

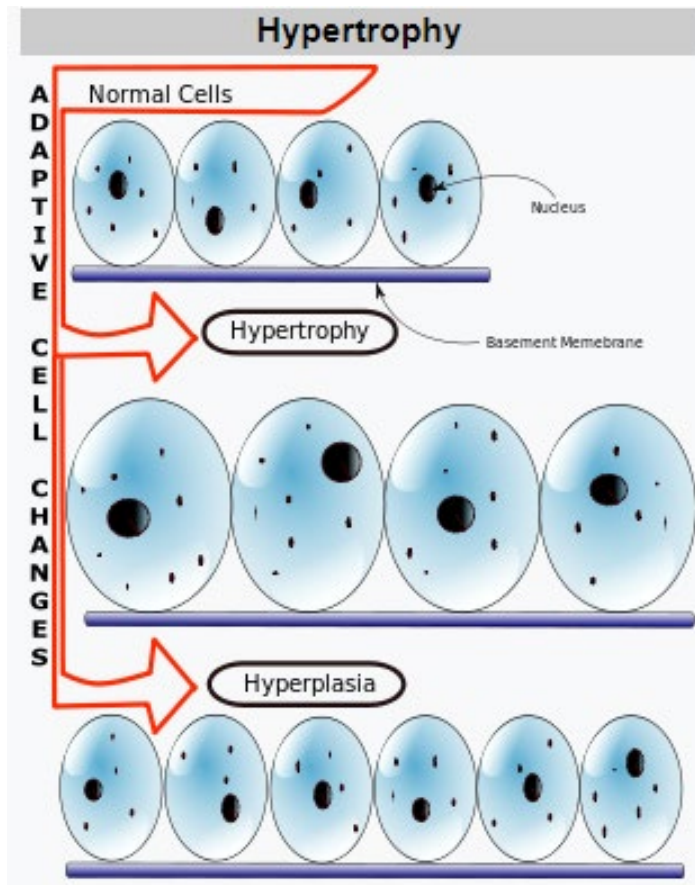
Hypertrophic Cardiomyopathy

Jonathan Ginns MD

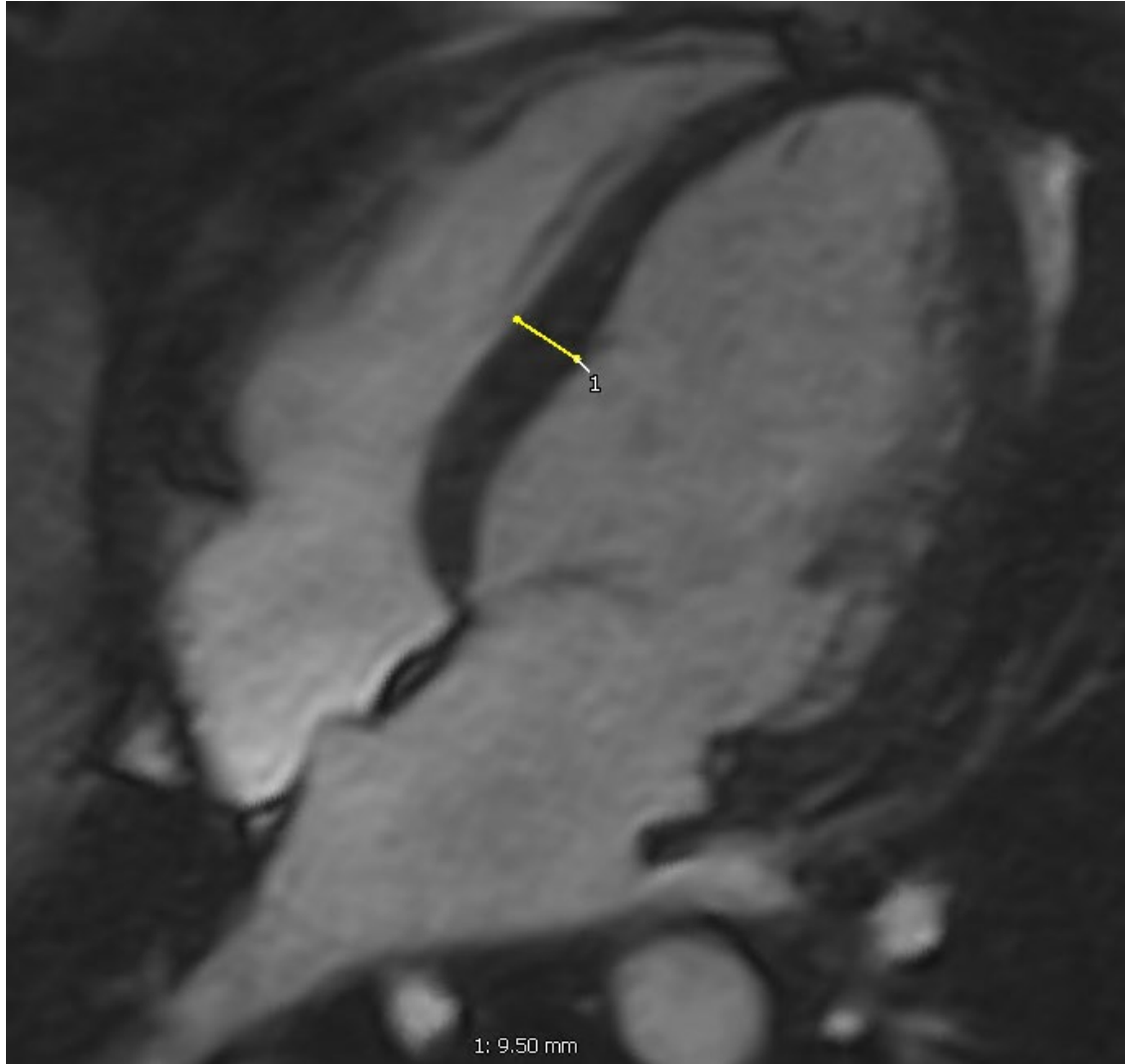
Cardiologist, Austin Heart and Heart Hospital of Austin

What is Hypertrophic Cardiomyopathy (HCM)?

