

How Hypertrophic Cardiomyopathy Became a Contemporary Treatable Genetic Disease With Low Mortality Shaped by 50 Years of Clinical Research and Practice

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Disclosures :
Medtronic (Grantee)
GeneDx (Consultant)

2° prevention

Cardiac arrest/sustained VT

1° prevention

Family history HCM-SD

Unexplained syncope

Multiple-repetitive NSVT (Holter)

Abnormal exercise BP response

LGE $\geq 15\%$ of LV mass

Massive LVH ≥ 30 mm



Rare subgroups/potential arbitrators

End-stage (EF < 50%)

LV apical aneurysm

Marked LV outflow obstruction (rest)

Modifiable

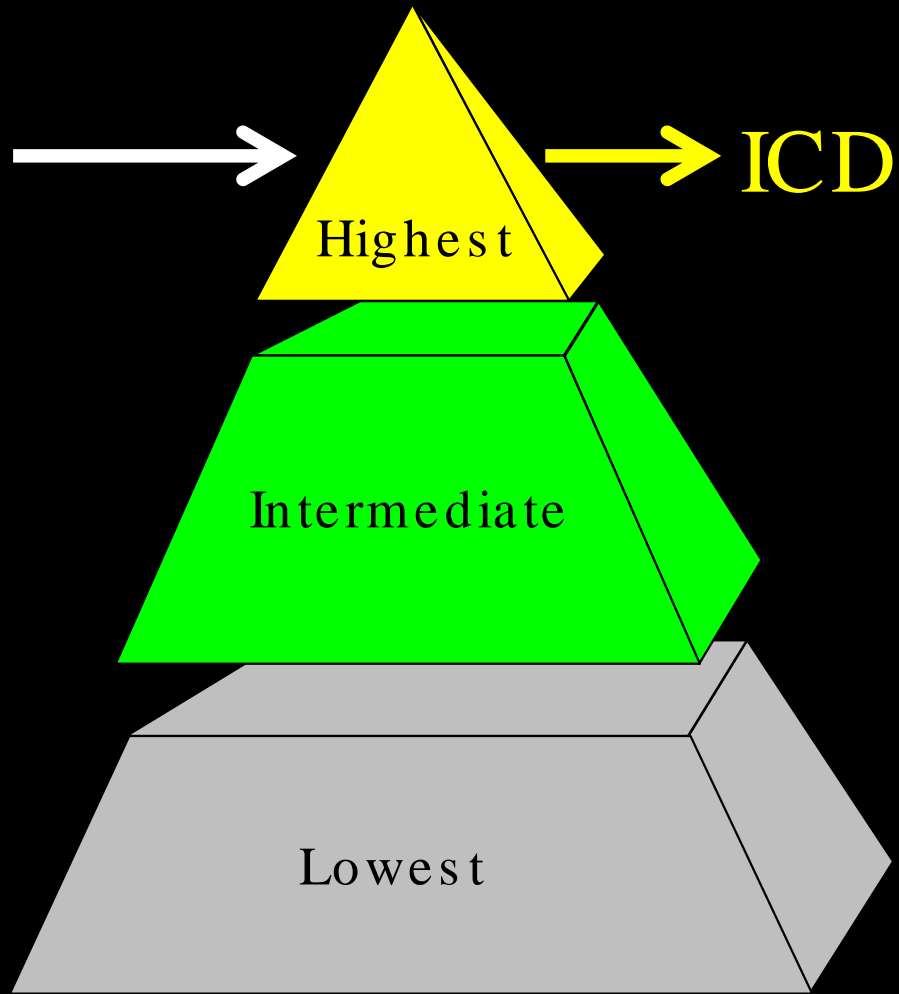
Intense competitive sports

CAD

LGE $\geq 15\%$ of LV mass

Age ≥ 60 y

Alcohol septal ablation (?)



