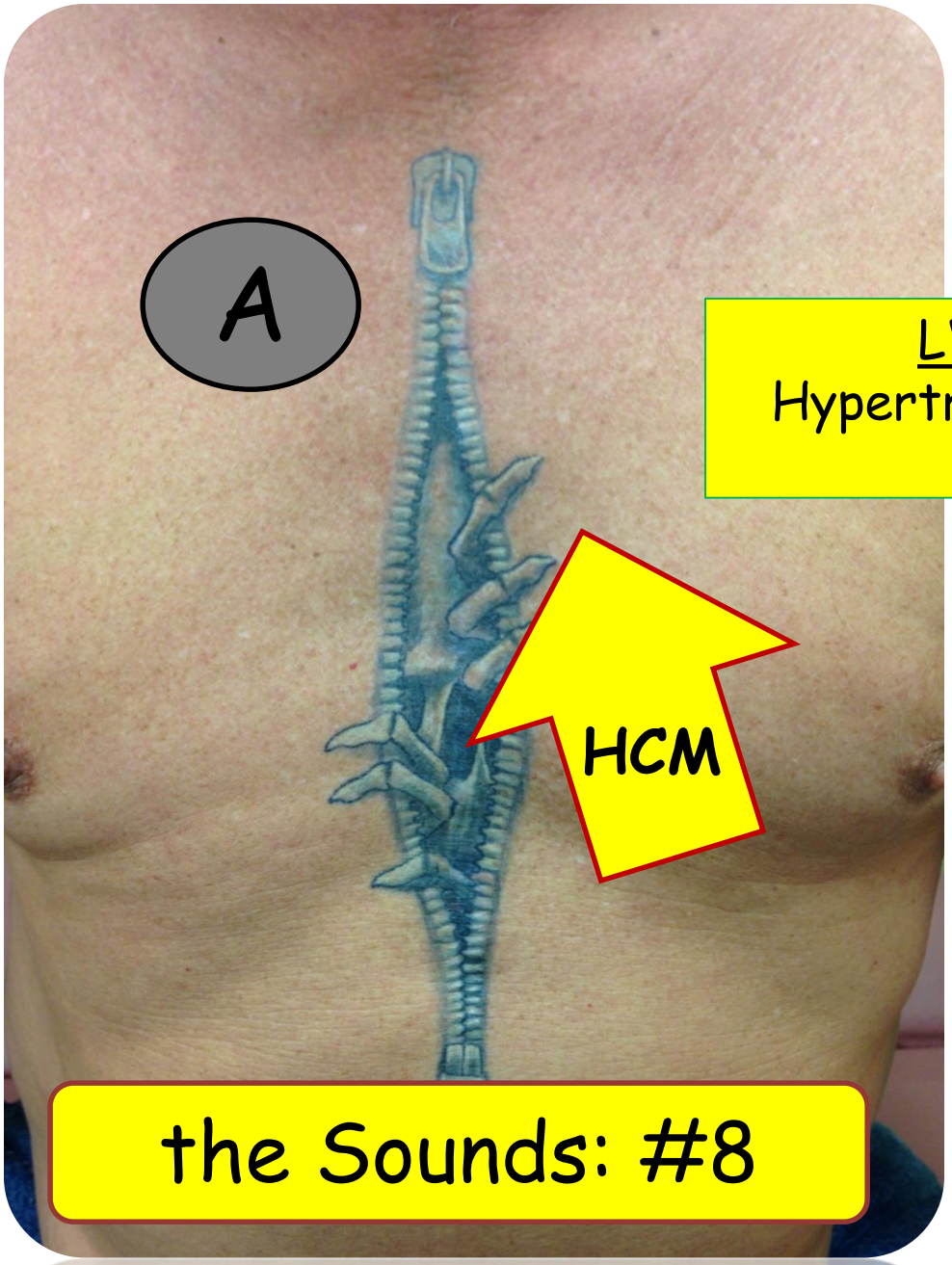

Cardiology

LV Outflow Obstruction:
Hypertrophic Cardiomyopathy (HCM)
(s/p Aortic Stenosis)

Howard J. Sachs, MD
www.12DaysinMarch.com
E-mail: Howard@12daysinmarch.com

HCM *(Hypertrophic Cardiomyopathy)*

- Background
  - Obstructive physiology → *Dynamic Outflow Gradient*
  - Pathology description: *Myocyte Disarray*
    - Key Derivative: *Syncope* or *SCD* (especially in the young athlete)
- Definition/Nomenclature/Pathology
  - HCM is…
    - Genetics
  - HCM is NOT…
- Physical Exam
  - Maneuvers
  - Applied physiology/pharmacology
- Questions/Vignettes
  - Bring this bad boy home…

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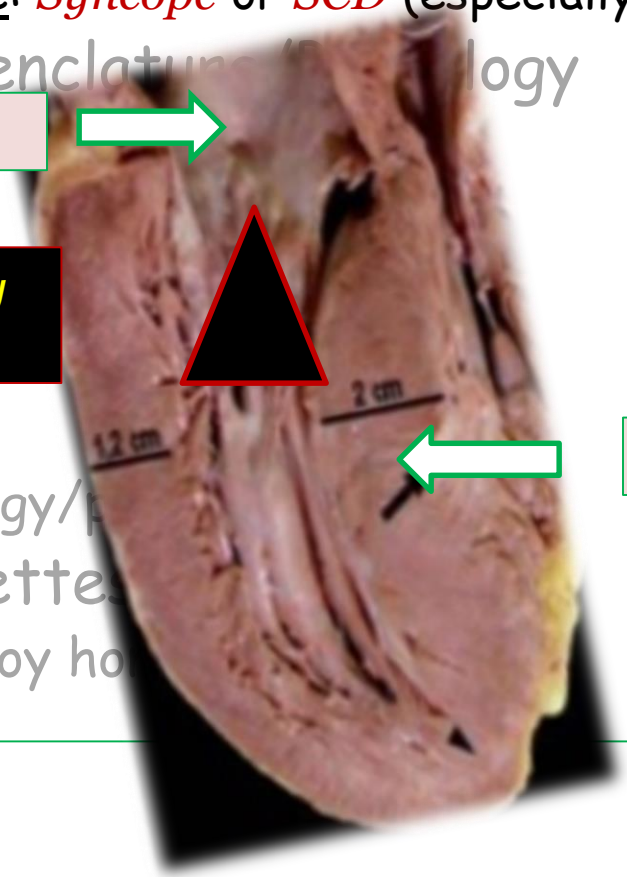
    - Key Derivative: *Syncope* or *SCD* (especially in the young athlete)
- Definition/Nomenclature/Epidemiology
  - Lower Pressures
    - Genetics
  - *Obstructive Physiology Creates Gradient*
  - Maneuvers
  - Applied physiology/p
- Questions/Vignettes
  - Bring this bad boy ho



High Pressures

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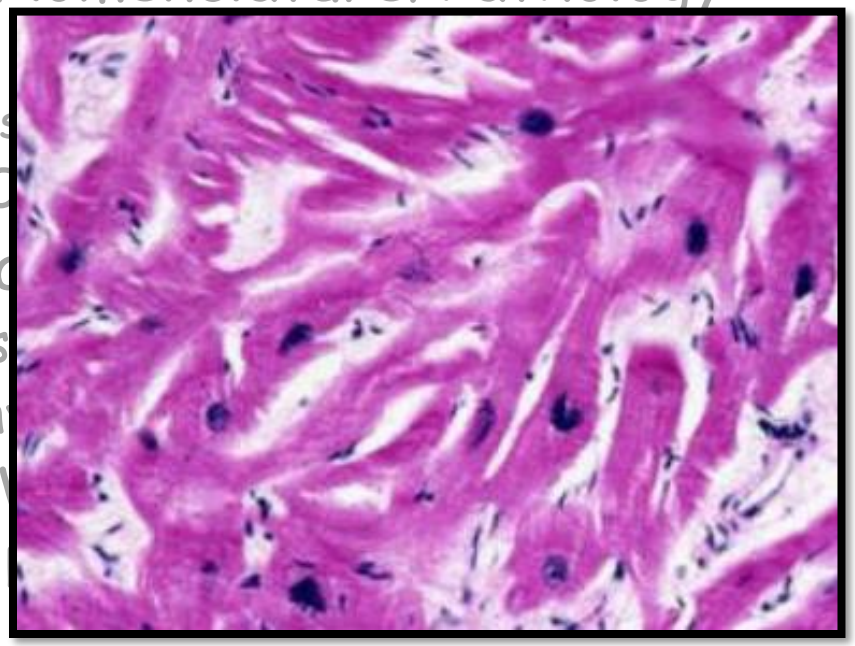
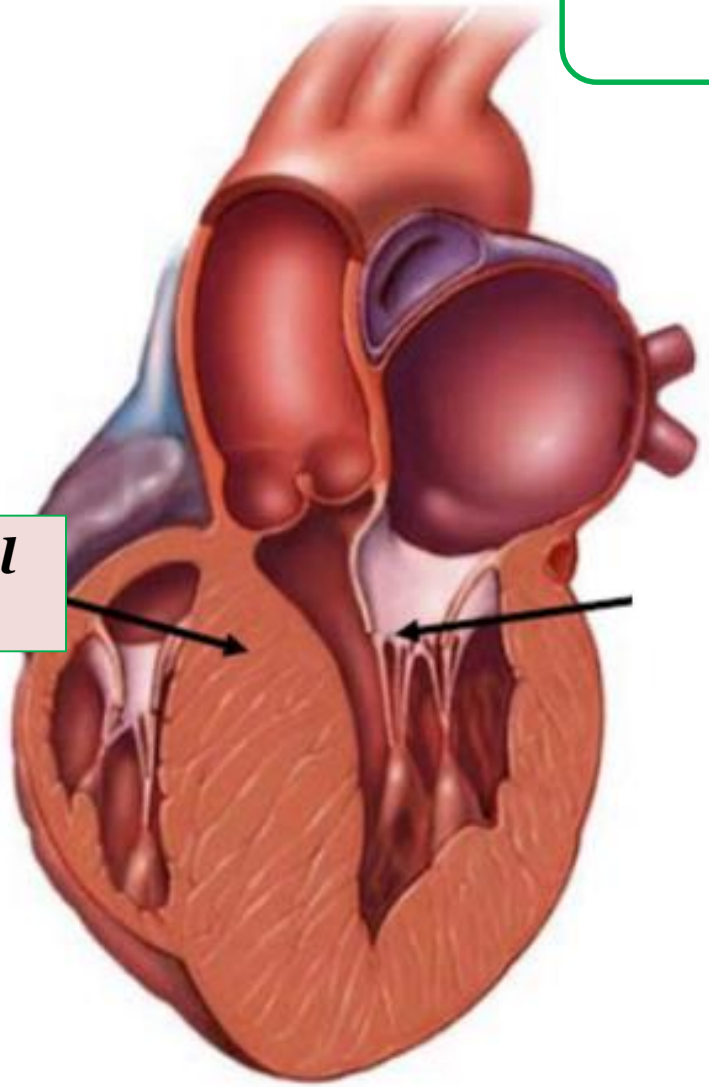
  - HCM is NC
- Physical Exa
  - Maneuvers
  - Applied ph
- Questions/
  - Bring this



