Constriction/ Restriction

PP16 Imaging Conference

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Differentiation of Constriction vs. Restriction

- **Why is it important?**

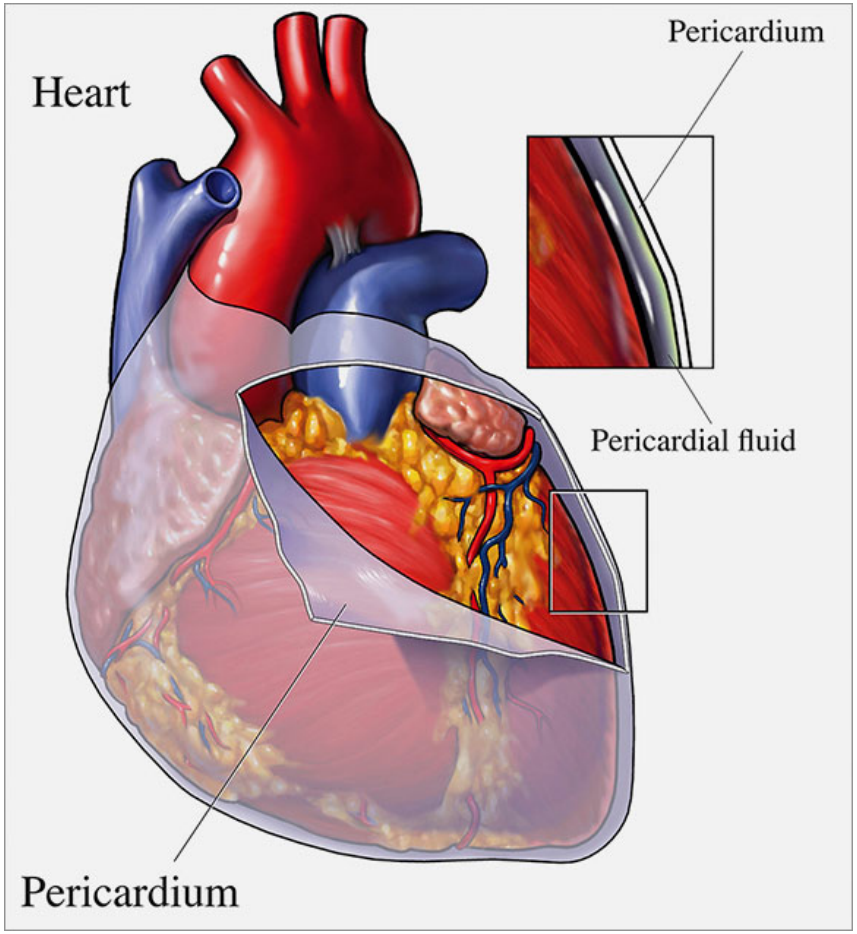
- Important therapeutic implications
  - Pericardiectomy
  - Heart failure management and treatment of underlying disease