U.S./Canada: ACC/AHA: 2011

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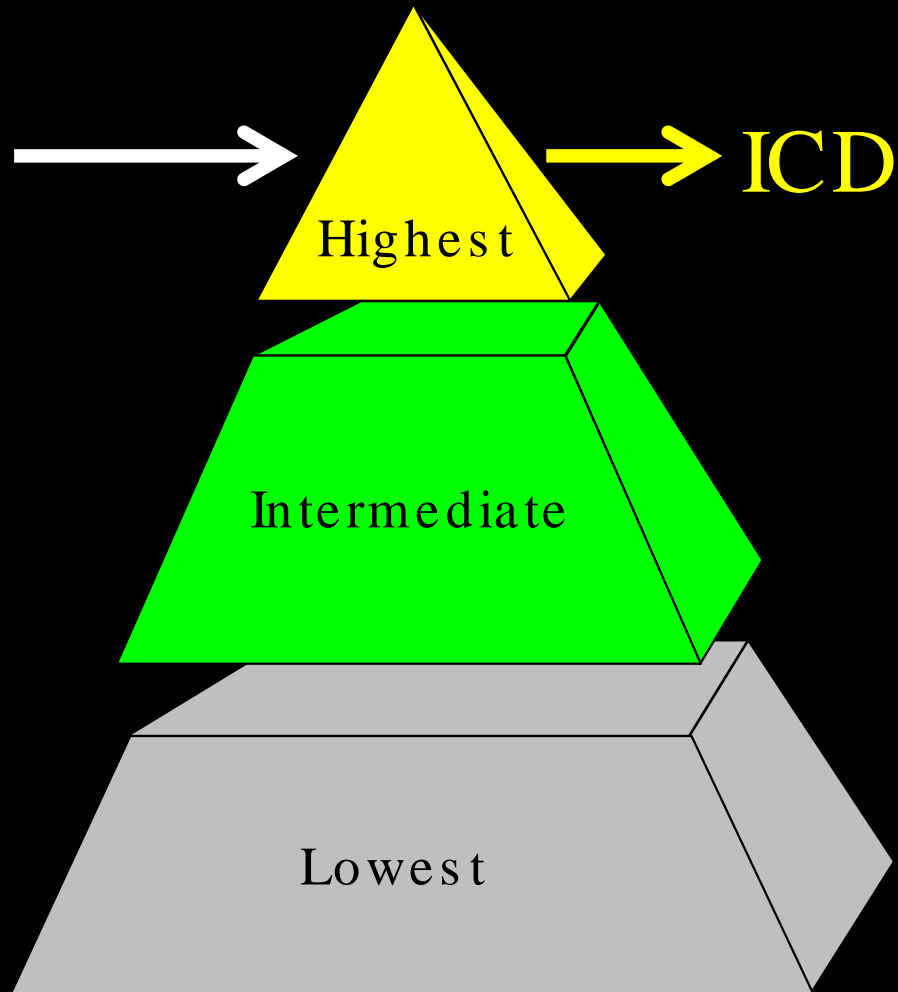
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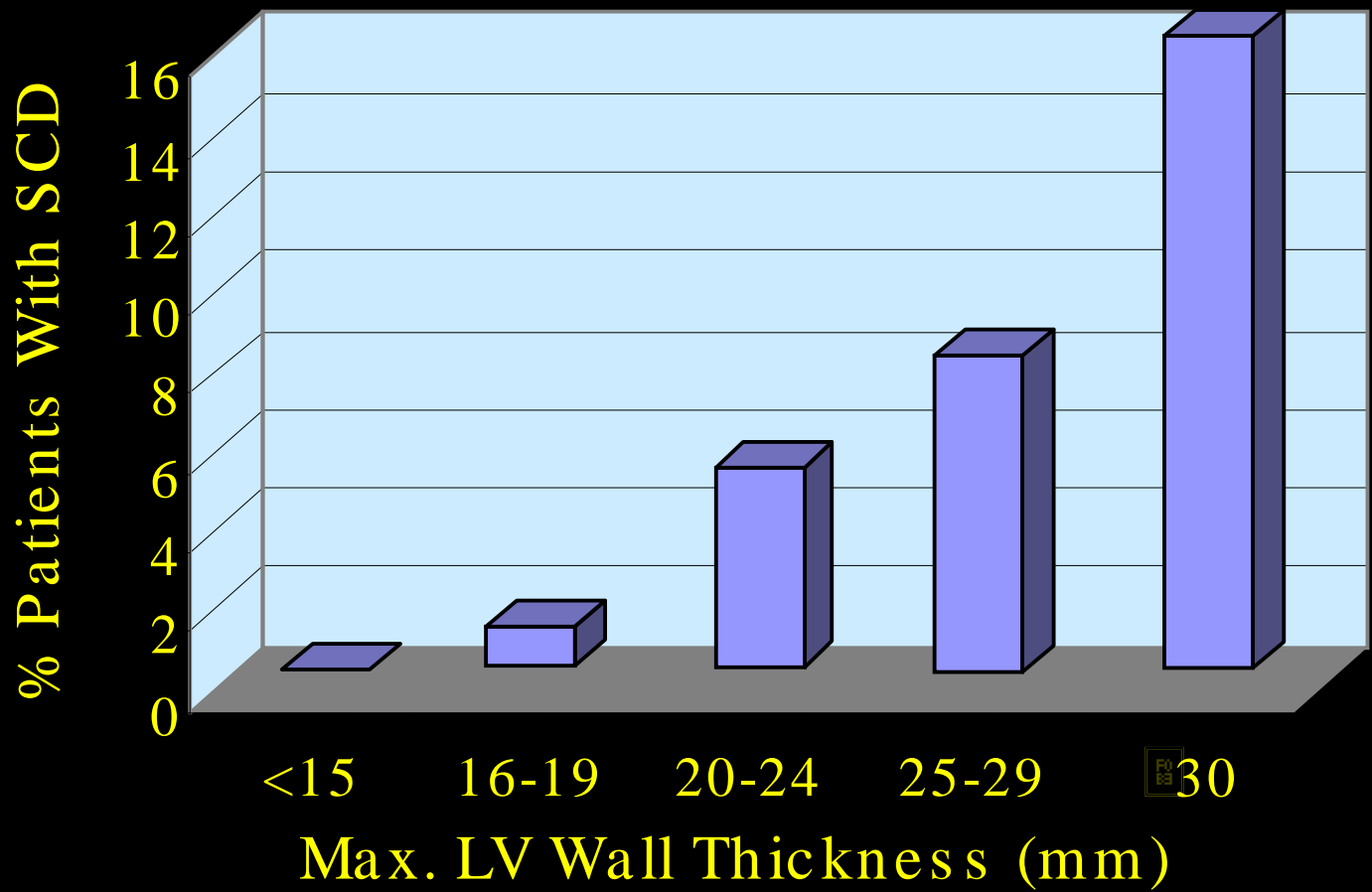


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Relation Between LV Thickness & SCD in 482 HCM Patients



