



Constriction/ Restriction

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Differentiation of Constriction vs. Restriction

- **Why is it important?**
- Important therapeutic implications
 - Pericardiectomy
 - Heart failure management and treatment of underlying disease



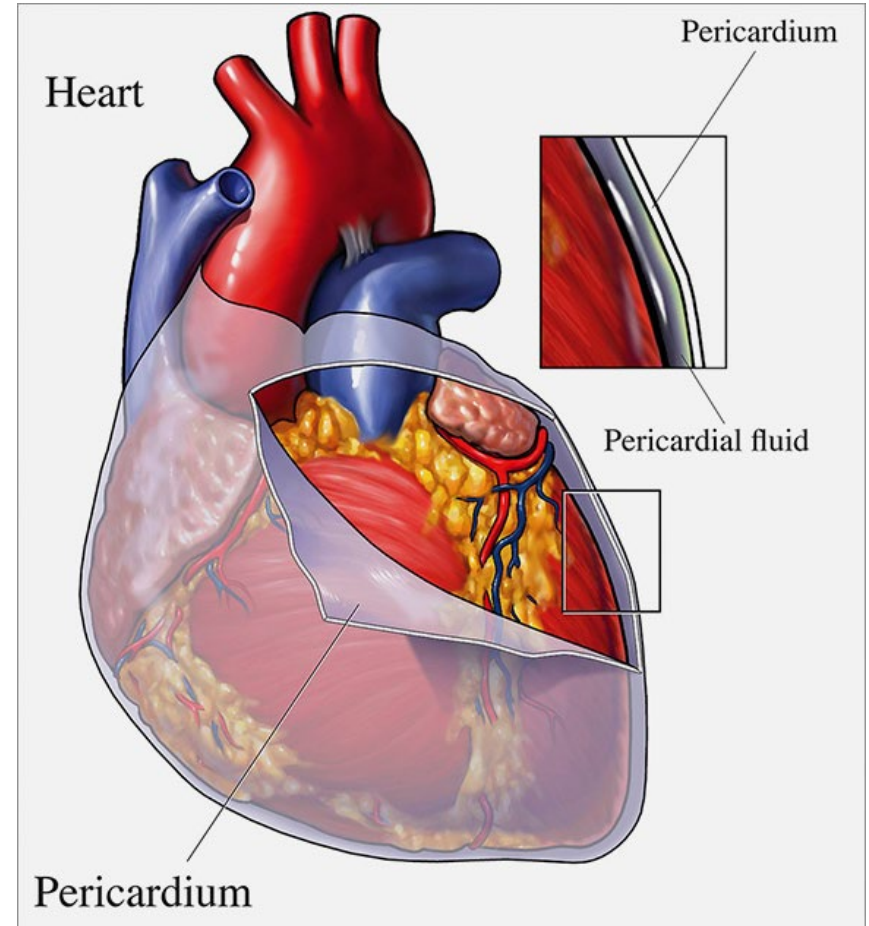
Outline

- **Normal pericardium**
- **Restriction cardiomyopathy**
- **Constrictive pericarditis**
- **Differentiating between the two processes**



Pericardium

- **Fibroelastic sac surrounding the heart**
- **Two layers**
- **Physiologic pericardial fluid <50 mL**





Normal Pericardium

Inspiration



Intrathoracic pressure decreases



Increase in venous return to the right heart



**Transient increase in RV size
(No impairment of LV filling)**



Normal Pericardium

Expire



Intrathoracic pressure increases



**Increase in pulmonary venous return to the
left atrium**



LV filling



Restrictive Cardiomyopathy



Restrictive Cardiomyopathy

- Disease of the myocardium
- Predominant diastolic, rather than systolic, dysfunction
- Pulmonary systolic pressure usually is moderately to severely elevated



Clinical Manifestations of Restrictive Cardiomyopathies

- Exercise intolerance
 - Impaired ability to augment cardiac output with tachycardia because diastolic restriction of filling
- Peripheral edema, hepatomegaly, ascites, anasarca
- Highly prone to developing atrial fibrillation



Restrictive Cardiomyopathies

Noninfiltrative

- Idiopathic
- Hypertrophic
- Radiation
- Eosinophilic

Infiltrative

- Amyloidosis
- Sarcoidosis
- Gaucher disease
- Hurler disease

Storage Disease

- Hemochromatosis
- Fabry Disease
- Glycogen storage disease



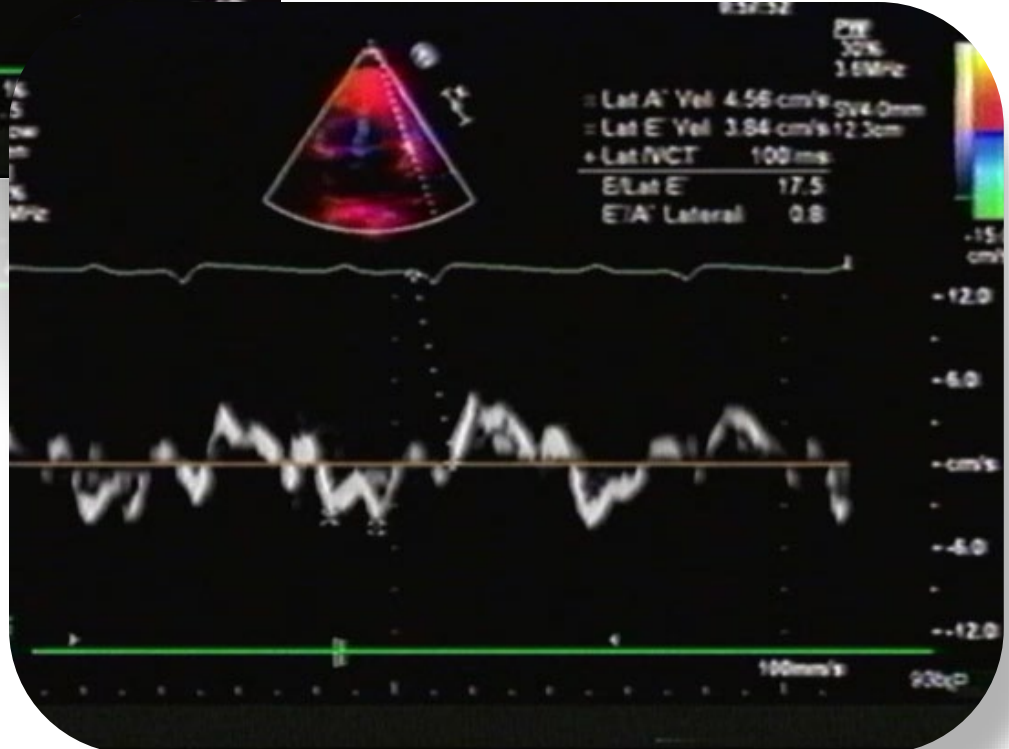
Typical Echocardiographic Features

- Non-dilated thick-walled LV
- Abnormal diastolic function
- RV free wall thickening
- Bi-atrial enlargement
- Elevation of pulmonary pressures
- Elevation of RA pressures



Doppler Features of Restrictive Cardiomyopathy

- Mitral inflow E/A ratio > 2.5
- DT of E velocity < 150 msec,
- IVRT < 50 msec
- Decreased septal and lateral e' velocities (3–4 cm/sec)
 - Lateral $>$ septal e' velocity
- E/e' ratio > 14
- Inc LA volume index > 50 mL/m²

$$E/E' = 17$$




Amyloid Heart Disease



Amyloid Heart Disease

- Disorder of protein metabolism which results in protein deposition in organs and tissues
- Amyloid deposition begins in sub-endocardium and extends within the myocardium between the muscle fibers

MAIN TYPES OF AMYLOIDOSIS

ISOLATED DEPOSITS

TYPE	SOURCE of AMYLOID	ORGANS INVOLVED
AL (Primary) Amyloidosis Amyloid Light-Chain	Bone Marrow (Light chains produced by plasma cells)	Kidneys, Heart, Liver, GI system, Nervous system
AA (Secondary) Amyloidosis Amyloid A Protein	Circulating inflammatory protein (Serum amyloid A)	Kidneys, Liver
TTR (Familial) Amyloidosis Mutant Transthyretin	Unstable, mutant transthyretin produced in the liver	Nervous system, Heart
SSA (Senile systemic) Amyloidosis Seniors	Wild-type (normal) transthyretin	Heart



Echocardiographic Features of Amyloid Infiltration

- 
- Increased LV wall thickness (“ground glass”)
 - Increased RV wall thickness

- 
- Small LV; normal or reduced systolic function
 - Pericardial effusion; big LA; thick atrial septum
 - Valve thickening; mild regurgitation

- 
- E/A ratio >2
 - Deceleration time <150 ms
 - PV: small S wave, large D wave; S/D ratio <0.5



Case 1

76 year old man with senile cardiac amyloidosis, Afib, ICD presents with generalized weakness

