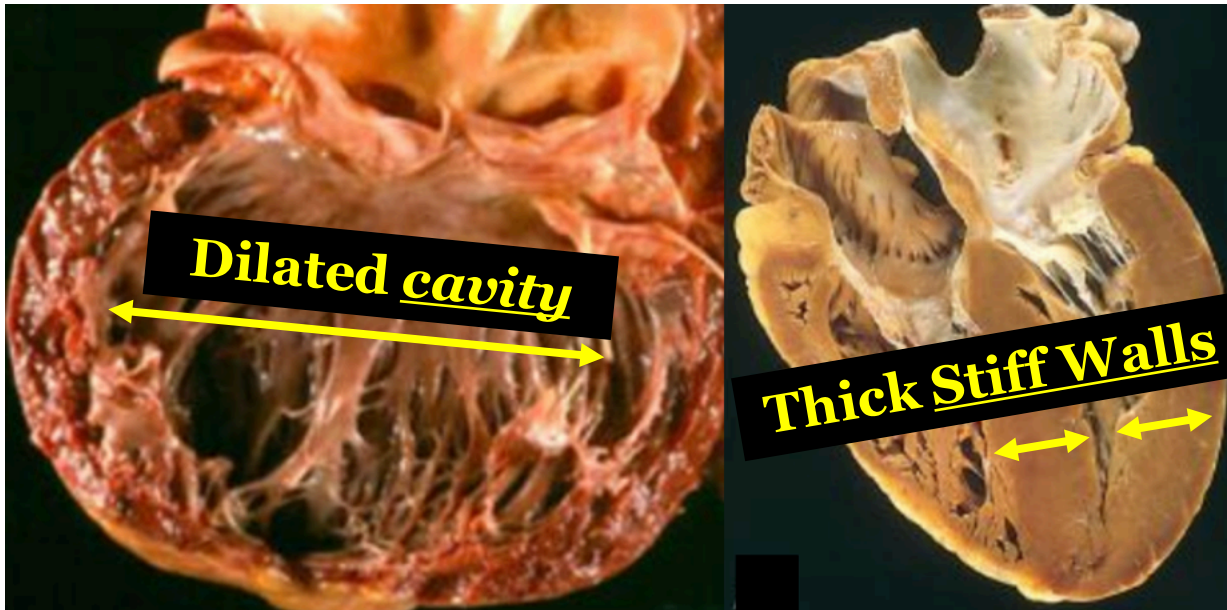


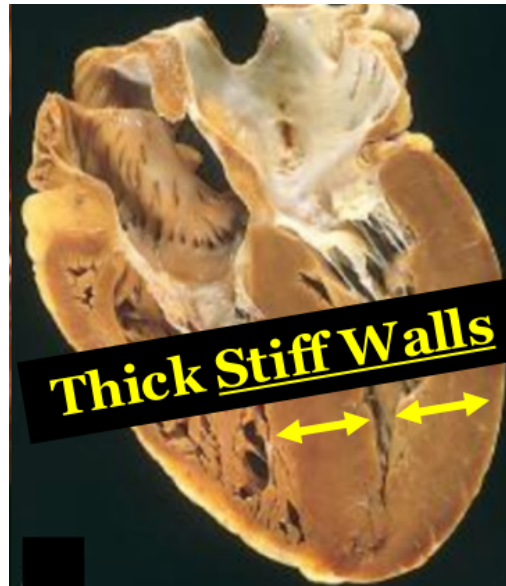
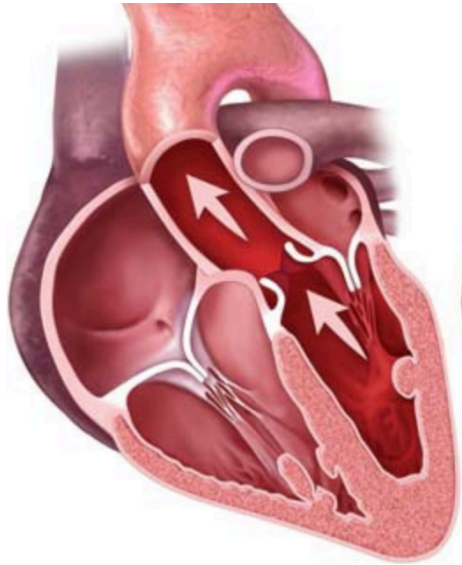
Heart Failure and the Cardiomyopathies for the USMLE Step One Exam:
Part IV: Restrictive Cardiomyopathy and Diastolic Heart Failure



HF^rEF

HF^pEF

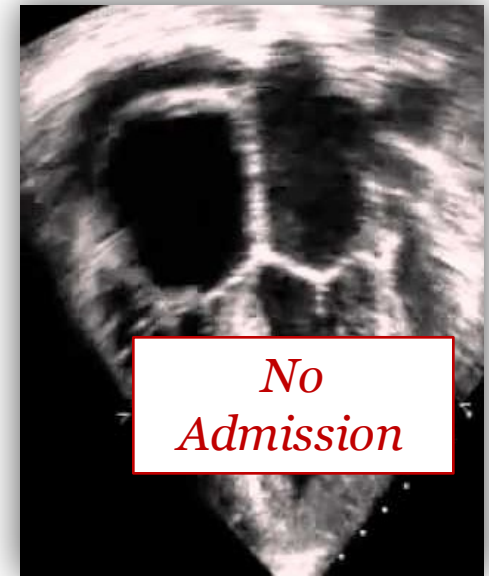
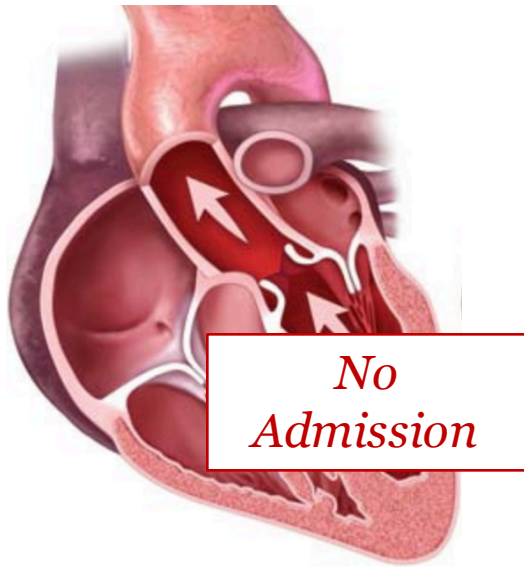
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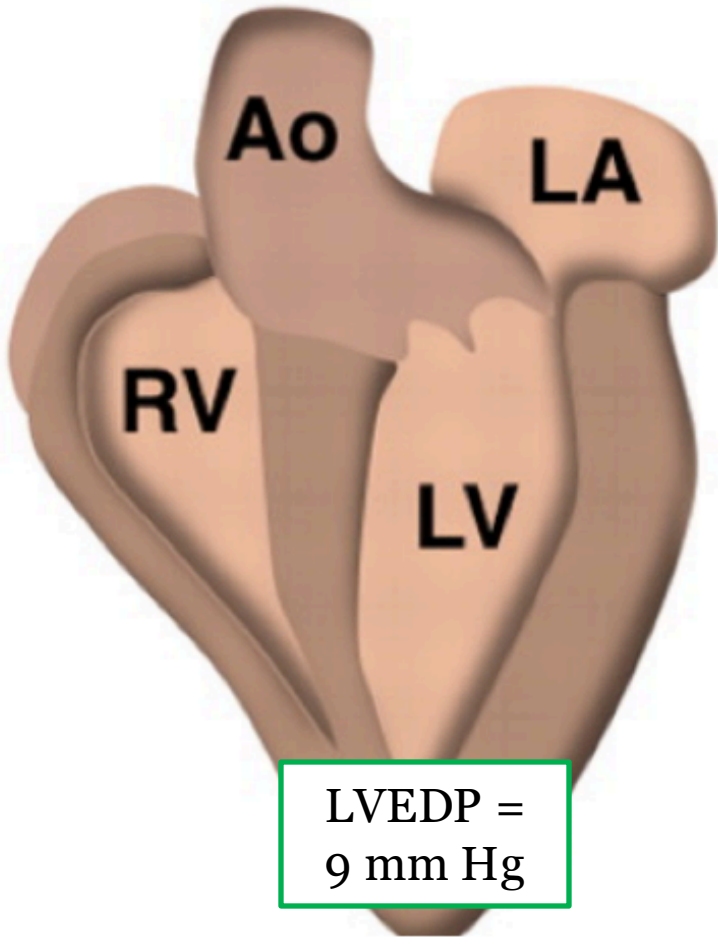
Restrictive Cardiomyopathy (HF_pEF) is a *Physiologic Disorder*

In combination with Diastolic (HF) Dysfunction, it is characterized by a narrow set of *hemodynamic parameters*

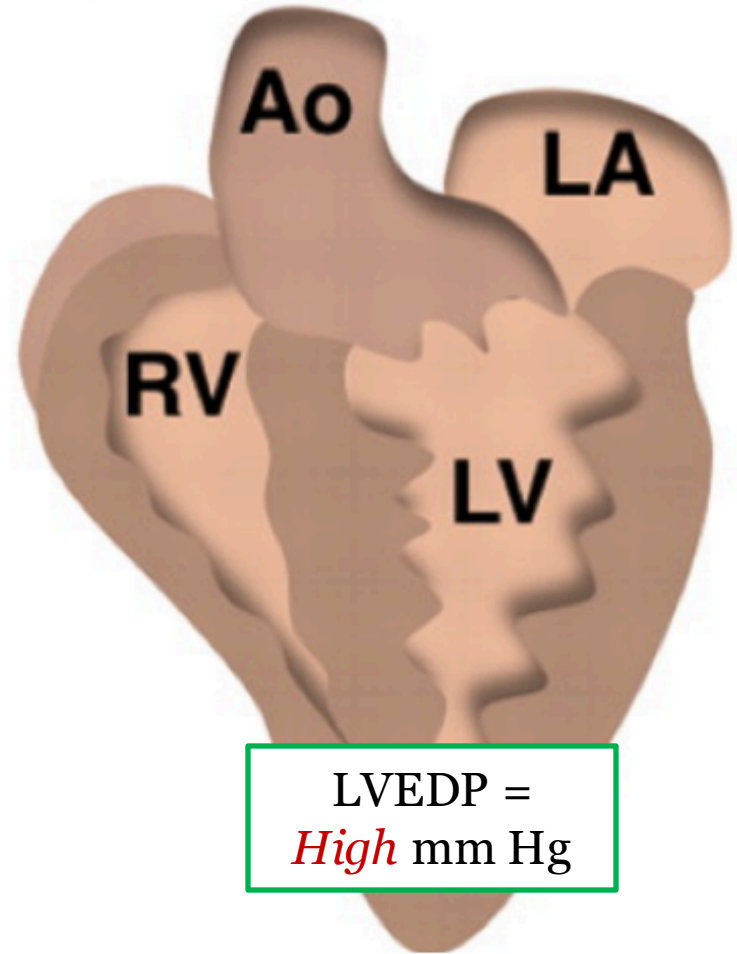
There are a *small number of conditions* that share these common *pathophysiologic derivatives*.



