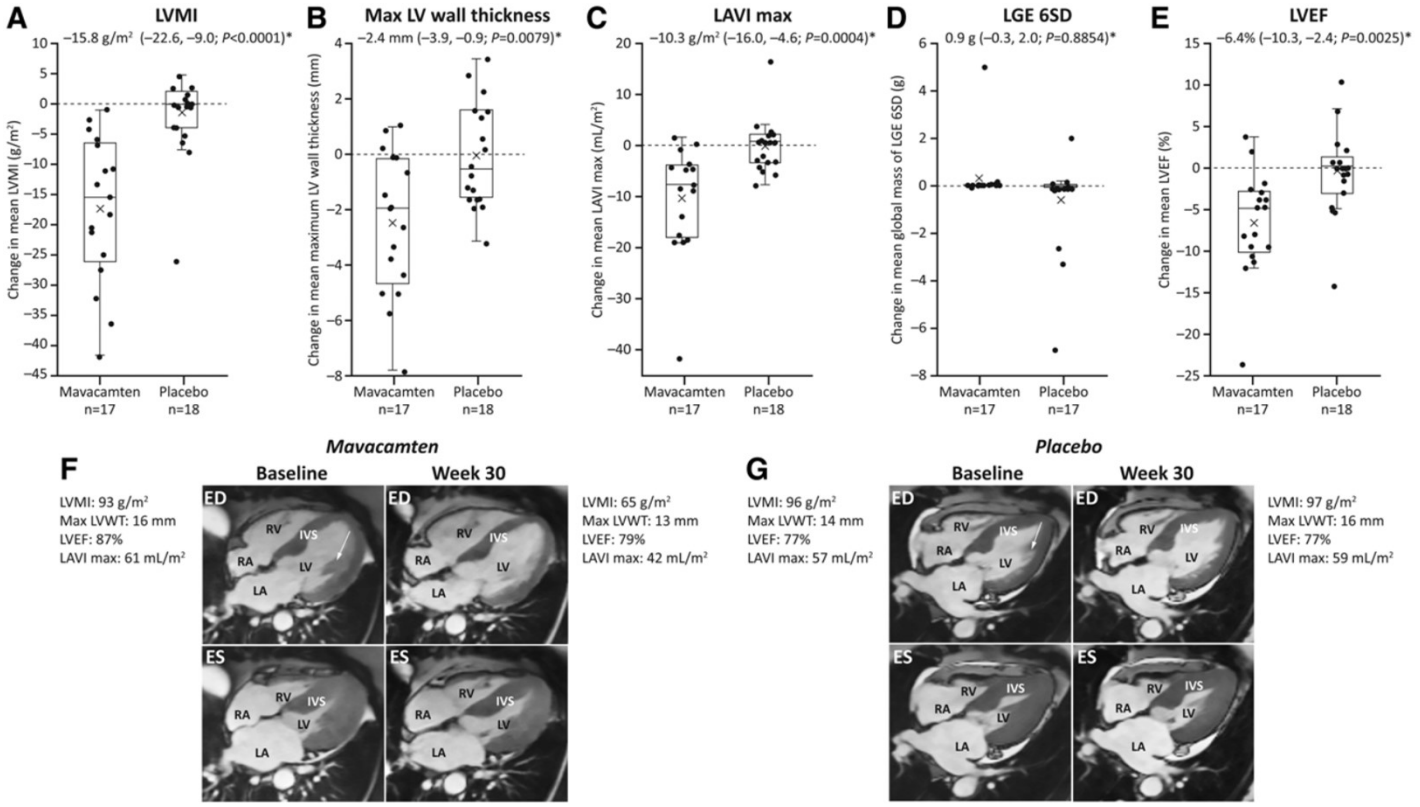
FIGURE 2 Overview of the SEQUOIA-HCM Study



PACE-CME

Coats CJ, et al. *JACC Heart Fail*. 2024;12(1):199-215.