- Abnormal heart muscle thickening (>1.5cm thick; normal ≤ 1.2 cm)- typically asymmetric (one part thicker than others)

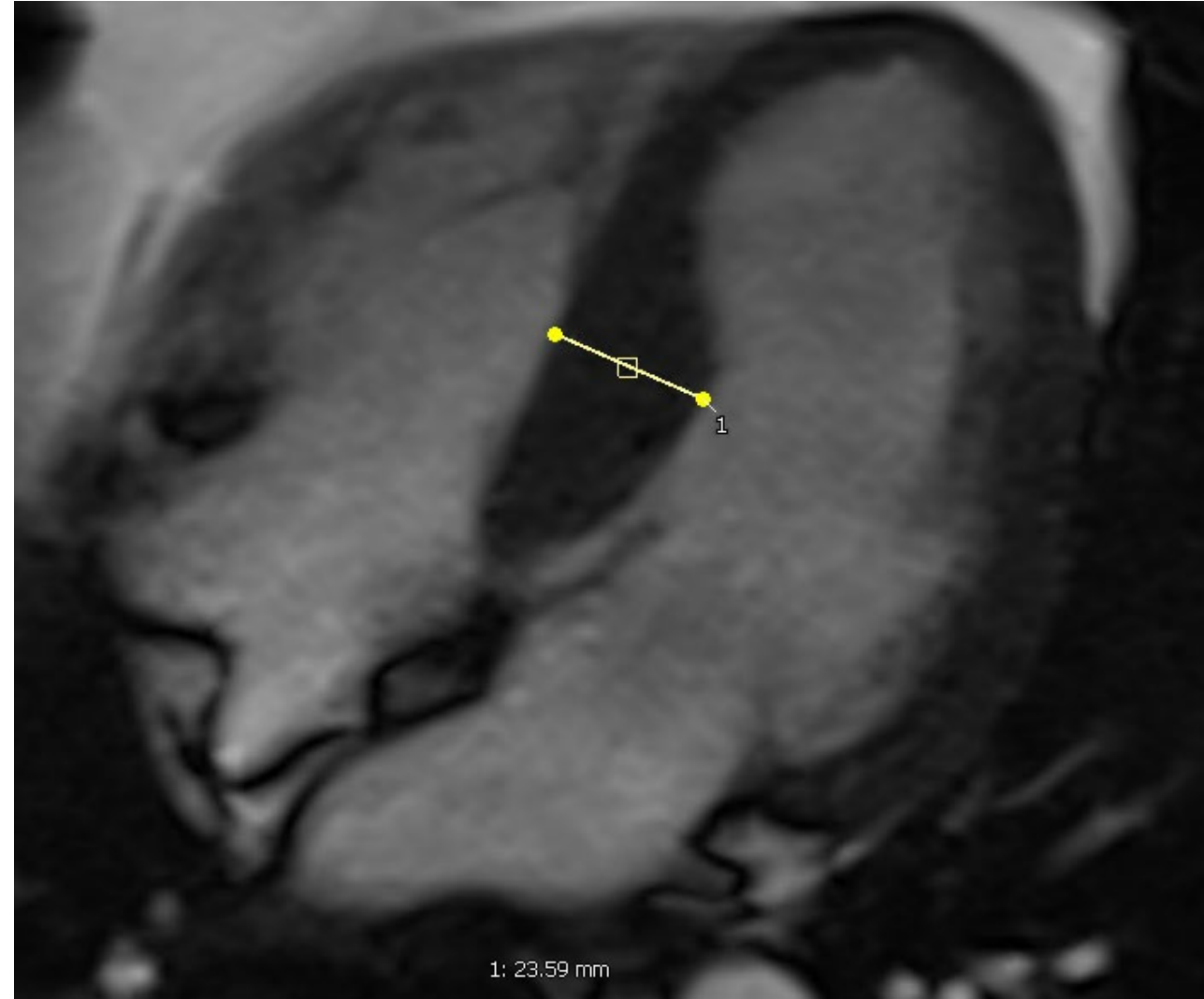


Normal versus...



