

Medical therapy and exercise guidelines in HCM

Athletes, Sudden Death and HCM
Stanford University
June 27, 2009

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The purpose of medical therapy

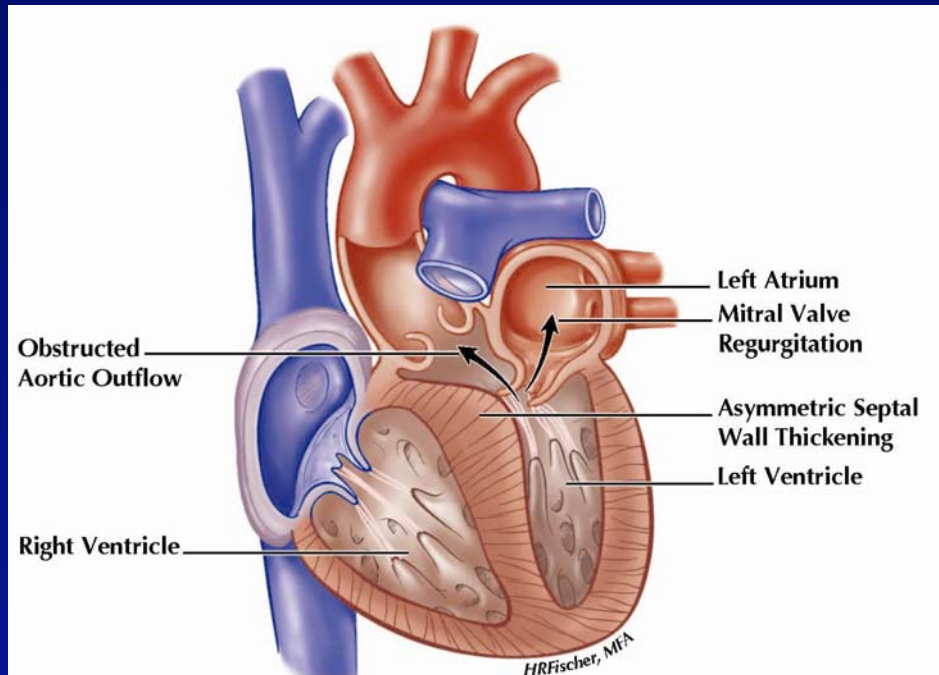
Cure disease → Not yet

Prolong life → ICD, myectomy

Prevent disease progression → ?

Improve quality of life (i.e. treat symptoms)

The principles of treating symptomatic patients with obstruction (>30 mmHg at rest or w/ provocation)



Shortness of breath

Chest pain

Dizziness

Narrow LV outflow tract

Increase size of LV cavity

Increase volume and peripheral resistance (i.e. drink water and avoid high dose diuretics and afterload reducers)

Contraction too forceful

Decrease strength of contraction

Use negative inotropes (i.e. beta blockers, calcium channel blockers, norpace) and avoid positive inotropes (i.e. digoxin)

The principles of treating symptomatic patients without obstruction (<30 mmHg at rest or w/ provocation)