MMI in Monitoring Response to CMI Therapy

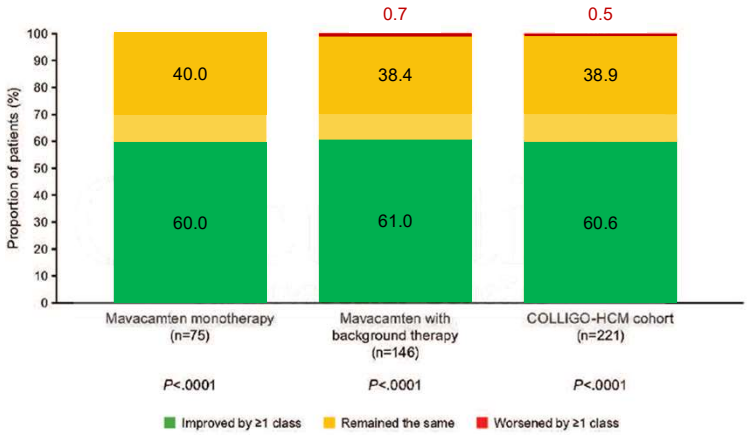


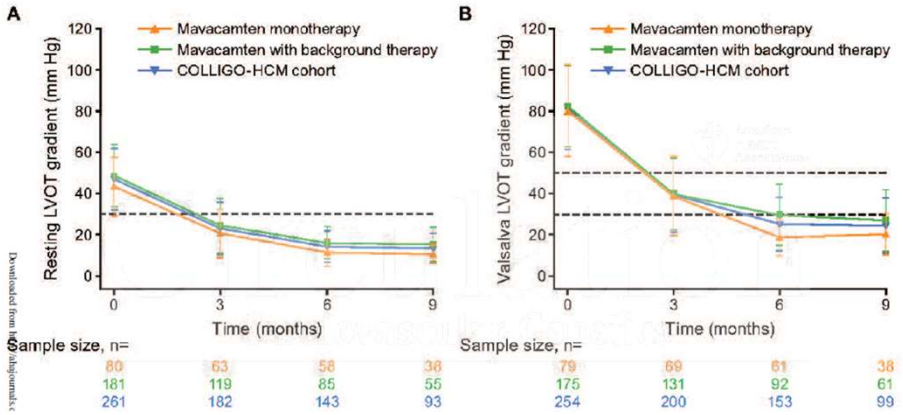
PACE-CME

Saberi S, et al. *Circulation*. 2021; 143(6):606-608.

Real-World Efficacy and Safety of Mavacamten: Evidence From COLLIGO-HCM

- Retrospective, observational, multicenter, international study
- 7 participating sites in 5 countries across 4 continents
- 278 patients receiving mavacamten included from April 2022 to February 2025
  - n = 88; mavacamten monotherapy
  - n = 190; mavacamten with background therapy (BB, CCB)





PACE-CME

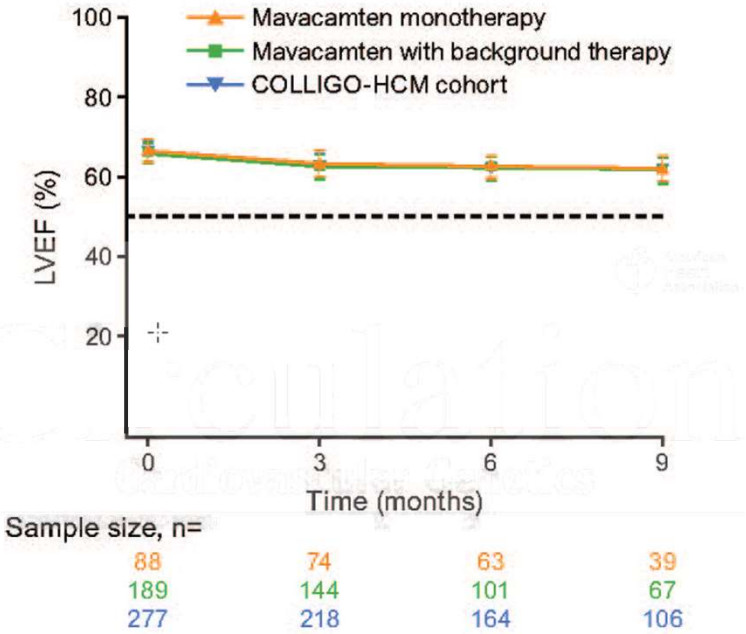
Bilen O, et al. *Circ Genom Precis Med*. Published online November 10, 2025. doi:10.1161/CIRCGEN.125.005502