47Hz

m

%

0

low

en

G

R

2.8

M3

FR 61Hz
17cm

2D
58%
C 50
P Low
HRes

G
P 2.1 R 4.2

M3

2D
58%
C 50
P Low
HRes

G
P 2.1 R 4.2

JPEG

1

7

0

m/s

M3

FR 37Hz
24cm

2D
84%
C 50
P Low
HGen

G
P 1.7 R 3.4

M3

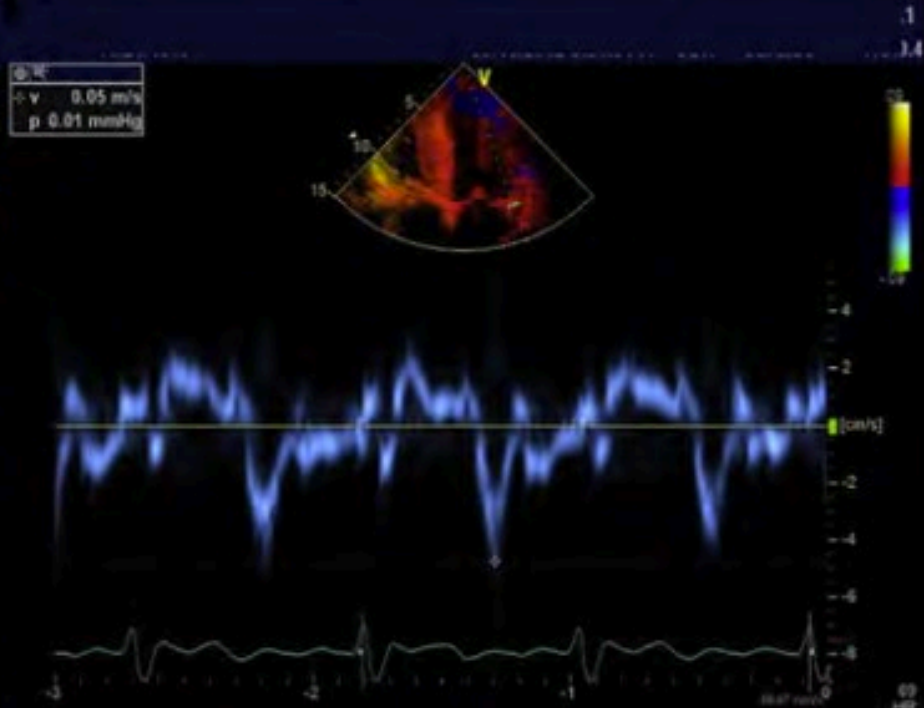
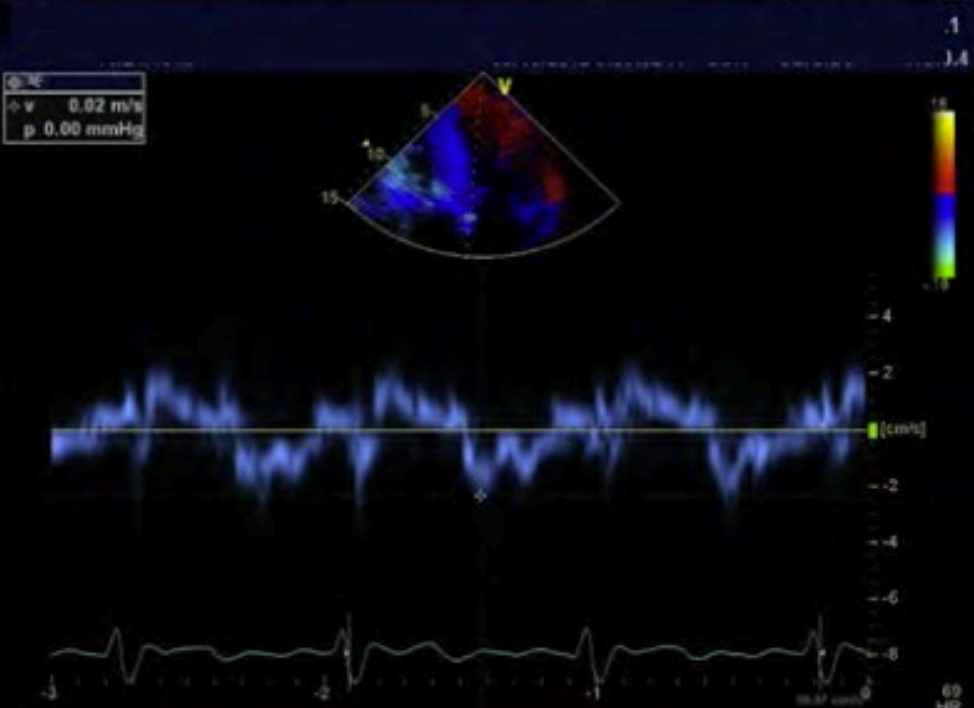
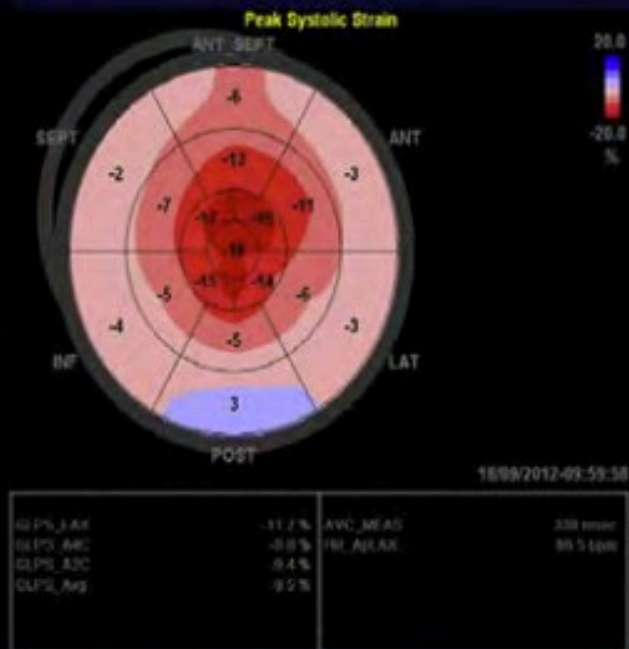
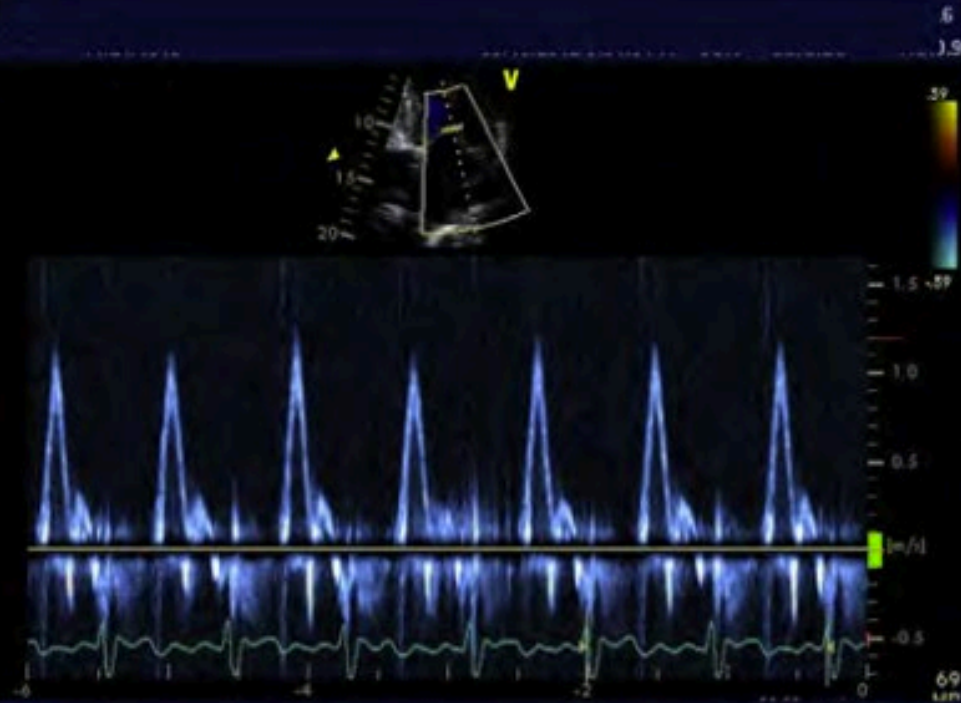
2D
84%
C 50
P Low
HGen