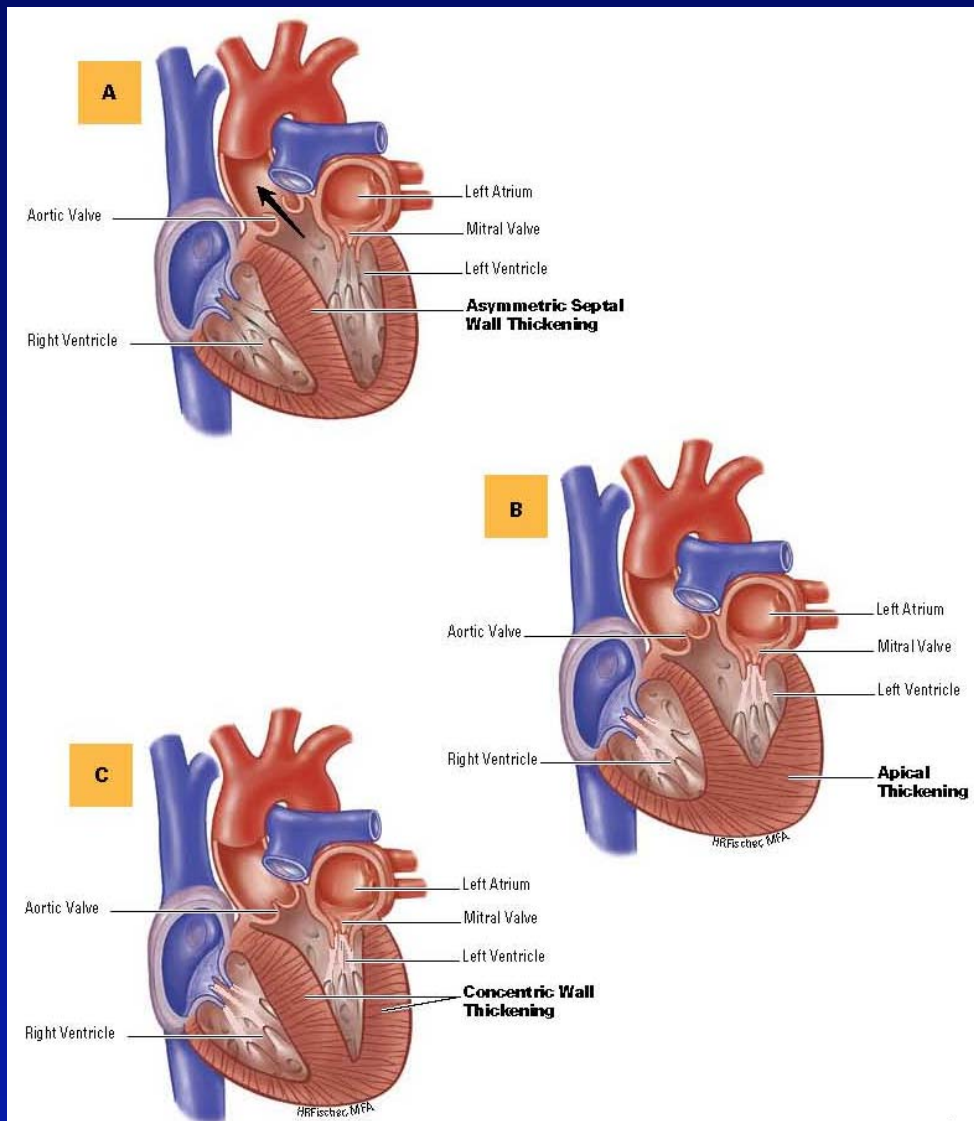
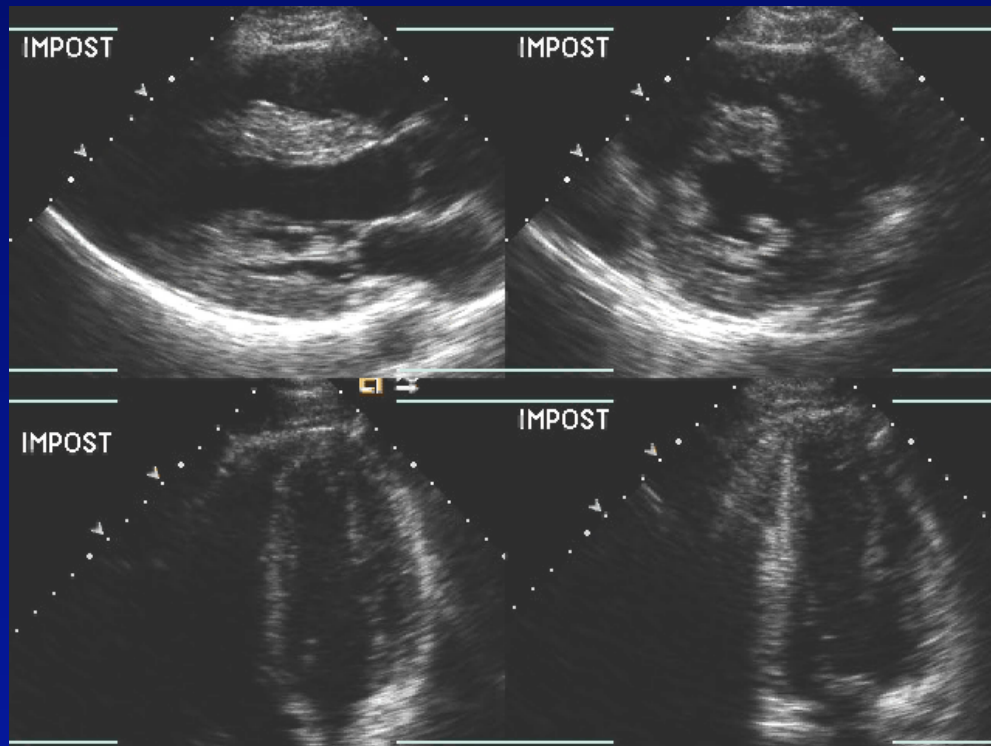


