HCM Summit VI

The ICD and Prevention of Sudden Death

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ASYMMETRICAL HYPERTROPHY OF THE HEART IN YOUNG ADULTS

BY

DONALD TEARE

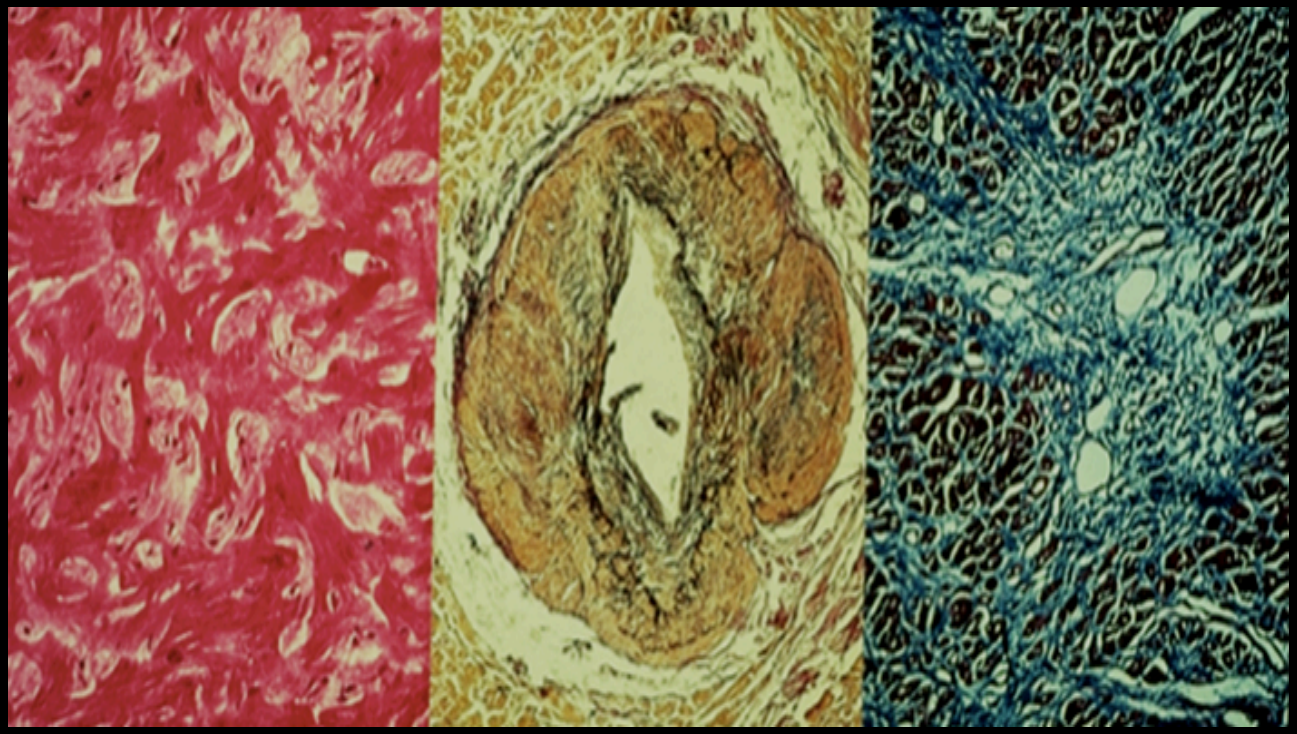
From the Department of Pathology, St. George's Hospital

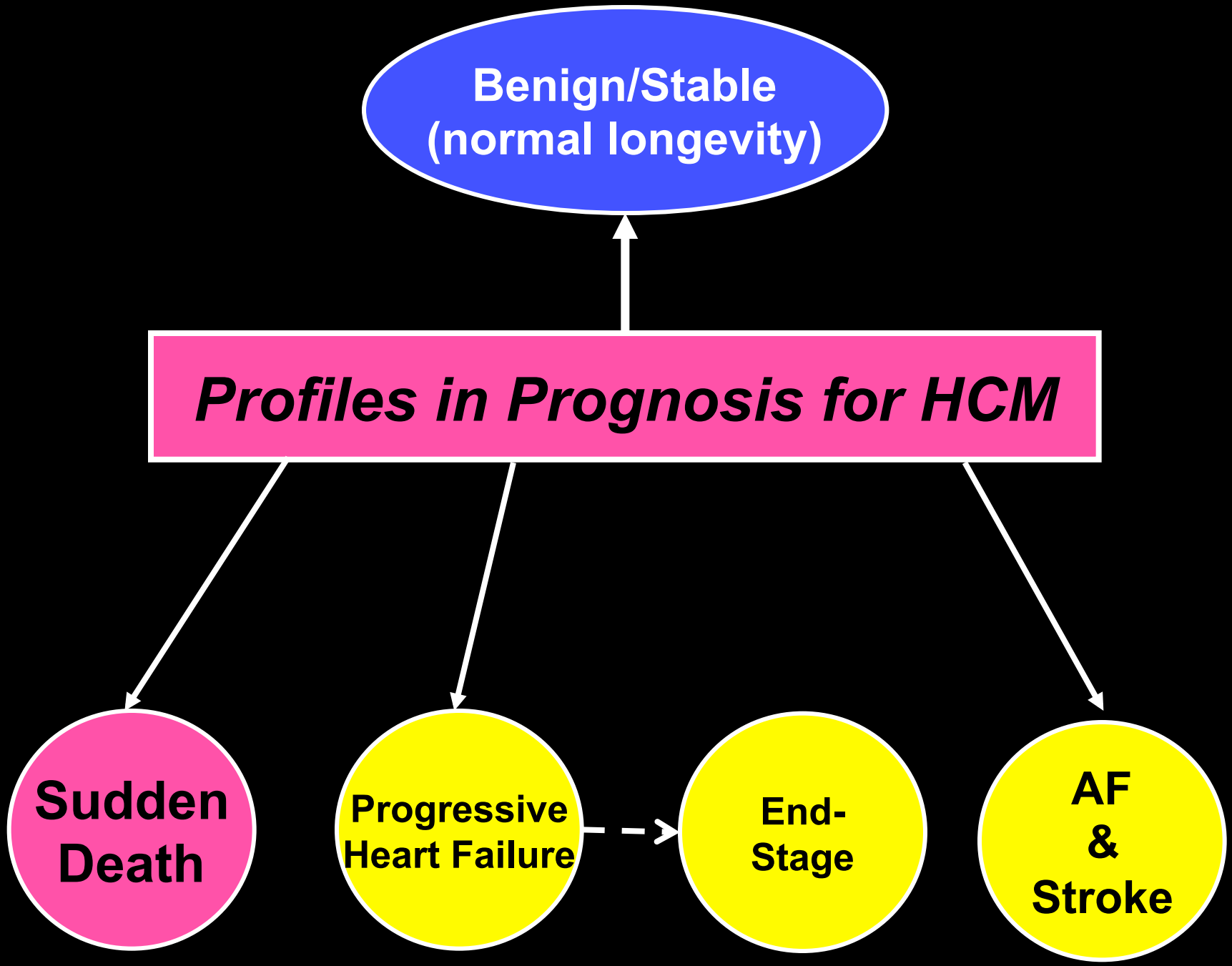
Received January 7, 1957

"Tumours of the heart and pericardium have evoked an extensive literature out of all proportion to their uncommon incidence and their relative unimportance as a cause of clinical heart disease." This opening sentence of Friedberg's chapter on cardiac tumours in *Diseases of the Heart* (Friedberg, 1949) fills a pathologist with diffidence in reporting eight cases that have been seen in the last six years in a series of 16,000 autopsies.