Hypertrophic Cardiomyopathy

1. *Obstructive physiology*

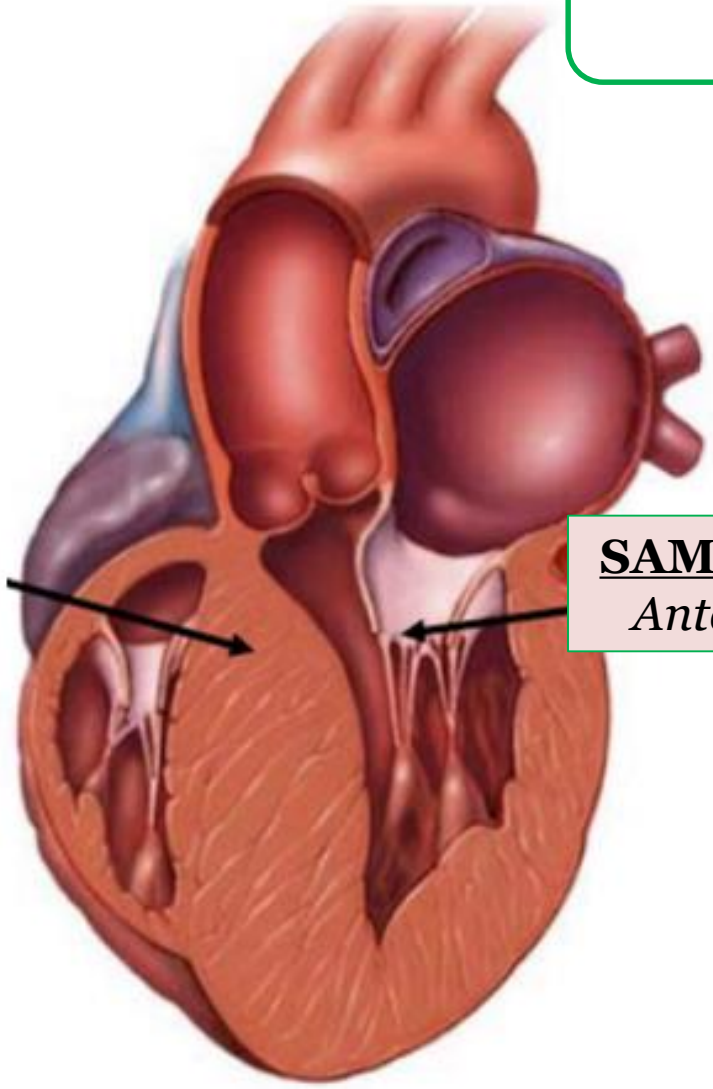


Asymmetric Septal
Hypertrophy

Hypertrophic Cardiomyopathy

1. *Obstructive physiology*

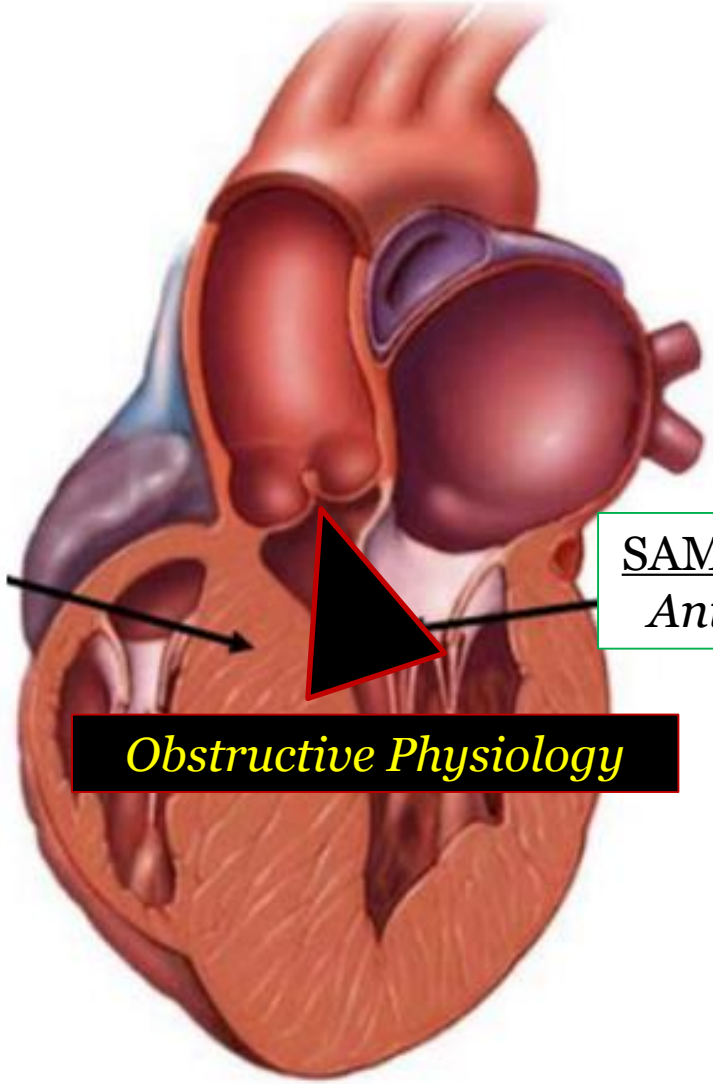
Asymmetric Septal Hypertrophy



SAM (Systolic Anterior Motion)*: Anterior Cusp of Mitral Valve*

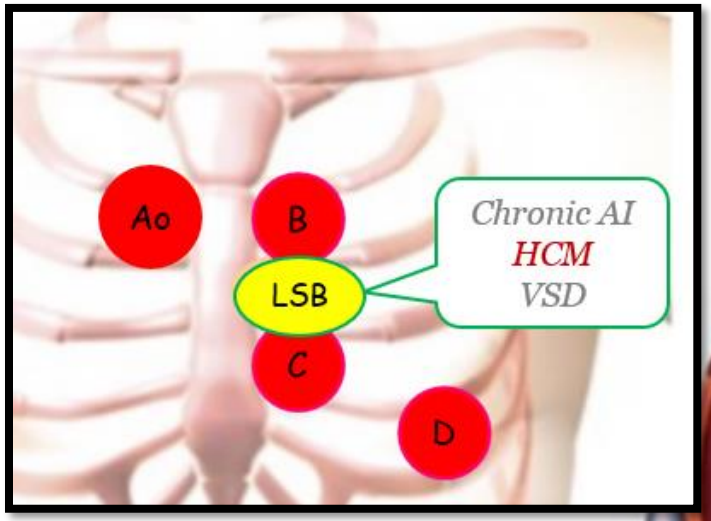
1

Asymmetric Septal Hypertrophy



2

<u>SAM (Systolic Anterior Motion)</u>*: Anterior Cusp of Mitral Valve*



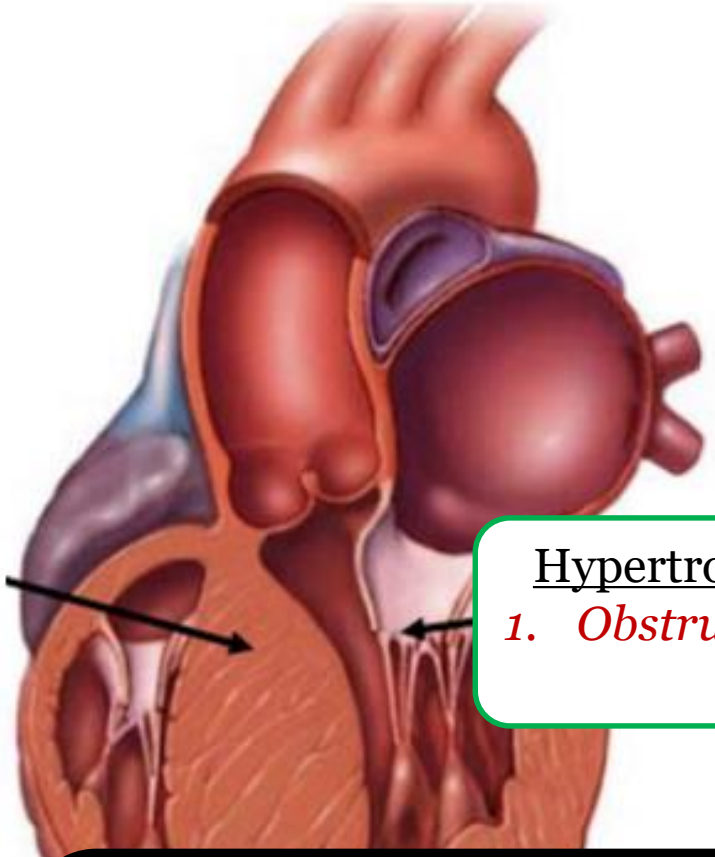
1

Asymmetric Septal Hypertrophy

2

<u>SAM (Systolic Anterior Motion)</u>*: Anterior Cusp of Mitral Valve*

<u>*Key Derivative*</u>:
Obstructive Physiology → **Syncope**



Hypertrophic Cardiomyopathy

1. *Obstructive physiology*

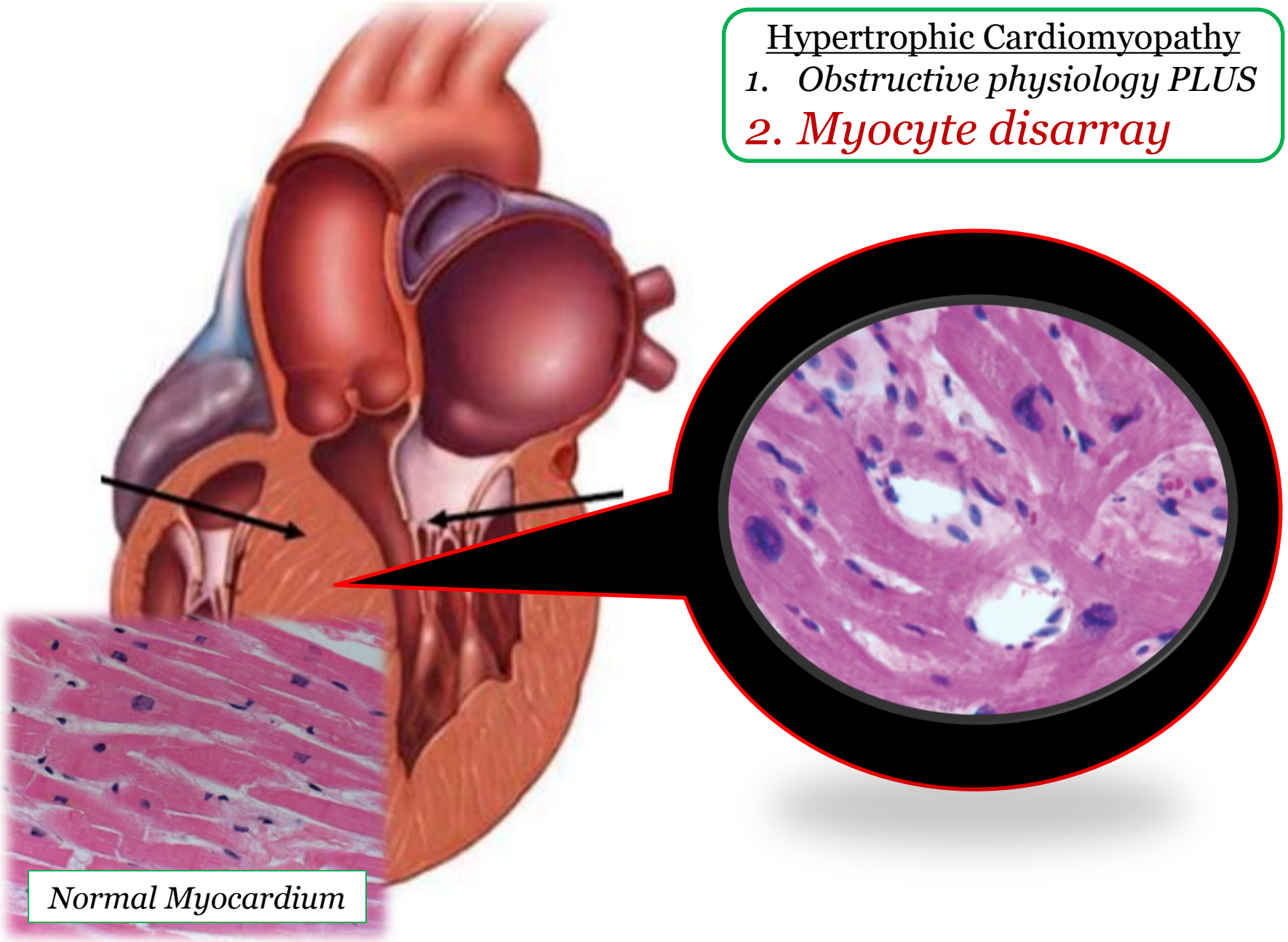
Other Names/Descriptions:

IHSS: Idiopathic Hypertrophic Subaortic Stenosis
ASH: Asymmetric Septal Hypertrophy

Implication is outflow obstruction (in combination with SAM)

Hypertrophic Cardiomyopathy

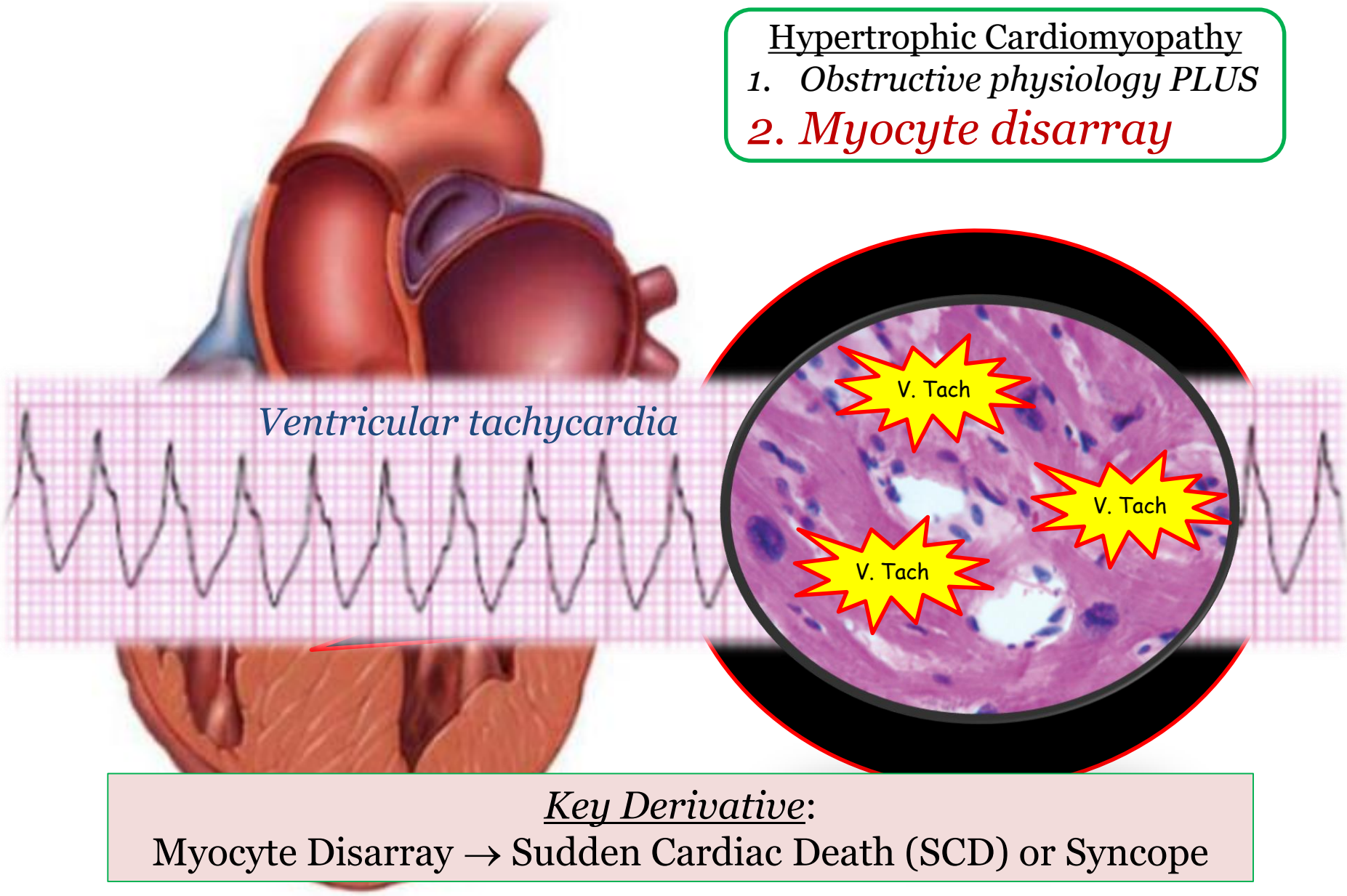
1. *Obstructive physiology PLUS*
2. *Myocyte disarray*



Normal Myocardium

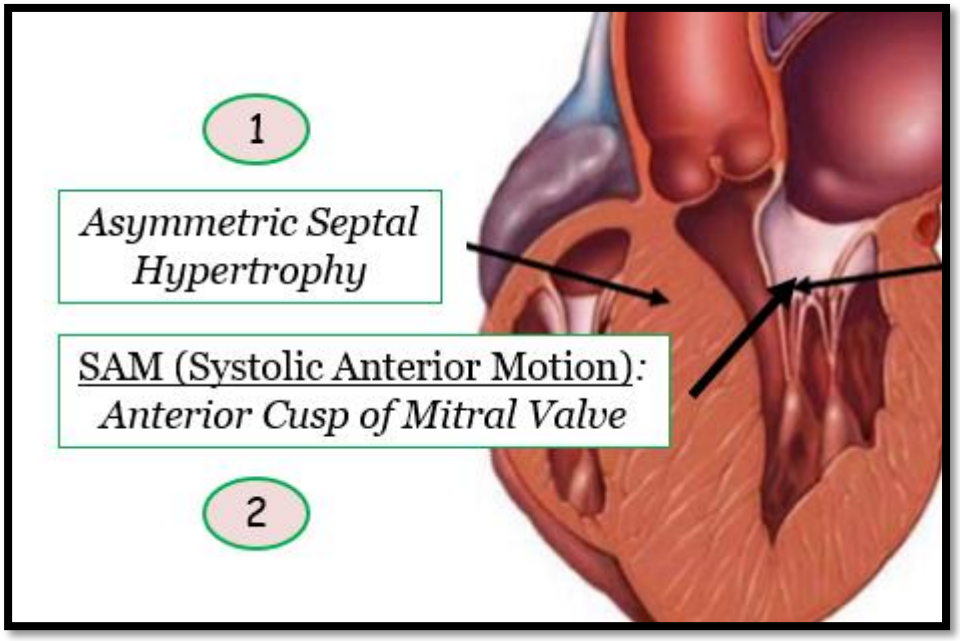
Hypertrophic Cardiomyopathy

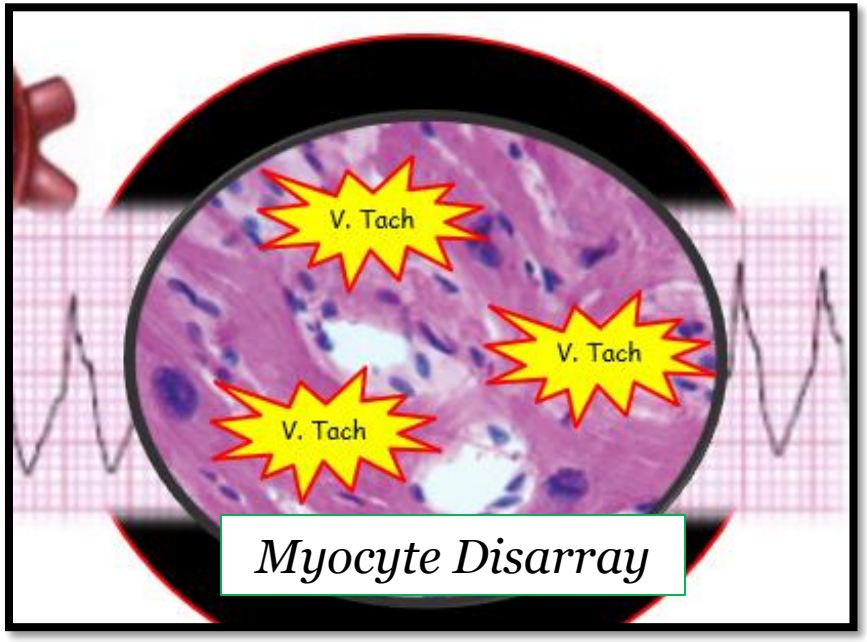
1. *Obstructive physiology PLUS*
2. *Myocyte disarray*



