Hypertrophic Cardiomyopathy
When and How to Follow?

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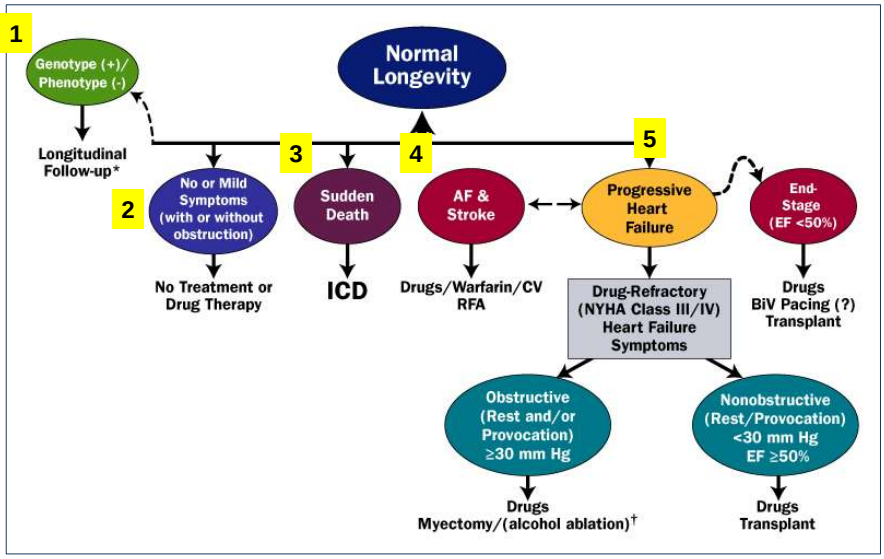
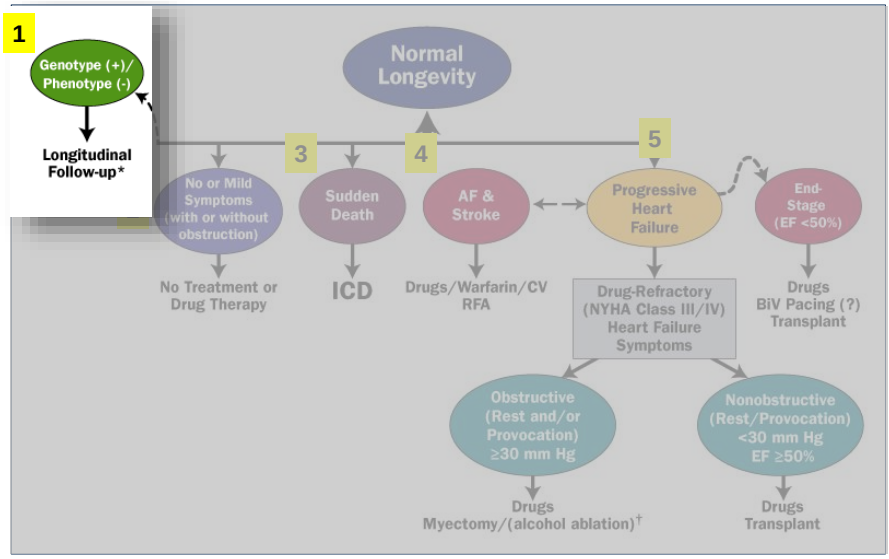
HCM: natural history



