

Bigger... Isn't ... Better :

Risks & Reasons for an Enlarged Heart

UVM Community Medical School
February 19, 2019



The University of Vermont
LARNER COLLEGE OF MEDICINE

Disclosures: None

-
- Financial Conflicts
 - Family Secrets
 - Pediatric Cardiologist



OBJECTIVES

1. *Experience*: UVM “Active Learning” Process
2. *Define*: Cardiomyopathy
3. *Evaluate*: Patients & Families
4. *Discuss*



Beginner



Intermediate



Expert



Terrain Park



The University of Vermont
LARNER COLLEGE OF MEDICINE

Active Learning

vs

Passive

STUDENT BASED

- Engage/ Construct Knowledge
- Demonstrate/Teach/Apply/Discuss
- Effective teamwork & communication

LECTURE BASED

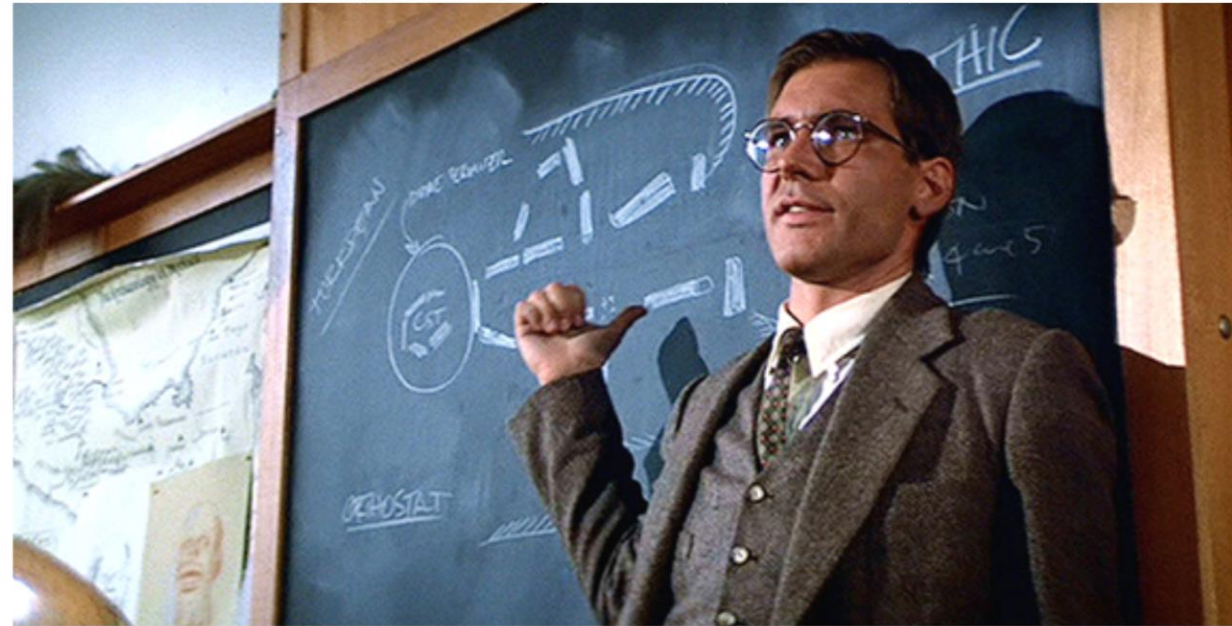
- Absorb facts from an expert
- Faculty transfer information
- Faculty talk/direct



Active Learning

vs

Passive Lectures



.... READ... BEFORE CLASS

Class = Team Based Learning

- Group pre/post test(s)
- Group Discussion
 - Concepts
 - Knowledge
 - Application

Online applicants

Submit your Vermont online application. Prior to submitting your application, you

- Be at least 18 years of age.
- Be a citizen or permanent legal resident of the United States.
- Be a Vermont resident of or have a place of employment or practice in Vermont.

2. Upload a scanned copy of your completed, signed and notarized Oath of Office. Please complete this form prior to filling out your online application.

Your non-refundable application fee of \$15.00 paid with credit card or by check (not from a checking account).

Vermont Secretary of State
Office of Professional Regulation
89 Main Street, 3rd Floor
Montpelier VT 05620-3402
www.sec.state.vt.us





Beginner

Direct Question

To become a Notary Public in the state of Vermont, which of the following is NOT a requirement:

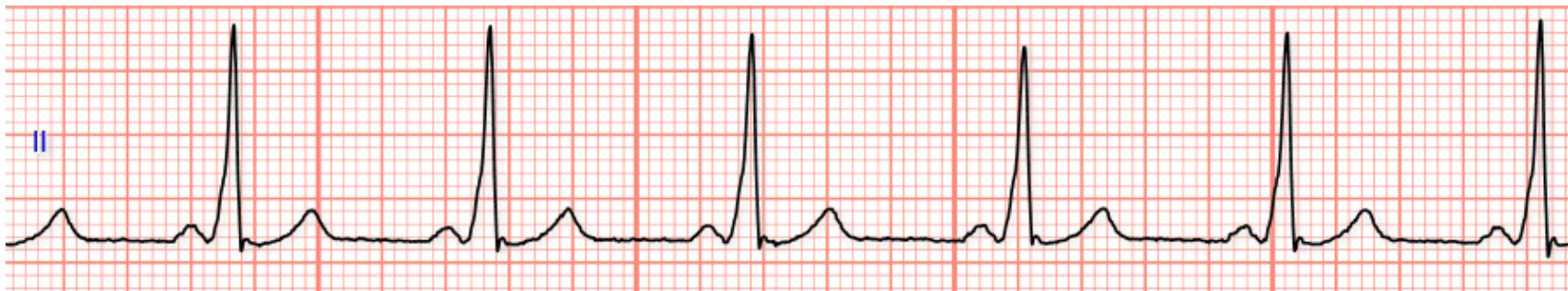
- A. Vermont resident or employment
- B. Citizenship (USA)
- C. ≥ 18 years of age
- D. Pediatric cardiology certification
- E. 15 dollars (\$)



Short Case

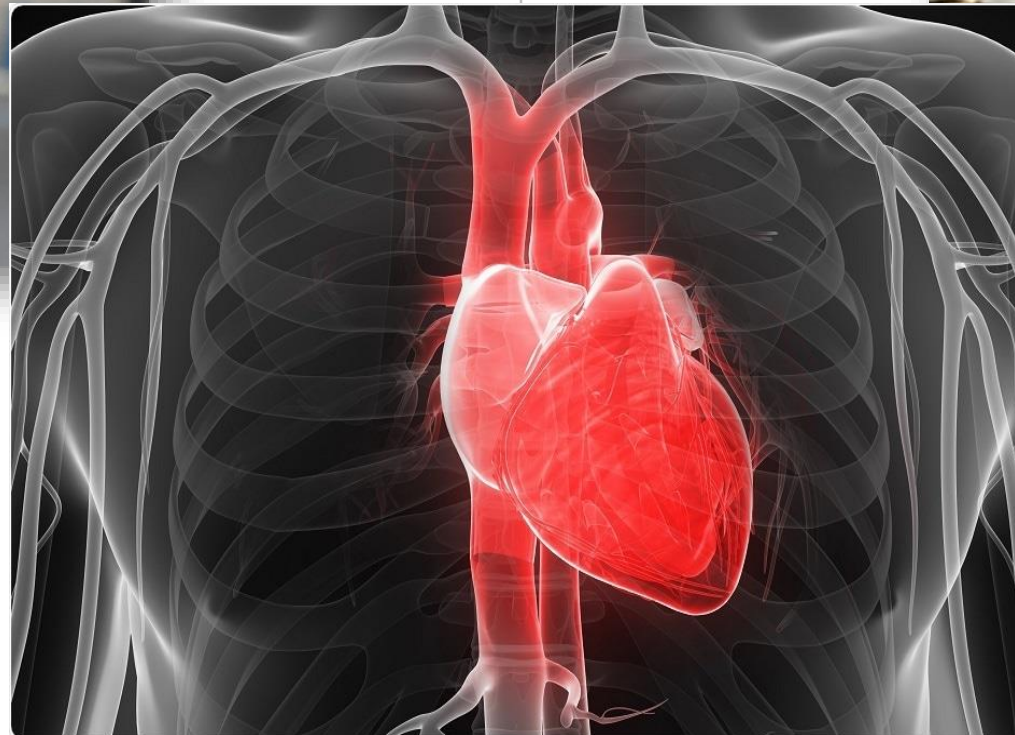
An EKG is obtained today for a 17 year old female with palpitations during English literature class.

What is the most likely diagnosis?



- A. Neurocardiogenic Syncope
- B. Wolff-Parkinson White Syndrome
- C. Long QT syndrome
- D. Shakespearean Comedy

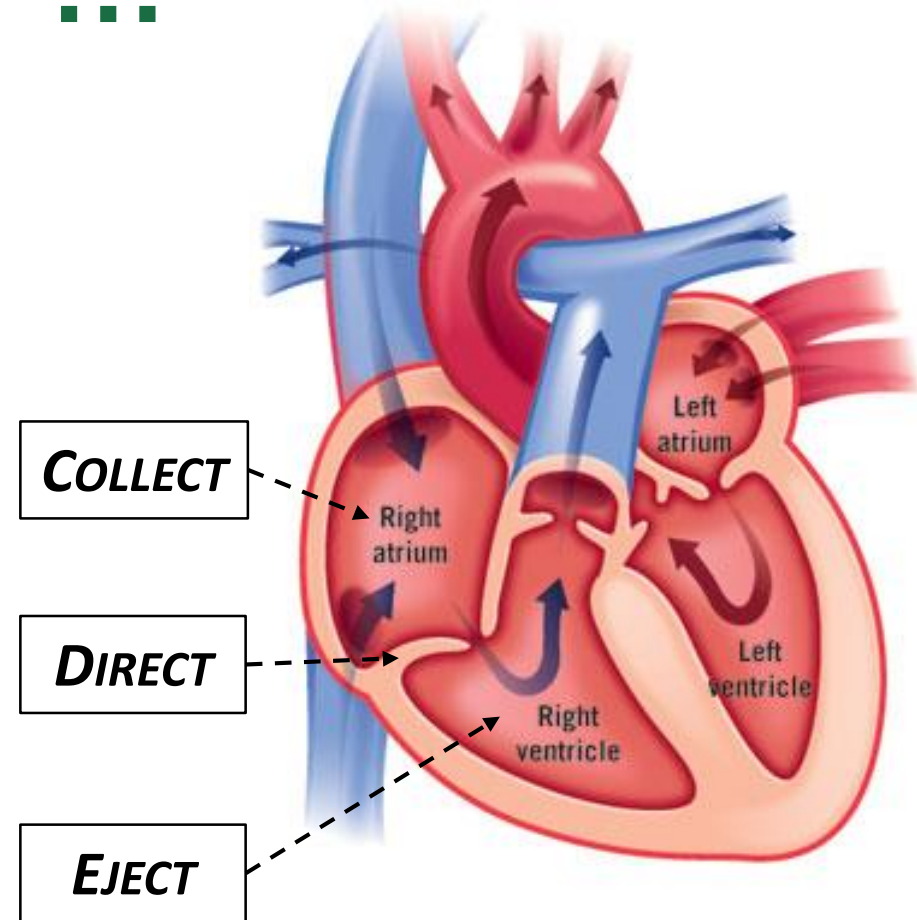
HEART = MUSCLE



PUMP... PUMP... PUMP...

MOVE : BLOOD

- Collect: Atria
- Direct: Valves & Walls
- Eject: Ventricles



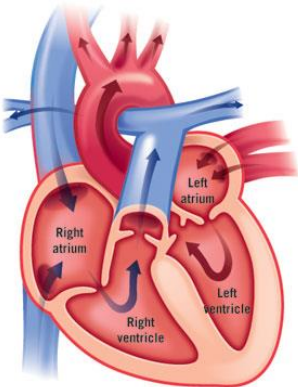
So... What's a Cardiomyopathy?



“An enlarged heart killed him. It grew three sizes *today*.”