Key Derivative:
Myocyte Disarray → Sudden Cardiac Death (SCD) or Syncope

Syncope:
Obstruction
Arrhythmia





SCD:
Arrhythmia

HCM *(Hypertrophic Cardiomyopathy)*

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- Definition/Nomenclature/*Pathology*
  - HCM is…
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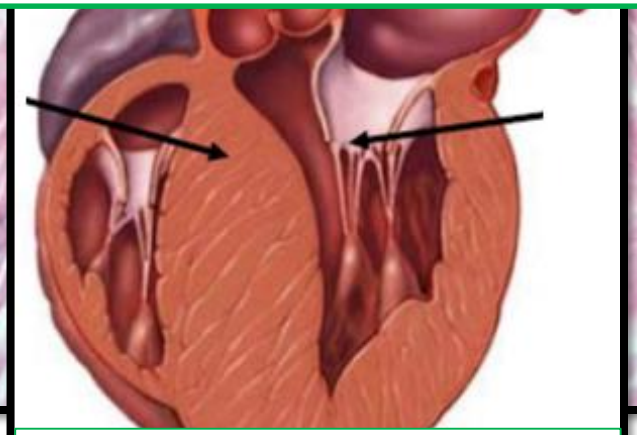
Hypertrophic Cardiomyopathy

Nomenclature

- Refers to a specific class of Genetic (or Familial) Disorders

  HCM Family of Disorders: *Implies dysfunction of the sarcomere*

- Distinct from the class of disorders that result in LVH (left ventricular hypertrophy) as an *adaptive response to increased afterload* (e.g. Aortic Stenosis, HTN).



Genetic: *Hypertrophic*

Adaptive: *Concentric*

Hypertrophic Cardiomyopathy

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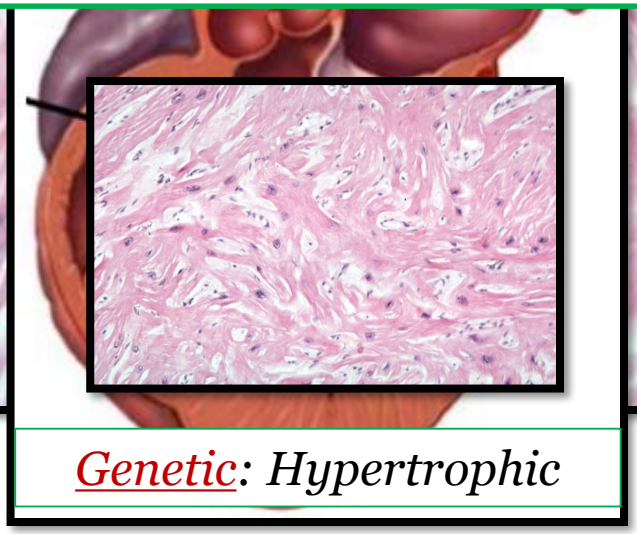

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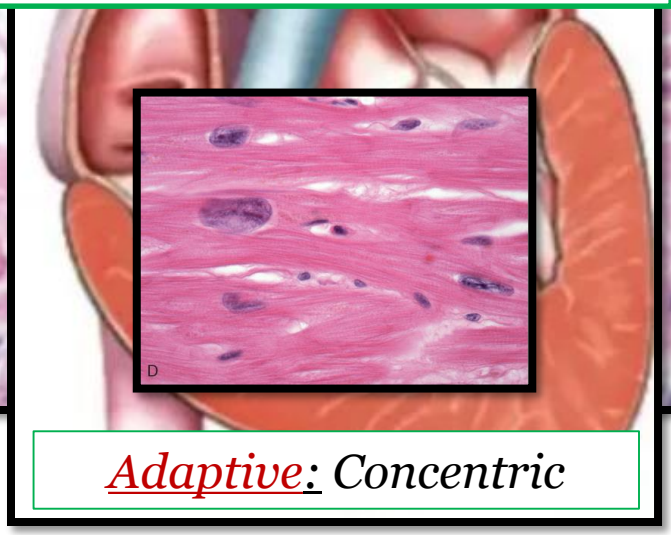
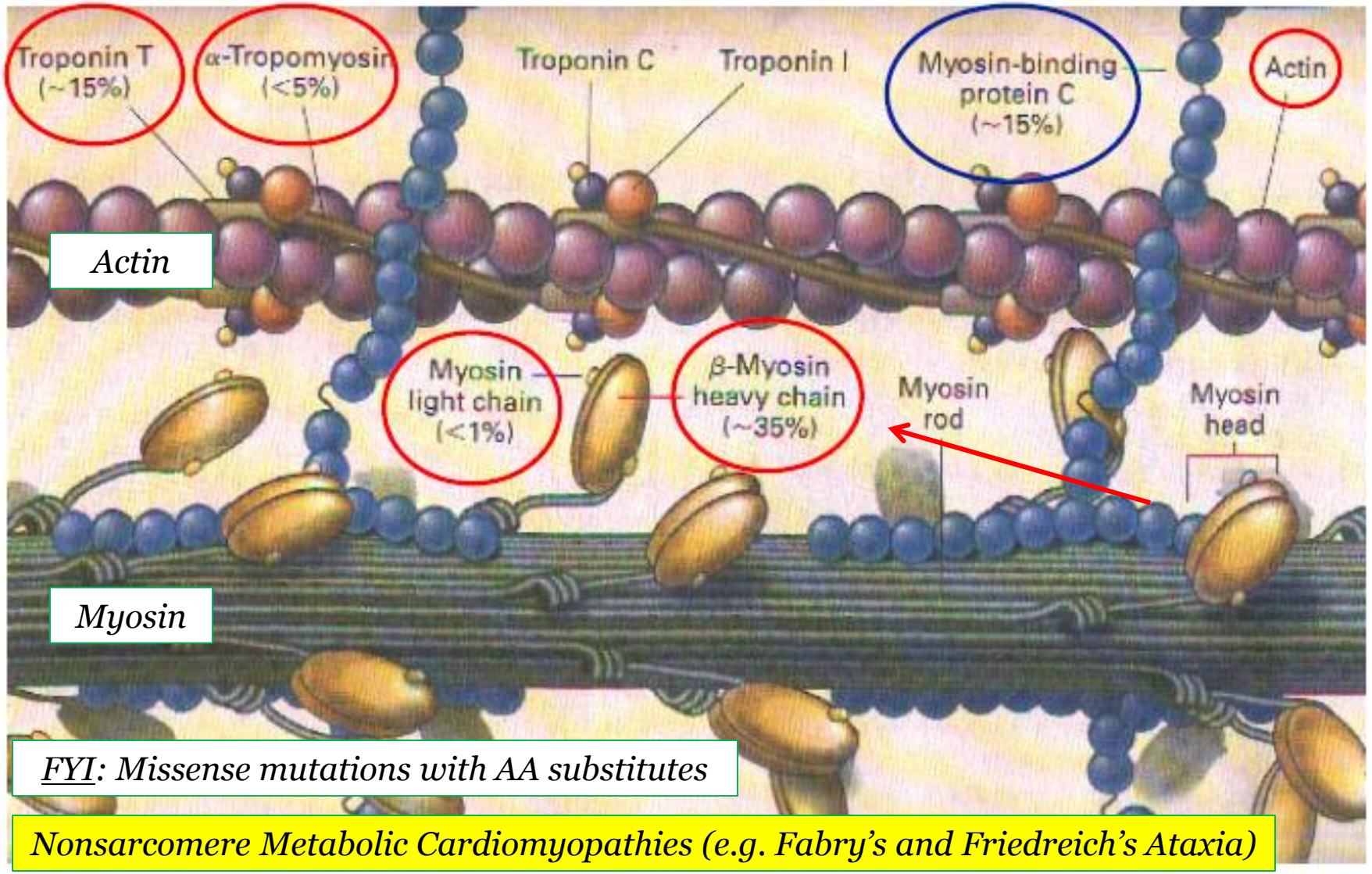
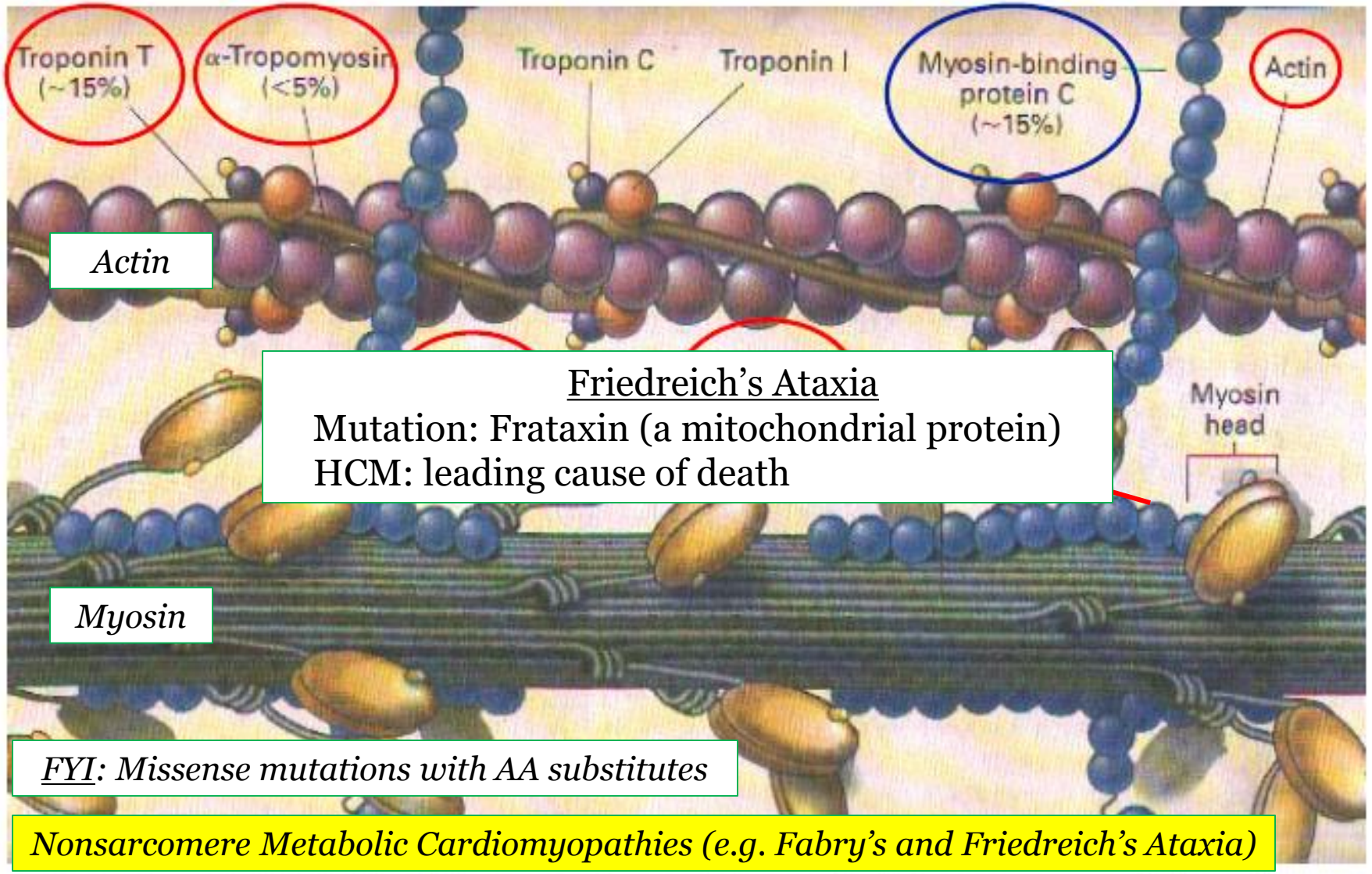


Adaptive: *Concentric*



FYI: Missense mutations with AA substitutes

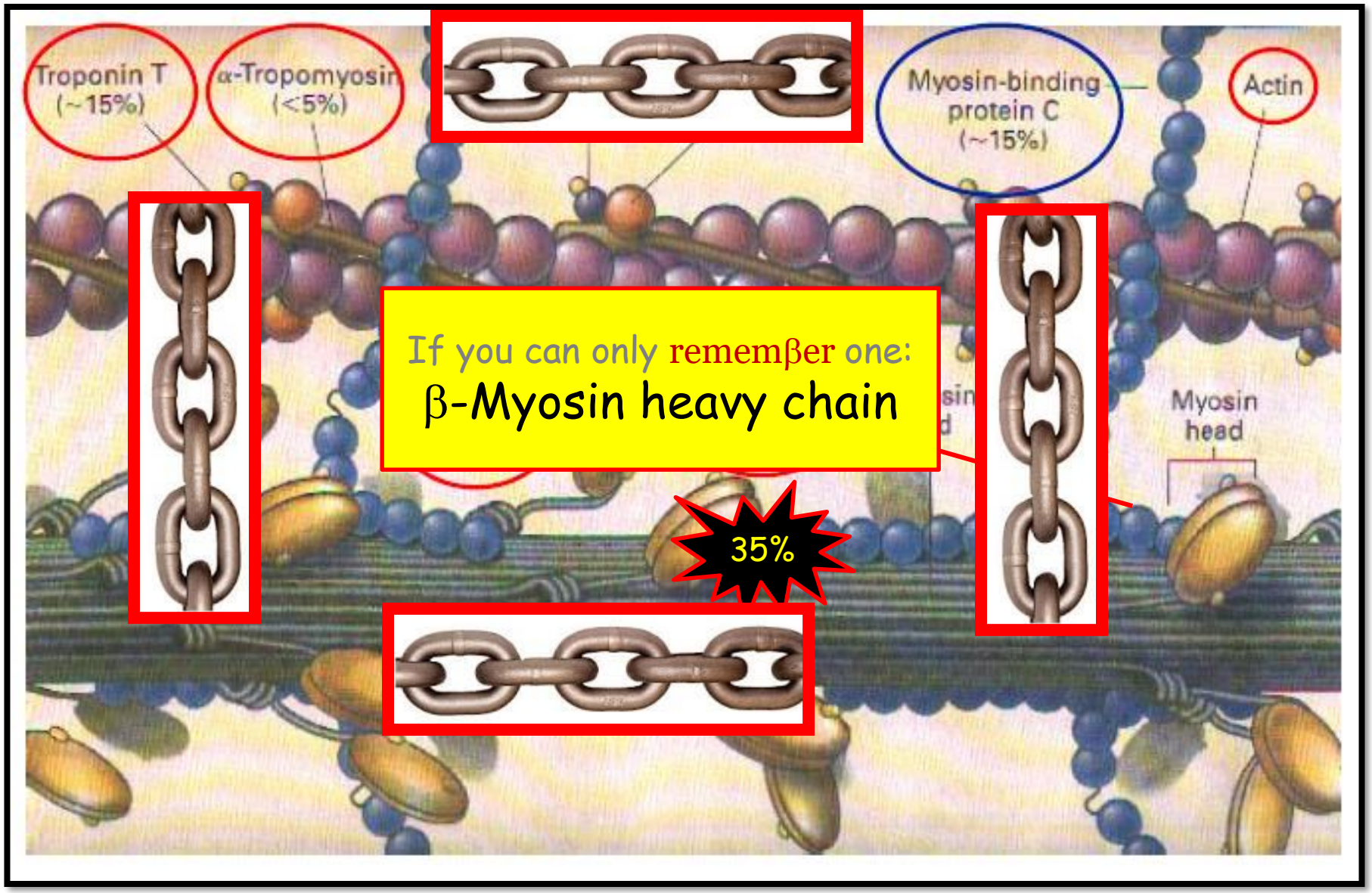
Nonsarcomere Metabolic Cardiomyopathies (e.g. Fabry's and Friedreich's Ataxia)