Real-World Efficacy and Safety of Mavacamten: Evidence From COLLIGO-HCM

Table 2. Summary of Safety Results

	Mavacamten monotherapy (n=88)	Mavacamten with background therapy (n=190)	COLLIGO-HCM cohort (n=278)
Mavacamten treatment interruption due to LVEF <50%	5 (5.7)	6 (3.2)	11 (4.0)
Recovery of LVEF to ≥50% after mavacamten treatment interruption	5	5*	10*
Resumed treatment after mavacamten treatment interruption	4	5	9
Mavacamten treatment discontinuation due to LVEF <50%	2 (2.3)	1 (0.5)	3 (1.1)
Recovery of LVEF to ≥50% after mavacamten treatment discontinuation	2	1	3

Data are presented as n (%).
*The LVEF of 1 patient had not recovered to ≥50% by the data extraction date.
COLLIGO-HCM indicates mavaCamten ObservationaL evIdence Global cOnsortium in HCM; and LVEF, left ventricular ejection fraction.



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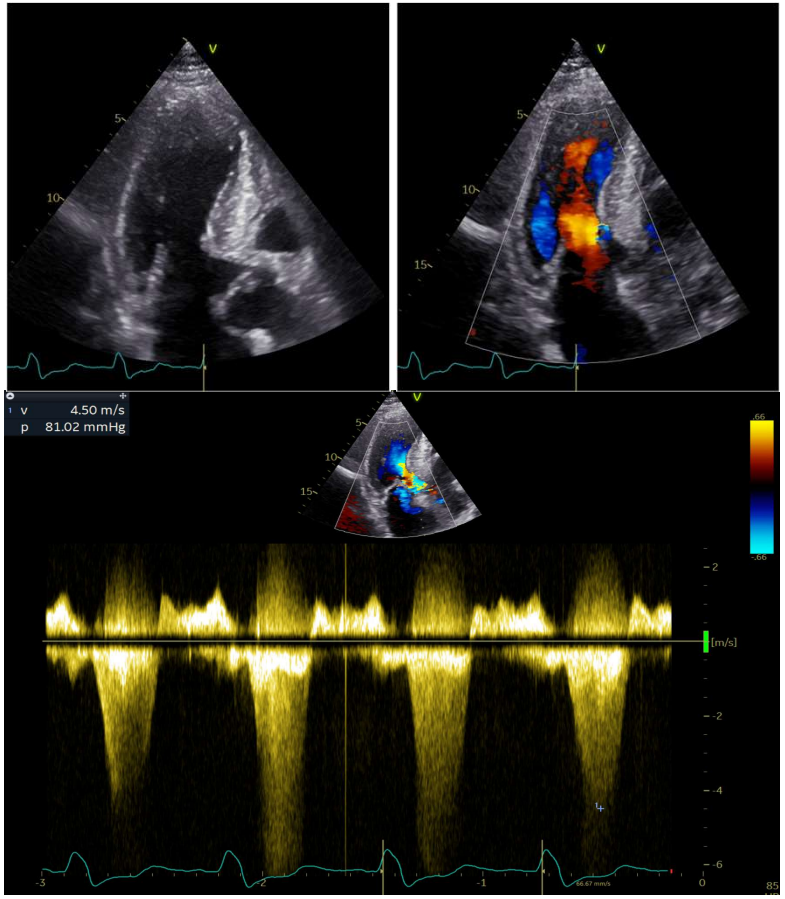
Bilen O, et al. *Circ Genom Precis Med*. Published online November 10, 2025. doi:10.1161/CIRCGEN.125.005502