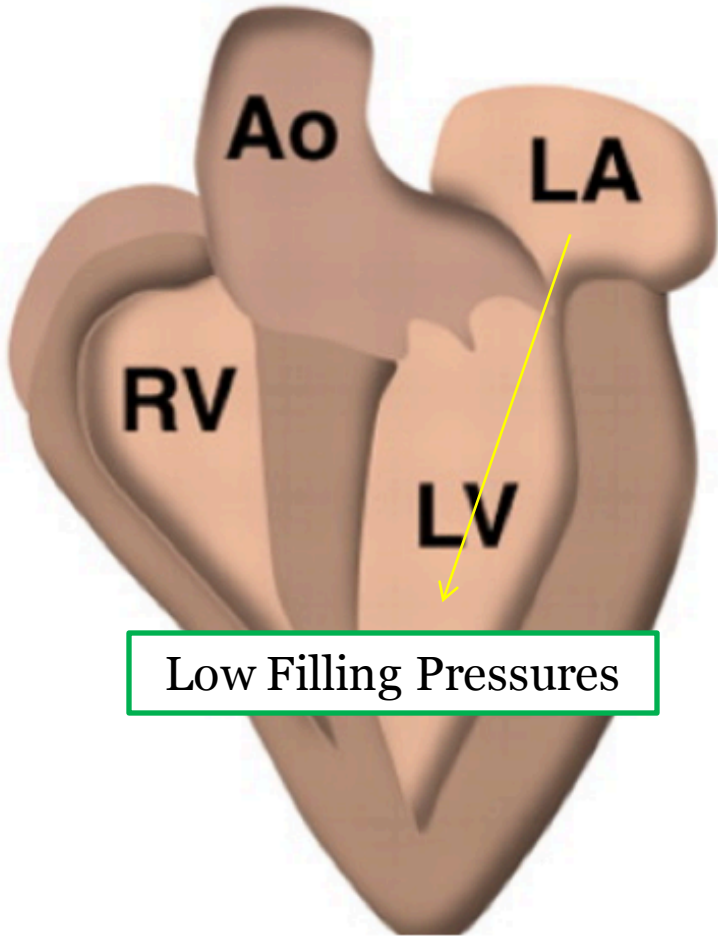
The Problem: an inability to fill the LV at normal pressures.
It's too stiff.



Normal Heart

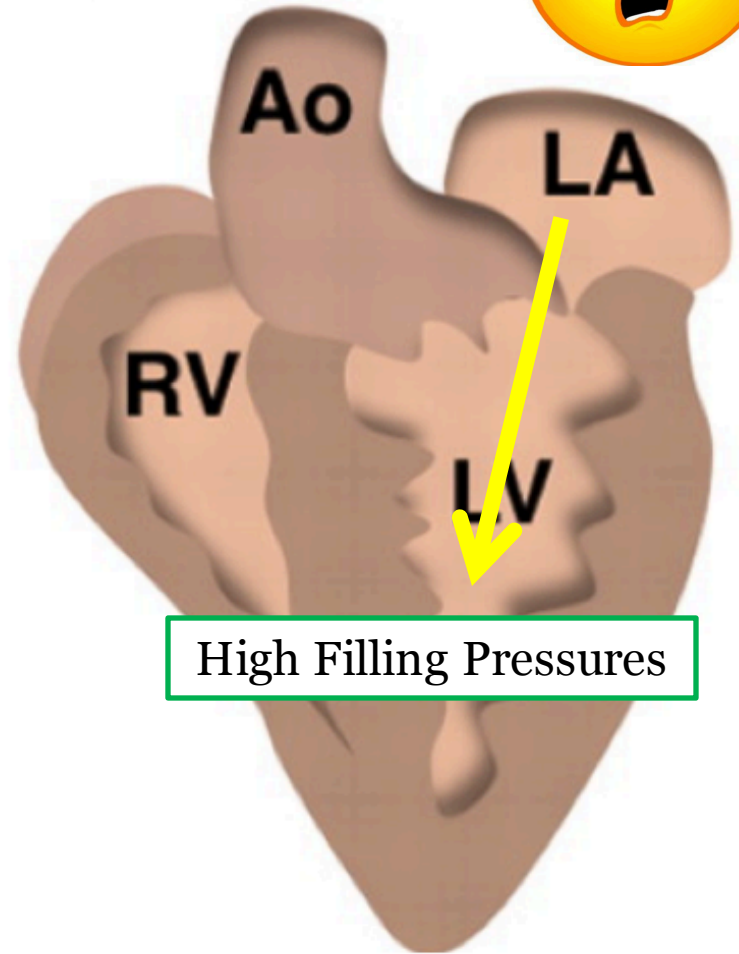


RCM/Diastolic HF



Low Filling Pressures

Normal Heart



High Filling Pressures

RCM/Diastolic HF



Admission Granted
but only under *high pressure*

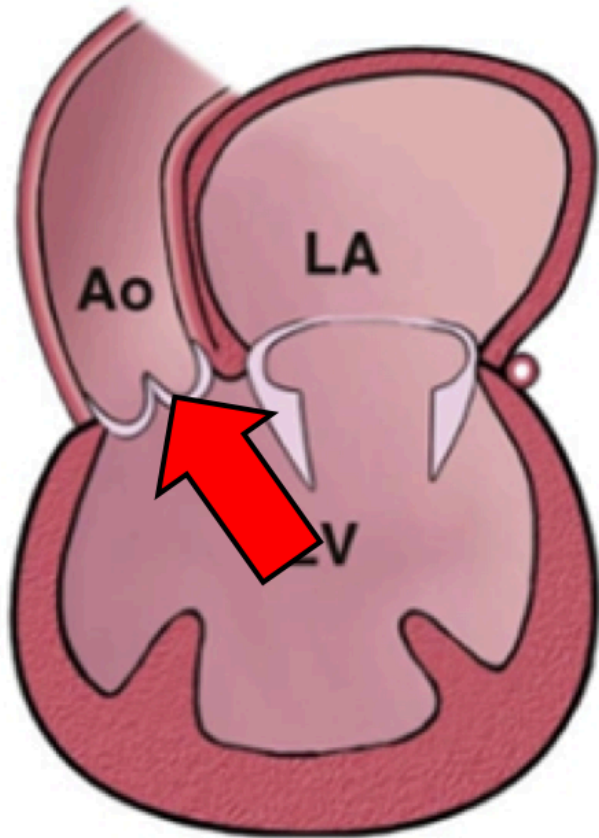


Admission Granted
but only under *high pressure*

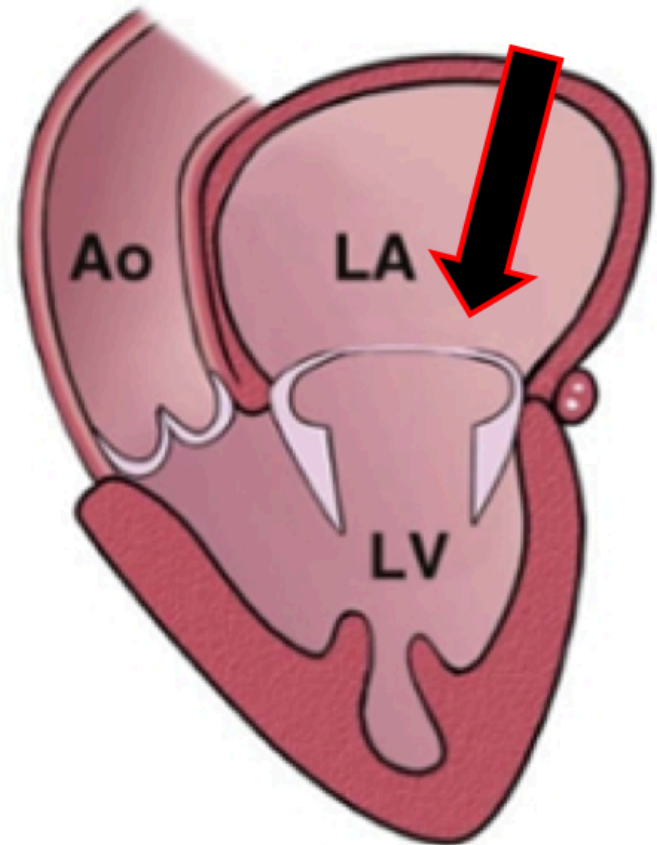
- LVEDV: normal (blood is forced into the LV under pressure)
- EF (HF_pEF): normal (the pump works; *it is just very stiff*)
- **LVEDP: *elevated*** (for any given volume of blood, the IV pressure is elevated)