Friedreich's Ataxia
Mutation: Frataxin (a mitochondrial protein)
HCM: leading cause of death

FYI: Missense mutations with AA substitutes

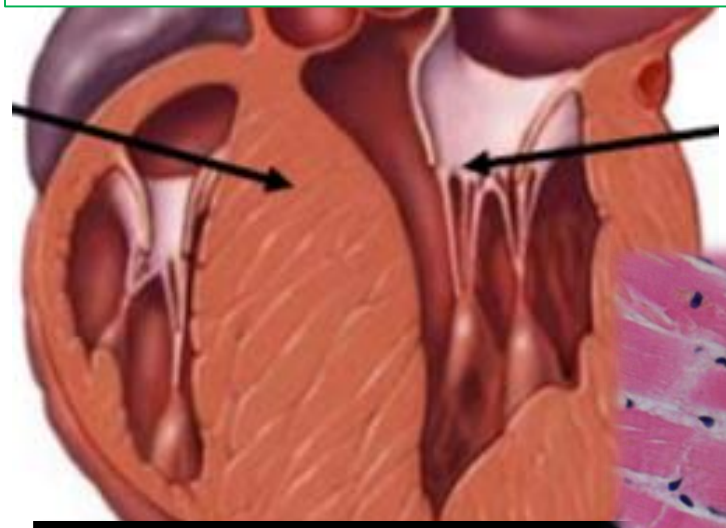
Nonsarcomere Metabolic Cardiomyopathies (e.g. Fabry's and Friedreich's Ataxia)

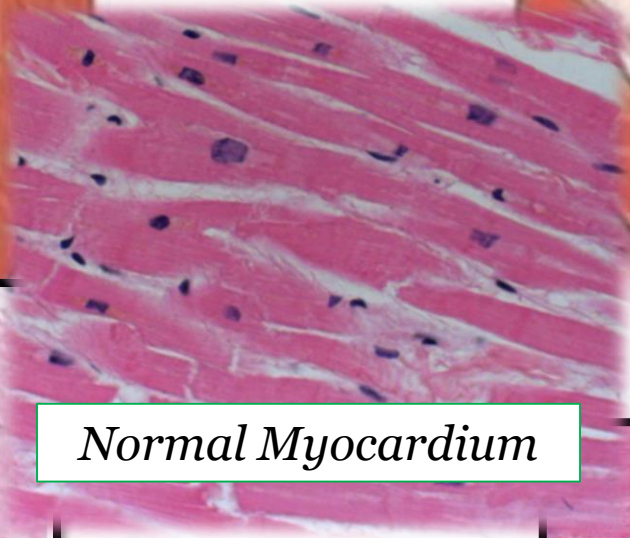


Variety of Genotypes: β-Myosin heavy chain is favorite!

Asymmetric Septal Hypertrophy

Increased Afterload (e.g. AS/HTN)





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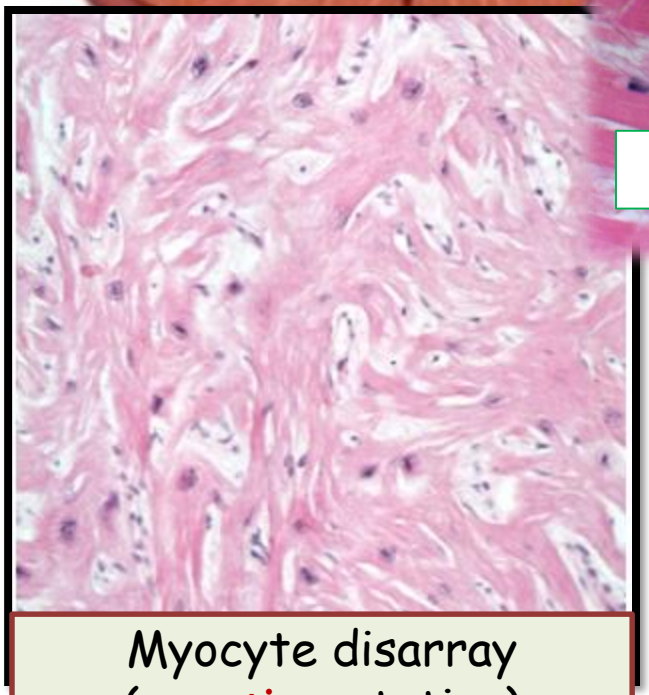
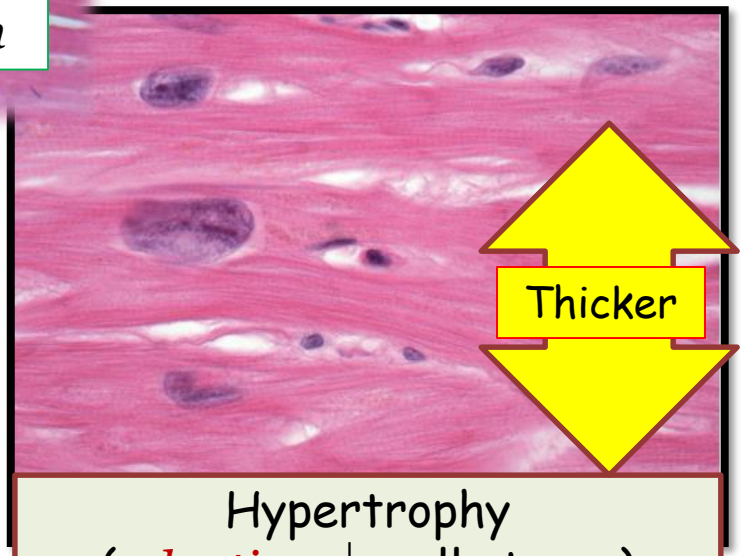


Myocyte disarray
(*genetic* mutation)



Hypertrophy
(*adaptive*: ↓ wall stress)

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  - Maneuvers
  - Applied physiology/pharmacology
- Questions/Vignettes
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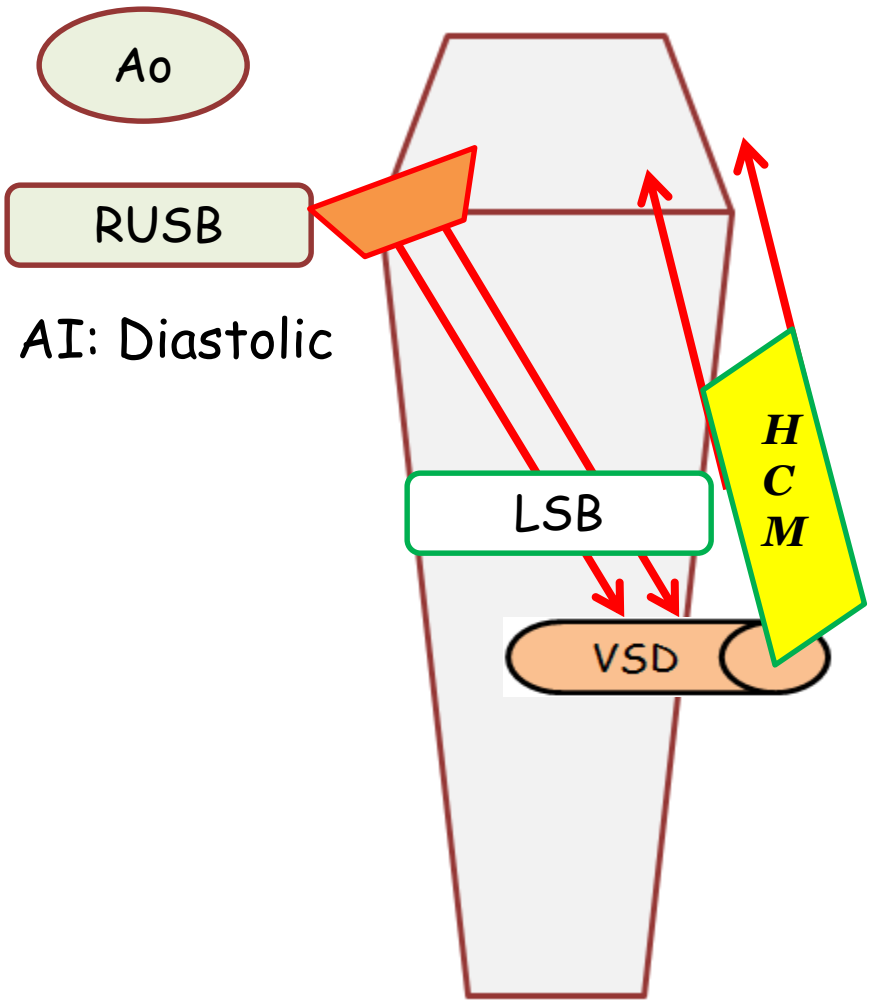
Physical Exam Findings at the LSB



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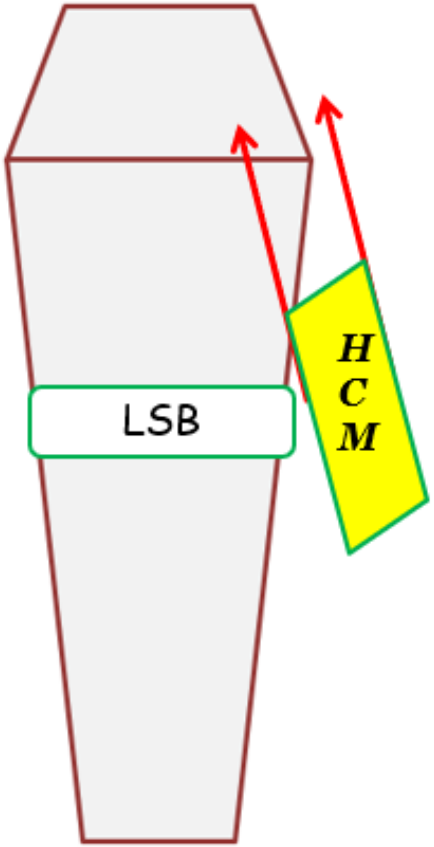

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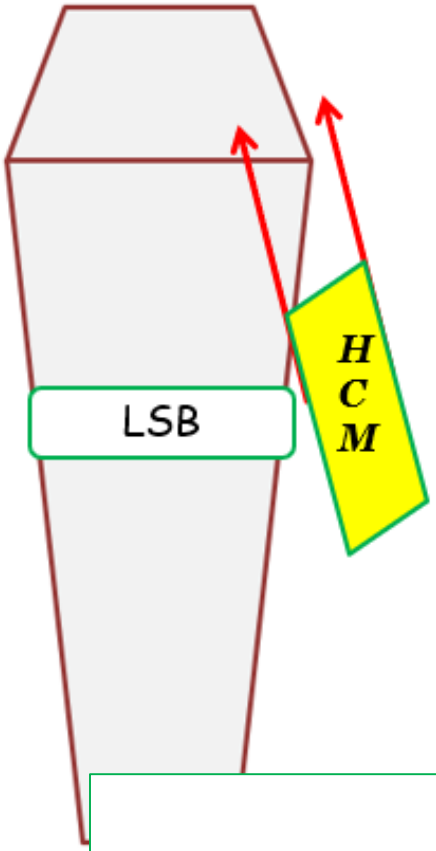
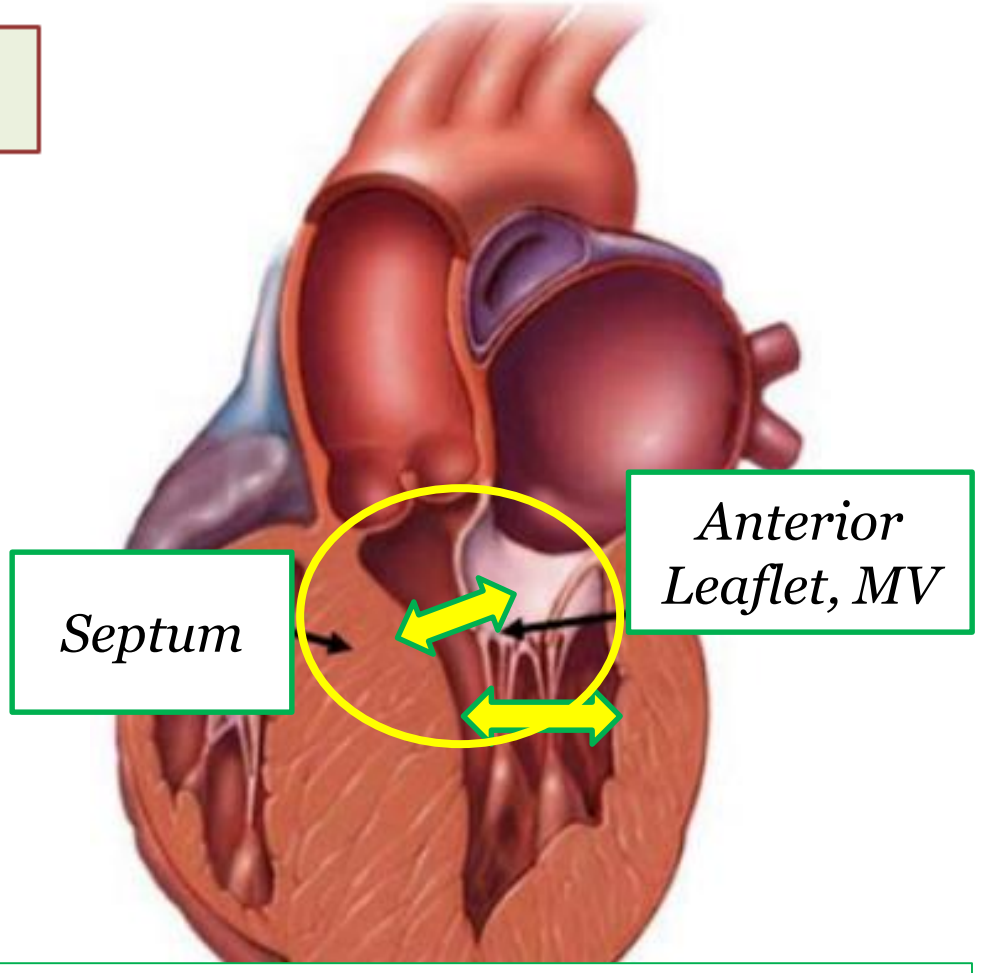
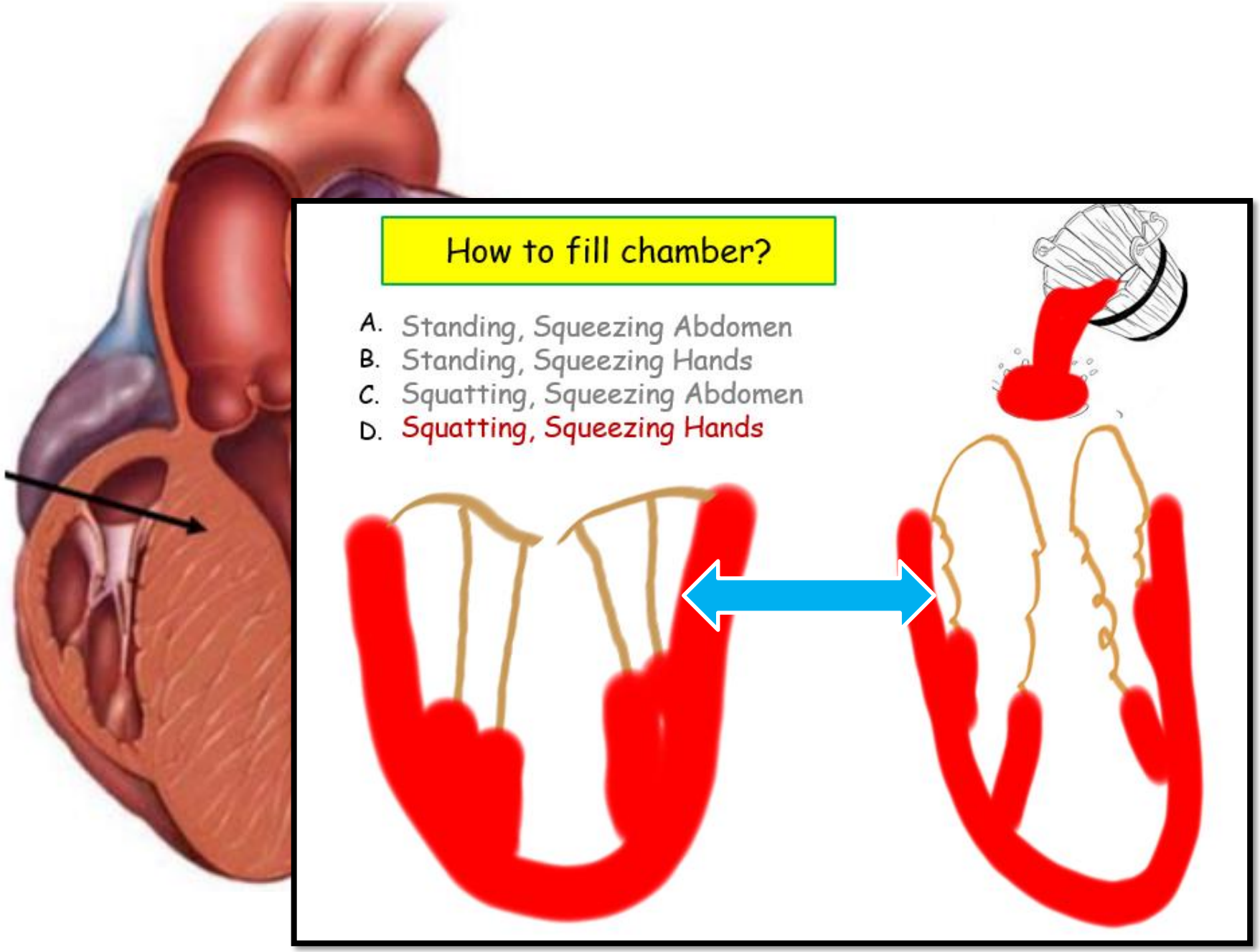