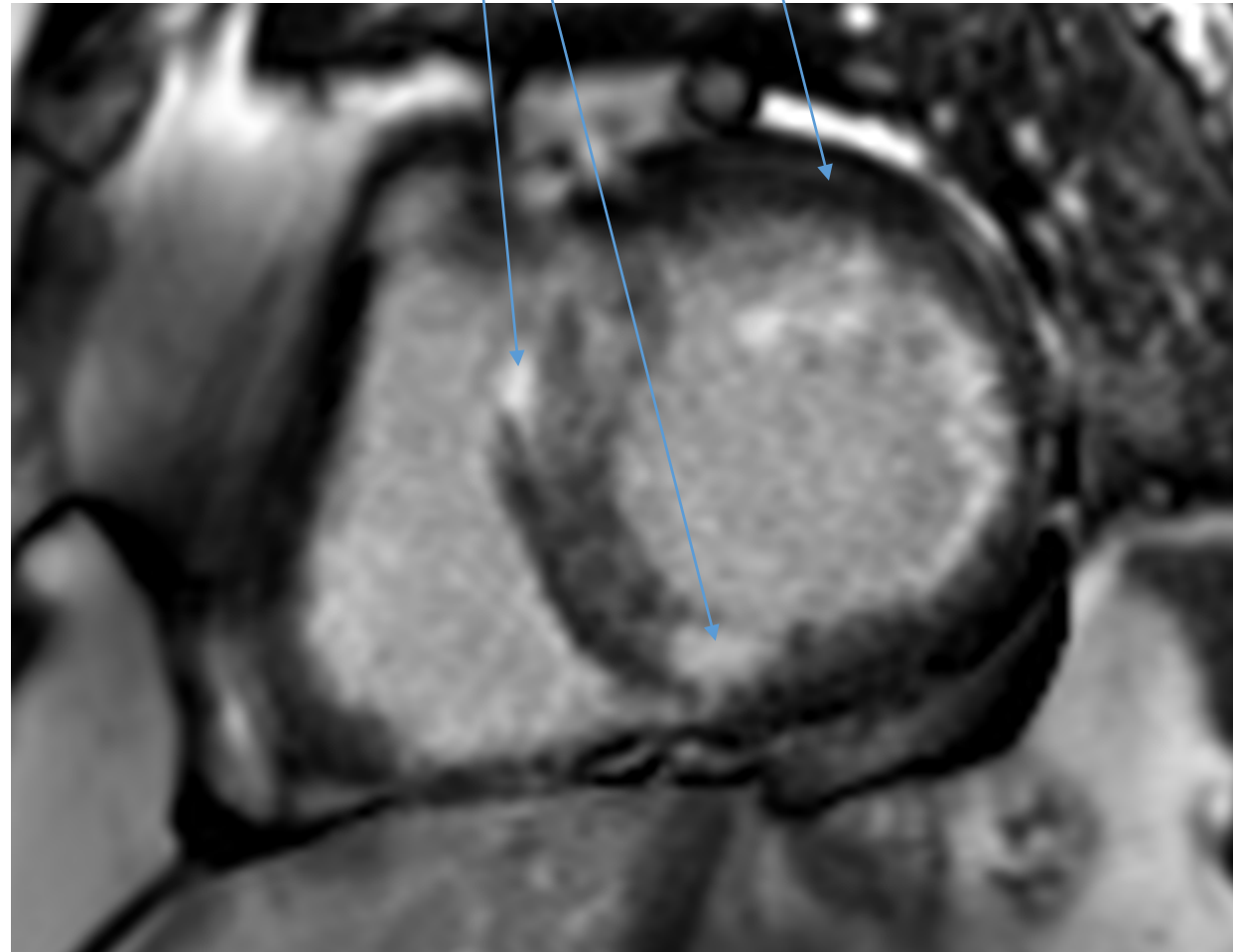
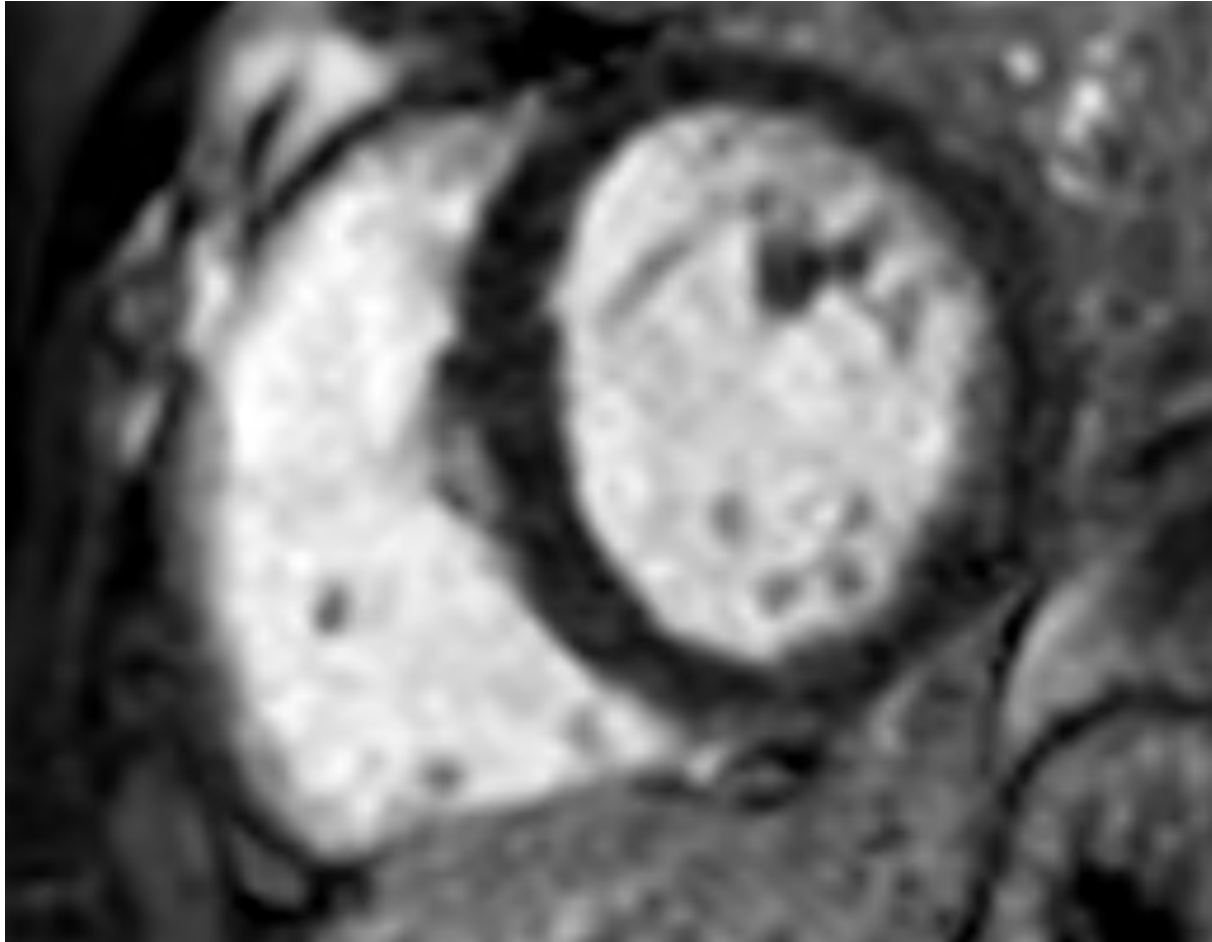
Wall 9.5mm thick