Dilated CM:
Pumping (contractility) problem

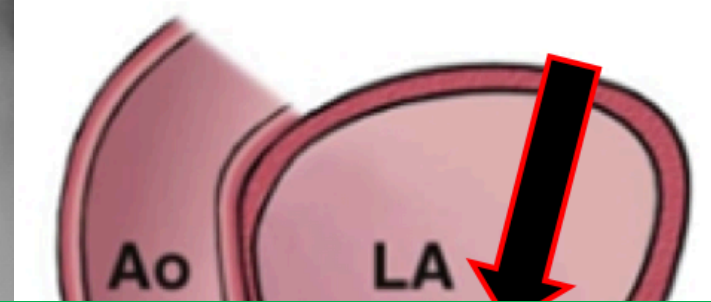


Restrictive CM:
Filling problem

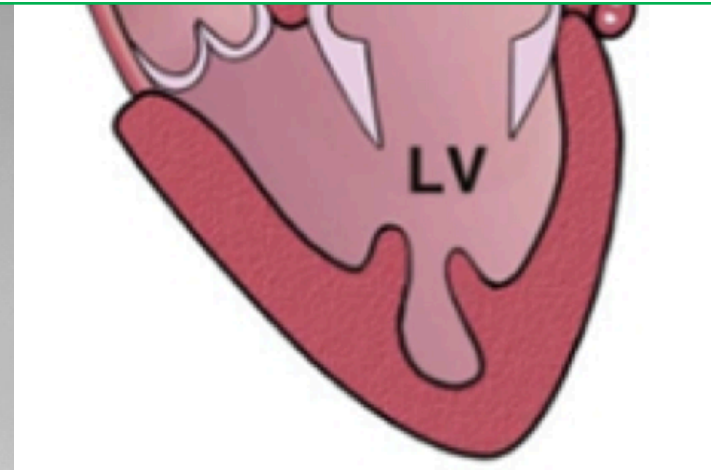
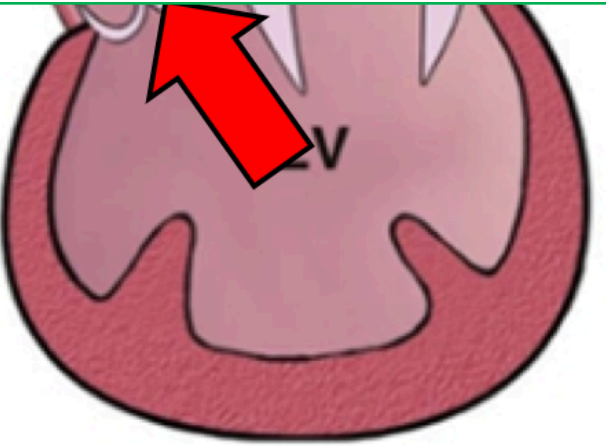


Dilated CM:
Pumping (contractility) problem

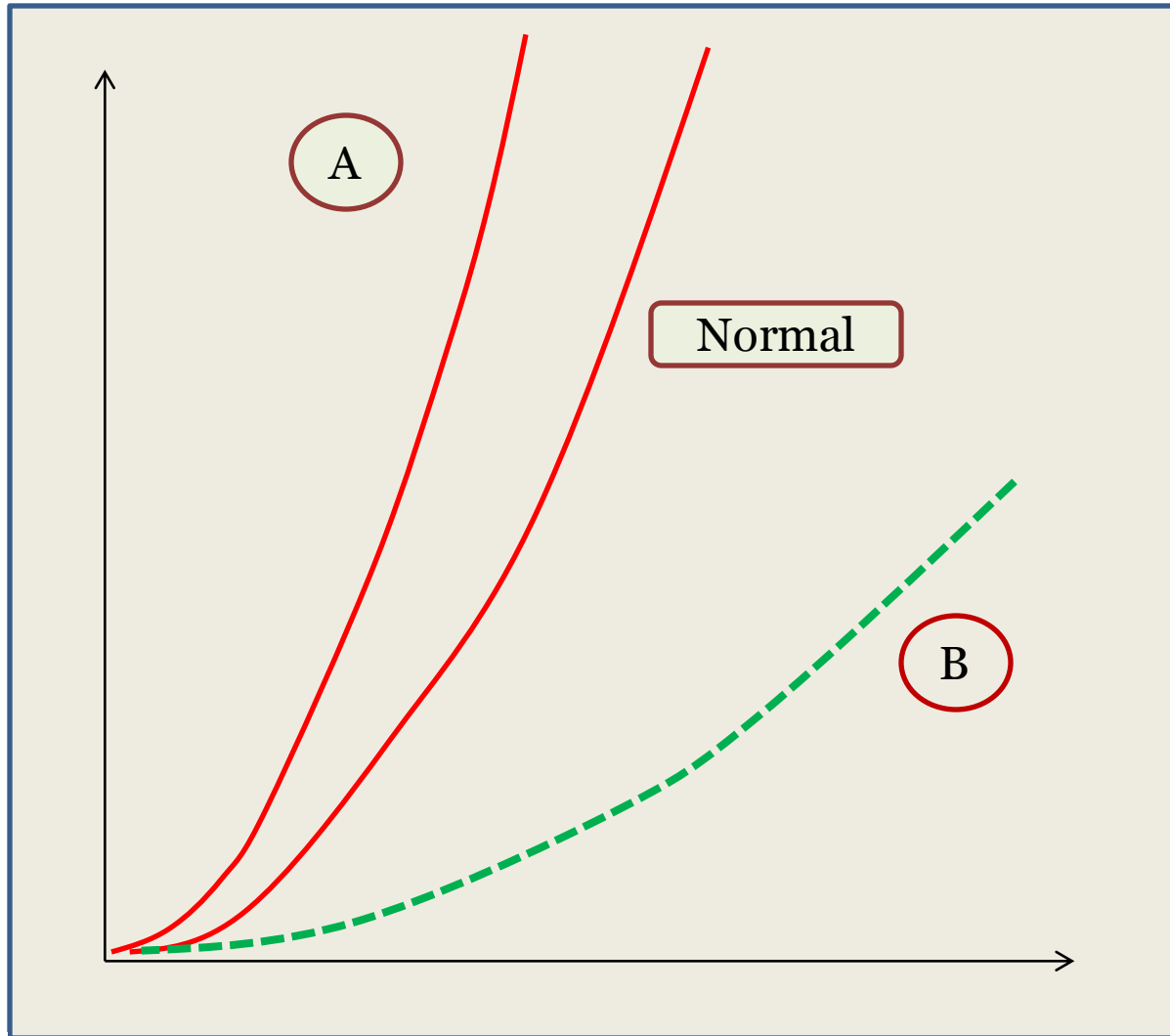
Restrictive CM:
Filling problem



Both result in *venous congestion* and *arterial hypoperfusion*



Pathophysiology: the Pressure-Volume (*Compliance*) Curve
‘Which curve is consistent with ‘restrictive’ disease?’



