5th iHEF CARD

The 5th Indonesian Symposium on Heart Failure and Cardiometabolic Disease

Indonesian Working Group on Heart Failure and Cardiometabolic Disease

WHFS World Heart Failure Society

HEART FAILURE SOCIETY (SINGAPORE)

Exploring Hypertrophic Cardiomyopathy: What Should We Do?

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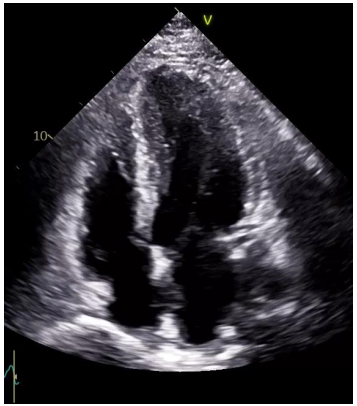
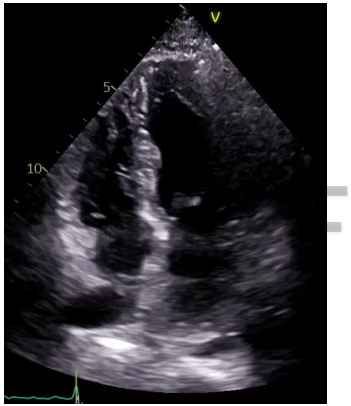
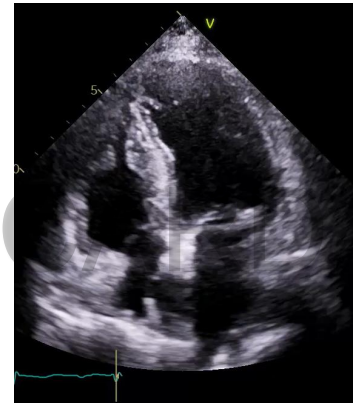
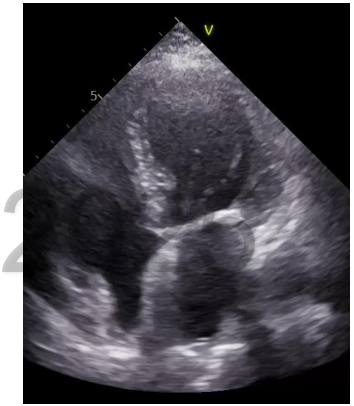
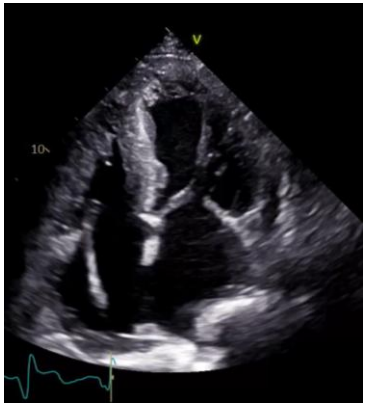
Quick Poll: Do you think this is HCM?

1 2 3 4 5

Learning Objectives

Understand	Understand HCM definition and classification
Explore	Explore key genetic and pathophysiologic mechanisms
Review	Review diagnostic modalities
Summarize	Summarize evidence-based management strategies

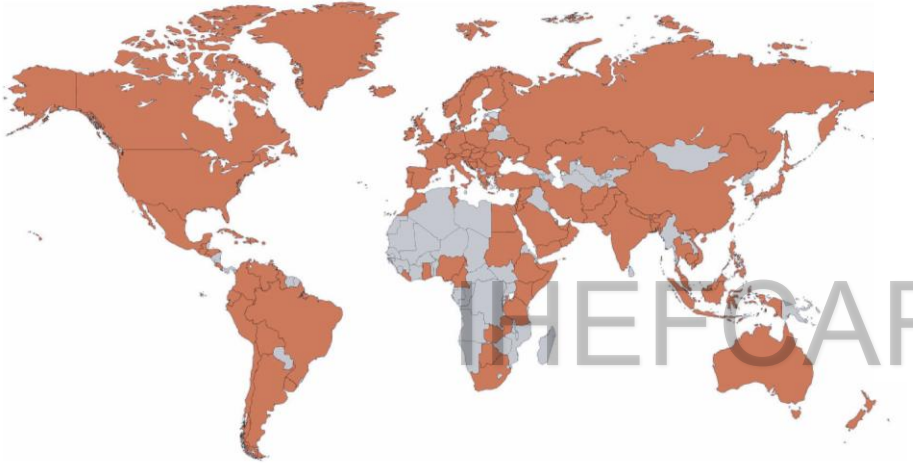
What is Hypertrophic Cardiomyopathy?

Definition: Unexplained LV hypertrophy (in the absence of another cardiac, systemic, or metabolic disease)

Distinguish from secondary causes (HTN, AS)

Sarcomeric disease: primary myocardial disorder

Epidemiology of HCM



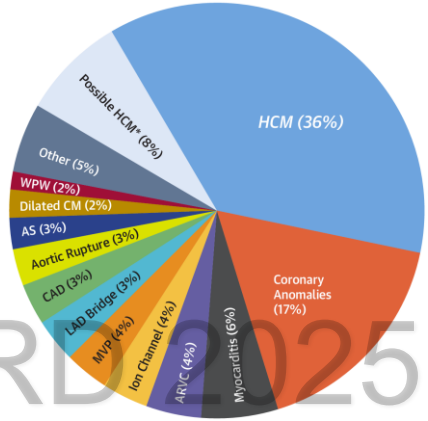
125 Countries involving 90% of world population

↓

Prevalence 1:200-1:500

↓

Estimated ~15-20 million affected worldwide



The most common cause of SCD in competitive athletes

Only 10–20% clinically diagnosed

Increasing recognition in aging populations

Male predominance in diagnosis, female symptom burden

Maron, B.J. et al. J Am Coll Cardiol. 2022;79(4):372–389

Types of HCM

- Obstructive HCM (oHCM)
- Non-obstructive HCM (nHCM)
- Apical and midventricular variants
- Genotype-positive, phenotype-negative individuals