HCM: natural history



Preclinical HCM

1. After Puberty 10-20 y : 1-2 y

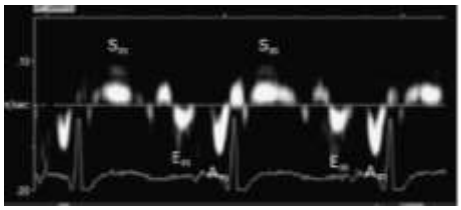
2. After Adolescent > 20 y: 2-5 y

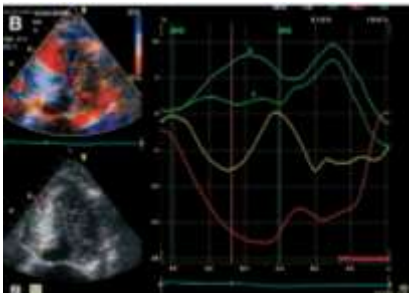
3. Before Puberty < 10 y

Malignant FH ; FH of early disease onset; SX; intense sports

- **Clinical assessment + ECG + Echocardiography**

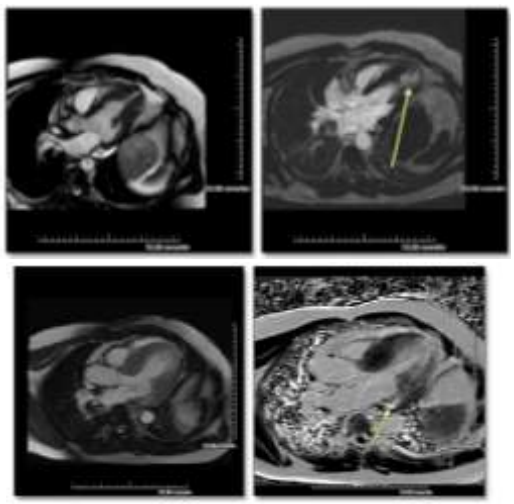
Preclinical HCM



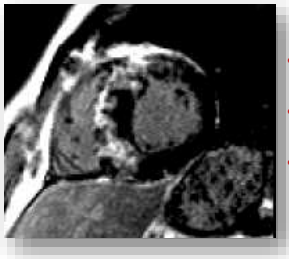


CMR: Diagnosis

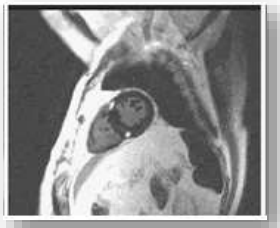
- Establish diagnosis (1)?? Echo ; (2) apical HCM ; (3) anterolateral LVH
- Rule out the others



CMR: Fibrosis



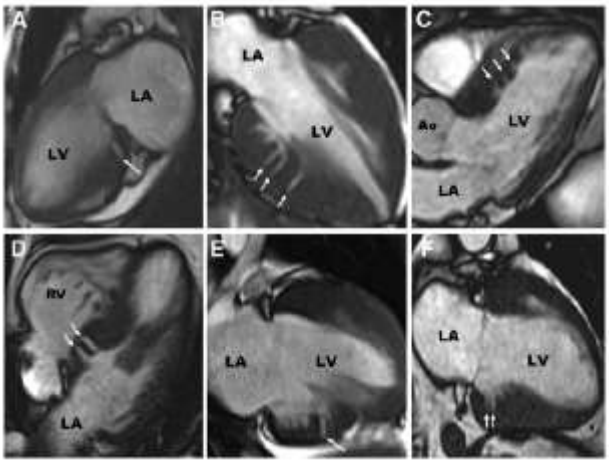
- Patchy distribution & Correlates with WT
- Subendocardium is not necessarily affected (unlike IHD)
- LGE typically more diffuse ; not specific to mid-wall (unlike DCM)



