SCD Prevention in HCM, Recent Guidelines and Decision-Making

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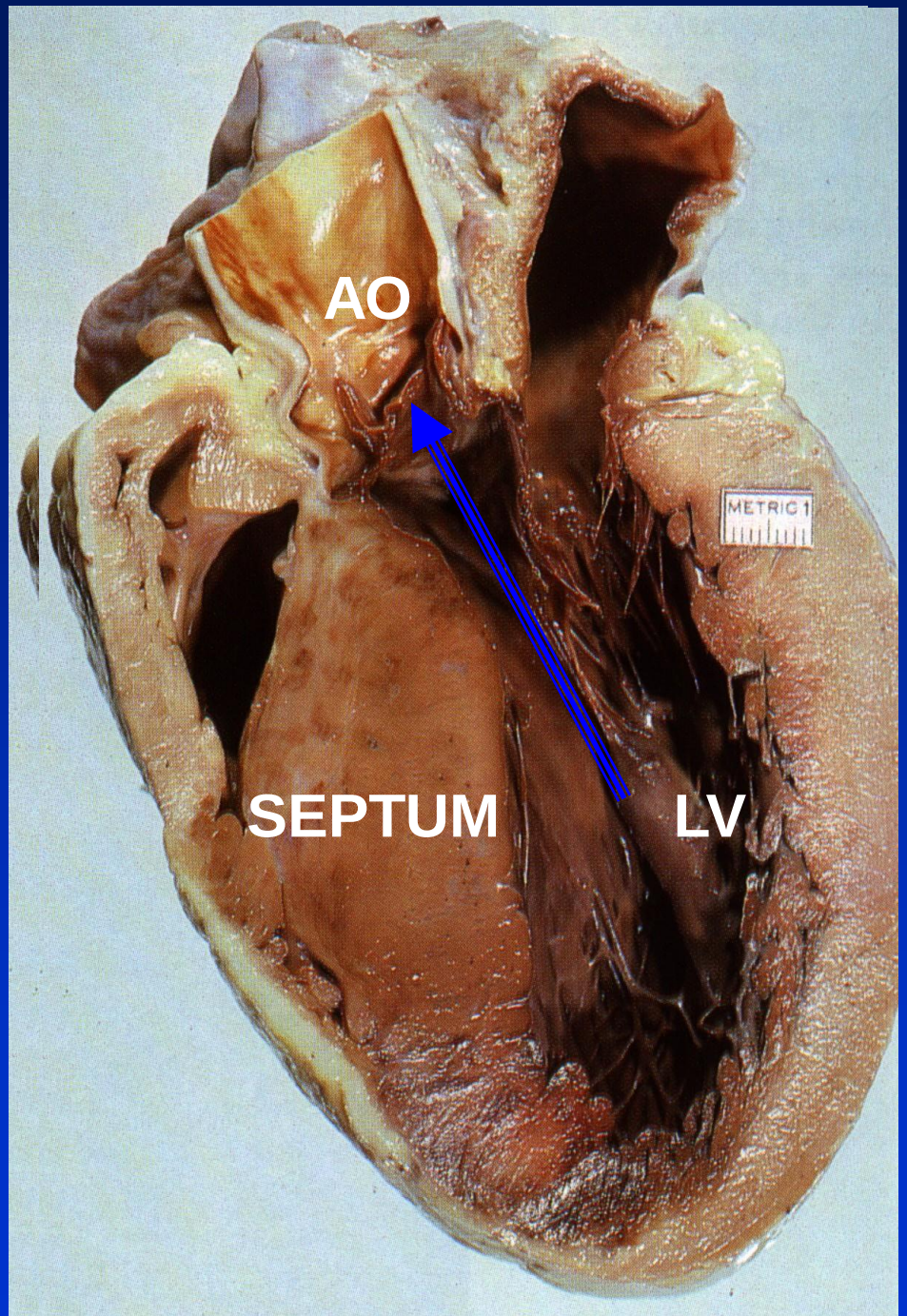
30.6.2022

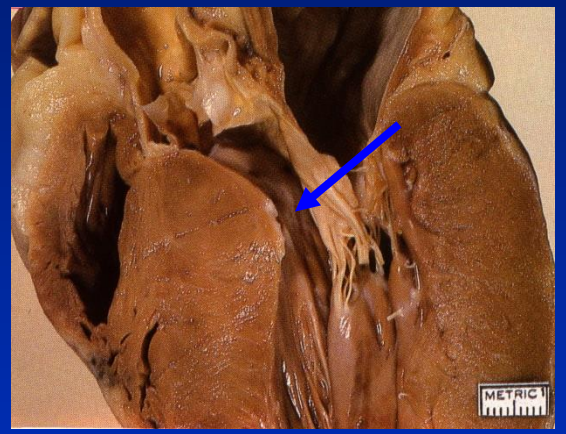
Potential Conflicts of Interest

No conflicts of interest related to this lecture

Lecturing / Consulting: Novartis, Bayer, Vifor-Pharma, CTP, Pfizer, Boehringer Ingelheim, BMS



