Risks & Reasons for an Enlarged Heart

UVM Community Medical School
February 19, 2019
Disclosures: None

- Financial Conflicts
- Family Secrets
- Pediatric Cardiologist
OBJECTIVES

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

- Group pre/post test(s)

- Group Discussion
  - Concepts
  - Knowledge
  - Application
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. $\geq 18$ 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
hypertrophy noun

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

2. Exaggerated growth

dilation noun

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

<table>
<thead>
<tr>
<th></th>
<th>Hypertrophy</th>
<th>Dilation</th>
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<tr>
<td><strong>Hypertension</strong></td>
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<td>+++</td>
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#### Diagram:
- **Obstruction**
- **Valve**
- **Communication**
- **Opening**

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**Notes:**
- Hypertrophy
- Dilation
- Hypertension
- Obstruction
- Communication
- Cardiomyopathy
CARDIOMYOPATHY

HYPERTROPHIC

DIABOLIC 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

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Hypertrophic Cardiomyopathy
- Normal
- Dilated Cardiomyopathy

- Hypertrophic
- Left ventricle
- Enlarged
- Thick Walls
- Enlarged Thin Walls

- Aorta
- Left atrium
- Right atrium
- Right ventricle
- Left ventricle
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
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
Hypertrophic Cardiomyopathy

Genetic Cause: 34% of HCM\textsuperscript{2}

- Autosomal Dominant
- >11 genes
- >1500 mutations

2. Geske et al. *HCM Clinical Update*. JACC HF 2018
Hypertrophic Cardiomyopathy

Most Common: Heritable Cardiomyopathy
- Historically: 1/500
- Muscle: Contractile Proteins
- 122 Countries (Map: Red)¹

Unexplained: Left Ventricular Hypertrophy
- > 15mm
- ≥ 13mm (+ First Degree Relative)

1. Maron et al. *Global Burden of HCM. JACC HF 2018*
2. Geske et al. *HCM Clinical Update. JACC HF 2018*
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Thicker Muscle: Slows You Down?

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 Failure\(^1\)
  - Heart: Function ≠ Needs
  - Range: Mild ↔ Severe
- < 1% Annual Sudden Cardiac Death\(^2\)
  - Ventricular Fibrillation

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

+  

=  

LUB  

DUB  

ATRIA  

VENTRICLES
EKG: PHOTO HUNT

Lub  Dub

Normal

HCM

ATRIA

VENTRICLES
Ultrasound (ECHO)
NORMAL

HYPERTROPHIC CARDIOMYOPATHY

RIGHT ⬅️ LEFT
VENTRICULAR SEPTUM

NORMAL

HYPERTROPHIC CARDIOMYOPATHY

RIGHT

LEFT VENTRICLE
Hypertrophic Cardiomyopathy
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)**

- **Screened**: 11,168
  - **Abnormal**: 830
    - SCD + Disorder: 42
    - Non-SCD Cardiac Dx: 225
    - No Diagnosis: 276
    - Not Abnormal: 287
  - **Normal**: 10,338
    - Non-Cardiac Deaths: 15
      - Cancer
      - Traffic
      - Drug OD
      - Suicide
    - Cardiac Deaths: 6
      - 1 HCM

Malholtra et al “Outcomes of Screening in Soccer Players” NEJM 2018
Hypertrophic Cardiomyopathy
Family & Patient Centered Care

PATIENT-CENTERED CARE

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Hypertrophic Cardiomyopathy
Family & Patient Centered Care

CLINICIANS

NURSING

SOCIAL WORK NUTRITION

FAMILY

RESEARCHERS

MEDICAL & HOSPITAL STAFF

PHYSICAL RESPIRATORY OT/PT

COMMUNITY
Hypertrophic Cardiomyopathy

Family & Patient Centered Care

Left Ventricular Hypertrophy

Medical Focus

Shared Discussion

Echo/EKG  Genetics  Lifestyle  Medication  Surgery 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.”
THANK YOU

The University of Vermont
LARNER COLLEGE OF MEDICINE

COMMUNITY MEDICAL SCHOOL