RV insertion point enhancement

CMR: Diagnosis

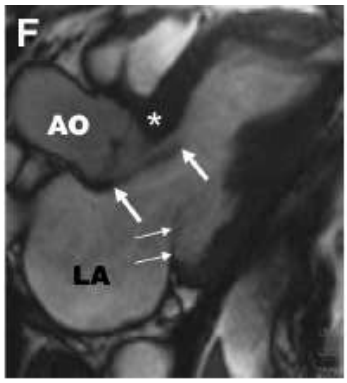
Myocardial crypts



Circ Cardiovasc Imaging. 2012;5:441-447

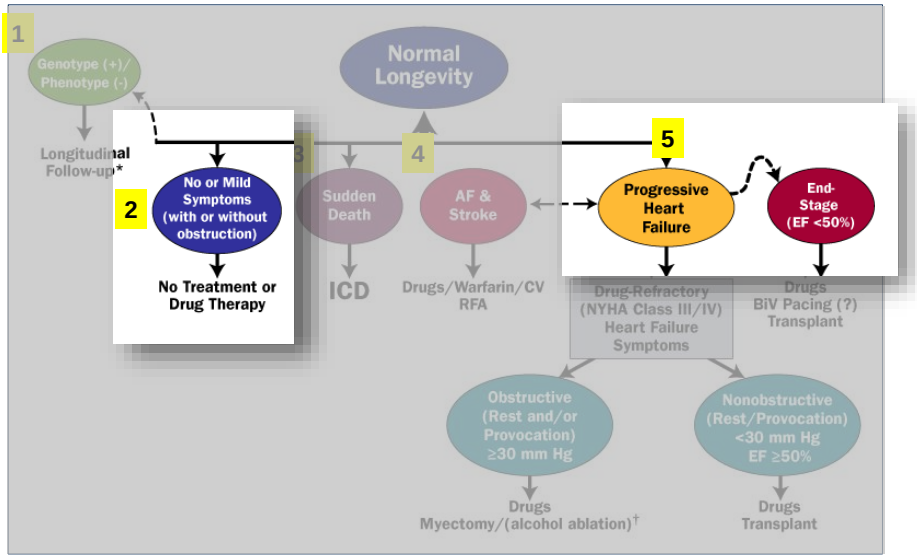
CMR: Diagnosis

Elongated MV Leaflets



Circulation. 2011;124:40-47.

HCM: natural history



Evaluation - Investigations

	Baseline	FU
ECG	✓ (75 - > 90%)	○ Every 1-2 year ○ Worsening of SX
TTE	✓	○ Every 1-2 year ○ Change in clinical status
24-48-Holter	✓	○ Every 1-2 year ○ Every 6-12 month (NSR + LA > 4.5 cm) ○ SX
Treadmill exercise	○ Functional capacity ○ Risk stratification ○ + echo: > 30 - > 50 mmHg	○ Every 2-3 years ○ Progressive SX
C. Cath	○ Not needed (rarely in cases with discrepancy: TTE / clinical)	
SPECT	○ Reversible defects (50%) ○ Fixed defects (scar) ○ False defects in non-hypertrophied segments	

SOB

- LVOTO ; MR ; diastolic dysfunction ; ischemia; DCM
- Mitral flow Doppler : little value
- E/E' ratio: not reliable
- BNP: little value

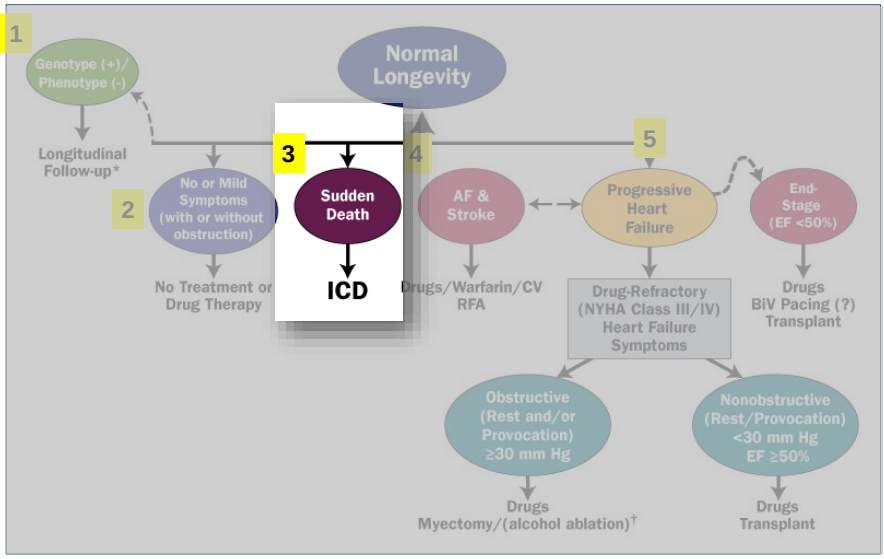
Counseling

- **Pregnancy:**
  - Generally safe; Class III: Severe SX LVOTO / Severe systolic dysfunction
  - Risk : > 50 mmHg OR symptoms (referred to high risk obstetrician
  - Issue of counseling
- **Exercise:**

  a. Low-intensity aerobic exercise (healthy lifestyle)

  b. avoiding strenuous activity or competitive athletics
- **Medications:** VDs; + diuretics ; digoxin ; NE; (use: phenylaphrine)
- **Occupations:** # : commercial motor vehicle driver ; pilot

