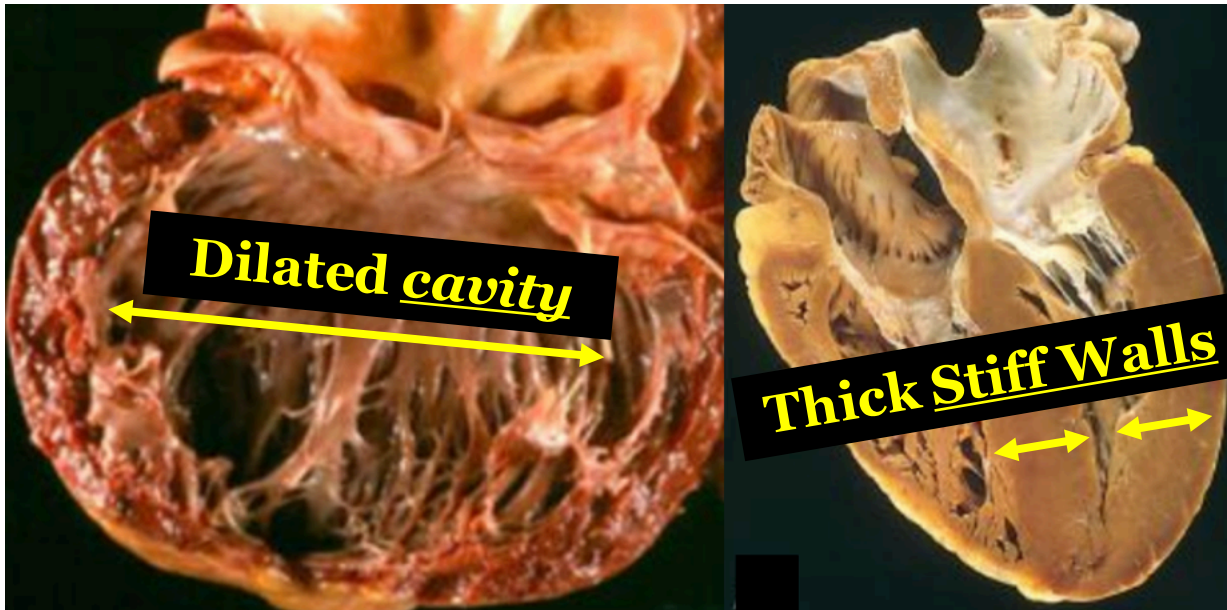


Heart Failure and the Cardiomyopathies for the USMLE Step One Exam:  
*Part III: Dilated Cardiomyopathy and Test Derivatives*

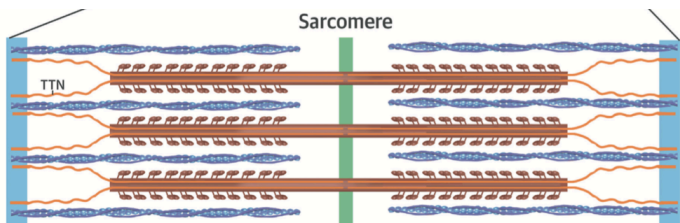


HF<sup>r</sup>EF

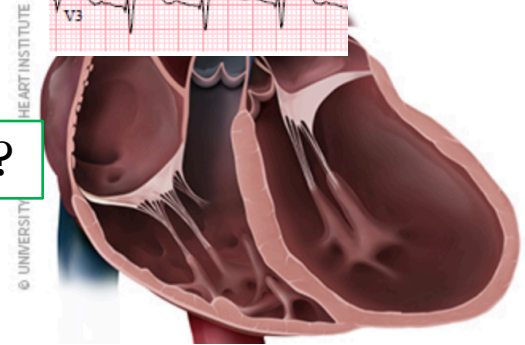
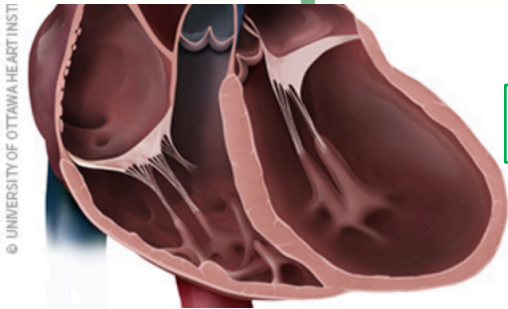
HF<sup>p</sup>EF

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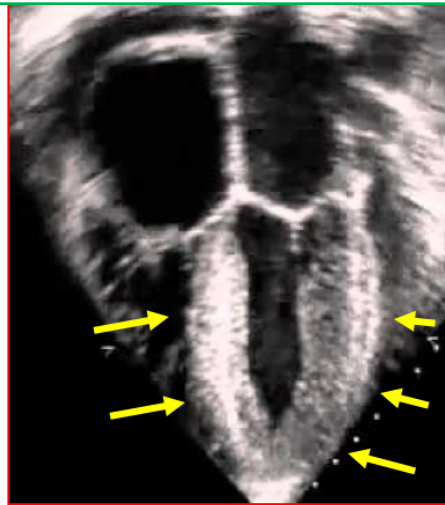
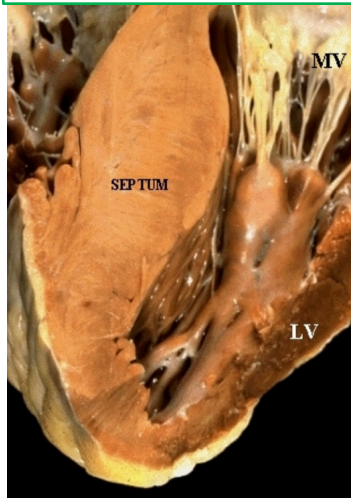
# The Cardiomyopathies?: *Clarification*



Dilated CM v Ischemic CM?



Hypertrophic CM v Hypertensive HD?



## The Cardiomyopathies (CM): *Clarification*

*Don't memorize...*this is a 'clarification'

### Definition:

"A myocardial disorder in which the heart muscle is *structurally* and *functionally* abnormal in the absence of *coronary artery disease, hypertension, valvular disease and congenital heart disease sufficient to explain the observed myocardial abnormality*"

## The Cardiomyopathies (CM): *Background Info*

### Definition:

"A myocardial disorder in which the heart muscle is *structurally* and *functionally* abnormal in the absence of *coronary artery disease, hypertension, valvular disease and congenital heart disease sufficient to explain the observed myocardial abnormality*"

### 'Clarification':

'...categorized as a distinct set of conditions with a functional and structural phenotype AND **often** with a **genetic component**. *In discussions of CM, ischemia, HTN and valvular causes are excluded although they do share hemodynamic and pathophysiologic parameters.*'

- Dilated
- Hypertrophic (100%)
- Restrictive...not so much



## The Cardiomyopathies (CM): *Background Info*

### Definition:

"A myocardial disorder in which the heart muscle is *structurally* and *functionally* abnormal in the absence of *coronary artery disease, hypertension, valvular disease and congenital heart disease sufficient to explain the observed myocardial abnormality*"

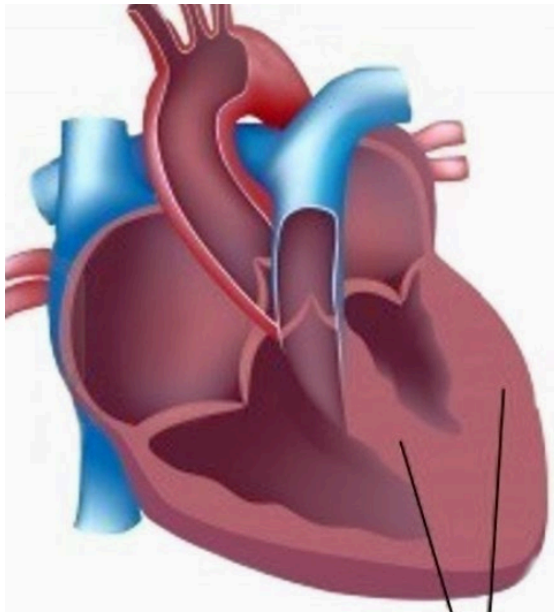
### 'Clarification':

'...categorized as a distinct set of conditions with a functional and structural phenotype AND **often** with a **genetic component**. *In discussions of CM, ischemia, HTN and valvular causes are excluded although they do share hemodynamic and pathophysiologic parameters.*'

### Dilated (Non-Genetic) Examples:

- Thiamine deficiency
- Endocrinopathies
- Infectious (Chagas)

## *The Cardiomyopathies (CM)*

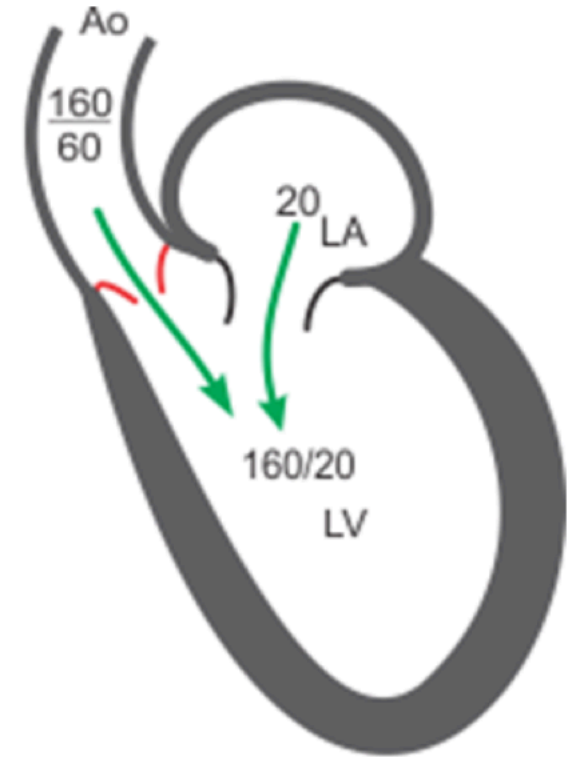


LVH (HTN)

*Concentric Hypertrophy*



Aortic Stenosis

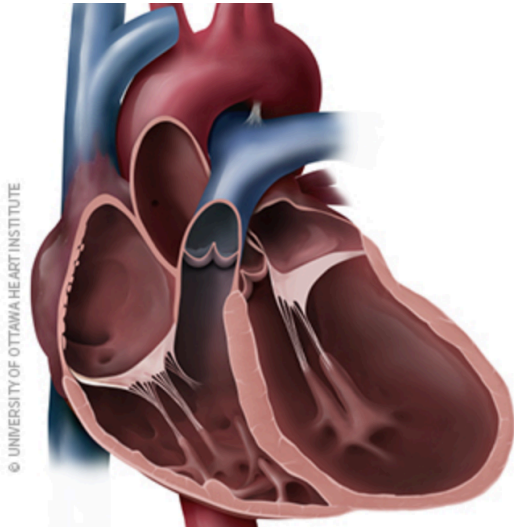


Aortic Regurgitation

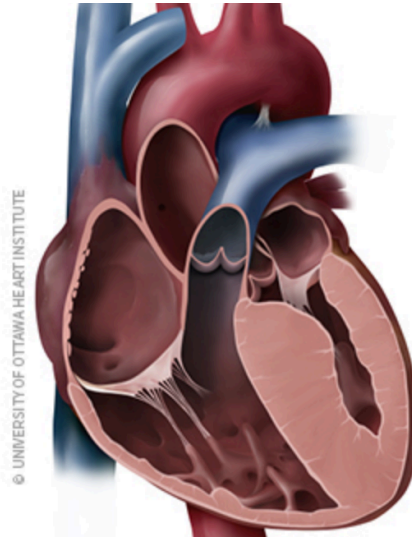
*Eccentric Hypertrophy*

Guilty with Explanation (*i.e. not genetic*)