Case Study

58-year-old man

- oHMC diagnosed in 2024
- NYHA II, NT-proBNP 946 ng/L
- Bisoprolol 2x5 mg
- Echocardiography 04/2025 → mavacamten initiation 5 mg
  - Maximum LV wall thickness 19 mm mid inferior septum
  - LVEF 58%
  - Resting gradient 63 mmHg, Valsalva 81 mmHg, exercise (squats) 100 mmHg
  - SAM, mild-moderate MR



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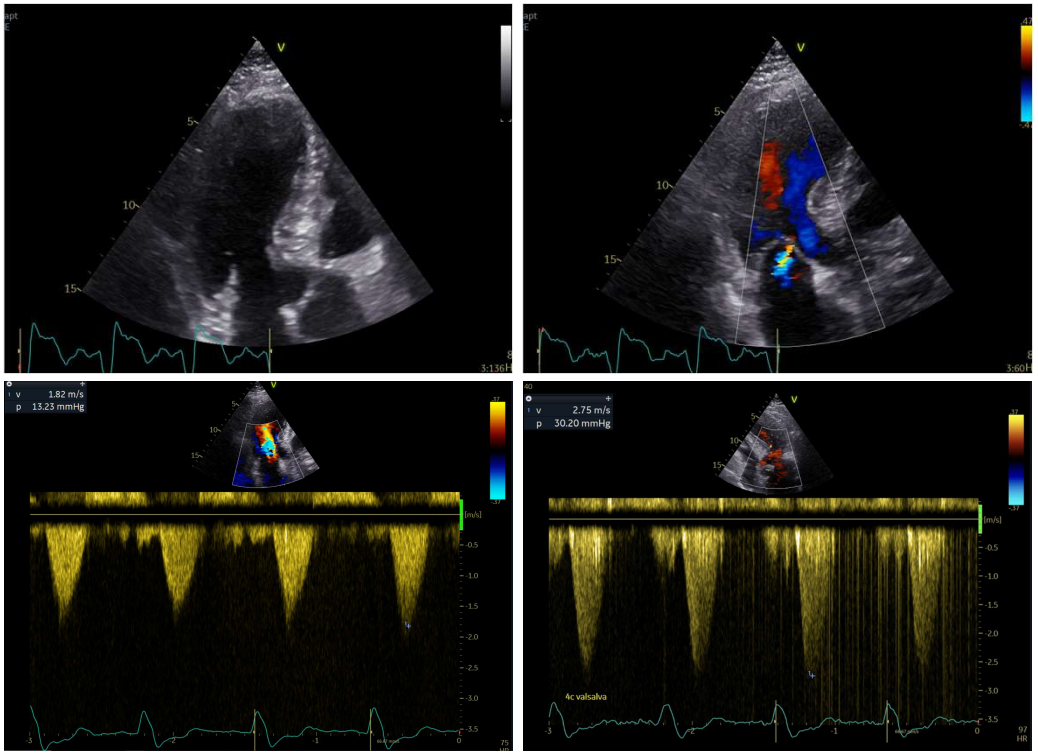
Mavacamten 5mg 12w

- Mild improvement of symptoms
- NT-proBNP 196 ng/L
- LVEF 55%
- Resting LVOT gradient 13 mmHg
- Valsalva 30 mmHg



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How Would You Proceed?

A. Maintain 5 mg
B. Up-titrate to 10 mg
C. Decrease to 2.5 mg
D. Interrupt treatment

PACE-CME

How Would You Proceed?