HCM

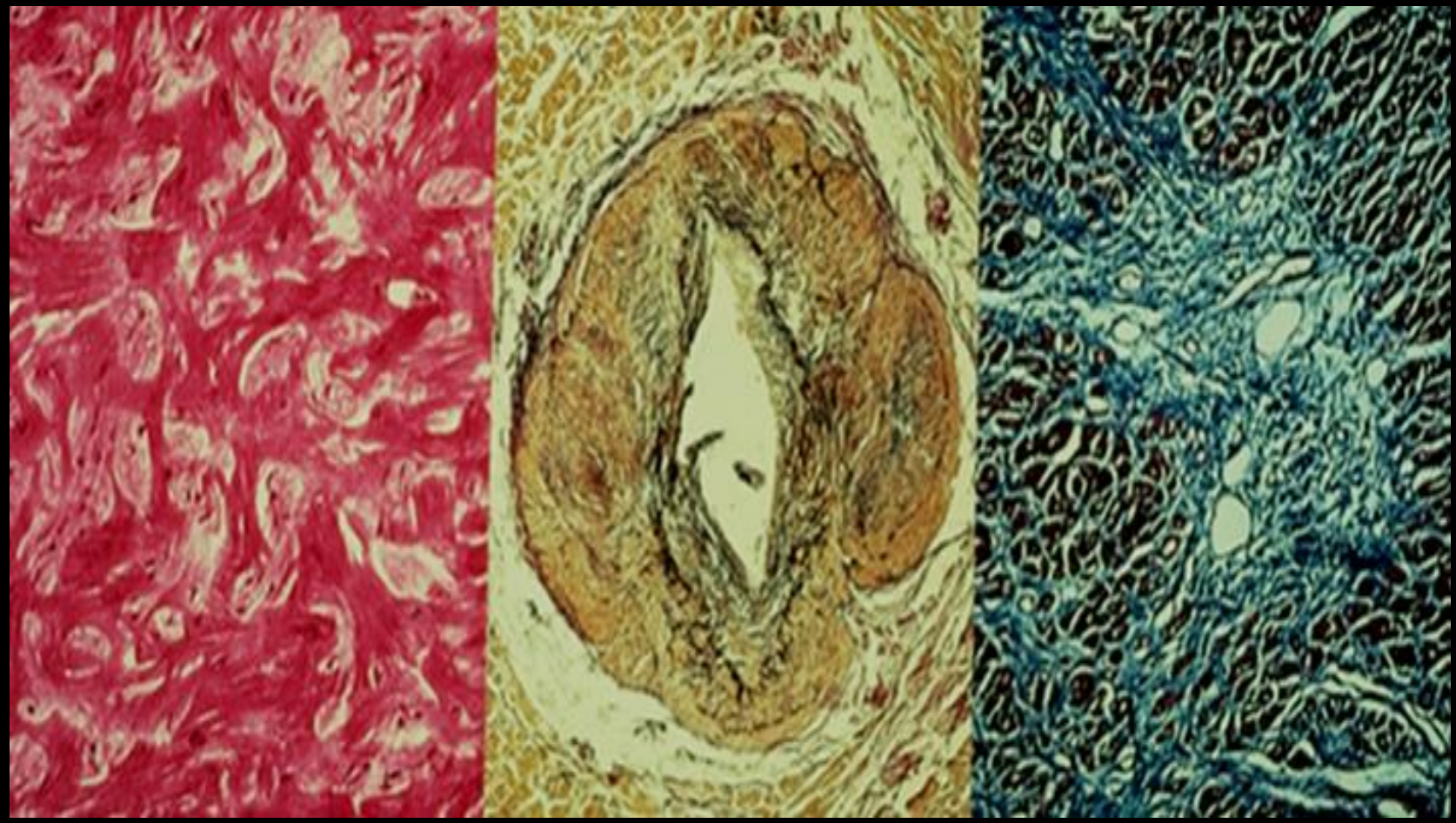


Sudden cardiac death in HCM

- Rare, but devastating event for the family, community

Courtesy of Maron BJ

Arrhythmogenic Myocardial Substrate in HCM

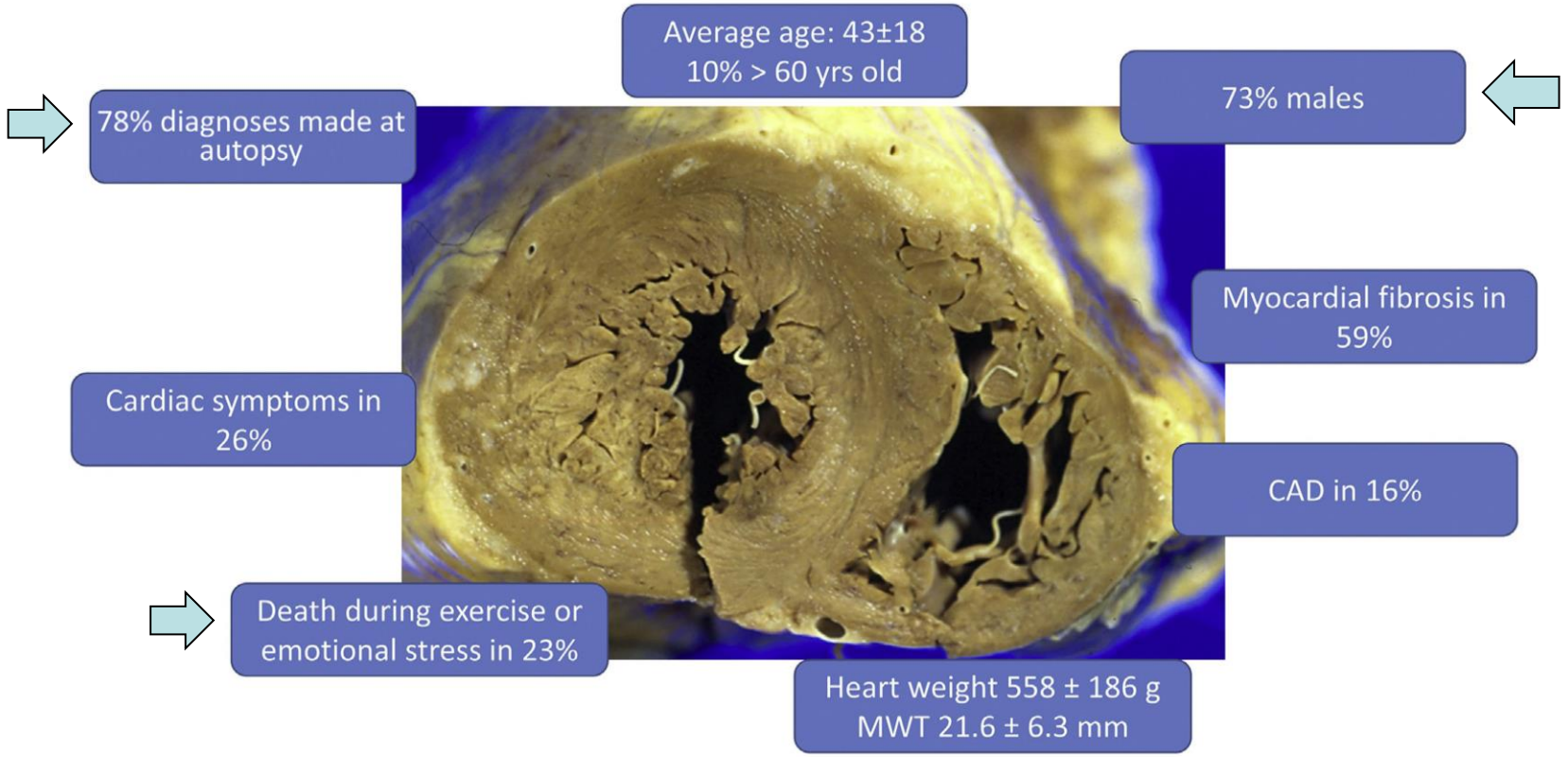


Courtesy of Maron BJ

Changing concepts on SCD in HCM

Sudden Cardiac Death in HCM

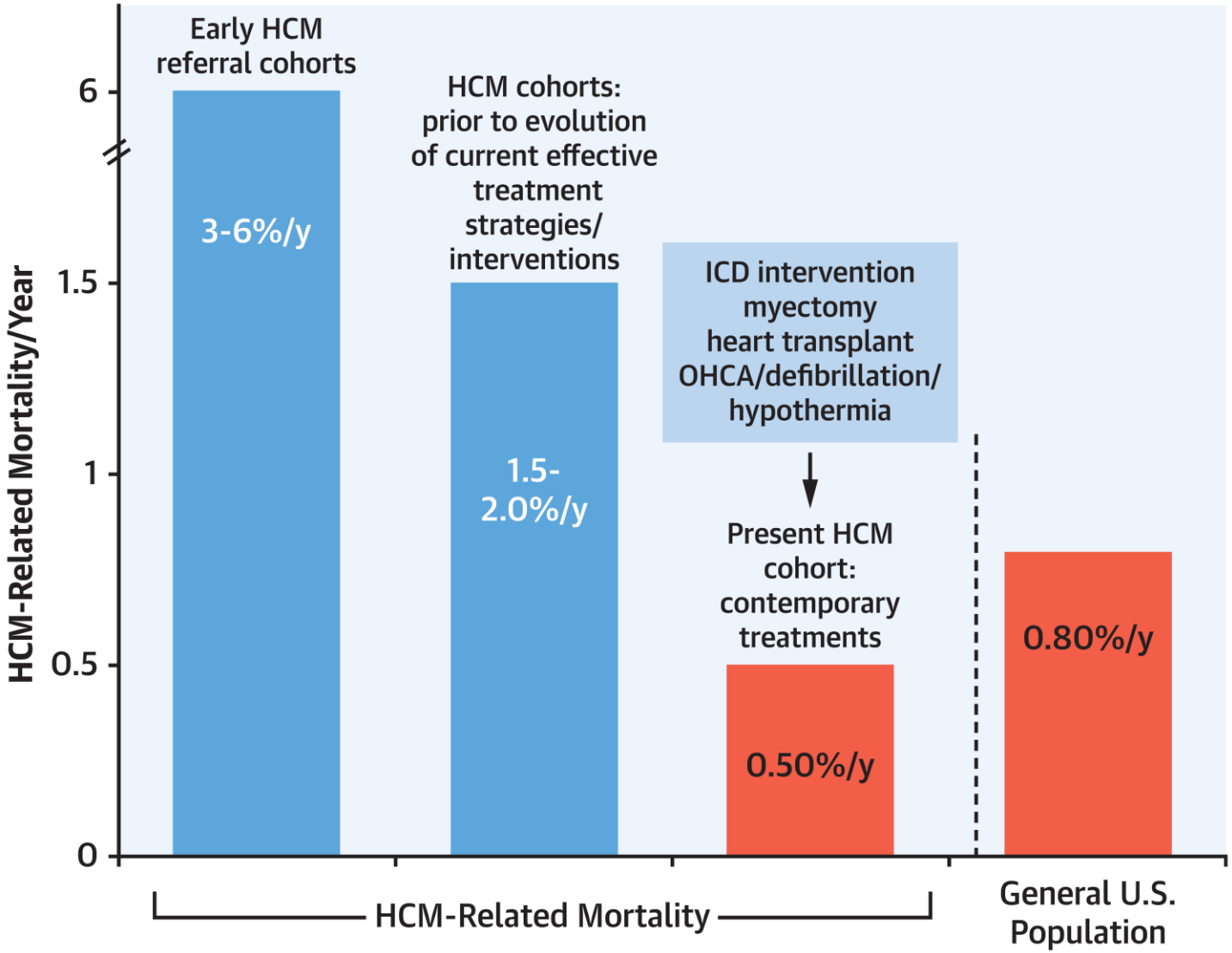
- The most prevalent inherited heart disease, 1:200-1:500 population
- SCD can be the first manifestation of the disease



Finocchiaro G, JACC Clin Electrophysiol 2019;5:252-4; UK autopsy registry of 194 pts with HCM&SCD