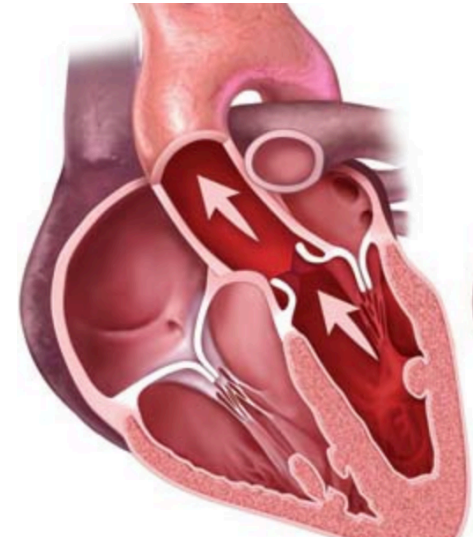
# The Cardiomyopathies (CM)



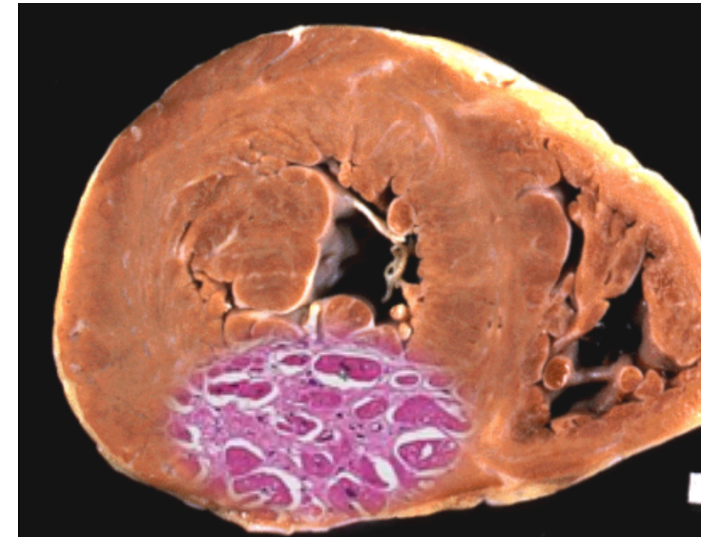
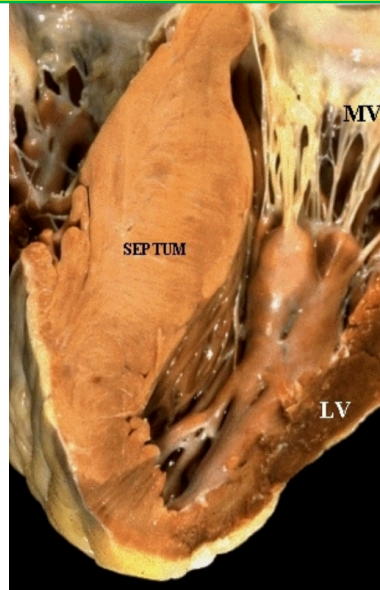
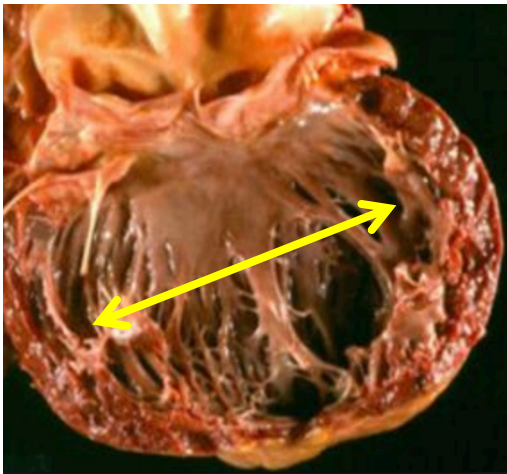
Dilated



Hypertrophic

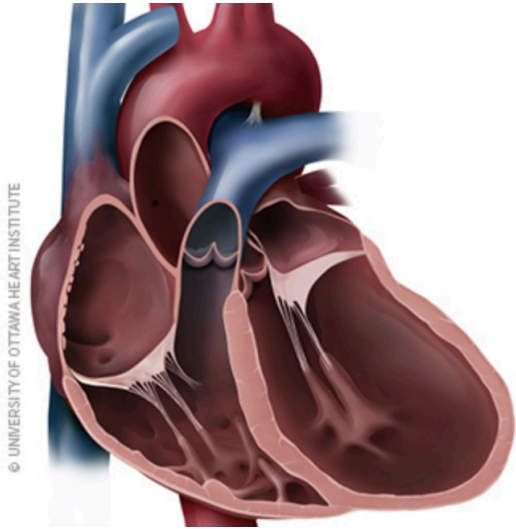


Restrictive

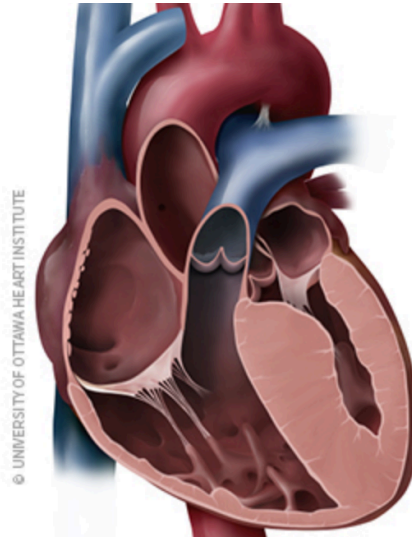




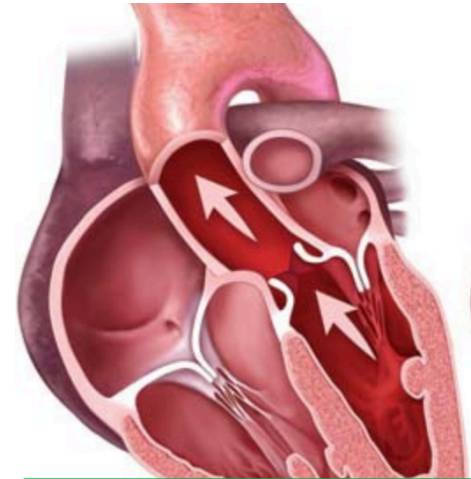
# The Cardiomyopathies (CM)



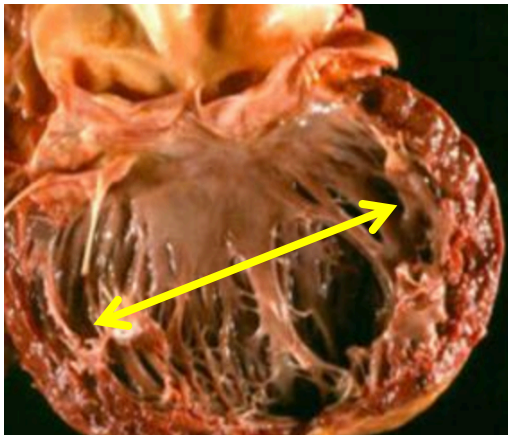
Dilated



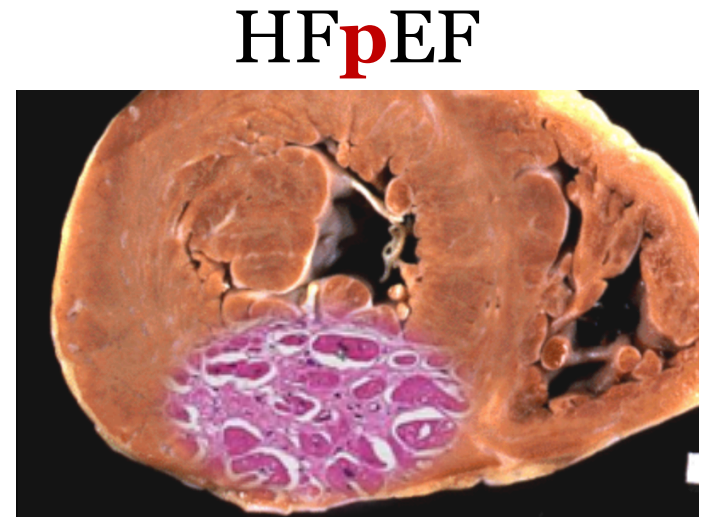
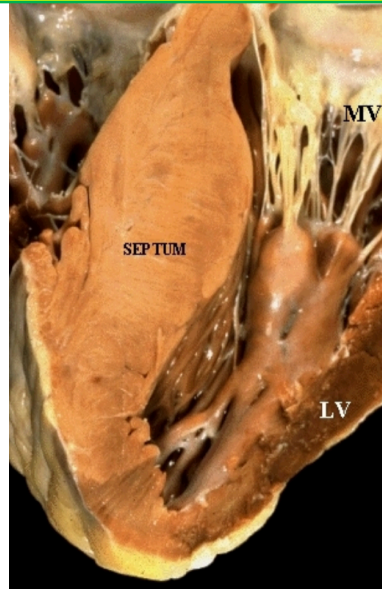
Hypertrophic