Figure out what is causing symptoms

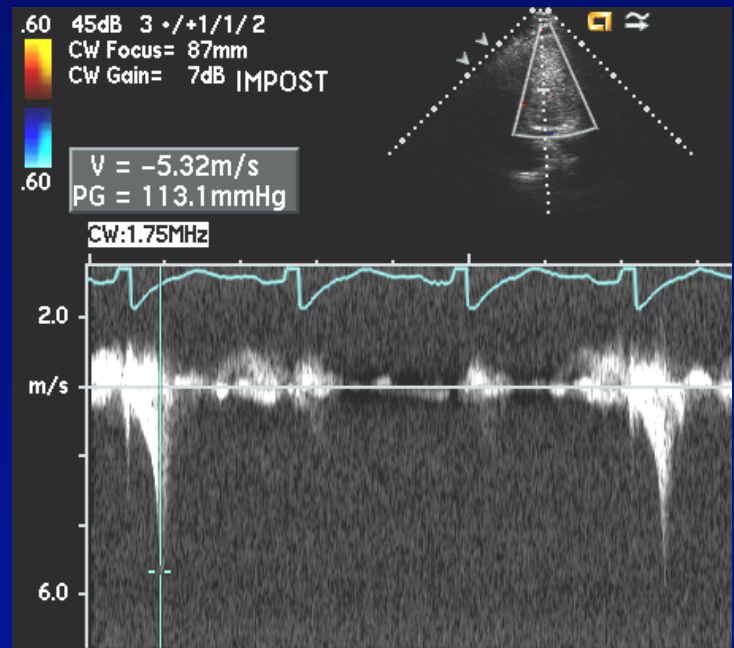
Can non-obstructive patients still have obstruction?

- Small ventricular cavities
- Forceful contraction
- Mid cavity or apical obstruction may occur, particularly with dehydration and exercise
- Symptoms stemming from this mechanism may respond to same therapy used to treat outflow obstruction