CARDIOMYOPATHY

CARDIO



HEART

Myo



MUSCLE

PATHOS



SUFFERING

BIG

hypertrophy noun

hy·per·tro·phy |

1. Biology: excessive organ development
 - Increase in bulk, without multiplication of parts
 - Thickening of muscle fibers
 - *Cardiac hypertrophy*
2. Exaggerated growth

dilation noun

di·la·tion |

1. Action of stretching or enlarging an organ or part of the body

BIGGER...examples

Hypertrophy

Dilation

Hypertension

++++

Obstruction

++++

Communication

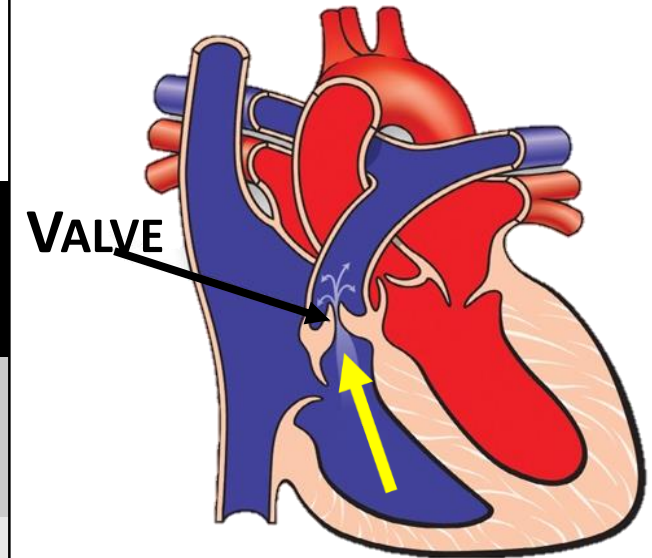
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Cardiomyopathy

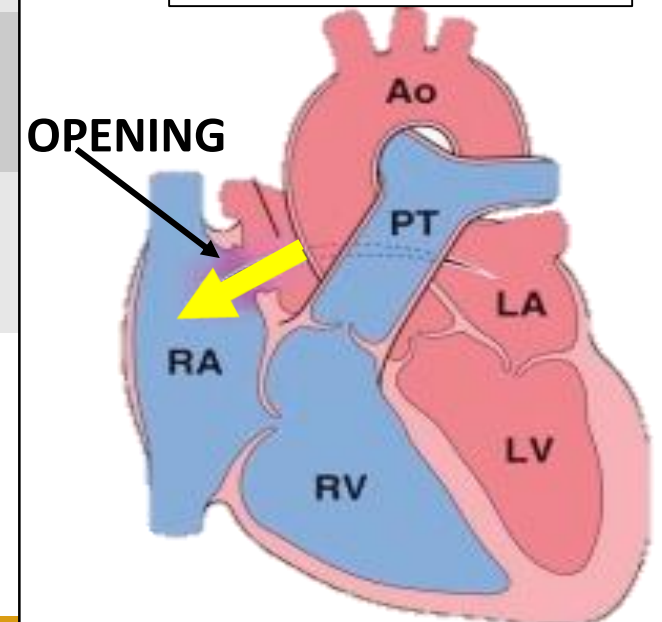
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OBSTRUCTION



COMMUNICATION



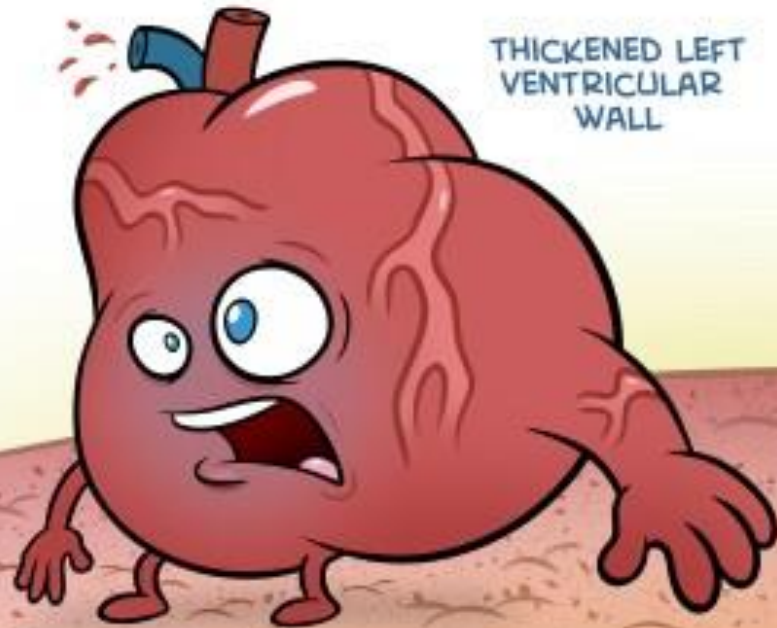
CARDIOMYOPATHY

HYPERTROPHIC

DIASTOLIC
DYSFUNCTION

RISK OF SUDDEN
DEATH IN YOUNG
ATHLETES

THICKENED LEFT
VENTRICULAR
WALL



DILATED

ENLARGEMENT
OF ALL CARDIAC
CHAMBERS

SYSTOLIC
DYSFUNCTION

MOST
COMMON
TYPE



RESTRICTIVE

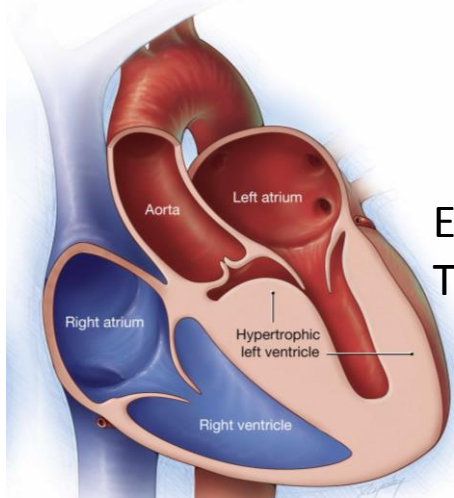
RIGID
VENTRICULAR
WALLS

DIASTOLIC
DYSFUNCTION

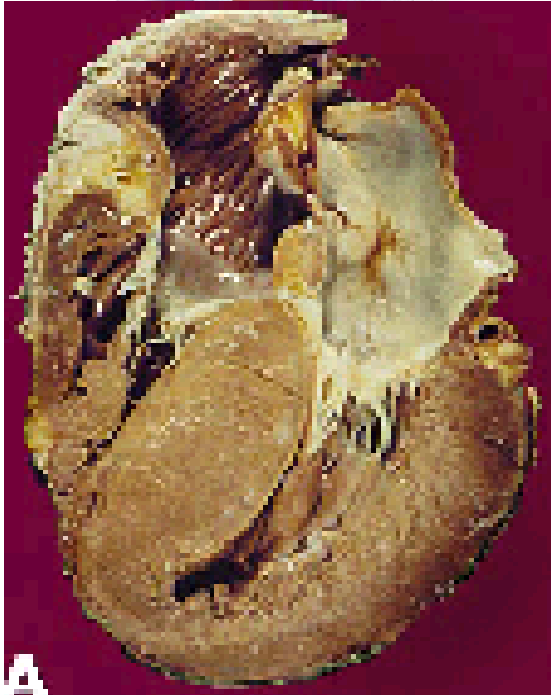
LEAST
COMMON
TYPE



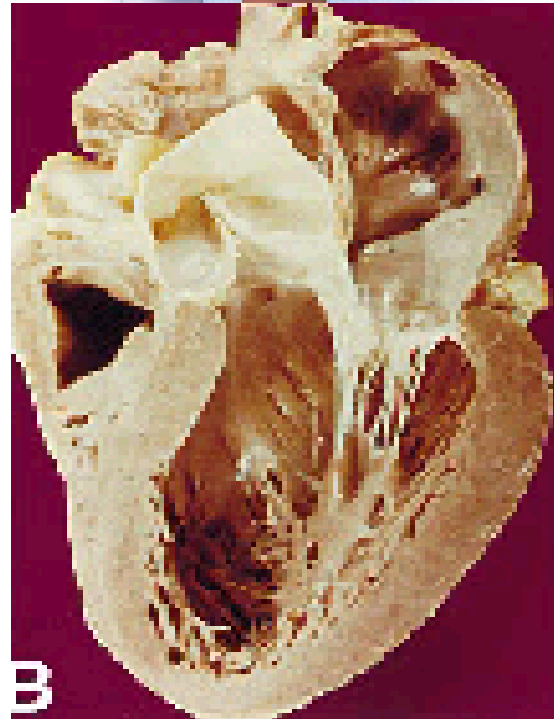
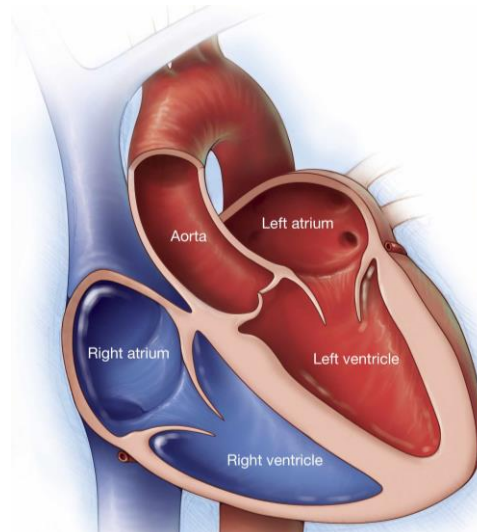
Hypertrophic Cardiomyopathy



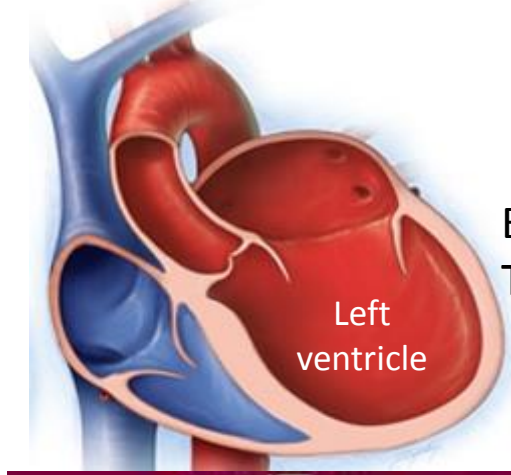
Enlarged
Thick Walls



Normal



Dilated Cardiomyopathy



Enlarged
Thin Walls

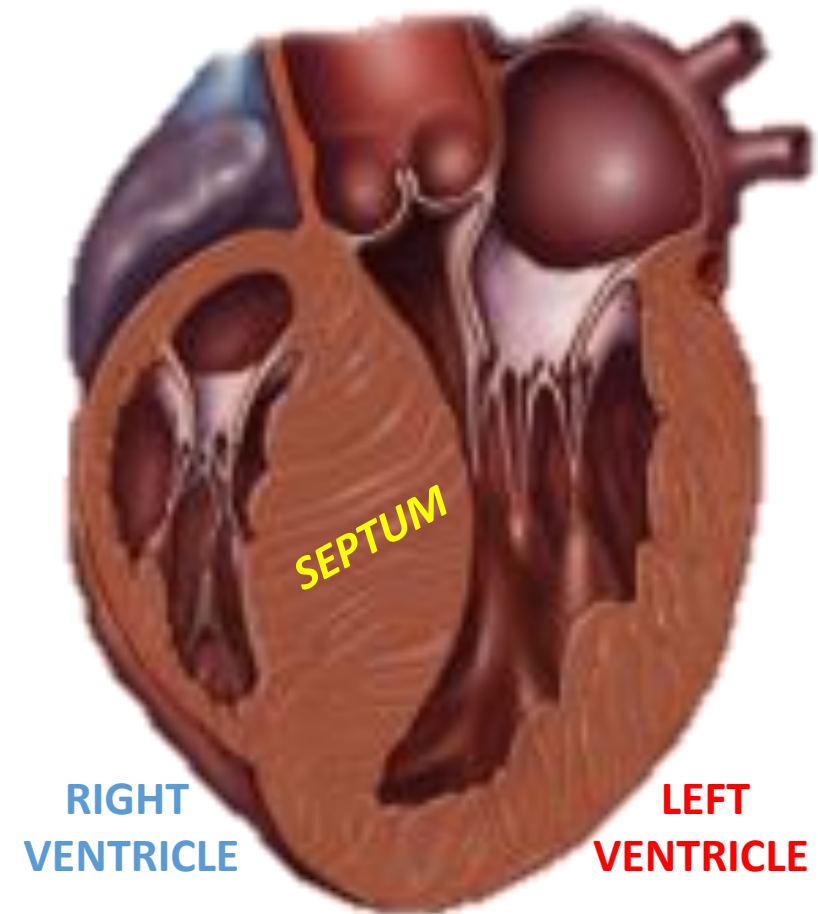




A 5 year old child has a genetic defect that causes the heart muscle to progressively thicken.