Maneuvers attempt to:

1. Quiet murmur through separation of walls (and MV), or
2. Accentuate murmur by bringing walls into close proximity

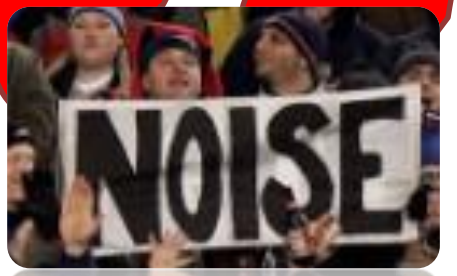

How to fill chamber?
A. Standing, Squeezing Abdomen
B. Standing, Squeezing Hands
C. Squatting, Squeezing Abdomen
D. Squatting, Squeezing Hands

Valsalva

Standing

'...the apposition of the hypertrophic septum to the free ventricular wall...'

Valsalva
Standing
(empty chamber)

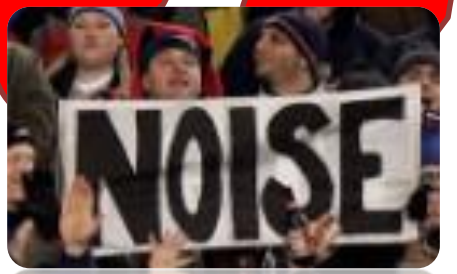


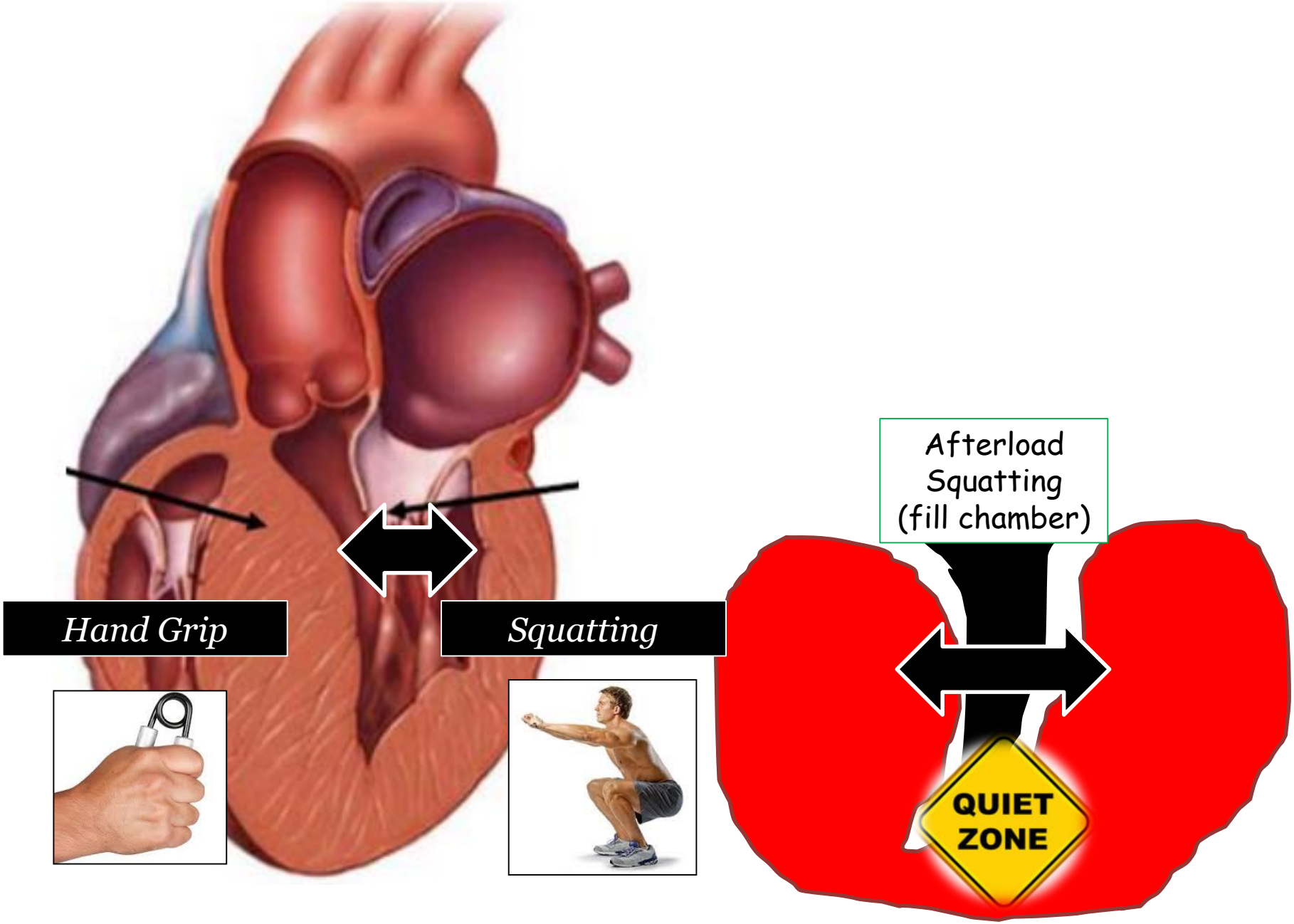
Valsalva

Standing

Decreased LV EDV → worsens outflow gradient

Valsalva
Standing
(empty chamber)

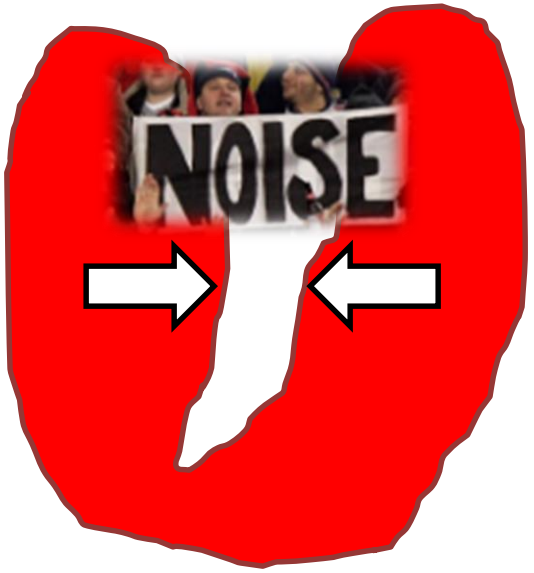



Hand Grip
Squatting
Afterload
Squatting
(fill chamber)
QUIET
ZONE

Valsalva

↓ EDV

Standing

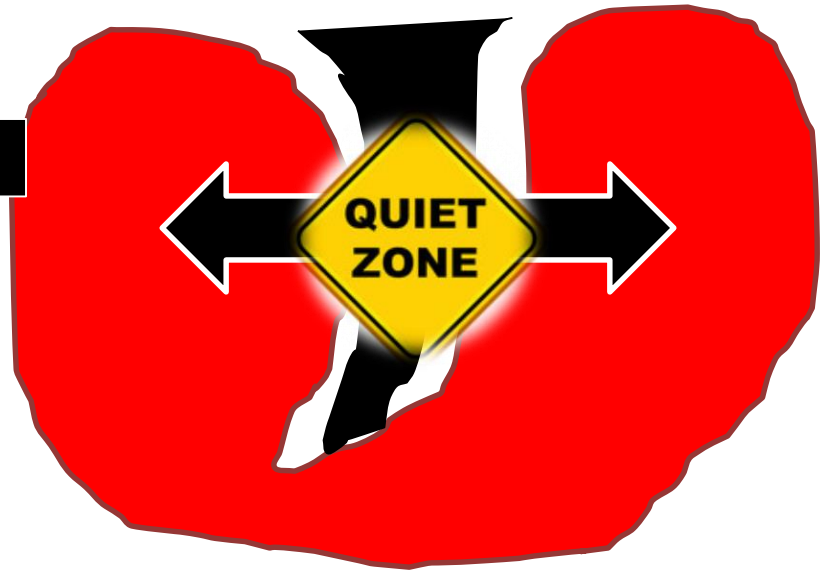


Outflow (Obstruction) Gradient

Hand Grip

↑ EDV

Squatting



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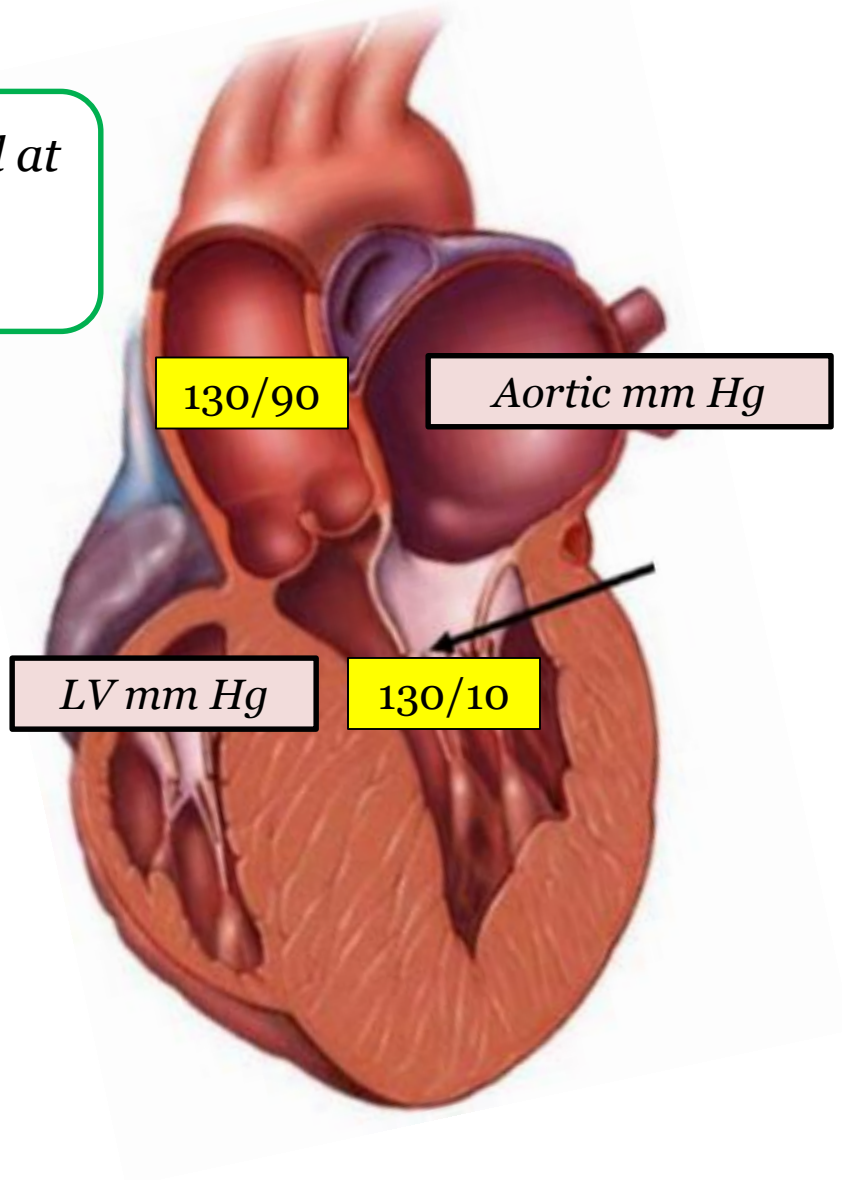
- Questions/Vignettes
  - Bring this bad boy home…