Post-exercise echocardiogram



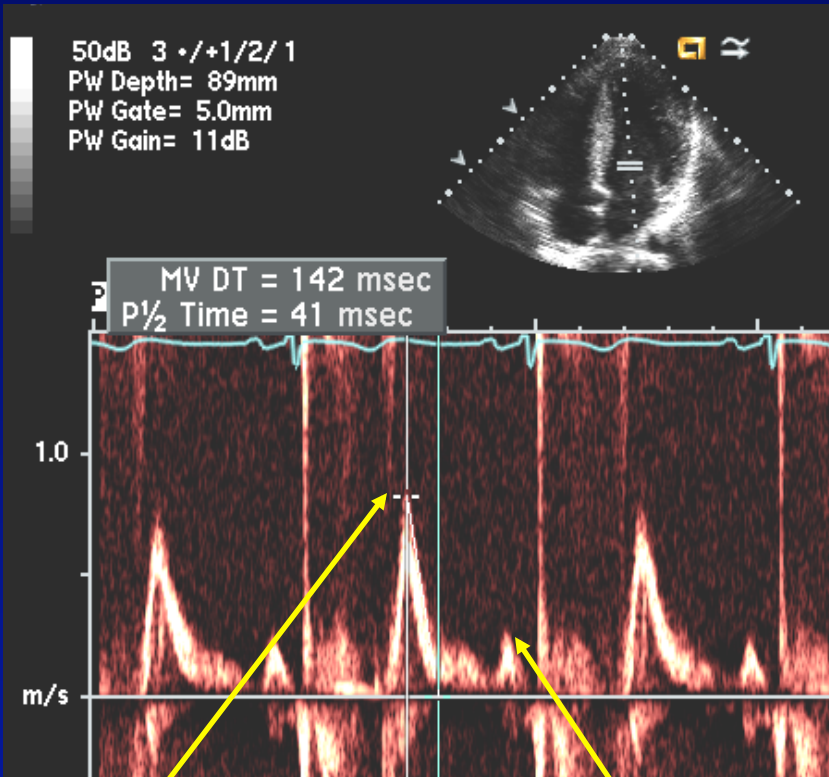
Mid-cavity gradient = 113 mmHg



Diastolic dysfunction

- Thickened ventricle is stiff and slow to relax
- Increased pressure is required to fill the heart with blood
- Not that simple
- Notoriously hard to objectively measure

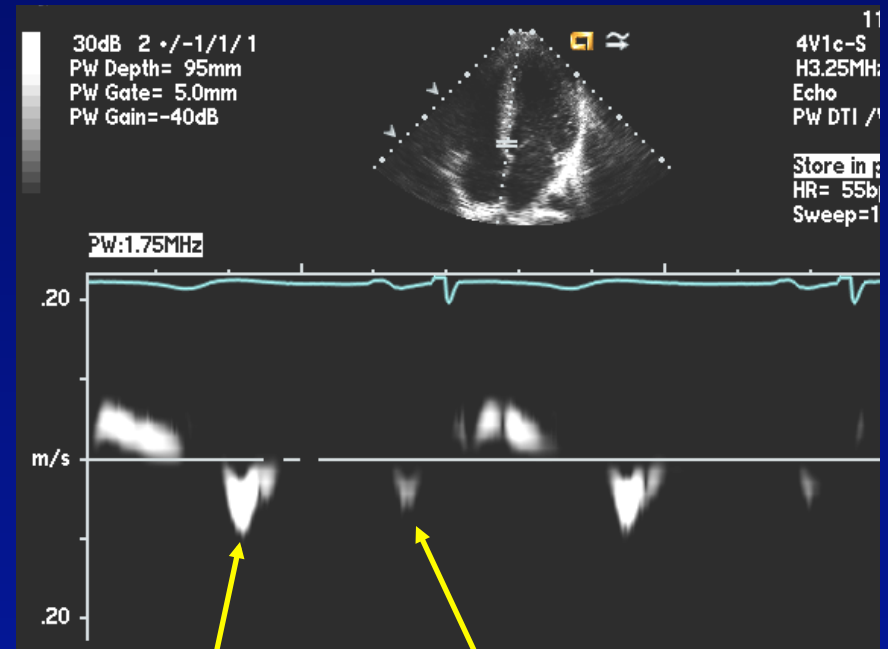
Mitral valve inflow



E = early (passive)
filling

A = atrial contraction
filling

Doppler tissue imaging of the mitral valve annular velocity



E'

A'

Principles of treating diastolic dysfunction

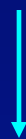
Prolong diastolic filling time, i.e. slow heart rate