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

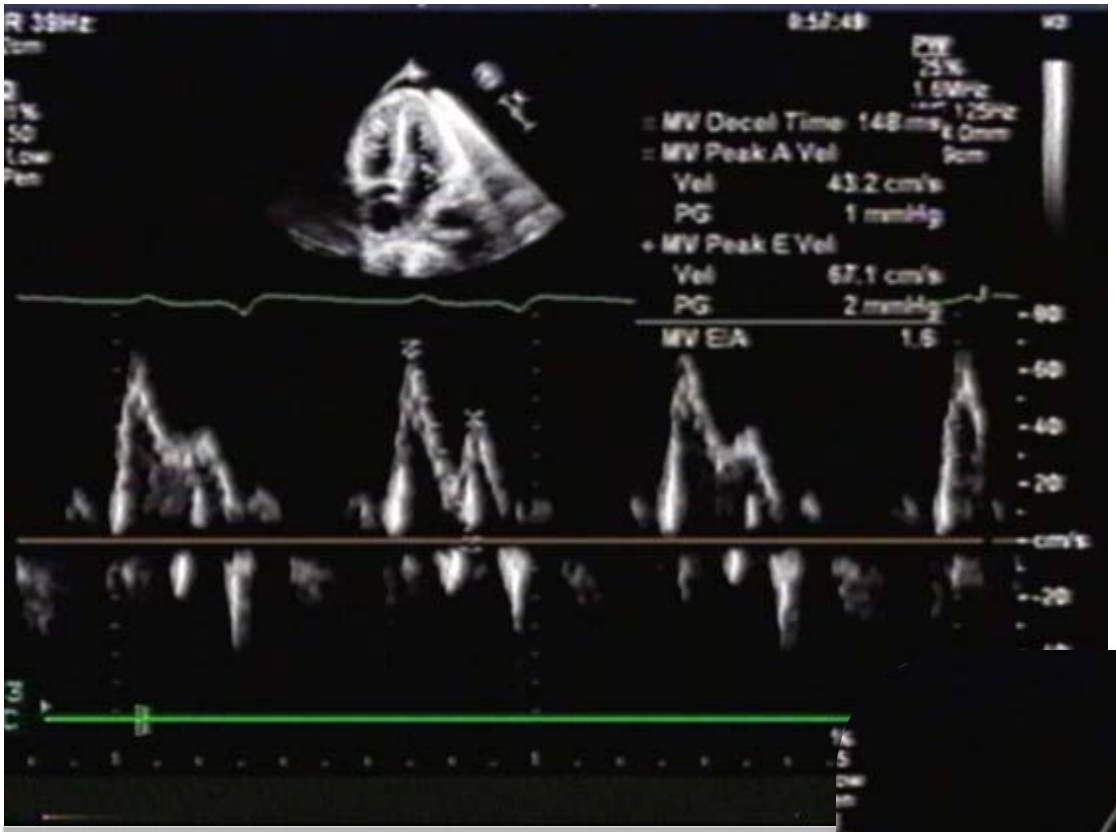
- Hemochromatosis
- Fabry Disease
- Glycogen storage disease