G
P 1.7 R 3.4

JPEG

88 bpm

94
BPM





Case 2

**35 year old man being evaluated
for heart/liver transplant for history
of hemochromatosis**

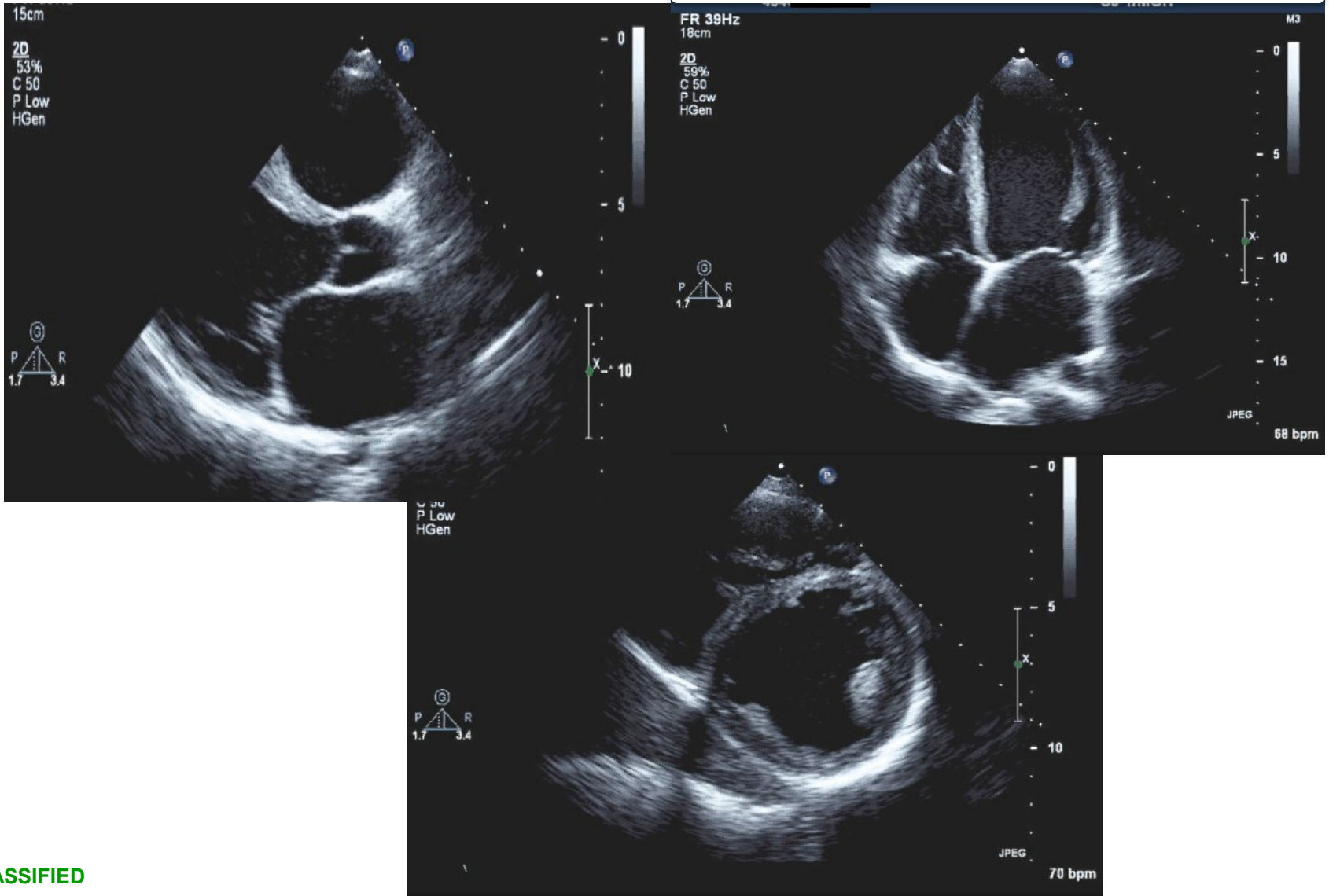


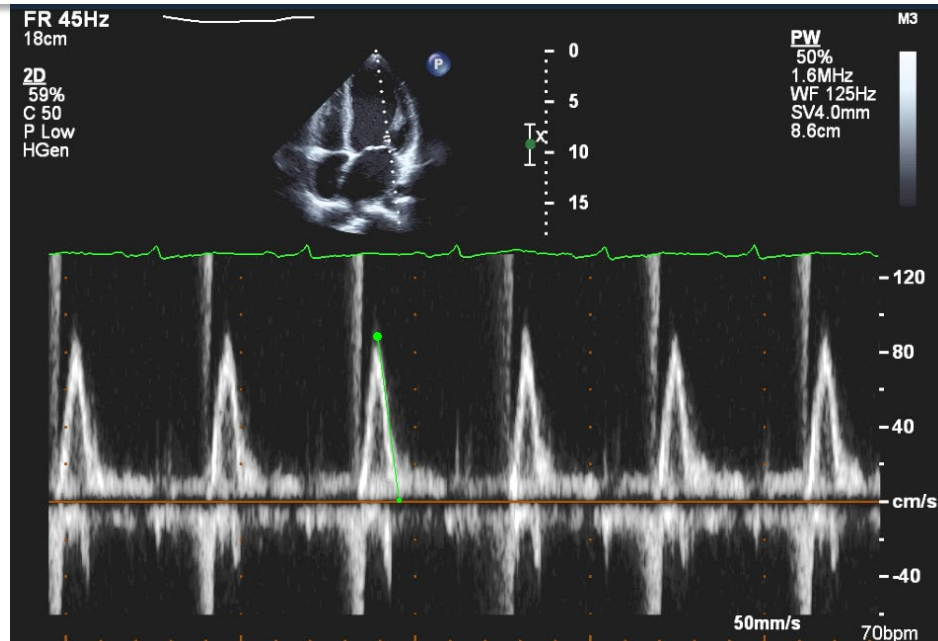
Hemochromatosis

- Initially characterized by diastolic dysfunction and arrhythmias and in later stages by dilated cardiomyopathy
- Diagnosis of iron overload is established by elevated transferrin saturation ($>55\%$) and elevated serum ferritin (>300 ng/mL)
- Genetic testing for mutations in the HFE gene

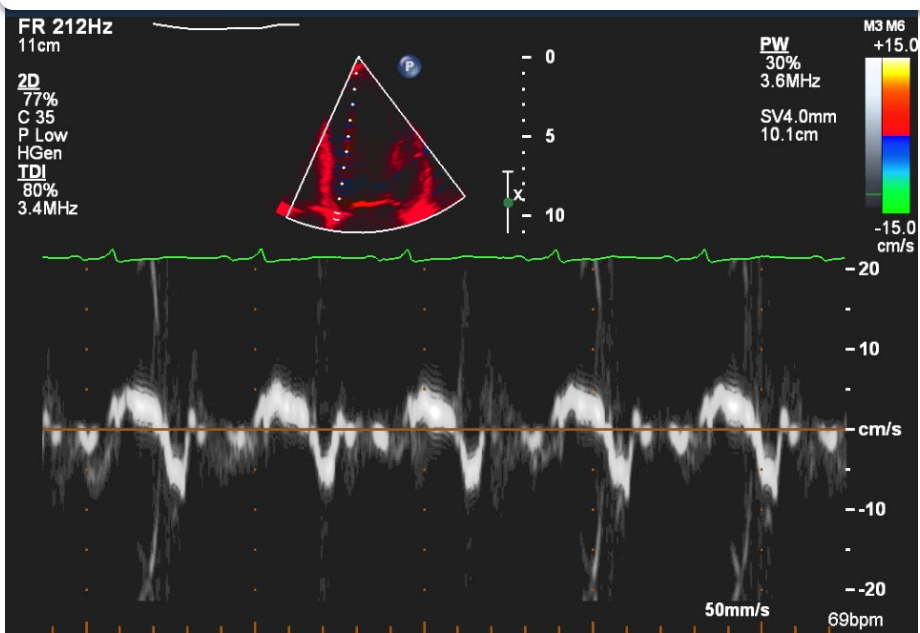


Hemochromatosis

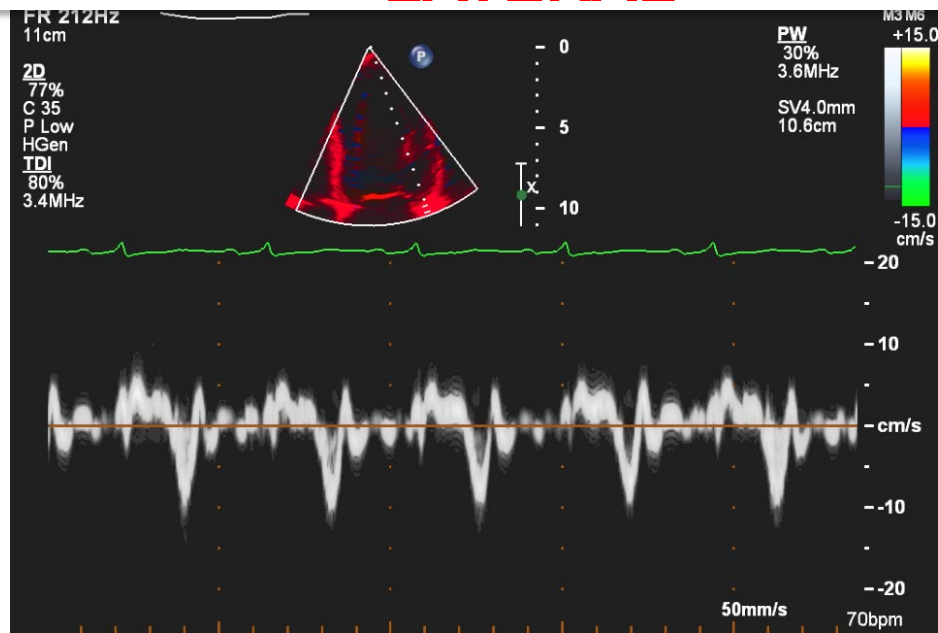


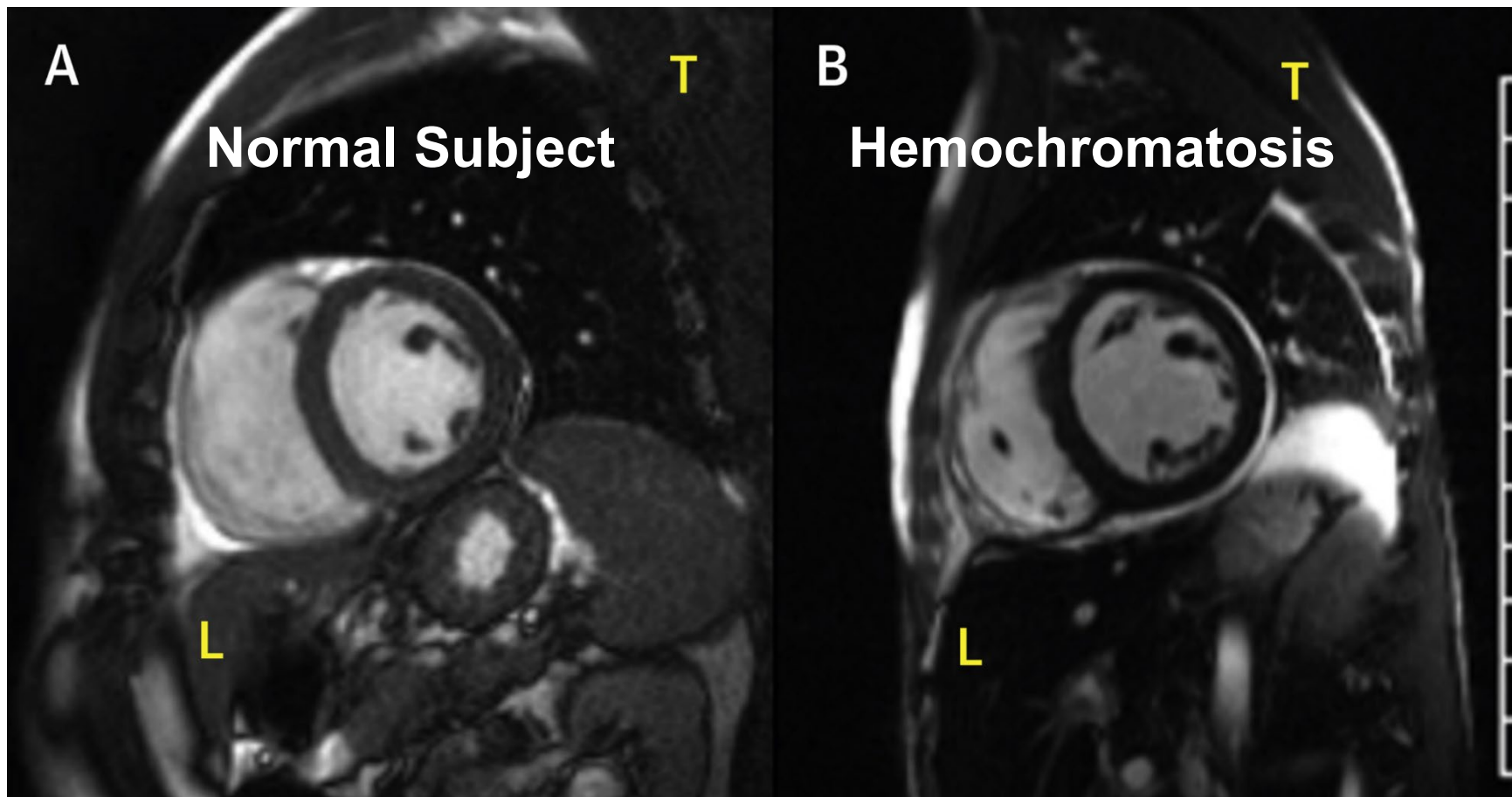


SEPTAL



LATERAL





CMR T2-Weighted Images. Both the myocardium & liver (L) show decreased signal intensity compared to trapezius (T) skeletal muscle