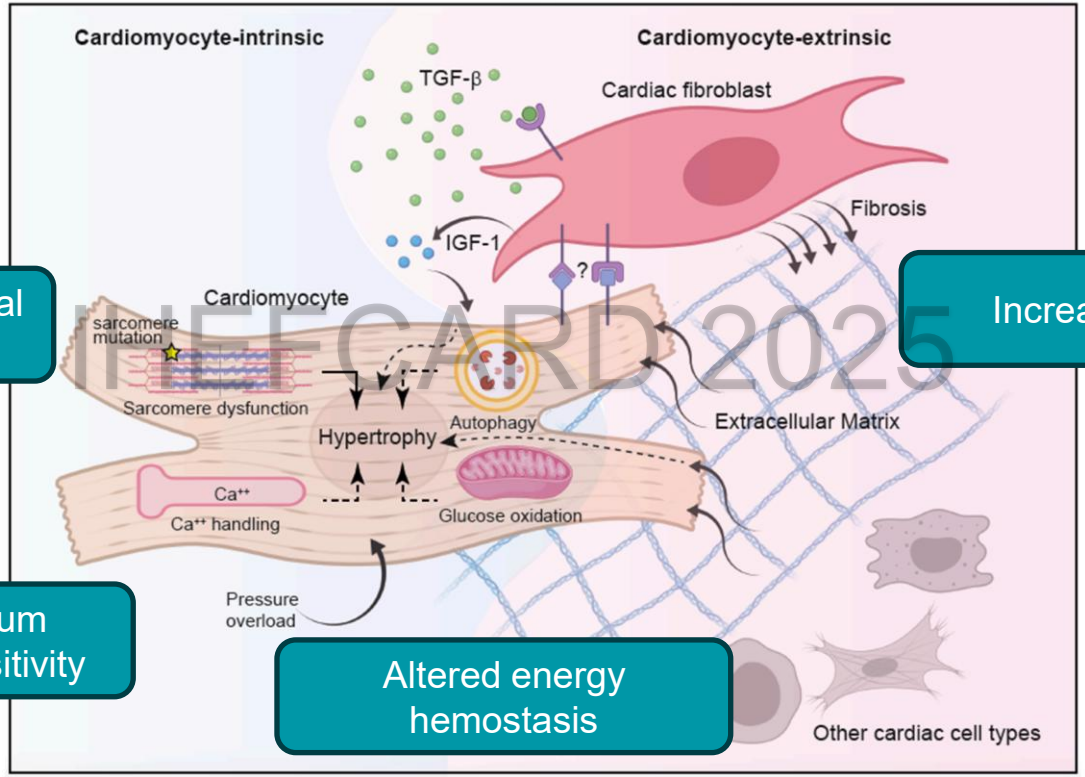
Genetic Basis of HCM

Autosomal dominant mutations

Common genes: MYH7, MYBPC3, TNNT2, TNNI3

Over 2,000 known mutations

Pathophysiology of HCM



Impaired biomechanical stress sensing

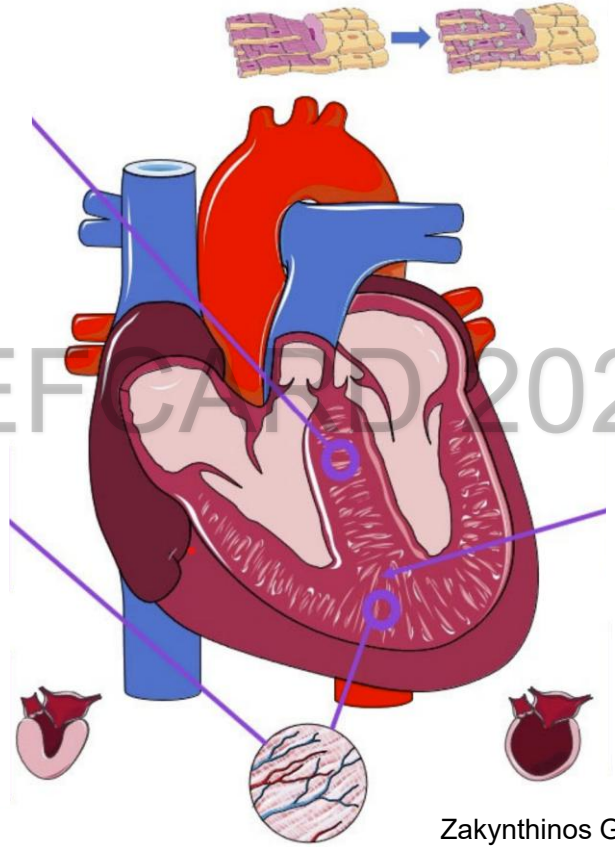
Increase fibrosis

Impaired calcium cycling and sensitivity

Altered energy hemostasis

Chou C, et al. Int. J. Mol. Sci. 2021, 22, 8933.

Key Pathophysiological Changes in HCM



Zakynthinos GE, et al. J. Cardiovasc. Dev. Dis. 2024, 11, 401.

Dynamic LVOT obstruction

Complex interplay of:

- MV and subvalvular apparatus abnormalities
- septal hypertrophy
- narrowing of the LVOT
- steep and/or anteroseptal angulation of the outflow tract

Classification HCM on the basis of obstruction:

- resting obstruction (LVOT gradient ≥30 mm Hg)
- latent obstruction (<30 mmHg at rest, ≥30 mm Hg with provocation)
- non-obstructive (<30 mmHg at rest and with provocation)

Initial Clinical Evaluation and Testing Algorithm for Patients With or Suspected of Having Hypertrophic Cardiomyopathy

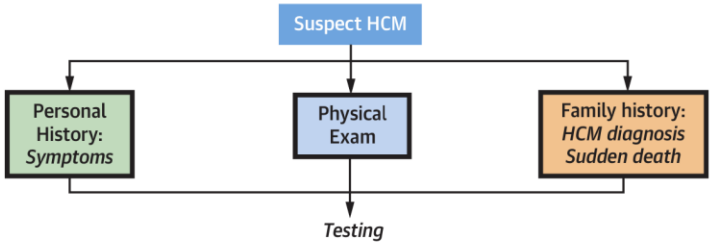


TABLE 1 Guide to Clinical Evaluation and Noninvasive Testing in HCM

Test	Initial Evaluation	Follow-Up
History taking and examination	+	Annual
Echocardiogram	+	Annual
Contrast CMR[a]	+	Every 3-5 y[b]
Stress (exercise) echocardiography[c]	+	Individualized
Ambulatory ECG[d]	+	1-3 y[e]
12-lead ECG	+	Annual

[a]Optional in patients >65 years of age. [b]Or more frequently when there is concern for increased late gadolinium enhancement or development of suspected left ventricular apical aneurysm in adults, or increasing wall thickness in young patients. [c]When gradient at rest is absent or <30 mm Hg. [d]A 24- to 48-hour Holter or ≥2-week wireless patch with continuous recording. [e]Based on presence or absence of arrhythmia.

CMR = cardiac magnetic resonance; ECG = electrocardiography.

Maron, B.J. et al. J Am Coll Cardiol. 2022;79(4):372-389.