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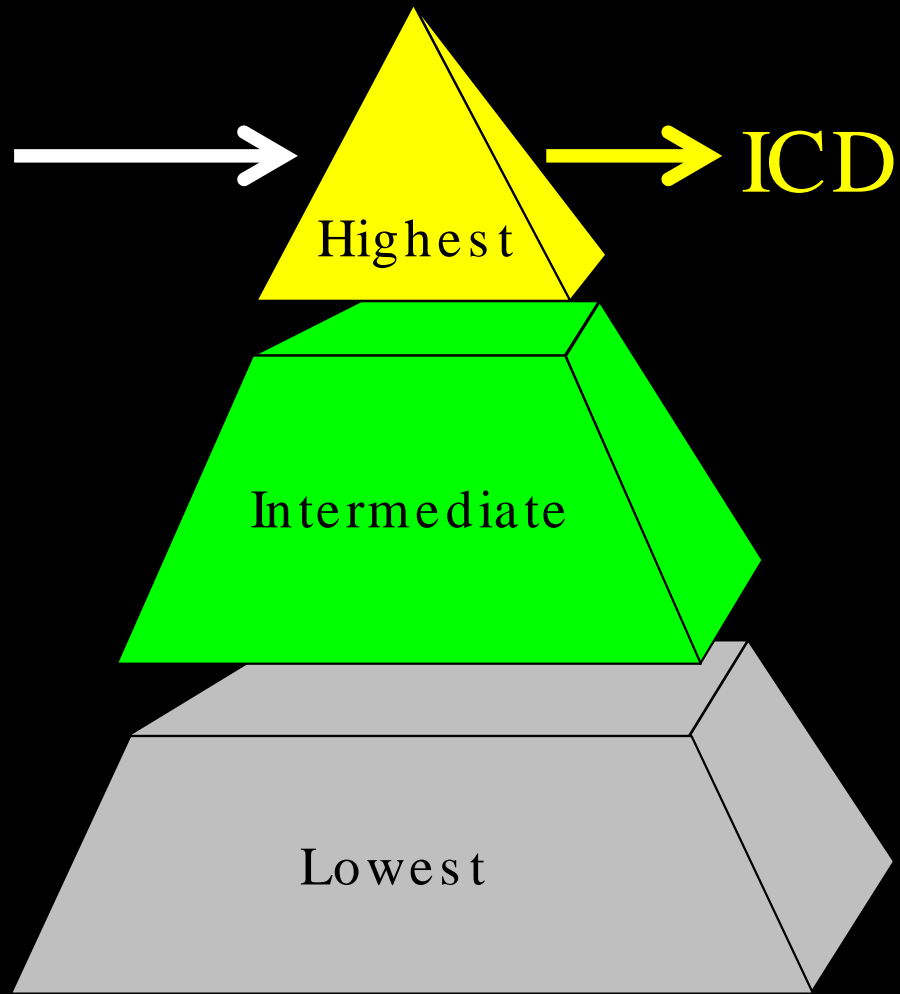
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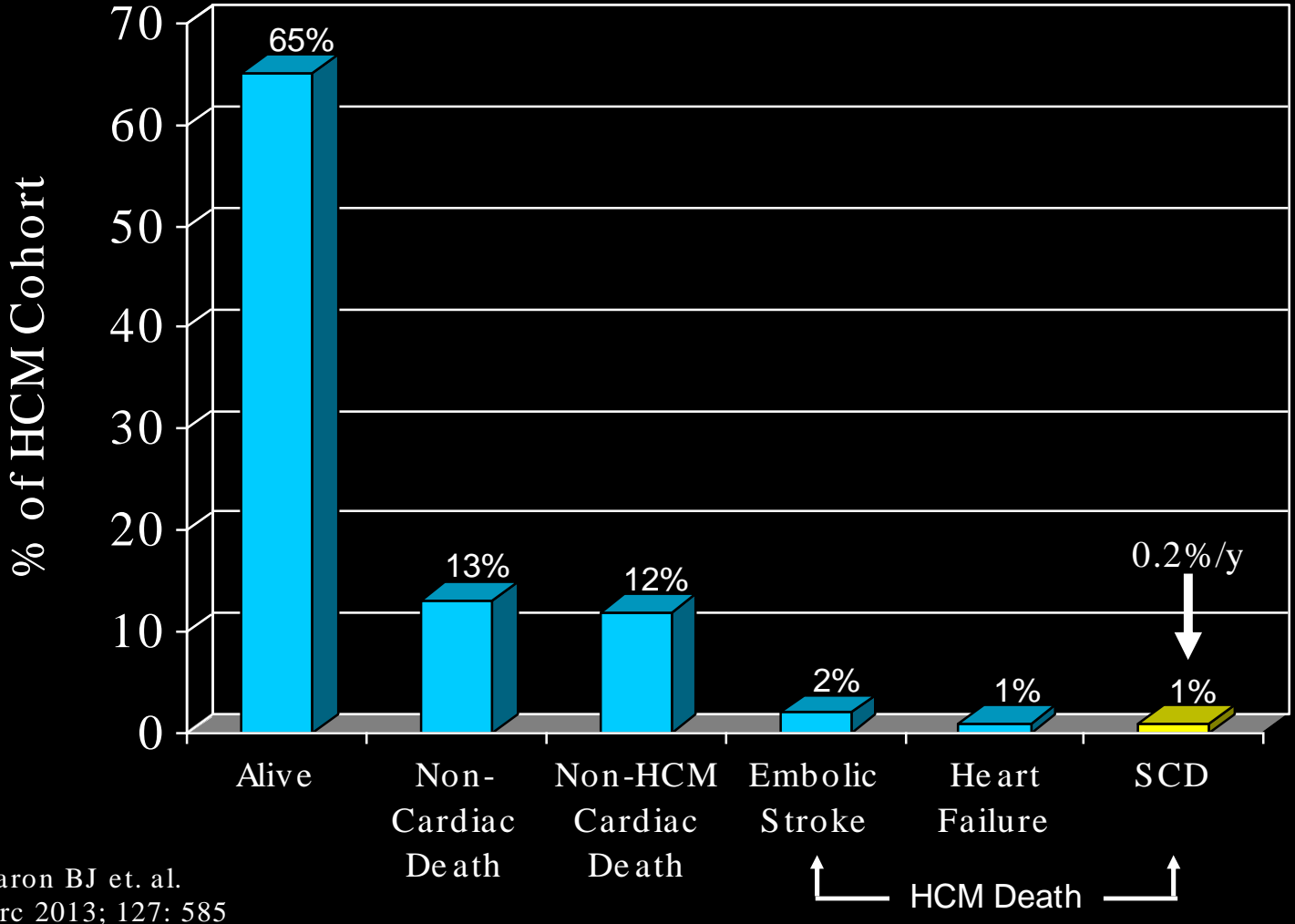
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Outcome of HCM Patients First Evaluated ≥ 60 Years