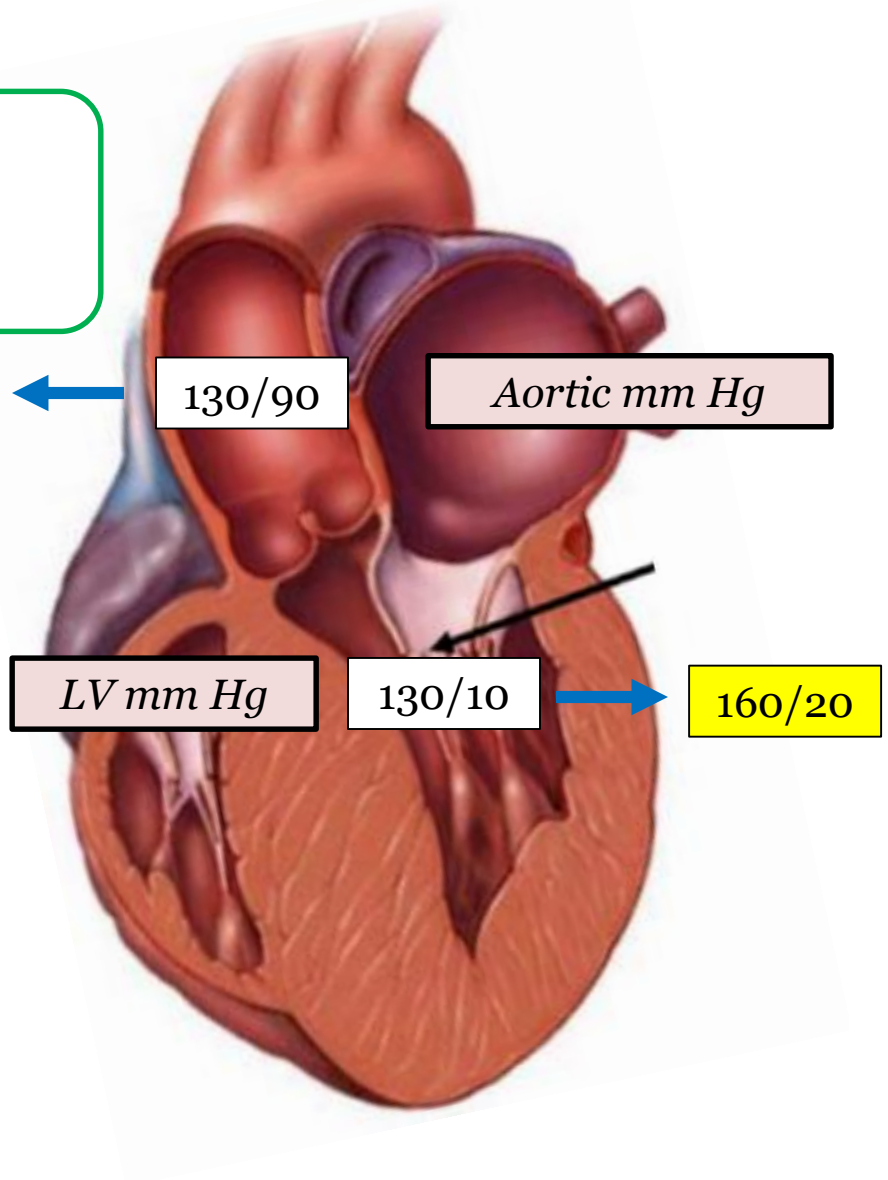
Understanding the *Outflow Gradient*: A Nice Question Assessing Hemodynamics

Patient with *systolic murmur located at the LSB* is taken to the cath lab.
Resting pressures recorded.



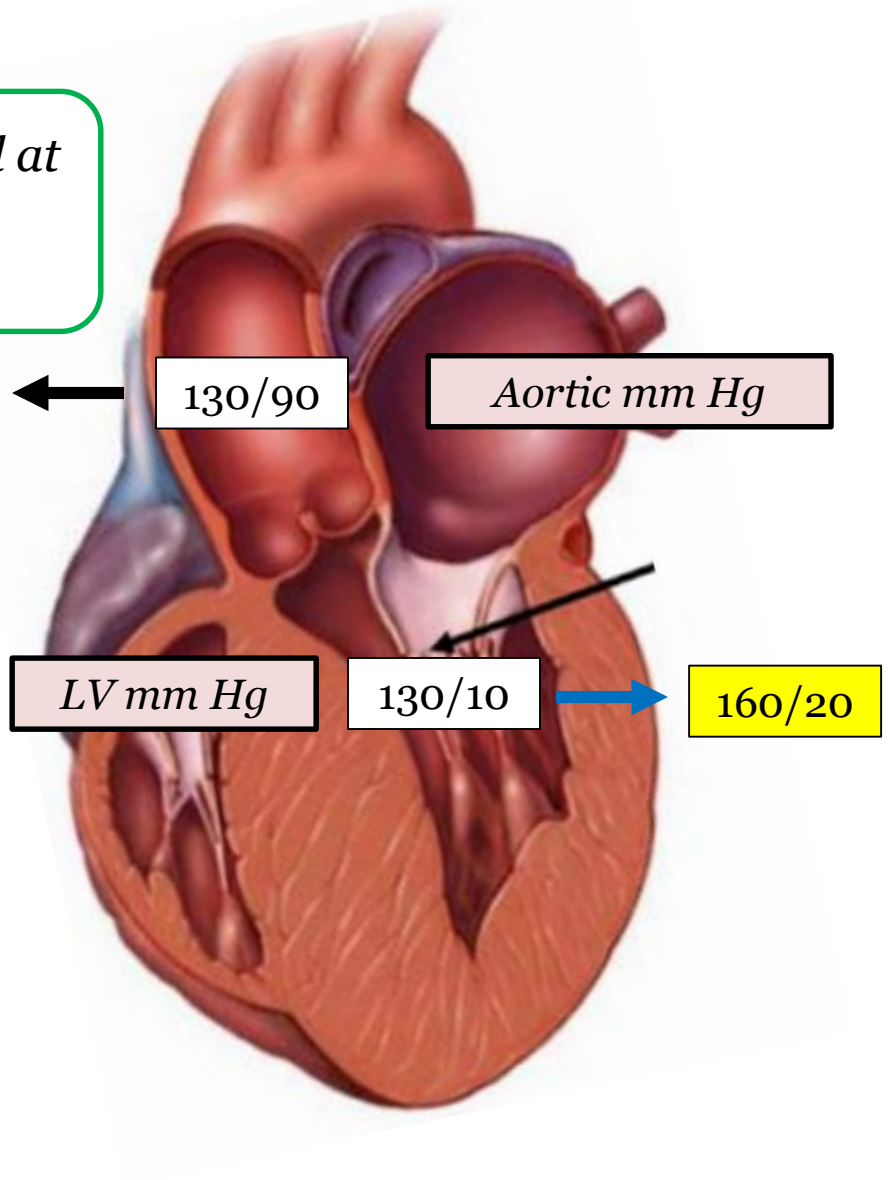
Outflow Gradient

A *normal* patient is now taken into the cath lab.
An *inotropic agent* is infused.
Pressures are recorded.



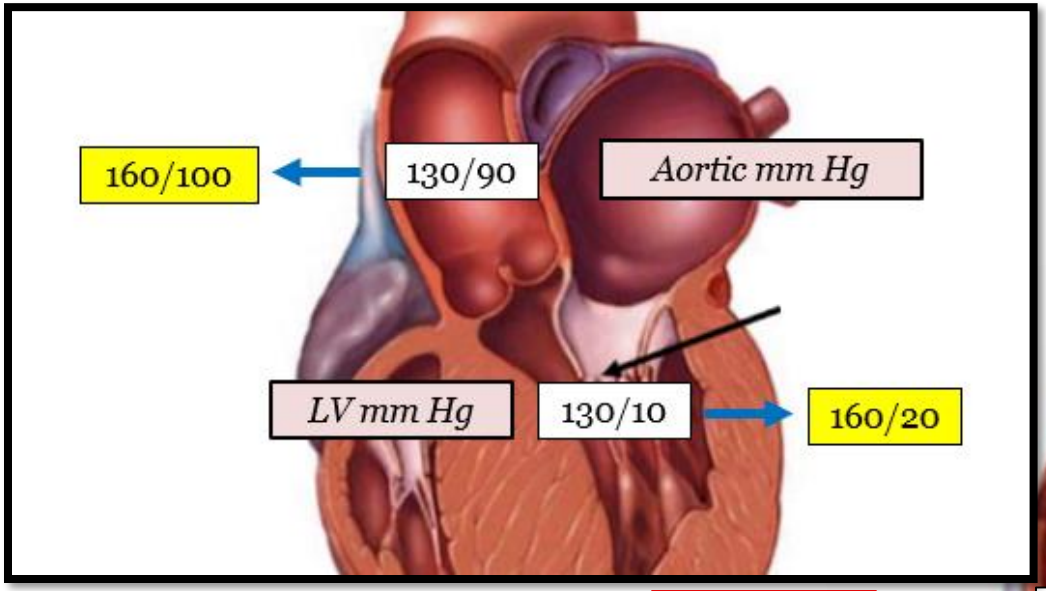
Outflow Gradient

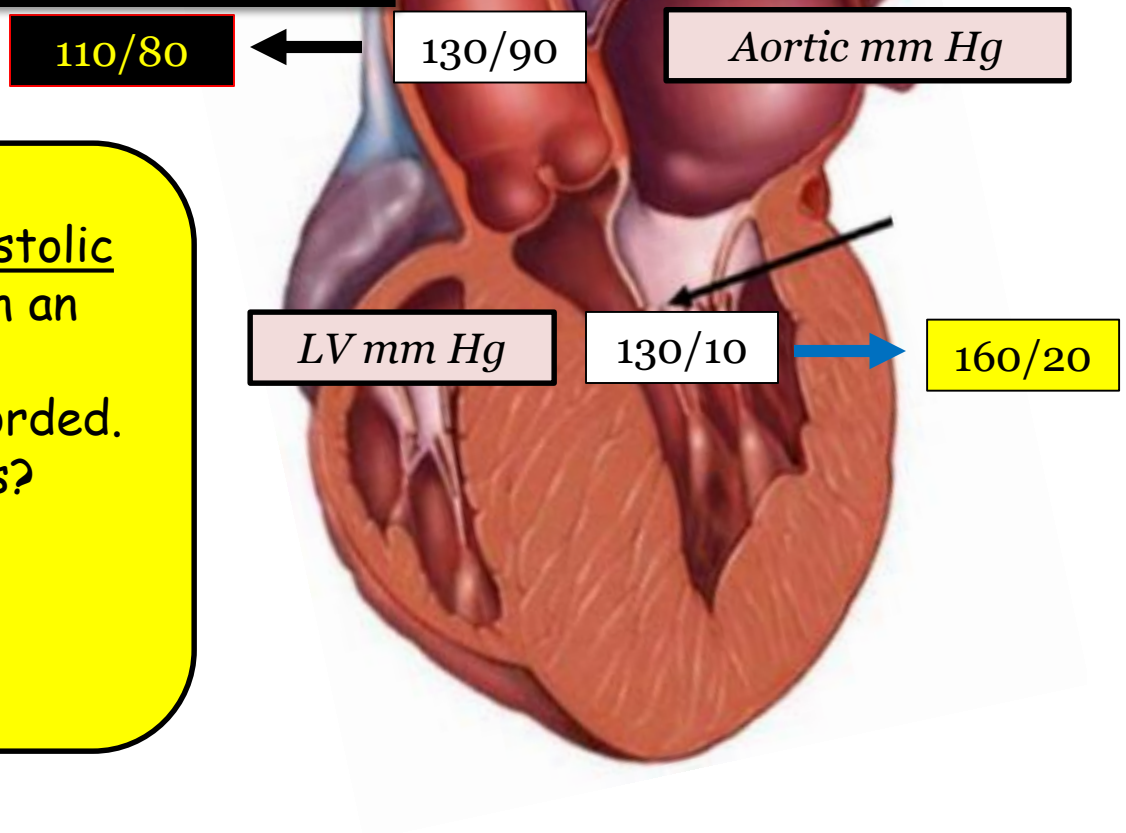
Patient with *systolic murmur located at the LSB* is taken to the cath lab.



Our patient <u>with the systolic murmur</u> is infused with an inotropic agent.
Pressures are again recorded.
Choose her diagnosis?

1. Aortic Stenosis
2. HCM





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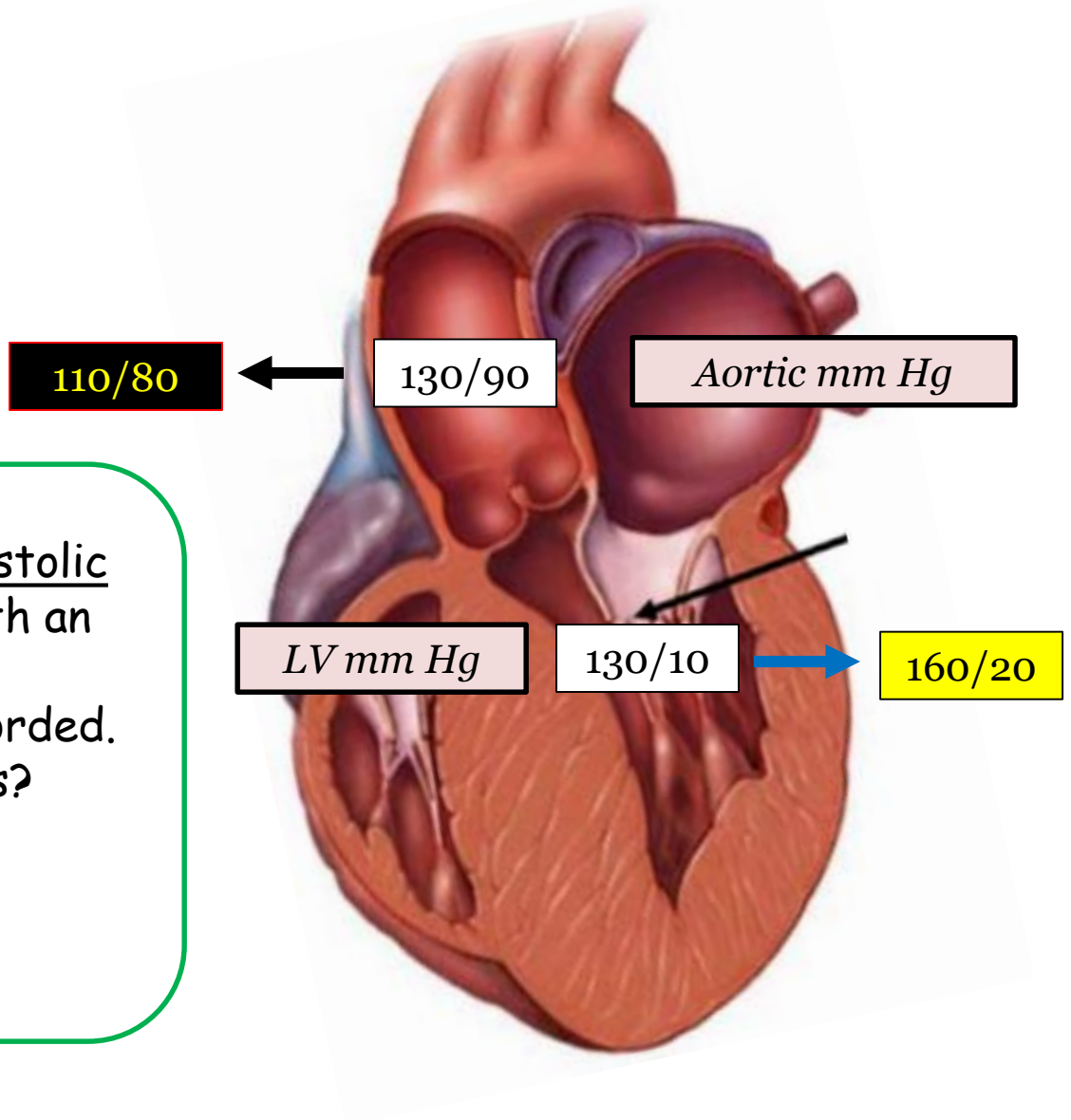
Outflow Gradient