Beta-blockers
Calcium channel blockers

Caveat: May directly impair relaxation and contractility

Treat volume overload



Diuretics

Caveat: Can drop stroke volume

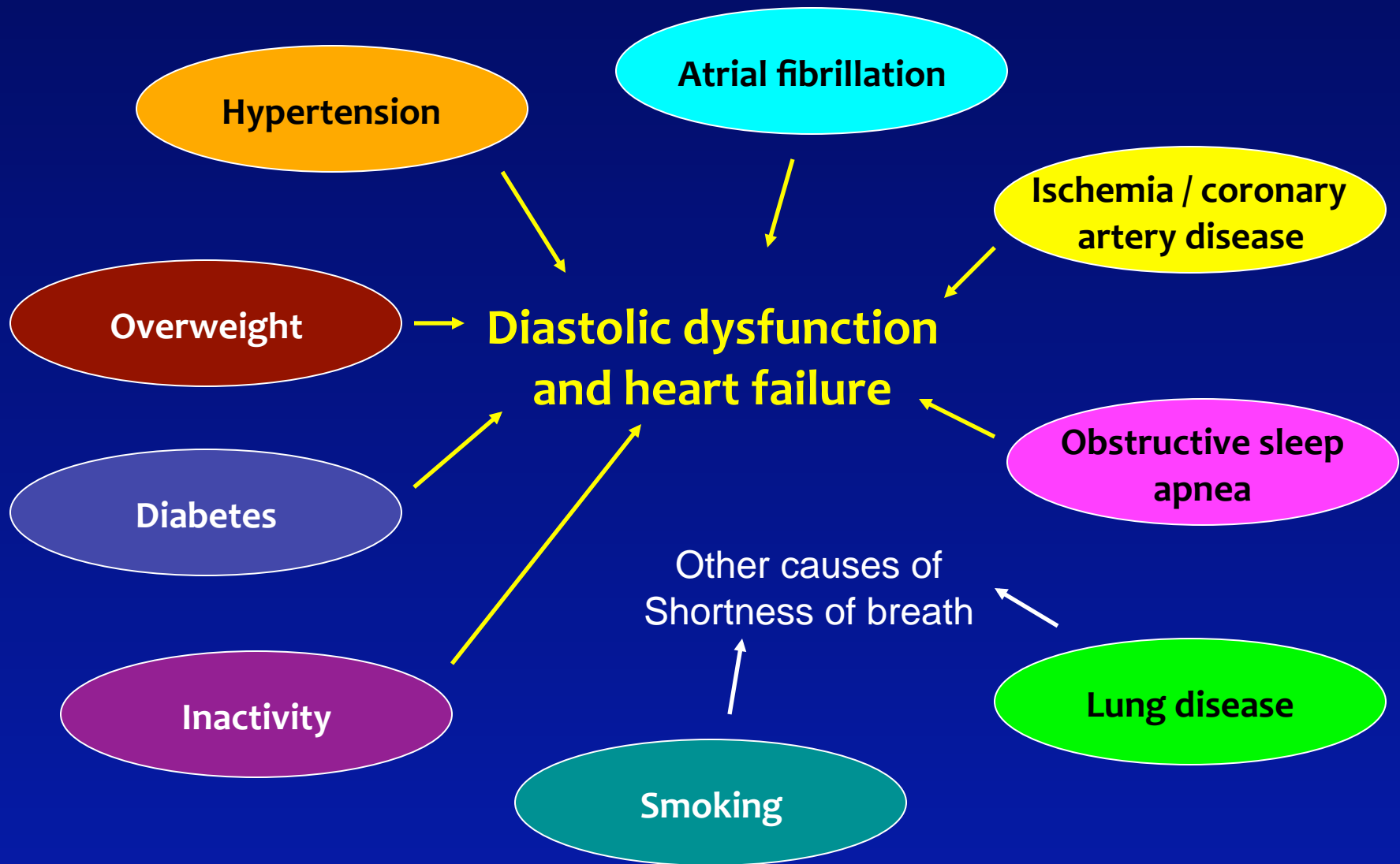
Reduce hypertrophy and fibrosis



ACE inhibitors
Angiotensin receptor blockers
Spironolactone

Caveats:
-Not much data in HCM so far
-No mortality benefit of ARBs in large randomized trials of patients with diastolic heart failure

Treating co-morbidities



Take home points on medical management of HCM

- Current therapies are targeted primarily to symptom relief
- While medical therapy for obstruction is well established, treatment of symptoms in non-obstructed patients is more challenging
 - *Diastolic dysfunction*
 - *Heart failure*
 - *Co-morbid conditions*
- Major focus needs to be placed on developing treatments that slow/prevent progression of disease and prolong life



What about exercise?