Case 3

**37 year old man with hypertension,
end stage renal disease on
dialysis, and Fabry's disease on
RV biopsy**



Fabry's Disease

- **X-linked, lysosomal storage disease**
- **Dysfunctional metabolism of sphingolipids**
- **Mutation in GLA gene**
 - **Makes enzyme α -galactosidase A**
 - **Build up of globotriaosylceramide**
- **Symptoms/clinical findings**
 - **Acroparesthesias**
 - **Angiokeratomas**
 - **Hypohidrosis**
 - **Corneal opacity**
 - **Tinnitus and hearing loss**
 - **Progressive kidney damage**
 - **Cardiomyopathy**
 - **Stroke**

FR 35Hz
20cm
2D
62%
C 50
P Low
HPen

G
P R
1.4 2.8



JPEG
20
83 hnn

FR 35Hz
20cm
2D
39%
C 50
P Low
HPen

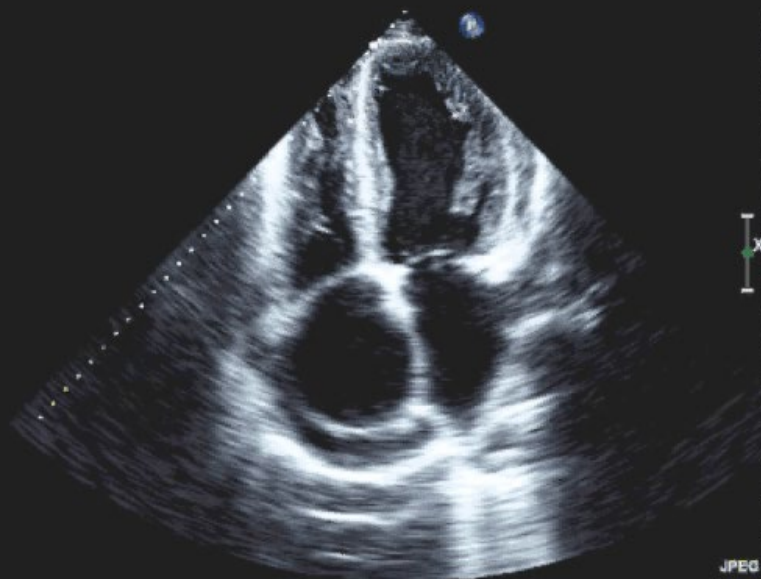
G
P R
1.5 3.0



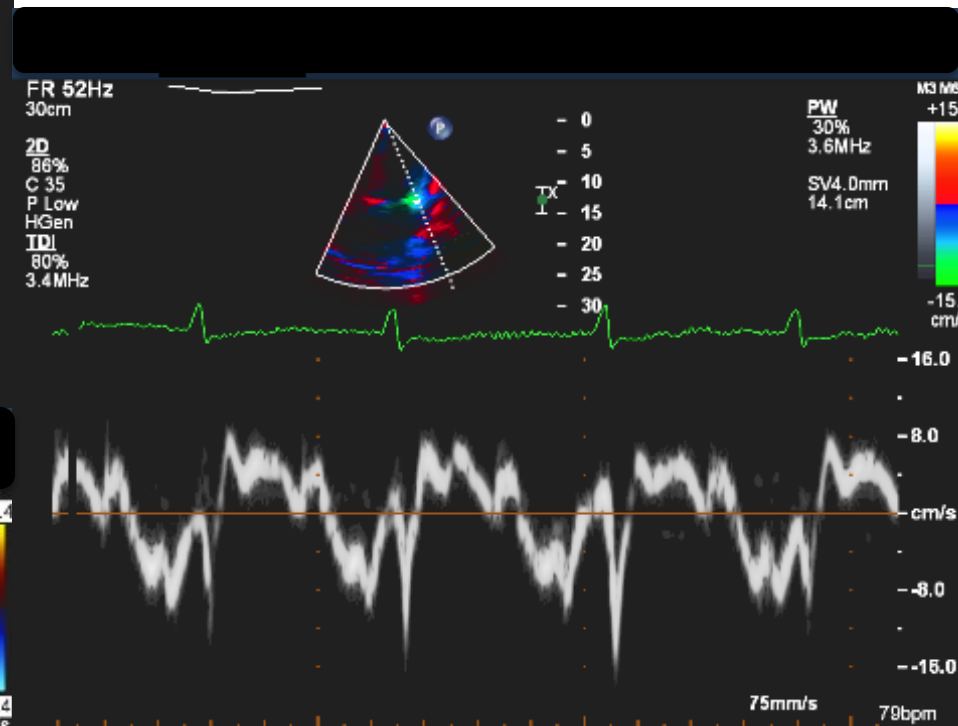
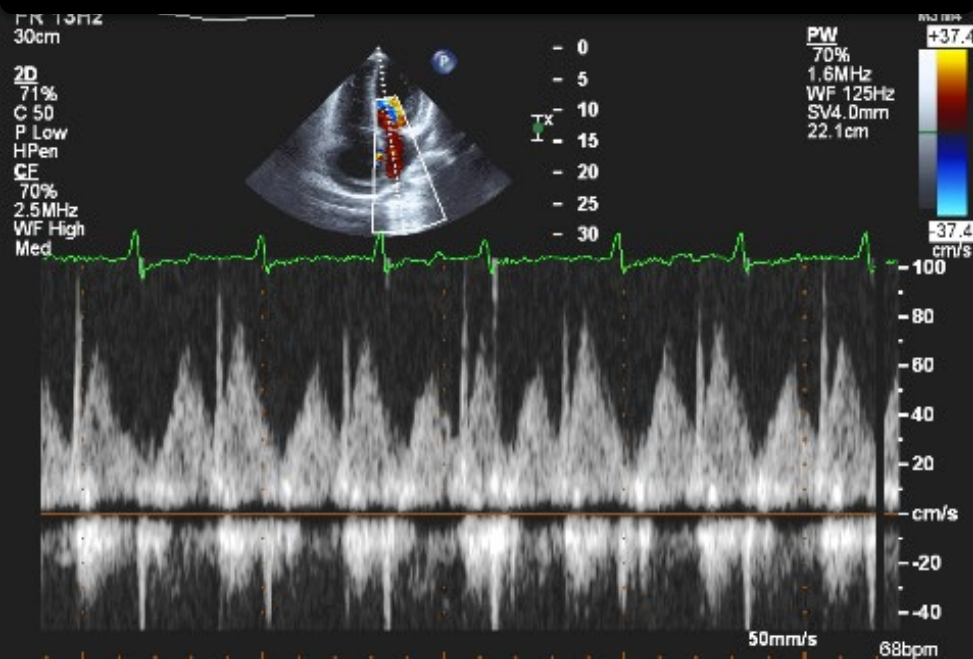
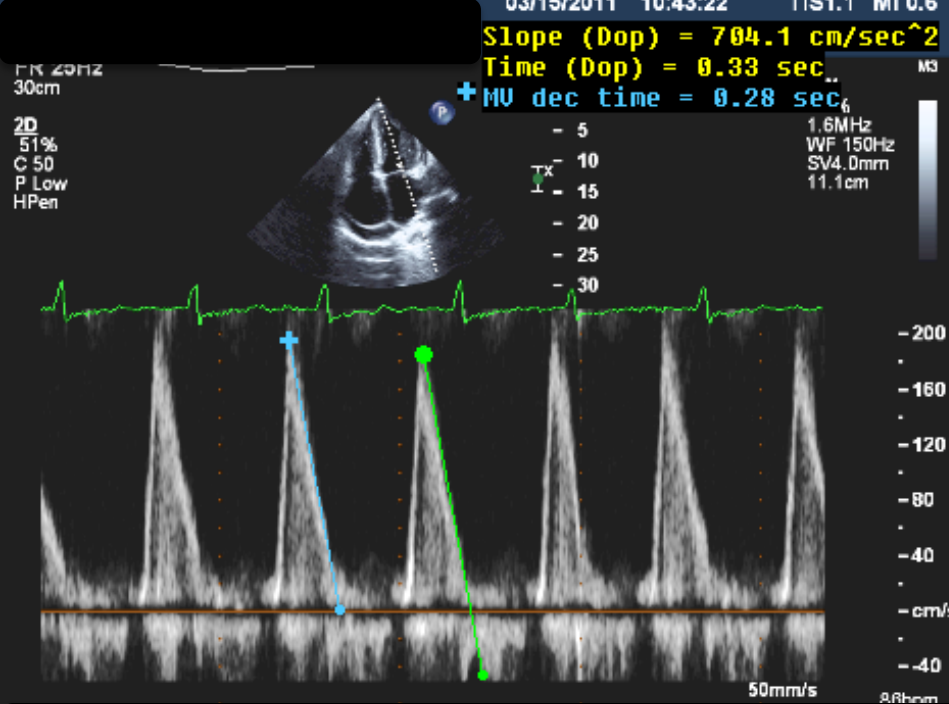
JPEG
90%

FR 25Hz
30cm
2D
54%
C 50
P Low
HPen

G
P R
1.5 3.0



JPEG
80%





Constrictive Pericarditis



Constrictive Pericarditis

- **Long-standing inflammation that leads to pericardial scarring with thickening, fibrosis, calcification**
- **Loss of normal elasticity of the pericardial sac**
- **Characteristic hemodynamic changes occur from changes in intrathoracic respiratory pressure with a fixed end-diastolic ventricular volume**