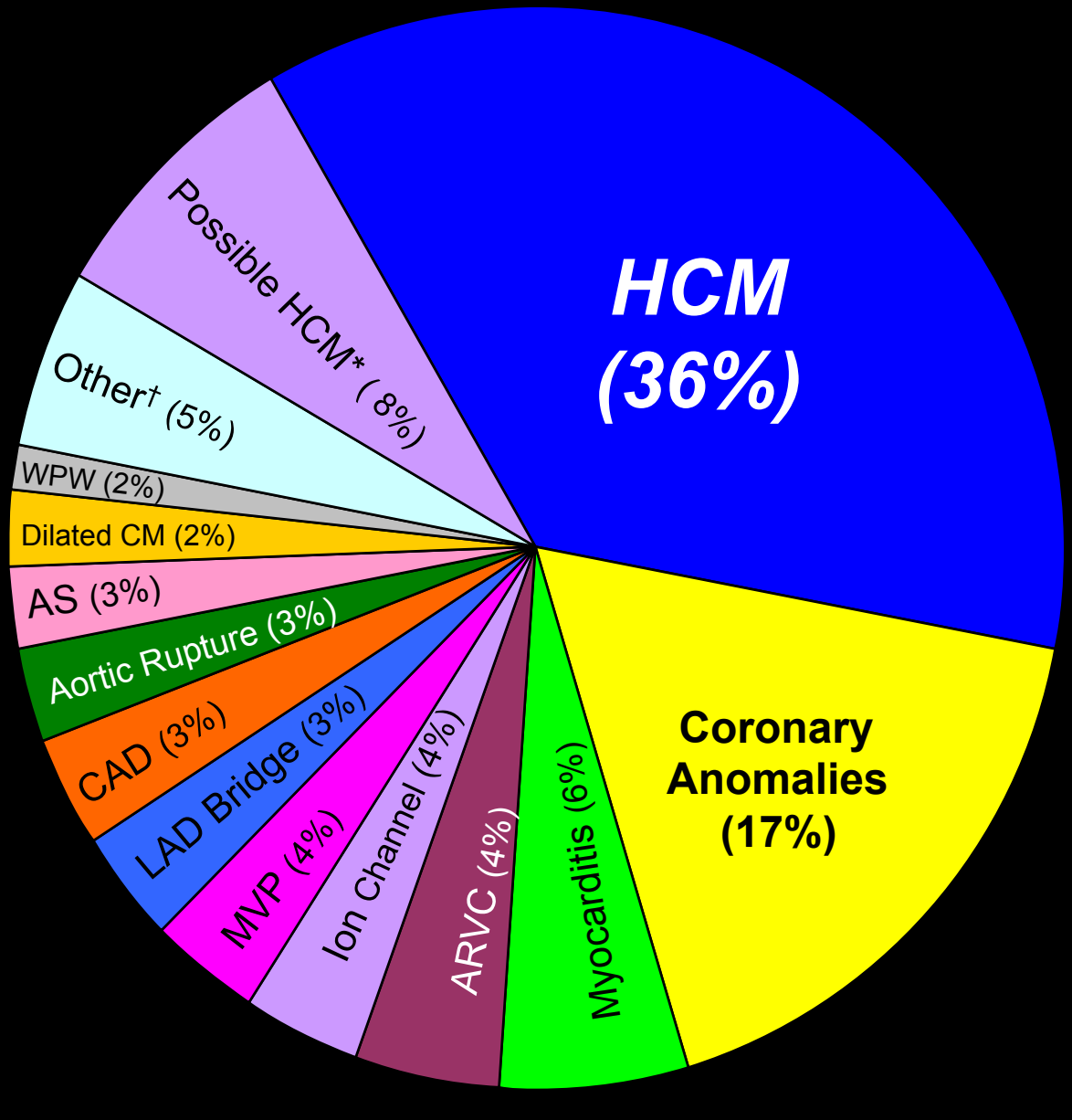
Primary tumours of the heart are undoubtedly a rarity and according to Mahaim (1945) 413 had been recorded up to 1945. There is little justification for recording rarities in young adults unless they have some relation to fitness for military service or confuse the differential diagnosis, particularly of conditions that may respond to cardiac surgery. These eight cases of asymmetrical hypertrophy or benign tumour of the heart have occurred in a large group where sudden death and indeed cardiac incapacity, particularly among men, is rare.

Arrhythmogenic (Unstable) Myocardial Substrate in HCM



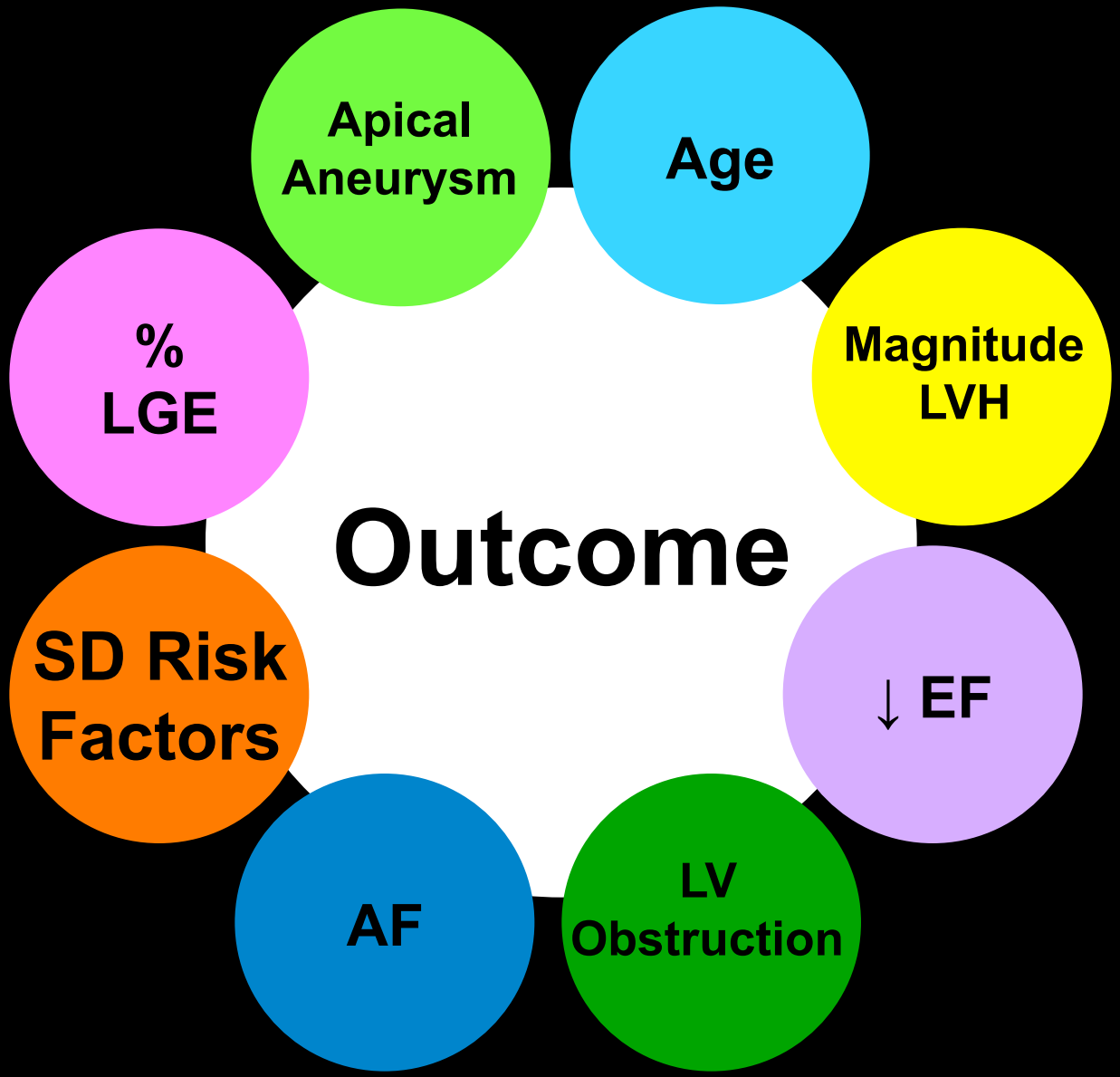

Benign/Stable
(normal longevity)
Profiles in Prognosis for HCM
Sudden Death
Progressive Heart Failure
End-Stage
AF & Stroke