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/m^2

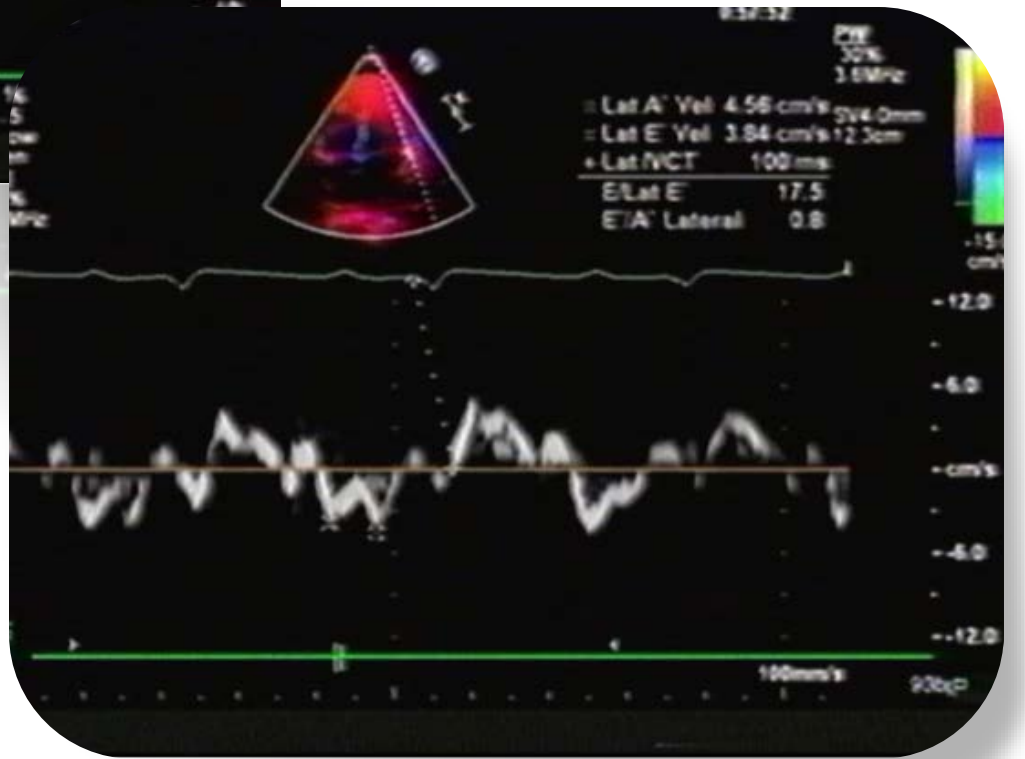


E/A = 1.7

DT = 140

E’ = 4

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

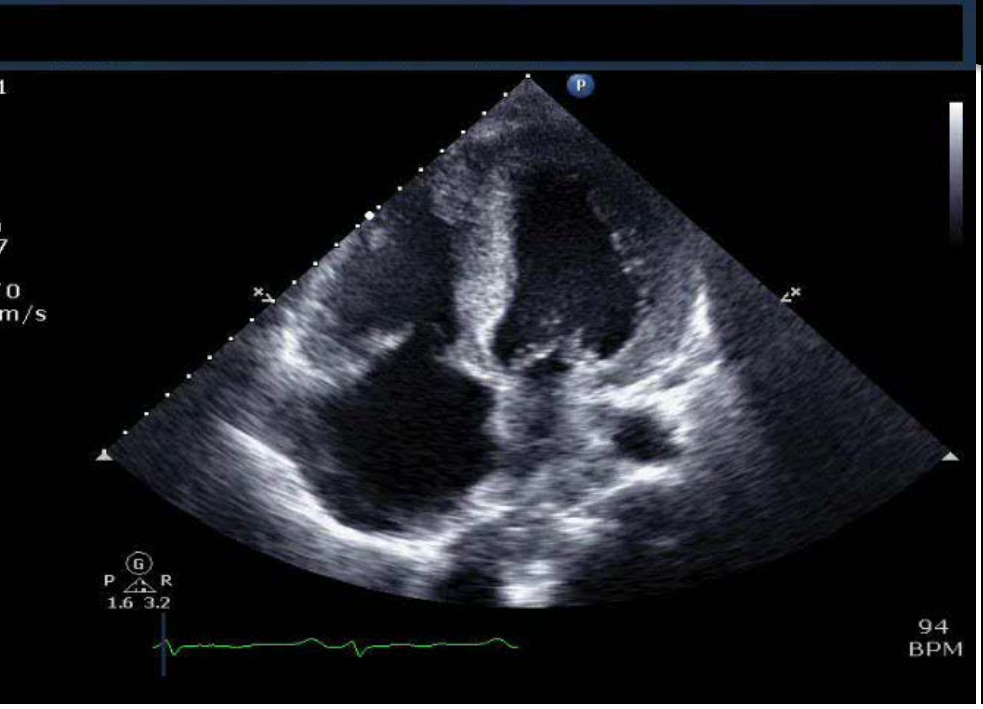
- E/A ratio >2
- Decleration time <150 ms
- PV: small S wave, large D wave; S/D ratio <0.5