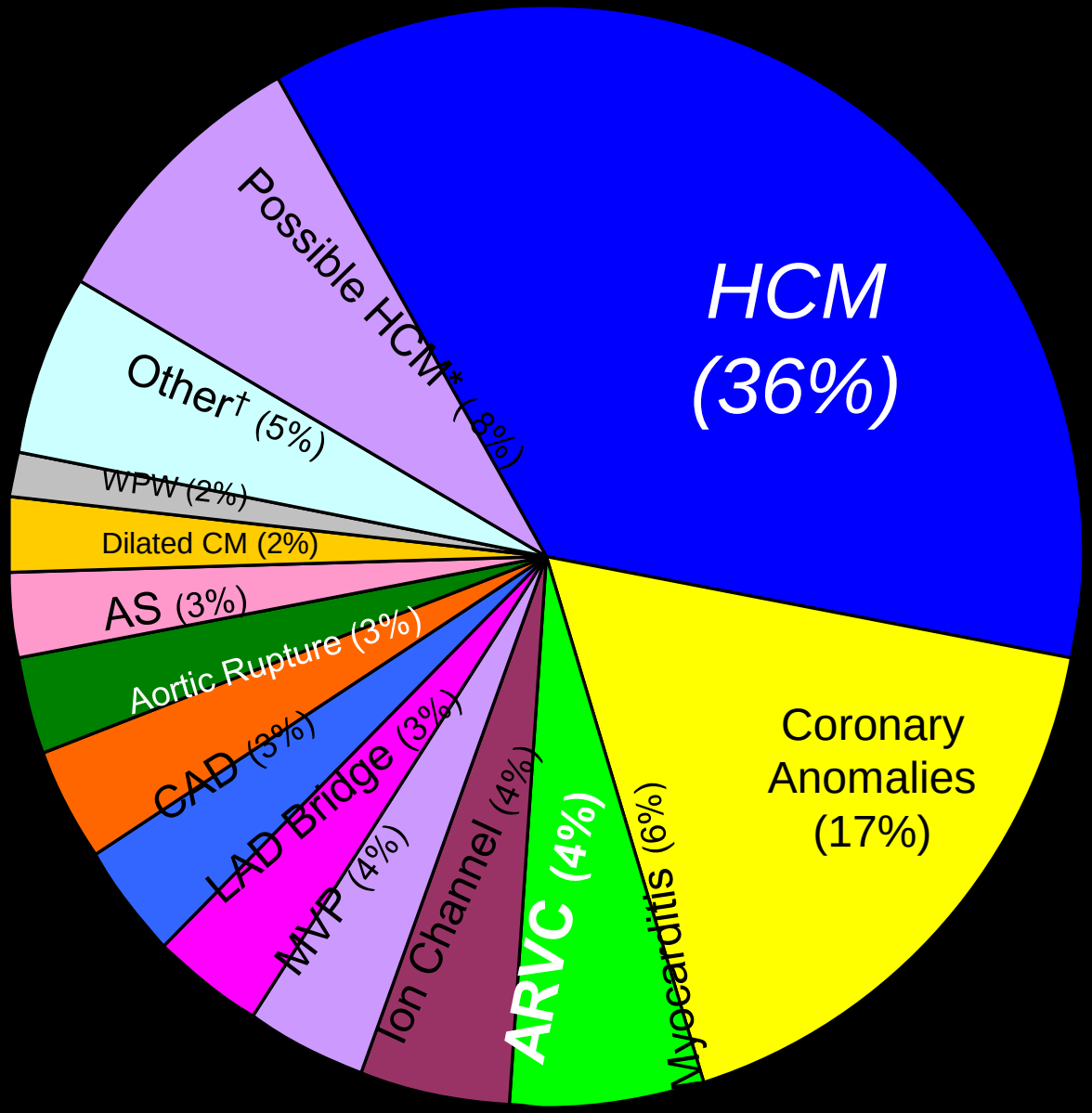
McKenna WJ, Nat Rev Cardiol 2021;18:22-36; Elliott PM, 2014;35:2733-2779; Ommen SR, J Am Coll Cardiol. 2020;76(25):e159-e240. Maron BJ, J Am Coll Cardiol. 2022;79(4):390–414.

Decrease in SCD rates over 60 years



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

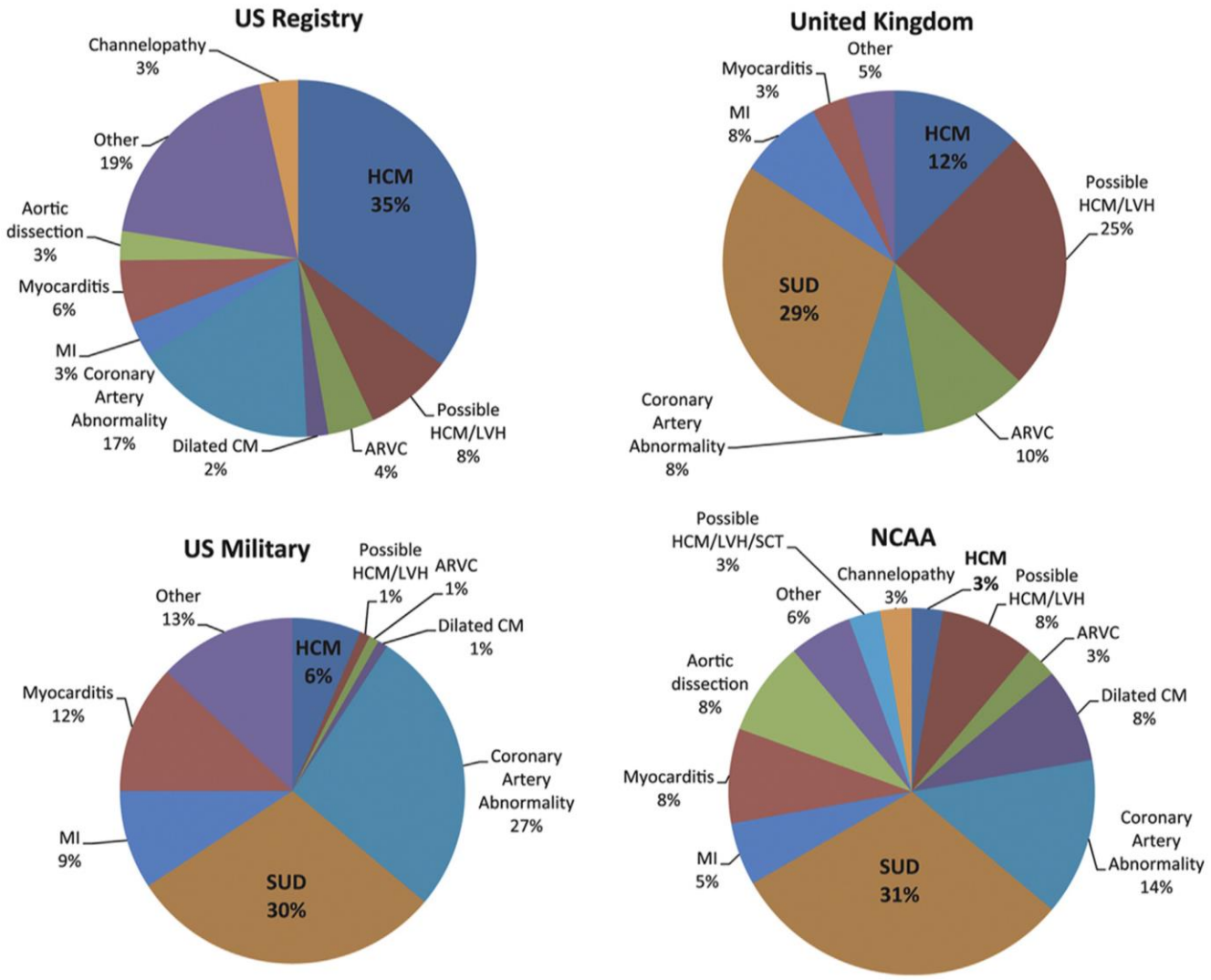
HCM – the leading cause of SCD in athletes and in the young



SCD in 1866 Young Athletes

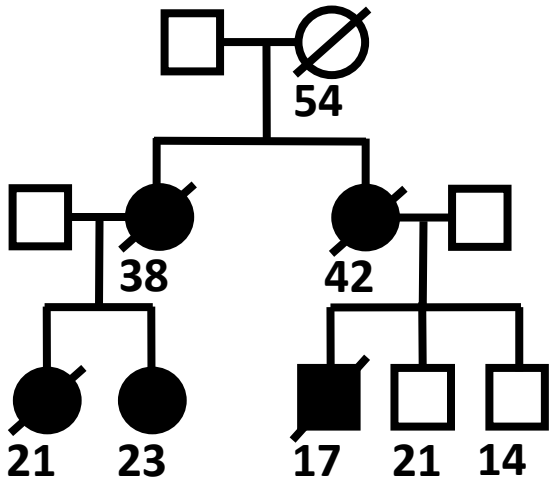
Maron, BJ et. al. Circulation 2009; 119:1085-1092