Sudden Death in Young Athletes



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

What Counts in HCM



2011 US/CANADA ACC/AHA Guidelines For HCM

2° prevention

Cardiac arrest/sustained VT

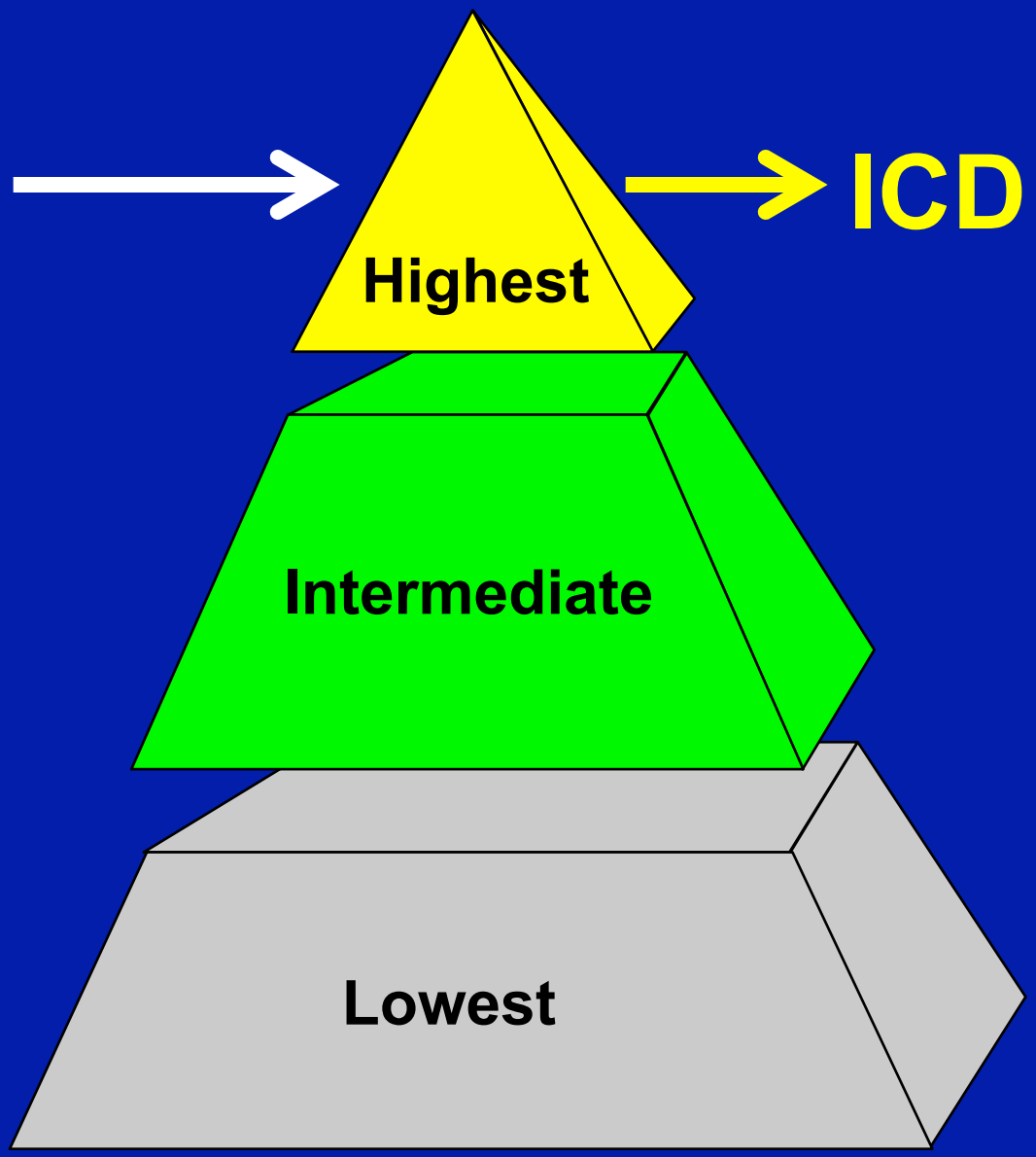
1° prevention

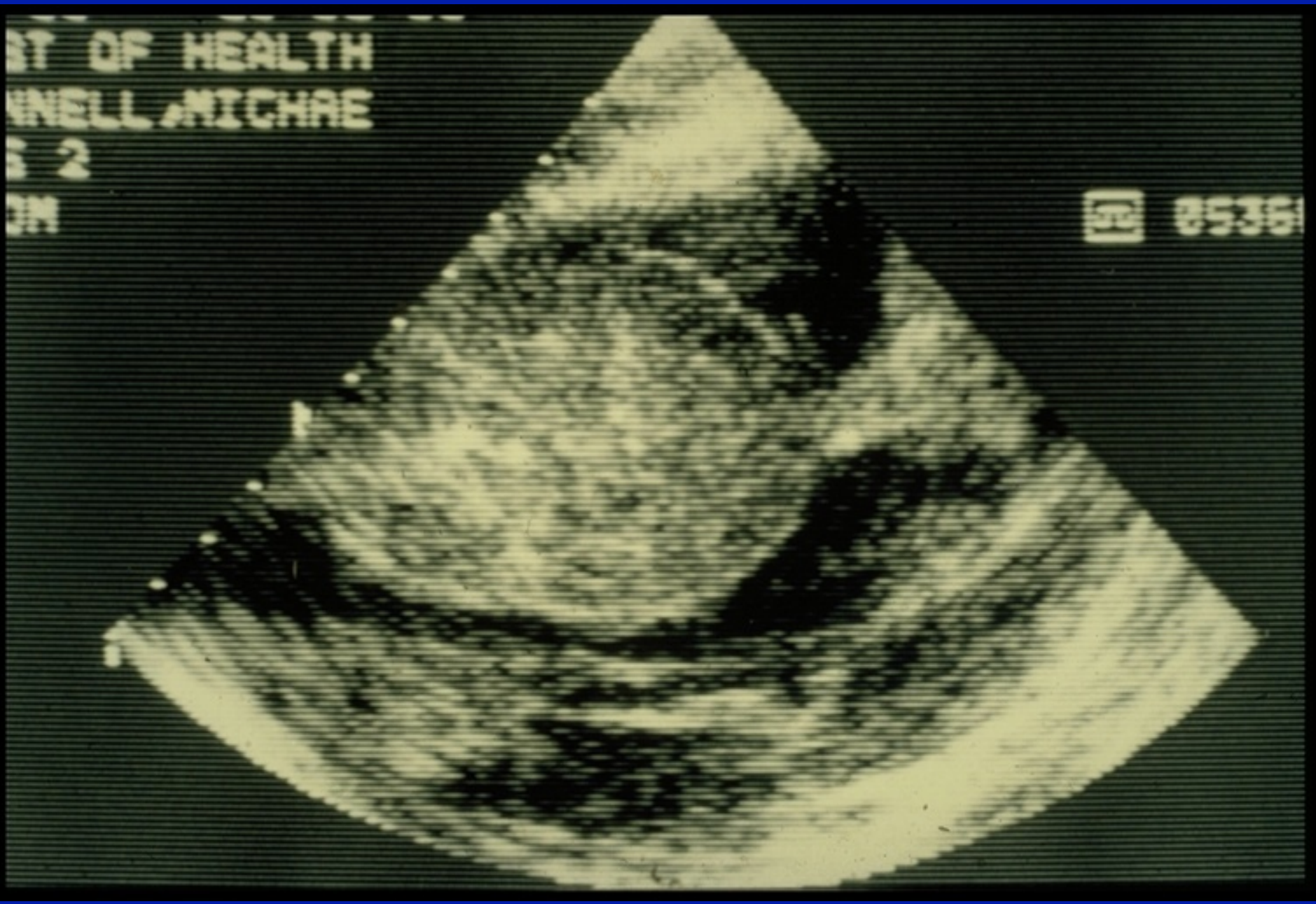
Family history HCM-SD
Unexplained syncope
Multiple-repetitive NSVT (Holter)
Abnormal exercise BP response
LGE ≥ 15% of LV mass
Massive LVH ≥ 30 mm
LV apical aneurysm

Rare subgroups/potential arbitrators

End-stage (EF < 50%)
Marked LV outflow obstruction (rest)
Modifiable
- **Intense competitive sports**
- **CAD**

LGE ≥ 15% of LV mass
Age ≥ 60y
Alcohol septal ablation (some pts)

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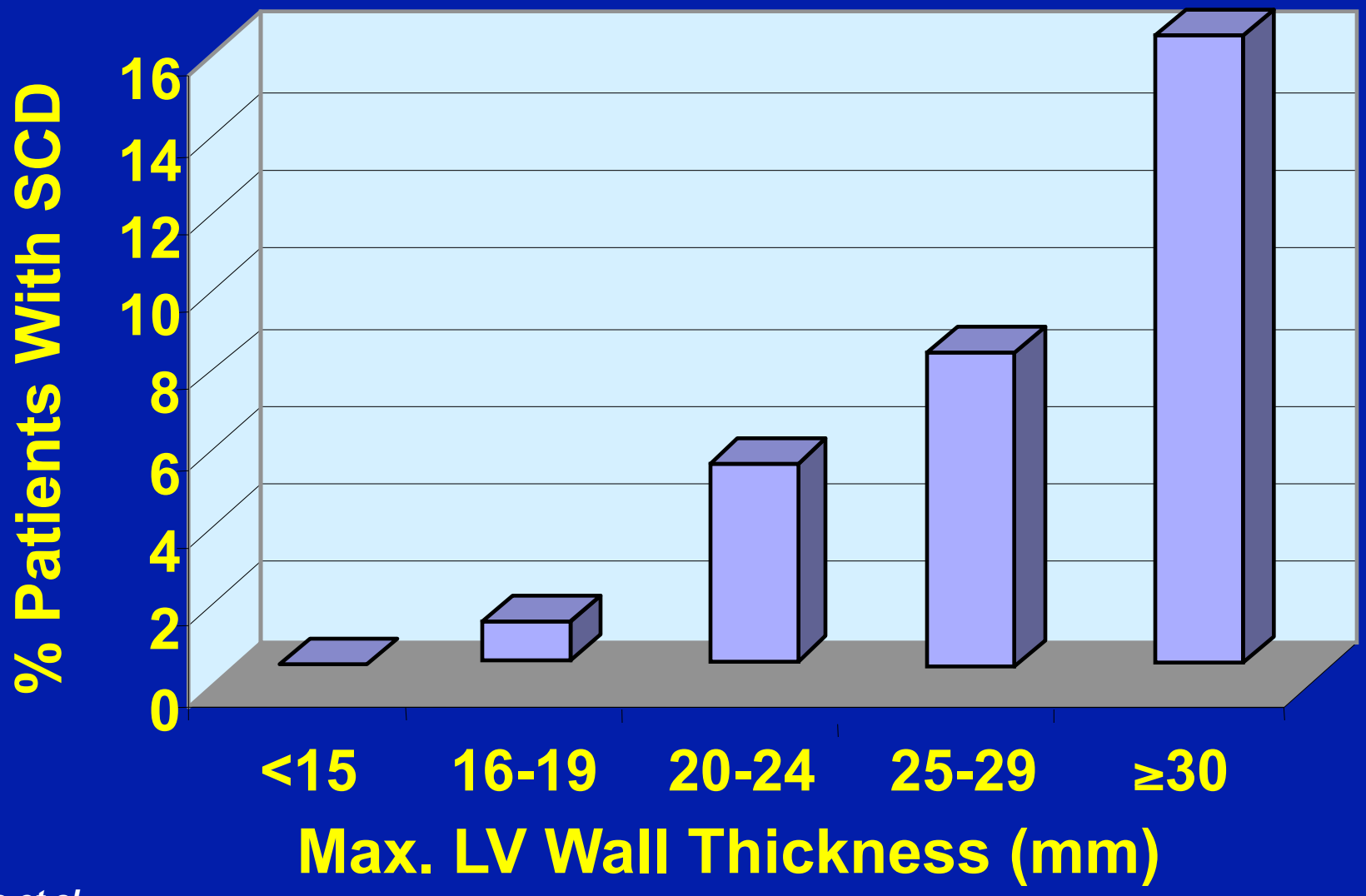
Hypertrophy Counts in HCM:
Relation Between LV Thickness & SD