HCM: natural history

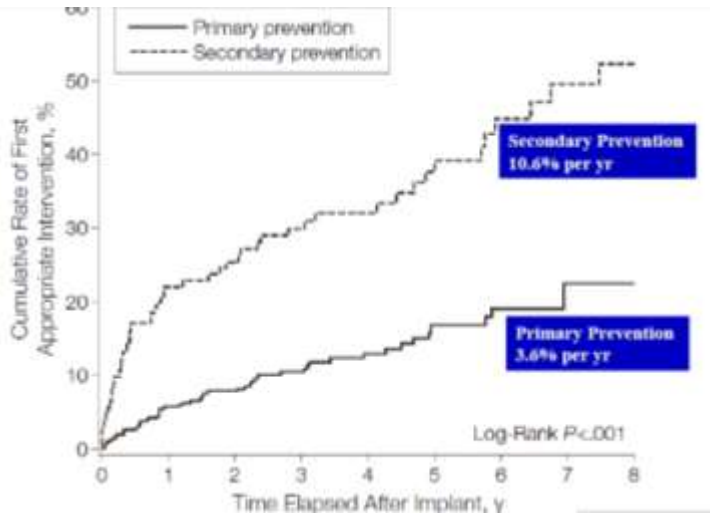


SCD in HCM: Challenges

- Young patients

- Without warnings
- Asymptomatic, mildly symptomatic
- Most occur during mild exertion or rest.

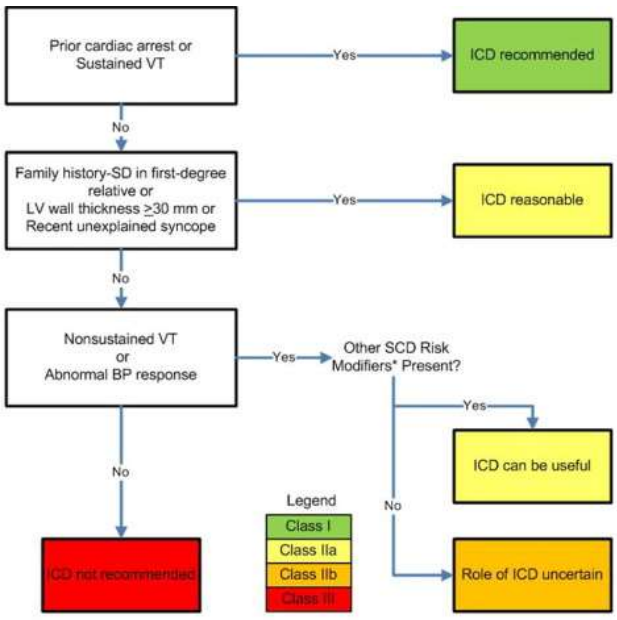
SCD in HCM: Importance



Maron BJ, et al., JAMA 2007;298:405

RFs of SCD in HCM

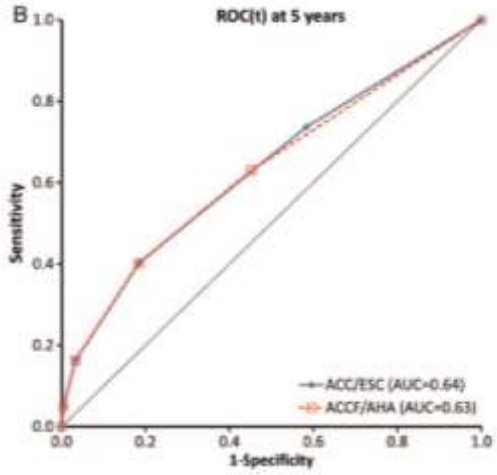
- Prior cardiac arrest (VF)
- Spontaneous sustained VT
- Family history of SCD
- Unexplained syncope
- **LV thickness ≥30 mm**
- Abnormal exercise BP
- NSVT
- **LV outflow obstruction**
- **Apical aneurysm**
- **Late gadolinium enhancement**
- High risk mutations
- AF
- Myocardial ischaemia
- End-stage phase
- ASA
- Competitive physical exercise
- Paced ventricular electrogram fractionation
- Surface ECG score
- **Diastolic dysfunction**
- **Myocardial bridge**