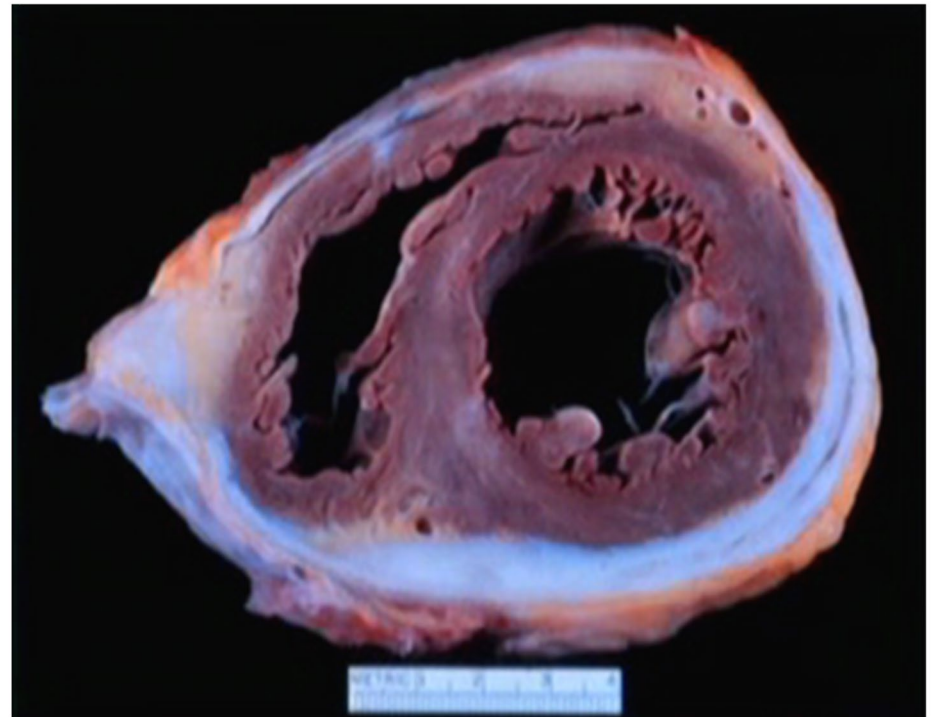
Constrictive Pericarditis

- **Etiology**

- Idiopathic
- Post-infectious: TB, viral, other
 - Recurrent pericarditis
- Cardiac surgery
- Radiation
- Trauma
- Malignancy

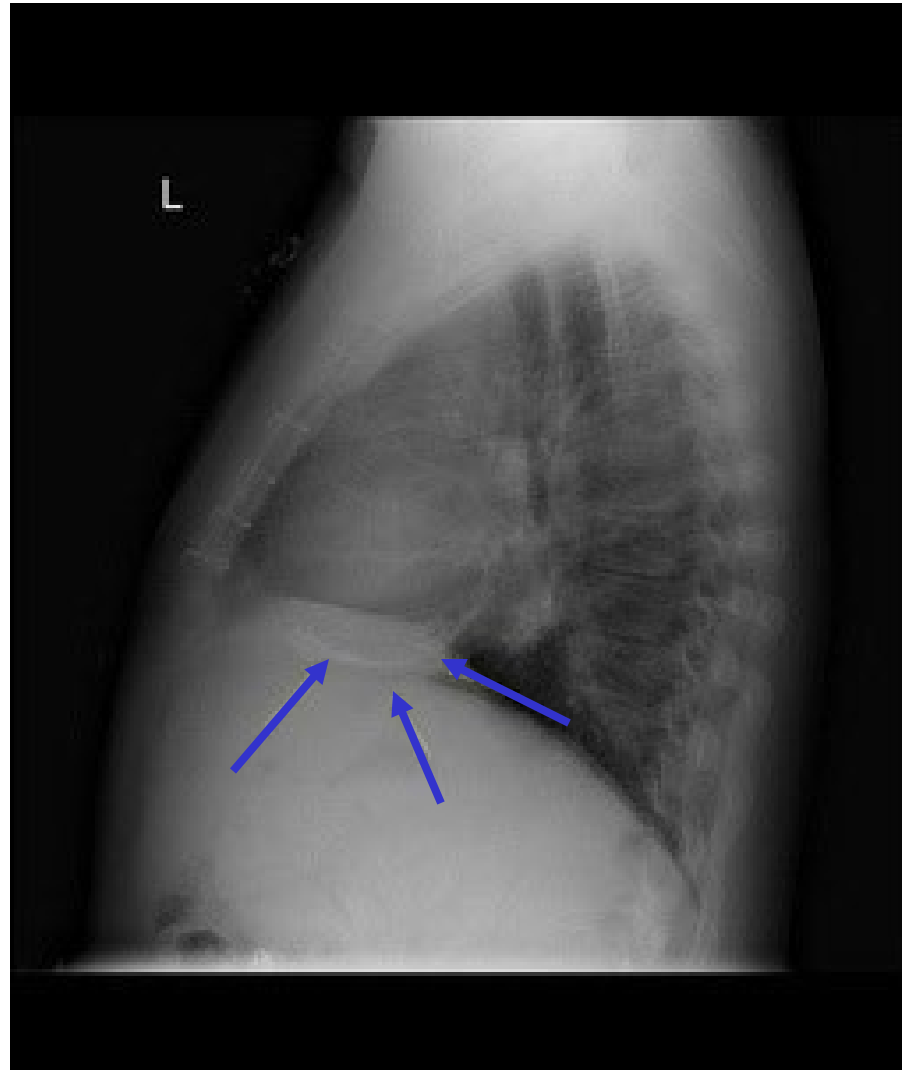
- **Signs**

- Right >> Left HF
- Elevated JVP with prominent Y descent
- Kussmal's sign
- Pericardial knock
- Edema
- Ascites





Calcified Pericardium



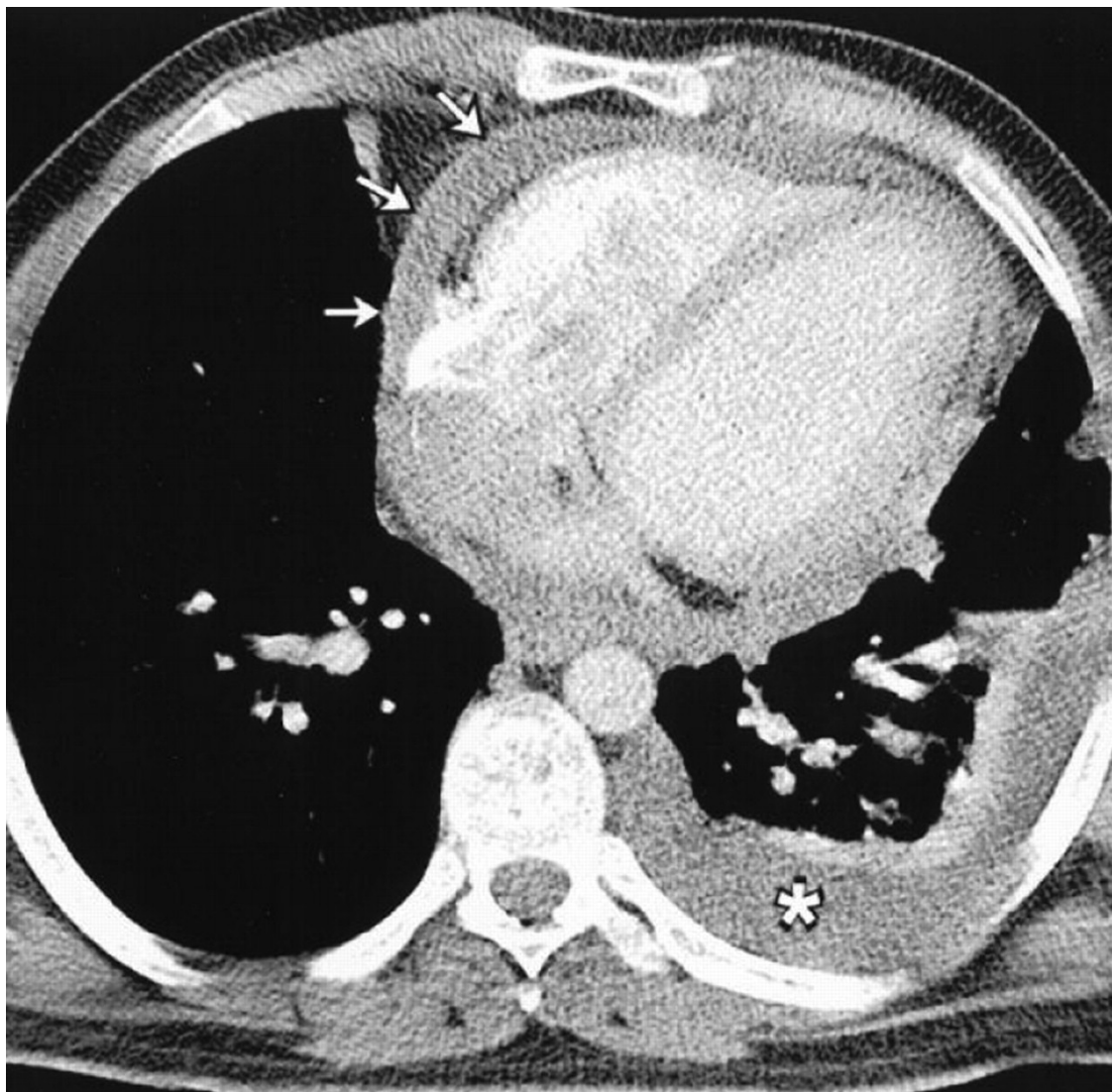
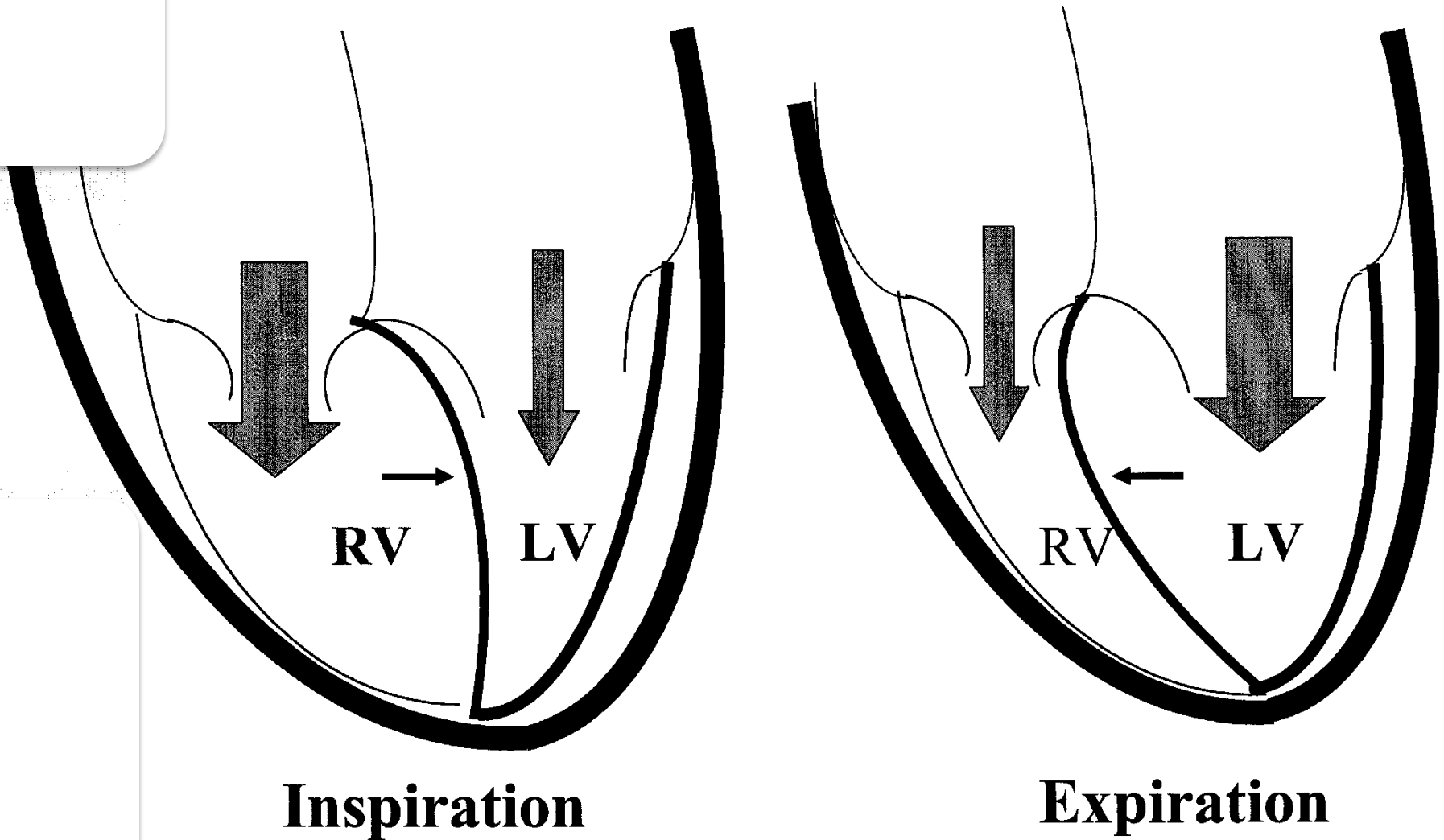