Clinical presentation

01

Asymptomatic to symptomatic

02

Dyspnea
Chest pain
Palpitations
Syncope

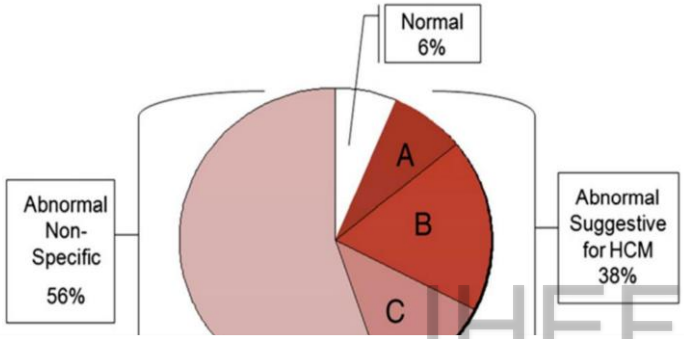
03

History of SCD in family

04

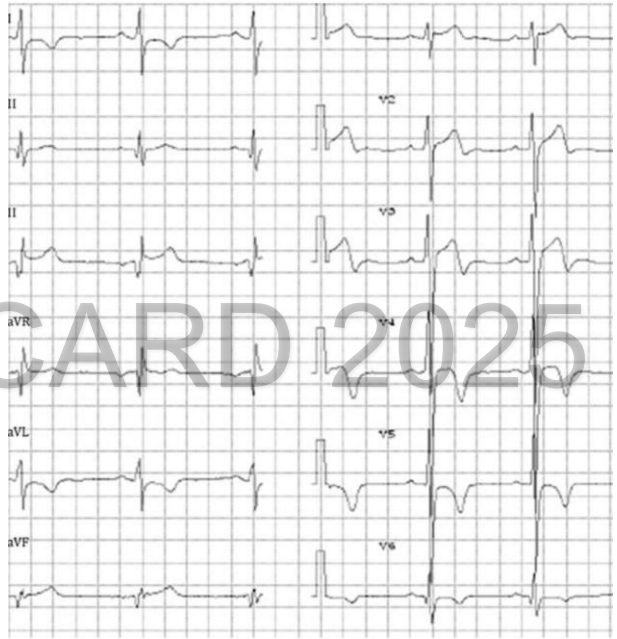
Murmur: late-peaking, dynamic

ECG in HCM



A. Giant T Waves
B. inferolateral Q-waves, LVH, repolarization abnormalities
C. inferolateral Q-waves, LVH

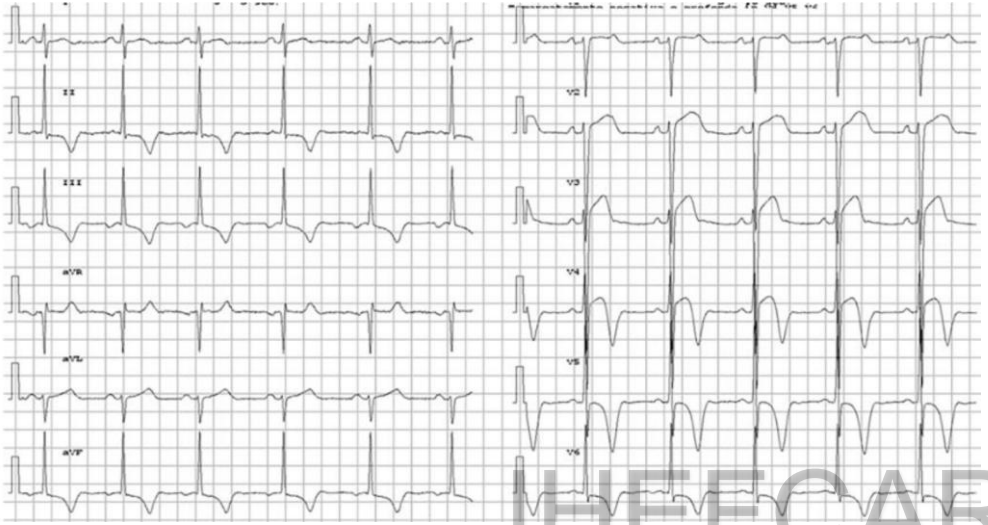
- LVH, Q waves, ST-T abnormalities
- Atrial enlargement
- Not specific, but often abnormal



Male, 21 years old, with sarcomeric HCM.

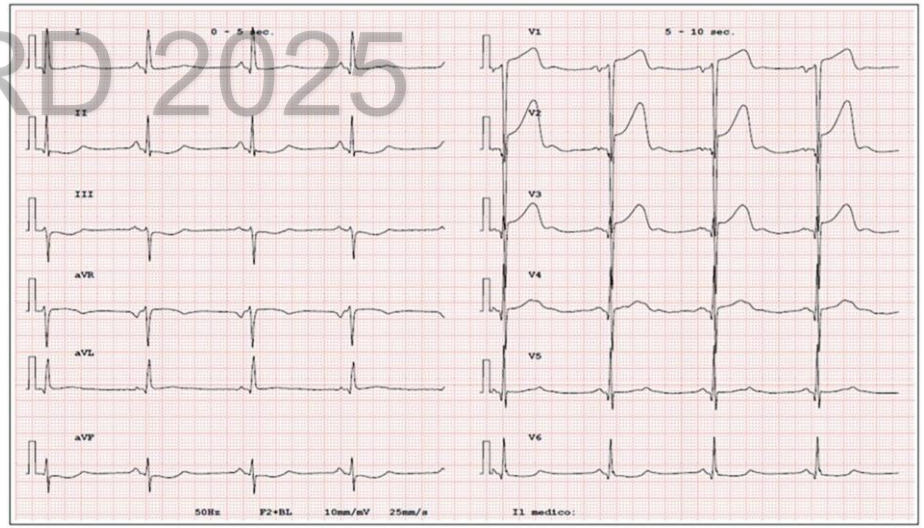
Inferior Q-waves, anterolateral T-wave inversion, ST depression in aVL, deep S-waves in V3–V5.

Bernardini A, et al. European Heart Journal Supplements (2023) 25 (Supplement C), C173–C178



Male, 37 years old, with apical HCM. Giant negative T-waves in V4–V6 and inferior leads, ST segment elevation (pseudo-STEMI pattern) in V2–V3.

Male, 47 years old, with obstructive HCM. QS pattern in V1–V2, marked ST elevation V2–V3, deep S-waves in V1–V4, ST depression in inferior leads, QTc prolongation



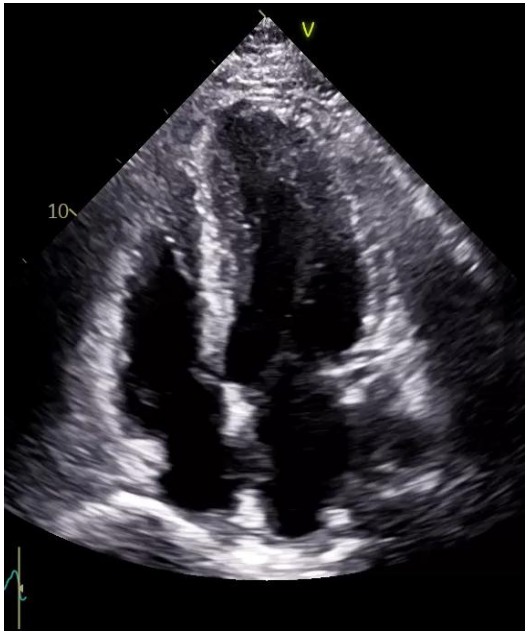
Bernardini A, et al. European Heart Journal Supplements (2023) 25 (Supplement C), C173–C178

