



Hypertrophic Cardiomyopathy

A Legacy of Clinical Acumen Embraced by Modern Medical Science

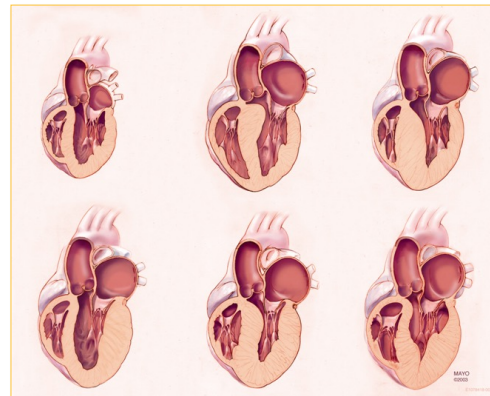
Steve R. Ommen, MD FACC
Professor of Medicine
Director, Mayo Hypertrophic Cardiomyopathy Clinic

No Disclosures

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Hypertrophic Cardiomyopathy



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Presentation

Cardiac symptoms (often varying from day to day) in young patients or in patients without other CV disease

May be asymptomatic with or without a murmur

ECG is abnormal in 95%
(LVH and/or ST-T)

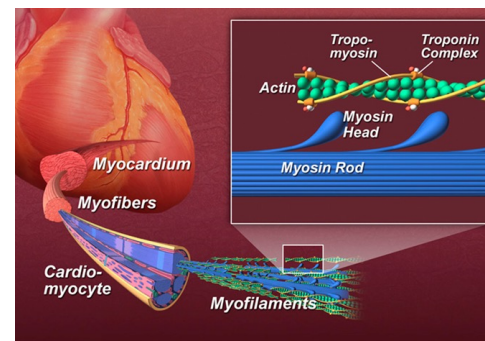
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HCM Micro-pathophysiology

Excess active myosin-actin bonds

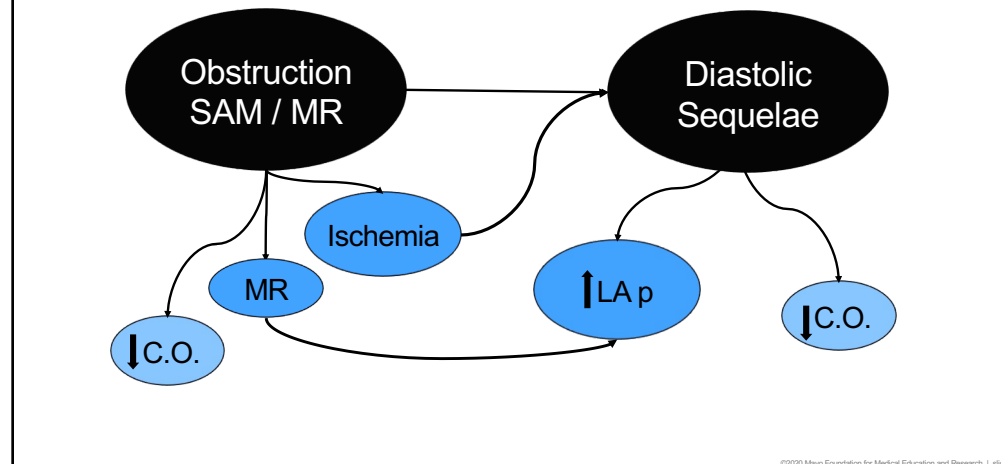
- Metabolic/Energetic abnormalities
- Hypercontractility
- Impaired relaxation
- Disordered sarcomeres
- Fibrosis



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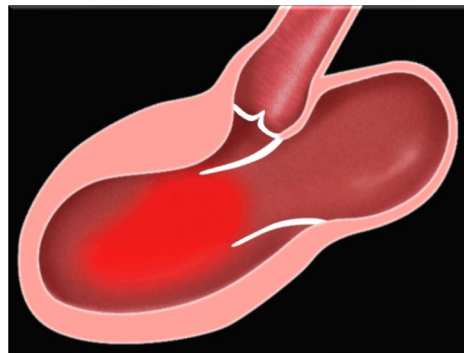
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Macro-Pathophysiology of HCM