Figure 5. Chest CT from a patient with pericardial constriction showing thickened pericardium (arrows) and a left pleural effusion.

Constrictive Pericarditis



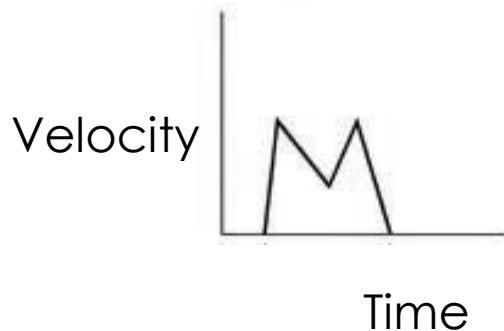
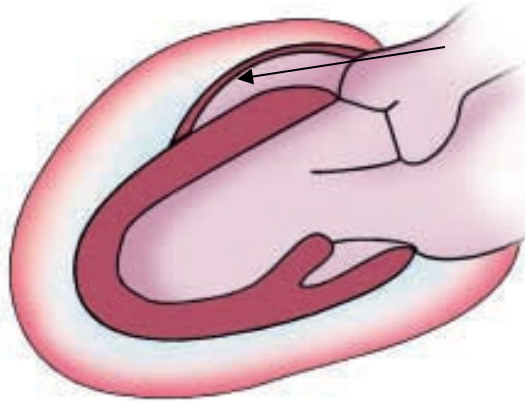


Constrictive Pericarditis: Pathophysiology

- **Marked restriction of filling**
- **Elevation and equilibration of filling pressures in all heart chambers**
 - **Early to mid diastole ventricular filling is abrupt and rapid**
 - **This filling abruptly ceases when the intracardiac limit reaches its set limit**
- **Systemic venous congestion leads to hepatic congestion, peripheral edema, ascites, and sometimes anasarca**

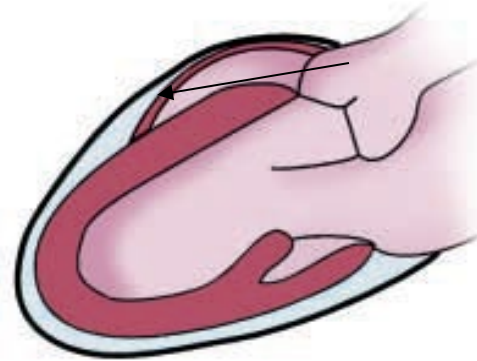
With tamponade, diastolic filling is impaired in both early and late diastole due to the elevated pericardial pressures “compressing” the heart.

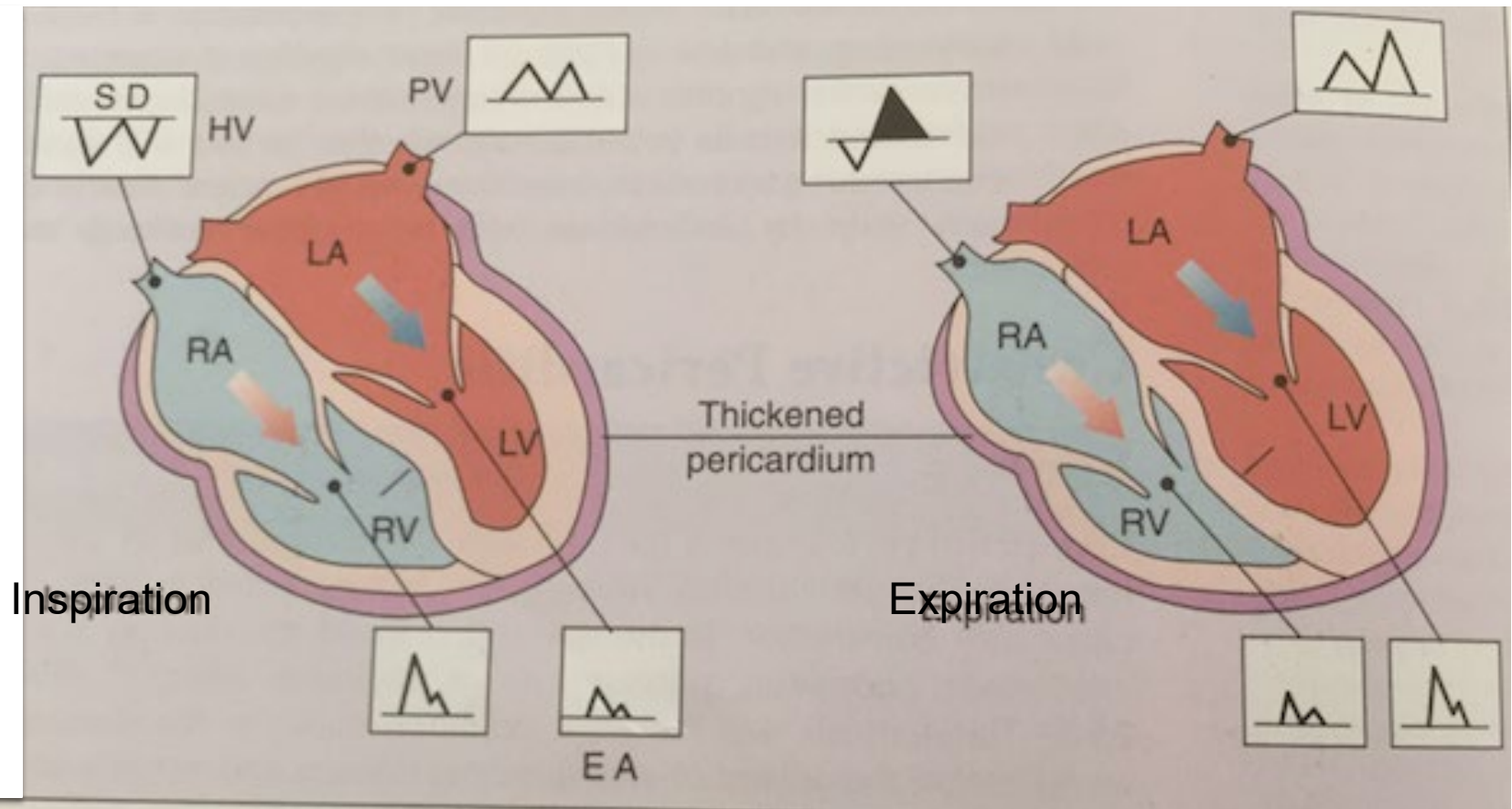
TAMPONADE



With constriction, early diastolic filling is rapid but ends abruptly when the volume limits of the rigid pericardial space are reached

CONSTRICTION







Constriction: Hemodynamics

- **RA: elevated pressure with “M” or “W” pattern**
- **RV: “dip and plateau” or “square root” sign**
- **Diastolic equalization of LV and RV pressures**
 - Volume loading
- **Systolic **discordance** of LV and RV pressures**

Tamponade



Constriction

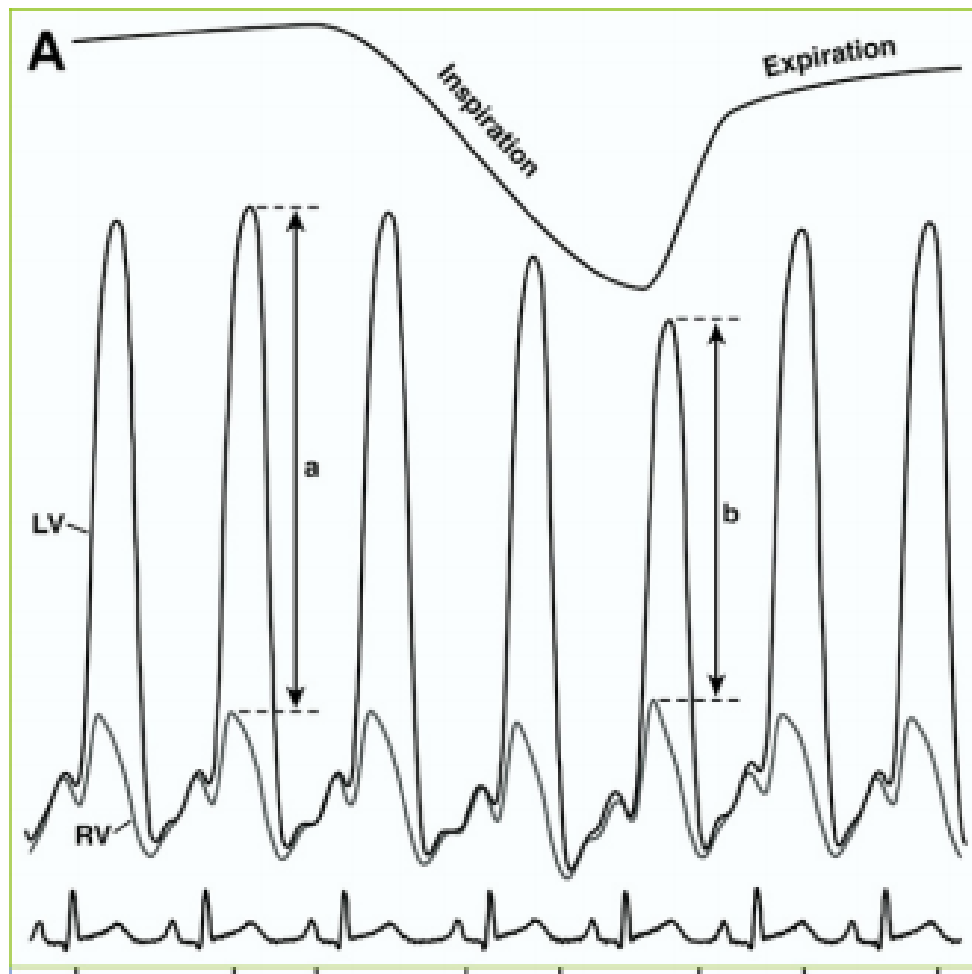




Constriction: Diastolic Equalization



“Dip and plateau”