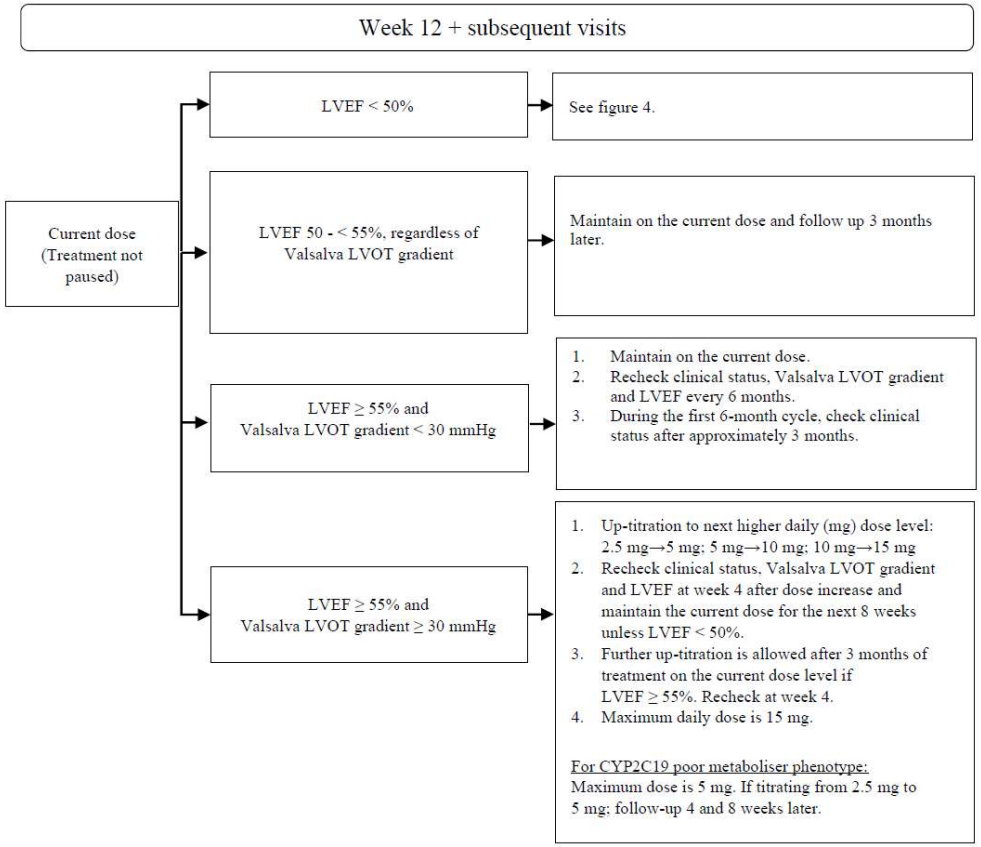
A. Maintain 5 mg

B. Up-titrate to 10 mg

C. Decrease to 2.5 mg

D. Interrupt treatment

Figure 3: Maintenance phase



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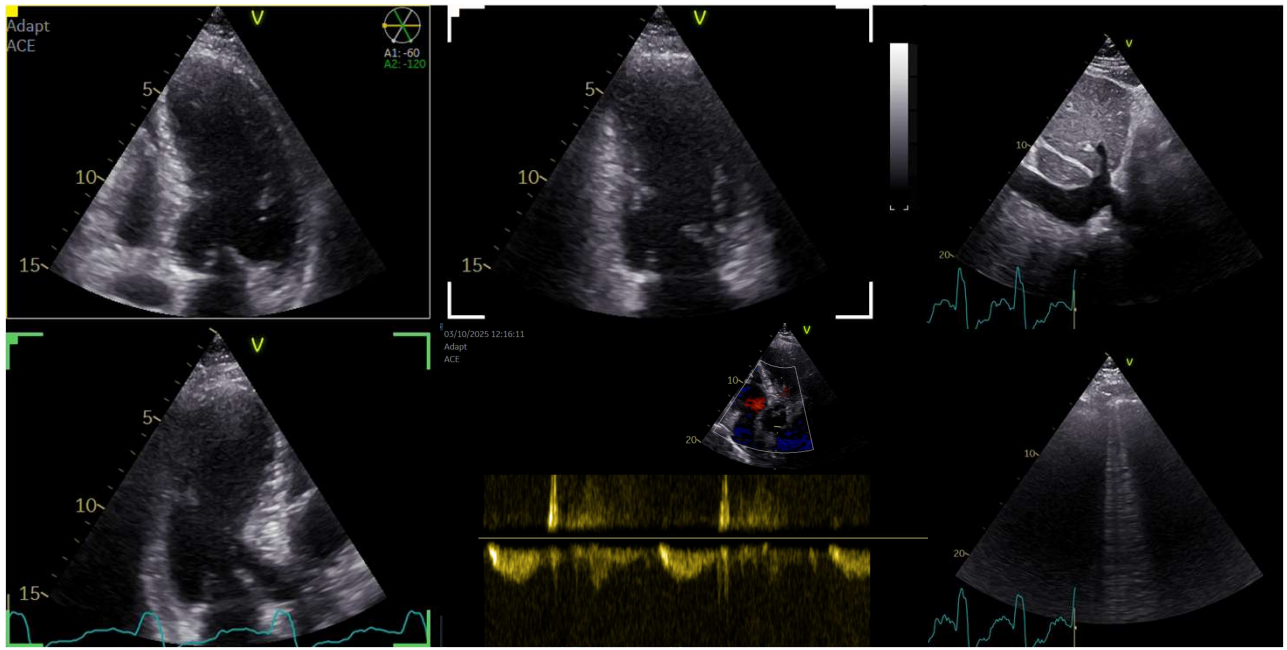
Mavacamten 10 mg 8w

- Severe dyspnea (NYHA III)
- Signs of hypervolemia
- LVEF 29%
- Resting gradient 4 mmHg



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How Would You Proceed?

A. Maintain 10 mg and reassess after 4 weeks
B. Decrease to 5 mg and reassess after 4 weeks