This defect may cause ventricular:

- A. Hypertrophy
- B. Dilation
- c. Shakespearean Comedy



Genetics: Overview

Humans: 30,000 – 40,000 genes

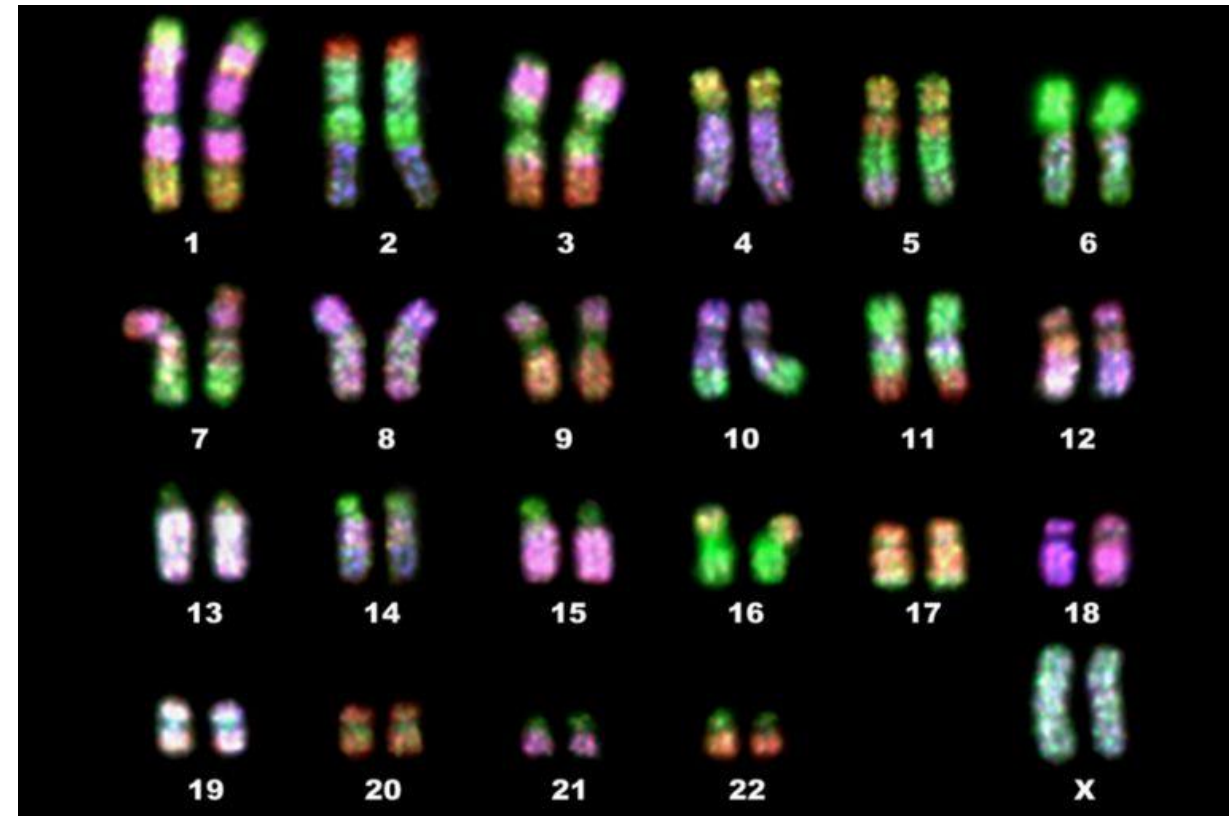
Genes: Code Proteins

Proteins: Make Muscle

Human Genome Project (2003)

- >1,800 disease genes
- >2,000 genetic tests for human conditions

Chromosome Pairs: 23

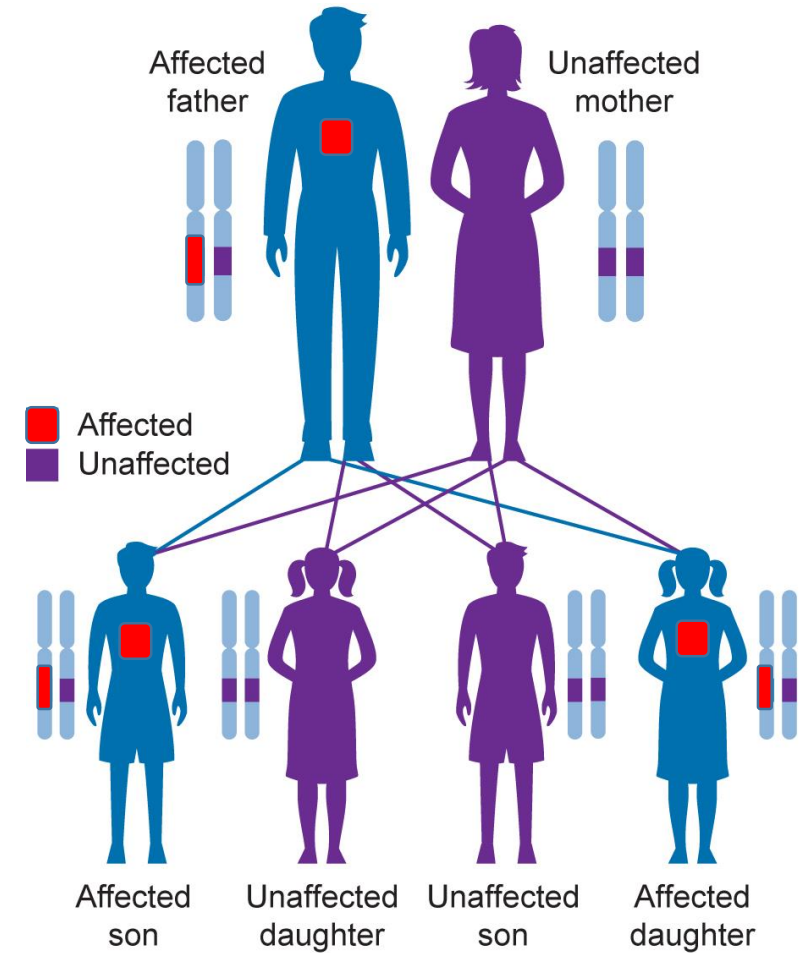


Hypertrophic Cardiomyopathy

Genetic Cause: 34% of HCM²

- Autosomal Dominant
- >11 genes
- >1500 mutations

Autosomal Dominant Inheritance



Hypertrophic Cardiomyopathy

Most Common: Heritable Cardiomyopathy

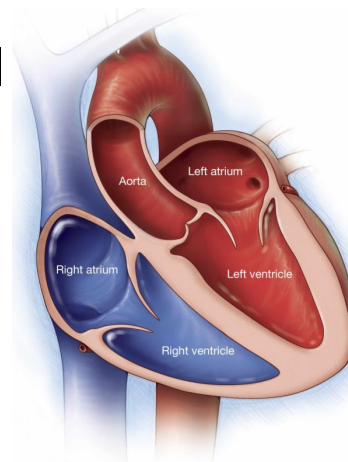
- Historically: 1/500
- Muscle: Contractile Proteins
- 122 Countries (Map: Red)¹



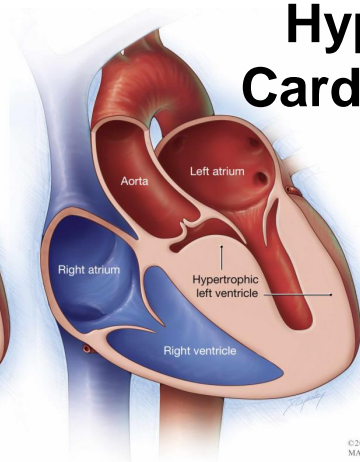
Unexplained: Left Ventricular Hypertrophy

- $> 15\text{mm}$
- $\geq 13\text{mm}$ (+ First Degree Relative)

Normal



**Hypertrophic
Cardiomyopathy**



1. Maron et al. *Global Burden of HCM*. JACC HF 2018
2. Geske et al. *HCM Clinical Update*. JACC HF 2018

Thicker Muscle: Slows You Down?

COLLECT

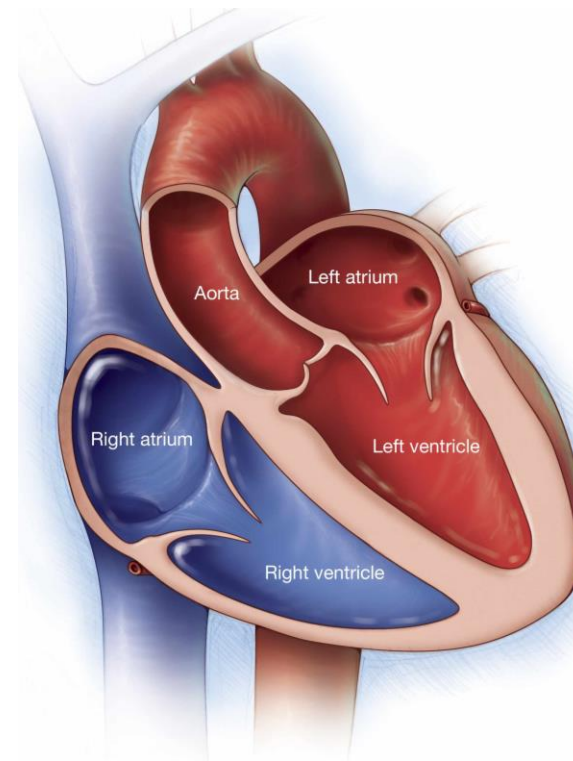
- Atria Upstream

DIRECT

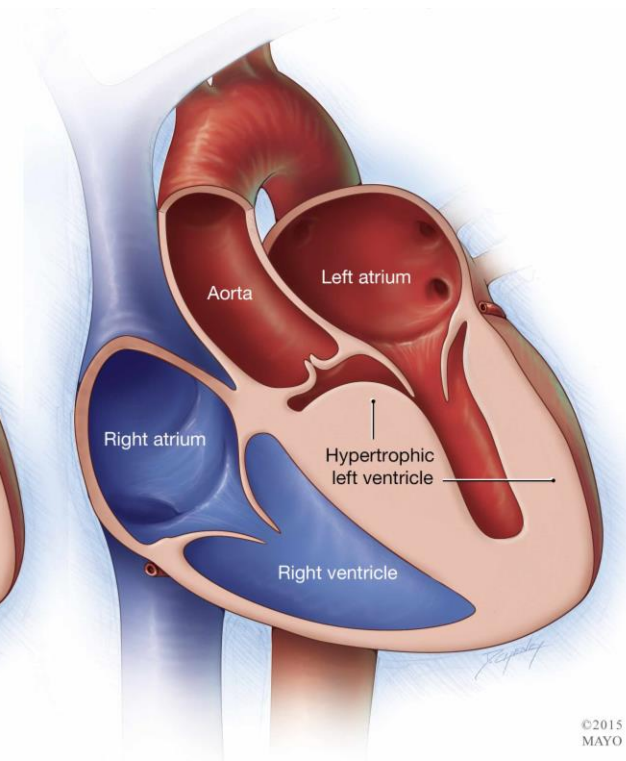
- Valves: Leak
- Septum: Thickened (Obstruction)

EJECT

- Ventricular Function: Relax/Squeeze
- Obstruction: Blood Flow (Heart/Body)
- Disorganized/Scar: Arrhythmias



Normal



**Hypertrophic
Cardiomyopathy**



Thicker Muscle: Slows You Down?