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Macro-Pathophys = Dynamic Obstruction

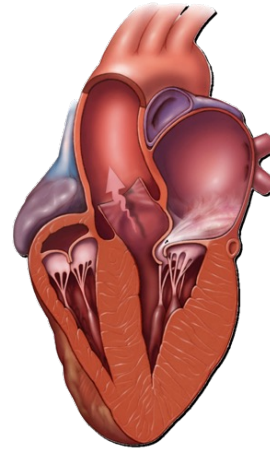


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Obstruction Gets Worse If...

- More Vigorous Contraction
- Decreased Resistance
- Decreased Volume

All of these occur
with physical exertion



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HCM Management Pathways

Family

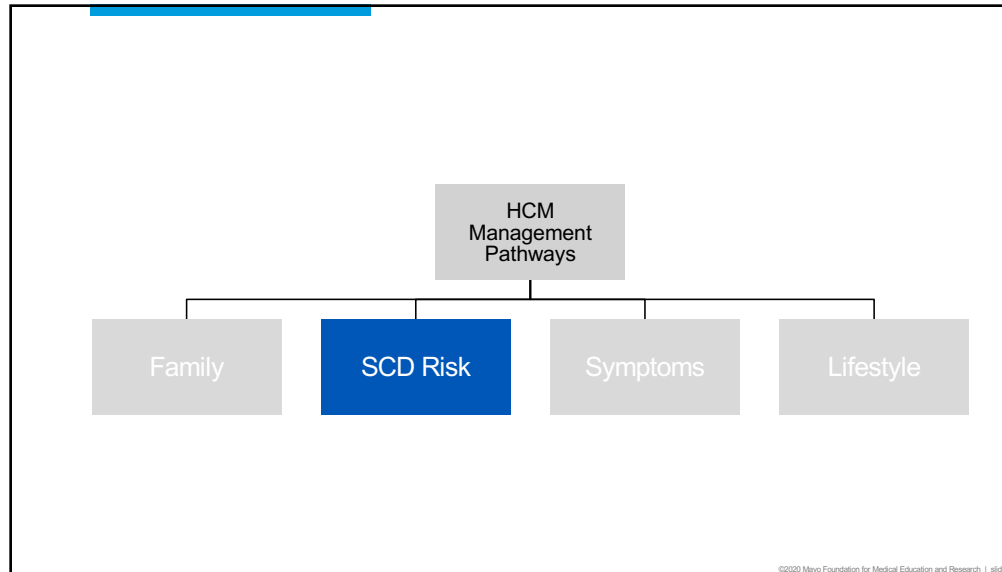
SCD Risk

Symptoms