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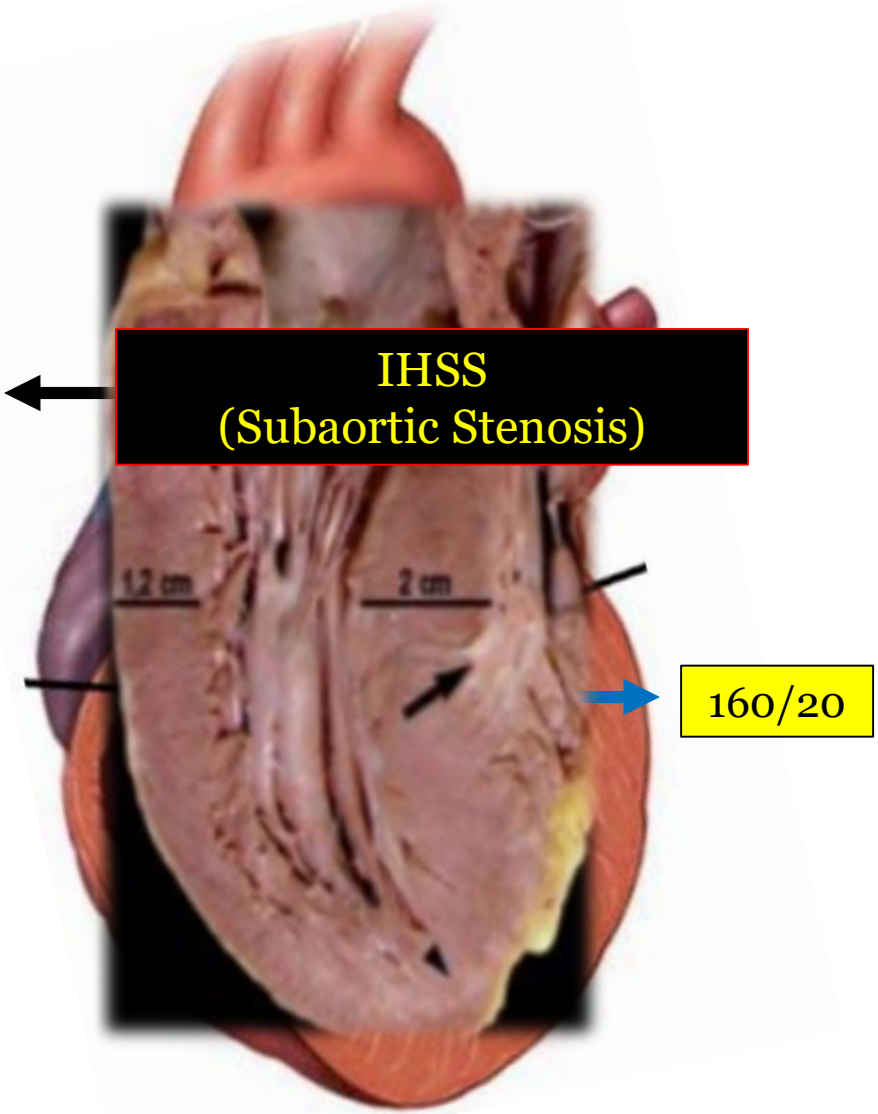

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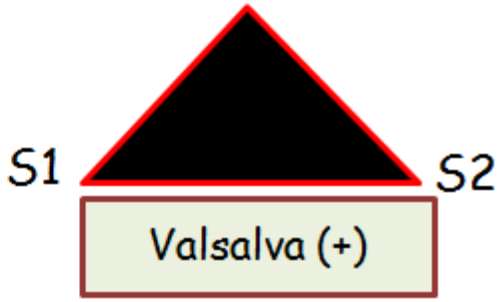
Understanding the *Outflow Gradient*: A Nice Question Assessing PE Maneuvers

You examine a patient and hear a systolic murmur depicted in the first image. You decide to test those maneuvers you've heard so much about. Lo and behold, after valsalva, the murmur sounded louder. Which of the following conditions is most likely present?



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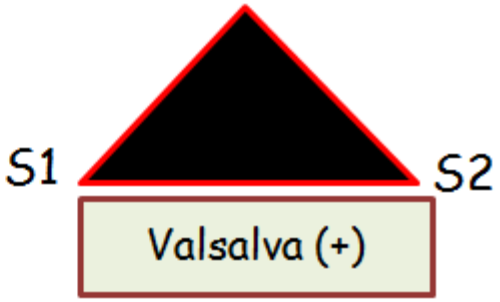

<u>To answer this question, you need to understand two principles</u>:

1. Venous return *decreases* with the Valsalva maneuver
2. The patient has a systolic murmur that *increased in intensity* with *decreased venous return (↓EDV)*

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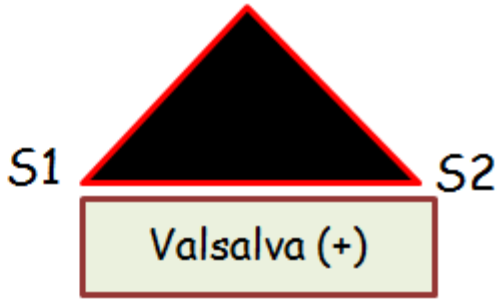
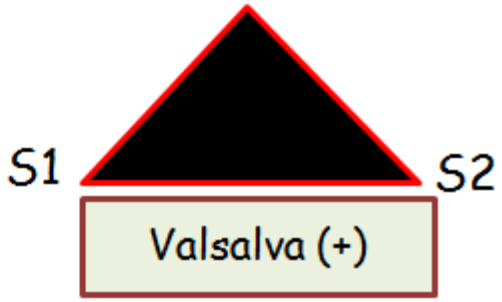
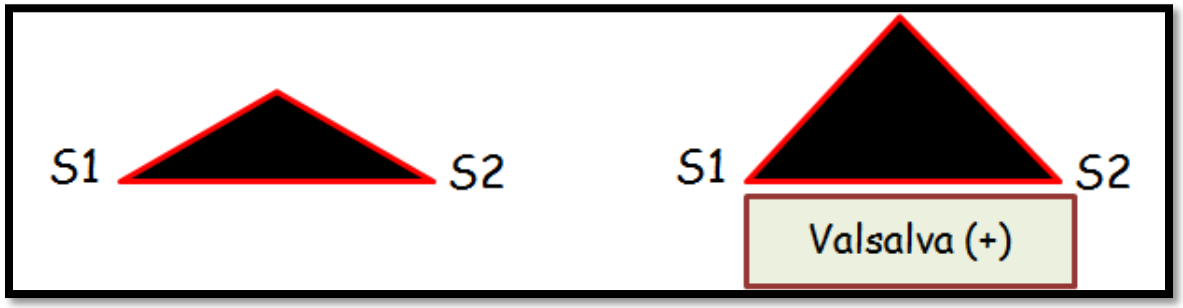


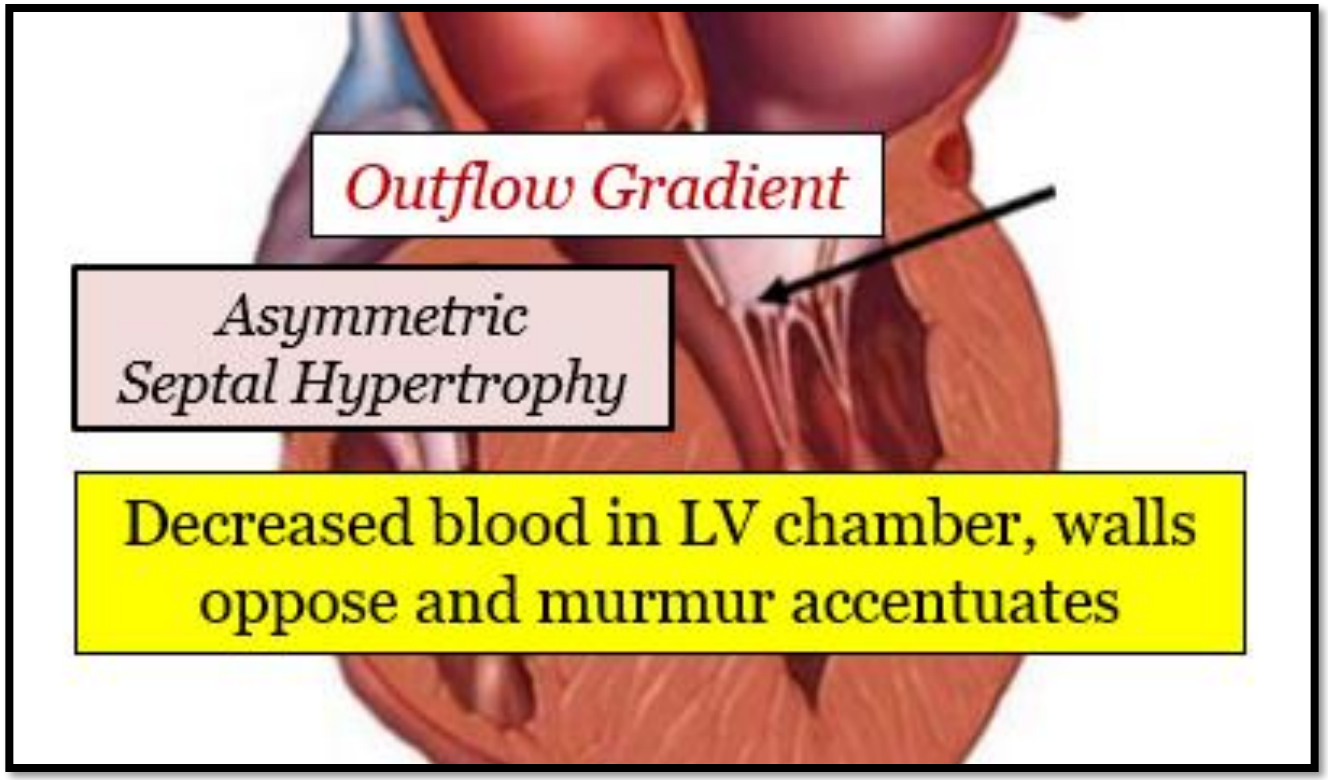
A. Mitral regurgitation
B. Aortic regurgitation
C. Asymmetric septal hypertrophy
D. Mitral valve prolapse
E. Aortic stenosis
F. Tricuspid regurgitation

You examine a patient and hear a systolic murmur depicted in the first image. You decide to test those maneuvers you've heard so much about. Lo and behold, after valsalva, the murmur sounded louder. Which of the following conditions is most likely present?



A. Mitral regurgitation
B. Aortic regurgitation
C. Asymmetric septal hypertrophy (aka HCM, IHSS)
D. Mitral valve prolapse (sound; mid-systolic click)
E. Aortic stenosis
F. Tricuspid regurgitation

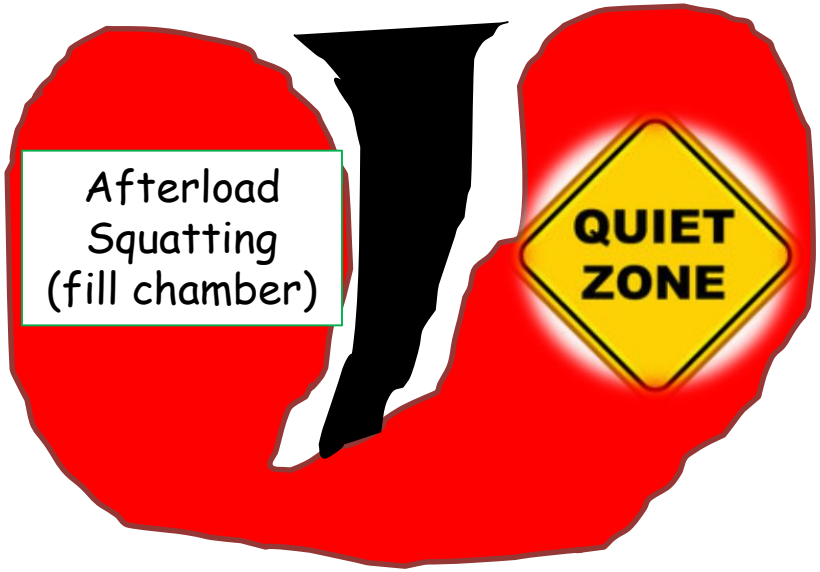

S1
S2
S1
S2
Valsalva (+)


Outflow Gradient
Asymmetric
Septal Hypertrophy
Decreased blood in LV chamber, walls
oppose and murmur accentuates

Match the Medication

Which combination of agents would be most likely to exacerbate the obstructive physiology?

A. Furosemide, Atenolol
B. Isosorbide mononitrate, Verapamil
C. Atenolol, Verapamil
D. Isosorbide mononitrate, Furosemide
E. Isosorbide mononitrate, Atenolol
F. Furosemide, Verapamil



Match the Medication

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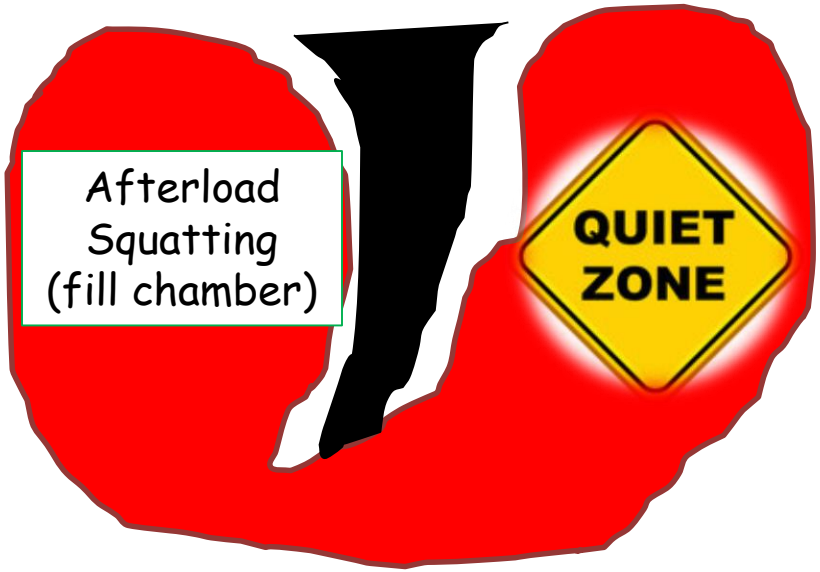
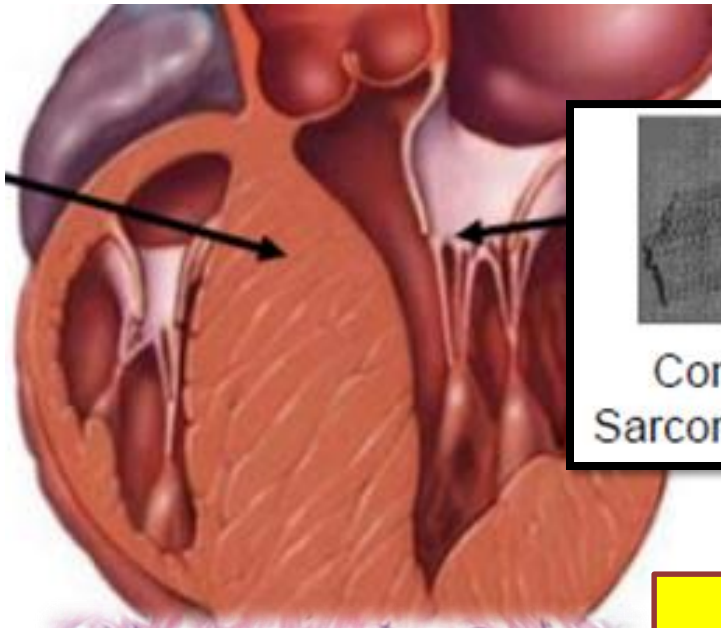
C. Atenolol, Verapamil
D. Isosorbide mononitrate, Furosemide
E. Isosorbide mononitrate, Atenolol
F. Furosemide, Verapamil