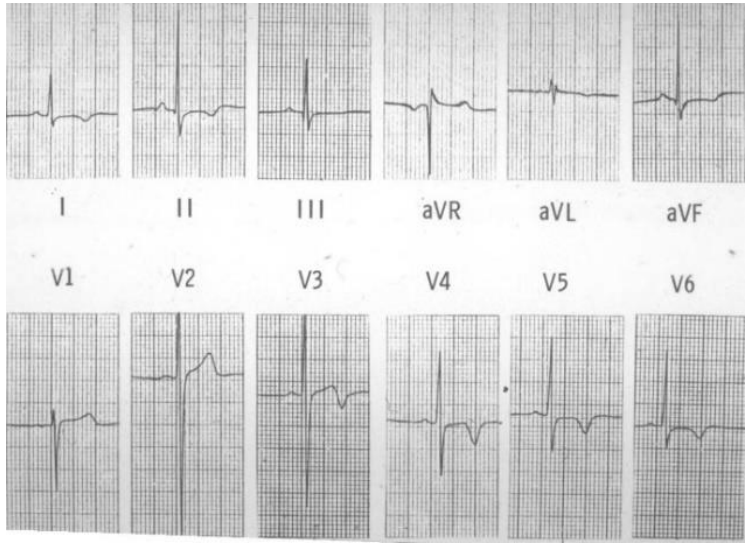
Pathogenesis of SCD in the young

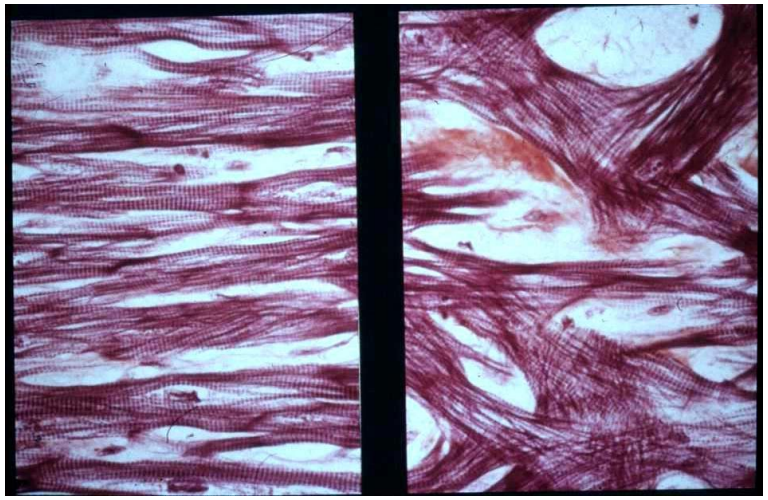


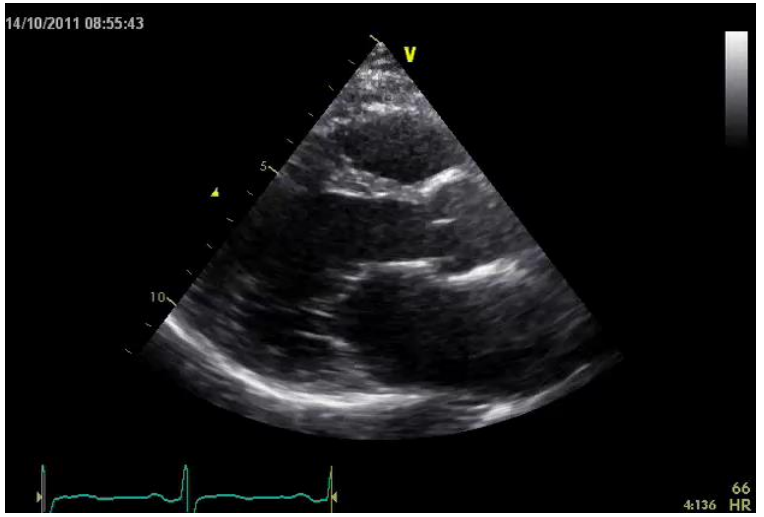
Maron BJ, Circulation 2009;119:1085-92; de Norontha SV Heart 2009;95:1409-14; Eckart RE Ann Intern Med 2004; 141:829-34; Harmon KG Circ Arrhythm Electrophysiol 2014;7:198-204.

Troponin T (Arg94Leu):
sudden death, myocardial disarray, but no hypertrophy







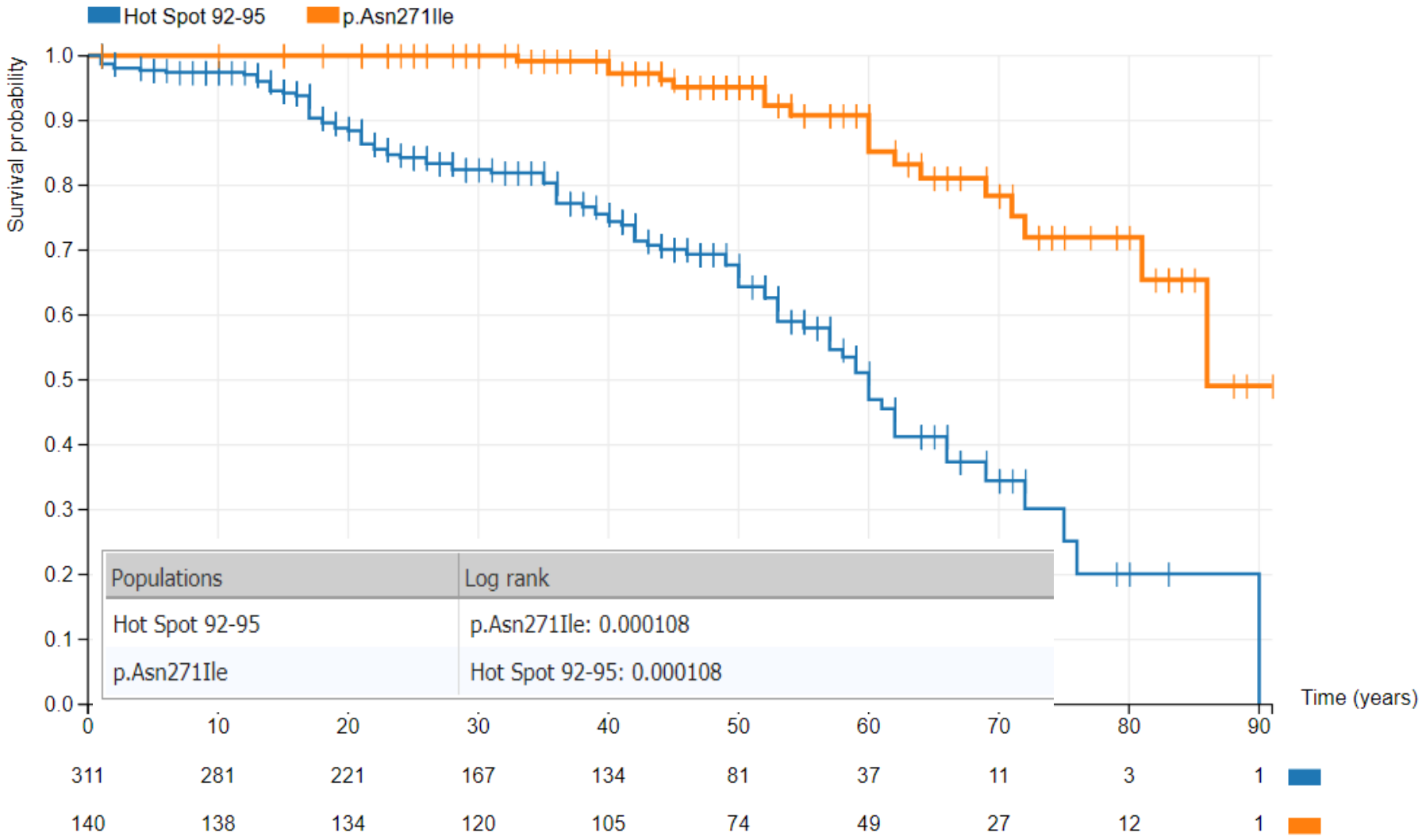


McKenna WJ, Stewart JT, Nihoyannopoulos P, McGinty F, **Davies MJ**. *British Heart Journal* 1990; 63:287-90

By courtesy of William McKenna

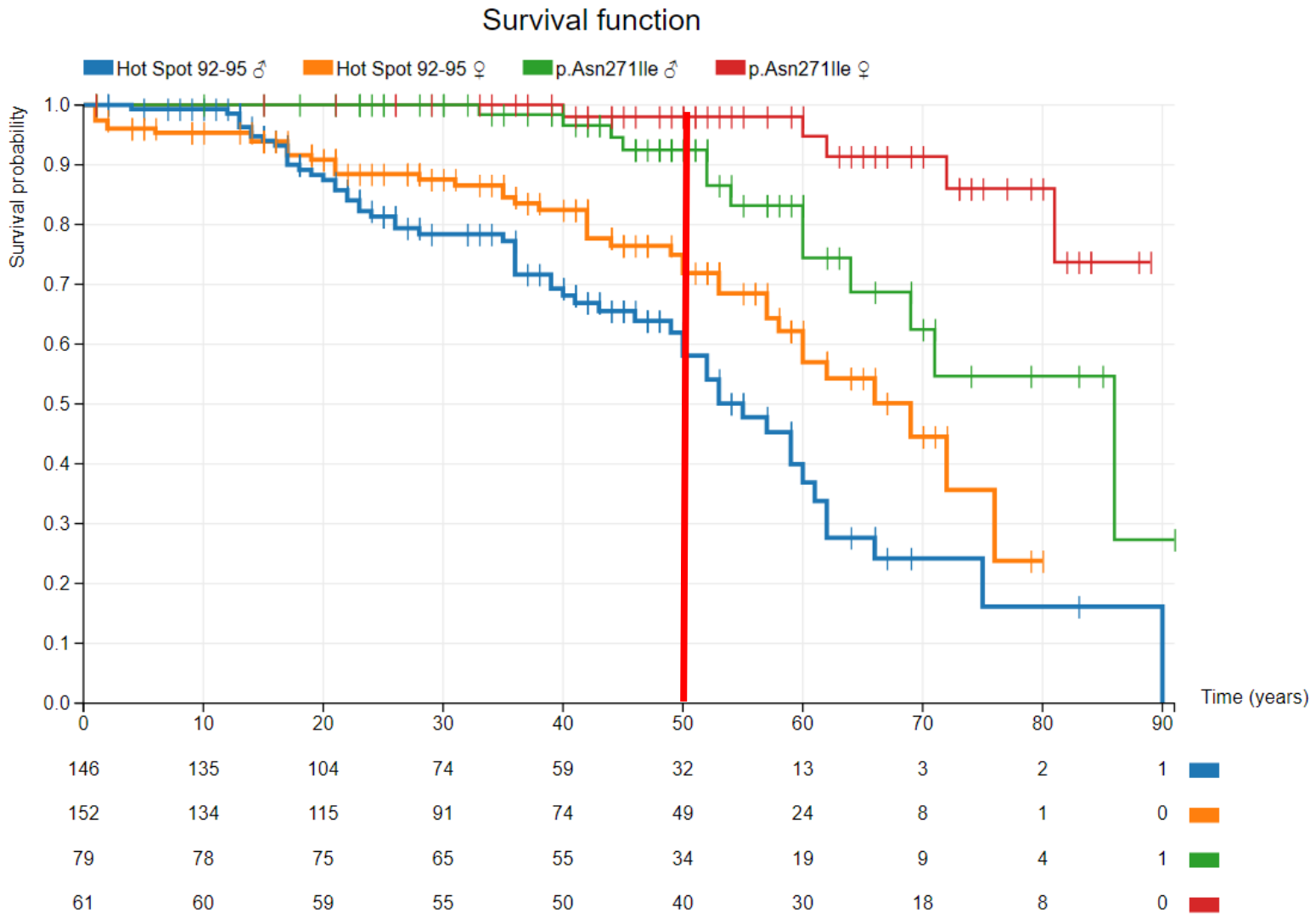
Genetic determinants of outcome

Kaplan Meier Survival – Troponin T Hot Spot 92-95 vs founder mutation at amino acid 271