SCD risk modifiers
1. LVOTO
2. LGE on CMR
3. Genetic mutations
4. Apical aneurysm

ACCF/AHA Hypertrophic Cardiomyopathy Guidelines
Circulation 2011;58.e212

A validation study of the 2003 American College of Cardiology/European Society of Cardiology and 2011 American College of Cardiology Foundation/American Heart Association risk stratification and treatment algorithms for sudden cardiac death in patients with hypertrophic cardiomyopathy



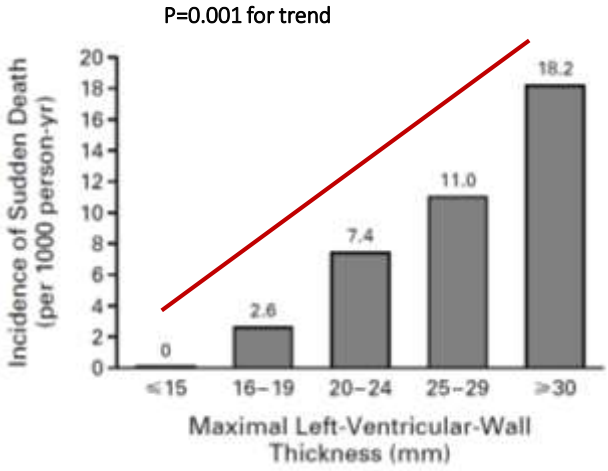
PPV = 9.8% @ 1 year
PPV = 10.5% @ 5 year

NPV = 94.1% @ 1 year
NPV = 94.6% @ 5 year

Constantinos O'Mahony Heart
Published Online First February, 2013

LV wall thickness and SCD

Maximal LV Wall Thickness and the Risk of SCD in 480 Patients



Paolo Spirito. N Engl J Med 2000;342:1778-85

Relation between severity of left-ventricular hypertrophy and prognosis in patients with hypertrophic cardiomyopathy

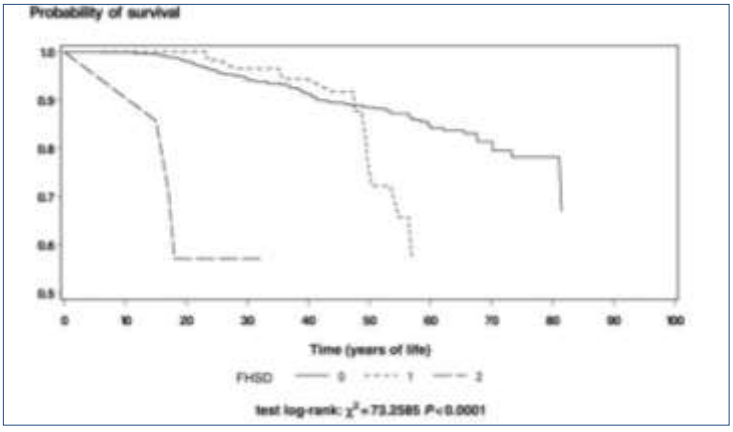
Findings 39 patients died suddenly or had an appropriate ICD discharge; nine died from progressive heart failure; 11 from other cardiovascular causes and 23 from non-cardiac causes. There was a trend towards higher probability of sudden death or ICD discharge with increasing wall thickness (p=0·029). Patients with wall thickness of 30 mm or more had higher probability of sudden death or ICD discharge than patients with

75% of those who died suddenly had a maximum wall thickness < 30 mm

Interpretation The risk of sudden death associated with a wall thickness of 30 mm or more in patients without other risk factors is insufficient to justify aggressive prophylactic therapy. Most sudden deaths occurred in patients with wall thickness less than 30 mm, so the presence of mild hypertrophy cannot be used to reassure patients that they are at low risk.