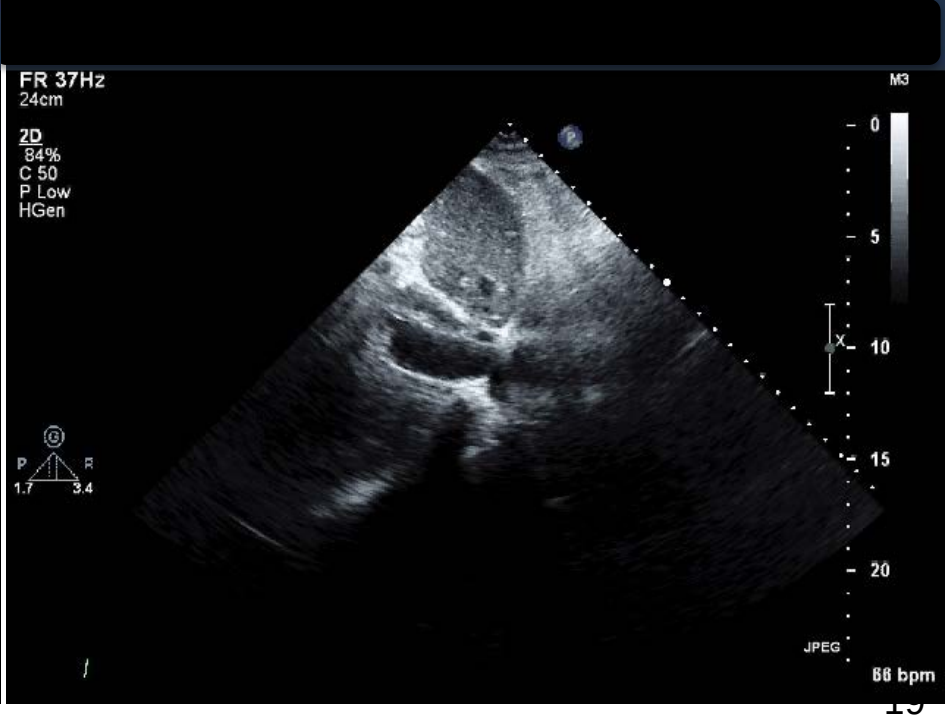
Case 1

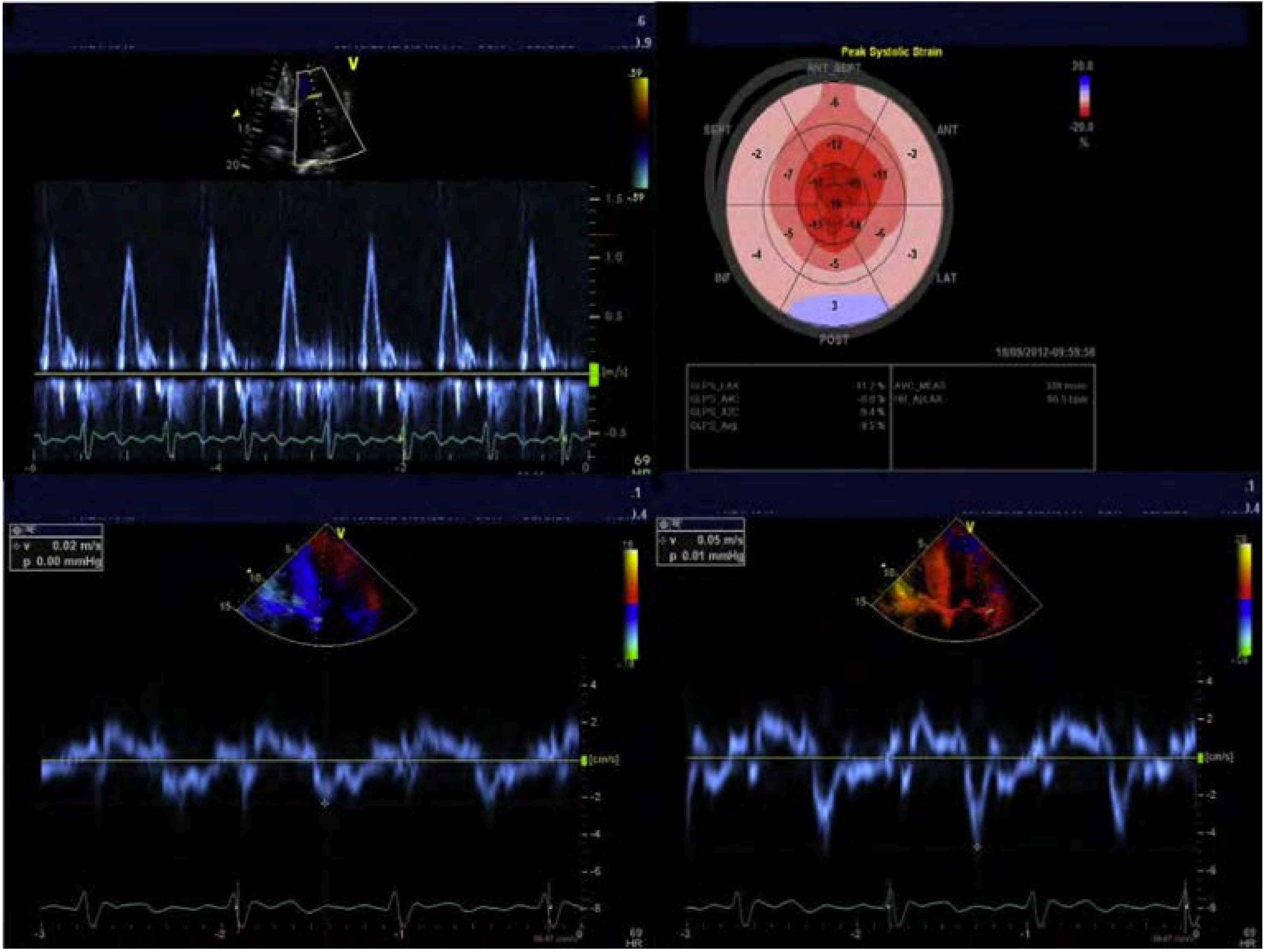
76 year old man with senile cardiac amyloidosis, Afib, ICD presents with generalized weakness