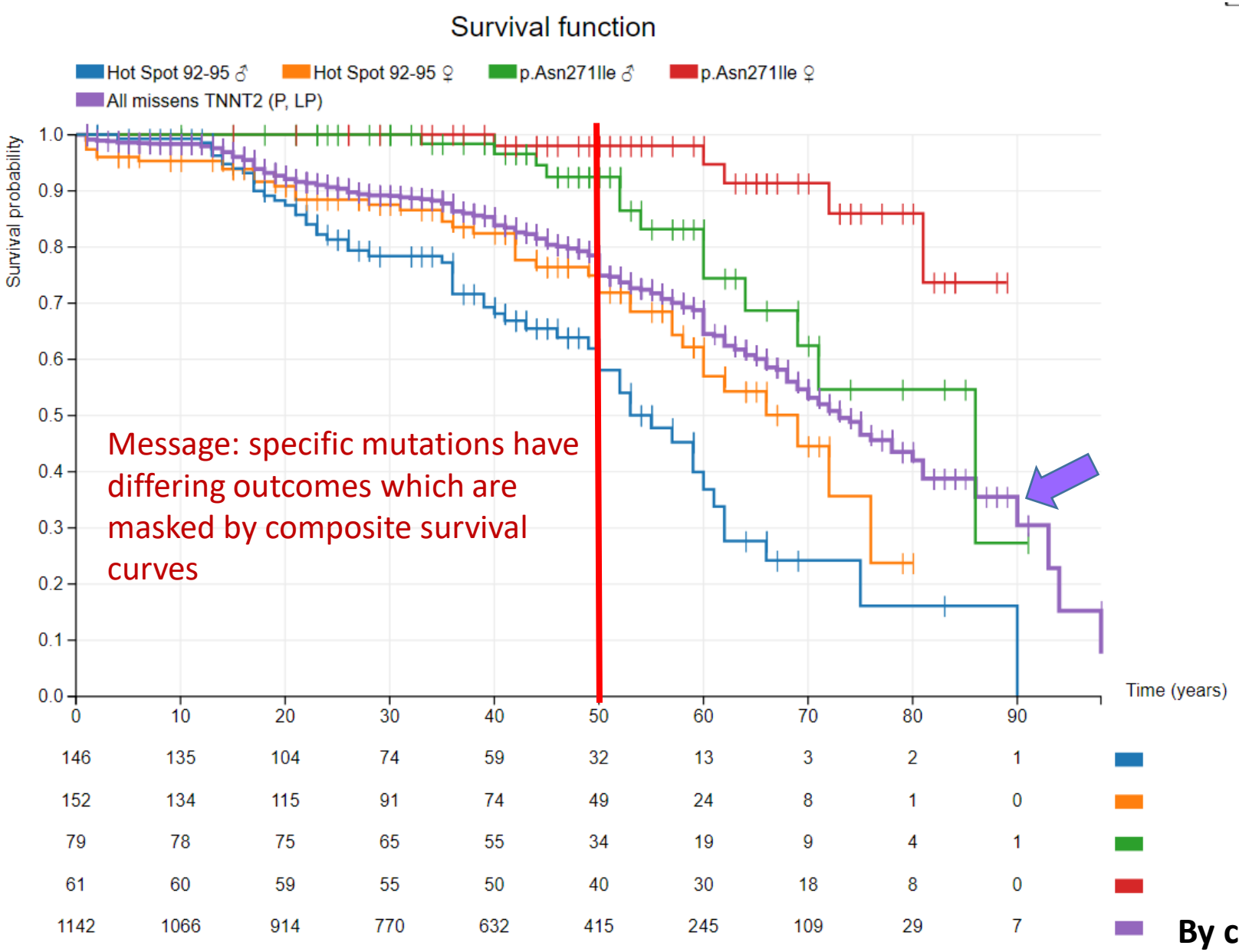
By courtesy of William McKenna

Kaplan Meier Survival – Troponin T Hot Spot 92-95 vs 271 – Male vs Female



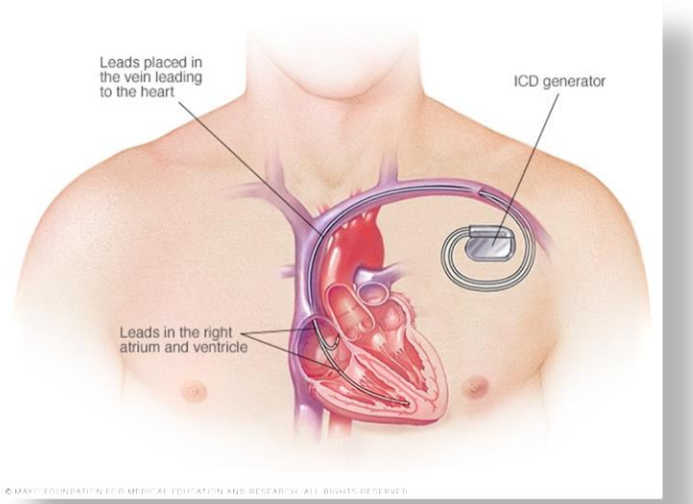
By courtesy of William McKenna

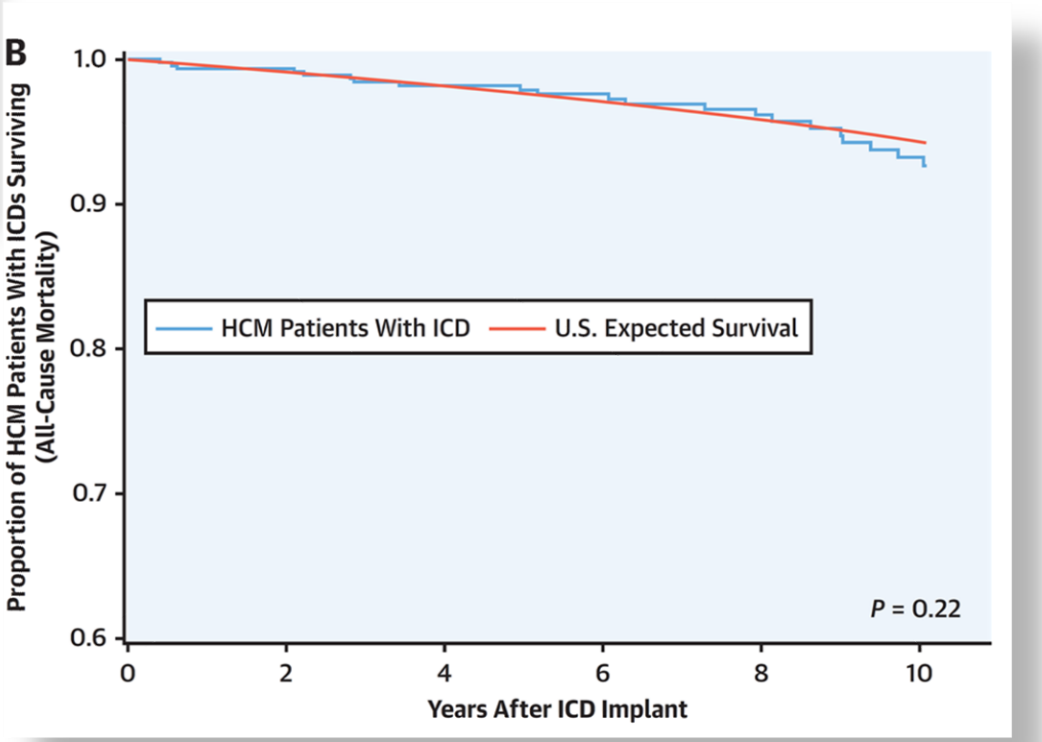
Kaplan Meier Survival – Troponin T Hot Spot 92-95 vs 271 – Male vs Female
Comparison with composite troponin T data