LV Volume

Normal

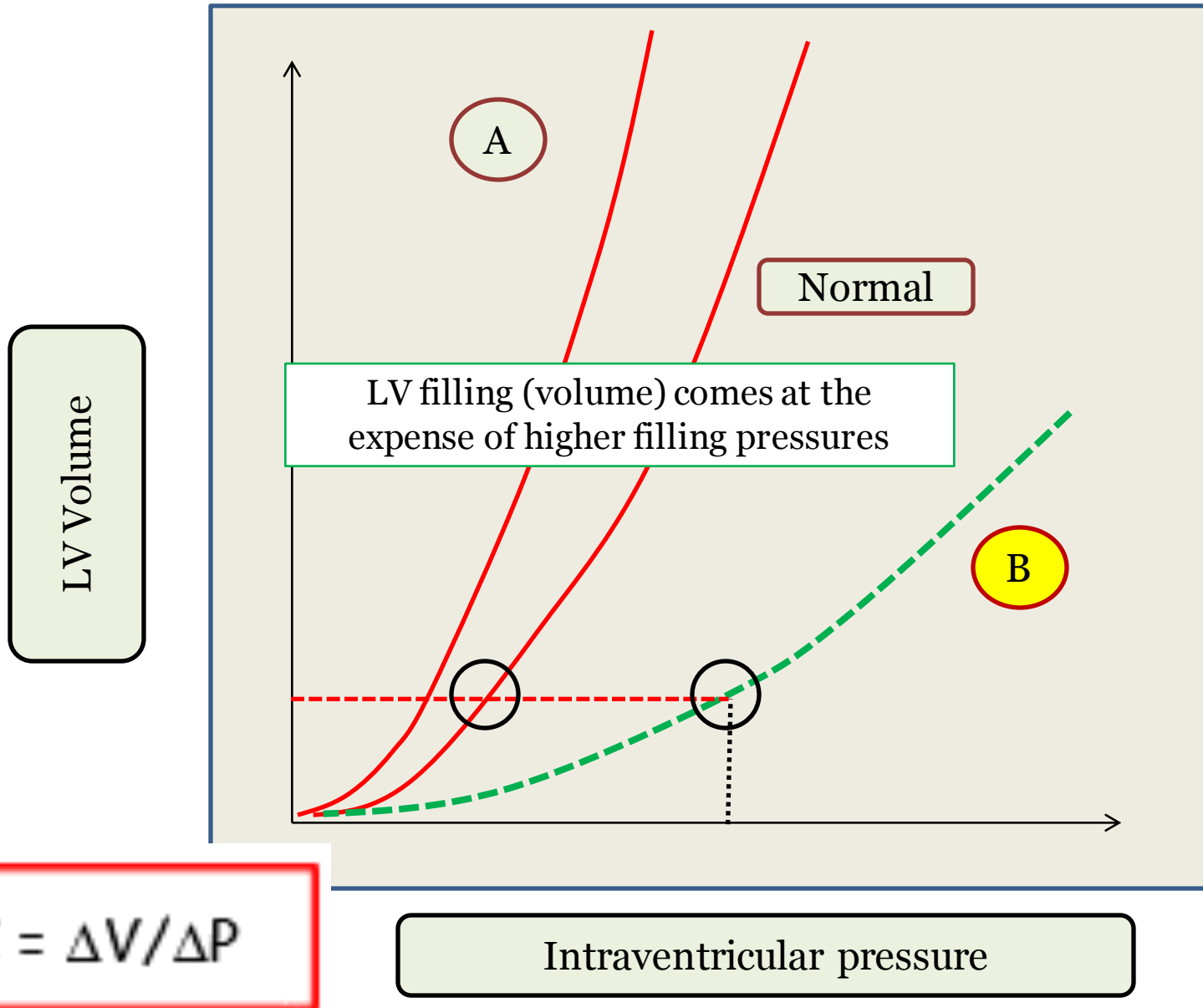
B

A

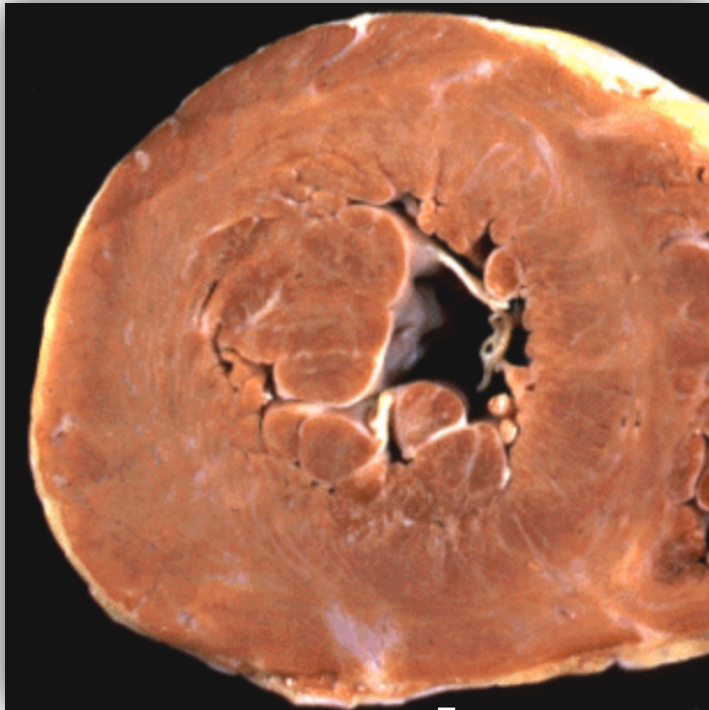
$$C = \Delta V / \Delta P$$

Intraventricular pressure

Restriction (stiff) = Poor Distensibility (*decreased compliance*)

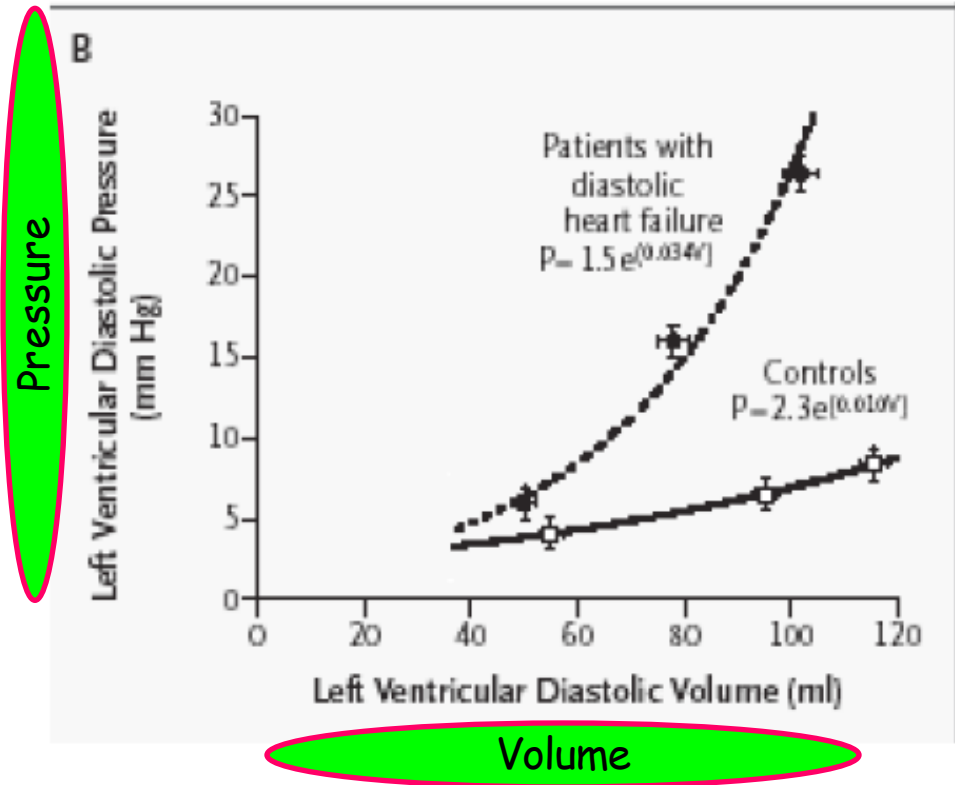
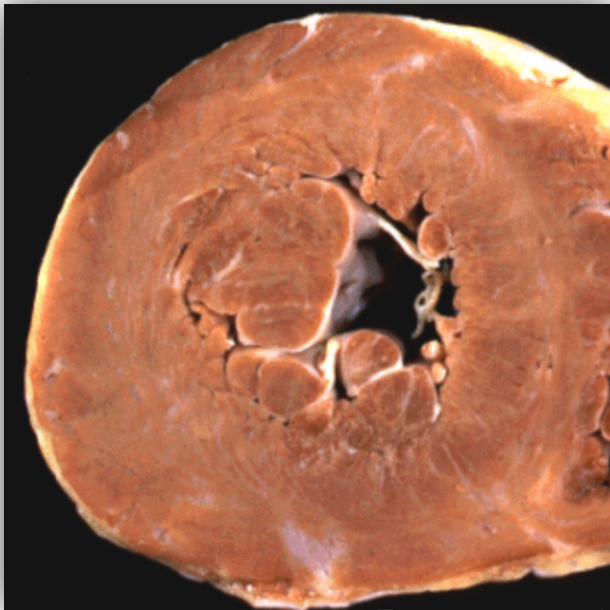


Restrictive Heart Disease: Diastolic Dysfunction
(HF_pEF)



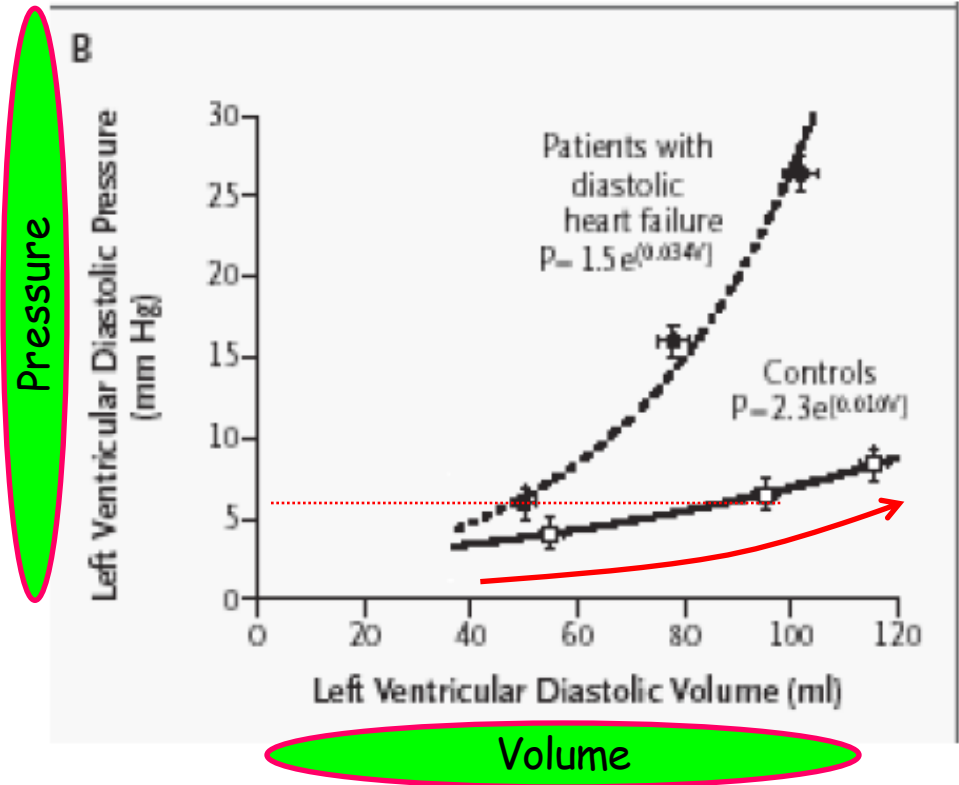
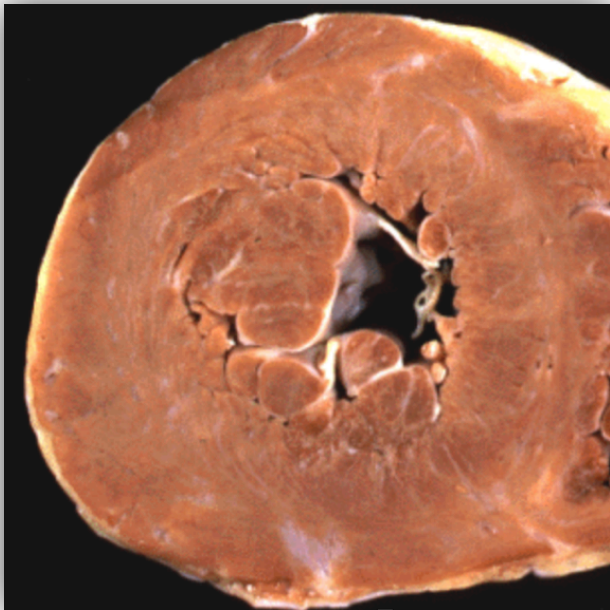
Restrictive Heart Disease: Diastolic Dysfunction (HF_pEF)