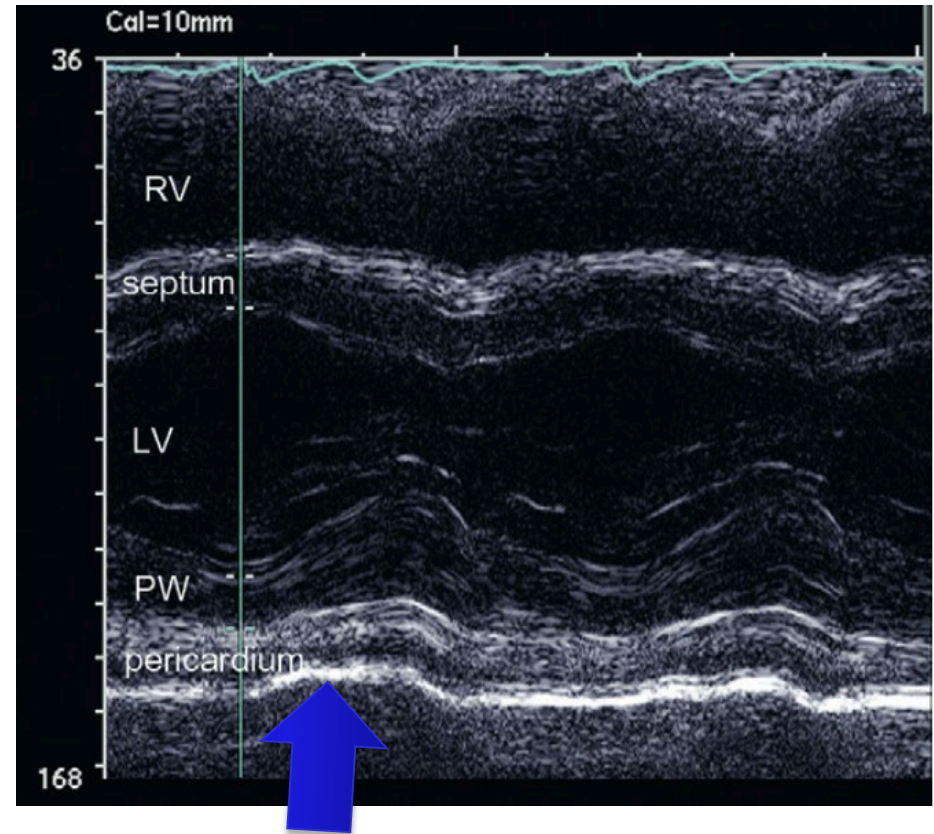
Echo findings in Constriction

- LV size and systolic function typically normal
- M-mode may show persistent pericardial thickening with low-gain settings
- >25% inc in MV inflow velocities seen with expiration
- Septal bounce
- IVC and hepatic vein plethora
 - Diastolic HV flow reversal
- Mitral E wave velocity usually < 160 msec
- Typically normal PASP



M-mode

Thick pericardium
that persists at low
gain settings





Doppler Findings in Constriction

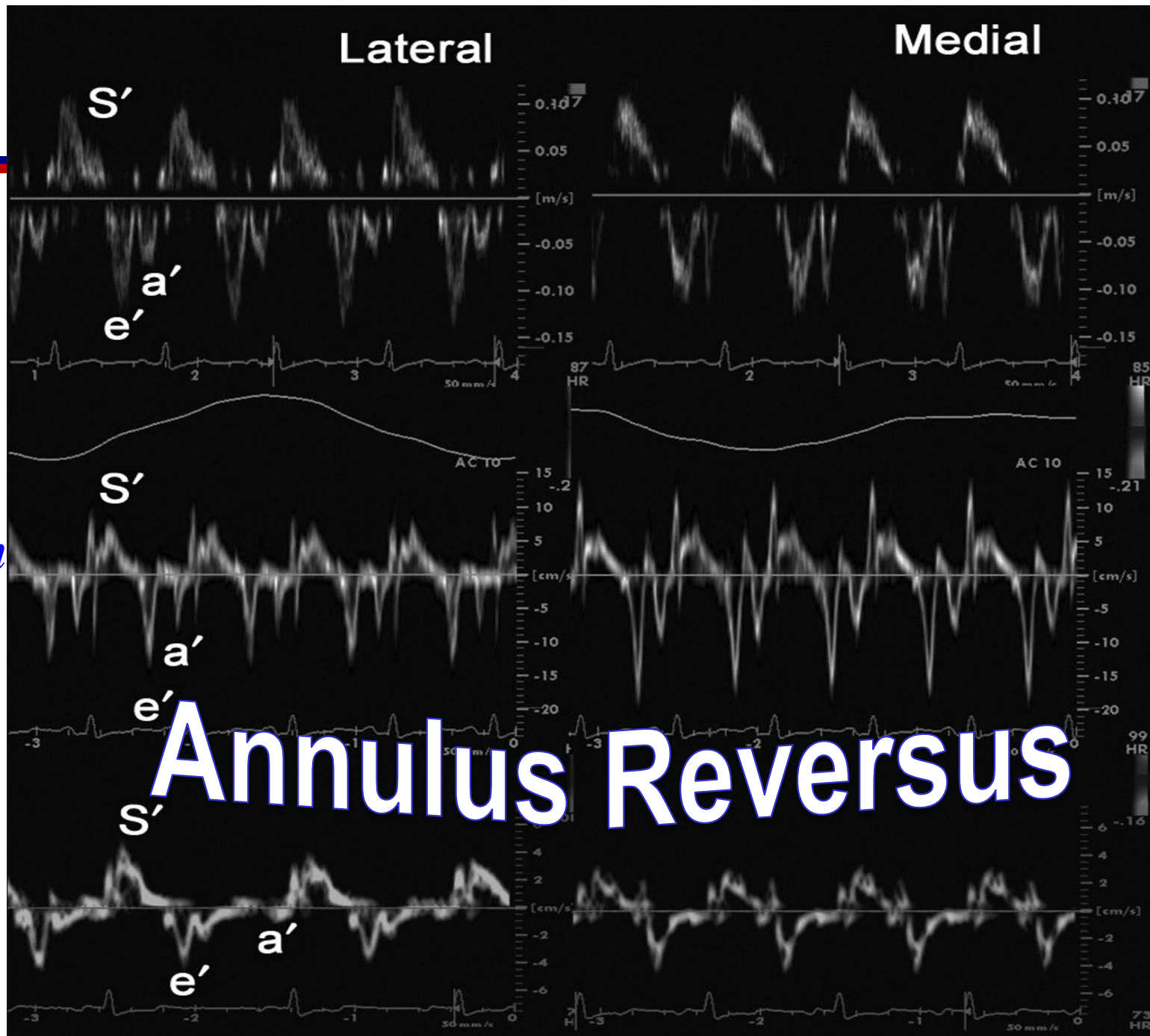
- Abnormal passive filling of the ventricles during early diastole → High E velocity
- Tissue Doppler: annulus reversus
- Respiratory variation in ventricular filling
 - Inspiration
 - MV inflow: decreases $\geq 25\%$
 - TV inflow: increases $\geq 40\%$
- Hepatic venous flow reversal with expiration