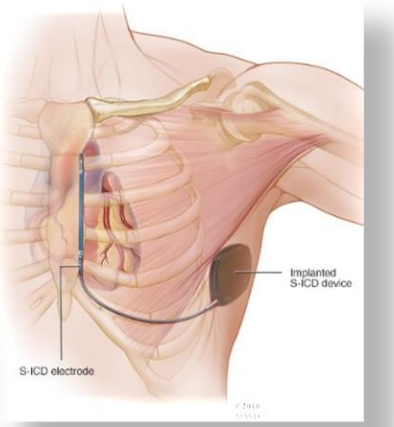


By courtesy of William McKenna

Improved ICD technologies and lower complication rates







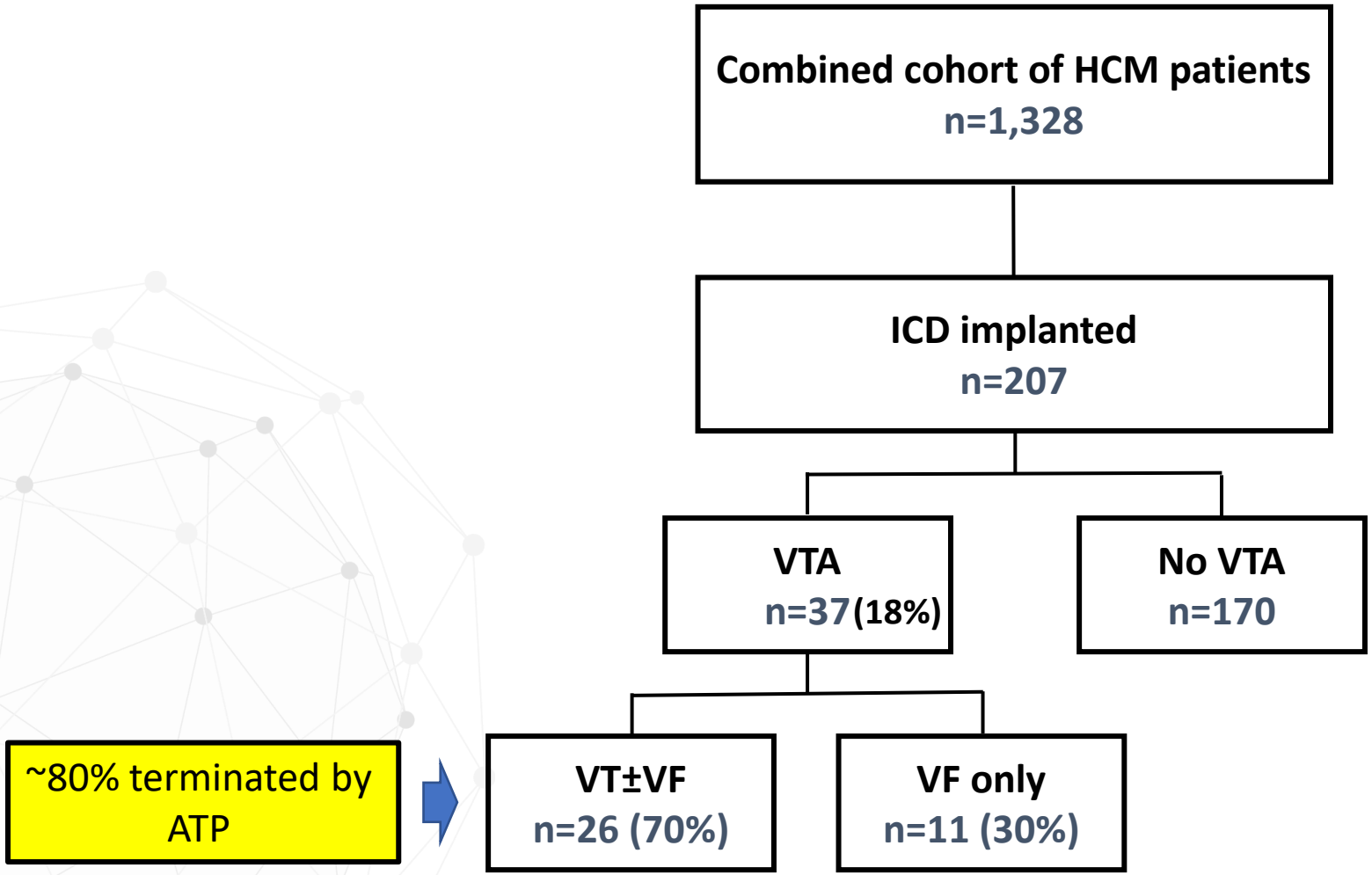
Maron MS, JAMA Cardiol 2019;4:644-657

The 69th Annual Conference of the Israel Heart Society
in association with the Israel Society of Cardiothoracic Surgery
May 25-26, 2022 | Expo Tel Aviv | Israel

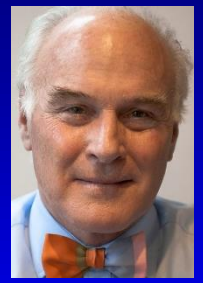
Ventricular arrhythmias in hypertrophic cardiomyopathy patients: prevalence, distribution, predictors and outcome

Amitai Segev[1], Yishay Wasserstrum[1], Michael Arad[1], Jose M. Larrañaga-Moreira[2], Cristina Martinez-Veira[2], Roberto Barriales-Villa[2], and Avi Sabbag[1]

1. Sheba Medical Center, Tel Hashomer, Israel
2. Complexo Hospitalario Universitario A Coruña, A Coruña, Spain


Combined cohort of HCM patients
n=1,328
ICD implanted
n=207
VTA
n=37 (18%)
No VTA
n=170
VT±VF
n=26 (70%)
VF only
n=11 (30%)
~80% terminated by ATP

How to apply the recommendations of the current guidelines?