HCM:



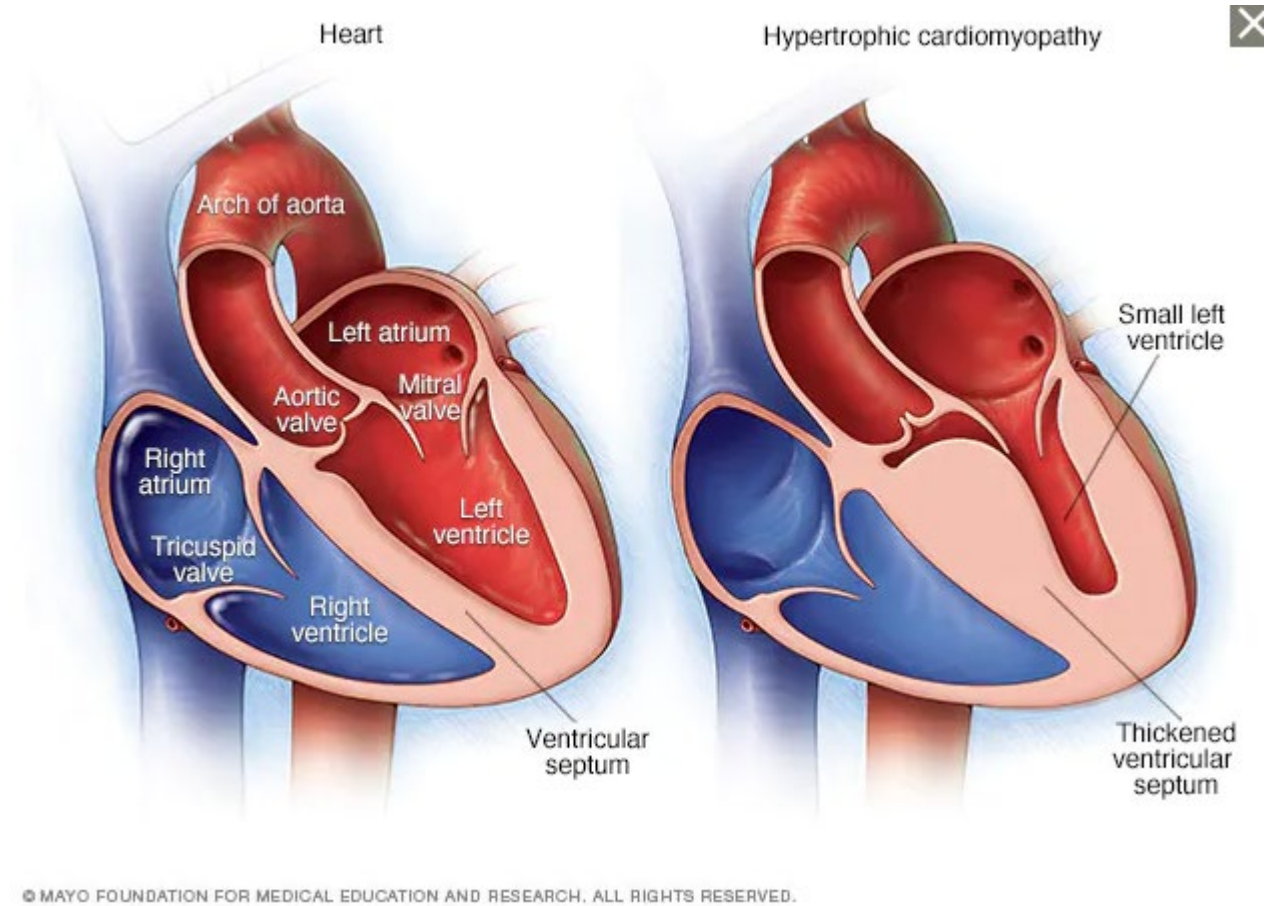
Wall 23.6mm thick

Scar tissue in HCM- can cause heart rhythm issues



Symptoms in HCM

- Shortness of breath
- Palpitations
- Chest pain
- Dizziness
- Fatigue
- Often asymptomatic and found on ECG/echo/screening



How many people suffer from HCM? (2020 guidelines)?

- HCM is a common genetic heart disease reported in populations globally.
- The distribution of HCM is equal by sex, although women are diagnosed less commonly than men. Children through to elderly
- The prevalence of unexplained asymptomatic hypertrophy in young adults in the United States has been reported to range from 1:200 to 1:500.
- Symptomatic hypertrophy based on medical claims data has been estimated at <1:3000 adults in the United States; however, the true burden is much higher when unrecognized disease in the general population is considered.