Restrictive

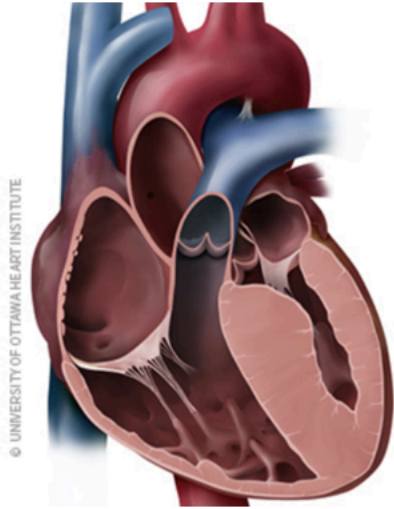


HF<sub>r</sub>EF

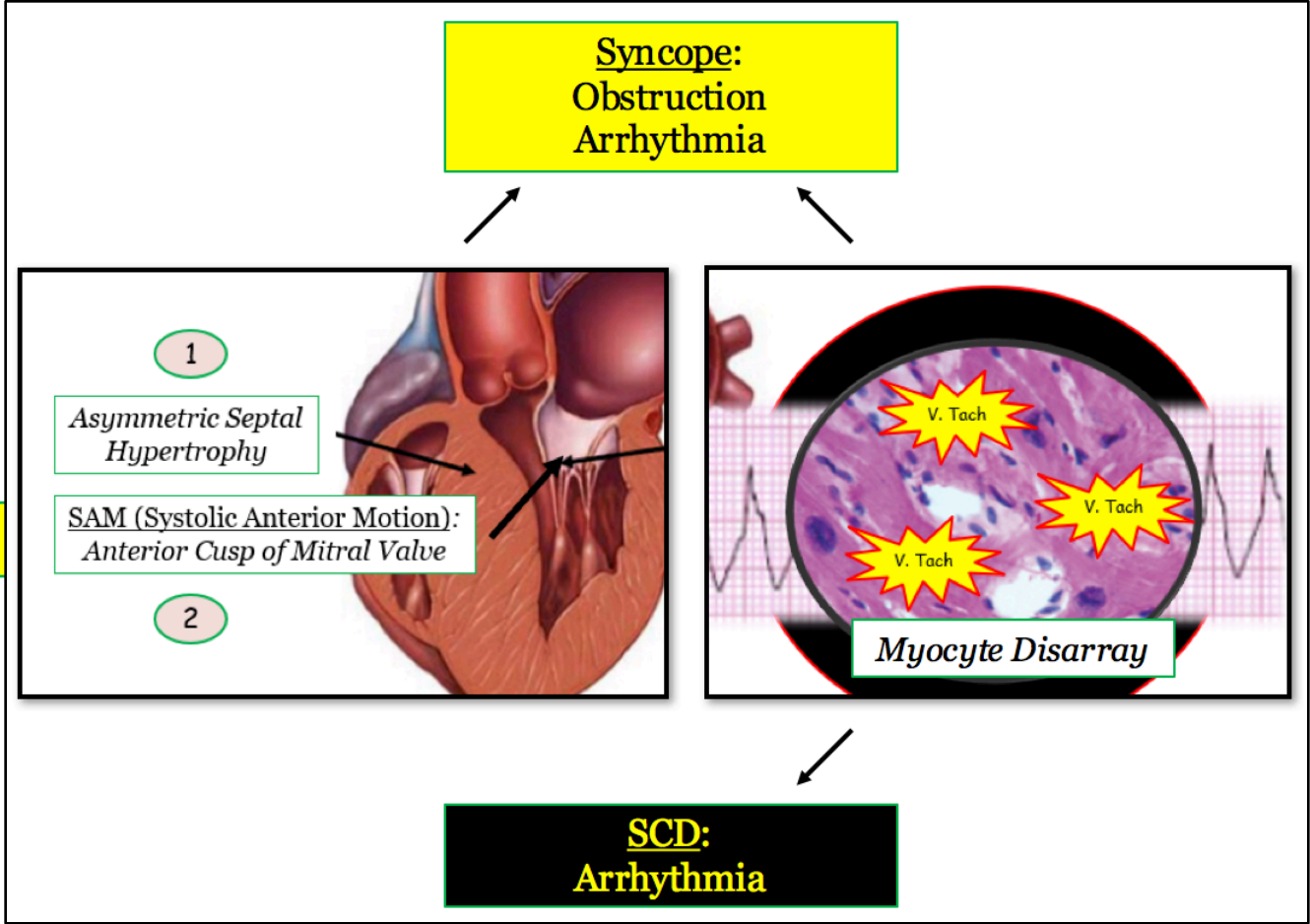
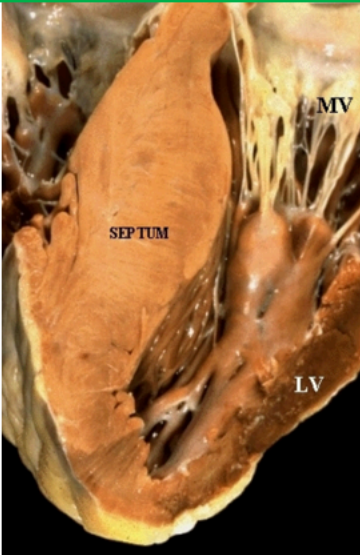


HF<sub>p</sub>EF

# The Cardiomyopathies (CM)



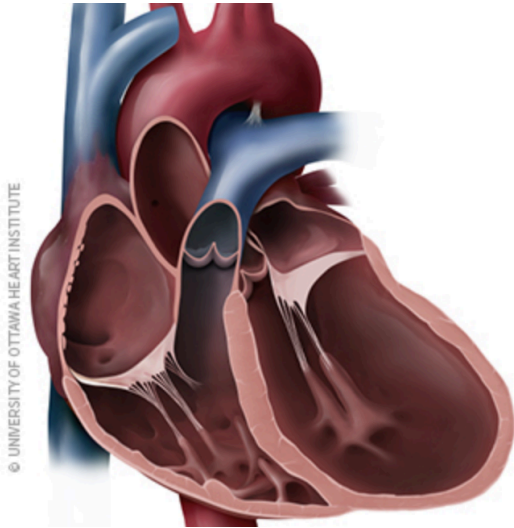
**Hypertrophic**



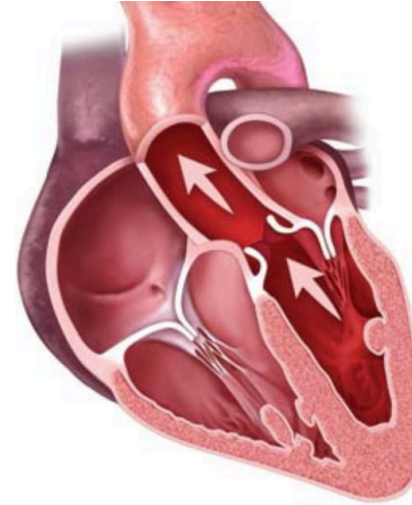
*Heart Sounds #8*



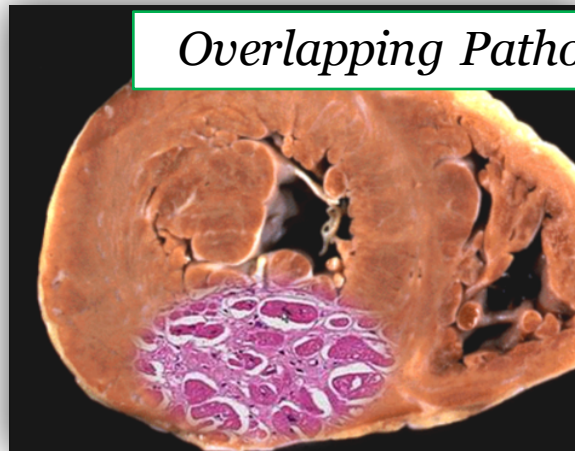
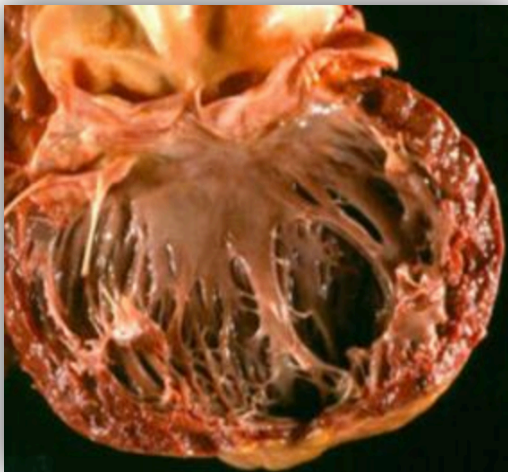
# The Cardiomyopathies (CM)



Dilated



Restrictive = Diastolic Heart Failure

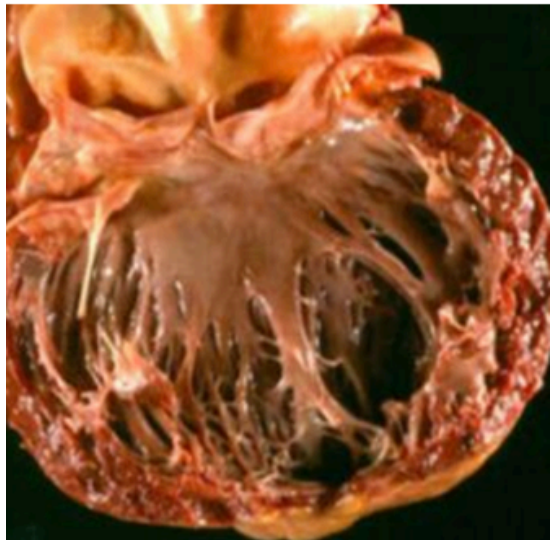
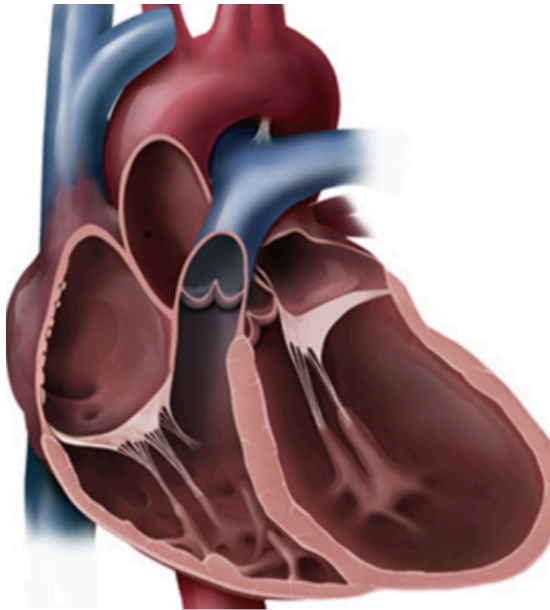


*Overlapping Pathophysiologic Principles*





# Dilated Cardiomyopathy



## Genetic

**Familial** (>30 mutations)

## Acquired (Secondary)

**Drugs:** doxorubicin, cocaine, Etoh

**Metabolic:** Thiamine, ↑ Iron,  
↑ Catecholamines

**Infectious:** Viral (coxsackie), Chagas  
(trypanasoma cruzi)

**Peripartum**

## 'Guilty with Explanation'

- Ischemic/Cardiogenic Shock
- Valvular (AI)

# Dilated Cardiomyopathy

## How They Play The Game:

*Common Language:* Patient with CHF (**S3**, rales)...



## Pathophysiology:

Pressure-volume loop...

Pressure-volume curve...



- Which condition does the poor dude have...???
- What caused this condition...???



## Genetic

***Familial*** (>30 mutations)

## Acquired (Secondary)

**Drugs:** doxorubicin, cocaine, Etoh

**Metabolic:** Thiamine, ↑ Iron,  
↑ Catecholamines

**Infectious:** Viral (coxsackie), Chagas  
(trypanasoma cruzi)

**Peripartum**

## Dilated Cardiomyopathy

*Reduviid* → Chagas



**Genetic**

***Familial*** (>30 mutations)

**Acquired** (*Secondary*)

**Drugs:** doxorubicin, cocaine, Etoh

**Metabolic:** Thiamine, ↑ Iron,  
↑ Catecholamines

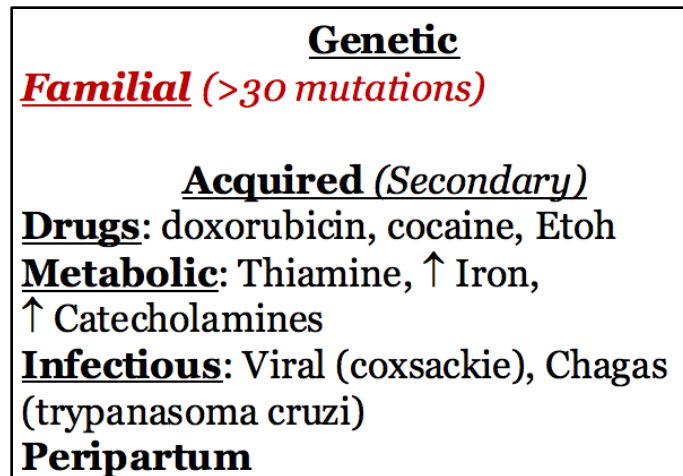
**Infectious:** Viral (coxsackie), Chagas  
(trypanasoma cruzi)

**Peripartum**

# Dilated Cardiomyopathy

How They Play The Game:

*Common Language:* Patient with CHF (**S3**, rales)...



Choose the correct graph/curve...