C. Decrease to 2.5 mg and reassess after 4 weeks
D. Interrupt treatment and reassess after 4 weeks
E. Permanently stop treatment and reassess after 4 weeks

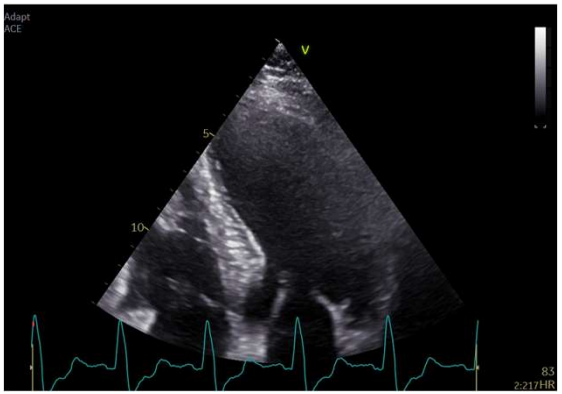
How Would You Proceed?

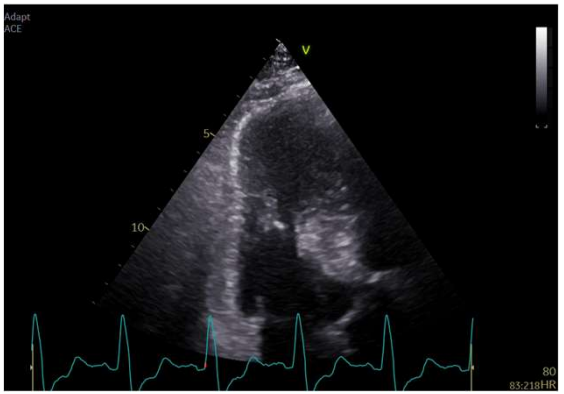
A. Maintain 10 mg and reassess after 4 weeks
B. Decrease to 5 mg and reassess after 4 weeks
C. Decrease to 2.5 mg and reassess after 4 weeks
D. Interrupt treatment and reassess after 4 weeks
E. Permanently stop treatment and reassess after 4 weeks