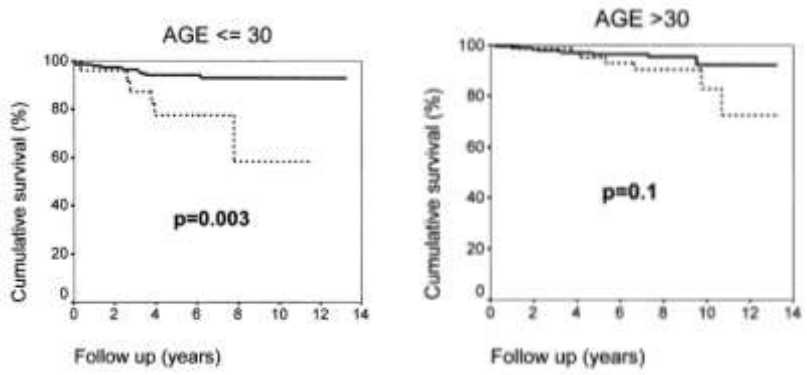
Perry M Elliott, Lancet 2001; 357: 420–24

FH and SCD



Paweł Petkow Dimitrow. European Heart Journal (2010) 31, 3084–3093

NSVT and SCD



RR: **4.35** (95% CI: 1.54 to 12.28; p 0.006

Lorenzo Monserrat, J Am Coll Cardiol 2003;42:87

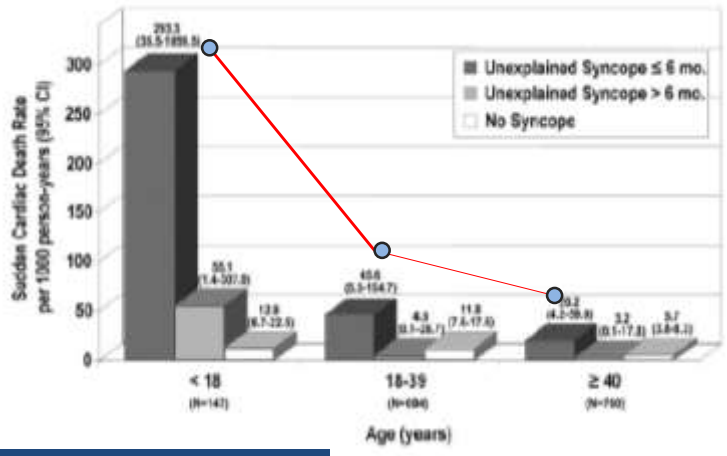
Syncope and SCD

Table 4. Multivariable Analysis of the Prognostic Importance of the Time Interval Between Unexplained Syncope and Initial Patient Evaluation at the Participating Institutions

Time between unexplained syncope and first patient evaluation			0.006
Without unexplained syncope‡	1349	1 (Reference)	
≤6 mo	53	4.89 (2.19–10.94)	
>6 to 12 mo	16	0 (No events)	—
>1 to ≤2 y	13	2.01 (0.27–14.80)	
>2 to 5 y	19	1.04 (0.14–7.57)	
>5 y	50	0.38 (0.05–2.74)	

Paolo Spirito, Circulation. 2009;119:1703-1710

Syncope and SCD



< 18 y: HR:8.01, 95% CI:2.07-31.45

Paolo Spirito, Circulation. 2009;119:1703-1710

SCD in patients ≥ 60 years

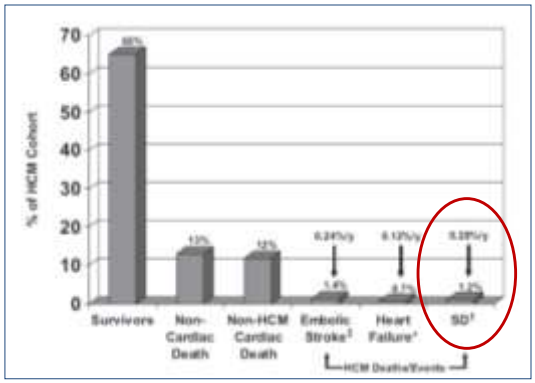
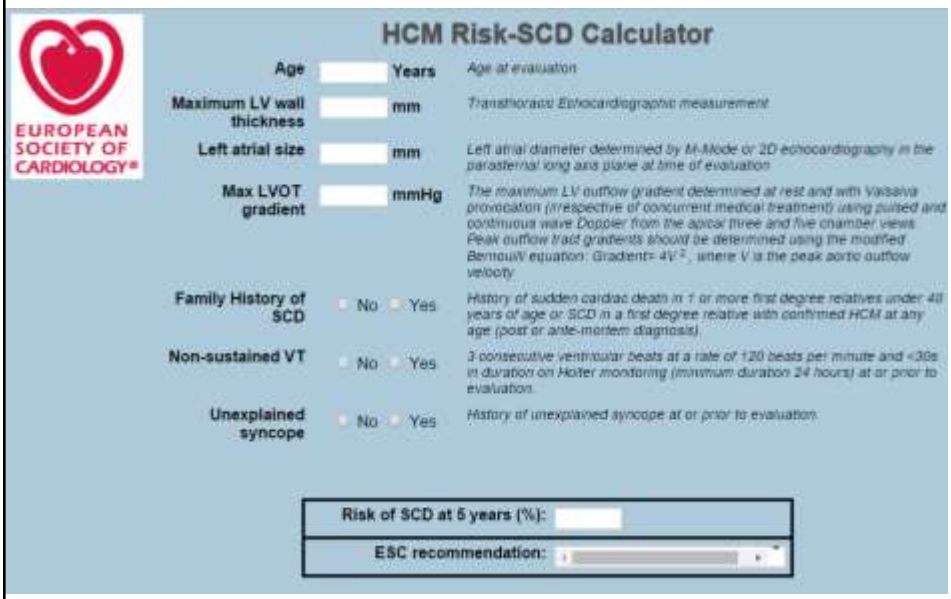