Guidelines for sports participation

- Expert opinion
- Many areas (i.e recreational, master's level athlete) in which there are little or no data
- Cannot predict individual SCD risk, so guidelines are same for all
- Generally conservative

36th Bethesda conference:

Eligibility for competitive sports

“Athletes with probable or unequivocal clinical diagnosis of HCM should be excluded from most competitive sports with the possible exception of those of low intensity” (golf, bowling, billiards, cricket, curling, riflery).

“This recommendation is independent of age, gender, and phenotypic appearance, and does not differ for those athletes with or without symptoms, LV outflow obstruction, or prior treatment with drugs or major interventions with surgery, alcohol septal ablation, pacemaker, or implantable defibrillator.”

Maron BJ, et al. Task Force 4: HCM and other cardiomyopathies, mitral valve prolapse, myocarditis, and Marfan syndrome. *J Am Coll Cardiol* 45: 1340-1345, 2005.

AHA Scientific Statement

Recommendations for Physical Activity and Recreational Sports Participation for Young Patients With Genetic Cardiovascular Diseases

Barry J. Maron, MD, Chair; Bernard R. Chaitman, MD, Cochair; Michael J. Ackerman, MD, PhD; Antonio Bayés de Luna, MD; Domenico Corrado, MD, PhD; Jane E. Crosson, MD; Barbara J. Deal, MD; David J. Driscoll, MD; N.A. Mark Estes III, MD; Claudio Gil S. Araújo, MD; David H. Liang, MD, PhD; Matthew J. Mitten, JD; Robert J. Myerburg, MD; Antonio Pelliccia, MD; Paul D. Thompson, MD; Jeffrey A. Towbin, MD; Steven P. Van Camp, MD; for the Working Groups of the American Heart Association Committee on Exercise, Cardiac Rehabilitation, and Prevention; Councils on Clinical Cardiology and Cardiovascular Disease in the Young

High	
Basketball	
Full court	0
Half court	0
Body building	1
Ice hockey	0
Racquetball/squash	0
Rock climbing	1
Running (sprinting)	0
Skiing (downhill)	2
Skiing (cross-country)	2
Soccer	0
Tennis (singles)	0
Touch (flag) football	1
Windsurfing	1

Moderate	
Baseball/softball	2
Biking	4
Modest hiking	4
Motorcycling	3
Jogging	3
Sailing	3
Surfing	2
Swimming (lap)	5
Tennis (doubles)	4
Treadmill/stationary bicycle	5
Weightlifting (free weights) [†]	1
Hiking	3
Low	
Bowling	5
Golf	5
Horseback riding	3
Scuba diving	0
Skating [#]	5
Snorkeling	5
Weights (non-free weights)	4
Brisk walking	5

Scale 0-5, with 0 being strongly discouraged, and 5 being OK

Maron et al., Circ 2004;109:2807-2816

How to follow a safe exercise program

Avoid:

- Competitive, strenuous, or “burst” exertion
 - basketball, football, hockey, sprinting
- Exercise programs that involve systematic training
- Exercising in extremely adverse environmental conditions
- Heavy weight lifting/bodybuilding

Do:

- Light-moderate aerobic exercise
 - walking, hiking, fitness machines, light jogging, biking, swimming
- Low weight, high repetition strength training
- Exercise at a conversational pace
- Stay well hydrated
- Be alert and aware of symptoms, willing to terminate or back off
- Know ICD settings