COLLECT

- Atria Upstream

DIRECT

- Valves: Leak
- Septum: Thickened (Obstruction)

EJECT

- Ventricular Function: Relax/Squeeze
- Obstruction: Blood Flow (Heart/Body)
- Disorganized/Scar: Arrhythmias

Irregular Heart Beat

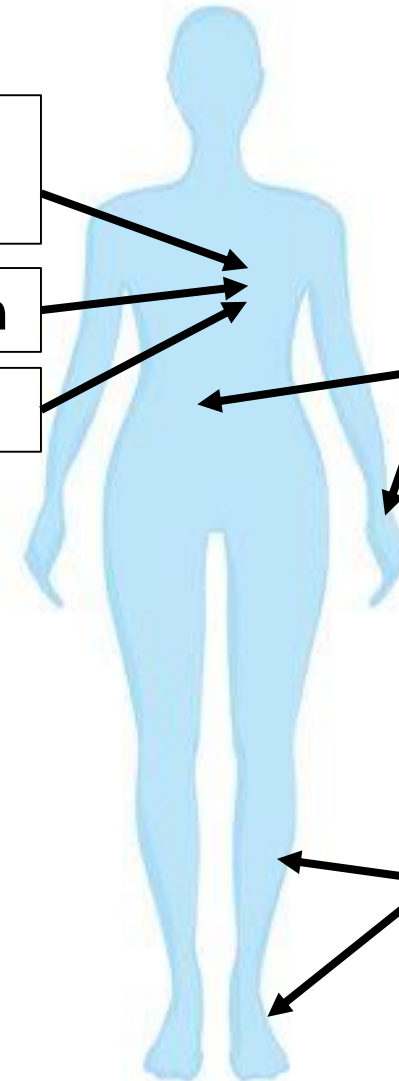
Short of Breath

Chest Pain

Tired
Dizzy
Fainting

Swelling

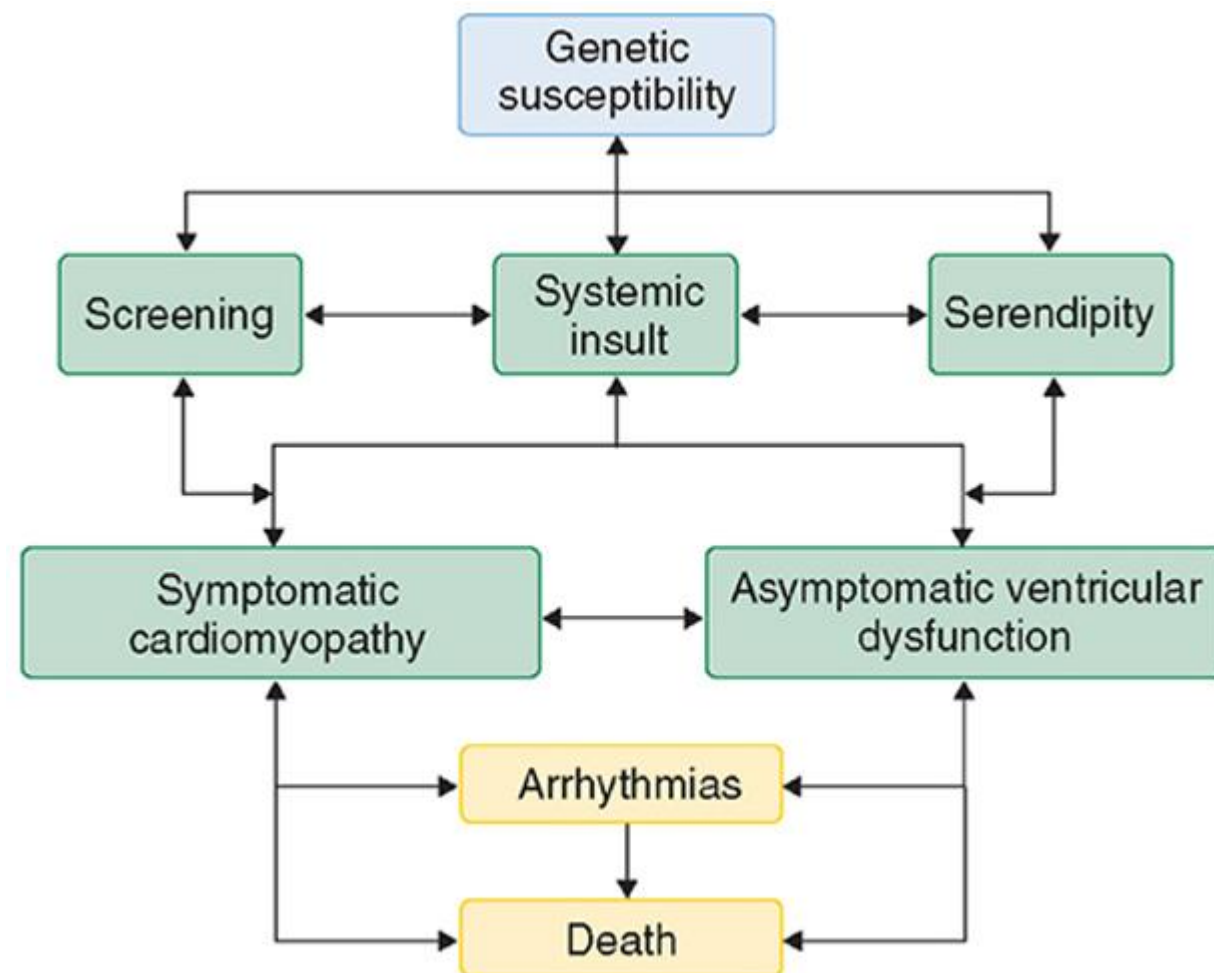
Swelling



Hypertrophic Cardiomyopathy

- Generally: Favorable Prognosis
- Most: Undiagnosed/Asymptomatic
- < 50% Heart Failure¹
 - Heart: Function ≠ Needs
 - Range: Mild ↔ Severe
- < 1% Annual Sudden Cardiac Death²
 - Ventricular Fibrillation

Patient Map



1. Maron et al. *Heart Failure in HCM*. JACC HF 2018

2. O'Mahony et al. *Sudden Death in Hypertrophic Cardiomyopathy*. Circ Arrhythmia Electrophysiology 2013

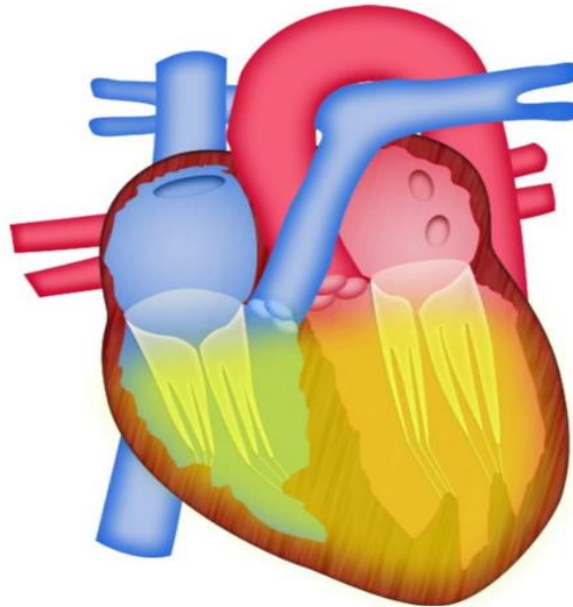
EKG

LUB

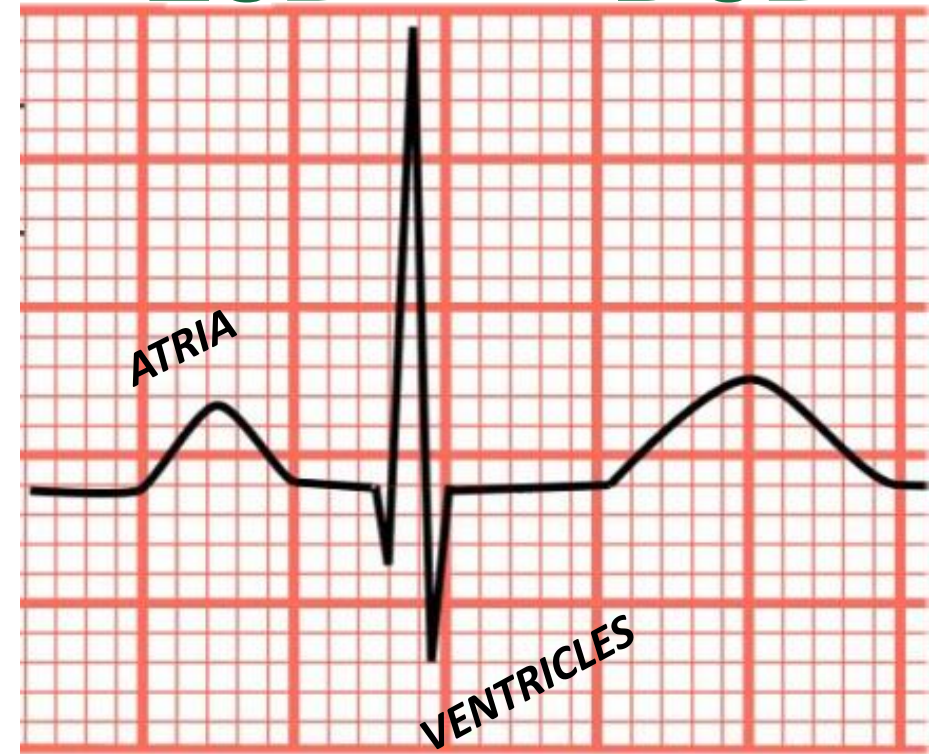
DUB



+



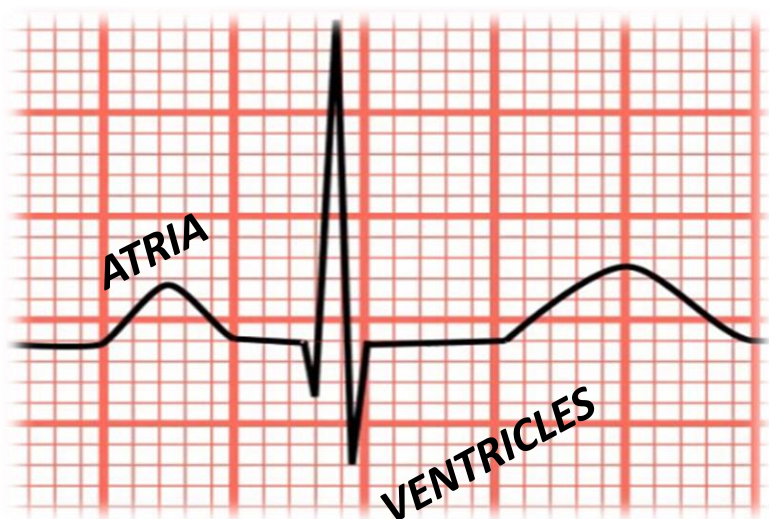
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LUB

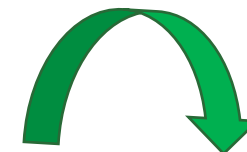
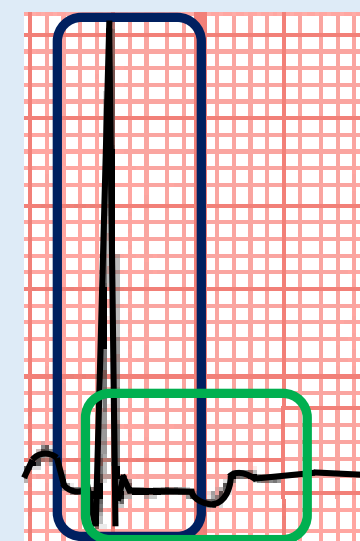
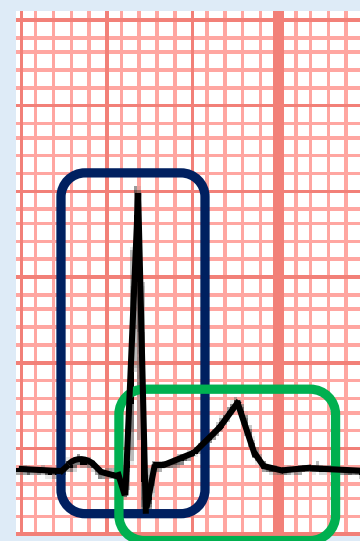
DUB