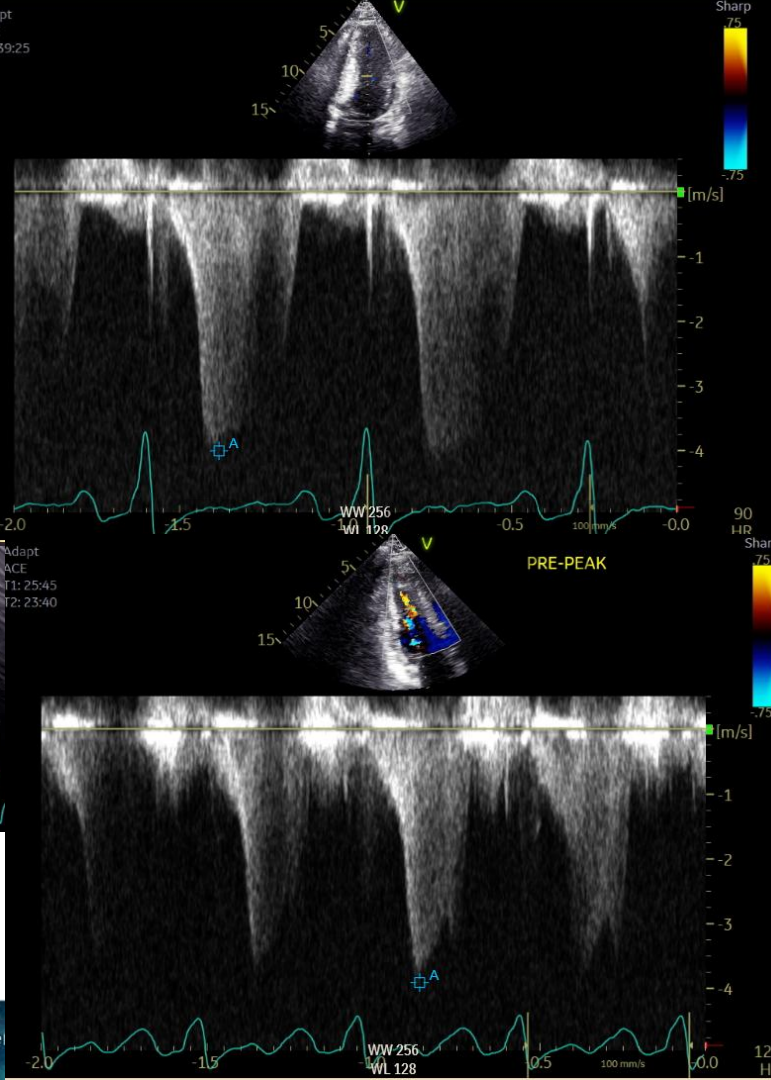
Echocardiography ACC/AHA Guidelines Recommendation (Class I-B)

TEE is recommended in the initial evaluation	In patients with no change in clinical status/events: repeat TEE every 1-2 years	In patients with a change in clinical status/new clinical event: repeat echo	If resting peak LVOT gradient <50 mmHg: TTE provocative maneuvers
Symptomatic patients who have no resting/provocable LVOT peak gradient >= 50 mmHg: exercise TTE	Patient undergo septal myectomy: TEE to assess mitral valve anatomy and function	TTE/TTE for intraprocedural or after procedure (3-6 months) evaluation	Screening: First degree relatives; genotype positive, phenotype negative

Ommen, SR et al. Circulation. 2024;149:e1239–e1311.