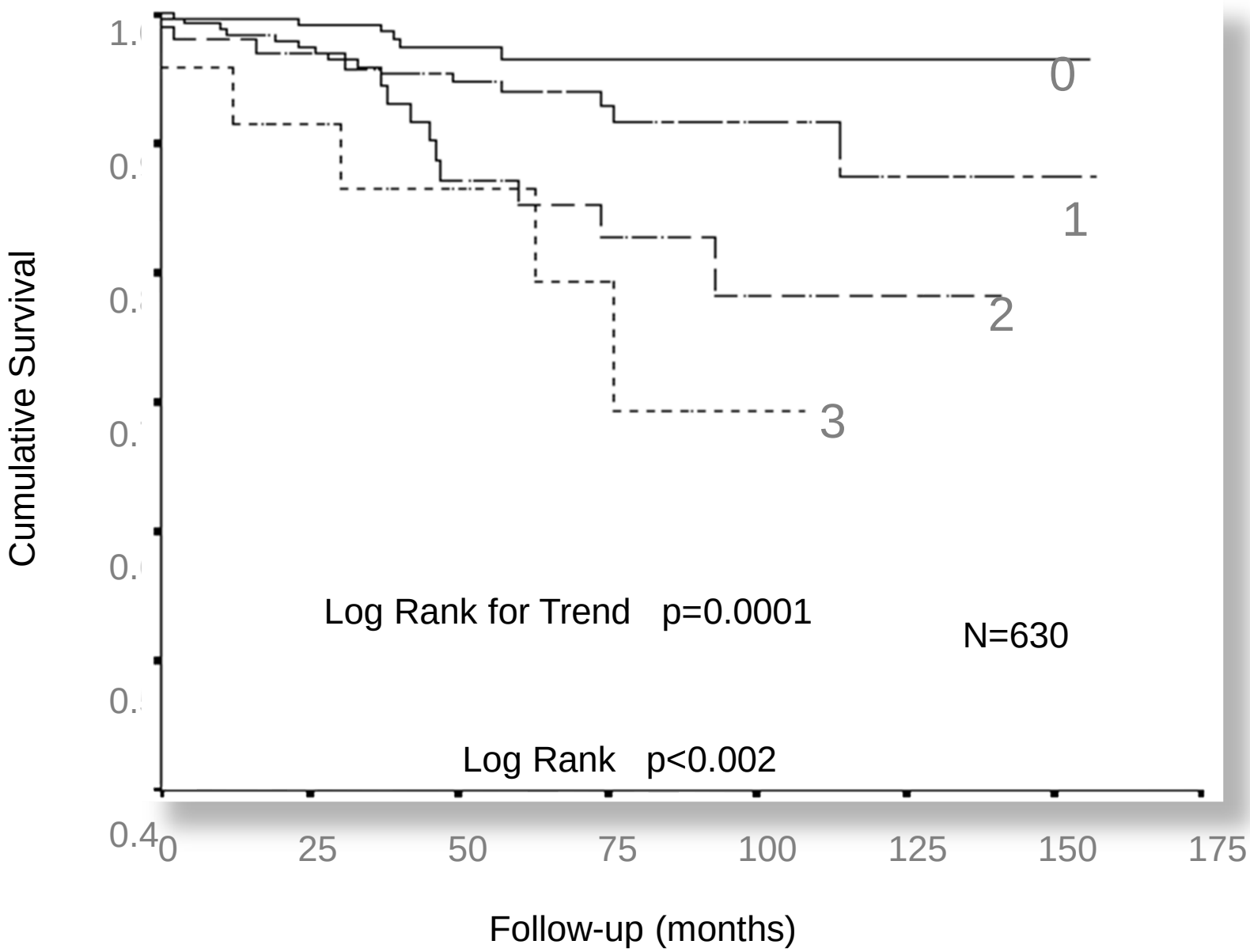



ESC 2014 Guideline "Euroscore" like principle

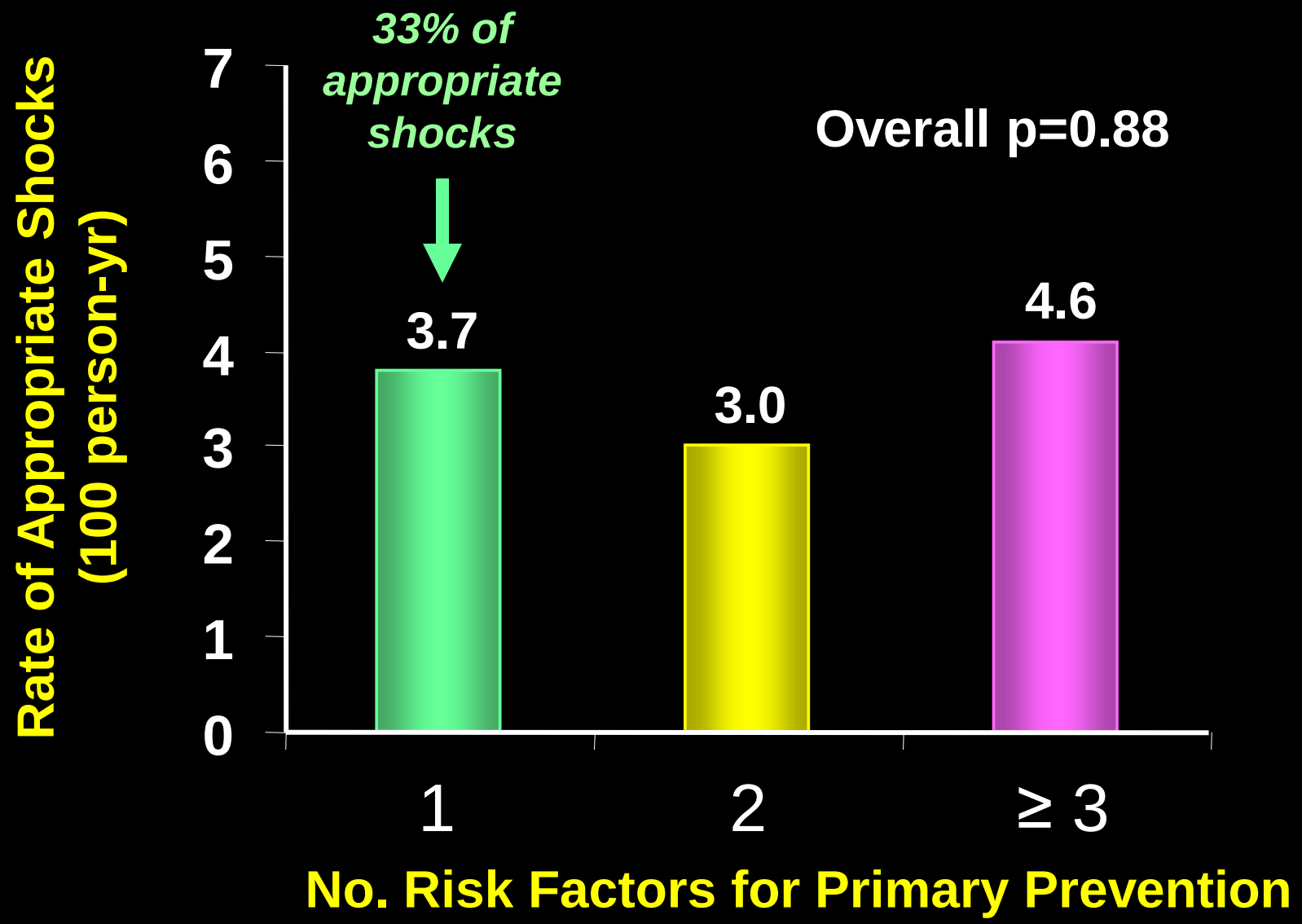
ACCF/AHA Guideline 2011/ 2020

ACC/ESC Consensus Document (2003)

More Risk Factors – Lower the Survival



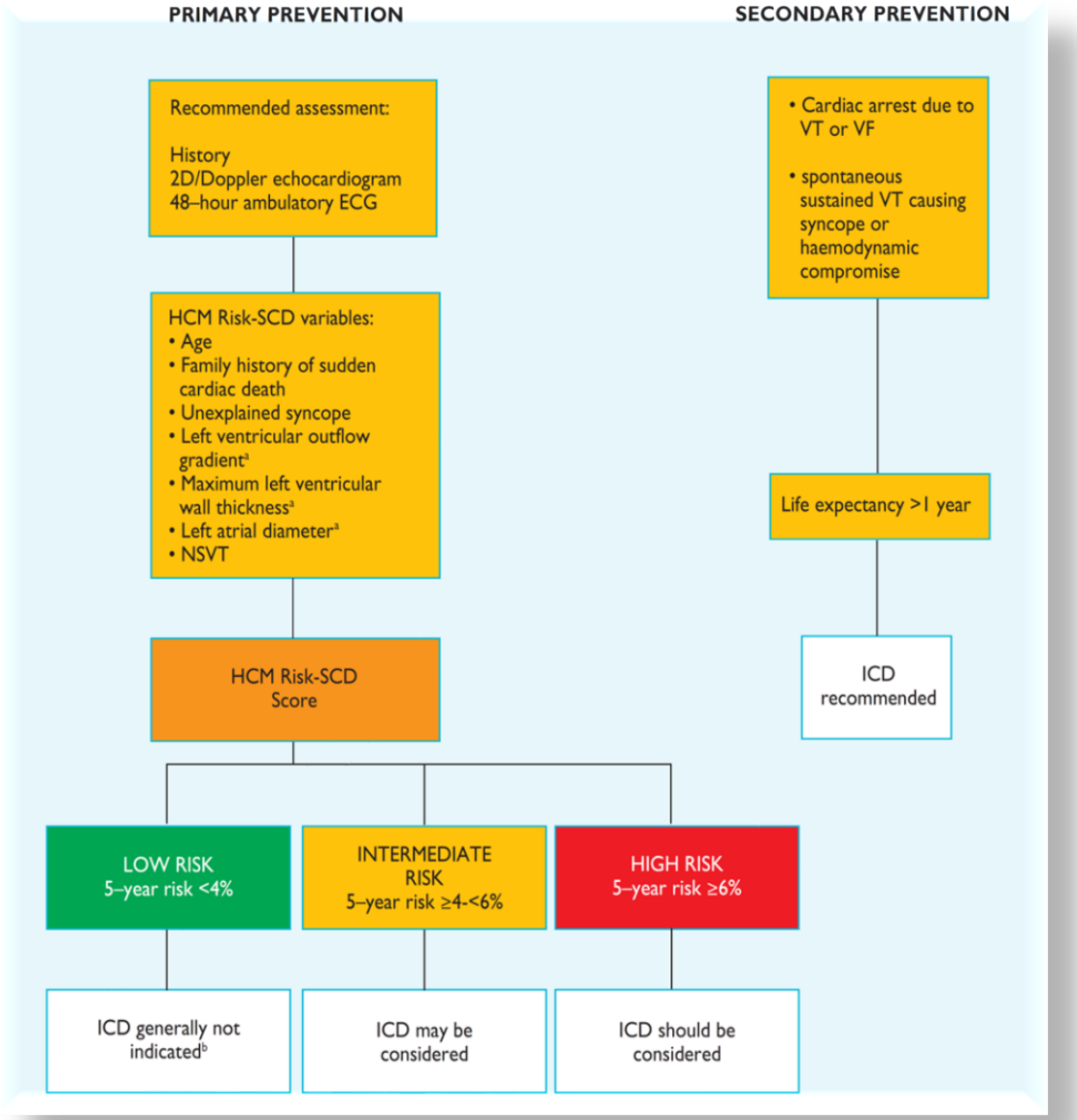
Elliott PM et al. Lancet 2001;357:420-24