**Pathophysiology:**  
Pressure-volume loop...  
Pressure-volume curve...

# DCM: *Pathophysiology* Derivatives

## **Genetic**

***Familial*** (>30 mutations)

## **Acquired (Secondary)**

**Drugs**: doxorubicin, cocaine, Etoh

**Metabolic**: Thiamine, ↑ Iron,  
↑ Catecholamines

**Infectious**: Viral (coxsackie), Chagas  
(trypanasoma cruzi)

**Peripartum**

## **'Guilty with Explanation'**

- Ischemic
- Valvular (AI)

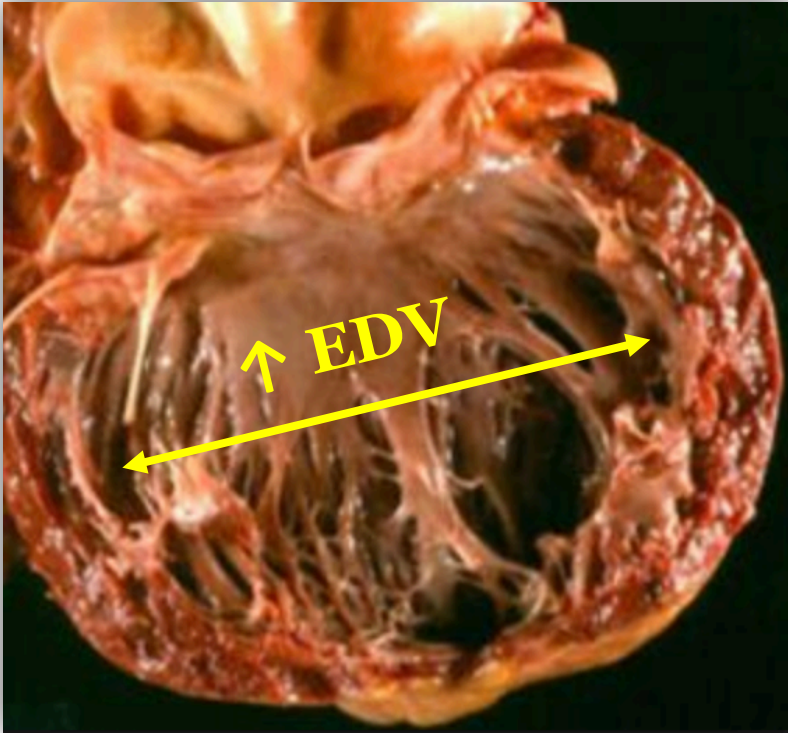
## **NBME Language:**

Cavity size?

Contractility?

Neurohumoral response?

## DCM (*all causes*): Pathophysiology Derivatives



Cavity size:  
↑ EDV (*preload*)

Contractility:  
↓ SV, EF, CO

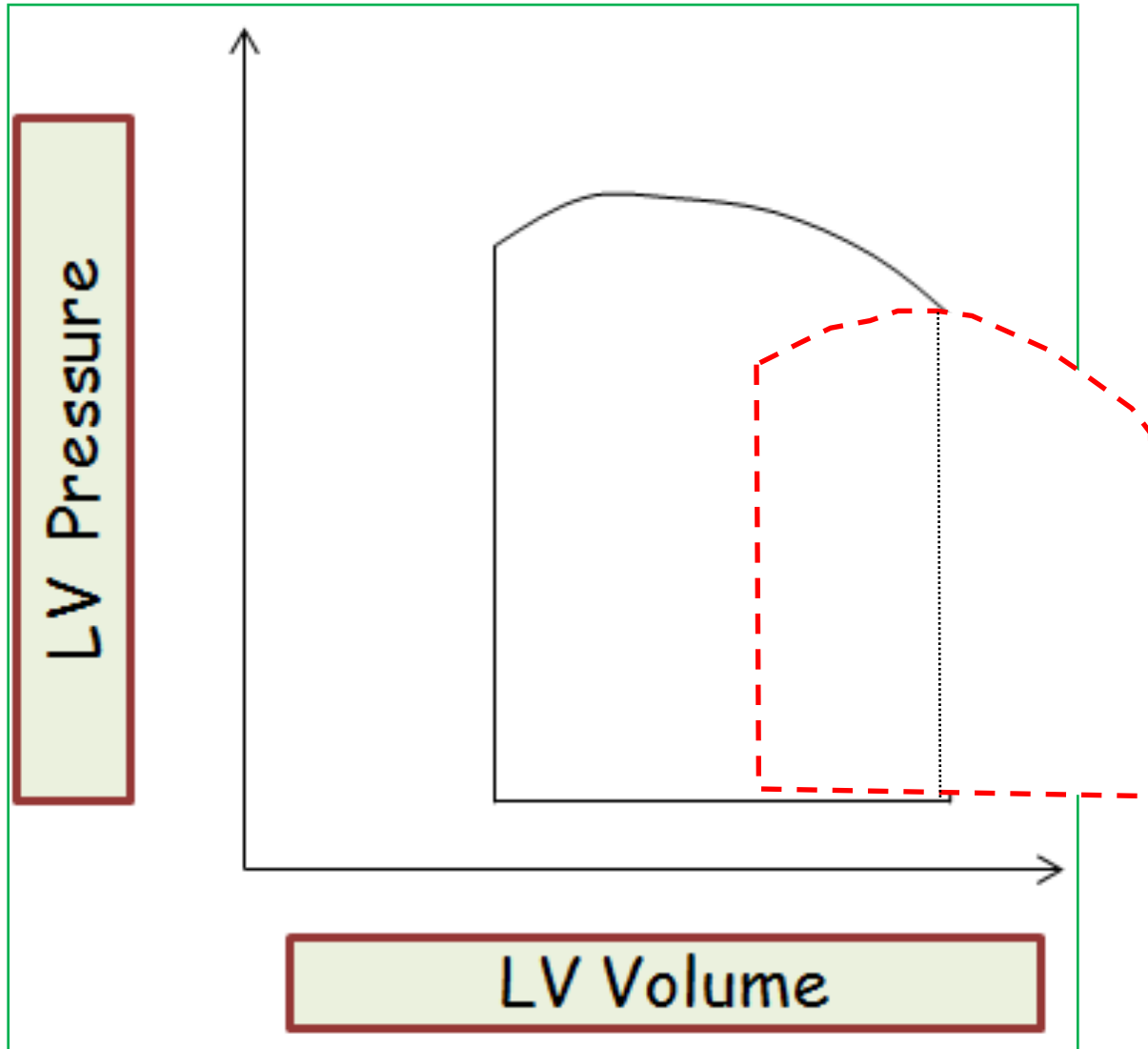
Neurohumoral response?

↑ SNS, RAA, ADH

└─→  $TPR_{\Omega}$ , VR



## Pressure-Volume Loop

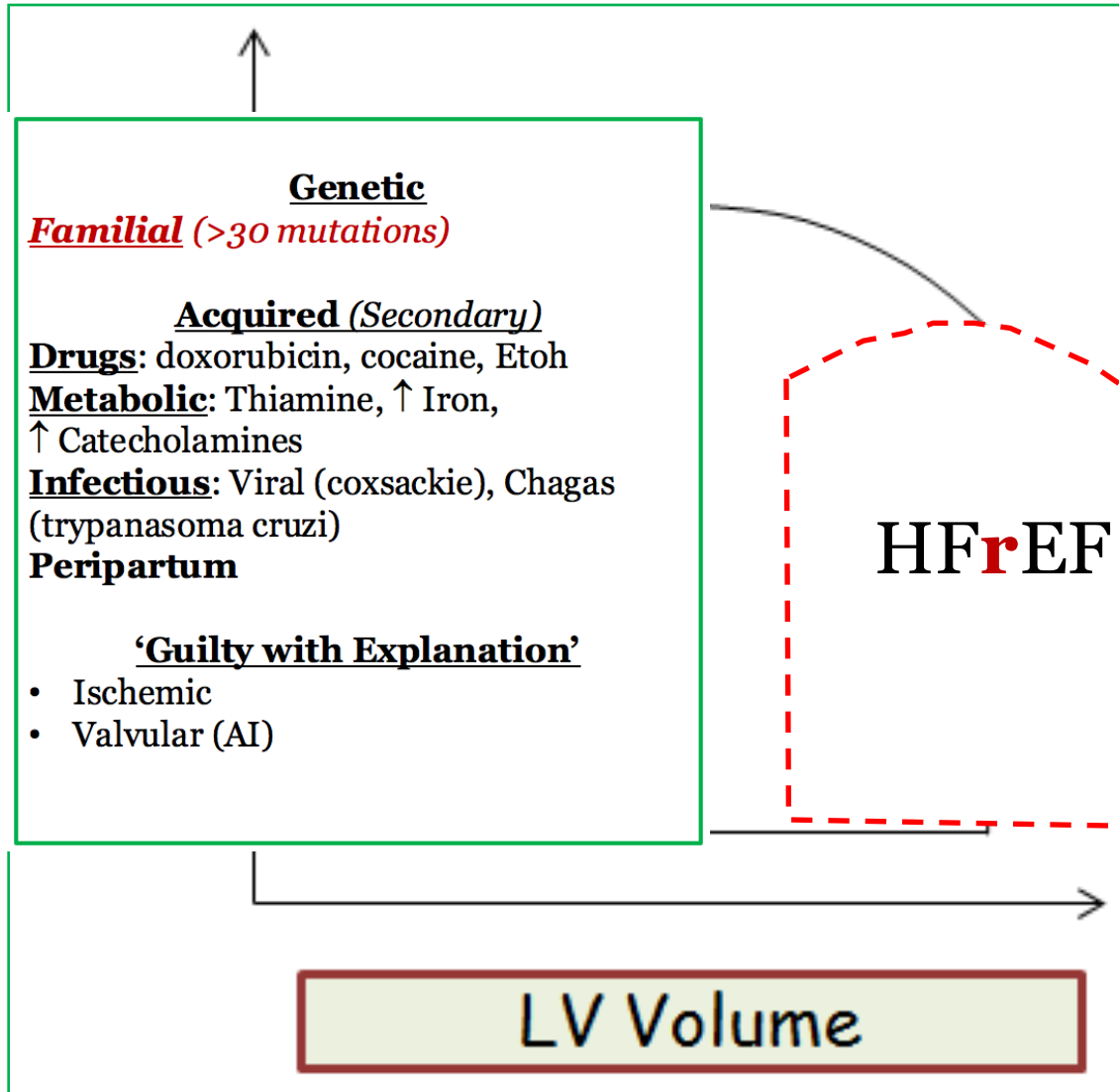


DCM:

EDV?  
Stroke Volume?  
ESV?

Which of the following conditions is the patient most likely to be suffering from?