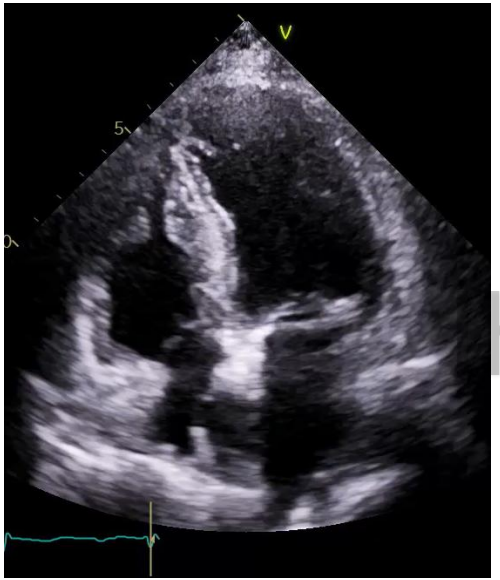
Exercise TTE



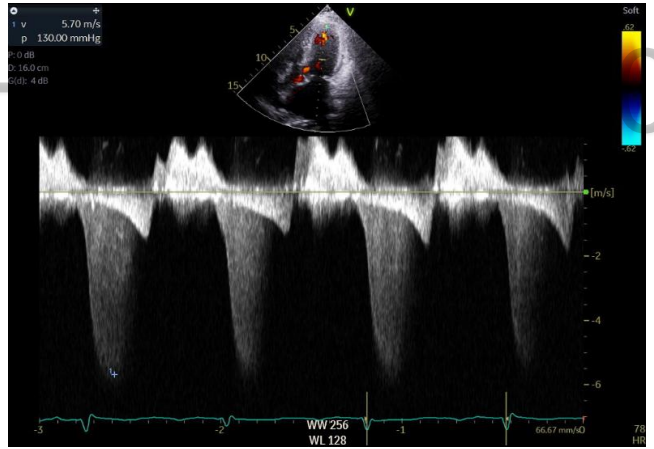


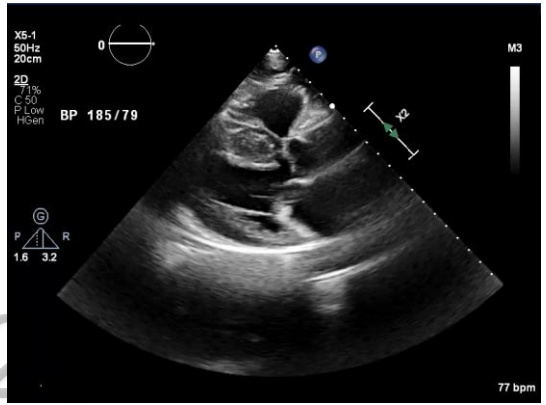
1 Male, 60 yo, symptomatic

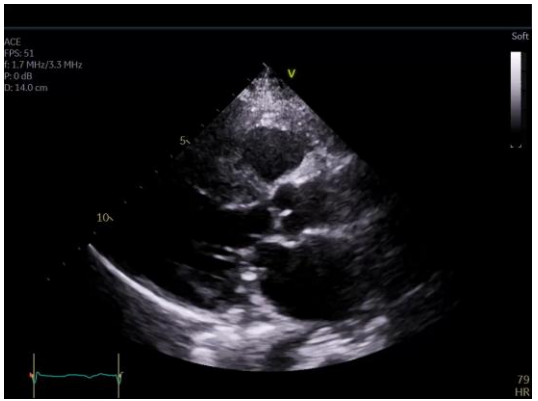
Mid-septal oHCM



66 yo-lady
Recurrent admission for ADHF
History of HT

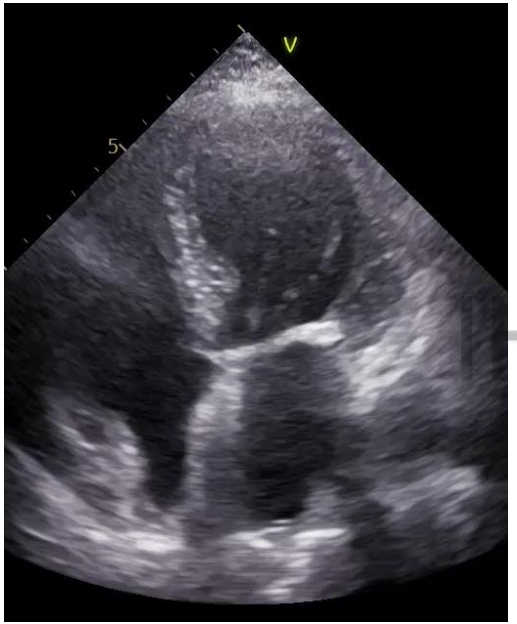




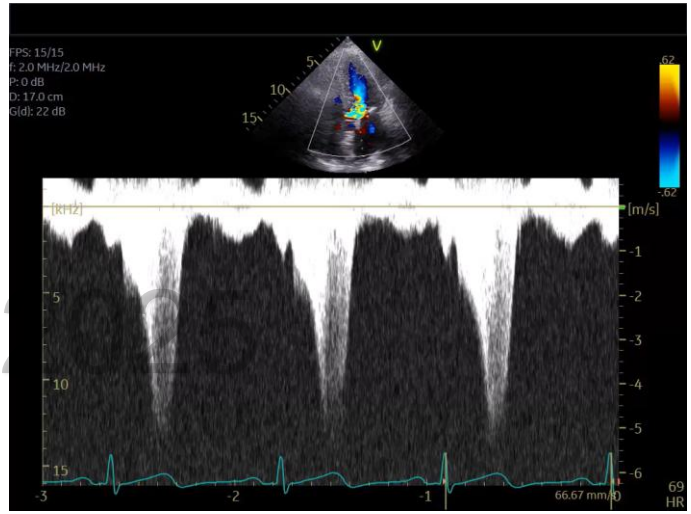


3

oHCM



73 yo-lady
History of myeloma
History of long-standing treated hypertension
No symptoms
Co-incidental findings upon orthopedic surgery



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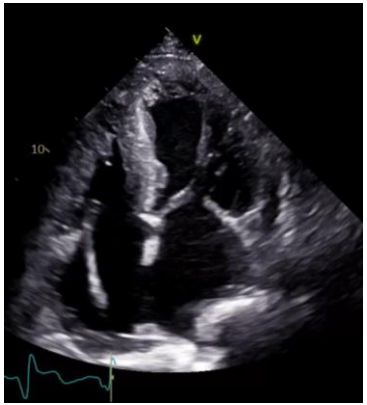
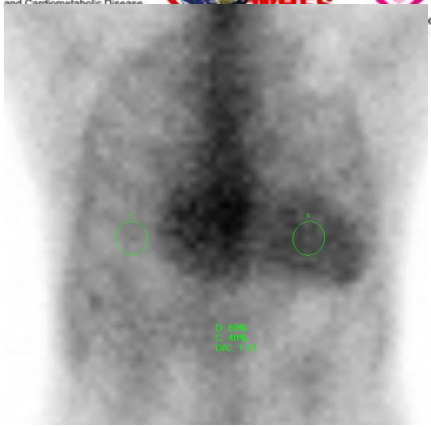


Red flags on echocardiogram: **sparkling myocardium, asymmetric hypertrophy**.

Tc-PYP scan: **Heart/CL ratio 1.51**, uptake similar to ribs → strongly suggestive of ATTR-CM.

Myeloma screen: Negative →

Confirmed wild-type ATTR.



5

2011 — Diagnosed with HCM, ICD placed.

2021 — **LVEF declined to 45%;** coronary arteries normal.

2022 — Tafamidis started

2022–2024 — Multiple heart failure admissions, ICD shocks, eventual start of Tafamidis.

CMR Imaging ACC/AHA Guidelines Recommendations (Class I-B)

01

Clarifications for the patients in whom echocardiography is inconclusive

02

HCM with suspicion of alternative diagnosis

03

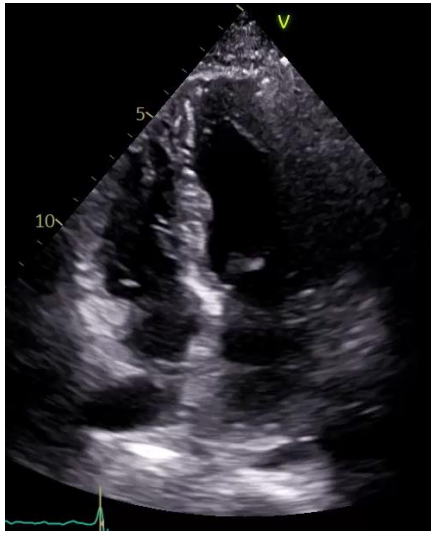
For whom a decision to proceed with ICD remains uncertain: access max LV wall thickness, EF, apical aneurysm, extent LGE

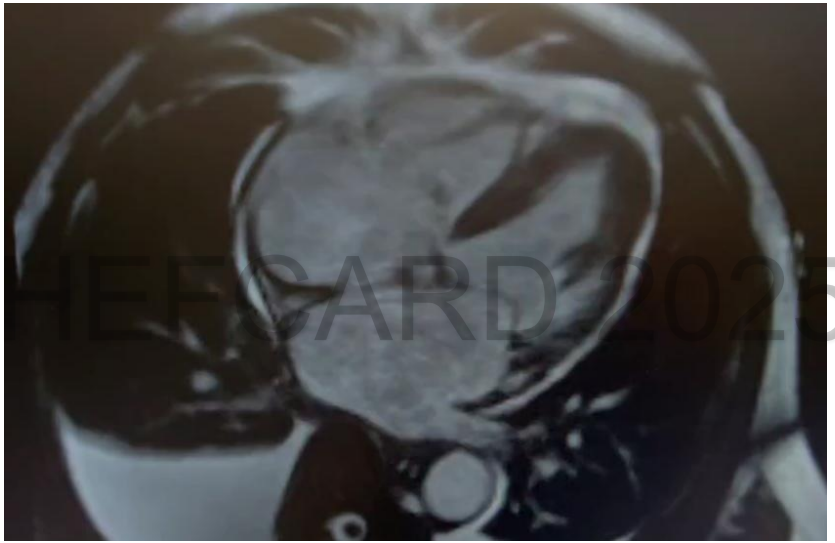
04

For oHCM in whom the anatomic mechanism of obstruction is inconclusive with echocardiography

Ommen, SR et al. Circulation. 2024;149:e1239–e1311.