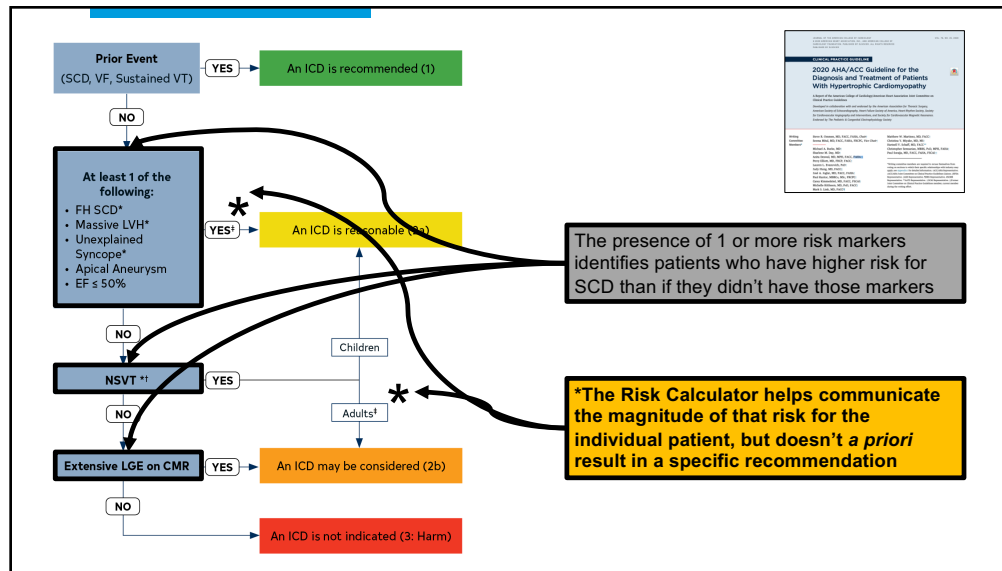
Lifestyle

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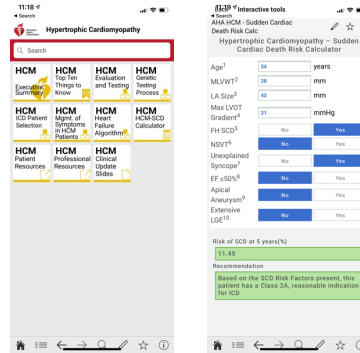


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Interactive Figures and SCD Risk Calculator



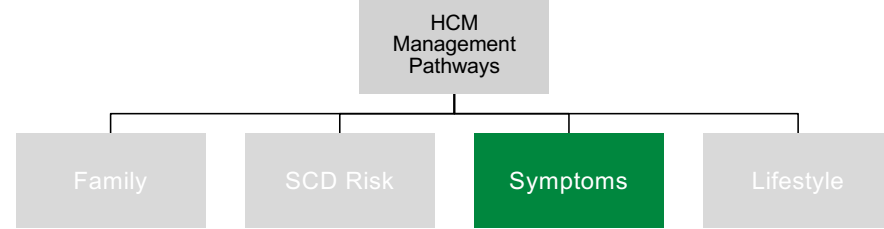
AHA Guidelines On the Go



ACC Guideline Clinical App

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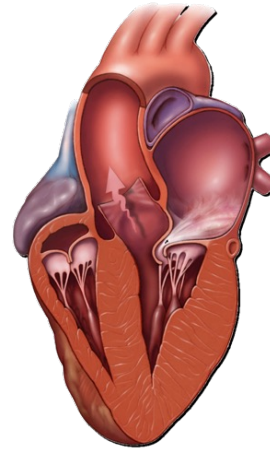
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Obstruction Gets Worse If...