Spirito et al
NEJM 2000;342:1778

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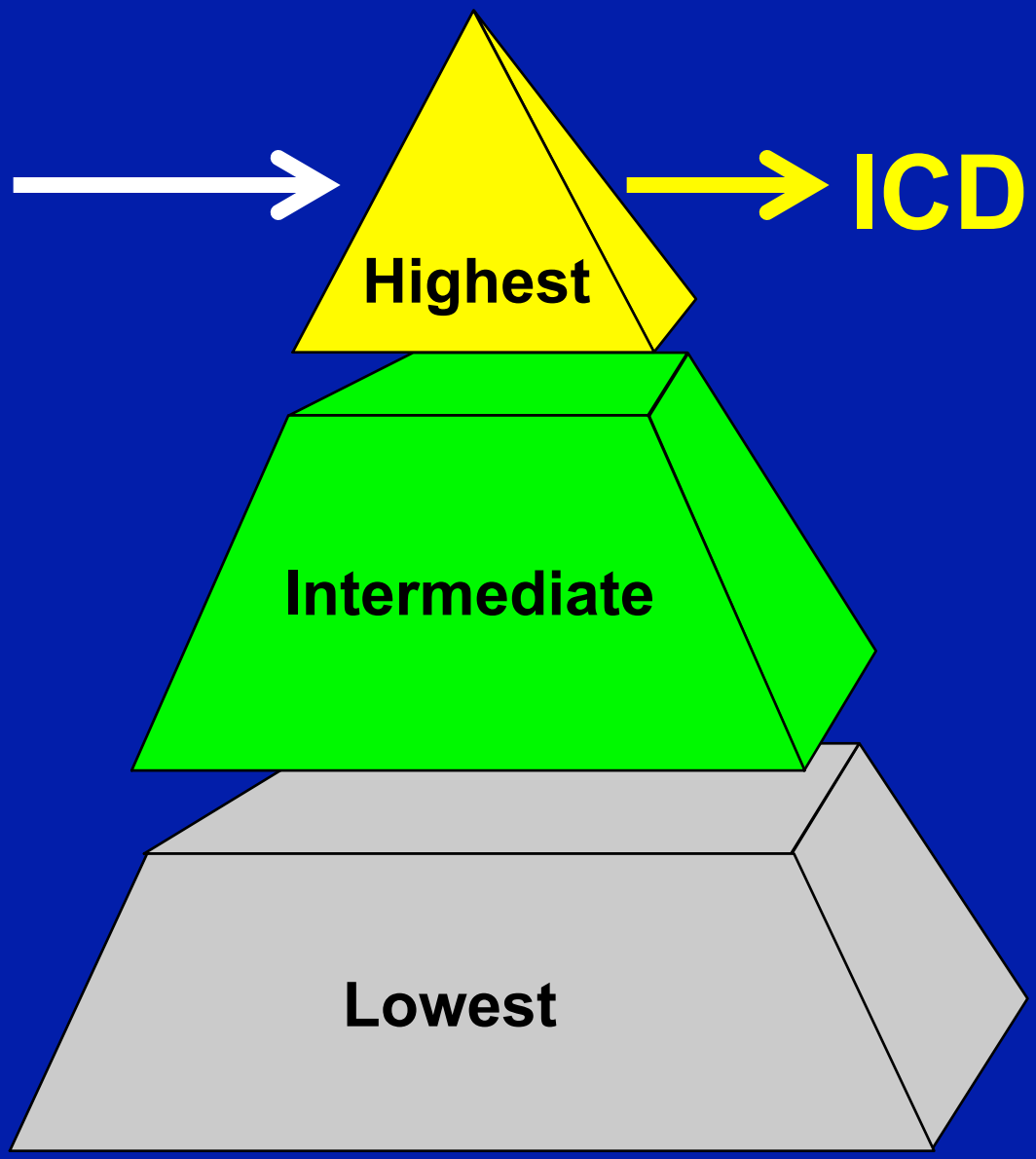
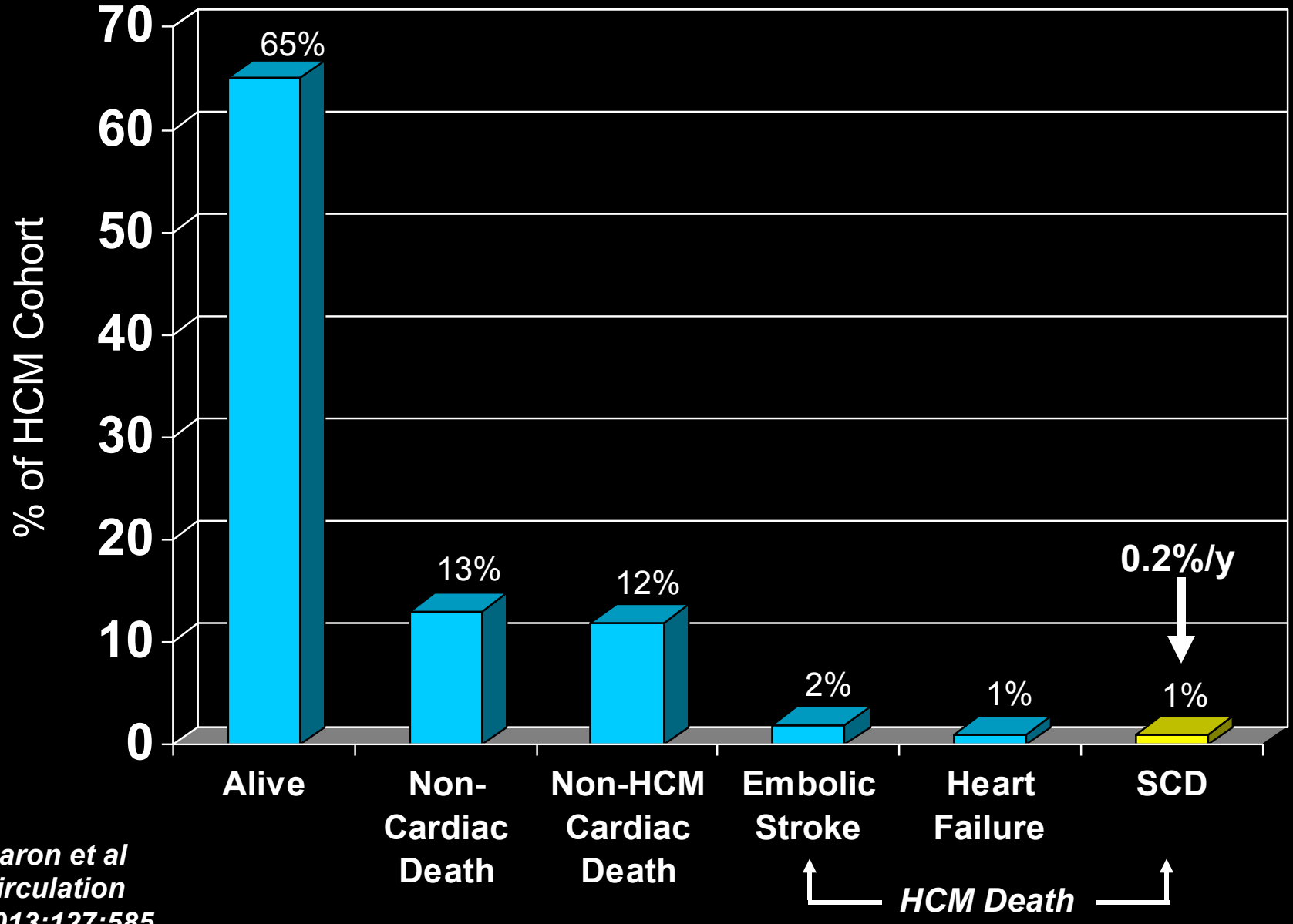


Outcome of HCM Patients First Evaluated ≥ 60 Years

Age Counts And Aging is Good in HCM



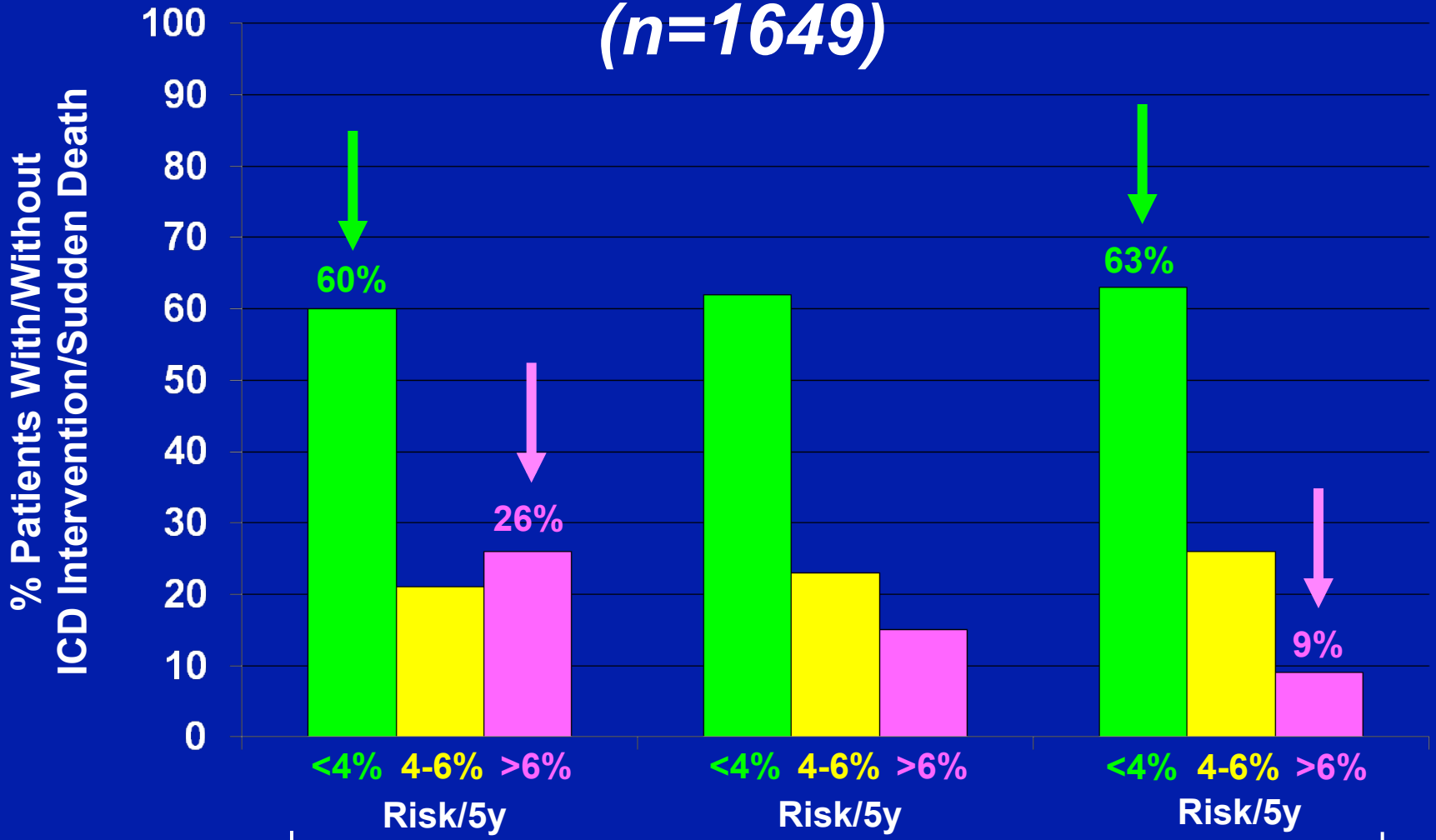
Maron et al Circulation 2013;127:585

The ESC-HCM prediction formula for SD is as follows:

$$\text{Probability}_{\text{SCD at 5 years}} = 1 - 0.998^{\exp(\text{Prognostic index})};$$

where Prognostic index = [0.15939858 x maximal LV wall thickness (mm)] – [0.00294271 x LV maximal wall thickness^2 (mm^2)] + [0.0259082 x left atrial diameter (mm)] + [0.00446131 x maximal (rest/Valsalva) LV outflow tract gradient (mm Hg)] + [0.4583082 x family history SCD] + [0.82639195 x NSVT] + [0.71650361 x unexplained syncope] – [0.01799934 x age at clinical evaluation (years)].

Limitations in ESC Sudden Death Risk Score (n=1649)



Maron et al
AJC 2015;116:757

Prevention of Sudden Death in HCM

Dr. Michele Mirowski

Circulation

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EDITORIAL

Implanted Standby Defibrilla

BERNARD LOWN
PAUL AXELROD

WHEN A PROBLEM gains wide social consciousness a diversity of practical and impractical solutions is engendered. This is now the case with the formidable problem of sudden death in patients with coronary heart disease.

Sudden death largely afflicts the ambulatory subject, prodromes are not distinctive, lead time is short, and death probably results from ventricular fibrillation (VF). Tragedy is magnified by the realization that the heart may

of survivors. The inexorable logic of the problem coerces a new direction, namely, identification and protection of the patient at high risk from sudden death.[1] One intriguing approach is to pre
implantation in t
automatic defibrilla

A completely implanted defibrillator can reverse VF in dogs.[2] A special transducer-tipped catheter, sensing pulsatile pressure, is introduced through a peripheral vein into the

Furthermore, there is no evidence that the individual who has a single bout of VF is likely to have recurrences.

If the patient with such an implanted device is found dead, numerous questions will loom including the gnawing doubt that electrocution may have been a factor.

There is serious question whether an indication can be spelled out for the use of an implanted standby defibrillator.

the implanted defibrillator system represents an imperfect solution in search of a plausible and practical application.

TERMINATION OF MALIGNANT VENTRICULAR ARRHYTHMIAS WITH AN IMPLANTED AUTOMATIC DEFIBRILLATOR IN HUMAN BEINGS

M. Mirowski, M.D., Philip R. Reid, M.D., Morton M. Mower, M.D., Levi Watkins, M.D., Vincent L. Gott, M.D., James F. Schauble, M.D., Alois Langer, Ph.D., M. S. Heilman, M.D., Steve A. Kolenik, M.S., Robert E. Fischell, M.S., and Myron L. Weisfeldt, M.D.

THE development of a clinically applicable, automatic, implantable defibrillator has been described previously.[1] This electronic device is designed to monitor cardiac electrical activity, to recognize ventricular fibrillation and ventricular tachyarrhythmias with a sinusoidal wave form, and then to deliver corrective defibrillatory discharges. It is intended to protect patients at particularly high risk of sudden death whenever and wherever they are stricken by these lethal arrhythmias.

After extensive preclinical testing,[2] a pilot study of this new technique was recently initiated at The Johns Hopkins Hospital. This article decribes the first three patients in whom the automatic defibrillator was implanted to manage recurrent ventricular tachyarrhythmias that were refractory to medical therapy. Our results suggest that the device can successfully identify and reverse these malignant arrhythmias in human beings.

Clinical Summaries

Case 1

A 57-year-old woman had an inferior myocardial infarction complicated by ventricular fibrillation eight years before the most recent admission; intractable angina associated with ventricular arrhythmias then developed. Coronary-artery bypass improved the angina but the arrhythmias remained refractory to propranolol, digitalis, quinidine, and procainamide. Two months before admission, ventricular fibrillation occurred outside the hospital and required multiple defibrillations. There was no evidence of acute myocardial in-

Case 2

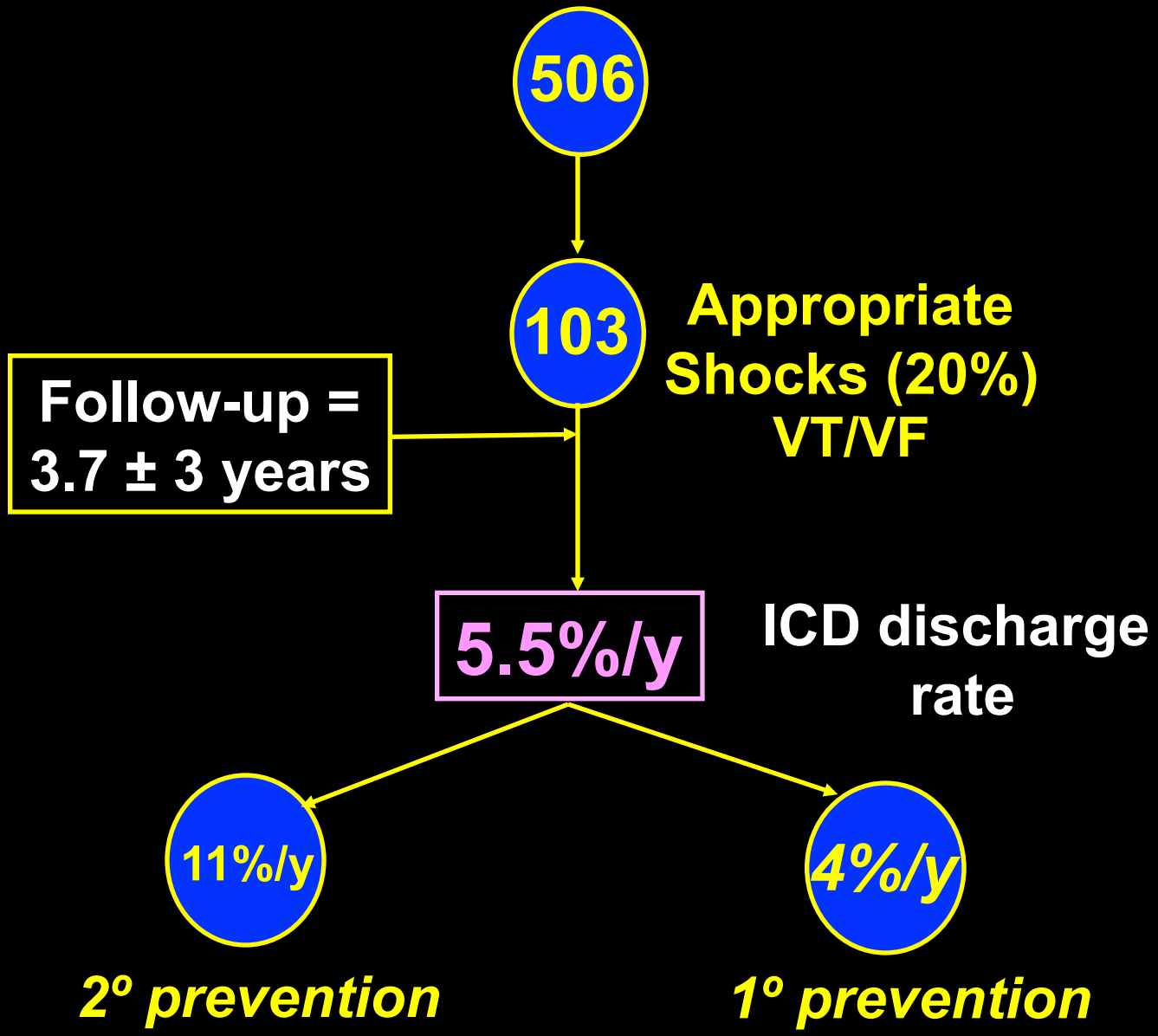
A 16-year-old boy was resuscitated from ventricular fibrillation four years before the most recent admission. Physical examination was unremarkable. Although the coronary arteries and left ventricular function were normal on cardiac catheterization, the papillary muscles were prominent. Ventricular tachycardia was induced during electrophysiologic testing. A demand pacemaker was implanted, and the patient was treated with quinidine, phenytoin, lidocaine, propranolol, procainamide, disopyramide, tocainide, and