Apical HCM



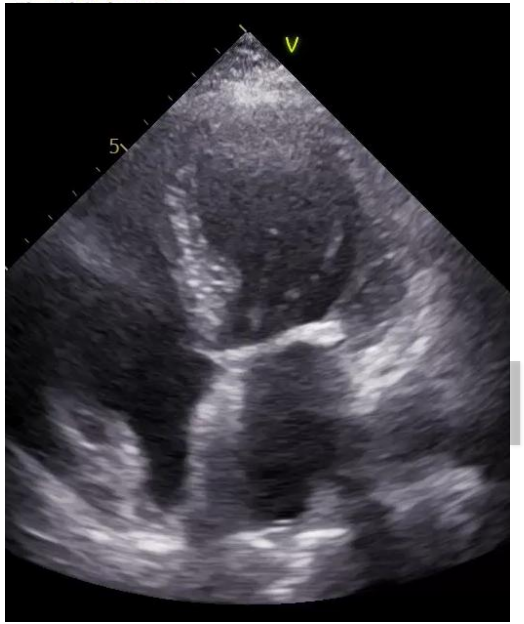


Hypertrophy of the apical myocardium, with the maximal wall thickness measuring up to 12 mm at the apical lateral wall (indexed wall thickness 6.9 mm/m2), which is greater than the threshold of 5.6 mm/m2 proposed by Hughes et al (JACC, 2024).

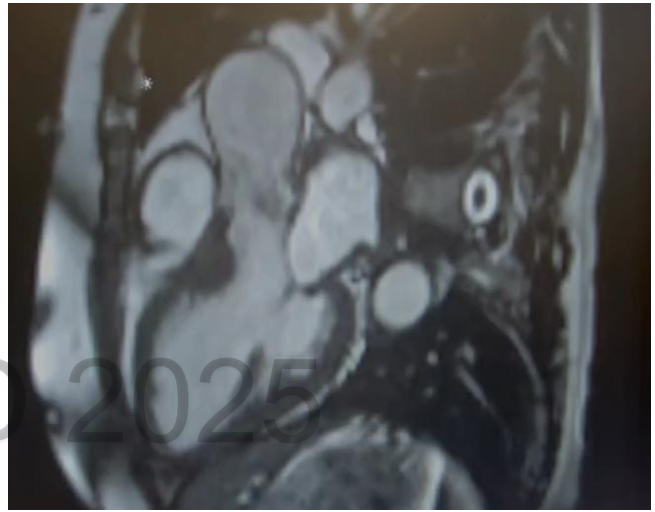
2

Focal LGE at the mid-cavity inferior RV insertion point is non-specific but may be related to underlying pulmonary hypertension.

oHCM



Kappa FLC (NUHS) 3.3 - 19.4 mg/L	658.0 ^
Lambda FLC (NUHS) 5.7 - 26.3 mg/L	14.4
Free Light Kappa to Lambda ratio (NUHS) 0.26 - 1.65	45.69 ^



- HCM with **asymmetric hypertrophy** of the basal anterior septum, anterior left ventricular myocardium and **LVOT obstruction**.
- **Minimal LGE** in the inferior right ventricular insertion point from the basal to mid-cavity level.
- **SAM of the anterior mitral leaflet** with mild mitral regurgitation.

4

Genetic Testing

🔍 Why Perform Genetic Testing?

- Confirms sarcomeric etiology in the proband
- Enables cascade screening of at-risk relatives
- Informs prognosis and risk stratification

👥 Who Should Be Tested?

- All patients with a clinical diagnosis of HCM
- Especially those with family history of HCM or SCD

🧬 Key Genes:

- MYH7, MYBPC3, TNNT2, TNNI3, ACTC1

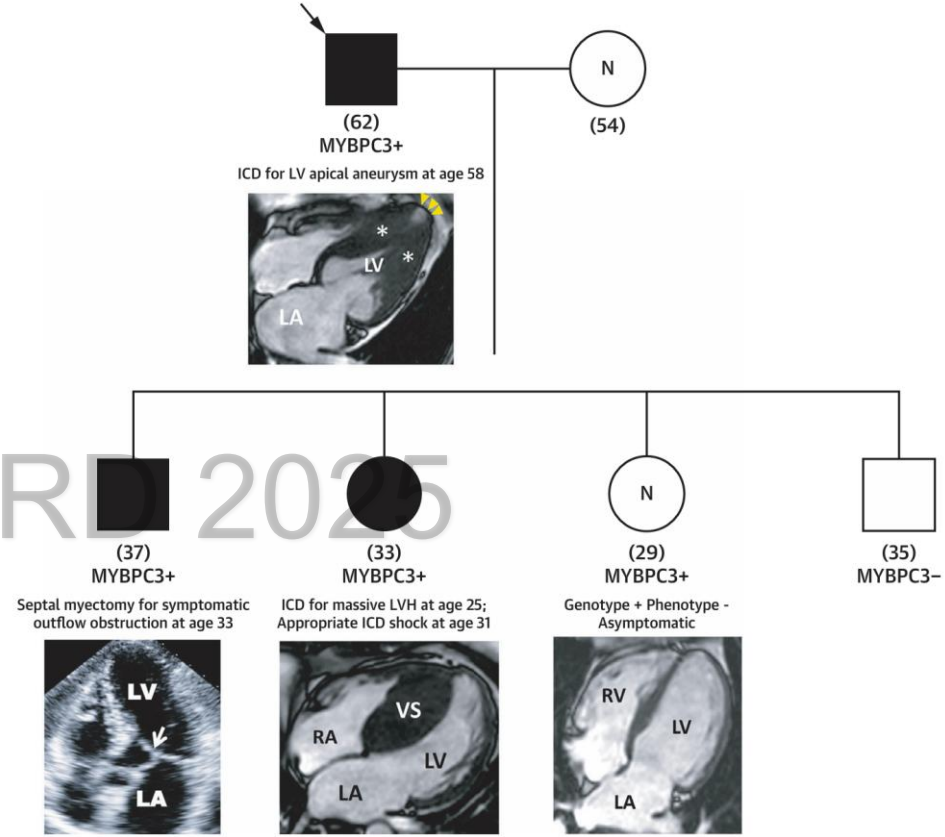
Family Screening

Family Screening Strategy:

- Genotype-positive, phenotype-negative relatives: regular follow-up + imaging every 1–3 years
- Children: Start screening by age 10–12
- Earlier if symptoms or family history of SCD

