Hypertrophic Cardiomyopathy

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Hypertrophic Cardiomyopathy Echocardiographic Diagnosis

Left Ventricular Hypertrophy > 15 mm
Left Ventricular Hypertrophy > 13 mm (+ FH)

↓

In absence of another cardiovascular or systemic disease associated with LVH or myocardial wall thickening

Gersh, BJ, et al. JACC 2011; 58: e212 ACC/AHA Guidelines

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Hypertrophic Cardiomyopathy Echocardiographic Diagnosis

NOT mandatory for diagnosis

Asymmetric Septal Hypertrophy (ASH)
Systolic Anterior Motion (SAM)
Dynamic LVOT obstruction

Gersh, BJ, et al. JACC 2011; 58: e212 ACC/AHA Guidelines

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Case

- 45 M with history of uncontrolled hypertension presented for evaluation
- Diagnosed hypertension at 20 years, did not take medicines
- One year ago, had paroxysmal atrial fibrillation and started medications

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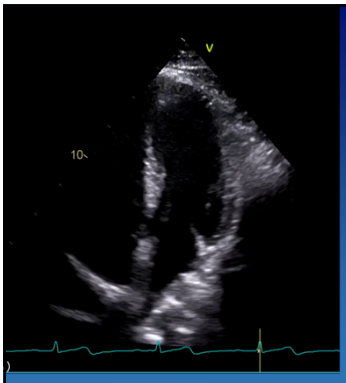

Wall thickness: 1.5-1.6 cm

SAM

LVOT gradient normal at rest Valsalva 56 mmHg

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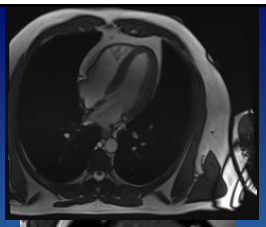

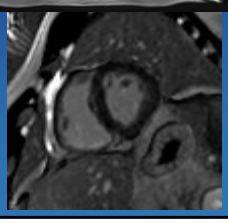

GLS – 15.7%

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What is the diagnosis?

1. Cardiac Amyloidosis
2. Hypertensive heart disease
3. Athlete's heart
4. Hypertrophic cardiomyopathy

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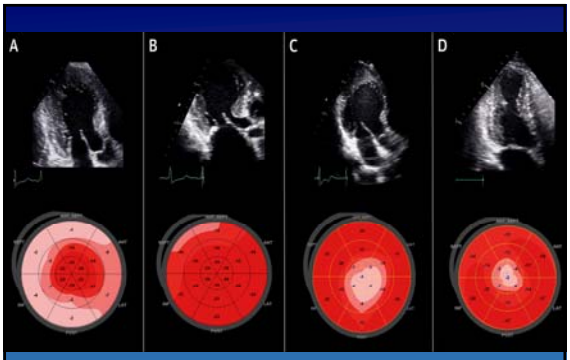

JACC 2017; 69 (8): Pages 1043-1056

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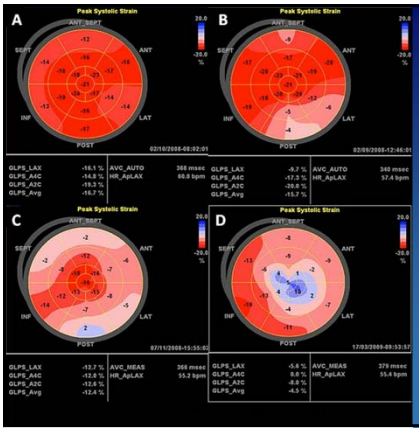

GLS
– 14.3% (sensitivity: 77% specificity:97 %,

- 11.5% Sensitivity 50-57% Specificity 99%

Alfonso et al. BMJ Open 2012;0:e001390

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Imaging data	HCM	Hypertensive heart disease
Echo, CMR, CCT		
LVH	Severe, asymmetric, IVS/PW >1.3 (1.5)	Moderate (<15 mm—except CRF and blacks), concentric, or mildly asymmetric IVS/PW <1.3 (1.5)
LVOTO	Frequent	Rare
'Sigmoid septum'	Rare	Frequent
Severe longitudinal systolic dysfunction	Frequent	Rare
Inhomogeneity (velocities and strain)	High	Low
Asynchrony (time intervals)	High	Low
CMR		
LGE	Frequent, RV insertion points, and intramural	Less frequent, non-subendocardial, no specific pattern