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HCM Management Pathways

Family

SCD Risk

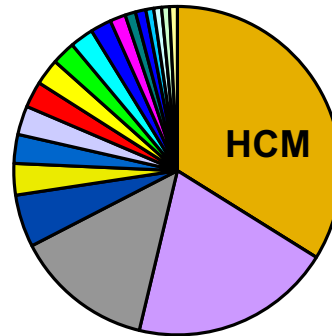
Symptoms

Lifestyle

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Causes of SCD in Athletes

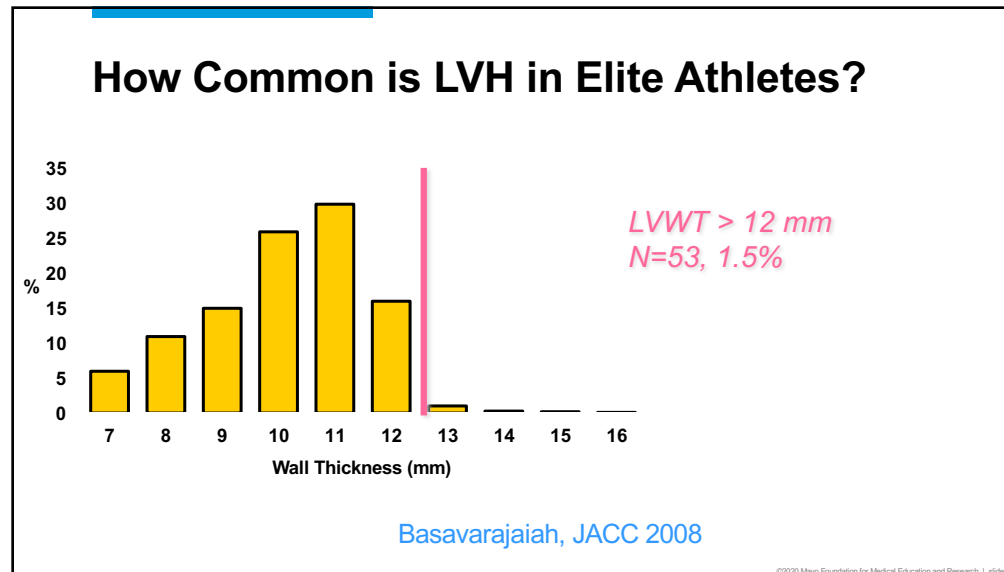


Maron, NEJM: 349(11) 11 September 2003 pp 1064-1075

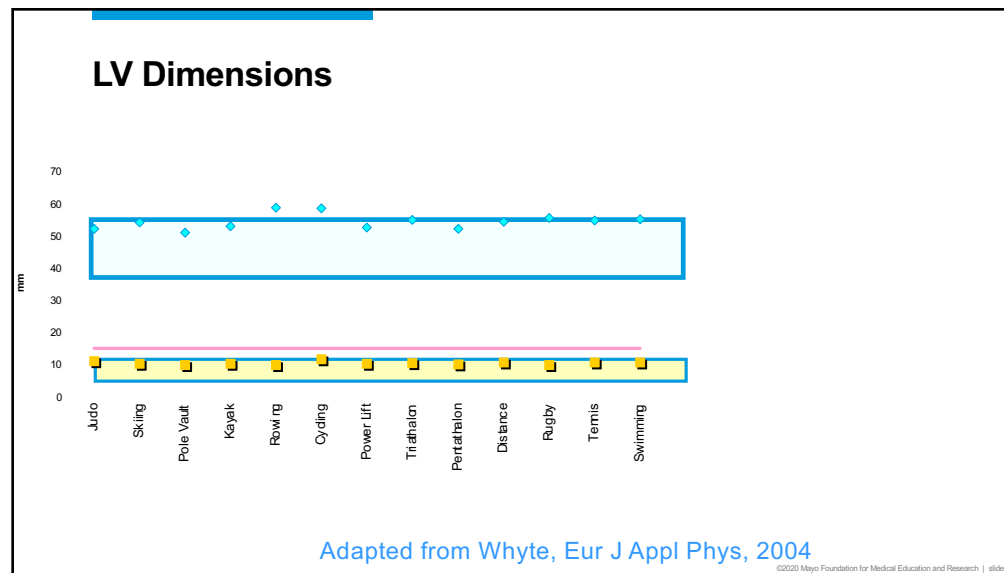
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HCM or Athletic Adaptation?

HCM

- Unusual LVH pattern
- LVEDD <45
- Marked LAE
- Bizarre ECG
- Abnormal LV filling
- Reduced TDI velocity
- Female