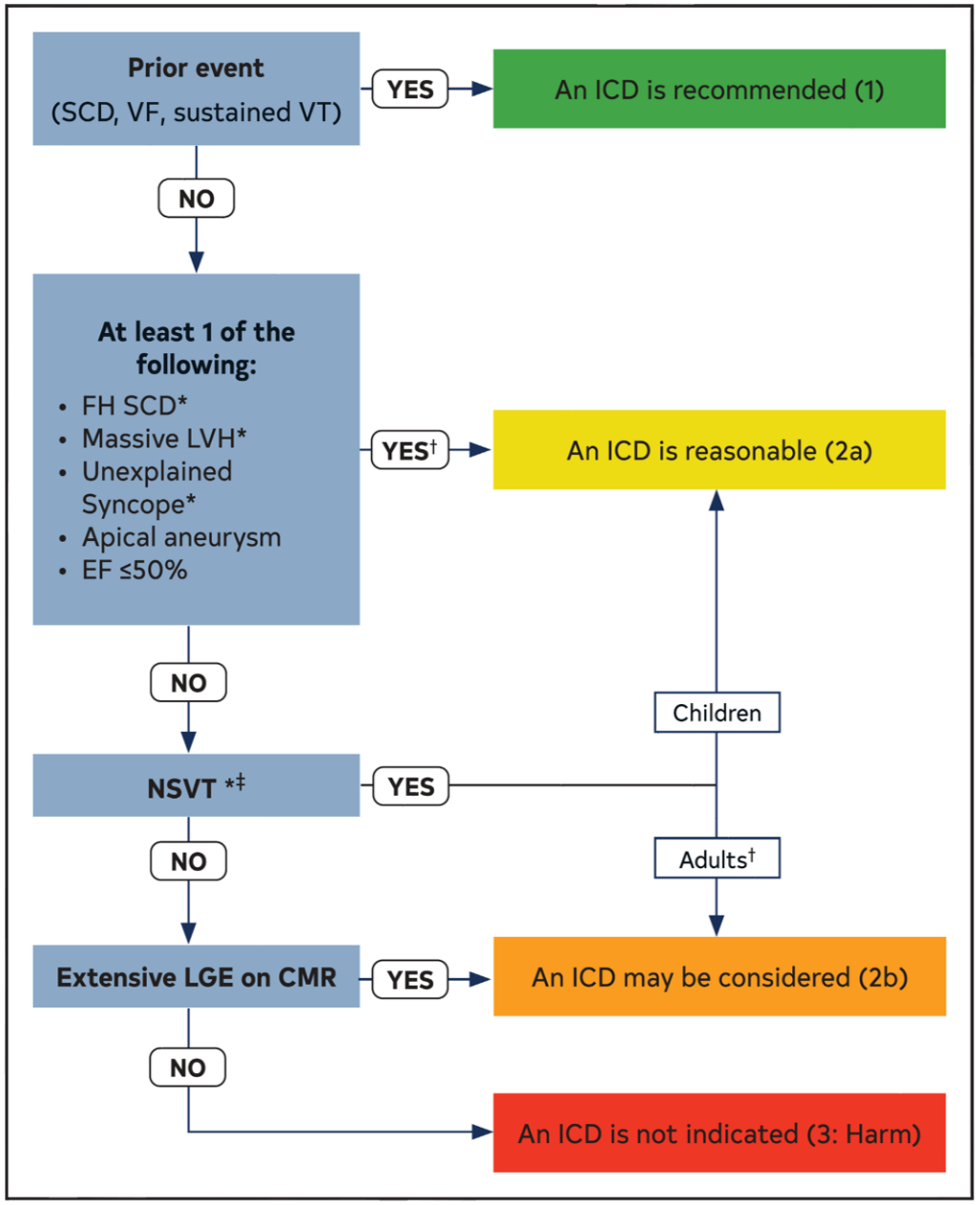
Courtesy of Maron BJ

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

Sudden Death Risk Definition in HCM



Elliott P, European Heart J 2014;39:2733-2779



2a	B-NR	3. For patients who are ≥ 16 years of age with HCM, it is reasonable to obtain echocardiography-derived left atrial diameter and maximal LVOT gradient to aid in calculating an estimated 5-year sudden death risk that may be useful during shared decision-making for ICD placement[2,22] (Table 7).

Ommen SR, AHA/ACC 2020 guidelines, J Am Coll Cardiol. 2020;76(25):e159-e240.

Missing Variables in SCD risk Assessment

- **ESC risk calculator (2014):**
  - Blood pressure response to exercise
  - End stage disease (LVEF ≤50%)
  - Apical aneurysm
  - Extensive late gadolinium enhancement on CMR

- **AHA/ACC 2020 guidelines**
  - Age
  - Blood pressure response to exercise
  - LVOT gradient
  - NSVT – not major risk category

- The implication of single major risk factor!

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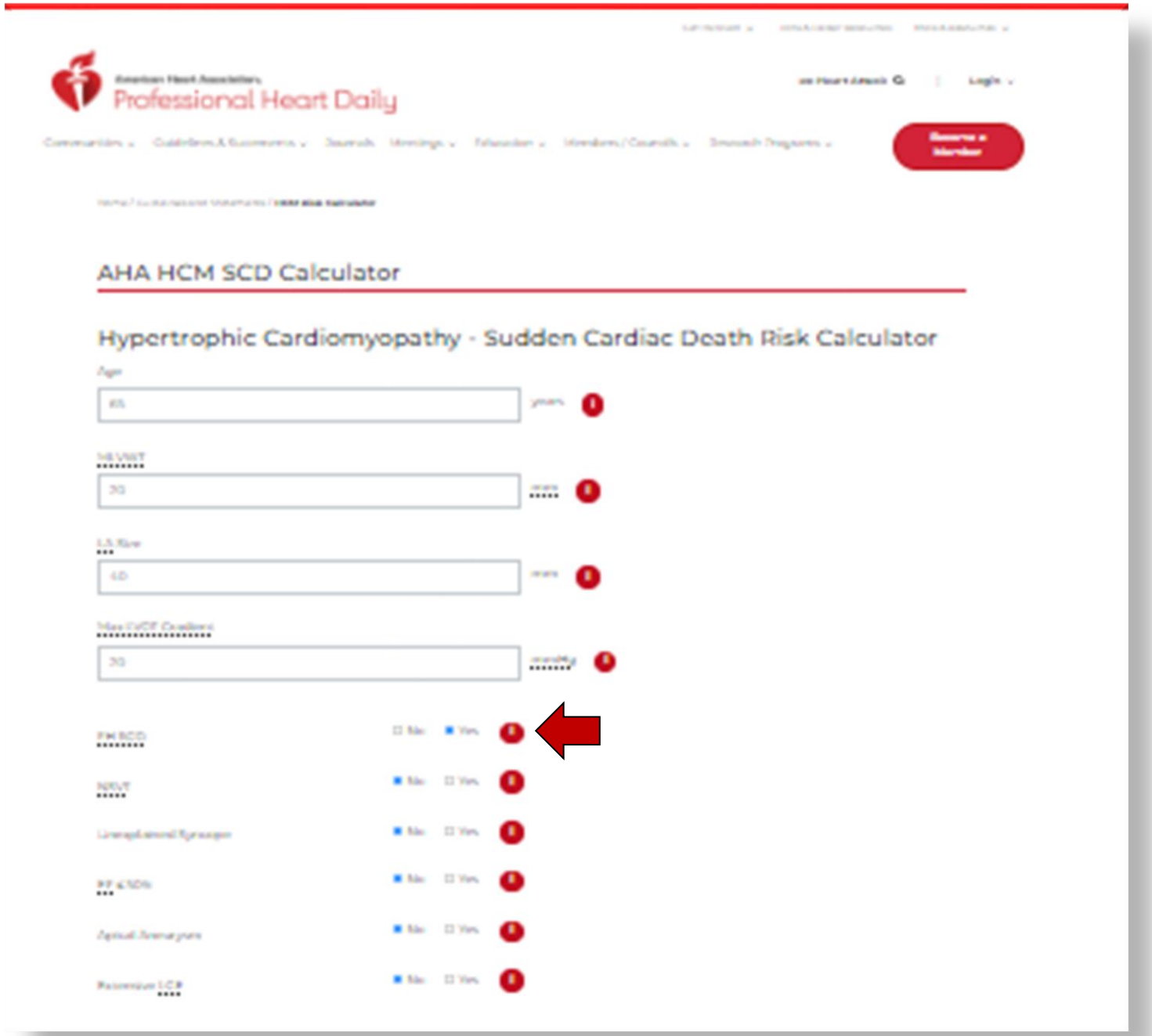
HCM Risk-SCD Calculator

Age	65	**Years**	*Age at evaluation*
Maximum LV wall thickness	20	**mm**	*Transthoracic Echocardiographic measurement*
Left atrial size	40	**mm**	*Left atrial diameter determined by M-Mode or 2D echocardiography in the parasternal long axis plane at time of evaluation*
Max LVOT gradient	20	**mmHg**	*The maximum LV outflow gradient determined at rest and with Valsalva provocation (irrespective of concurrent medical treatment) using pulsed and continuous wave Doppler from the apical three and five chamber views. Peak outflow tract gradients should be determined using the modified Bernouilli equation: Gradient= $4V^2$, where V is the peak aortic outflow velocity*
Family History of SCD	○ No	● Yes	*History of sudden cardiac death in 1 or more first degree relatives under 40 years of age or SCD in a first degree relative with confirmed HCM at any age (post or ante-mortem diagnosis).*
Non-sustained VT	● No	○ Yes	*3 consecutive ventricular beats at a rate of 120 beats per minute and <30s in duration on Holter monitoring (minimum duration 24 hours) at or prior to evaluation.*
Unexplained syncope	● No	○ Yes	*History of unexplained syncope at or prior to evaluation.*


Professional Heart Daily
AHA HCM SCD Calculator
Hypertrophic Cardiomyopathy - Sudden Cardiac Death Risk Calculator

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Risk of SCD at 5 years (%):	2.24
ESC recommendation:	ICD generally not indicated **

American Heart Association.

Risk of SCD at 5 years(%)

2.24

Recommendation

Based on the SCD Risk factors present, this patient has a Class 2A indication for ICD (is reasonable)

Current messages from the literature can be applied in the specific patient using sound clinical judgment

Shared Decision Making in HCM in Our Practice

Take-home messages

- **The rate of SCD in HCM is 0.5%/year**
- **Risk stratification should be based on ESC/AHA guidelines, clinical judgement and shared decision making**
- **Current technologies allow building an actionable mutations based genetic map for future use in evaluation of SCD risk**

When a thing ceases to be a matter of controversy it ceases to be a matter of interest

William Hazlitt 1778-1830


Thank
you