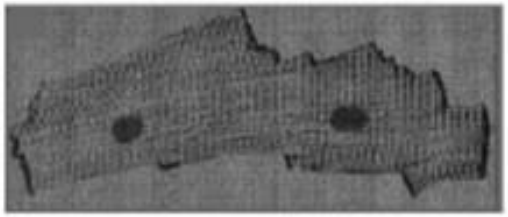
Worsen the Gradient:
Decreased Preload (nitrate)
Decreased LVEDV (diuretic)

Agents with negative chronotropic (and inotropic) properties, promote LV filling (and decreased outflow gradient)

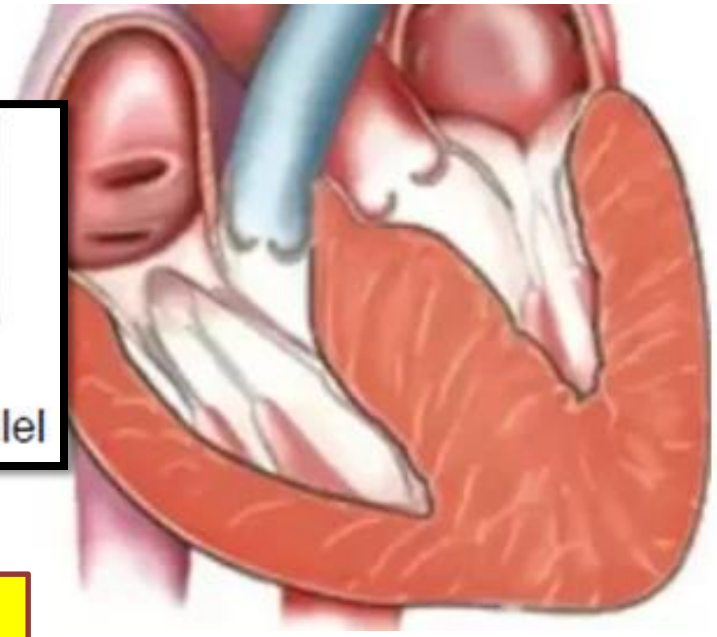
A. Furosemide, Atenolol
B. Isosorbide mononitrate, Verapamil
C. Atenolol, Verapamil
D. Isosorbide mononitrate, Furosemide
E. Isosorbide mononitrate, Atenolol
F. Furosemide, Verapamil



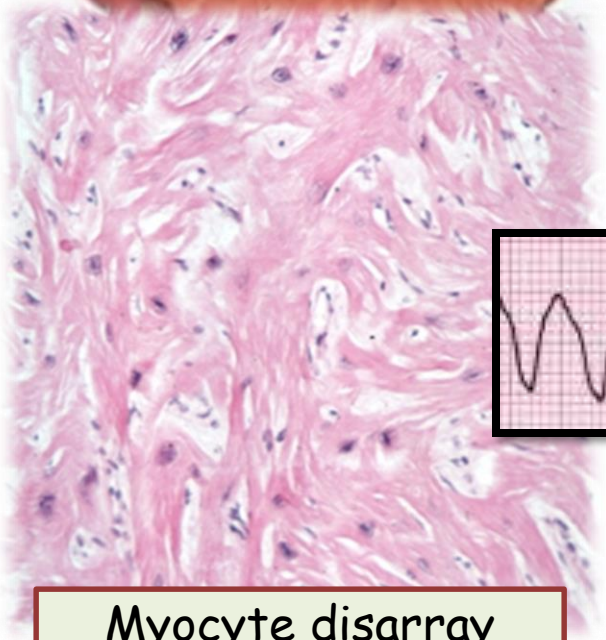




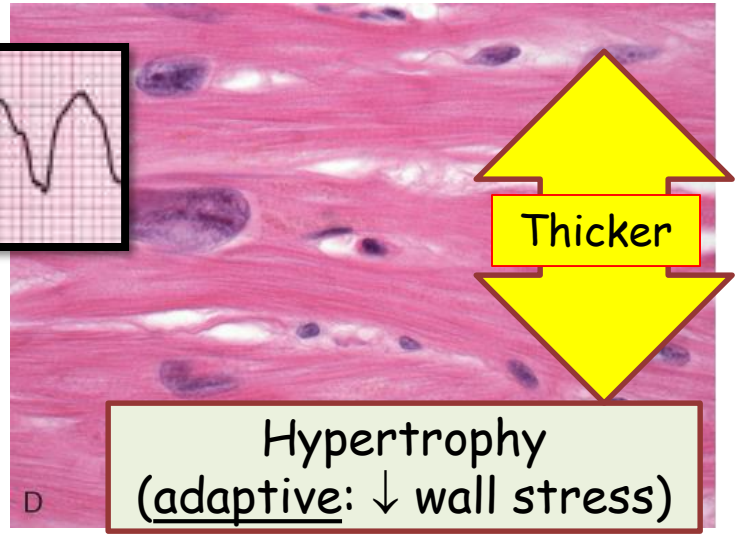
Concentric Hypertrophy
Sarcomeres added in Parallel



Pop Quiz:
Match the tracing.
Match the image.

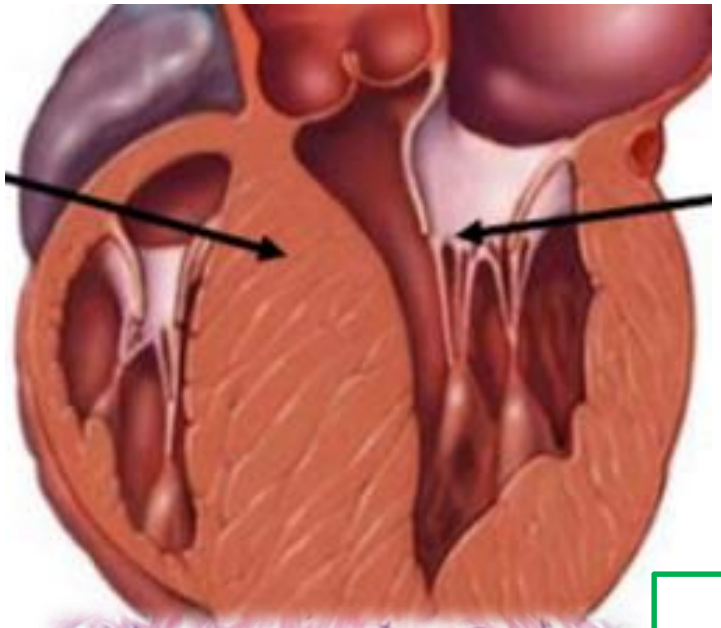


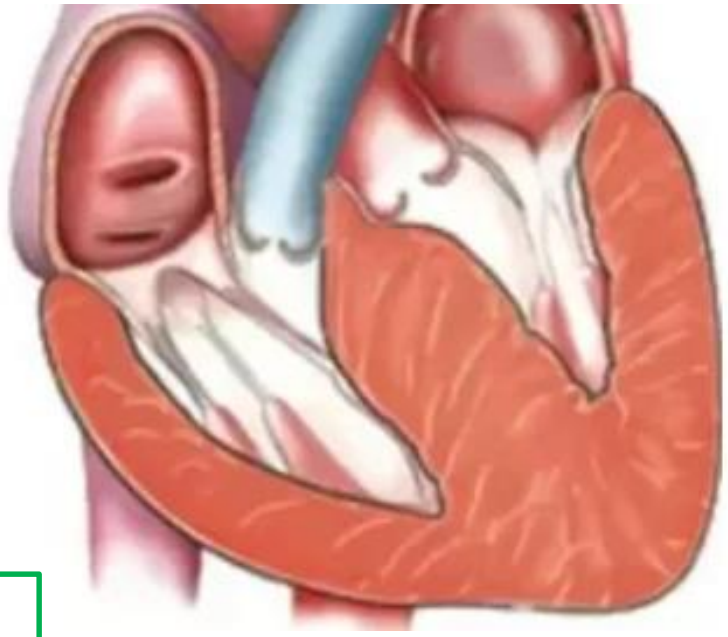
Myocyte disarray
(genetic mutation)



Hypertrophy
(adaptive: ↓ wall stress)

D





Pop Quiz:
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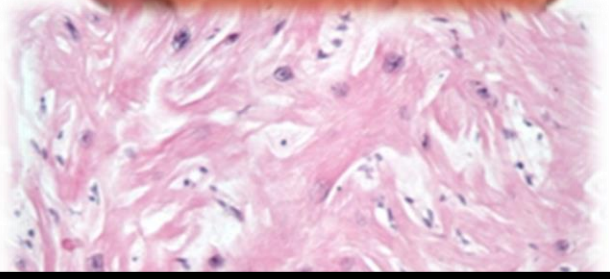
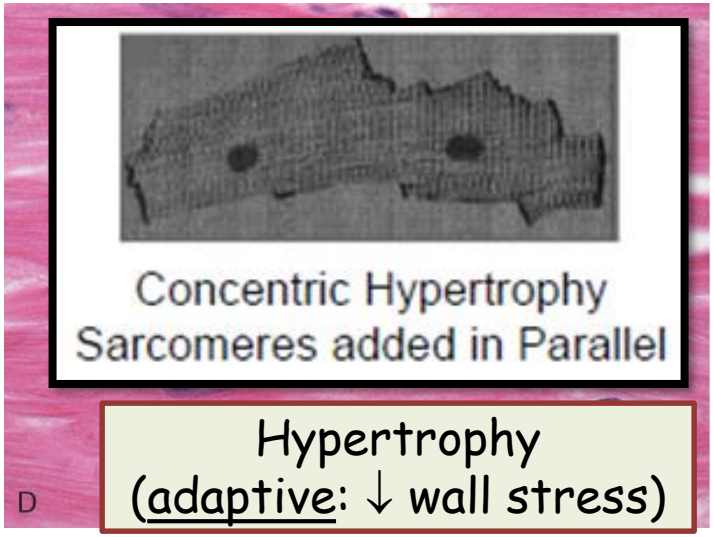


Myocardial Fibrosis

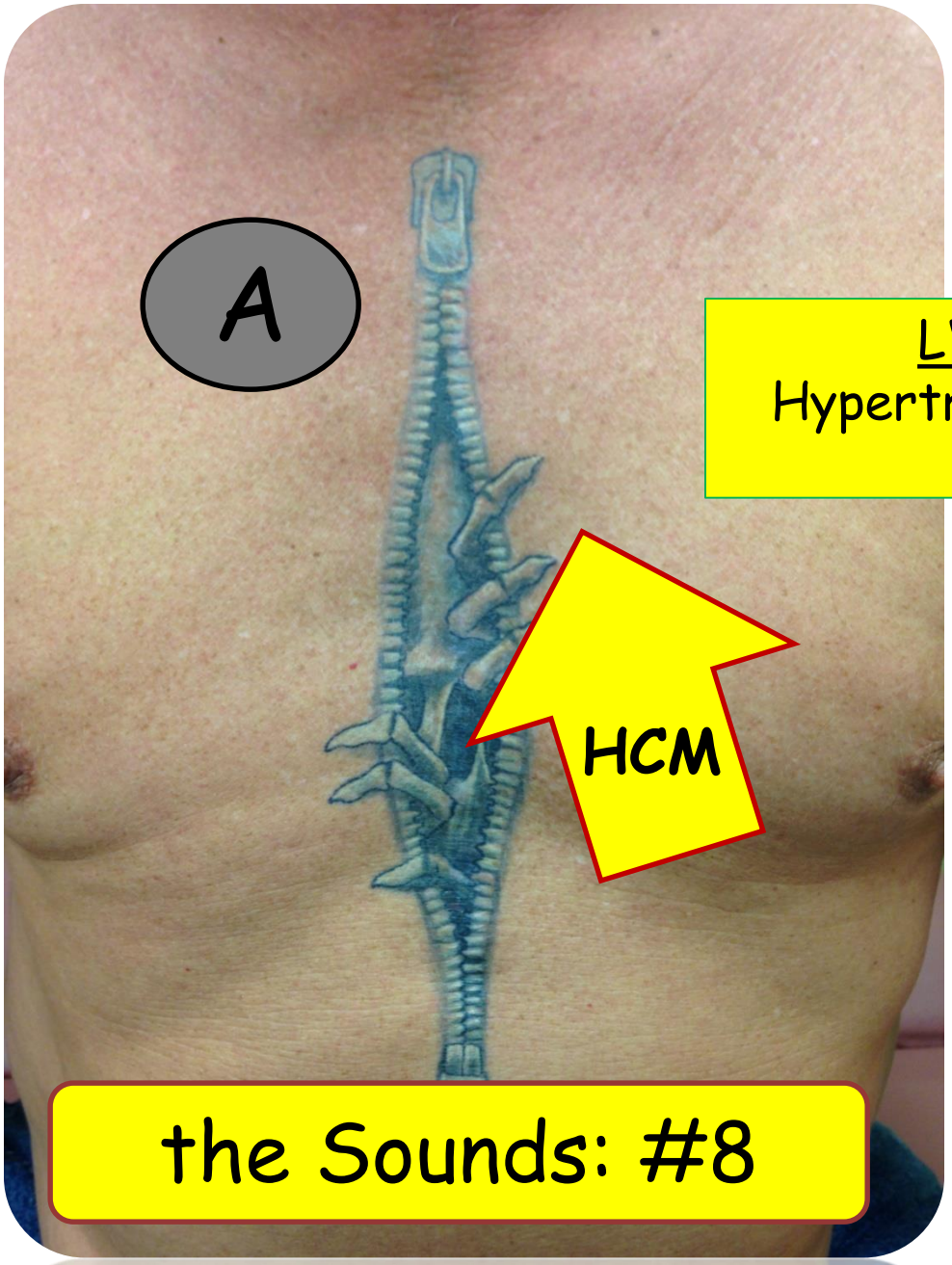
Myocyte disarray
(genetic mutation)



Hypertrophy
(adaptive: ↓ wall stress)

HCM *(Hypertrophic Cardiomyopathy)*

- Background
  - Obstructive physiology → *Dynamic Outflow Gradient*
  - Pathology description: *Myocyte Disarray*
    - Key Derivative: *Syncope* or *SCD* (especially in the young athlete)
- Definition/Nomenclature/Pathology
  - HCM is an inherited disorder
    - Sarcomere or Nonsarcomere Metabolic Dysfunction
  - HCM is *not* LVH (adaptation to disorders of afterload)
- Physical Exam
  - Maneuvers → *Outflow Gradient*
  - Applied physiology/pharmacology
- Questions/Vignettes
  - Bring this bad boy home…



Cardiology

LV Outflow Obstruction:
Hypertrophic Cardiomyopathy (HCM)
(s/p Aortic Stenosis)

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