European Heart Journal - Cardiovascular Imaging, Volume 16, Issue 3, March 2015, Page 280

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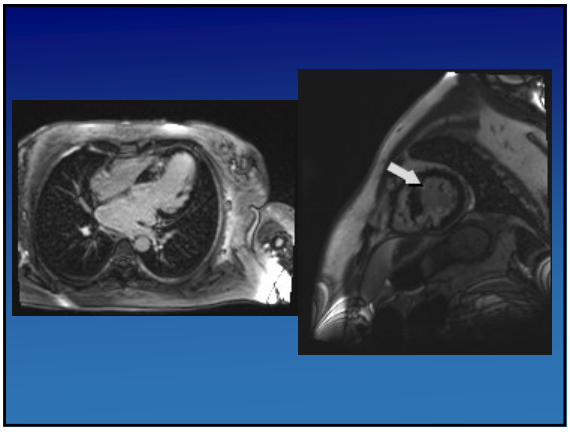

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You have diagnosed your patient with HCM. Your have consulted ACC and ESC guidelines and advised genetic testing. What is the yield of genetic testing in HCM?

A. 10%
B. 25%
C. 40%
D. 60-70%

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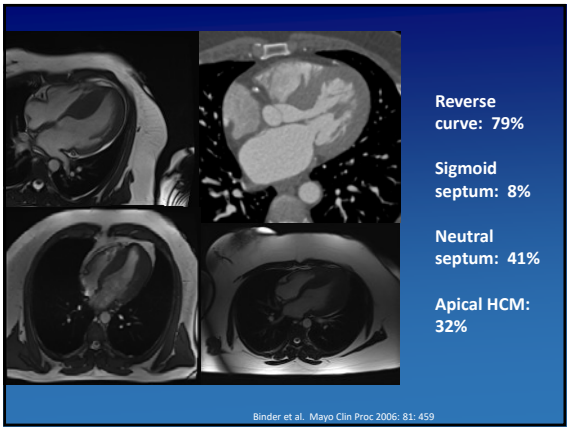

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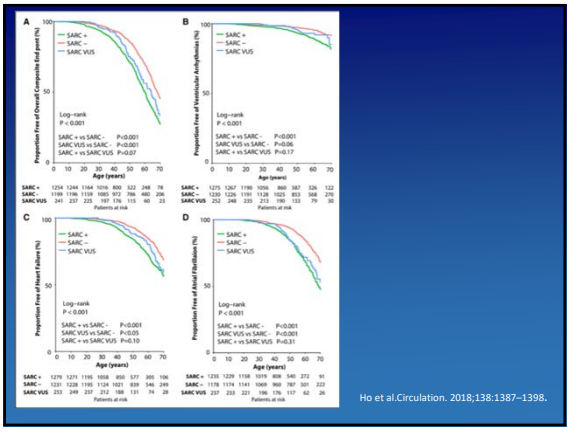

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Genetic testing in HCM

- Specific gene mutations are not associated with specific anatomic phenotypes
- Specific gene mutations not predictive of prognosis or sudden cardiac death

Van Driest SL, et al. Mayo Clin Proc 2005; 80: 739

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Family Screening of HCM by Echo

< 12 yrs old	Optional unless • Malignant family history • Cardiac symptoms • Competitive sports • Other signs or symptoms
12 to 18-21 yrs	Every 12-18 months
> 18-21 yrs old	• Every 5 years or as per clinical suspicion • More frequent if malignant course

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What is TRUE about sudden cardiac death in HCM?