47Hz
M3
0
5
10
15
2.8


FR 61Hz
17cm
2D
58%
C 50
P Low
HRes
M3
0
5
10
15
2.1
4.2
JPEG


1.6 3.2
94
BPM


FR 37Hz
24cm
2D
84%
C 50
P Low
HGen
M3
0
5
10
15
20
1.7
3.4
JPEG
88 bpm


Peak Systolic Strain
ANT_SEPT
SEPT
ANT
LAT
POST
[m/s]
v 0.02 m/s
p 0.00 mmHg
v 0.05 m/s
p 0.01 mmHg
[cm/s]
[cm/s]

Case 2

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

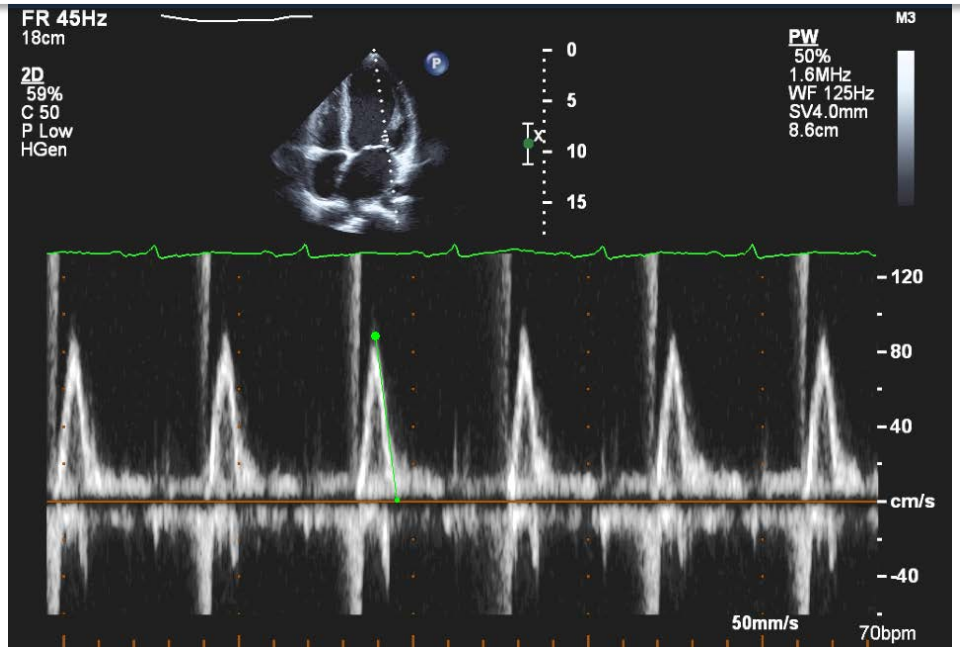
Hemochromatosis

- 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)

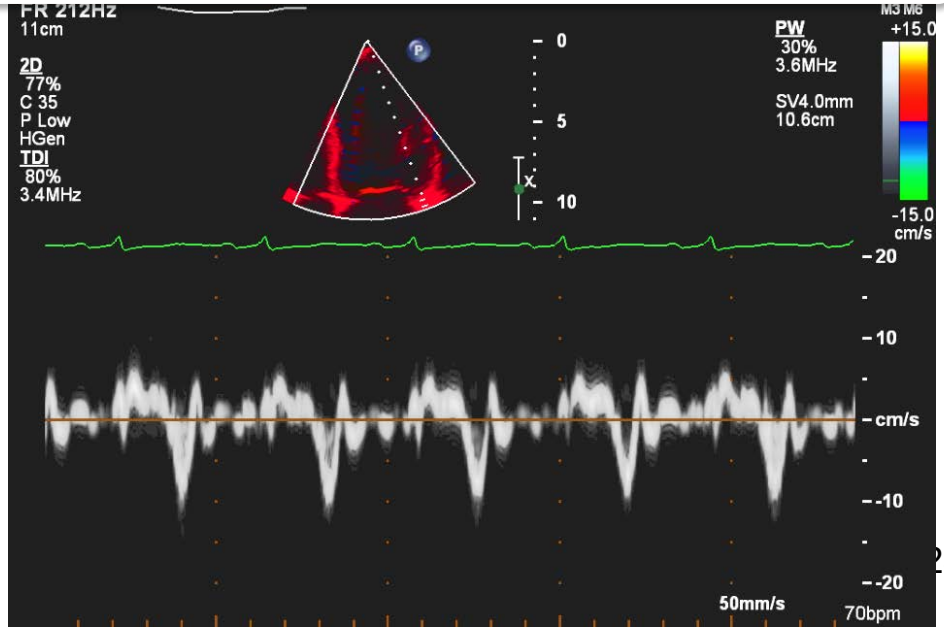
- Genetic testing for mutations in the HFE gene