An inability to
fill at normal
pressures



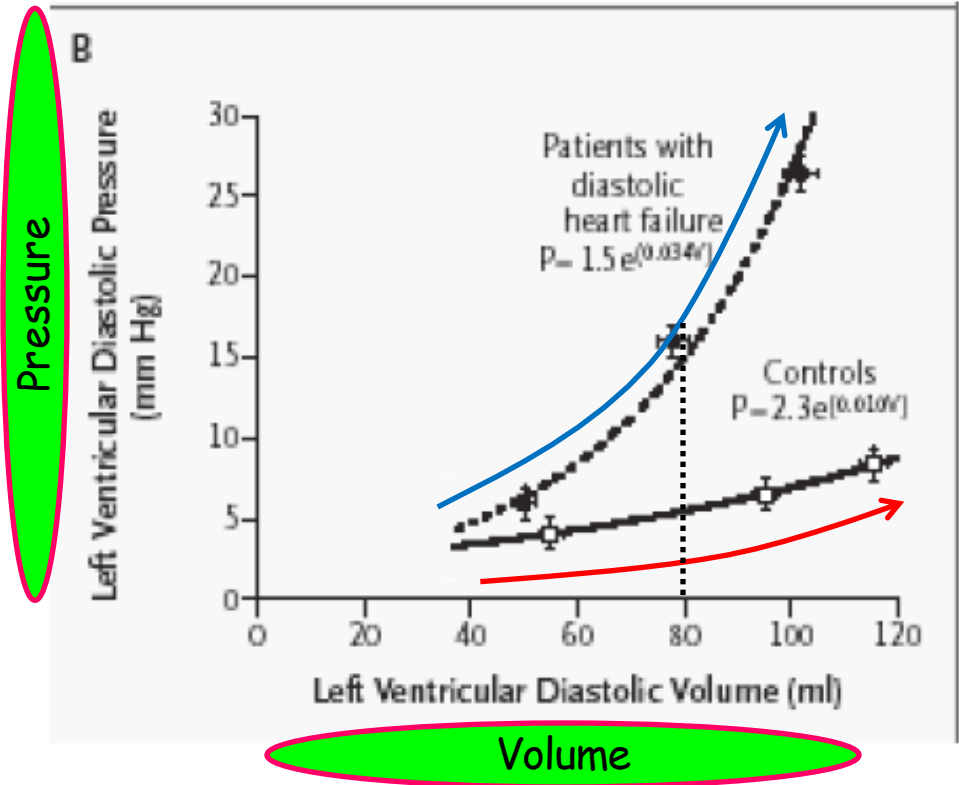
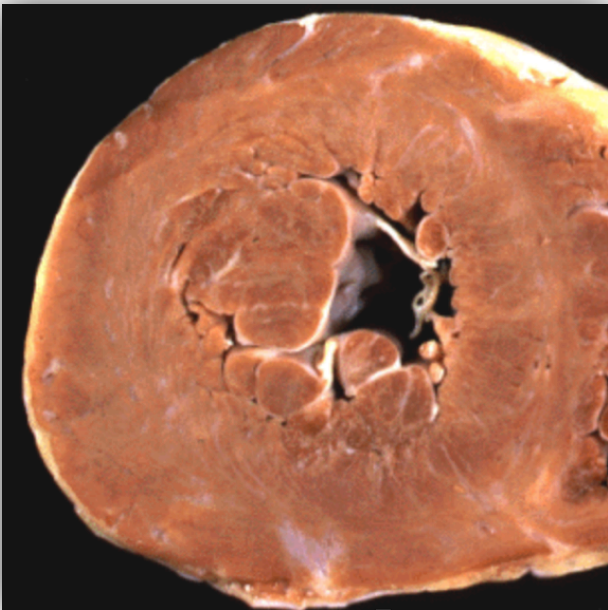
Diastolic Dysfunction (HF_pEF)

An inability to fill at normal pressures



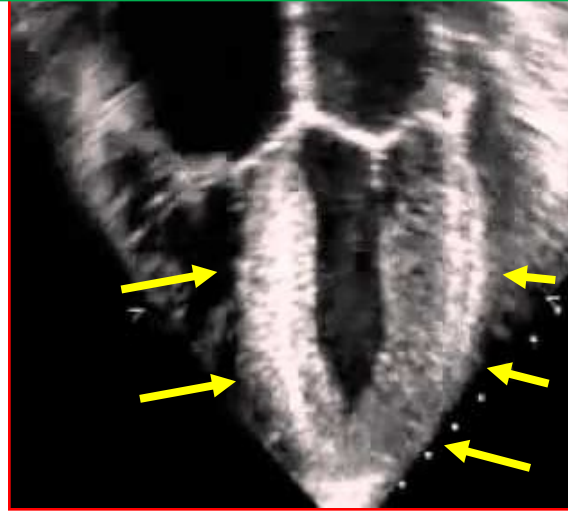
Diastolic Dysfunction (HF_pEF)

An inability to fill at normal pressures



Increased wall thickness with
outer dimensions remaining almost unchanged.

Result: narrow ventricular chamber size



Diastolic Dysfunction/RCM
Impaired ventricular relaxation
Poor compliance → Stiffness

- EF?
- LVEDV?
- LVEDP?





It's ***not*** a pumping problem, it's a filling problem

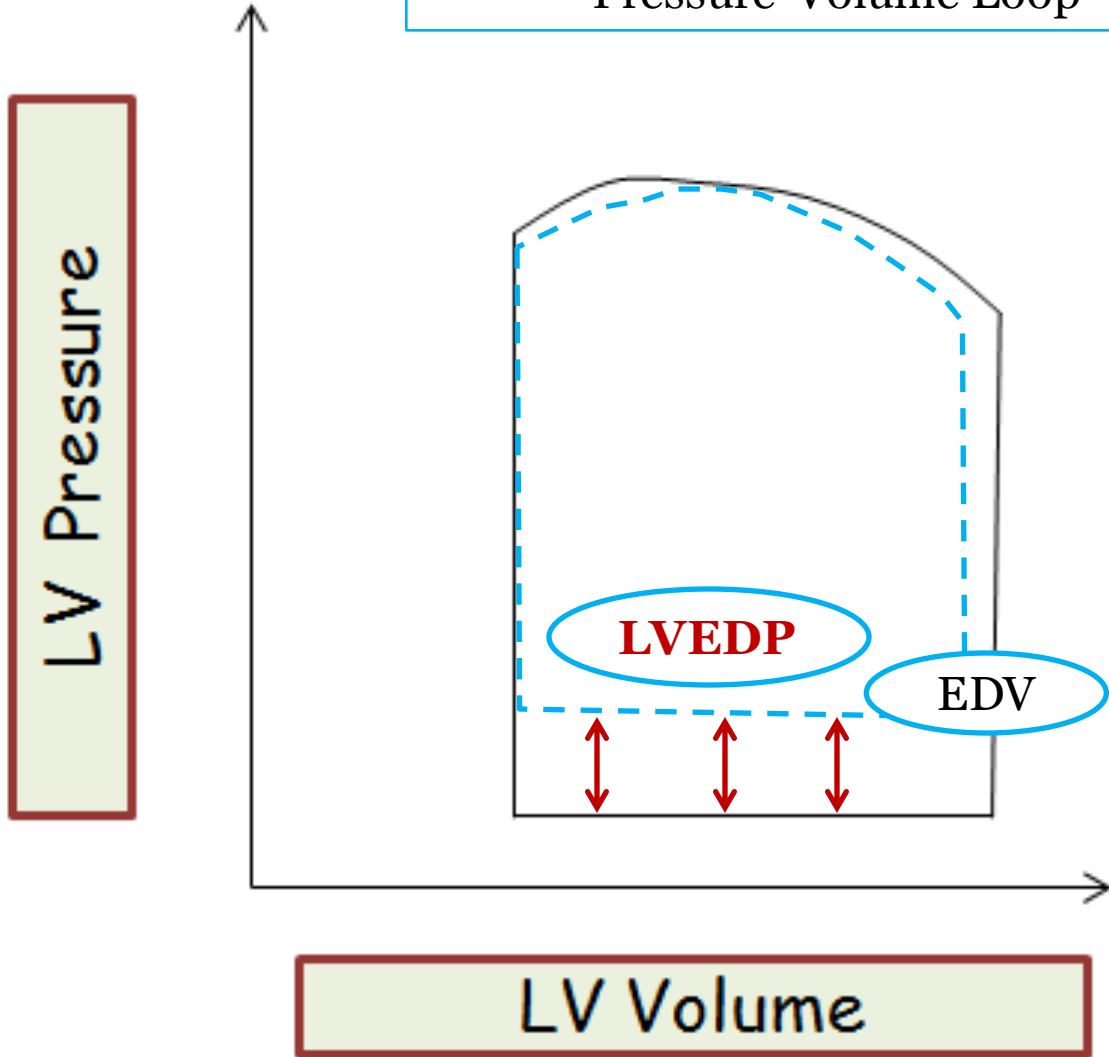
Diastolic Dysfunction/RCM

Impaired ventricular relaxation

Poor compliance → Stiffness

- EF: normal (or increased)
- LVEDV: normal
- LVEDP: ***increased***

Restrictive Heart Disease (all causes):
Pressure-Volume Loop



What is represented by the blue curve?

Adequate LVEDV comes at the expense of LVEDP.