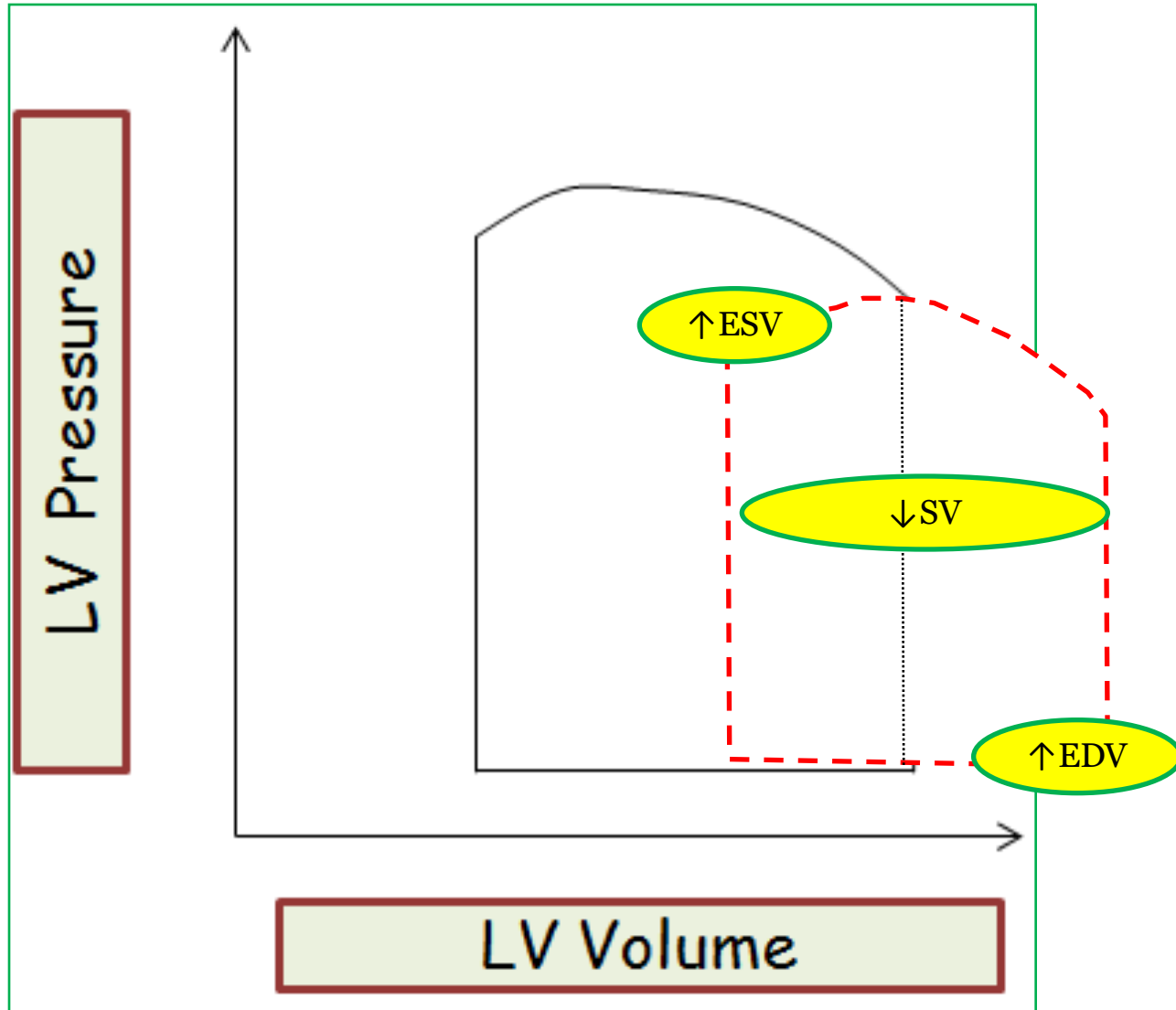
# Pressure-Volume Loop



DCM:  
EDV?  
Stroke Volume?  
ESV?

Which of the following conditions is the patient most likely to be suffering from?