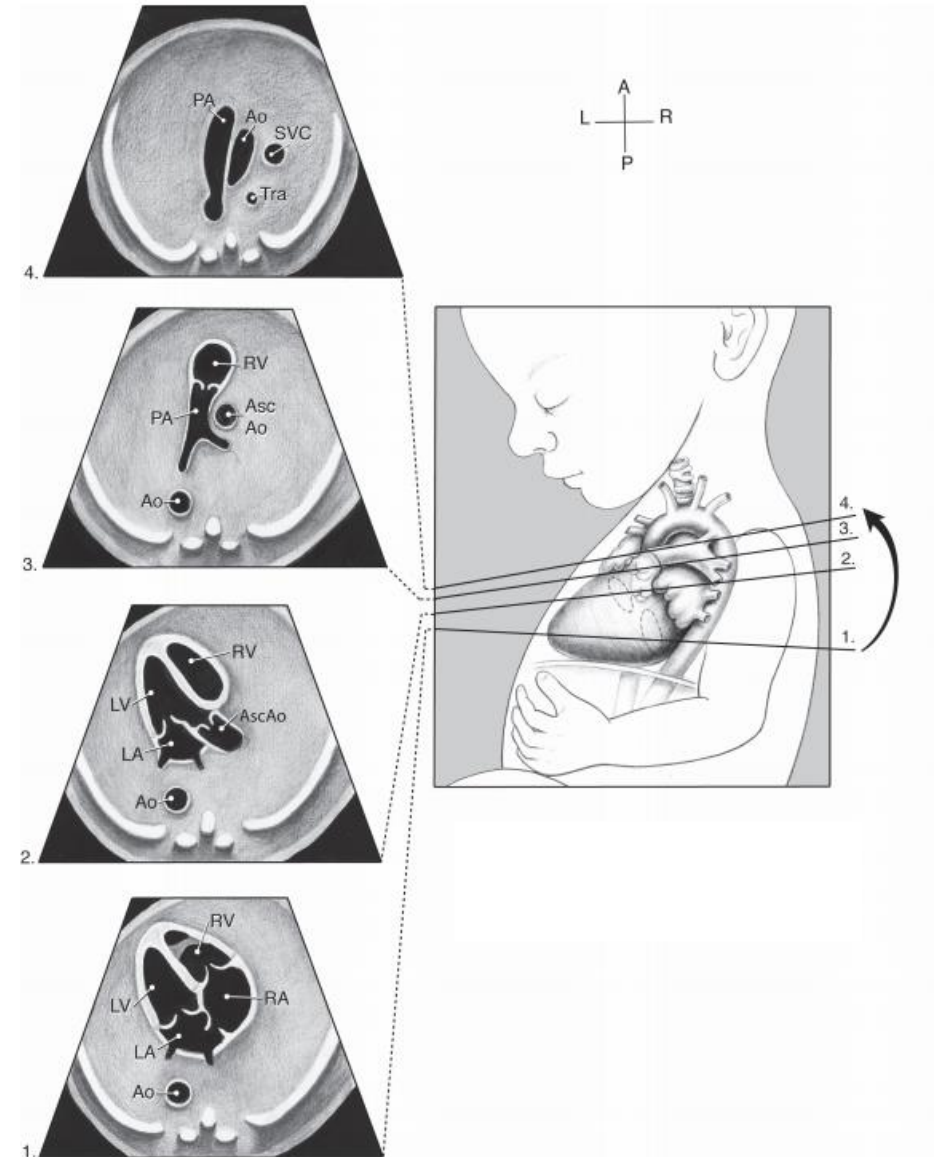
EKG: PHOTO HUNT

NORMAL

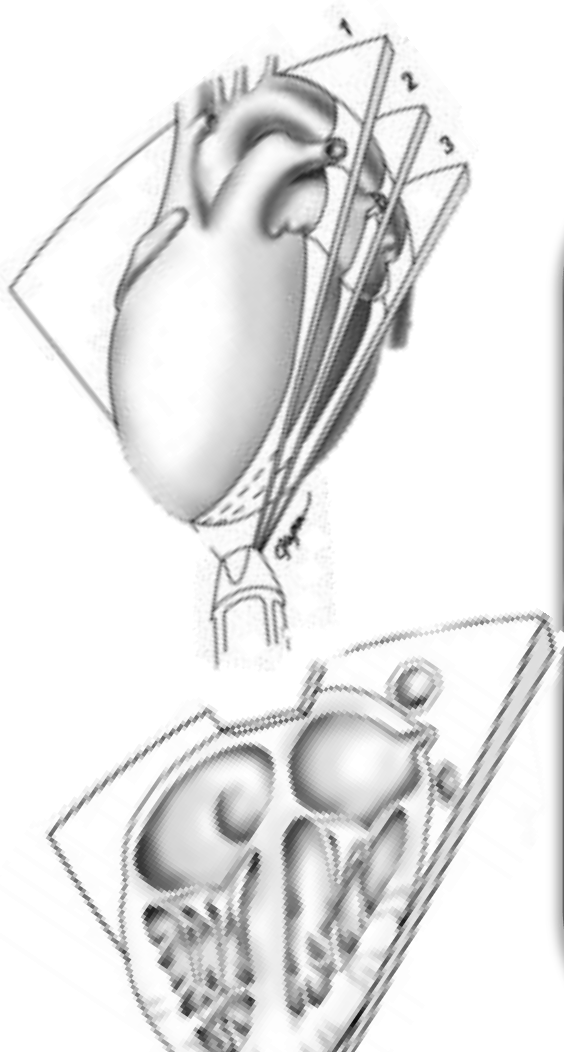
HCM



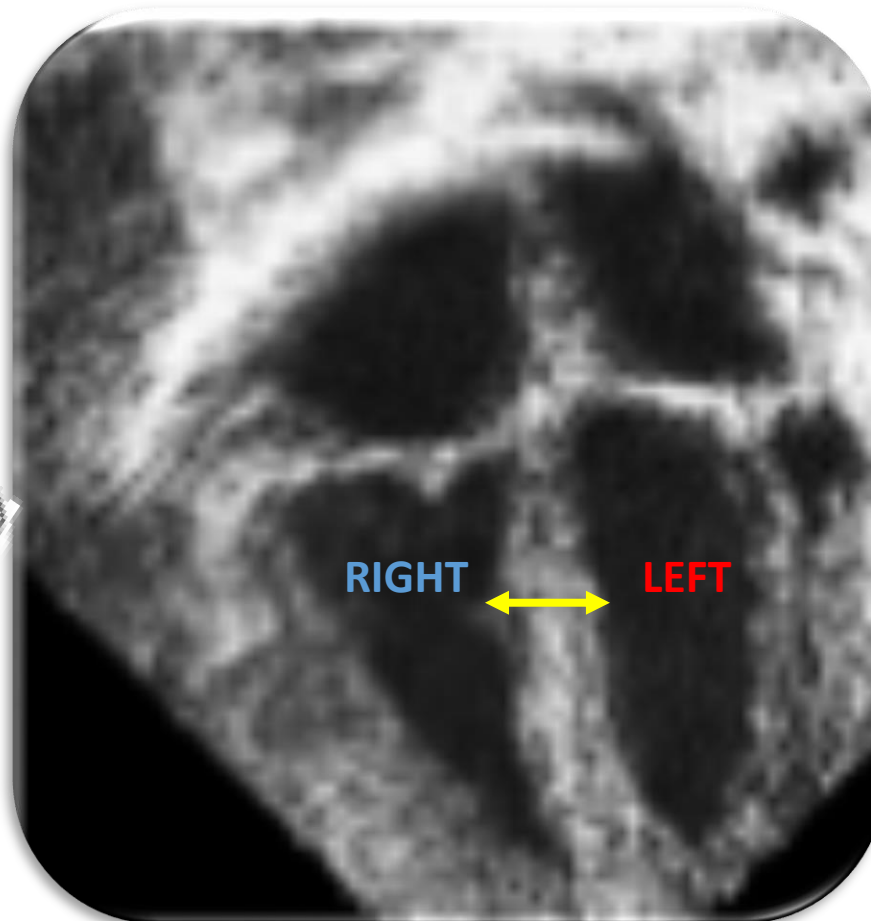
Ultrasound (ECHO)