Athletic Heart

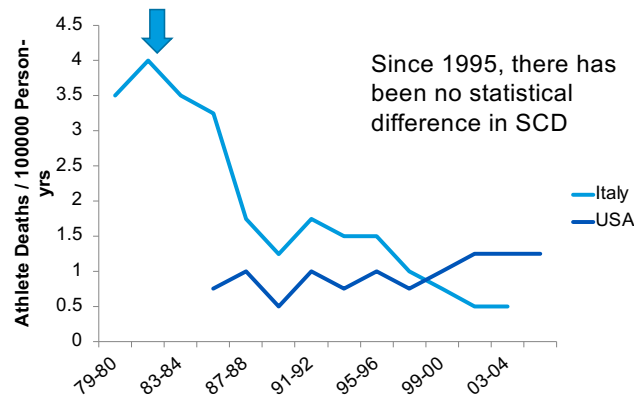
- LVEDD >55
- Responds to de-training
- Super-normal exercise capacity
- No MRI scar tissue or perfusion defects

Adapted from Maron: Heart 2005; 91:1380

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Trends in SCD rates 1979-2004



Maron et al. Am J Card. 2009.

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Christian Eriksen returns to Denmark squad for 1st time since collapse

Denmark plays a friendly away against the Netherlands on March 26 and hosts Serbia on March 29. Denmark prepares for the World Cup in Qatar in November.




Keyontae Johnson 2020 collapse



All underwent cardiac screening

Courtesy of Dr. Matthew Martinez



Bronny James

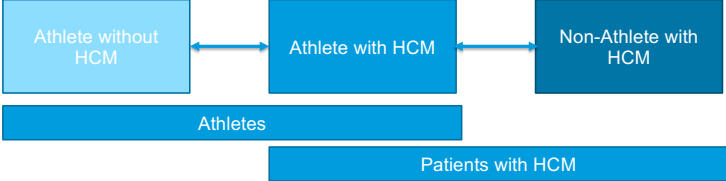


Damar Hamlin

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How risky is it to be an athlete with HCM?



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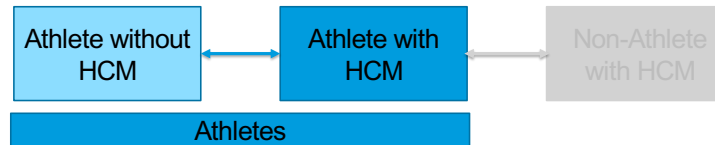
graph LR
    A[Athlete without HCM] <--> B[Athlete with HCM]
    B <--> C[Non-Athlete with HCM]
    subgraph Athletes
        A
        B
    end
    subgraph Patients_with_HCM
        B
        C
    end
            