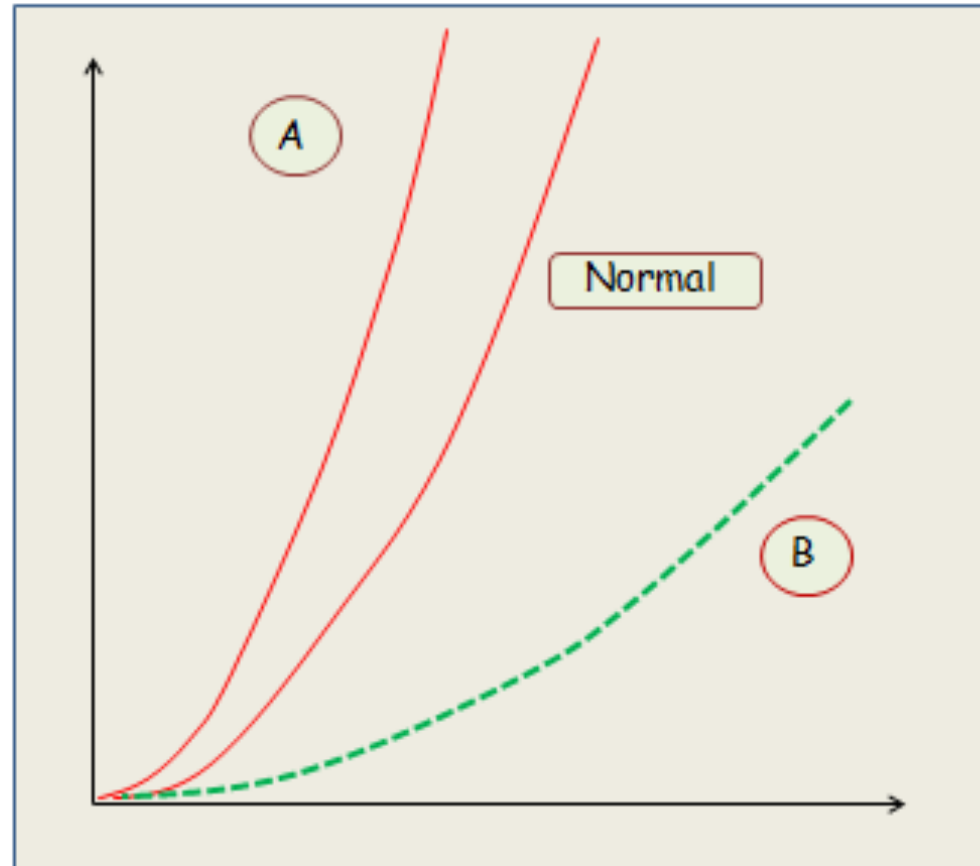
# HF<sub>r</sub>EF



## Pressure-Volume (*Compliance*) Curve

*Compliance = Distensibility*

LV Volume



Normal

A

B

Intraventricular pressure

$$C = \Delta V / \Delta P$$

# Pressure-Volume (*Compliance*) Curve

*Compliance = Distensibility*



# Pressure-Volume (*Compliance*) Curve

*Compliance = Distensibility*

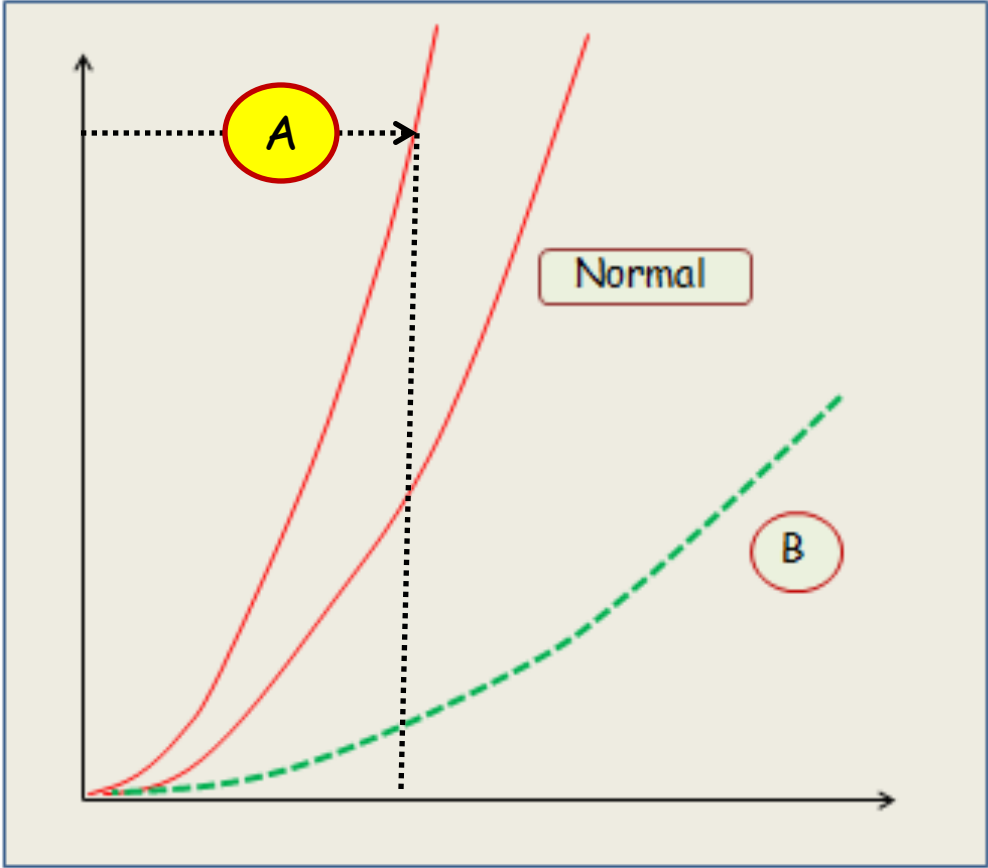




Pressure-Volume (*Compliance*) Curve

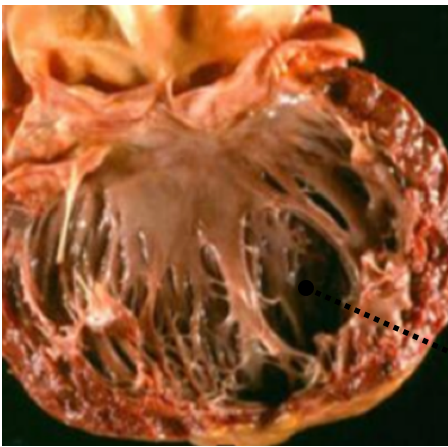
*Compliance = Distensibility*

LV Volume



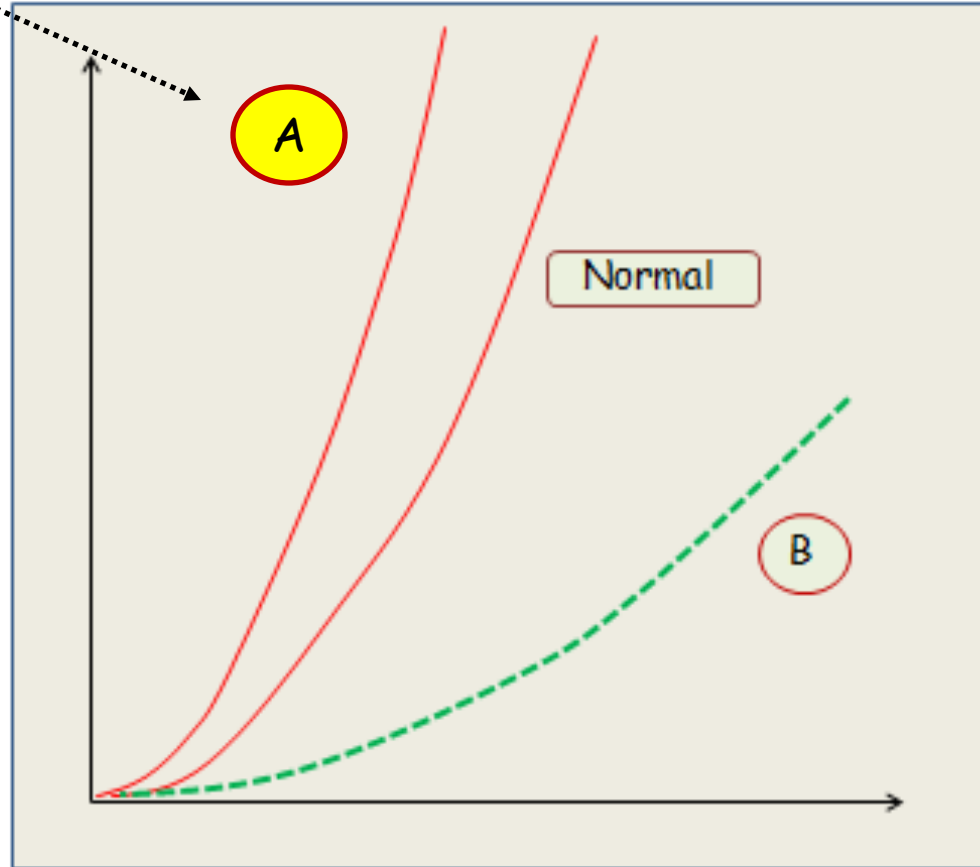
$$C = \Delta V / \Delta P$$

Intraventricular pressure



Curve A:  
Highly compliant (C) →  
High Volume at Low Pressure

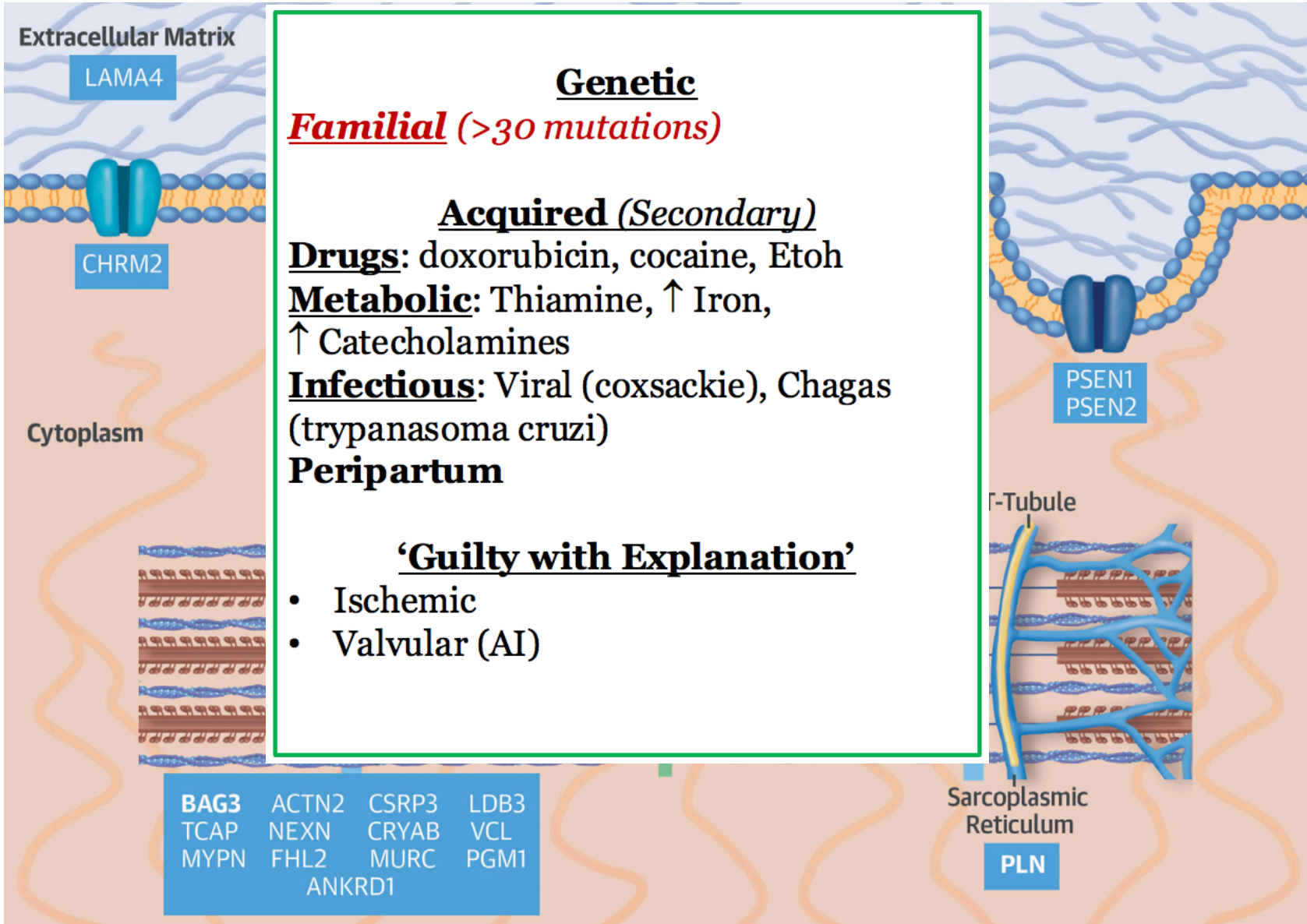
LV Volume



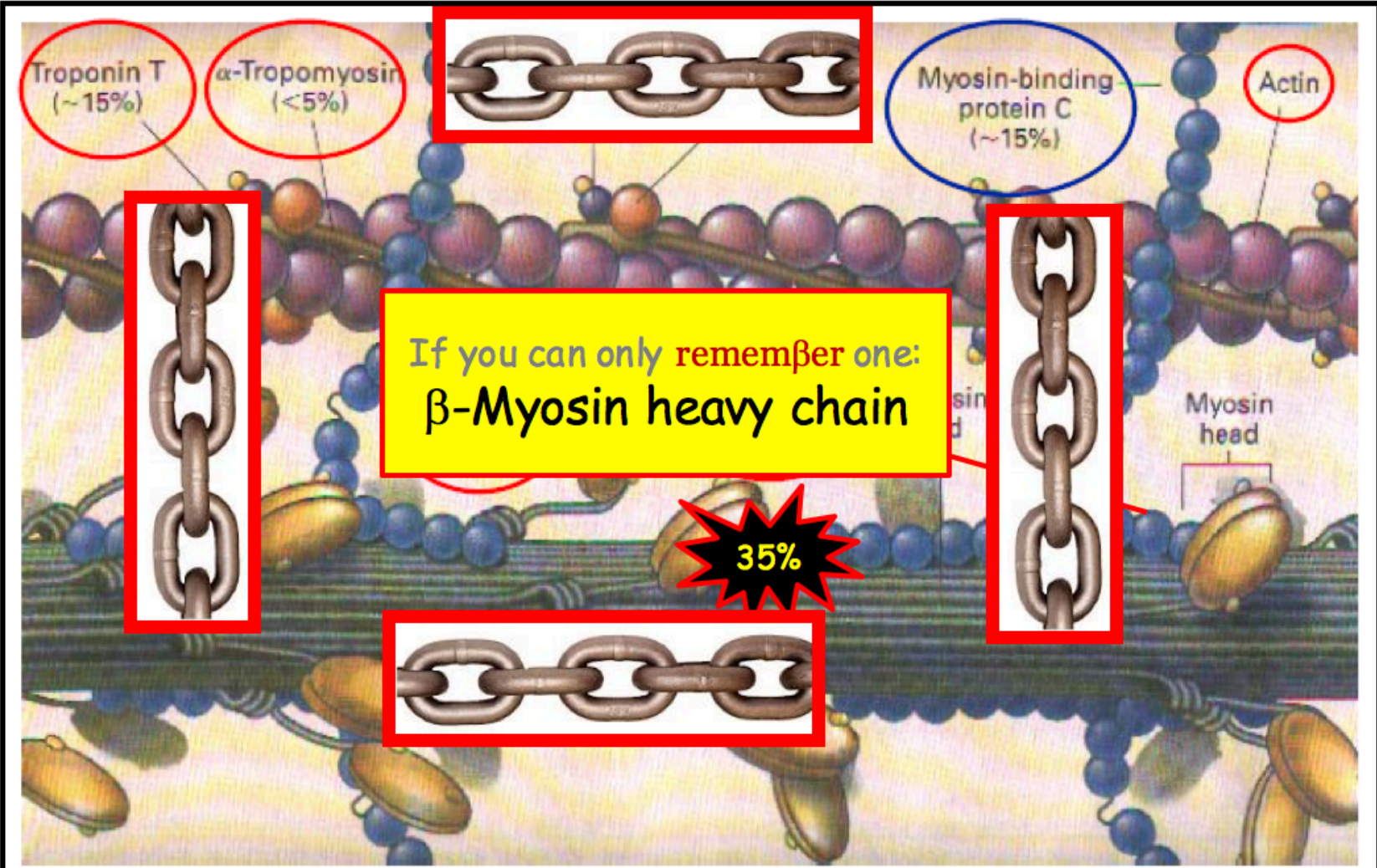
$$C = \Delta V / \Delta P$$