1. Most of the SCD is related to vigorous activity
2. Older patients are at a higher risk of HCM related SCD than younger patients.
3. Specific gene mutations are associated with higher risk of SCD
4. None of the above are true
5. All of above are true

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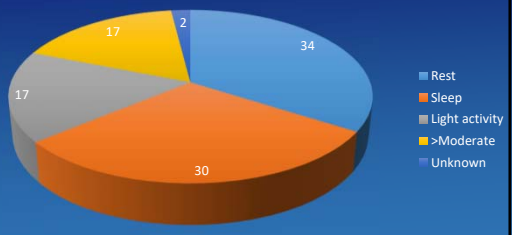
Most of the HCM deaths are not related to vigorous exercise



Weisler-Sneer et al. Circulation. 2019;140:1706–1716.

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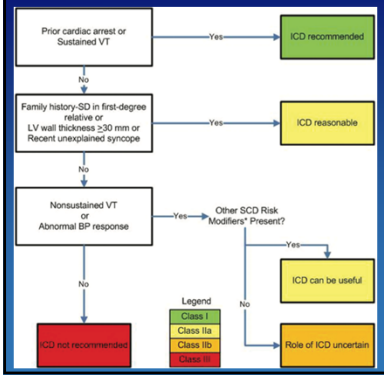
Recommendations for ICD use



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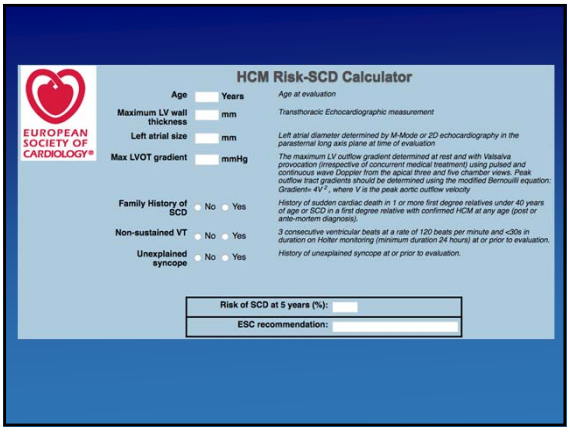

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Enhanced ACC/AHA risk score

- FH of SCD
- Thickness > 30 mm
- Unexplained syncope
- NSVT
- BP response
- LGE >15% of myocardium
- LV apical aneurysms
- End stage systolic HF, symptomatic

Maron et al. JAMA Cardiol. 2019;4(7):644-657

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	ACC/AHA	ESC high risk
Sensitivity of preventing SCD	95%	34%
Specificity	78%	92%
PPV	16%	12%
NPV	99%	97%
Number needed to treat	6.6	7.2

Maron et al. JAMA Cardiol. 2019;4(7):644-657

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Risk Stratification is imperfect!

A shared decision discussion often needed:

- Features that are associated with SCD
- Magnitude of the risk
- Risk/benefits of the ICD
- Allows patient to express their risk tolerance

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Therapeutic options

Obstructive

- Medical management: Betablocker / Diltiazem+ Disopyramide
- Septal ablation/ Septal myectomy

Non-obstructive: Diastolic Heart Failure

- Medical management
- MCS/Transplant

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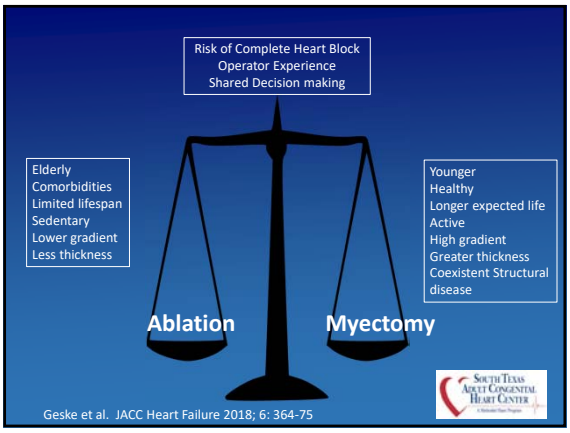
Risk of Complete Heart Block
Operator Experience
Shared Decision making
Elderly
Comorbidities
Limited lifespan
Sedentary
Lower gradient
Less thickness
Younger
Healthy
Longer expected life
Active
High gradient
Greater thickness
Coexistent Structural disease
Ablation
Myectomy
Geske et al. JACC Heart Failure 2018; 6: 364-75

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