Table 3. Markers of Increased Sudden Death Risk

Patient Subset	Patients, n	Risk Factors, n ≥1	1	2	>3
All patients	428	211	149	49	13
Survivors	279	135	96	33	6
All HCM deaths/events¶	16#	8	6	2	0
HCM SD/events**	5	3	2	1	0

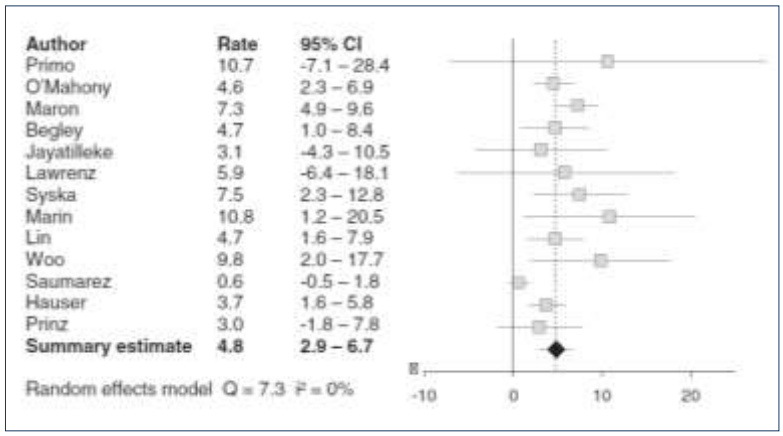
Conclusions—HCM patients surviving into the seventh decade of life are at low risk for disease-related morbidity/mortality, including sudden death, even with conventional risk factors. These data do not support aggressive prophylactic defibrillator implantation at advanced ages in HCM. Other cardiac or noncardiac comorbidities have a greater impact on survival than HCM in older patients. (*Circulation*. 2013;127:585-593.)