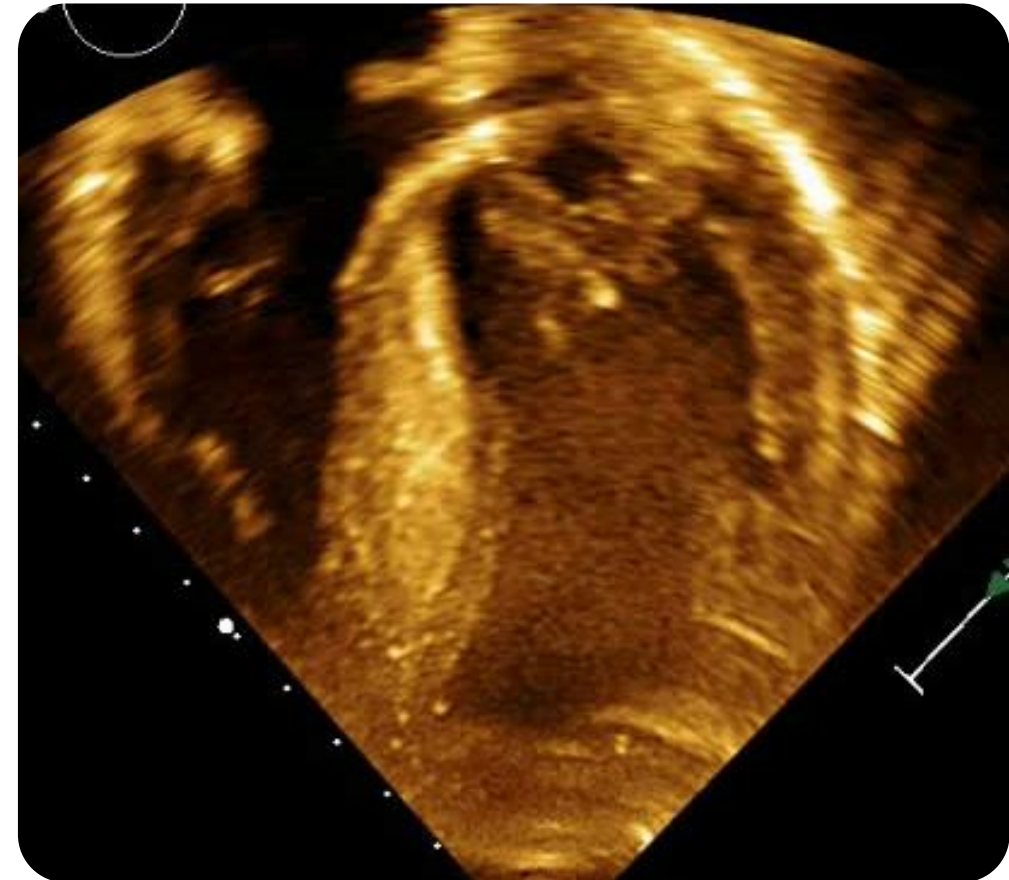
PUMP



NORMAL

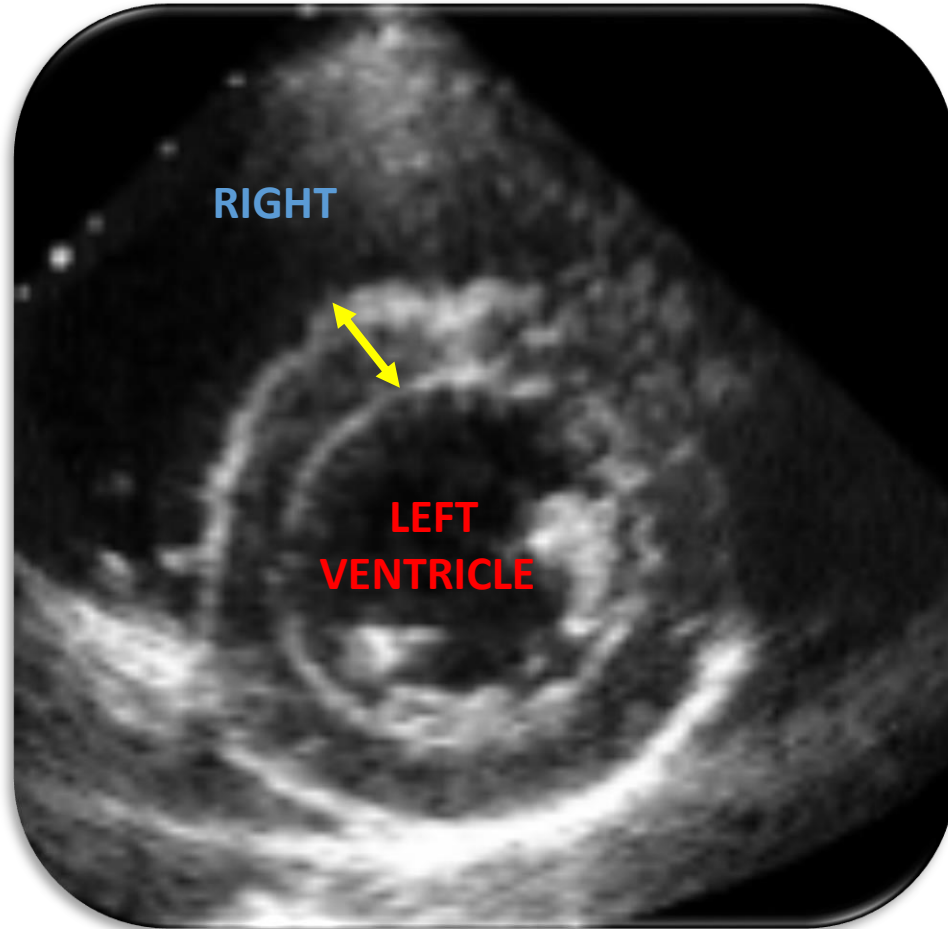


HYPERTROPHIC CARDIOMYOPATHY

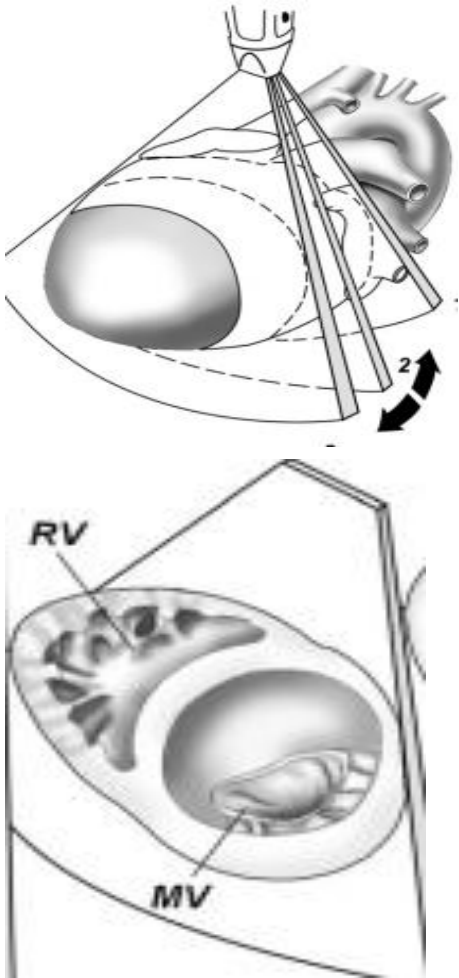


VENTRICULAR SEPTUM

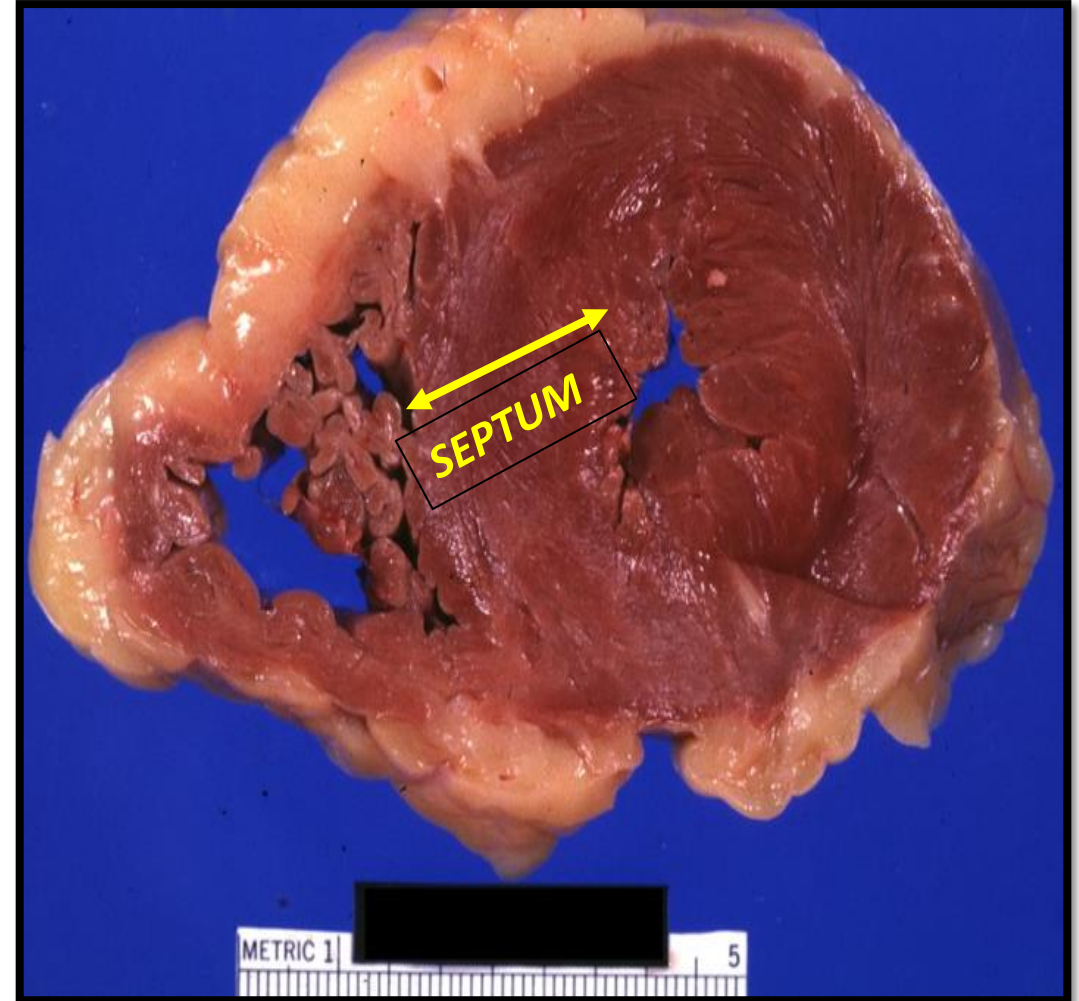
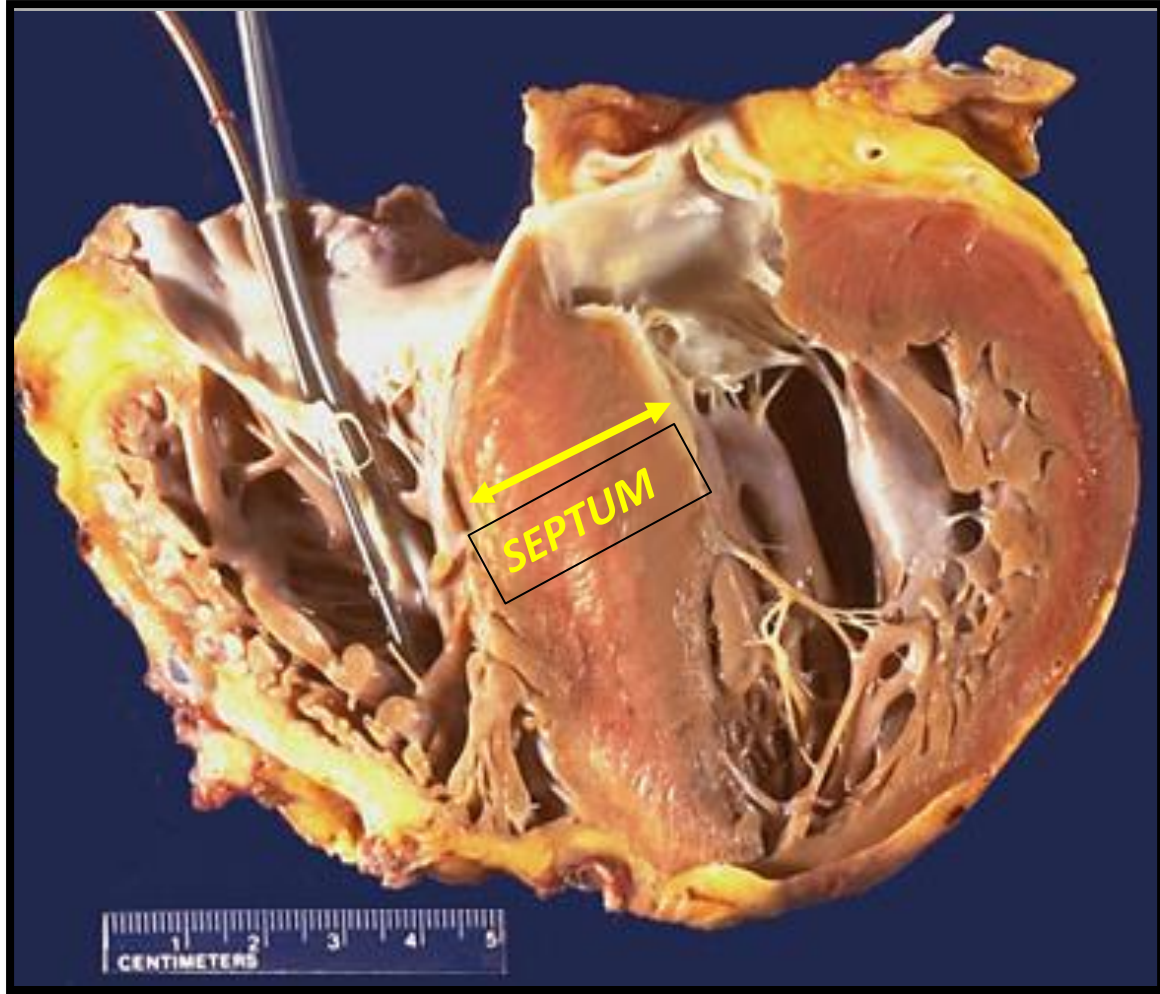
NORMAL