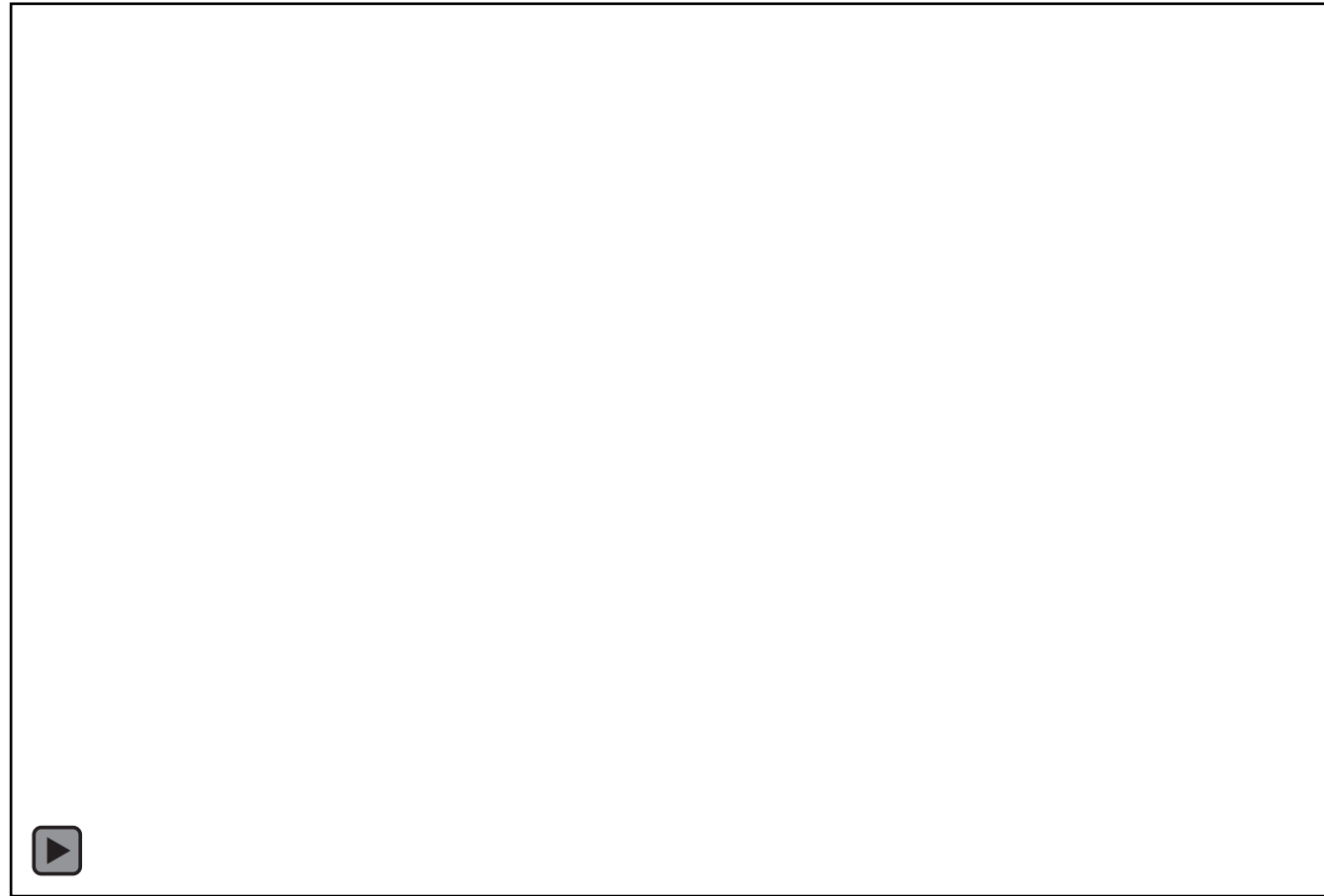
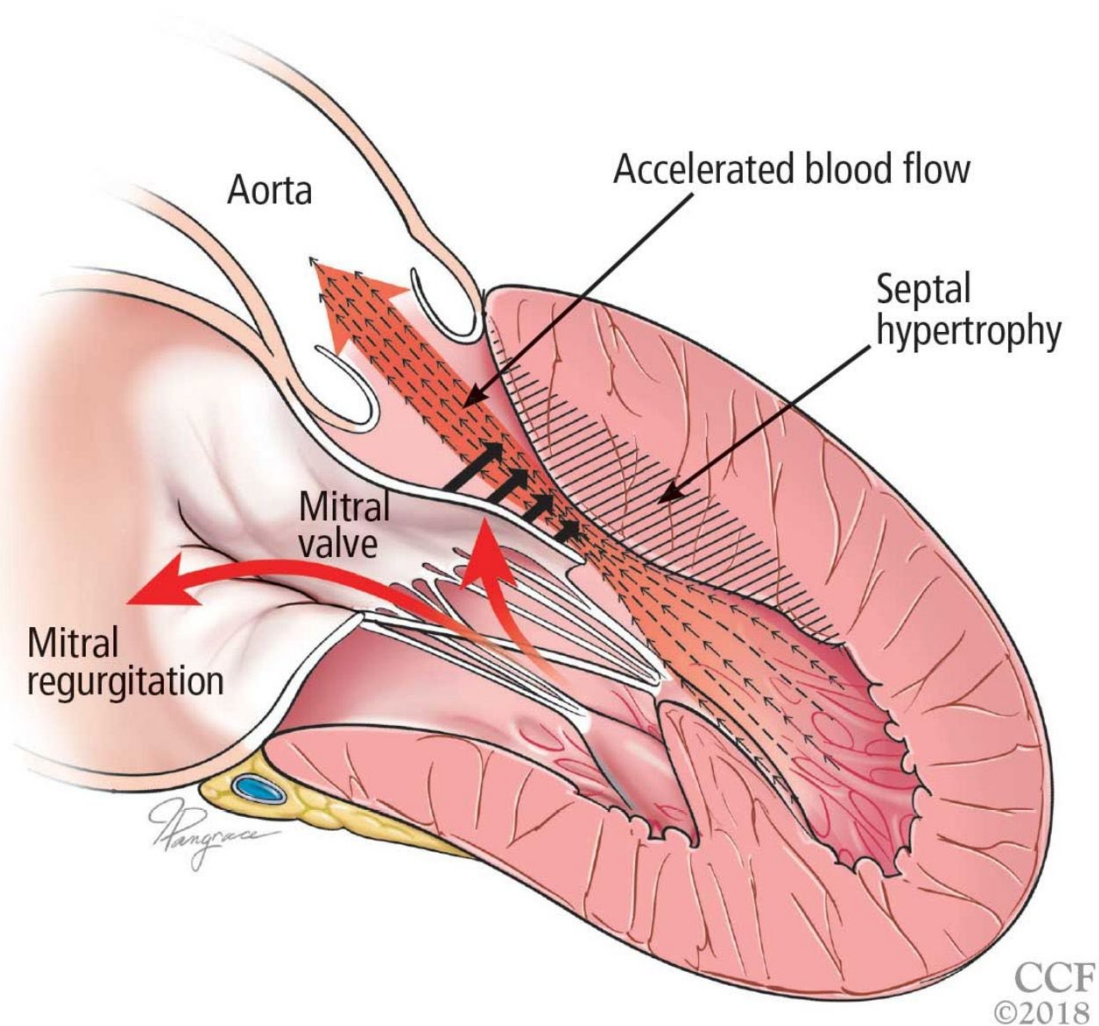
What causes HCM?