Barry J. Maron, Circulation. 2013;127:585-5



http://doc2do.com/hcm/webHCM.html

ICD in HCM: Complications



2190 HCM patients with ICD for 1^0 or 2^0 prevention (FU: 3.7 year)

Arend F.L. Schinkel. Circ Heart Fail. 2012;5:552-559

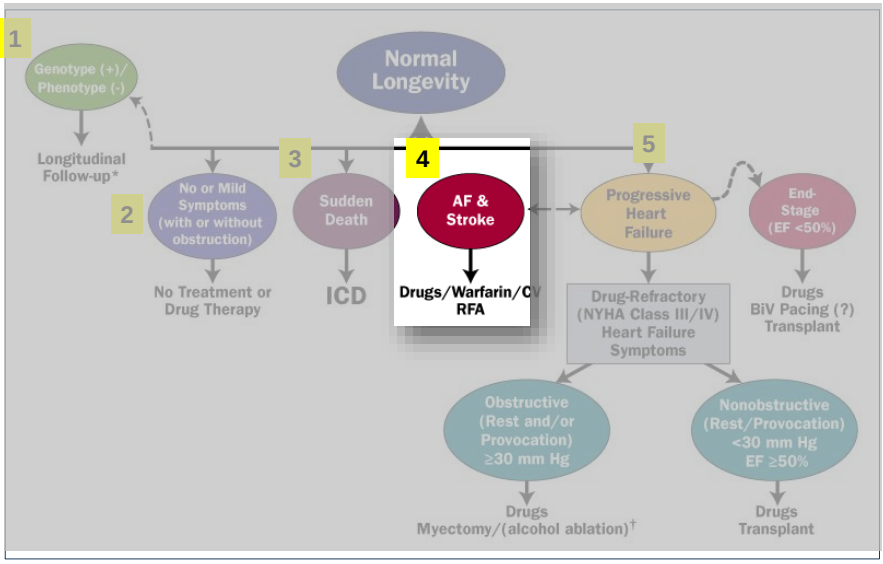
ICD in HCM: Complications

Table 2. Summary of Clinical Outcome

Cohort	Author	Year	Follow-Up, y	Appropriate Intervention, %	Inappropriate Intervention, %	Complications, %				
						Lead Malfunction	Infection	Lead Displacement	Psychological	Any
Event rate				13.7	19.0	4.2	3.1	2.7	3.8	14.9
(95% CI)				(9.9–17.5)	(12.6–25.4)	(4.1–8.3)	(1.2–5.0)	(1.6–3.9)	(0.5–7.1)	(9.9–19.9)
Annualized event rate (95% CI)				3.3 (2.2–4.4)	4.8 (2.9–6.7)	1.5 (0.9–2.1)	0.8 (0.1–1.6)	1.0 (0.5–1.4)	0.8 (−0.8 to 2.3)	3.4 (2.5–4.3)

Arend F.L. Schinkel. Circ Heart Fail. 2012;5:552-559

HCM: natural history



AF

- Common: 25%
- Clinical deterioration
- Thromboembolism ; stroke
- CHADVASc: no value
- OAC

AF: predictors in HCM`

Table 3 Univariable analysis for predictors of atrial fibrillation in hypertrophic cardiomyopathy

Univariable analysis			
Predictor			
Female			
Age (10 years increme			
NYHA II			
NYHA III,IV			
LA (5 mm increment)			
MWT (mm)			
FS (%)			
LVOT max (mm Hg)			
LVEDD (mm)			
LVESD (mm)			
Hypertension			
Diabetes			
Vascular disease			
Analyses with non-lin			
MWT (mm)			
MWT^2			
LA5	3.11	1.98 to 4.87	<0.001
LA5^2	0.97	0.94 to 0.99	<0.001

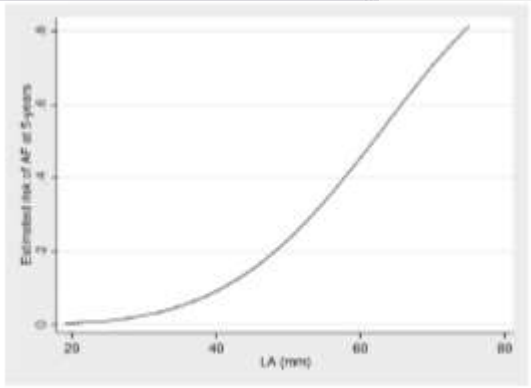
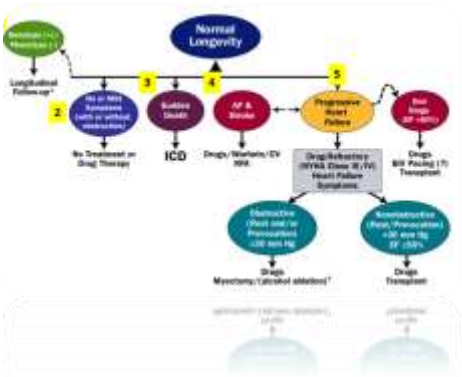


Figure 1 Graph showing the relationship of left atrial (LA) size with risk of atrial fibrillation (AF).

Heart 2017;**103:672–** 678.

HCM Team`

- Cardiologists
- Interventional cardiologists
- EP cardiologists
- Radiologists
- Cardiac surgeons
- Genetic counselors
- Clinical pharmacists
- Nurses



Thank You