Aging is Good in HCM



Maron BJ et. al.
Circ 2013; 127: 585

Risk Stratification for Sudden Death in HCM

Family history of sudden death

Extreme LVH

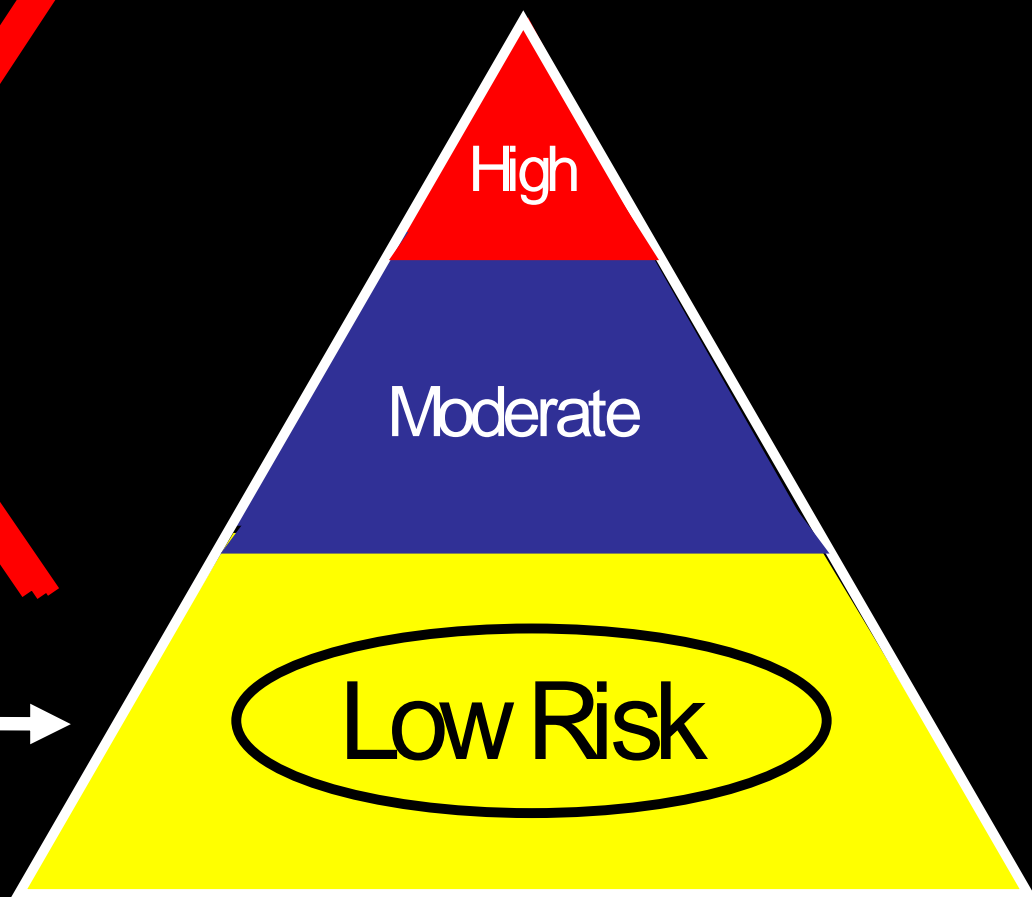
Nonsustained VT

Unexplained syncope

Abnormal BP response to exercise

No risk factors

0.5%/year



U.S./ Canada (ACC/AHA) 2011

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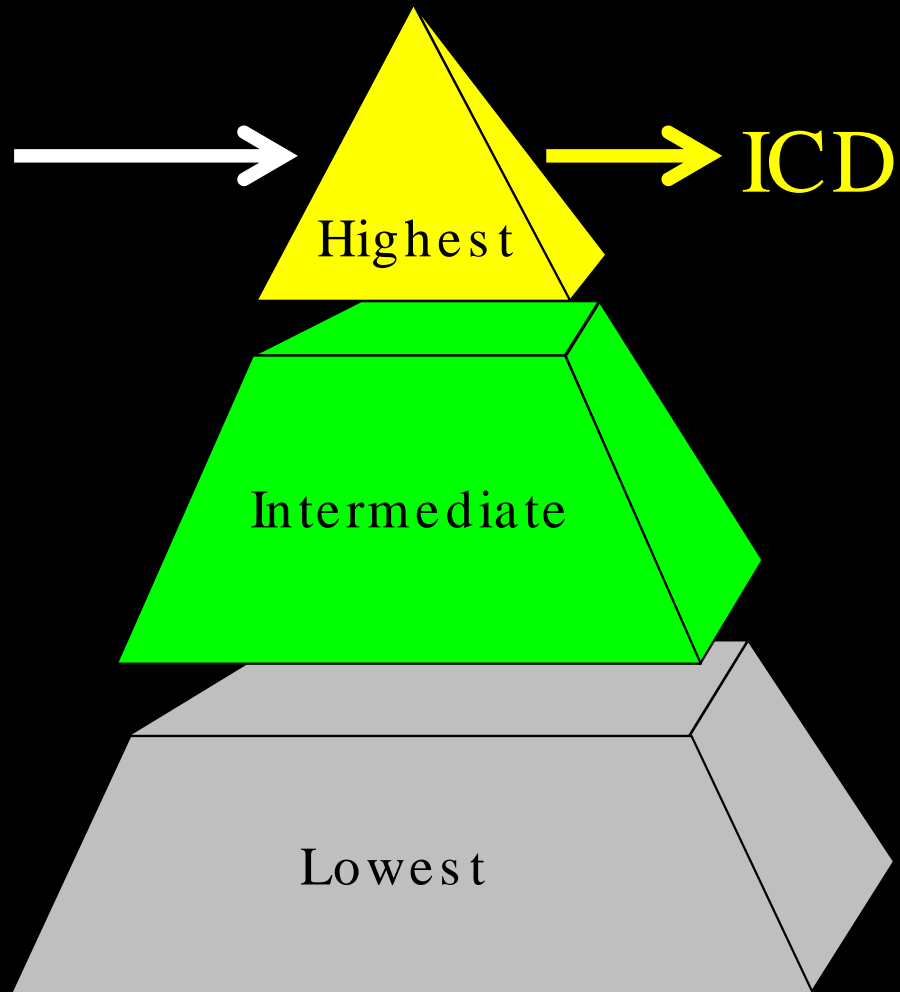