Normal

Constriction

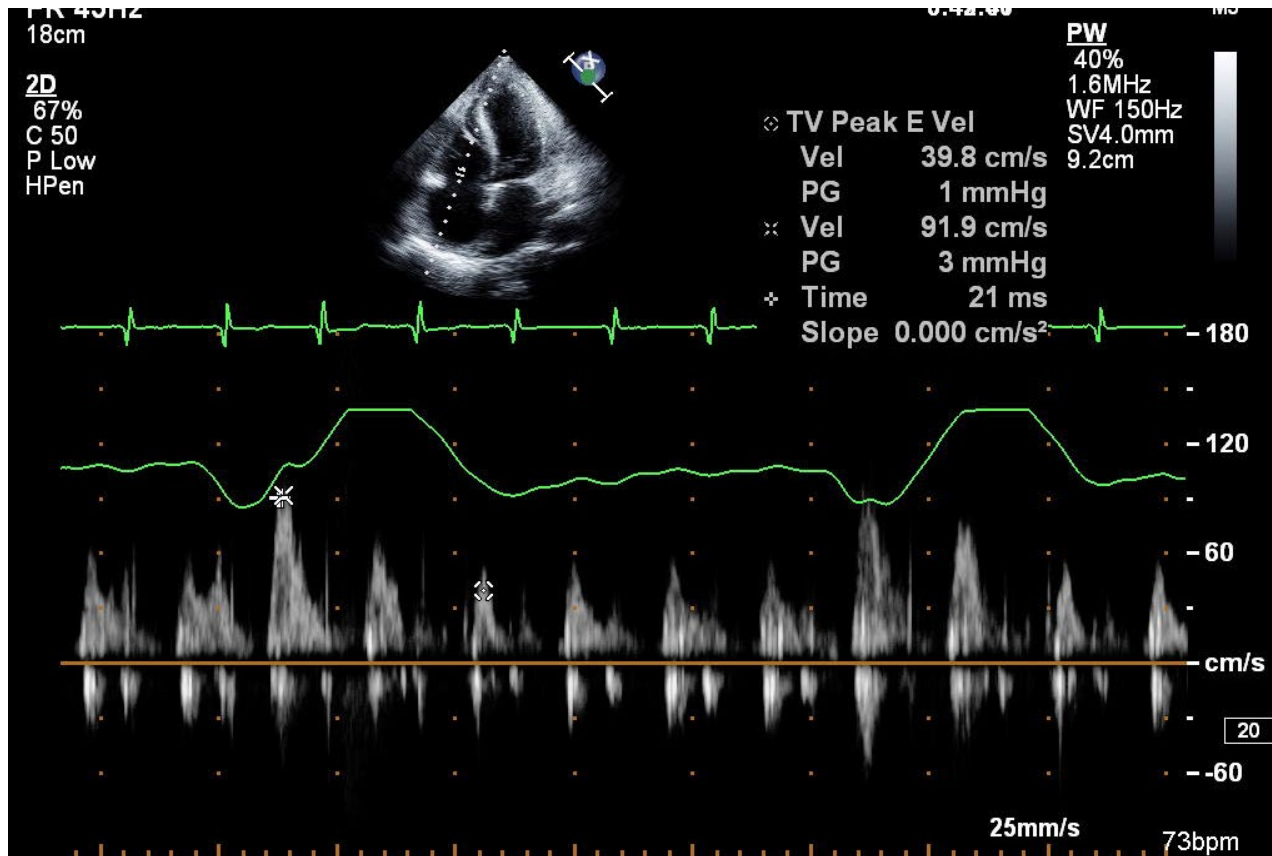
Restriction





Tricuspid Valve In-Flow

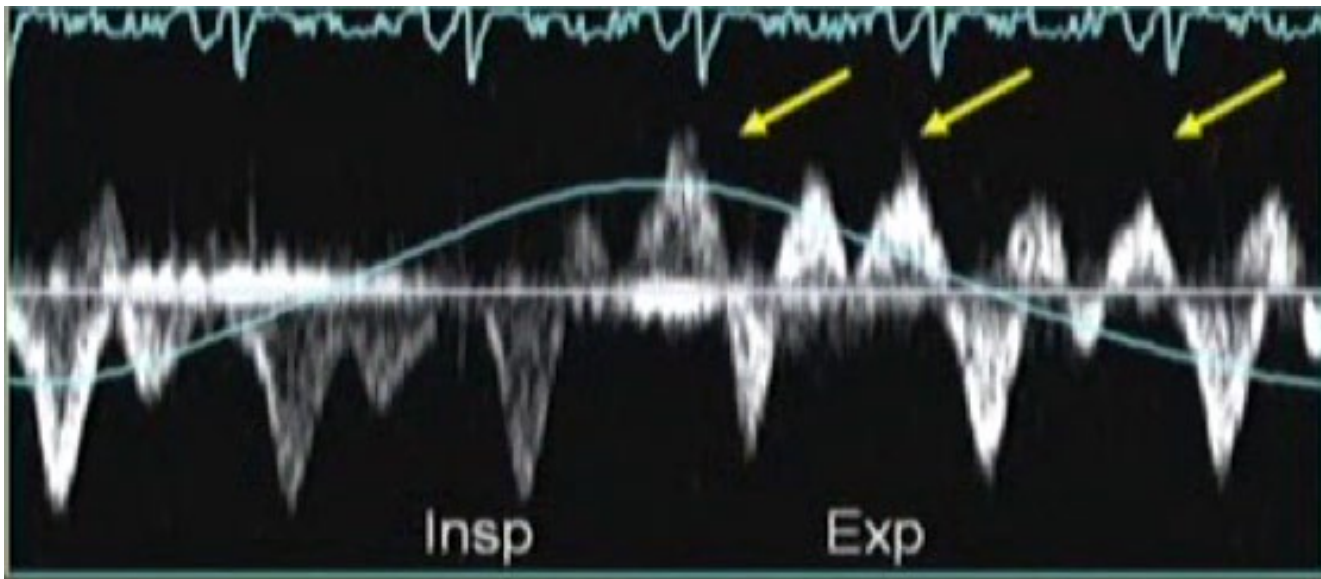
- Tricuspid inflow
 - E wave increase $\geq 40\%$ with inspiration





Doppler Findings: Constrictive Pericarditis

- Hepatic vein
 - **Expiration:** Enhanced diastolic flow reversal





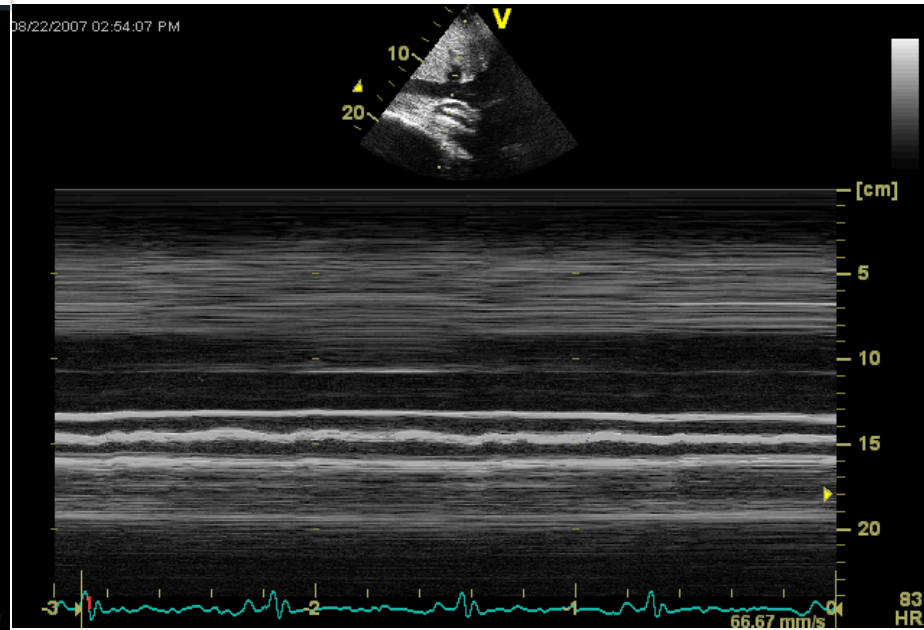
Diastolic Septal Bounce

Rapid filling during early diastole leads to asymmetrical filling of the RV and LV creating a fluctuating pressure gradient and an abrupt shift of the septum





Dilation and lack of respiratory variation in IVC





Differentiation of Constriction vs. Restriction

- Different etiologies



Differentiation of Constriction vs. Restriction

	Constrictive pericarditis	Restrictive cardiomyopathy
Mechanisms of disease	Cardiac volume constrained by inelastic pericardium resulting in impaired ventricular filling	Restriction of filling from impaired ventricular diastolic filling
Physiologic response	Changes in intrathoracic pressure not transmitted to cardiac chambers (obliteration of pericardial space)	Normal respiratory variation in intrathoracic pressure transmitted normally to cardiac chambers
Ventricular interaction	Greatly enhanced	Unchanged
Intrinsic myocardial function	Normal	Abnormal



Differentiation of Constriction vs. Restriction

- Different etiologies
- Similar clinical presentations
- Similar physical exam signs
- Thick pericardium is not necessary or sufficient to dx constriction
- Overlapping echo & hemodynamic features

	Constriction	Restriction
Prominent y decent in venous pressure	Present	Variable
Paradoxical pulse	~1/3 cases	Absent
Pericardial knock	Present	Absent
Equal Right and Left sided pressures	Present	Left at least 3-5 mmHg > right
Filling Pressures > 25 mmHg	Rare	Common
PASP > 60 mmHg	No	Common
Hepatic veins	Inc expiratory flow reversal	Inc inspiratory flow reversal
“Square root” sign	Present	Variable
Respiratory variation inflows velocities	Exaggerated	Normal
Ventricular wall thickness	Normal	Usually Increased
Atrial Size	Possible LA enlargement	Bi-atrial enlargement
Septal Bounce	Present	Absent
Tissue Doppler e' velocity	Increased	Reduced
Pericardial thickness	Increased	Normal



Deformation Imaging

- **Constriction**
 - Deformation of the LV and early diastolic recoil were attenuated in the **circumferential direction**
- **Restriction**
 - Attenuated in the **longitudinal direction**



Differentiation of Constriction vs. Restriction

Echocardiographic Parameters

Parameter	Constrictive pericarditis	Restrictive cardiomyopathy
Septal bounce	Yes	No
MV inflow respiratory variation	$\geq 25\%$	None
TV inflow respiratory variation	$> 40\%$	None
MVDT	Short	< 160 ms
Hepatic vein reversal	Diastolic reversal with expiration	No change
IVRT	Decrease during expiration Increase during inspiration	No change
TR duration	Increase	No change
E:e'	$< 8-10$	> 15
Myocardial mechanics with strain image	Normal longitudinal strain Decrease net-twist angle	Decrease longitudinal strain Normal net-twist angle

E: Peak transmitral flow velocity at early diastolic filling phase; *e'*: Peak early diastolic mitral annular velocity; *E:e'*: Ratio of *E* and *e'* velocities; *IVRT*: Isovolumic relaxation time; *MV*: Mitral valve; *MVDT*: Mitral valve deceleration time; *TR*: Tricuspid regurgitation; *TV*: Tricuspid valve.



Summary

- **Restrictive cardiomyopathy is a disease of the myocardium and may be due to various etiologies including noninfiltrative, infiltrative, and storage diseases**
- **Constrictive pericarditis is a disease of the pericardium leading to thickening and impairment of diastolic filling**



Summary

- **There are a numerous overlapping physical exam, echocardiographic and hemodynamic findings that overlap between the two disease states**



Summary

Think restrictive cardiomyopathy

- **Thick walls, biatrial enlargement**
- **Decleration time <150 ms**
- **Decreased septal and lateral e' velocities**
 - **Lateral E' velocity $>$ septal**
- **Elevated PASP**



Summary

Think constrictive pericarditis

- **Septal bounce**
- **Inspiratory drop in left heart velocities (MV, PV, LVOT)**
- **Thickened pericardium (not necessary)**
- **Dilated IVC**
- **Expiratory hepatic vein diastolic flow reversal**



Thank you!