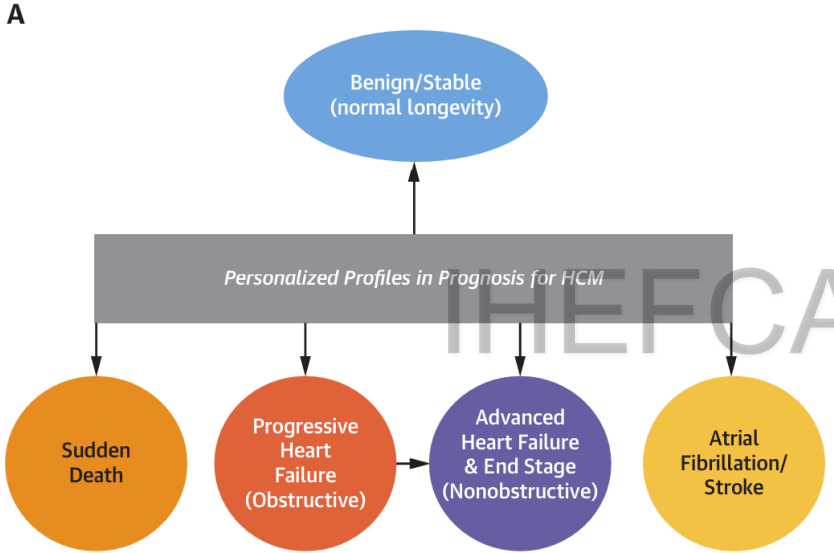
Benefits:

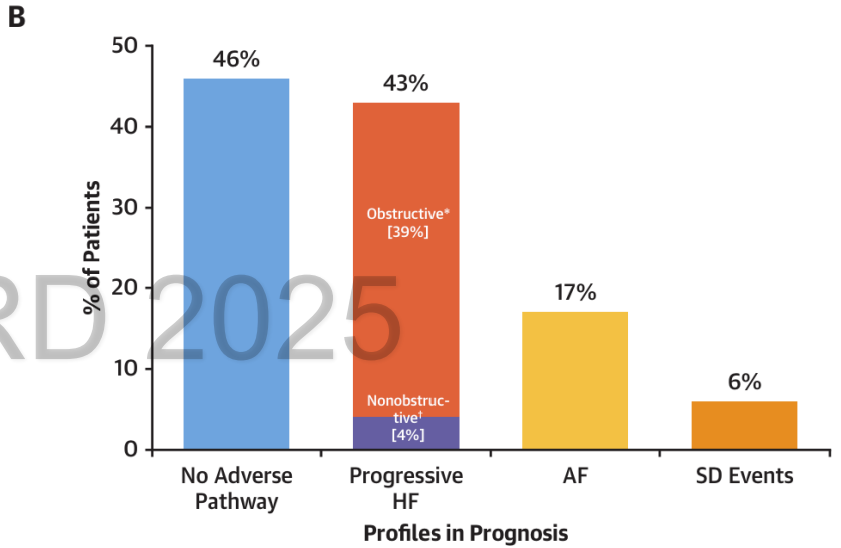
- Early detection before LVH appears
- Guides surveillance, lifestyle, and ICD decisions



Maron, B.J. et al. J Am Coll Cardiol. 2022;79(4):372–389

Prognostic Pathways





Individual patients may progress along 1 or more of these pathways, but along 2 or 3 pathways in only 10% patients.

Frequency of personalized HCM pathways in 1000-patient cohort

 Maron, B.J. et al. J Am Coll Cardiol. 2022;79(4):372–389

Risk for Sudden Cardiac Death

⚠️ Major Risk Markers for SCD:

- Family history of SCD (<50 years)
- Unexplained syncope (especially exertional)
- Max LV wall thickness ≥30 mm
- Apical aneurysm
- Extensive LGE on CMR (≥15% of left ventricular (LV) mass)
- NSVT on Holter
- LVOT obstruction ≥30 mmHg

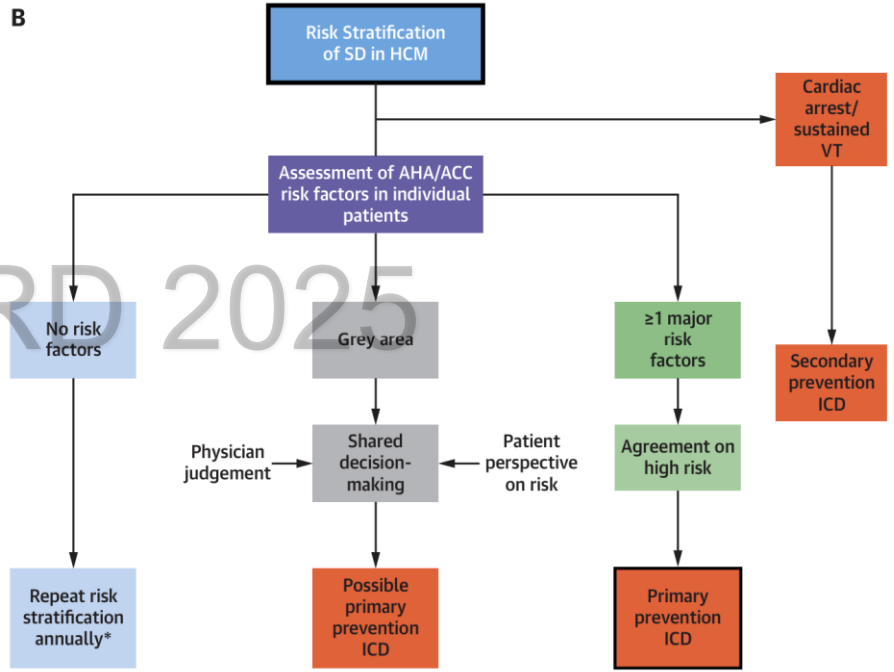
📈 Additional Risk Modifiers:

- Blunted BP response to exercise
- High-risk sarcomeric mutations
- Risk scores (e.g., HCM Risk-SCD calculator)

Ommen, SR et al. Circulation. 2024;149:e1239–e1311.