Key Causes: RCM

Endomyocardial Fibrosis
(*Fibrosis of the endocardium*)



Loeffler Endomyocarditis
(*EMF plus eosinophils*)



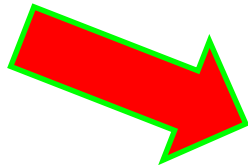
Endocardial Fibroelastosis
(*Fibroelastic thickening of the endocardium*)

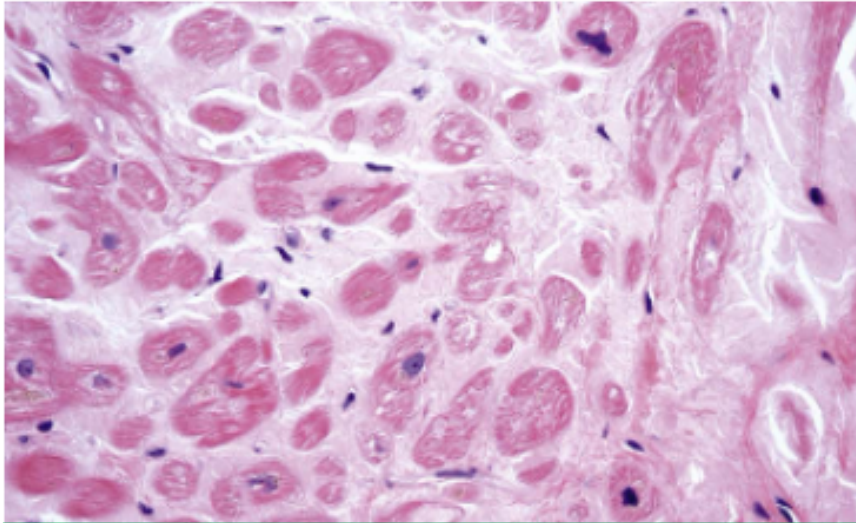
Radiation

Sarcoidosis

Amyloidosis

Prototypic

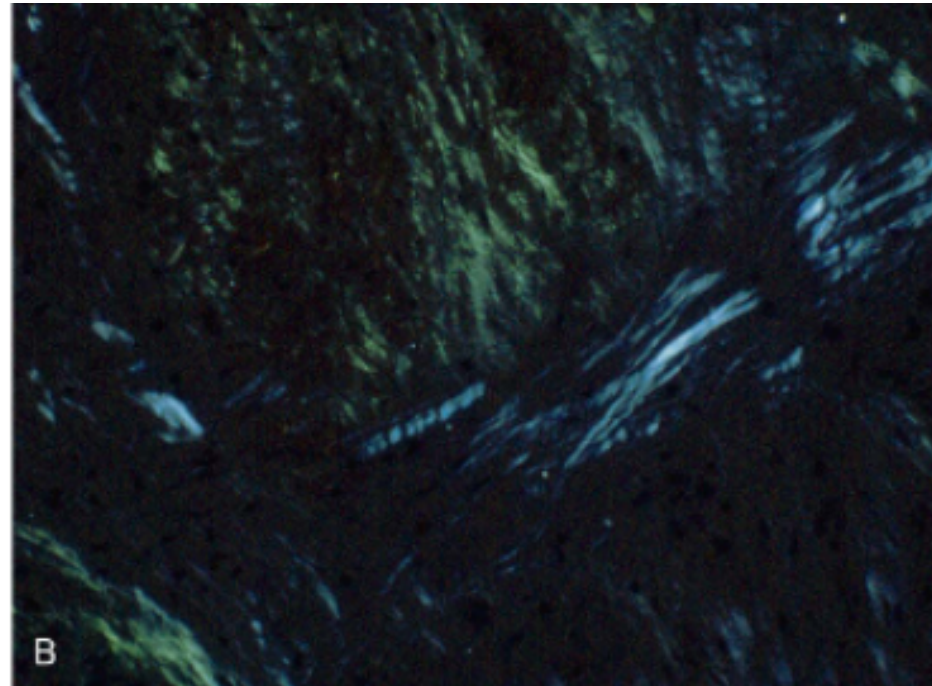




Eosinophilic deposits of amyloid in the interstitium → conduction disease

Congo Red Stain

Apple-green birefringence

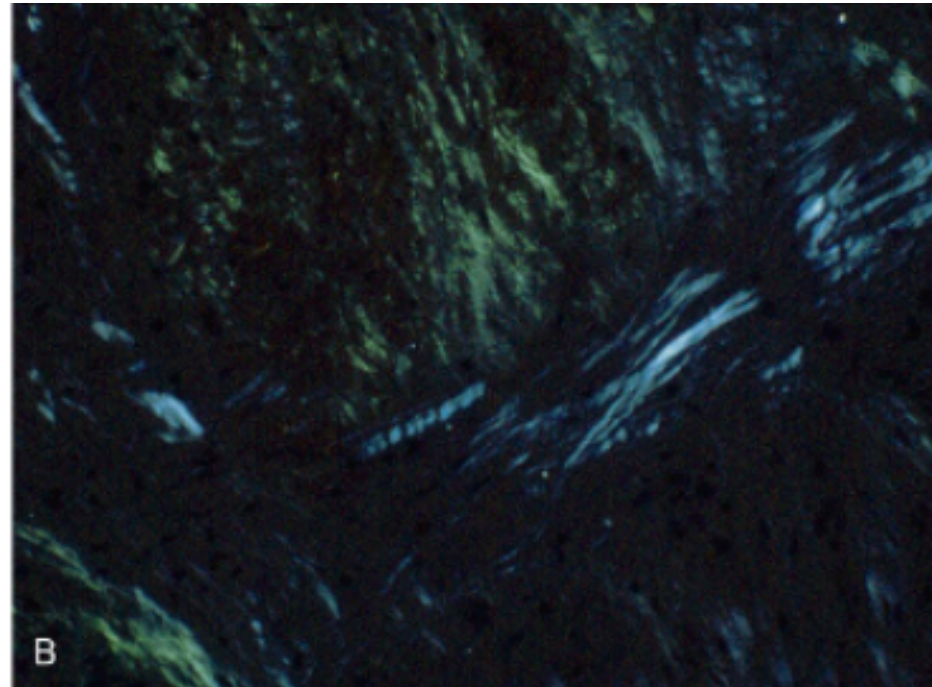


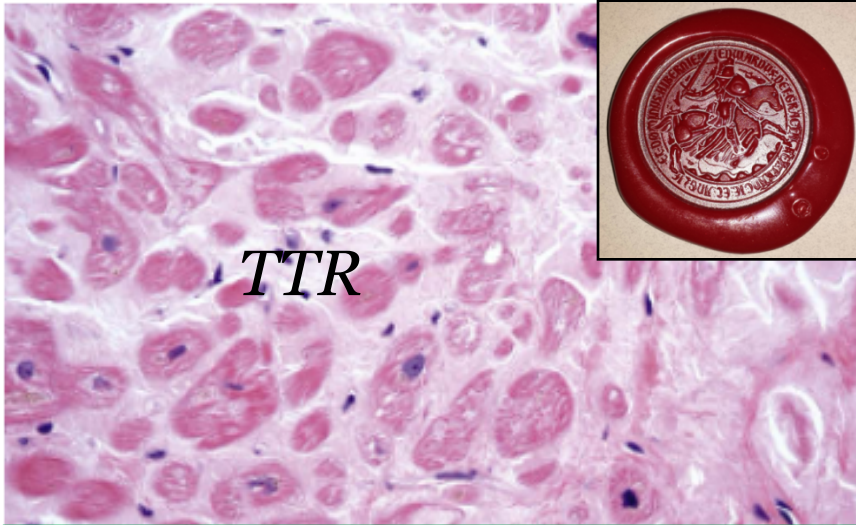


Nodules *resembling drops of wax* may be described on the *endocardial* surface

Congo Red Stain

*Apple-green
birefringence*



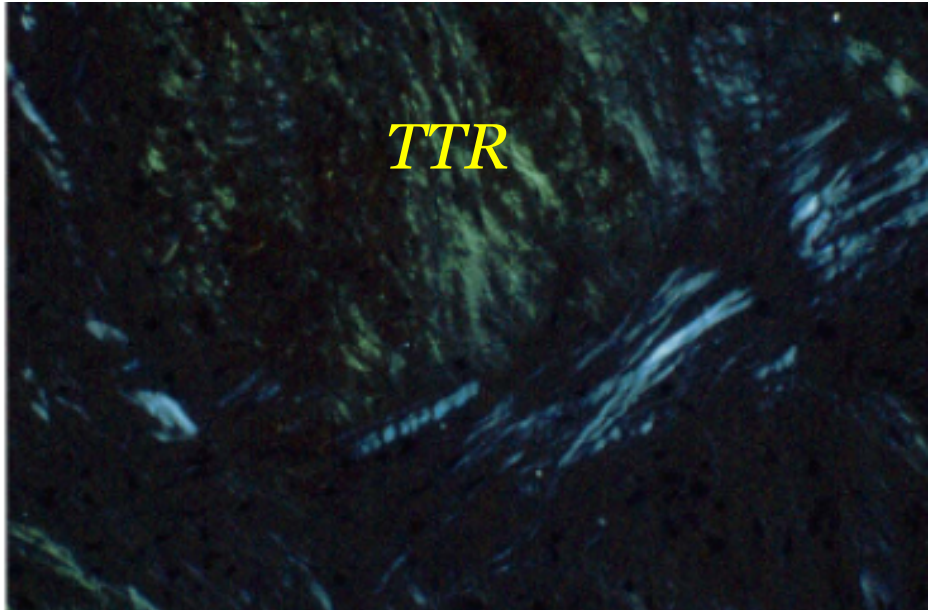


Congo Red Stain

*Apple-green
birefringence*

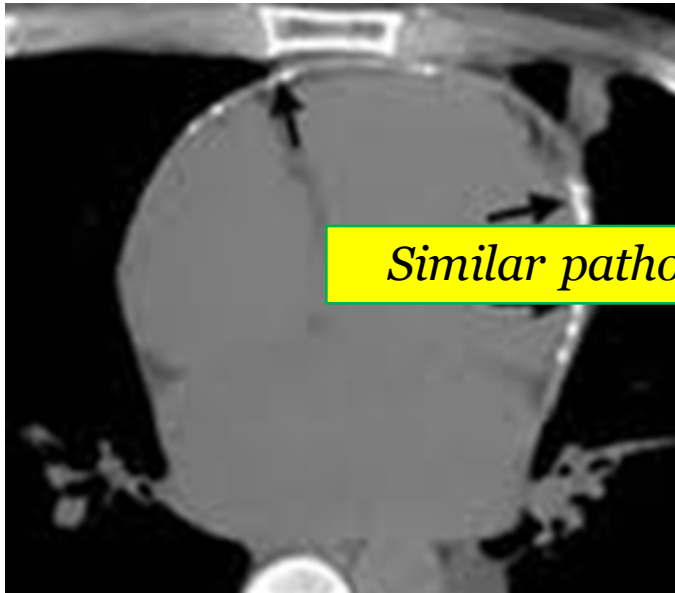
Eosinophilic deposits of amyloid in the interstitium → conduction disease