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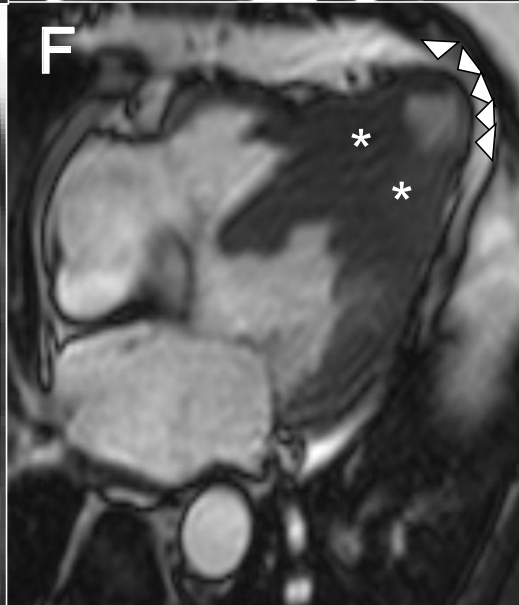
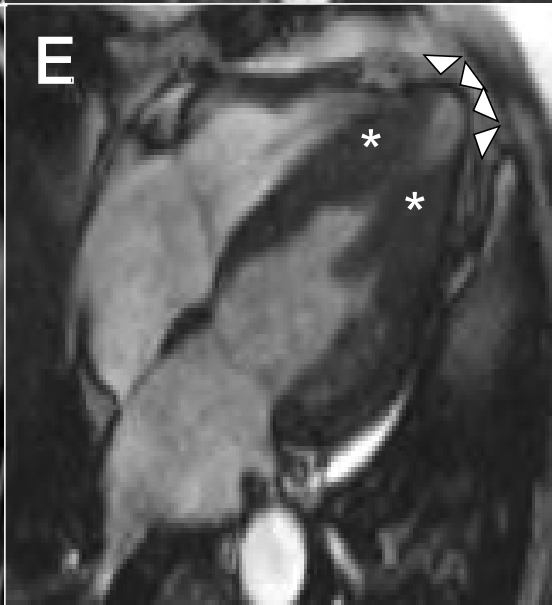
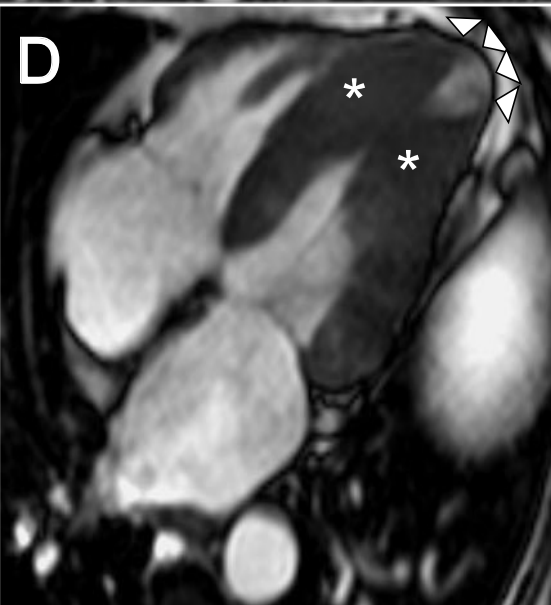
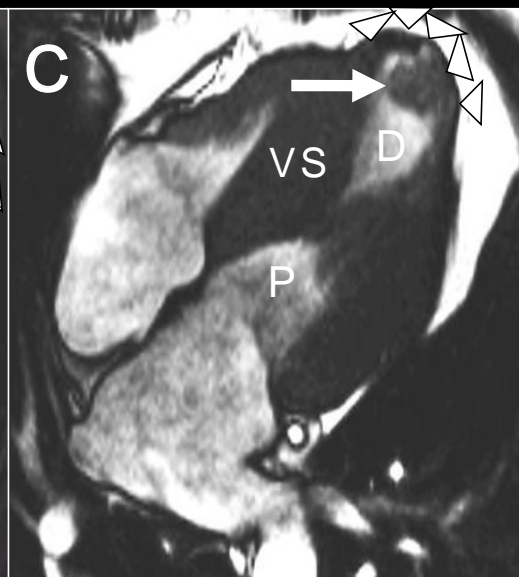
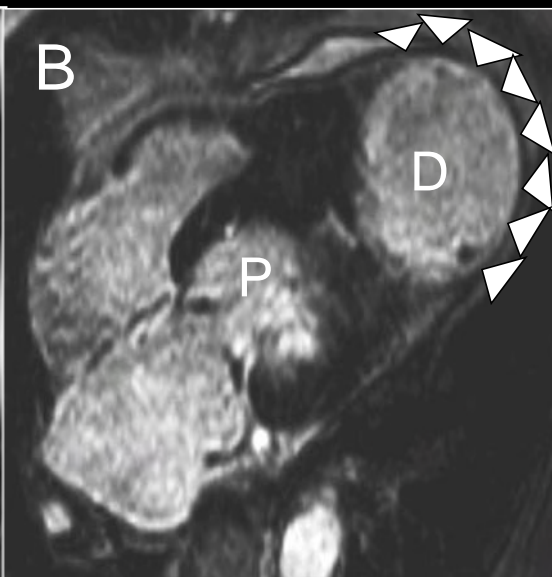
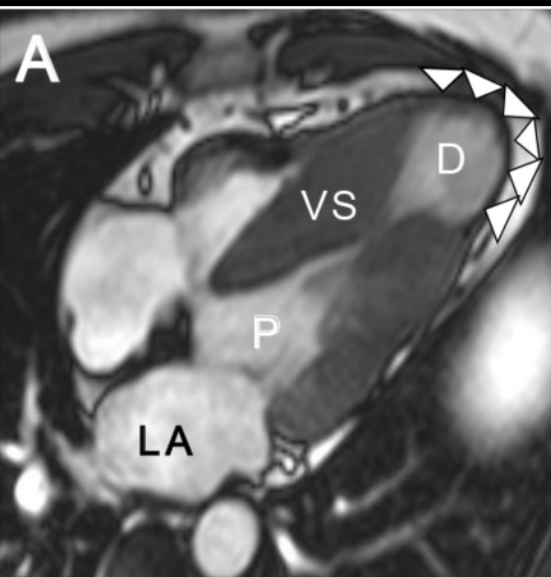
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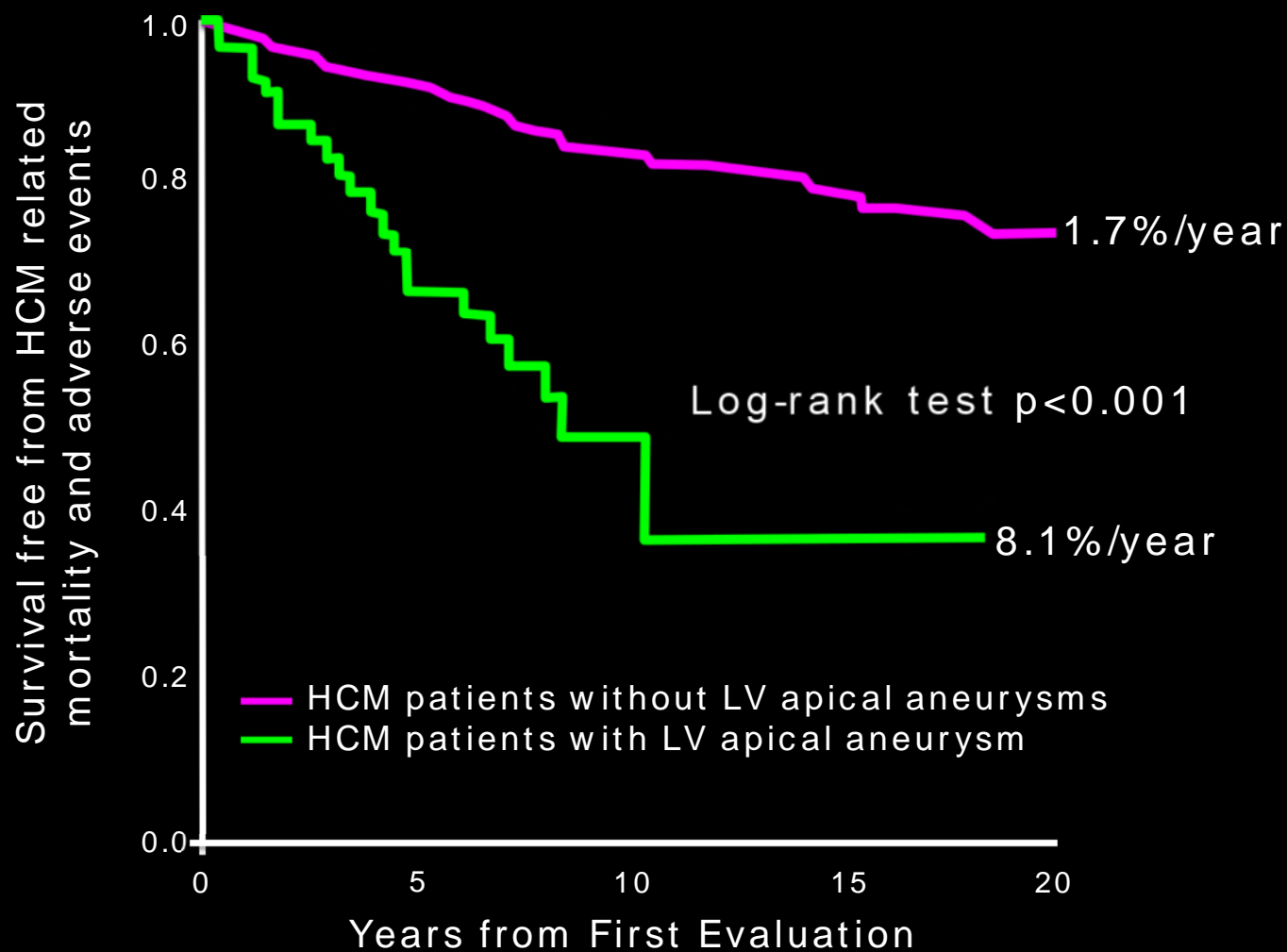