Intraventricular pressure

# DCM: Pathology

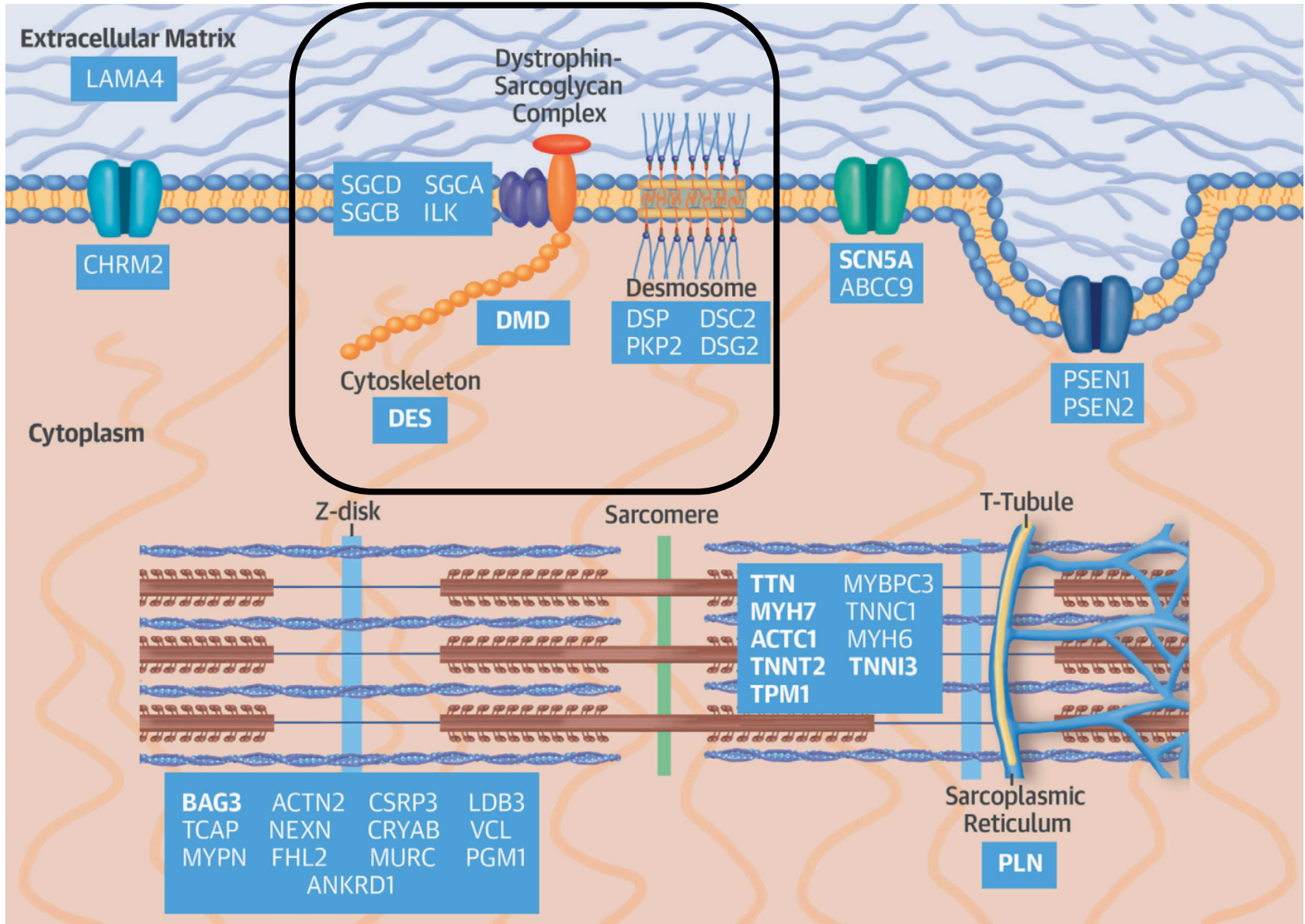


Dilated *Hypertrophic* Cardiomyopathy: **100% Genetic**

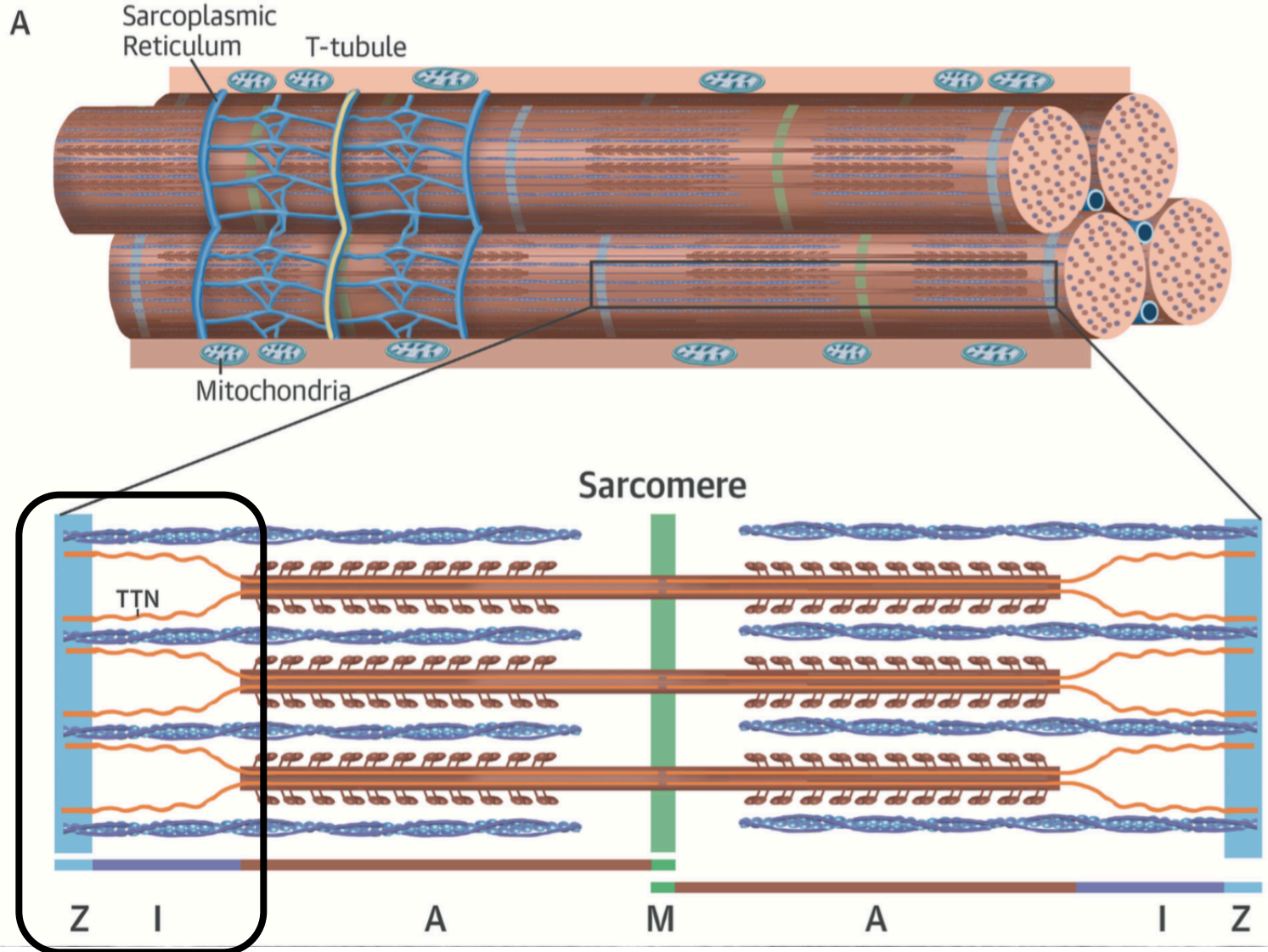




# Familial Dilated Cardiomyopathy Mutation: *Dystrophin*

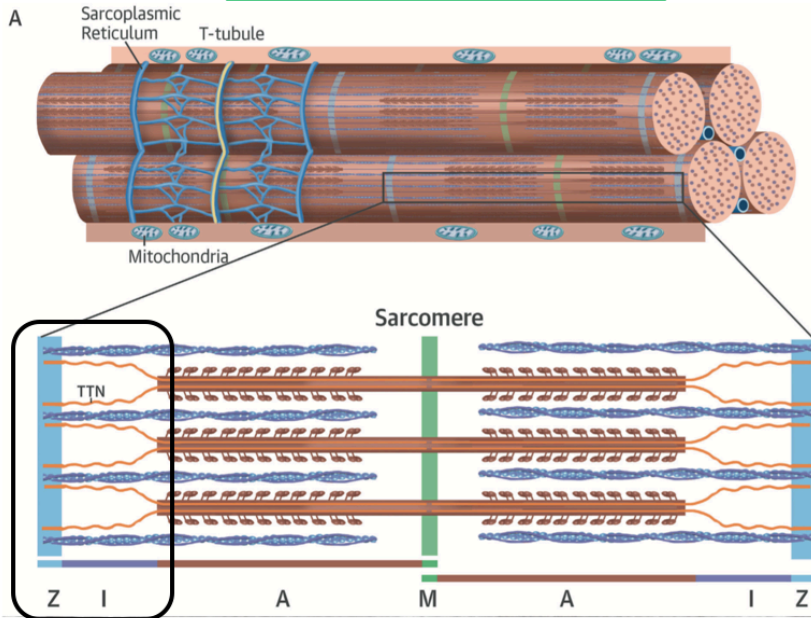


Familial Dilated Cardiomyopathy  
Mutation: *Titin*

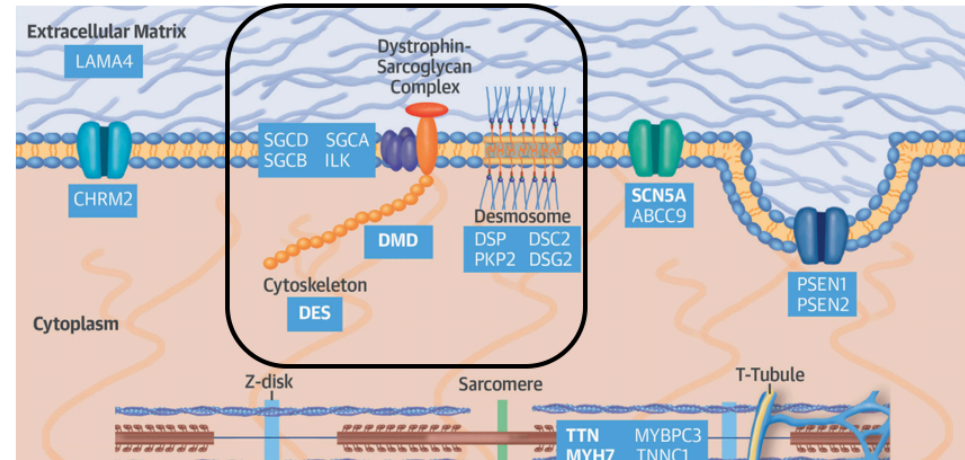




**Familial Dilated Cardiomyopathy**  
Mutation: *Titin*



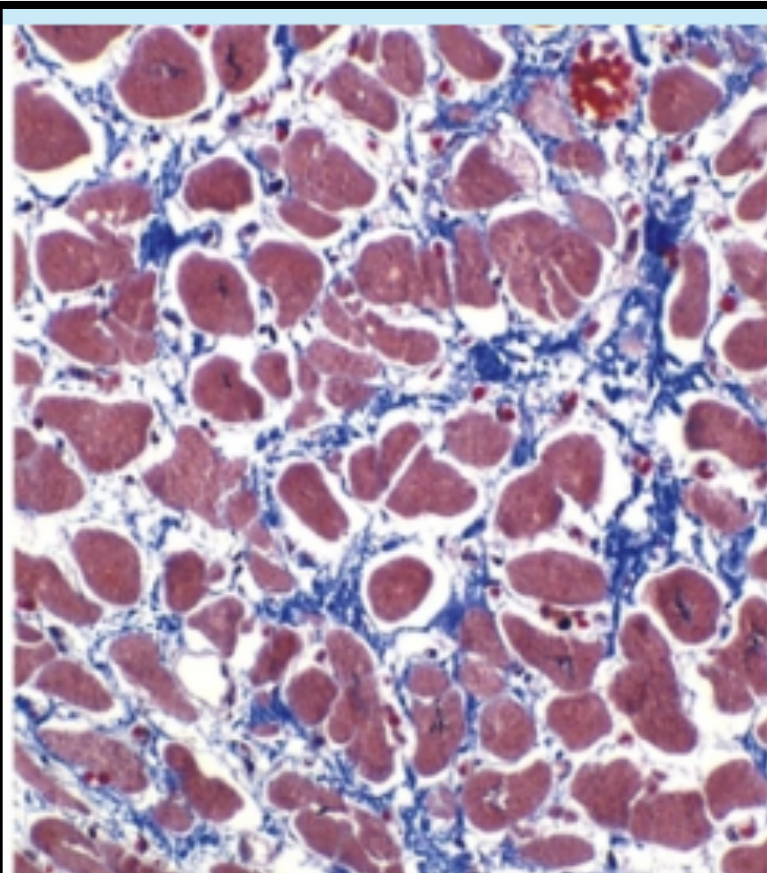
**Familial Dilated Cardiomyopathy**  
Mutation: *Dystrophin*



‘Clarification’:

‘...categorized as a distinct set of conditions with a functional and structural phenotype AND **often** with a **genetic component**. In discussions of CM, ischemia, HTN and valvular causes are excluded although they do share hemodynamic and pathophysiologic parameters.’