Case 3

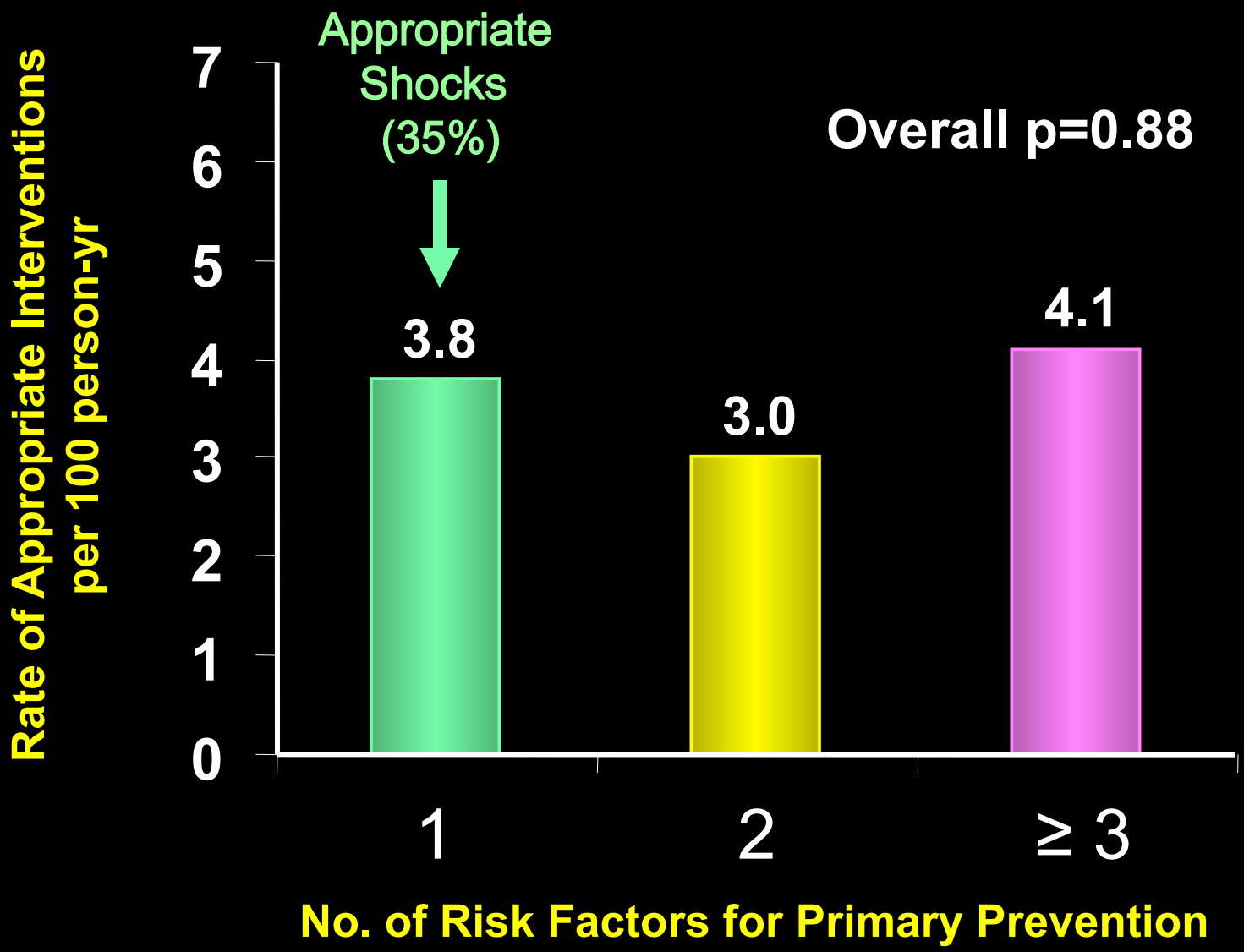
A 43-year-old man with a 10-year history of asymmetric cardiomyopathy had two episodes of ventricular fibrillation outside the hospital and was treated with propranolol, septal myectomy, and a pacemaker. Two months after the operation, progressive dyspnea developed and another episode of ventricular fibrillation occurred.

N Engl J Med 1980;303:322.