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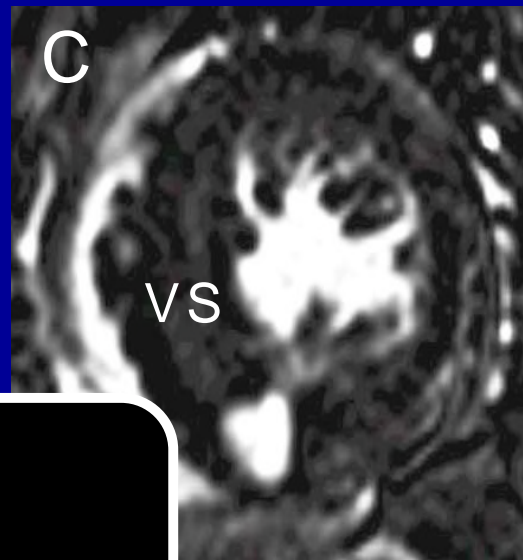
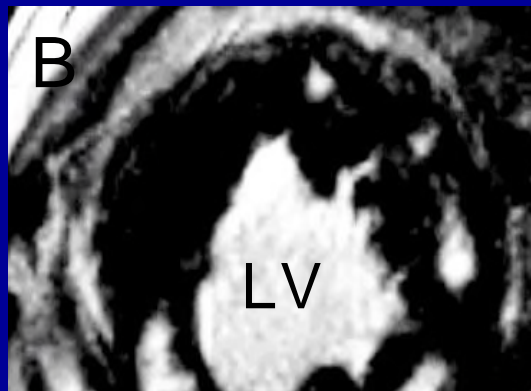
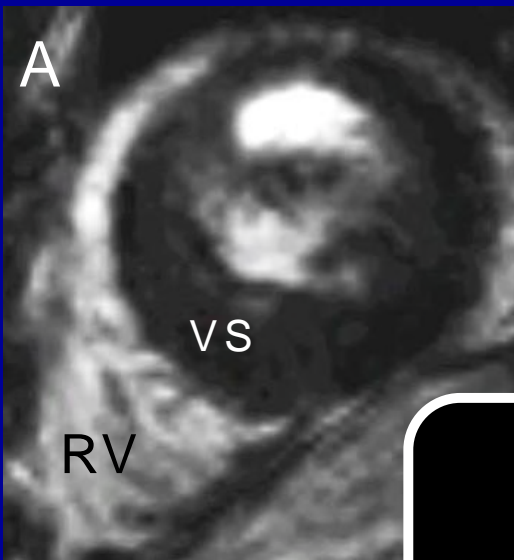
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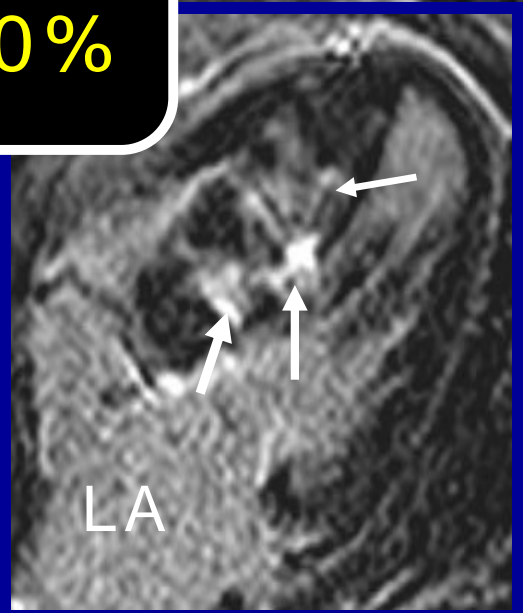
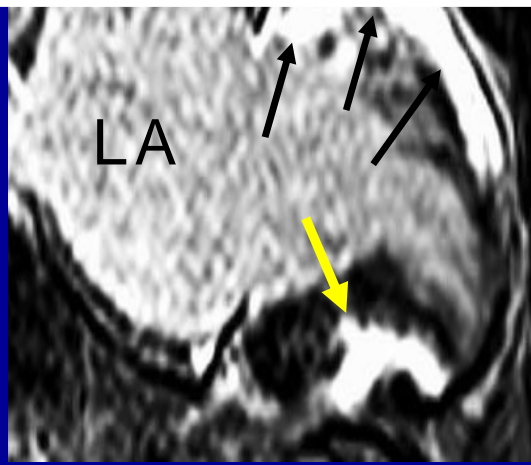
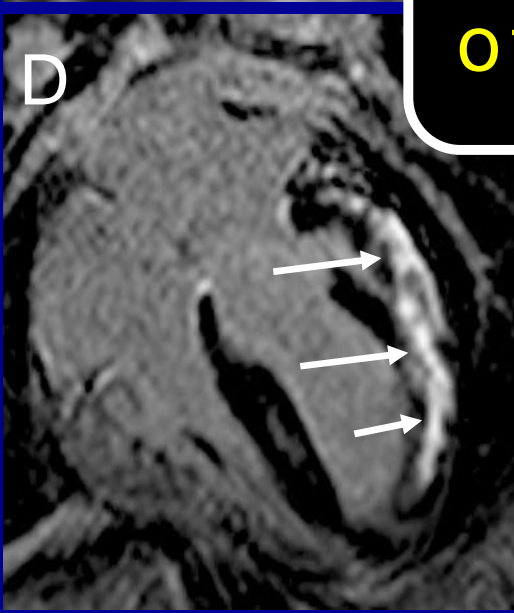