*Transthyretin
(or ANP)*



Amyloid: Pleated Sheets of *Protein*

Constrictive Pericarditis

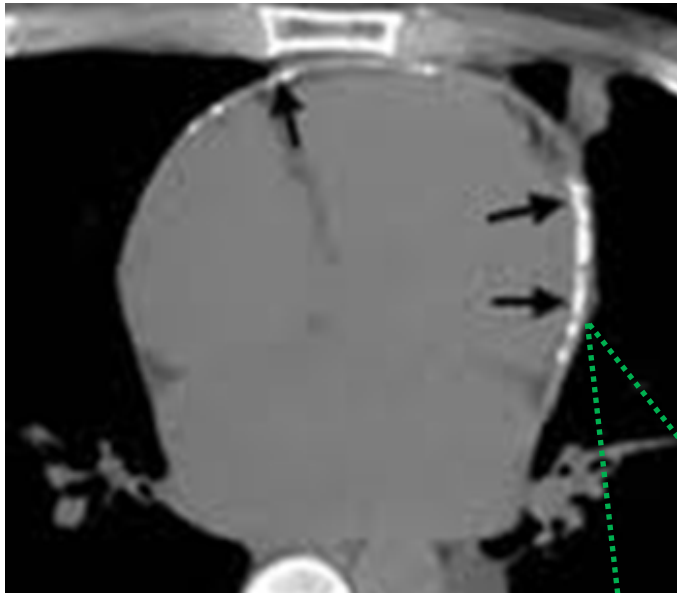


Restrictive Heart Disease

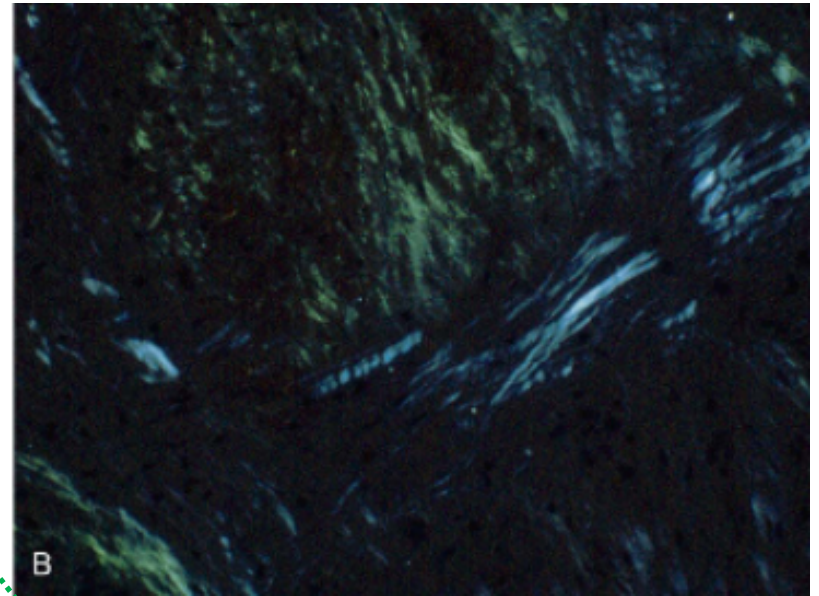


Similar pathophysiologic features

Constrictive Pericarditis

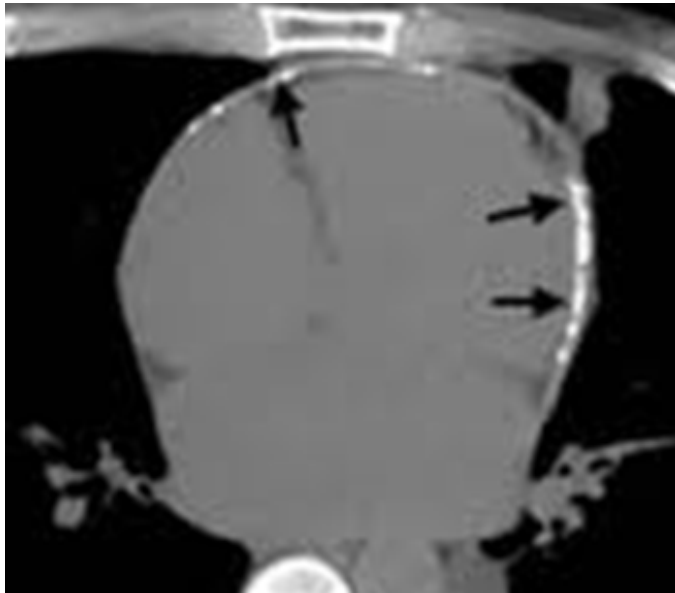


Restrictive Heart Disease

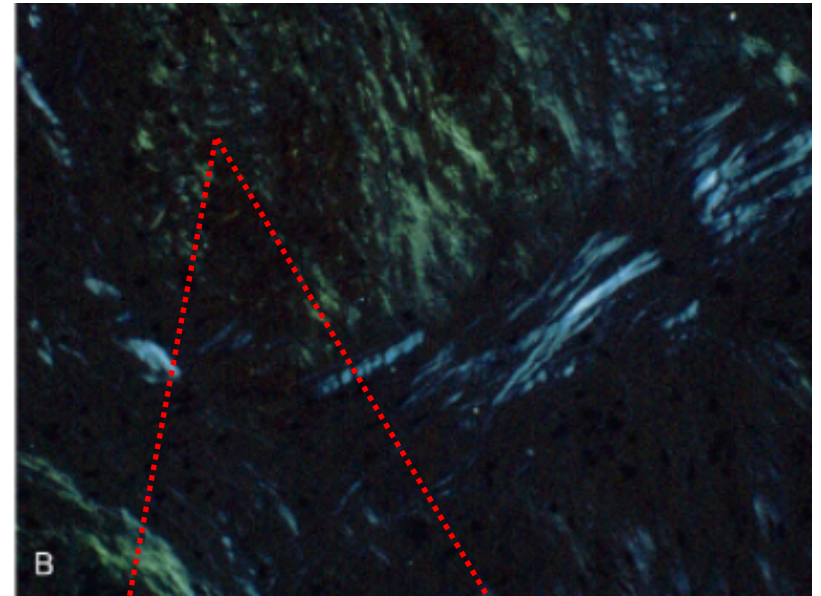


- PE: Pericardial knock, Kussmaul's sign
- Demographic: Radiation therapy '...30 yrs ago...'
- No evidence of a multisystem disorder

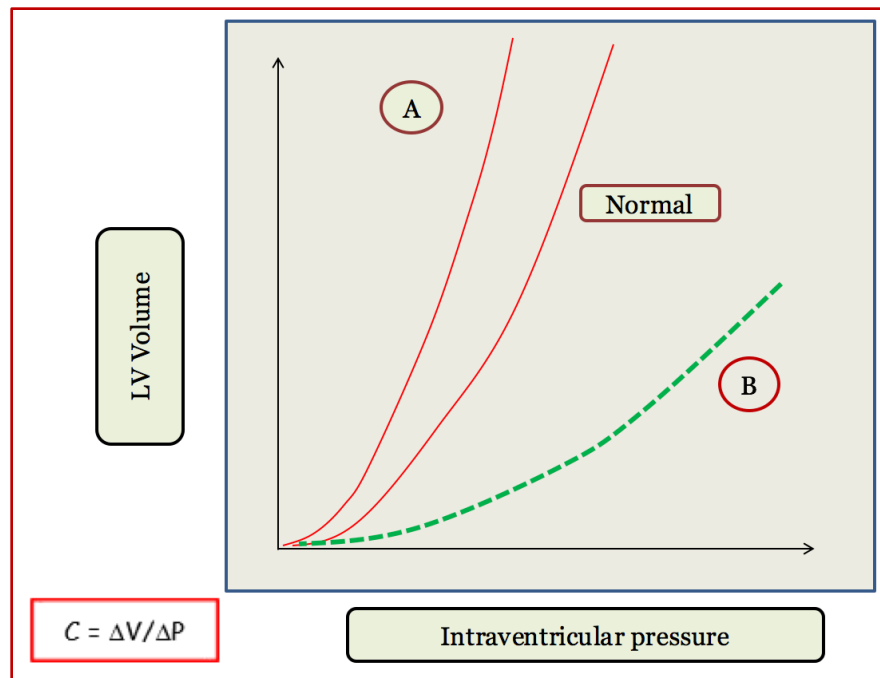
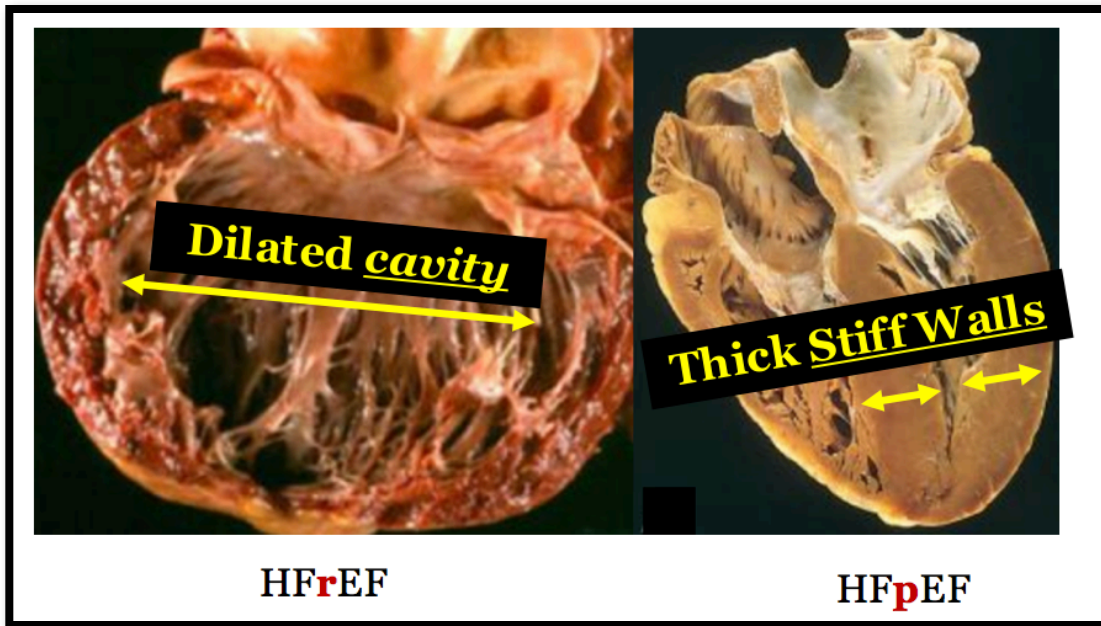
Constrictive Pericarditis