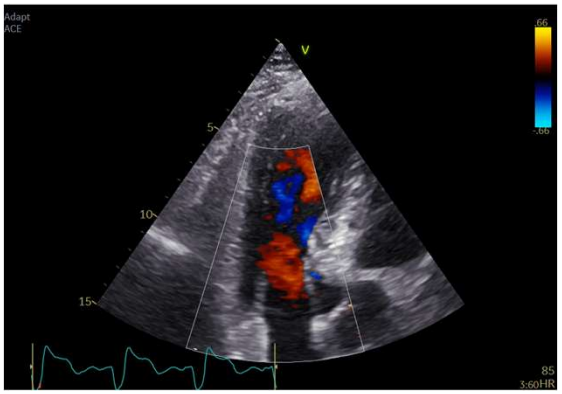
+ furosemide, sacubitril/valsartan, spironolactone

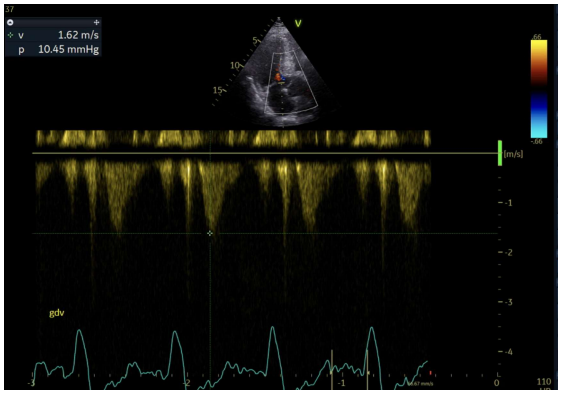
4 Weeks After Dose Interruption

- Improvement of symptoms
- No signs of hypervolemia
- LVEF 52%
- NT-proBNP 539 ng/L
- Resting gradient 5 mmHg,
- Valsalva 10 mmHg









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How Would You Proceed?

A. Interrupt treatment for another 4 weeks
B. Restart treatment with 10 mg and reassess after 4 weeks
C. Restart treatment with 5 mg and reassess after 4 weeks
D. Restart treatment with 2.5 mg and reassess after 4 weeks
E. Permanently stop treatment

How Would You Proceed?

A. Interrupt treatment for another 4 weeks

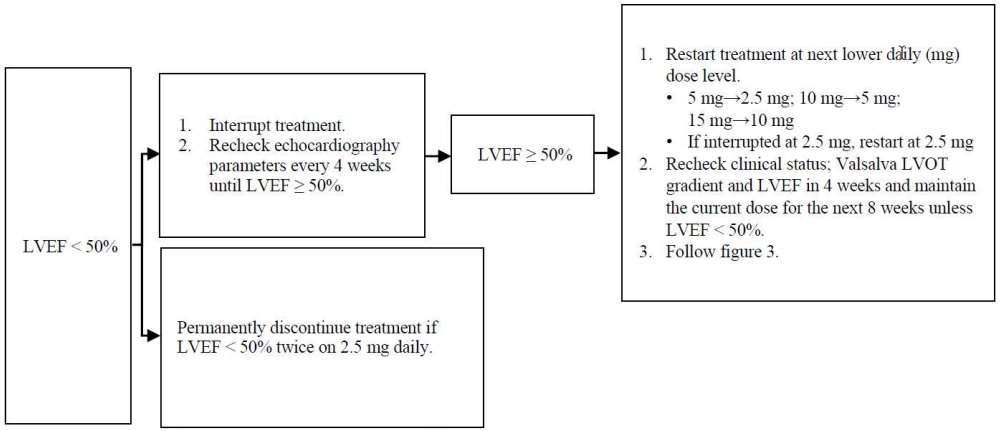
B. Restart treatment with 10 mg and reassess after 4 weeks