HCM Related Death or Adverse Clinical Events in 93 Patients with LV Apical Aneurysms

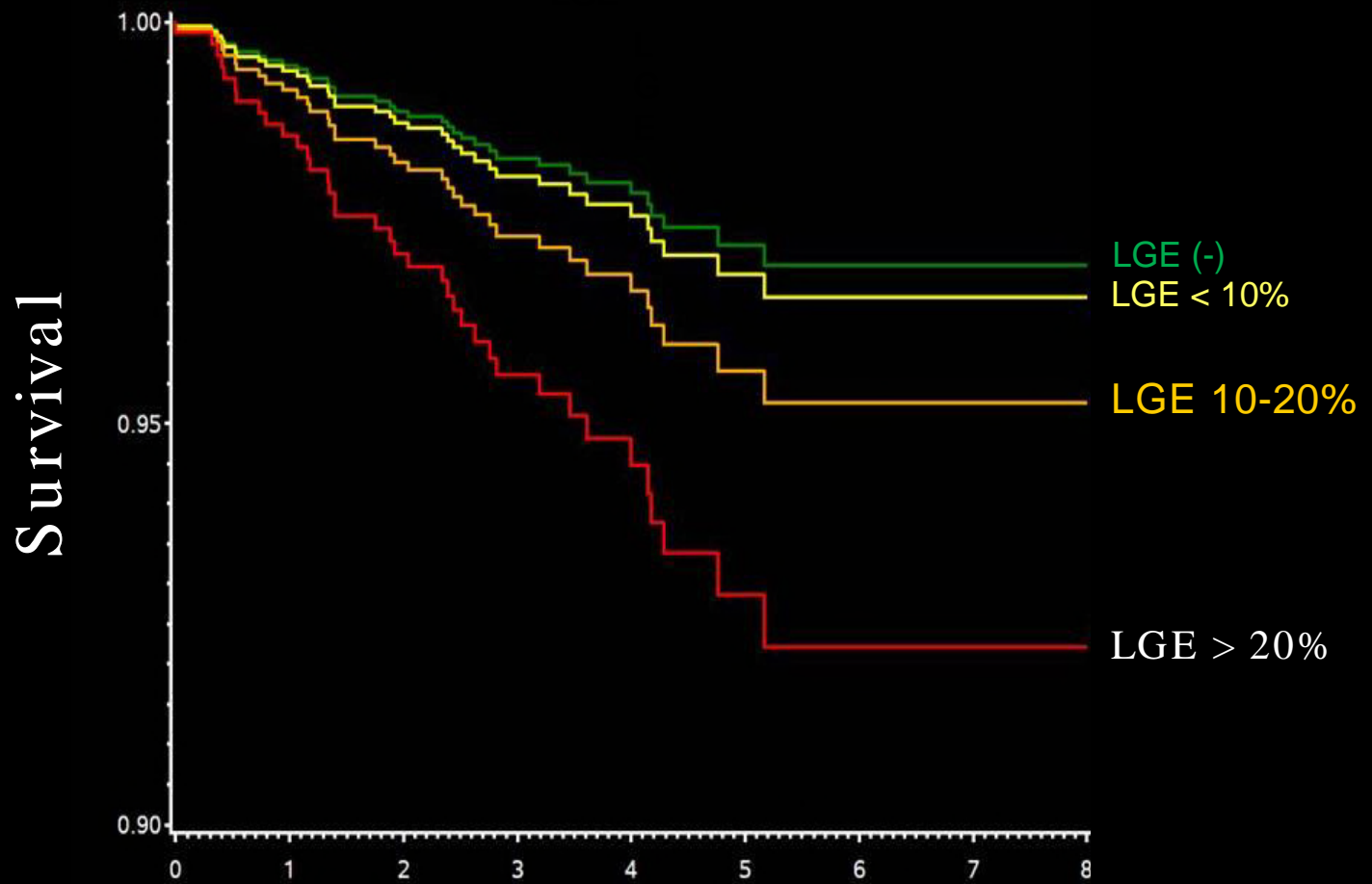




**Prevalence
of LGE = 55-70%**



Extent of LGE vs. Sudden Death Risk in HCM



Chan RH et. al.
Circ 2014; 130(6):
484-95

Follow-up (years)

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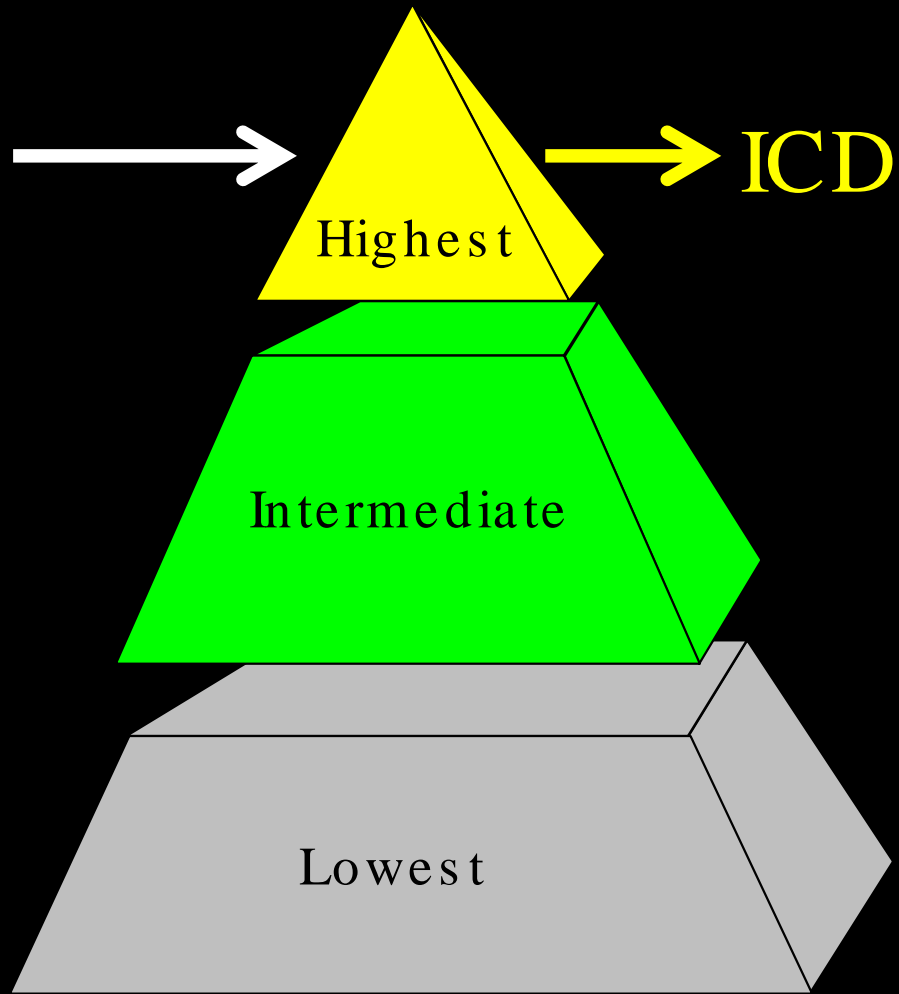
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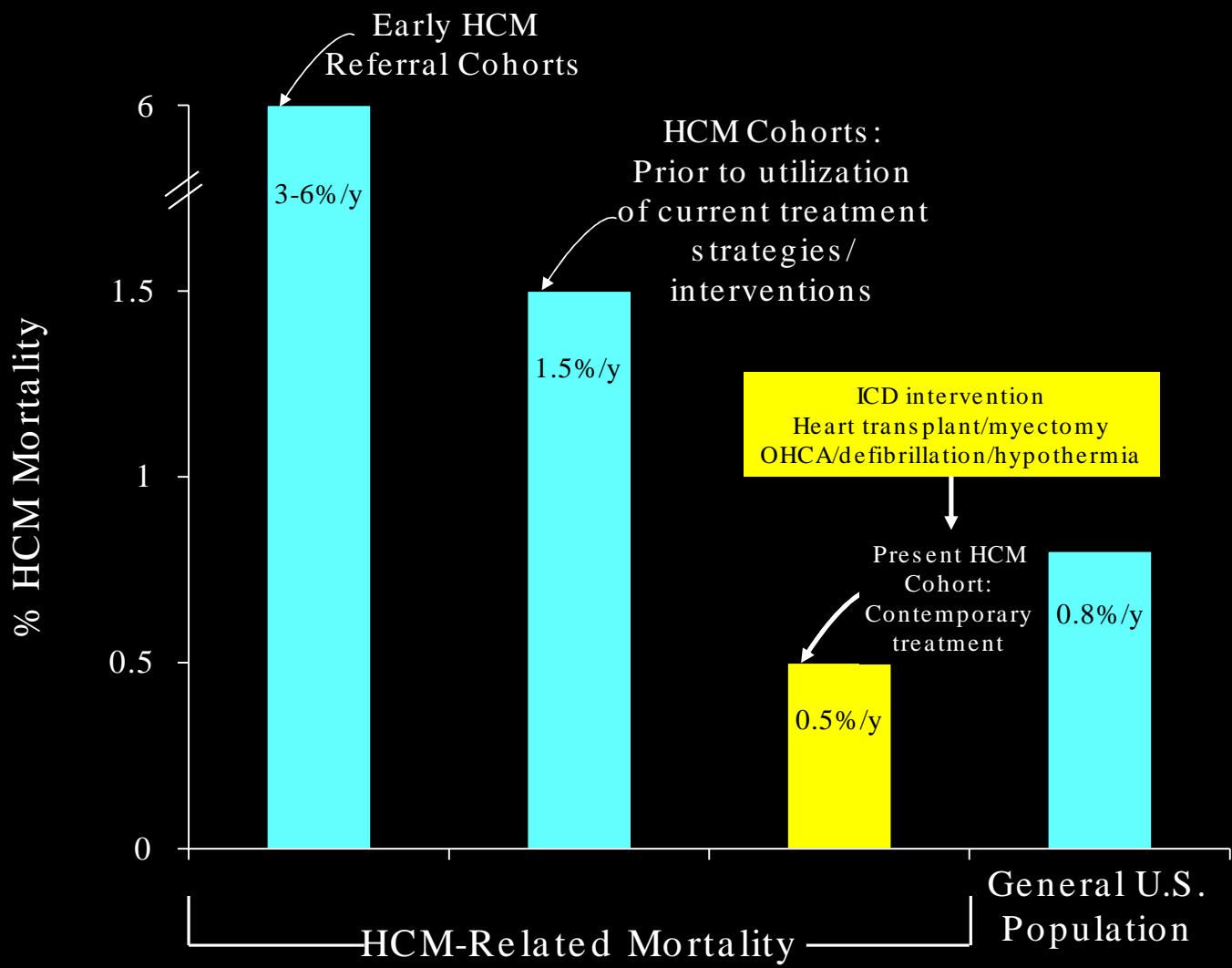
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Benign/Stable
(normal longevity)