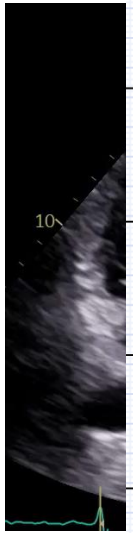
ICD Recommendation

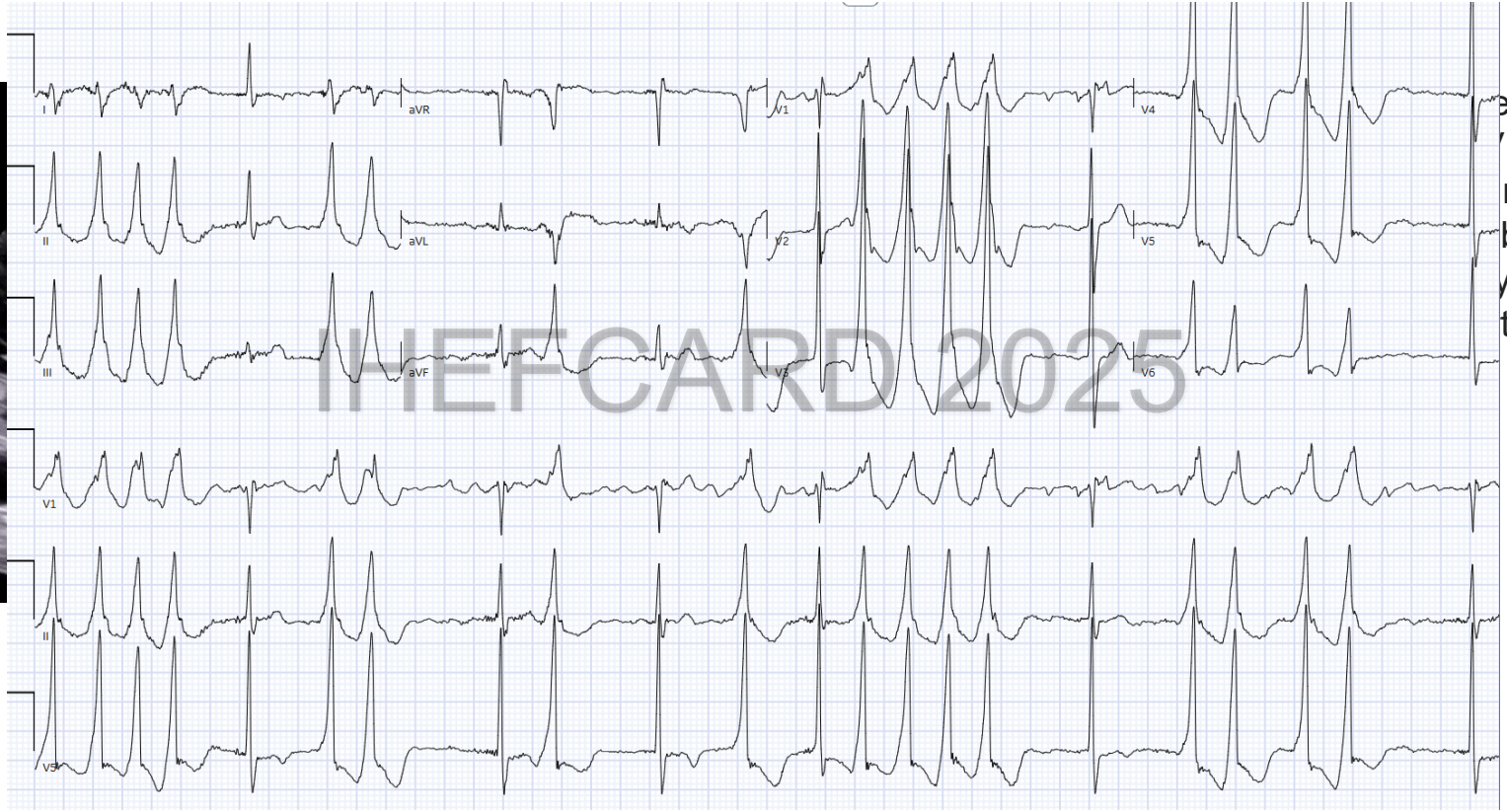
Class I	Prior cardiac arrest or sustained VT (secondary prevention)
Class IIa	One major risk factor
Class IIb	Multiple borderline risks or patient preference



Maron, B.J. et al. J Am Coll Cardiol. 2022;79(4):390–414.

Apical HCM: 74 yo, palpitation, near syncope





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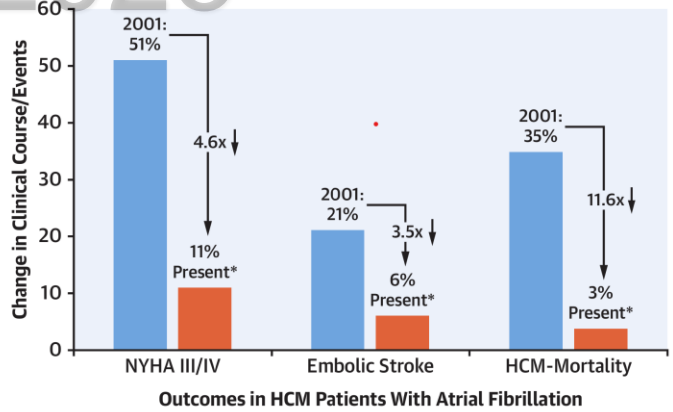
Management Guidelines for HCM

Benign/ Stable

Personalized Profiles of Prognosis in Hypertrophic Cardiomyopathy (HCM)

	Nonobstructive	*Provocable Obstruction*	*Rest Obstruction*
Proportion of Patients Who Evolved to NYHA Functional Class III/IV (red)	10% / 90% (NYHA I/II)	20% / 80% (NYHA I/II)	38% / 62% (NYHA I/II)
Rate of Progression to NYHA Functional Class III/IV (%/y)	1.6%/y	3.2%/y	7.4%/y
Treatment for Progression to NYHA Functional Class III/IV	Transplant (CRT)	Myectomy (ASA)	Myectomy (ASA)



Outcomes in HCM Patients With Atrial Fibrillation

Maron, B.J. et al. J Am Coll Cardiol. 2022;79(4):390–414.

Lifestyle & Monitoring in HCM

🧘 Lifestyle Recommendations

- Avoid dehydration, excessive alcohol, stimulants
- Encourage moderate exercise (e.g., brisk walking)
- Avoid high-intensity/competitive sports in high-risk patients
- Caution with heavy lifting (especially if obstructive)

👨‍⚕️ Routine Monitoring

- Annual cardiology follow-up
- Imaging (echo or CMR) every 1–3 years
- Assess wall thickness, obstruction, fibrosis (LGE)

🫀 Rhythm Monitoring

- ECG annually
- Holter or event monitor q1–2 years or if symptoms
- Consider implantable monitor in select cases

Cardiac Rehabilitation in HCM

Recommendation:	Supervised cardiac rehab is reasonable for selected patients Especially useful for deconditioned or post-procedure patients
Who Benefits:	Functional limitations or comorbidities (e.g., obesity, HF) Post-myectomy or sedentary lifestyle Need structured, safe exercise guidance
Precautions:	Avoid high-intensity or competitive training Customize programs to HCM-specific risk (e.g., SCD, obstruction)

Class IIa, Level of Evidence B

Ommen, SR et al. Circulation. 2024;149:e1239–e1311.

Conclusion

HCM is common and treatable

Early diagnosis and tailored management save lives

Genetic and molecular insights are transforming care

Multidisciplinary and personalized approach essential