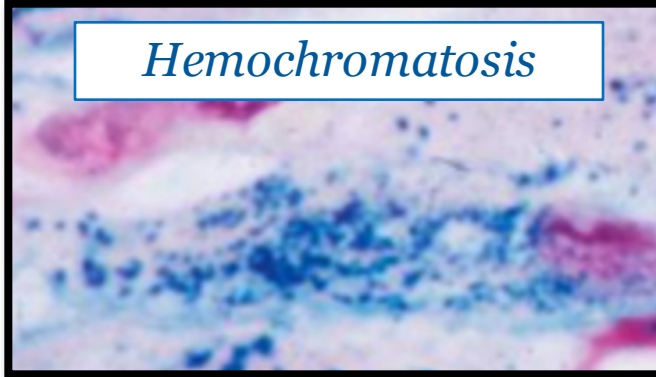
Familial (Genetic) DCM  
Histopathology is *nonspecific*



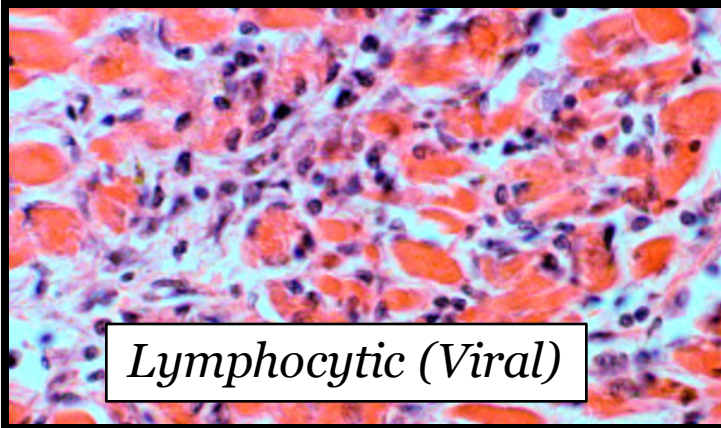
MM: Hypertrophy/Atrophy  
Fibrosis: Variable  
Morphology  $\neq$  Dysfunction

Acquired DCM  
*Pathology to be familiar with...*

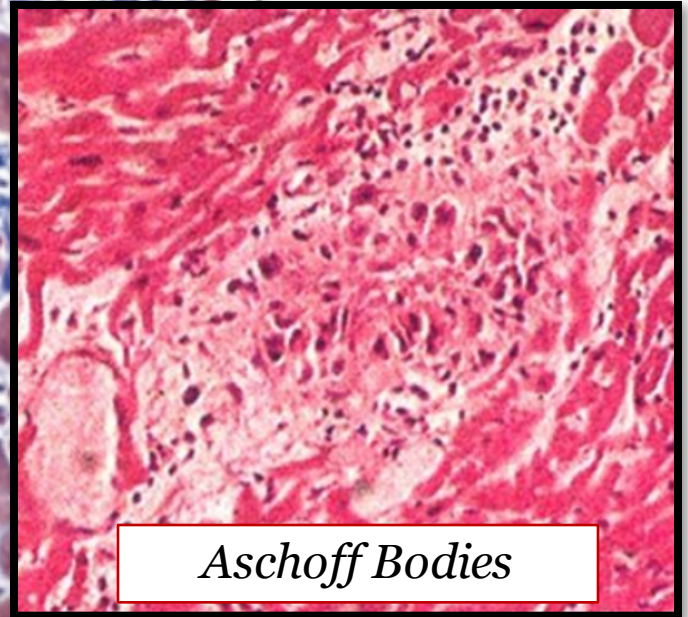
*Hemochromatosis*



*Lymphocytic (Viral)*

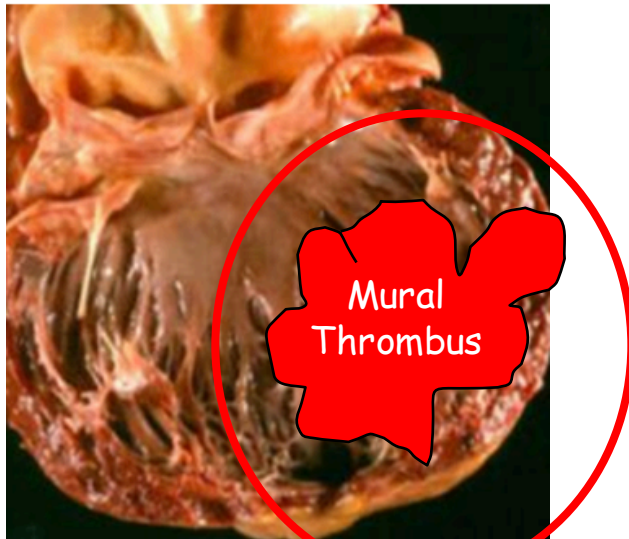
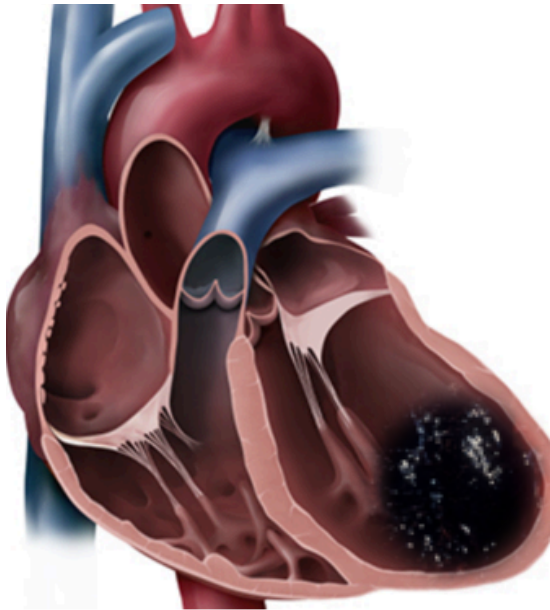


*Aschoff Bodies*





## Dilated Cardiomyopathy: *Complication*



Mural  
Thrombus

### Genetic

Familial (>30 mutations)

### Acquired (Secondary)

Drugs: doxorubicin, cocaine, Etoh

Metabolic: Thiamine, ↑ Iron,  
↑ Catecholamines

Infectious: Viral (coxsackie), Chagas  
(trypanasoma cruzi)

Peripartum

### 'Guilty with Explanation'

- Ischemic/Cardiogenic Shock
- Valvular (AI)

# Dilated Cardiomyopathy: *Complication*

How They Play The Game:  
*Common Language:* Patient with CHF (S3, rales)...

Pathophysiology:  
Pressure-volume loop...  
Pressure-volume curve...

*What complication?*

## Genetic

***Familial*** (>30 mutations)

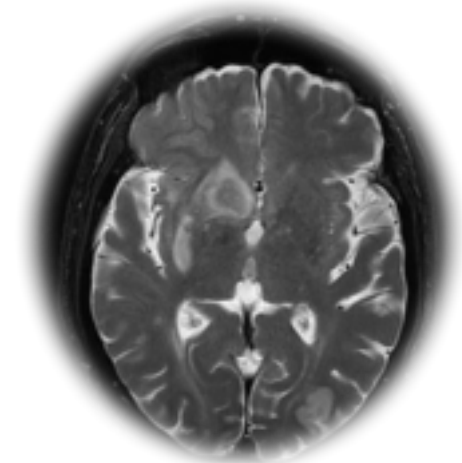
## Acquired (Secondary)

**Drugs:** doxorubicin, cocaine, Etoh

**Metabolic:** Thiamine, ↑ Iron,  
↑ Catecholamines

**Infectious:** Viral (coxsackie), Chagas  
(trypanasoma cruzi)

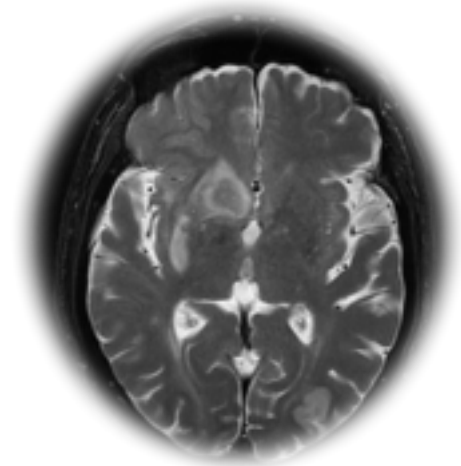
**Peripartum**





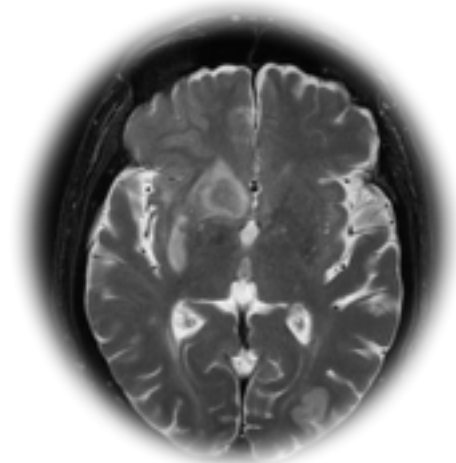
Here is the *short list* of Stroke PLUS Cardiac Disease:

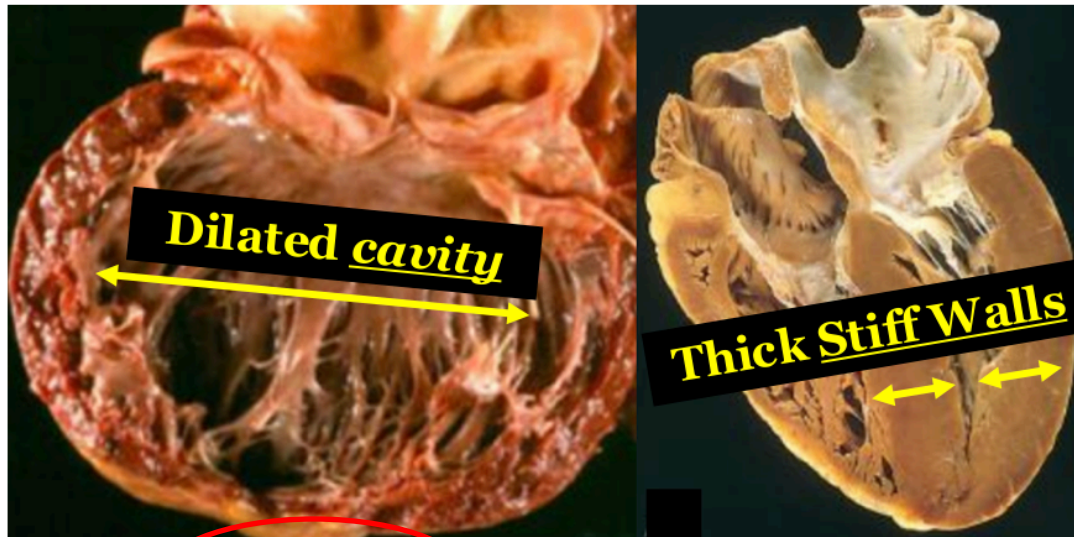
1. DCM (*all causes*)
2. Atrial fibrillation
3. Patent Foramen Ovale (PFO)



Here is the *longer list* of Stroke PLUS Cardiac Disease:

1. DCM (*all causes*)
2. Atrial fibrillation
3. Patent Foramen Ovale (PFO)
4. Valvular vegetations (all causes)
5. Atrial myxoma





HF<sub>r</sub>EF

HF<sub>p</sub>EF

In Summary,

1. the topic of *Dilated Cardiomyopathy* is simply about the **failing heart**
2. it is the heart with **reduced EF** (contrast with HF<sub>p</sub>EF)
3. it is about recognizing the *pathophysiologic derivatives* (i.e. interpreting curves)
4. the *pathology* is non-specific
5. the *genetic derivatives* are narrow
6. there are limited *complications* beyond those of the failing heart

Heart Failure and the Cardiomyopathies for the USMLE Step One Exam:  
*Part III: Dilated Cardiomyopathy and Test Derivatives*



HF<sup>r</sup>EF

HF<sup>p</sup>EF

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*University of Massachusetts Medical School*  
[www.12DaysinMarch.com](http://www.12DaysinMarch.com)  
*E-mail: Howard@12daysinmarch.com*