HYPERTROPHIC CARDIOMYOPATHY



Hypertrophic Cardiomyopathy



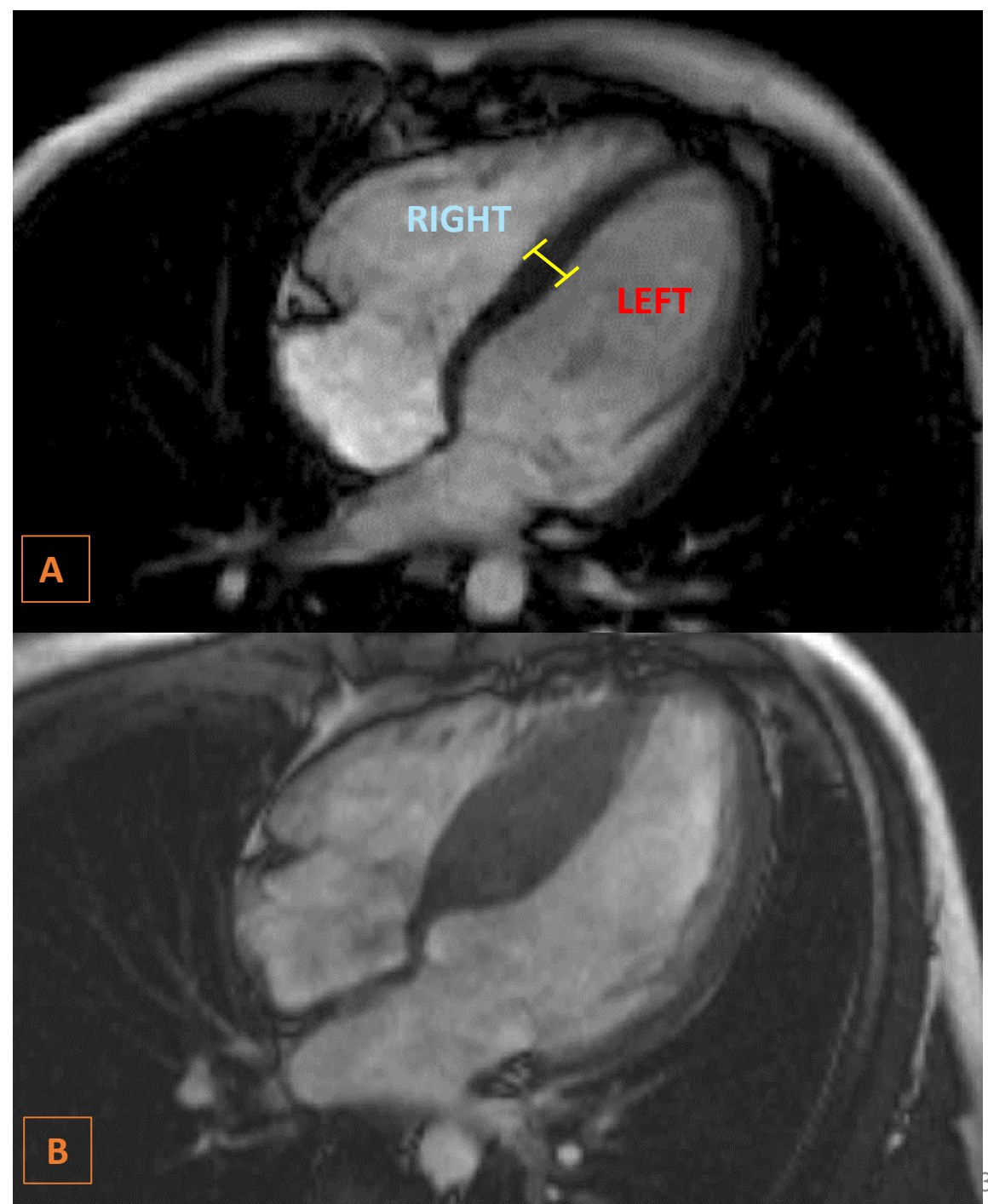


Expert

Active!

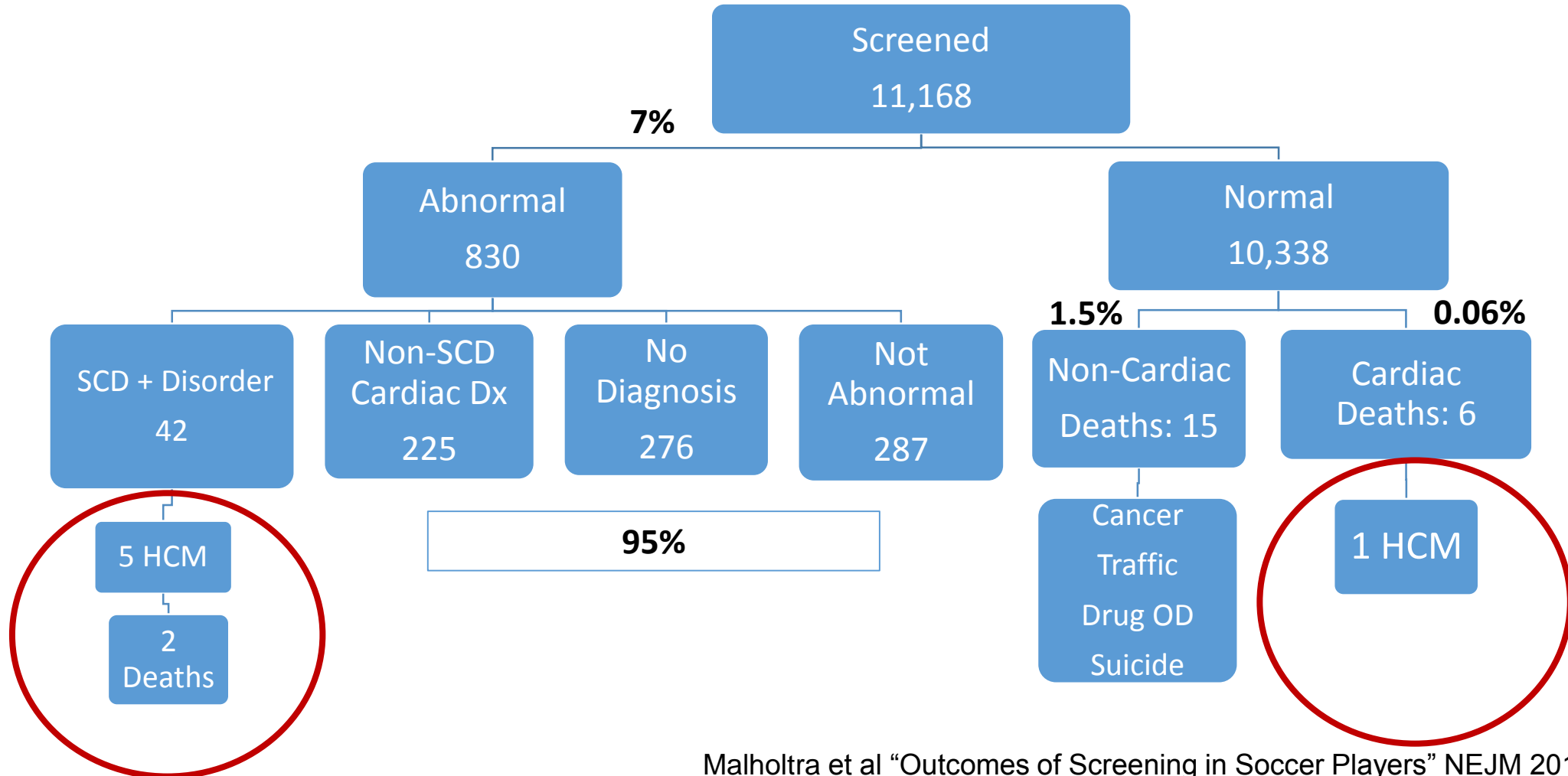
A 21 year old collapses suddenly while running. His EKG and ECHO show left ventricular hypertrophy. An uncle and cousin died suddenly. Genetic studies are pending.

Which cMRI best illustrates hypertrophic cardiomyopathy?



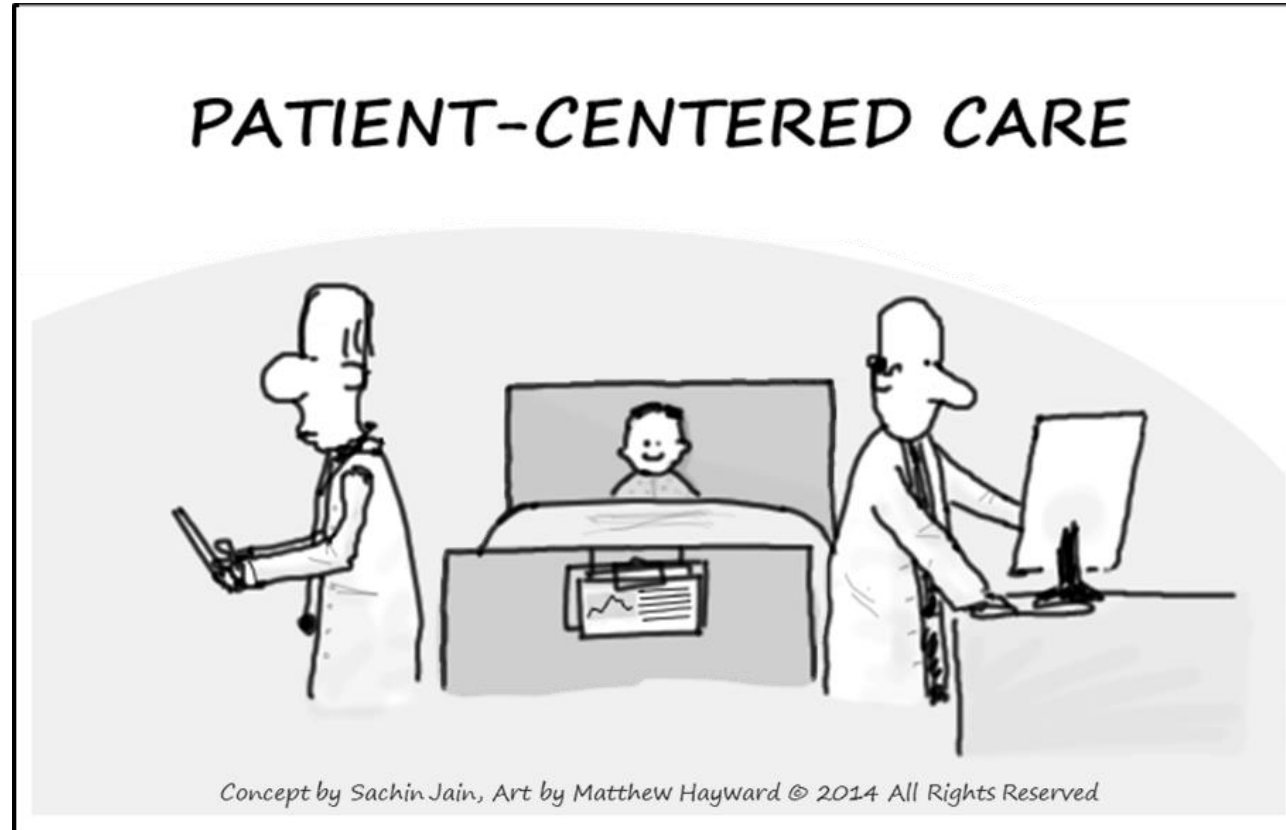
1996-2016: English Soccer Players (15-17yrs)

Screening for Sudden Cardiac Death (SCD)