C. **Restart treatment with 5 mg and reassess after 4 weeks**

D. Restart treatment with 2.5 mg and reassess after 4 weeks

E. Permanently stop treatment

Figure 4: Treatment interruption at any clinic visit if LVEF < 50%



LVEF = left ventricular ejection fraction; LVOT = left ventricular outflow tract

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Conclusion

- Echocardiography (with provocative maneuvers) is recommended in ALL patients with HCM to detect LVOTO
- Exercise stress echocardiography is recommended in SYMPTOMATIC patients
- TOE, CMR, or CT help in the assessment of the extent and distribution of hypertrophy and of mitral valve apparatus prior to SRT
- Myocardial contrast echocardiography is mandatory prior to alcohol septal ablation
- Echocardiography is of paramount importance during CMI administration, dose titration, and maintenance treatment

Thank You for Your Attention

JOURNAL ARTICLE ACCEPTED MANUSCRIPT

Role of multimodality cardiac imaging in the management of patients with hypertrophic cardiomyopathy in 2025. A Clinical Consensus Statement of the European Association of Cardiovascular Imaging (EACVI) of the ESC FREE

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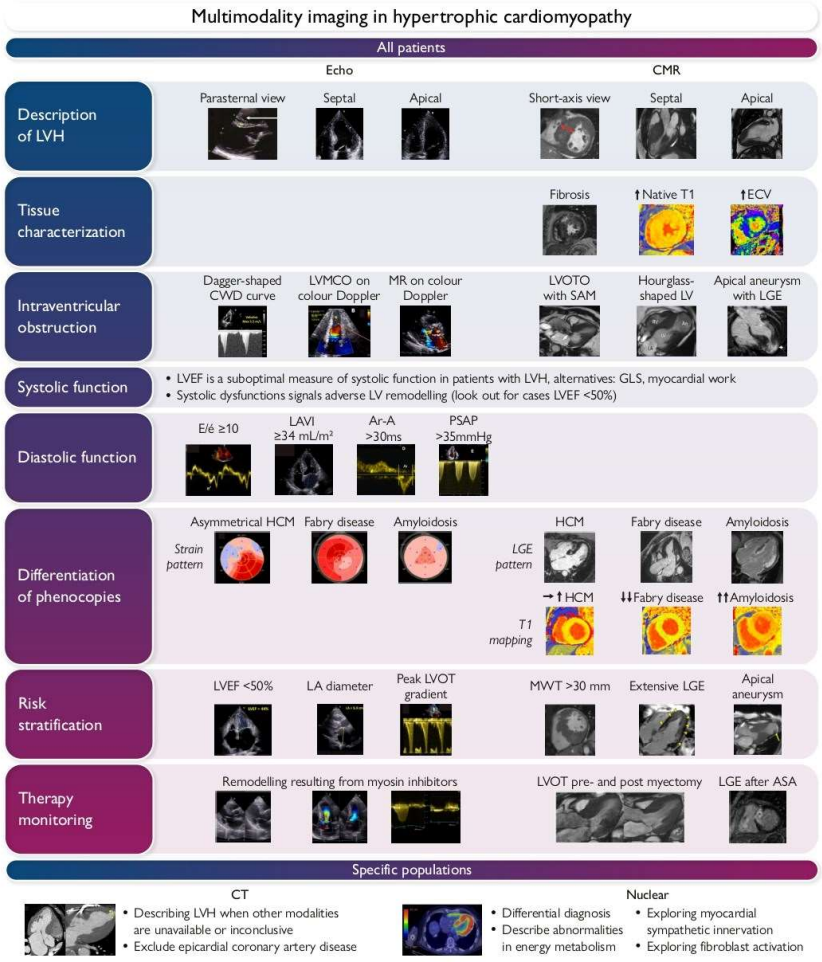

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