```

To determine the risk of competitive athletics, comparisons can be made to other athletes (e.g. those without HCM), and/or to other individuals with HCM who are not athletes

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How risky is it to be an athlete with HCM?



Among athletes who die suddenly,

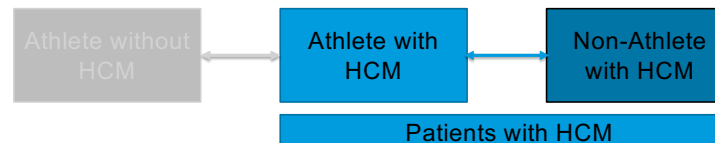
- HCM is the attributable cause in 36%
- 36% is far higher than the prevalence of HCM
- **Therefore, being an athlete with HCM is higher risk than being an athlete without HCM**

N.B. Being a human with HCM is higher risk than being a human without HCM

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How risky is it to be an athlete with HCM?



If being an athlete with HCM is higher risk than being a non-athlete with HCM, then the incidence of SCD should be higher for those individuals with HCM who participate in sports

Let's compare rates of expected vs observed SCD in athletes with HCM and see if it differs from the expected rate of SCD in the general population (0.8%)

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How risky is it to be an athlete with HCM?



Model 1

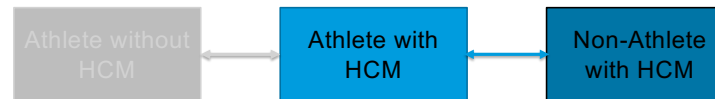
Patients with HCM

NCAA athletes per year =	400000
SCD Rate among NCAA athletes =	0.002%
Predicted deaths among NCAA athletes =	8
36% should be attributable to HCM =	2.88
Observed HCM-related SCD in athletes =	4 (Maron et al)

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How risky is it to be an athlete with HCM?



Model 2

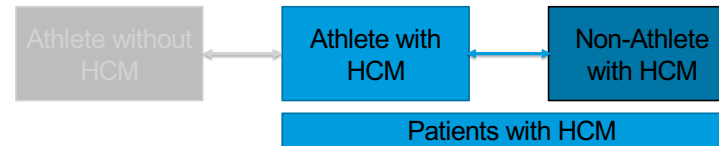
Patients with HCM

NCAA athletes per year =	400000
Prevalence of HCM among NCAA athletes =	0.08%
NCAA athletes with HCM =	320
SCD Rate in HCM =	0.8%
Expected deaths from HCM SCD =	2.56
Observed HCM-related SCD in athletes =	4 (Maron et al)

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How risky is it to be an athlete with HCM?



Are 3 or 4 deaths/year different than the baseline rate of SCD in the HCM population?

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Sample Size Calculation

- Compare rate of SCD in general population of patients with HCM to expected and observed rates of SCD among athletes with HCM
- Assume general (non-athlete) population HCM SCD rate = 0.8%
- Power to detect the difference – 80% at $p=0.05$

HCM Deaths in Athletes	Athlete HCM SCD Risk (%)	Non-athlete HCM SCD Risk (%)	N needed in <i>each</i> study group	Anticipated years to complete
3 (predicted)	0.938	0.8	33389	100 years
4 (observed)	1.25	0.8	3550	10+ years

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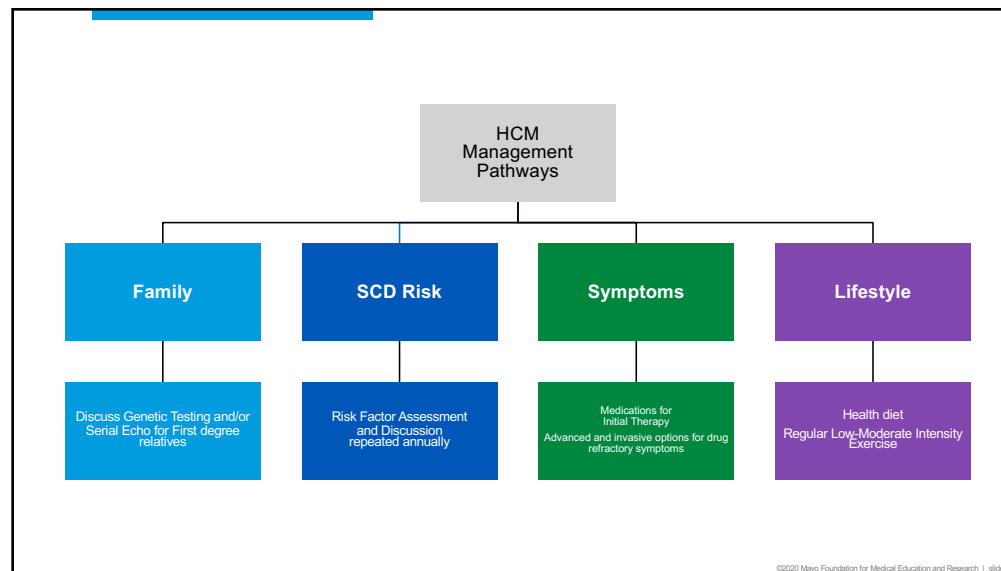
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Exercise in HCM

- LIVE-HCM: Among patients with HCM, those exercising at least 6 METS for more than 60 hours per year, did not experience worse outcomes than those exercising less than that [JAMA Cardiol. 2023 Jun 1;8(6):595-605]
- RESET-HCM: a randomized controlled trial to address exercise training in HCM, demonstrated a significant improvement in fitness with moderate intensity exercise. [JAMA. 2017; 317:1349–1357]

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