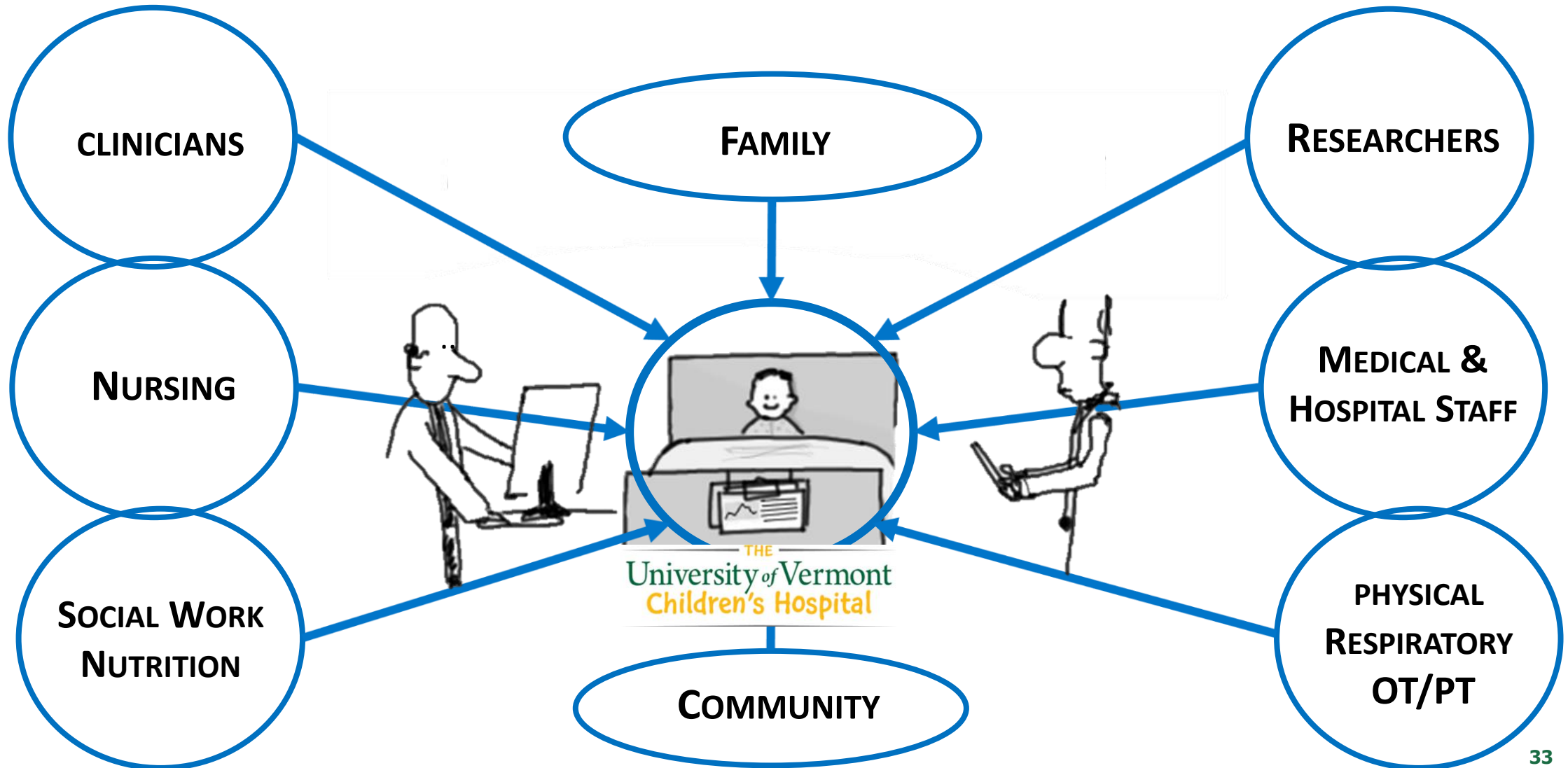
Hypertrophic Cardiomyopathy

Family & Patient Centered Care



Hypertrophic Cardiomyopathy

Family & Patient Centered Care



Hypertrophic Cardiomyopathy

Family & Patient Centered Care

2

Left Ventricular
Hypertrophy

HCM

Medical Focus

Family
Screening

Heart Failure:
Symptoms / Therapy

Sudden
Cardiac
Risk

Shared Discussion

Echo/EKG

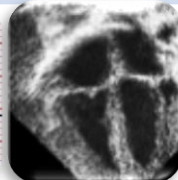
Genetics

Lifestyle

Medication

Surgery¹
vs
Ablation

ICD



PATIENTS & STORIES



A 66 year old woman has her first heart attack.

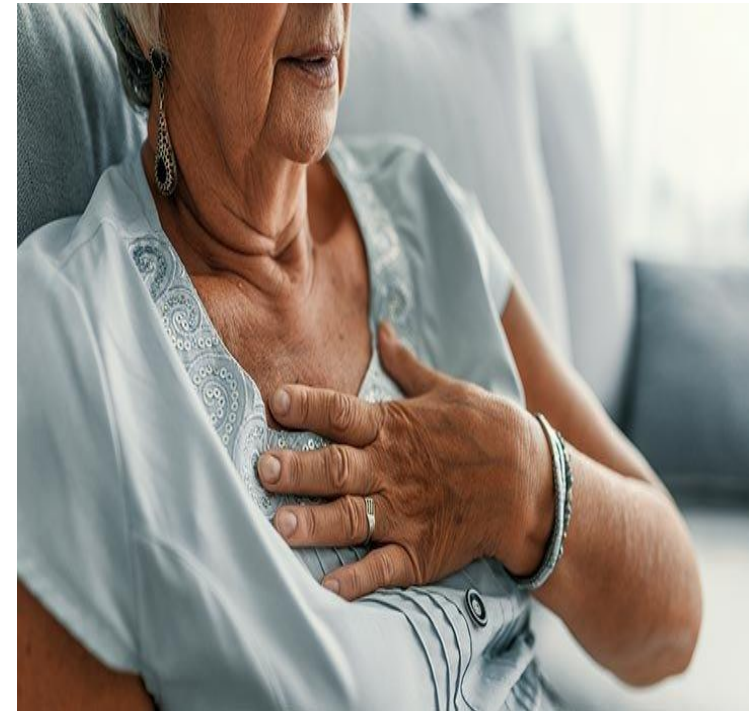
She gets to the hospital.

Then her doctors find another heart condition.



What about my daughter?

...My granddaughter?



**A 16 year old loves adventure.
He wants to talk about backcountry skiing.
You need to talk safety.**

What about my life?

...My sister? Parents? Grandparents?



A 36 year old woman is pregnant.

She talks about her happy 5 year old son.

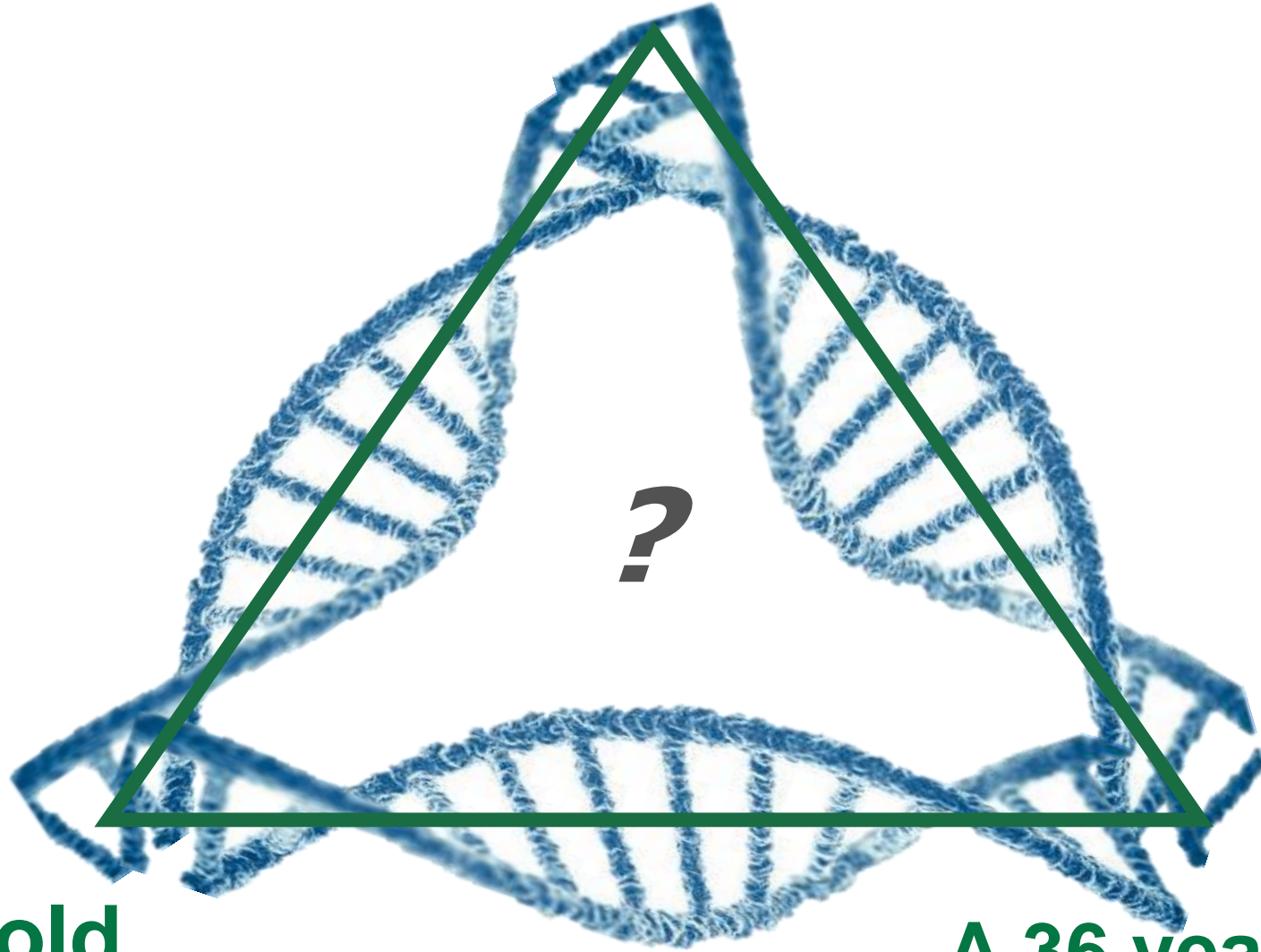
Tattoos cover a scar on her chest.

What about my child?

...My pregnancy?

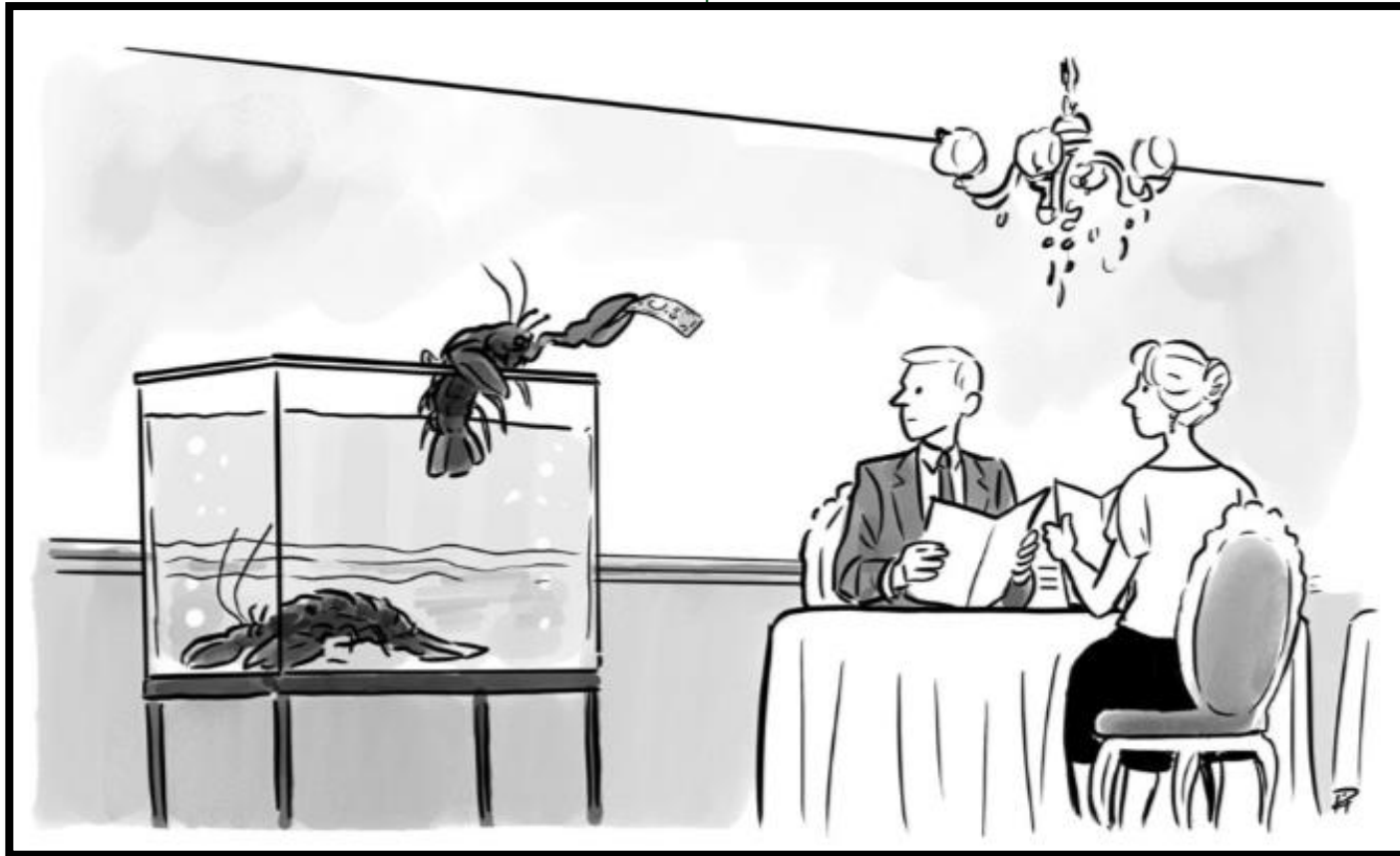


**A 66 year old woman has
her first heart attack.**



**A 16 year old
loves adventure.**

**A 36 year old woman
is pregnant.**



**“I’ll cover your meal if you
pick the other guy.”**

THE
NEW YORKER

February 18 & 25, 2019

THANK YOU



The University of Vermont

LARNER COLLEGE OF MEDICINE