- Genetic: Currently, variants in 1 of 8 or more genes encoding proteins of the cardiac sarcomere (or sarcomere-related structures) have been implicated in causing HCM.
- Among patients with HCM, 30% to 60% have an identifiable pathogenic or likely pathogenic genetic variant.
- A substantial proportion of patients with HCM are currently without any evidence of a genetic etiology to their disease, including a subgroup (up to 40% of patients in 1 study) who also have no other affected family members (ie, “non-familial” HCM)
- Mimicking conditions:- high blood pressure, cardiac amyloid, Fabry’s disease, “athlete’s heart”, Noonan syndrome

What does care look like in HCM?

- Interdisciplinary team + shared decision making
- Cardiologist- sometimes specialist in HCM
- Yearly or 6 monthly followup
- ECG, echo, Holter monitor, genetic testing, MRI
- Screening family members- 1st degree relatives
- Other specialties may include geneticist, heart surgeon, electrophysiologist

Obstruction



Treatment for obstruction:

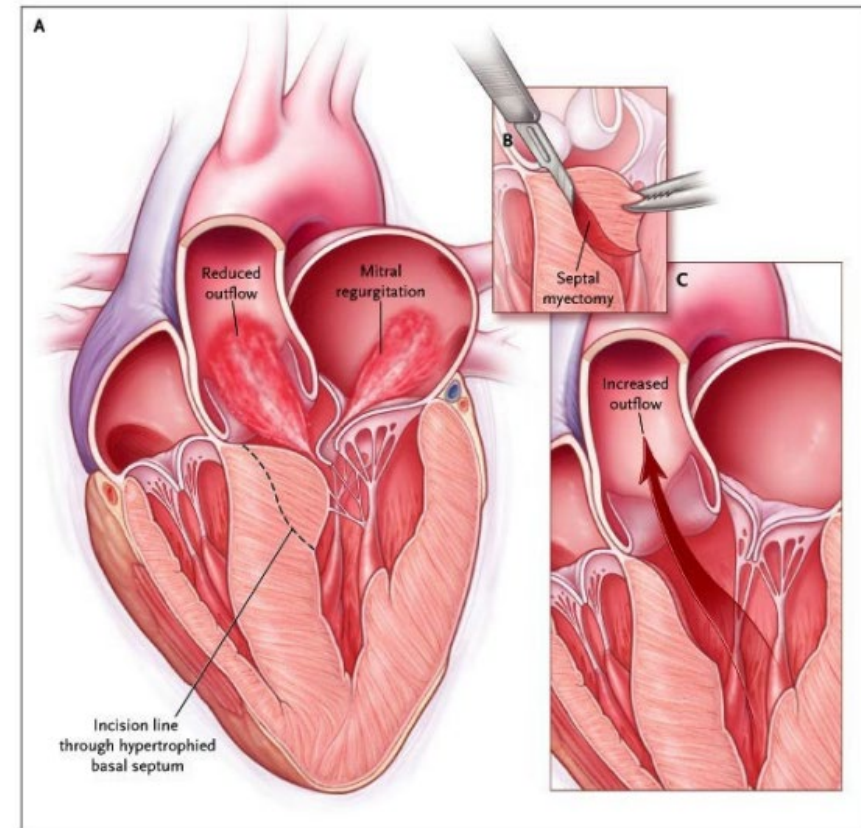
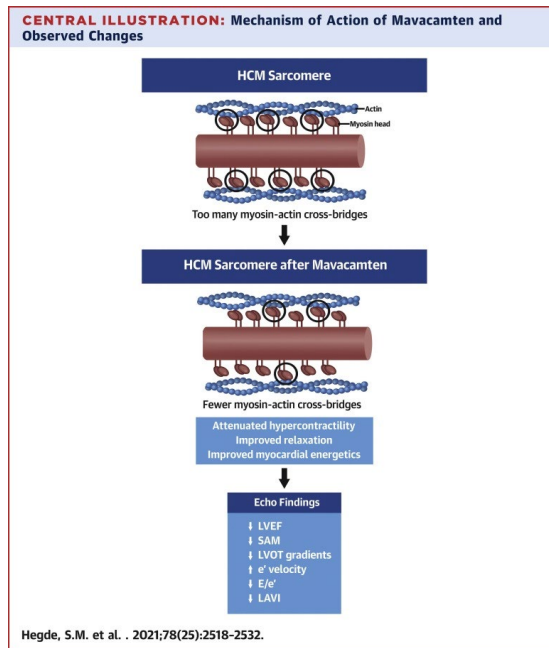
- Medical