ICD Performance in HCM

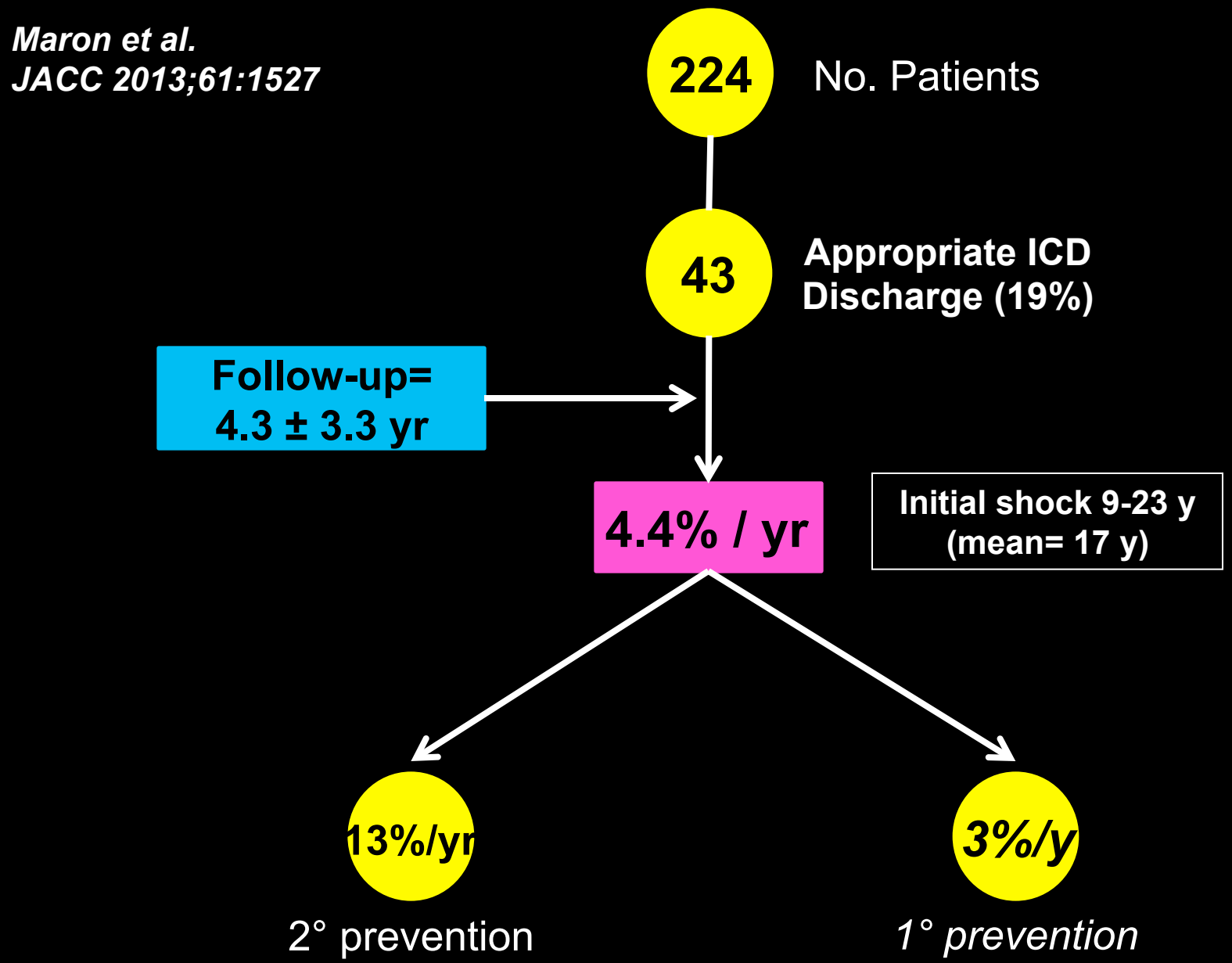


Maron et al
JAMA 2007;298

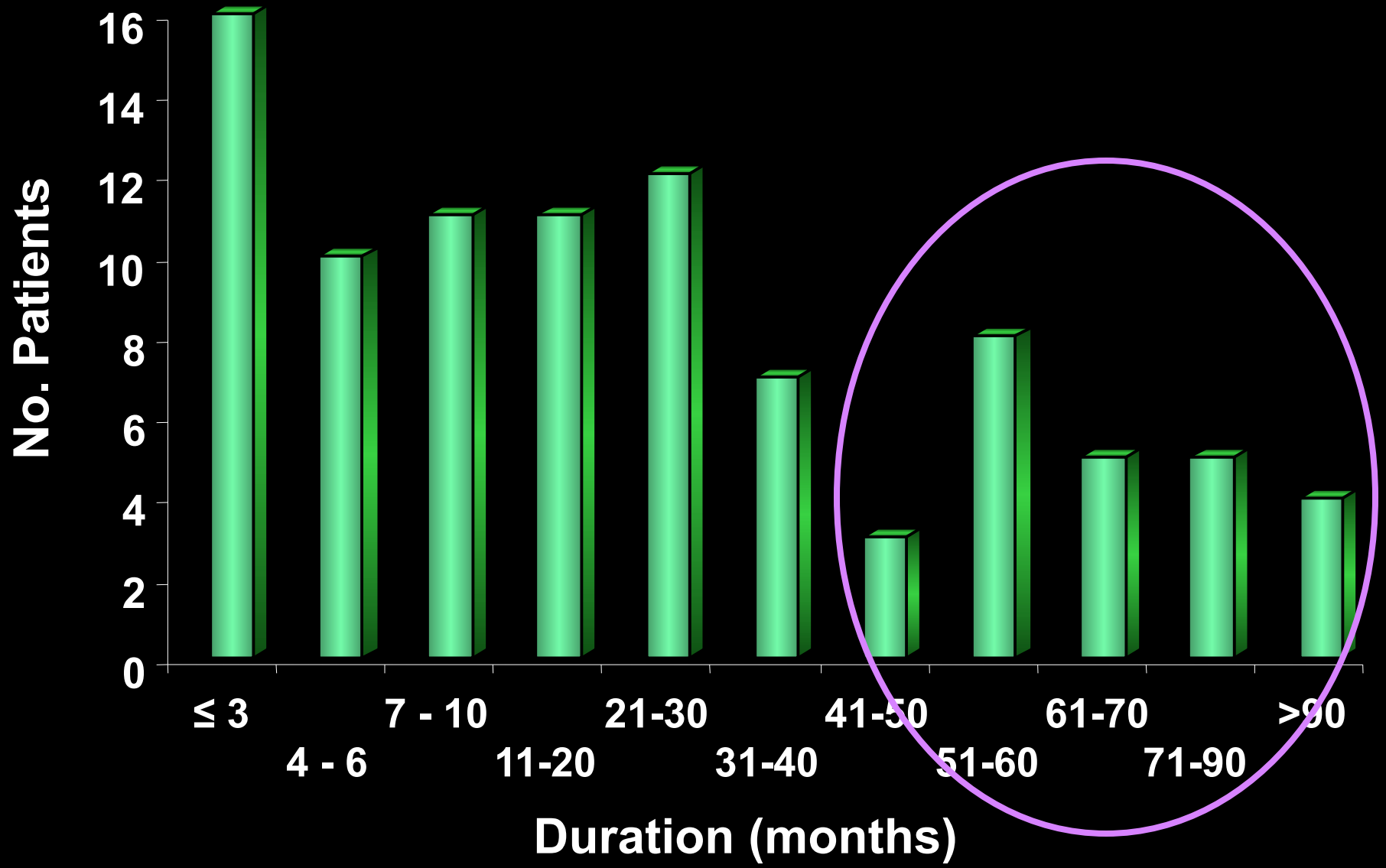


Maron et al
JAMA 2007;298:405

ICD in HCM for Children / Adolescents



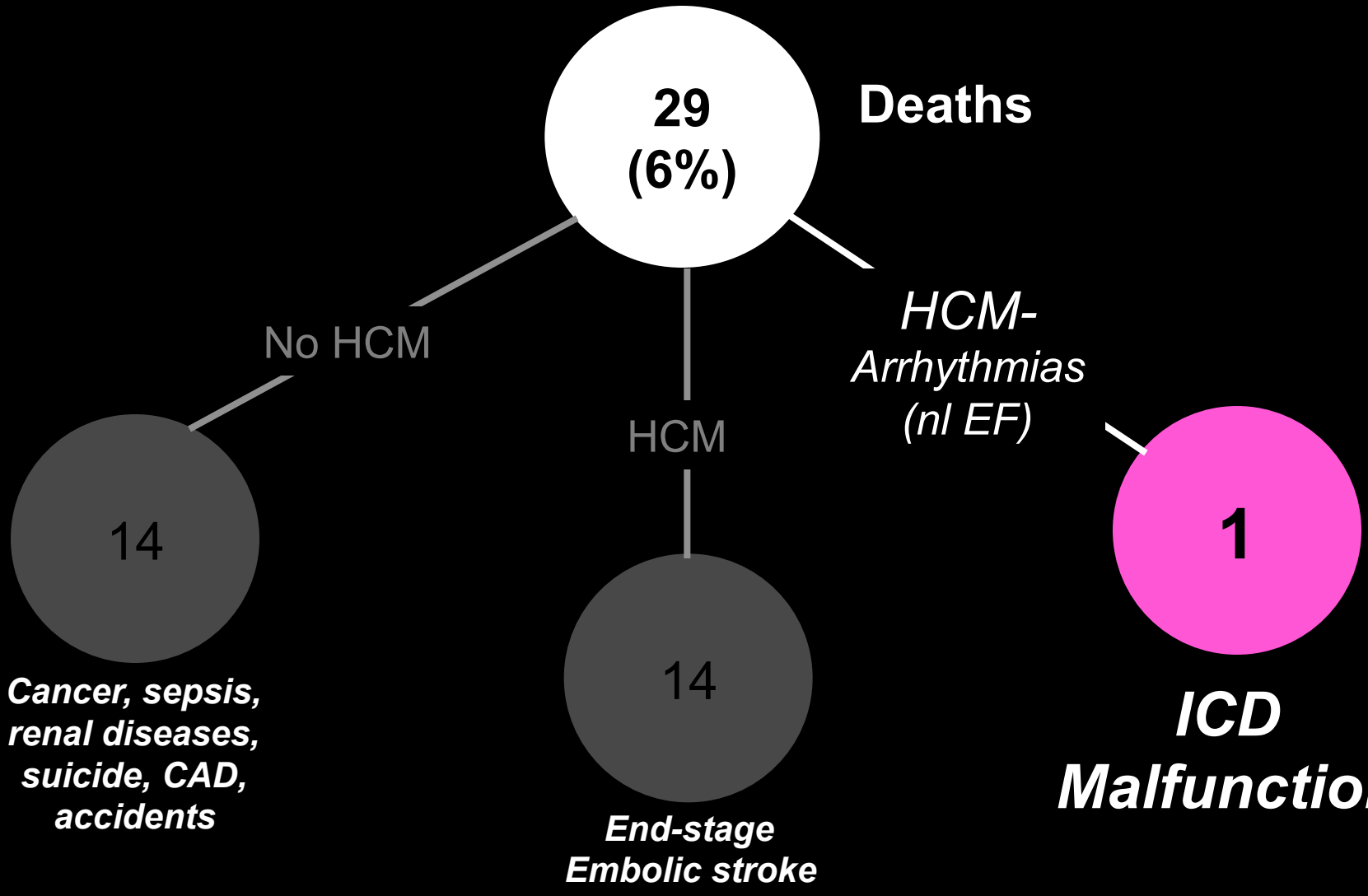
ICD in HCM: Time to First Shock



Maron et al
JAMA 2007;298:405

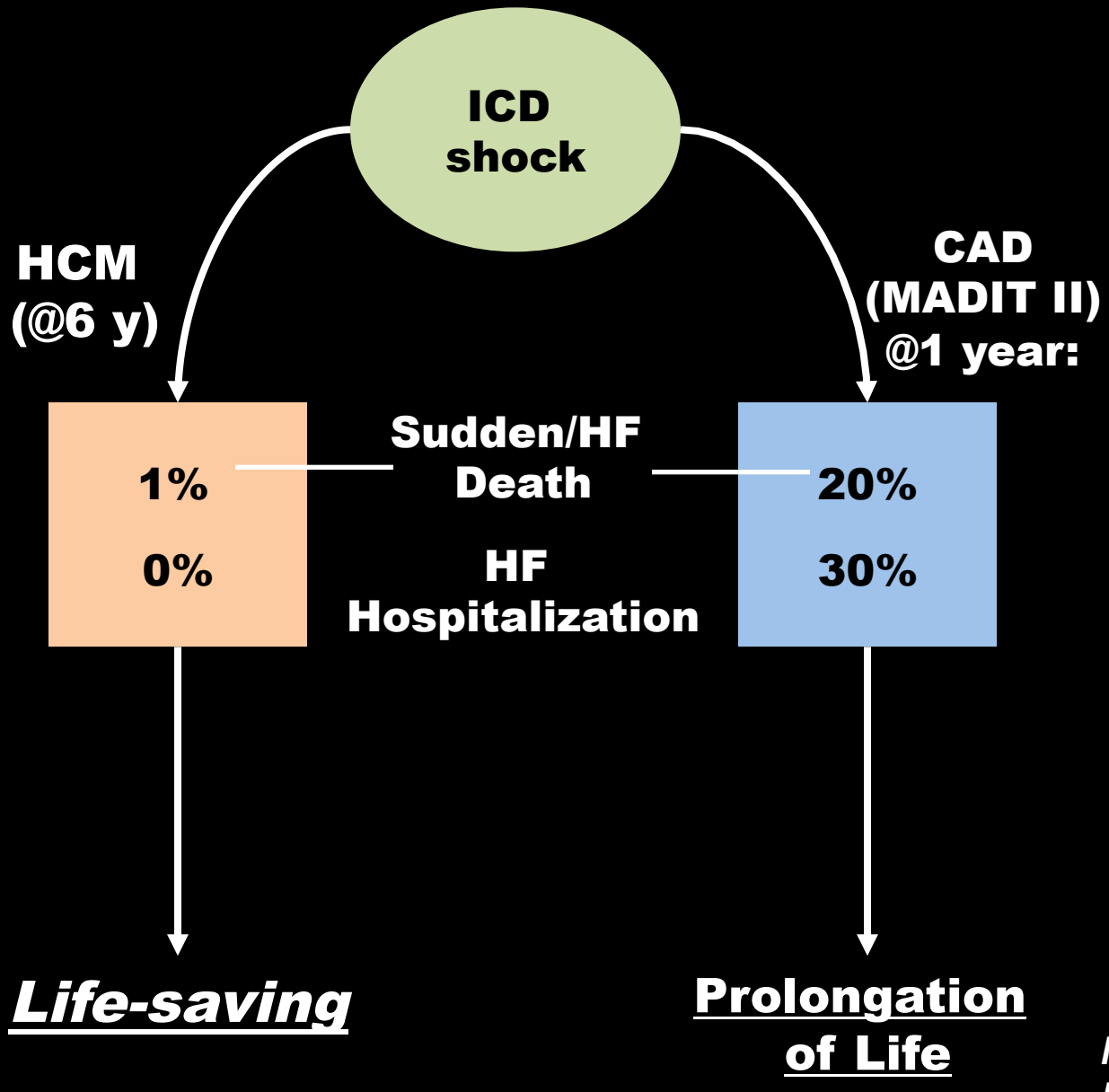
HCM is *Unpredictable*

HCM—ICD Registry



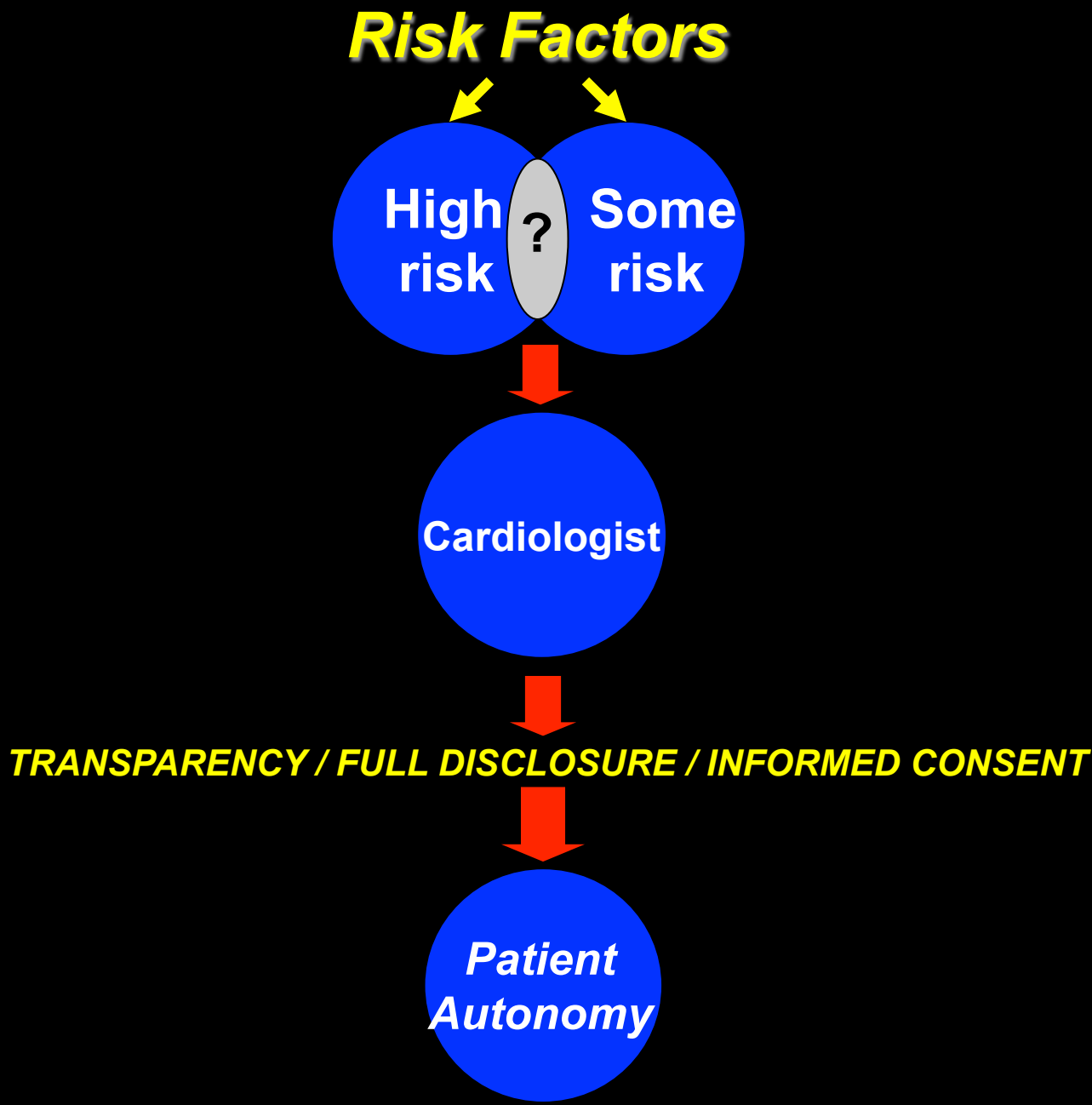
Maron, BJ et. al. JAMA 2007;298:405

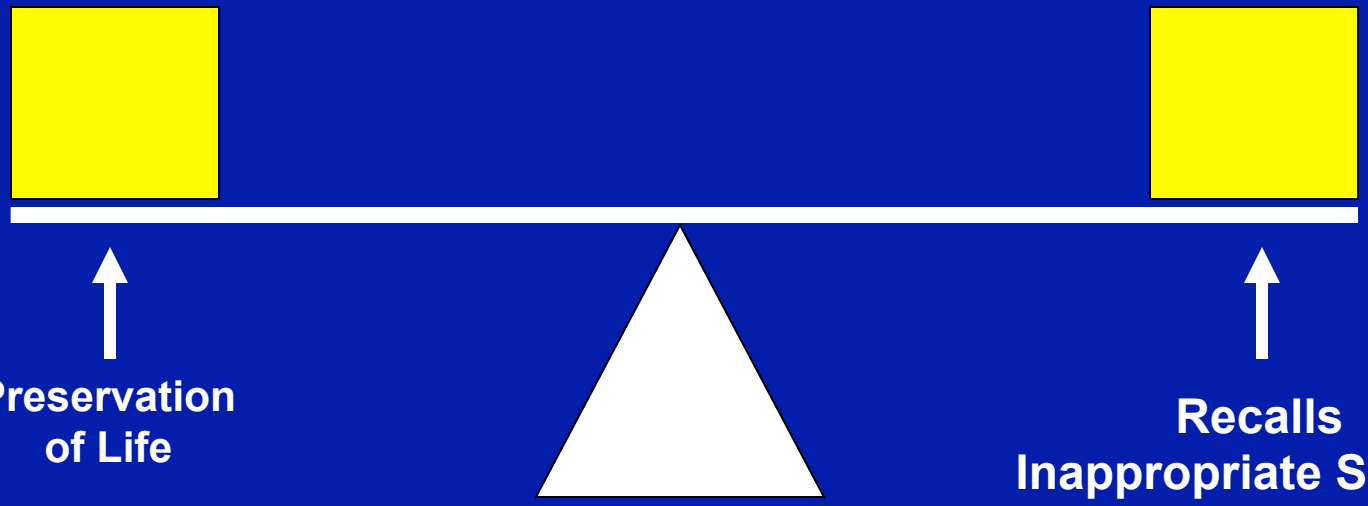
Expectations For HCM Patients After ICD Shocks

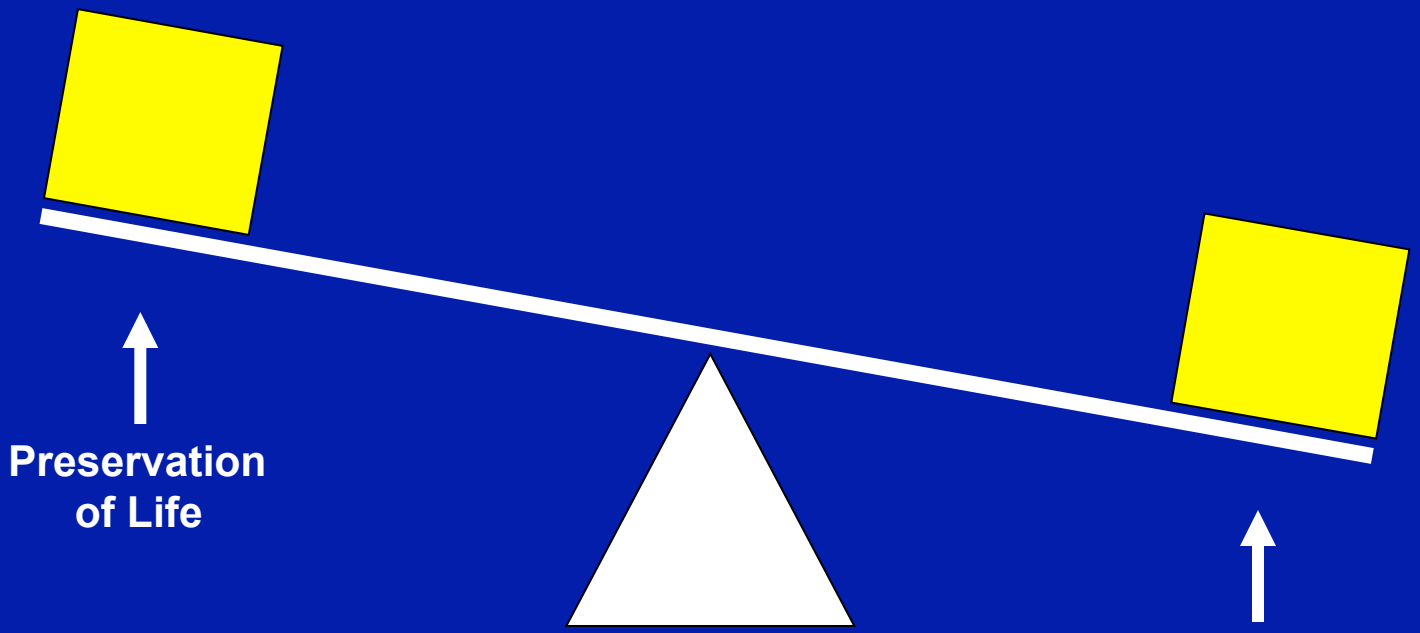


Maron BJ et al In press

Primary Prevention Decision Tree: ICD In HCM




Preservation
of Life
Recalls
Inappropriate Shocks
Infection
Thrombosis
Lead Complications
(5%/y)


Preservation
of Life
Recalls
Inappropriate Shocks
Infection
Thrombosis
Lead Complications
(5%/y)

Contemporary CV treatment options offer HCM patients a reasonable aspiration for reduced mortality and extended longevity. The ICD has altered clinical course for many patients creating the possibility of normal life expectancy even for those @ high risk .

Decrease in Annual HCM Mortality Over 50 Years

4-6%

(1975)

↓

1.5-2.0%

(2000)

↓

0.5%

(2017)