Profiles in Prognosis for HCM

Sudden
Death

ICD

Progressive
Heart
Failure
(obstructive)

Drugs

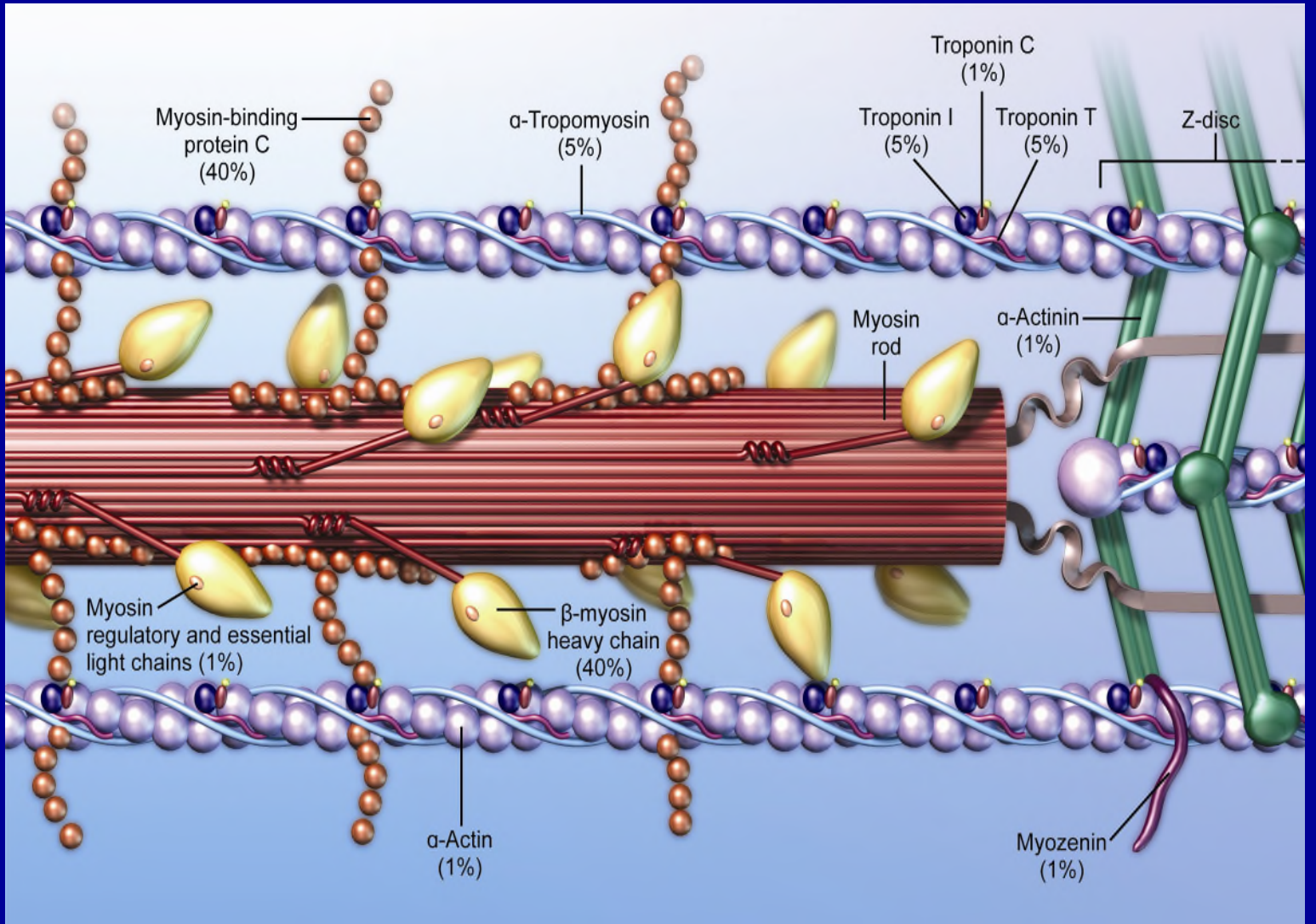
Septal Myectomy
(Alcohol Ablation)

Advanced
Heart Failure
& End Stage
(non-
obstructive)

Transplant

AF
&
Stroke

Drugs
Anticoagulants
Ablation



25-Year Contemporary Initiatives in Hypertrophic Cardiomyopathy

Genetic (molecular)
Single sarcomere mutation
hypothesis

"Clinicians"



Lives Saved → 0

Thousands

Improved Quality of Life → 0

Many thousands