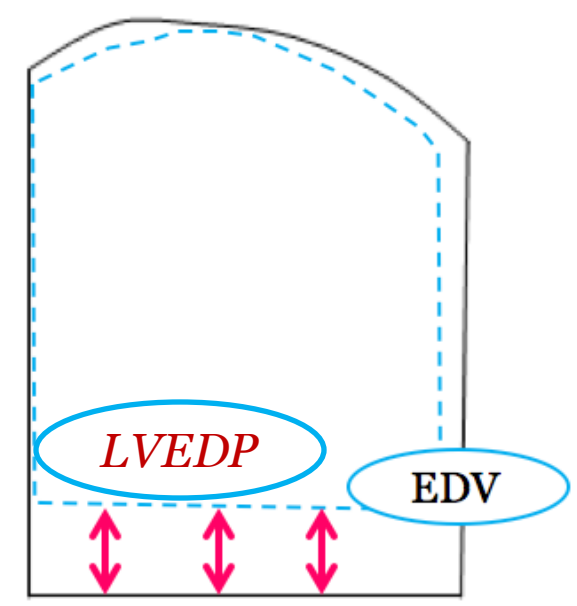
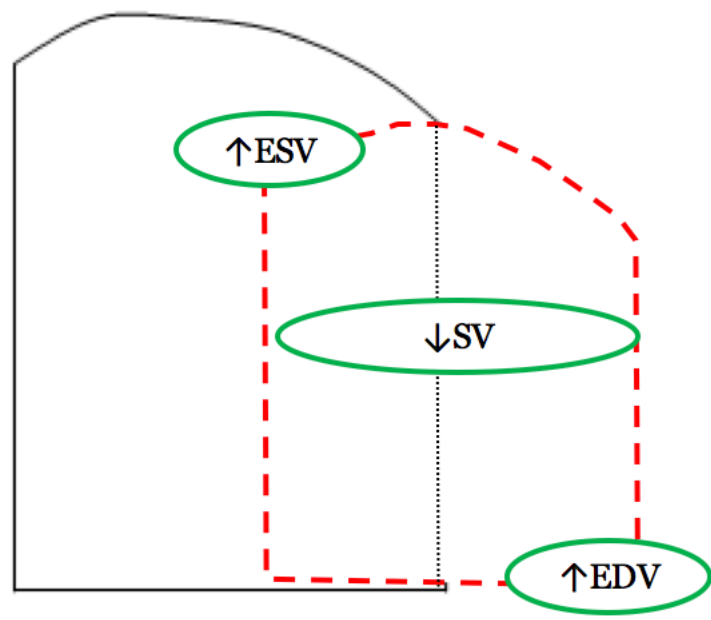
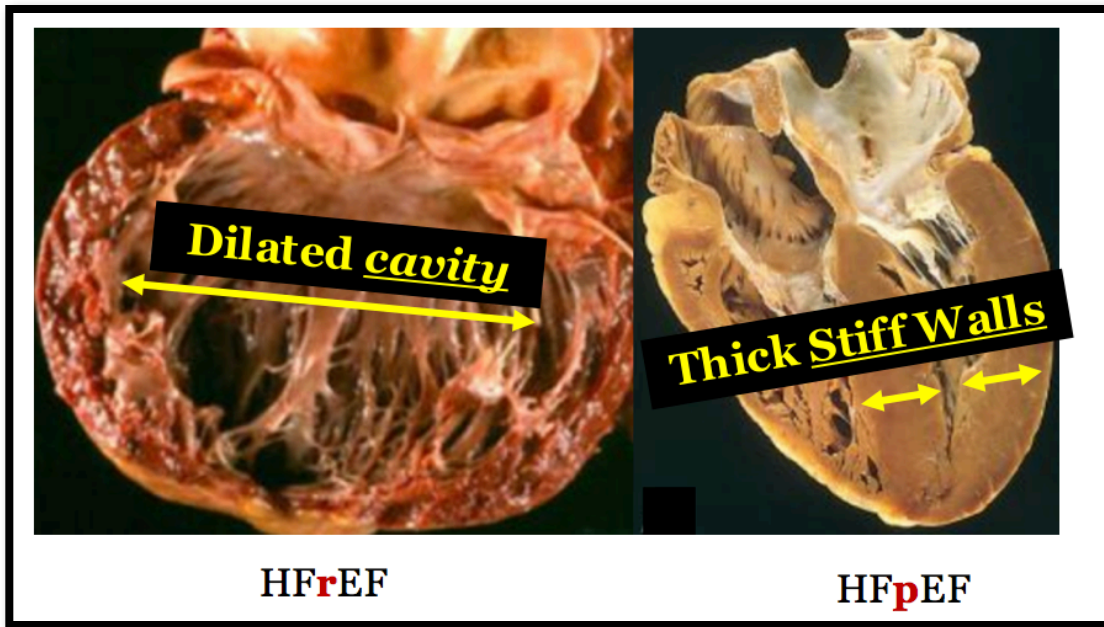


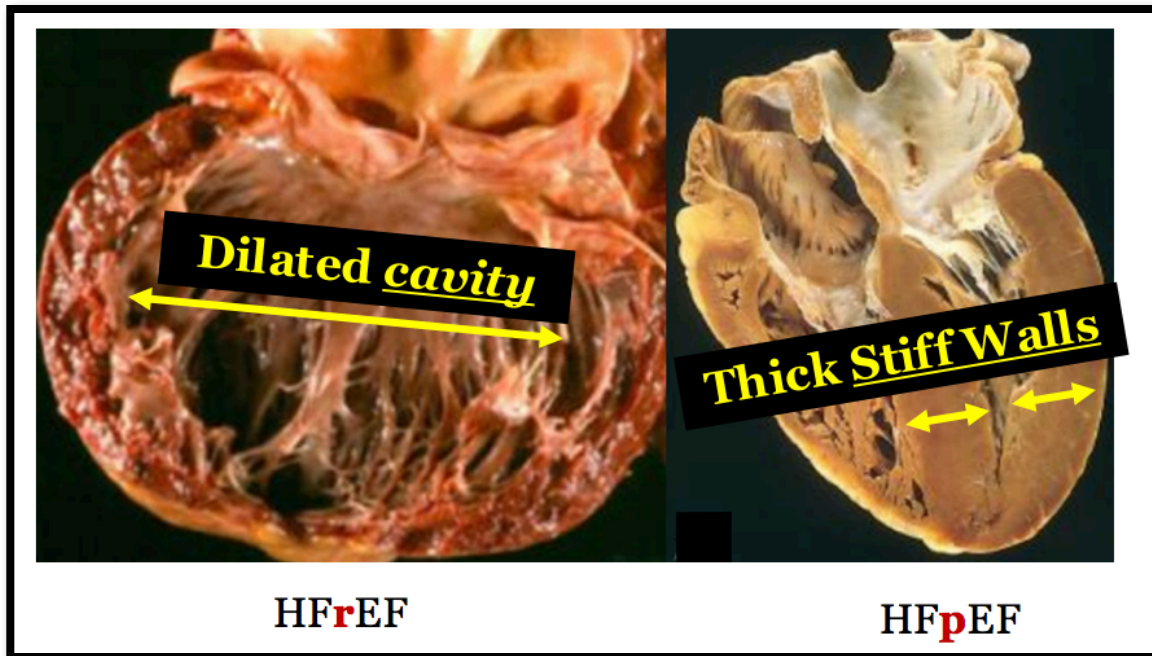
Restrictive Heart Disease



- Demographic: underlying cause for amyloid
- Exam: JVD, Kussmaul's possible
- Note: the stem usually describes a multisystem disorder (not limited cardiac amyloid)







| | DCM |
|----------------|----------------------|
| <i>EDV/ESV</i> | Increased |
| <i>EF</i> | Decreased |
| <i>SV/CO</i> | Decreased |
| <i>TPR/VR</i> | Increased |
| <i>Sounds</i> | S₃ |

| | RCM/Diastolic Dysfunction |
|---------------|---------------------------|
| <i>EDV</i> | Normal |
| <i>EDP</i> | Increased |
| <i>EF</i> | Normal |
| <i>Sounds</i> | S₄ |

Heart Failure and the Cardiomyopathies for the USMLE Step One Exam:
Final Edition: the Cardiomyopathies and Test Derivatives



HF^rEF

HF^pEF

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