- Beta-blockers

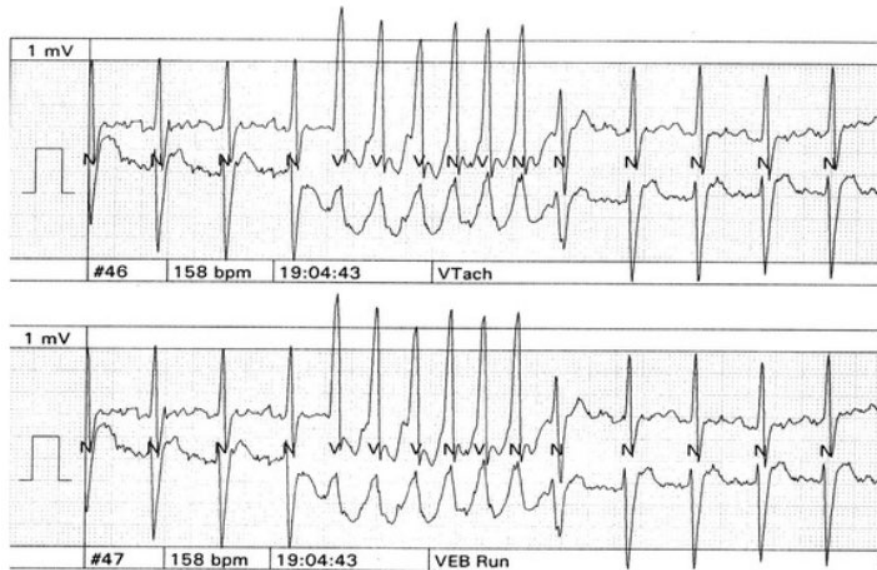
- Calcium channel blockers

- Mavacamten

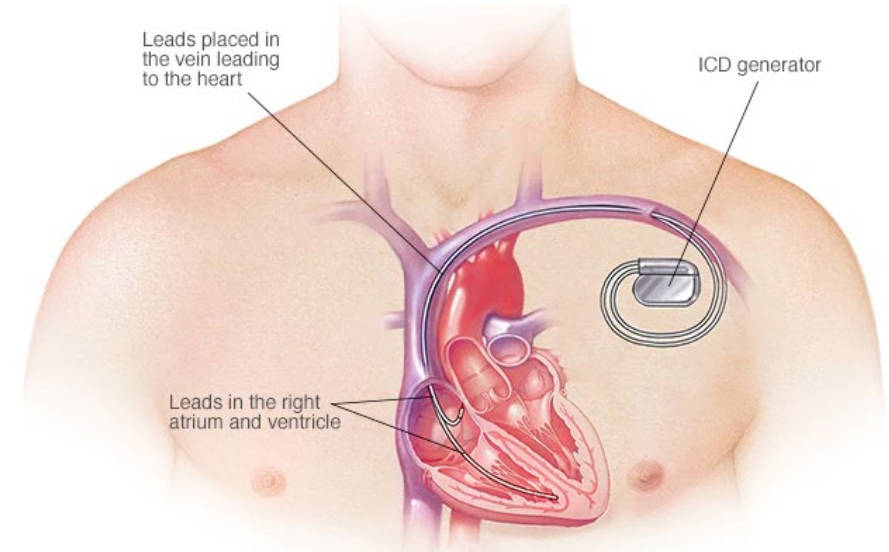
- Invasive



Arrhythmias



The patient still showing non sustained ventricular tachycardia on 24 hour holter monitor.



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Implantable cardioverter-defibrillator (ICD)

An ICD works to control the heartbeat by delivering shocks to the heart when the device detects an irregular heartbeat.

- Blood thinning meds
- Antiarrhythmics
- Ablation

Heart failure- “HFPEF”

Caused by heart muscle being “too stiff” which causes a backup of fluid into the lungs

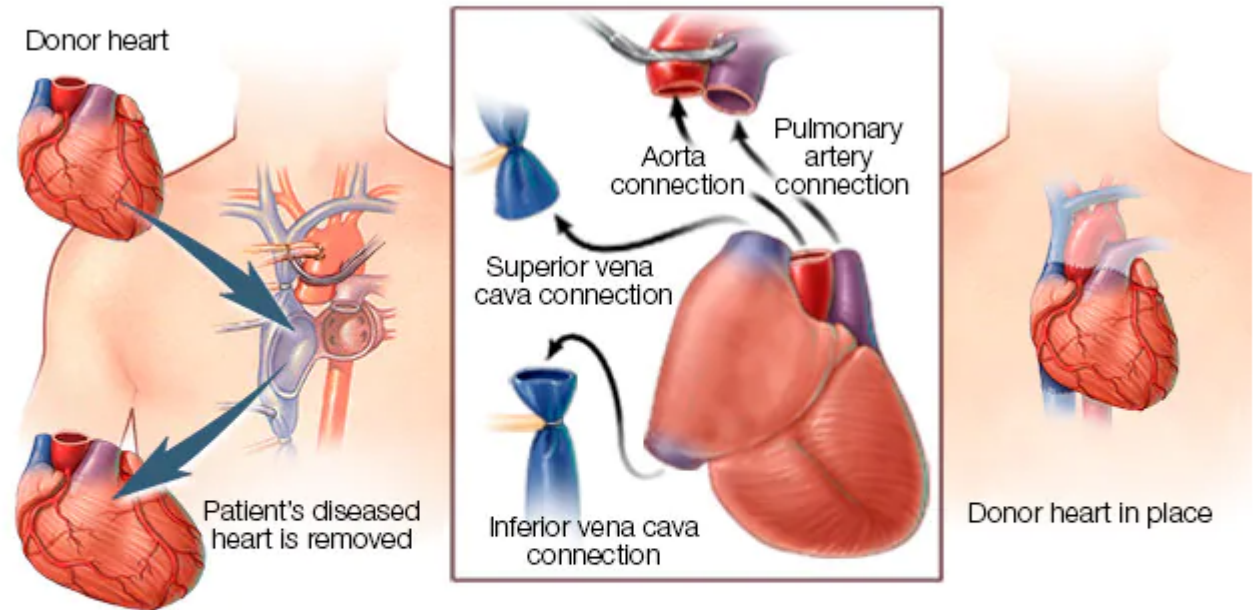
Treatment:

Medications- diuretics, SGLT-2 inhibitors, ?mavacamten

Consideration of LVAD (mechanical heart pump)- uncommon in HCM

Consideration of heart transplant

Heart transplant procedure

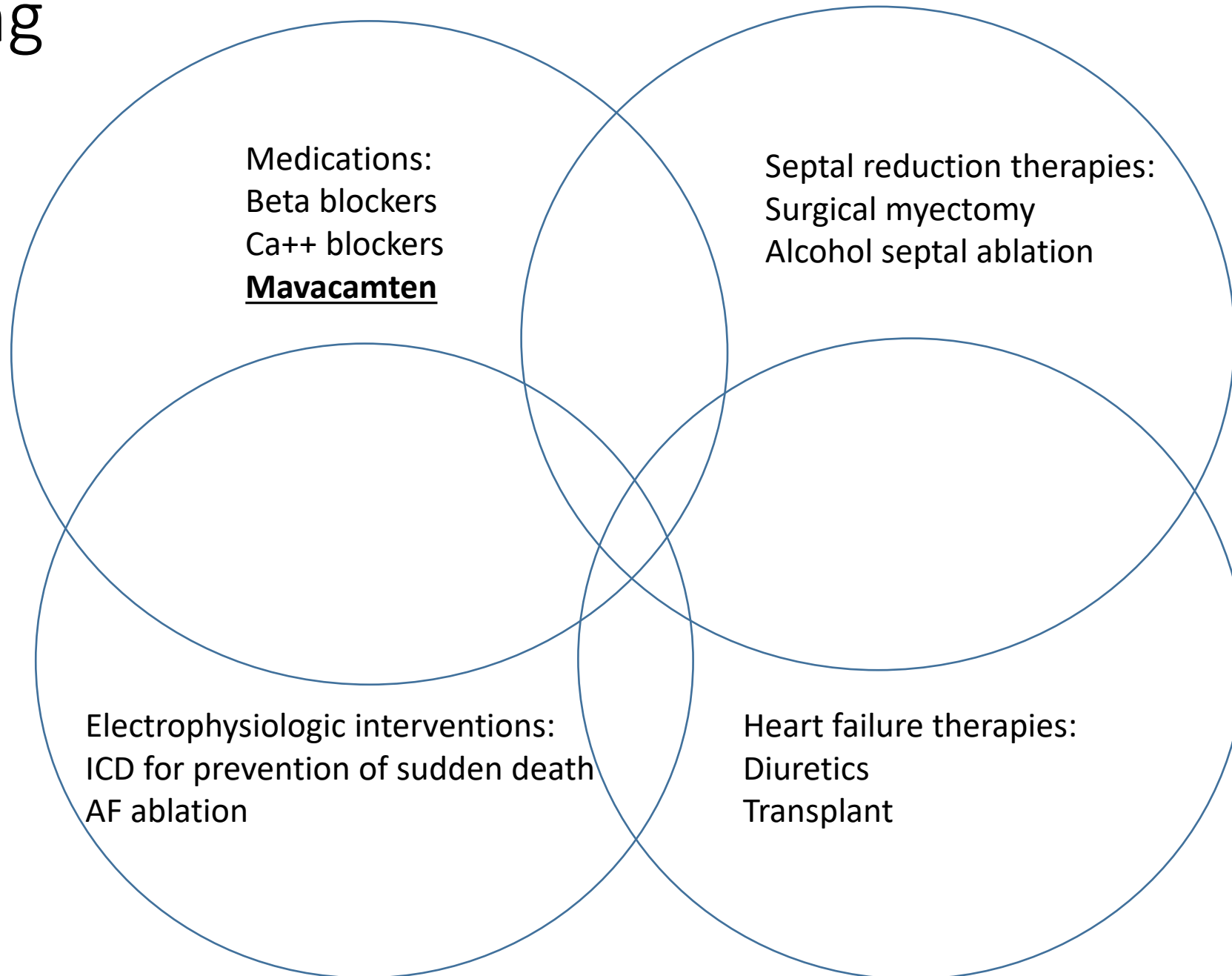


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Heart transplant procedure

In a heart transplant procedure, a surgeon removes the diseased heart and sews the donor heart in place. He or she then attaches the major blood vessels to the donor heart.

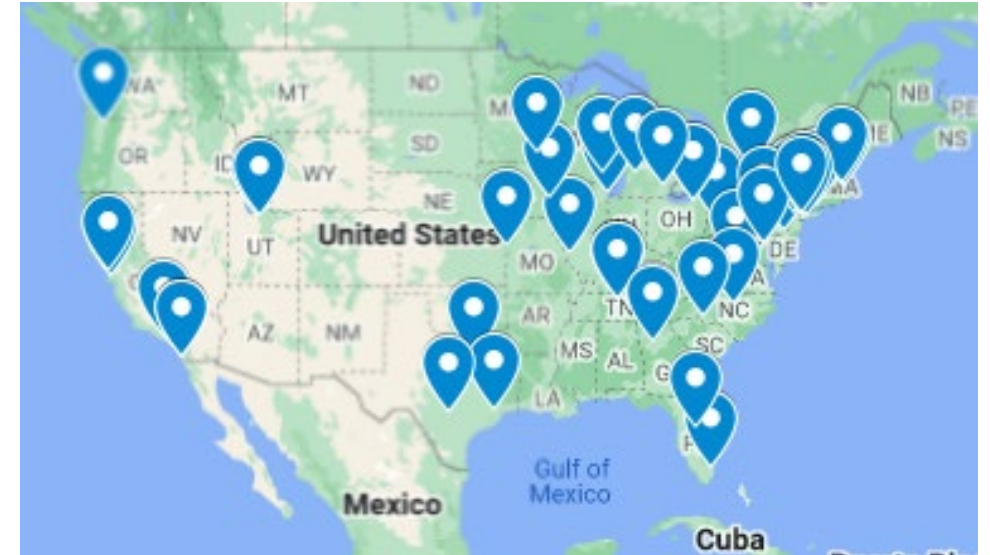
Treating HCM



Where are we now?

- Improving outcomes, comparable to age matched controls when cared for at a center of excellence
- Increased awareness has led to improved recognition and standardized therapies for patients with HCM
- Texas Healthy Cardiac Monitoring Act

Centers of excellence:



What does the future hold?

- Additional medical therapies for obstruction and heart failure
- Better devices for detection and treatment of arrhythmias
- Gene therapies?
- Increased awareness and better screening

Thank you!