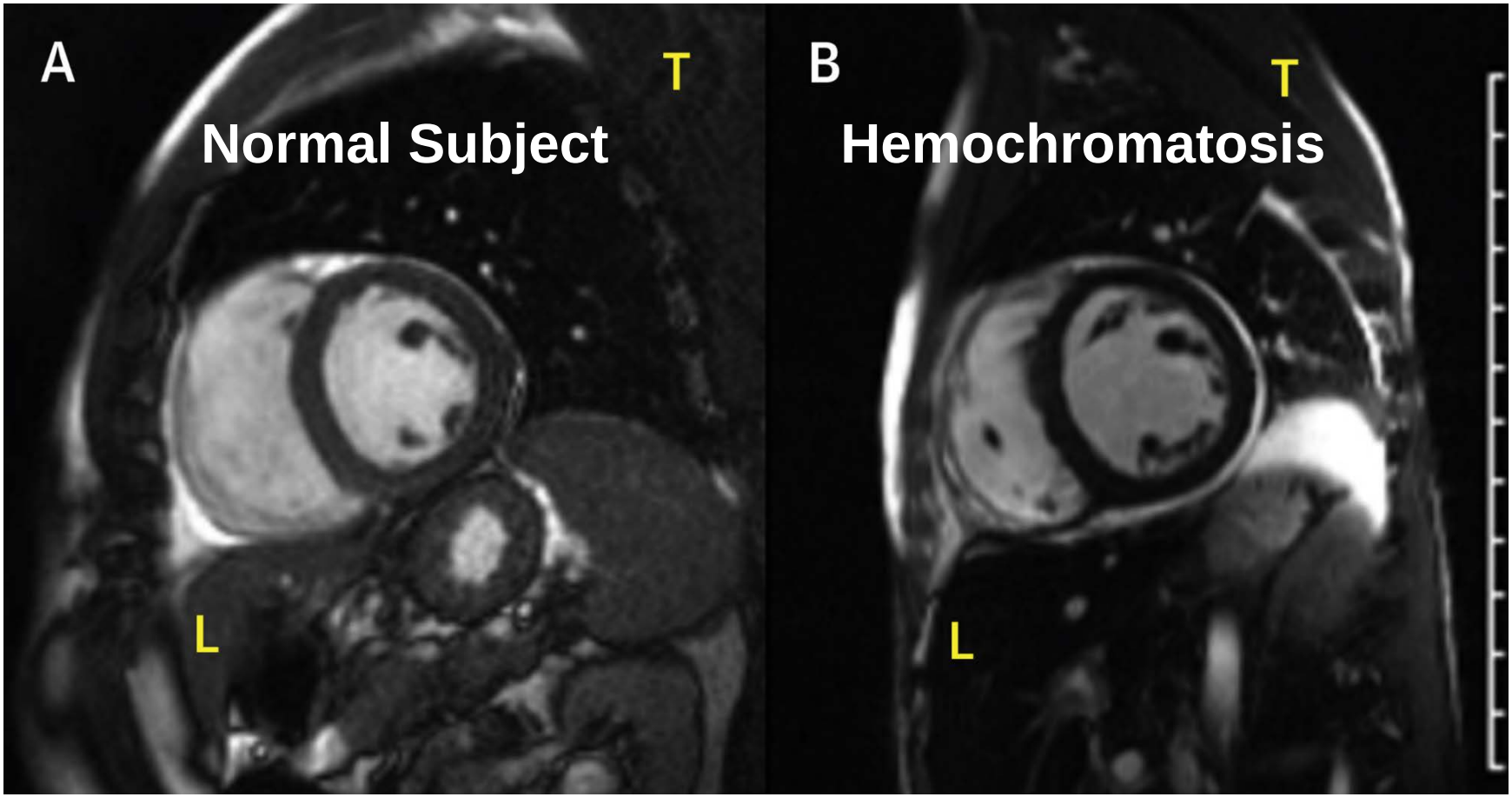
Hemochromatosis



SEPTAL

LATERAL





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

J Am Coll Cardiol. 2016;67(17):2061-2076

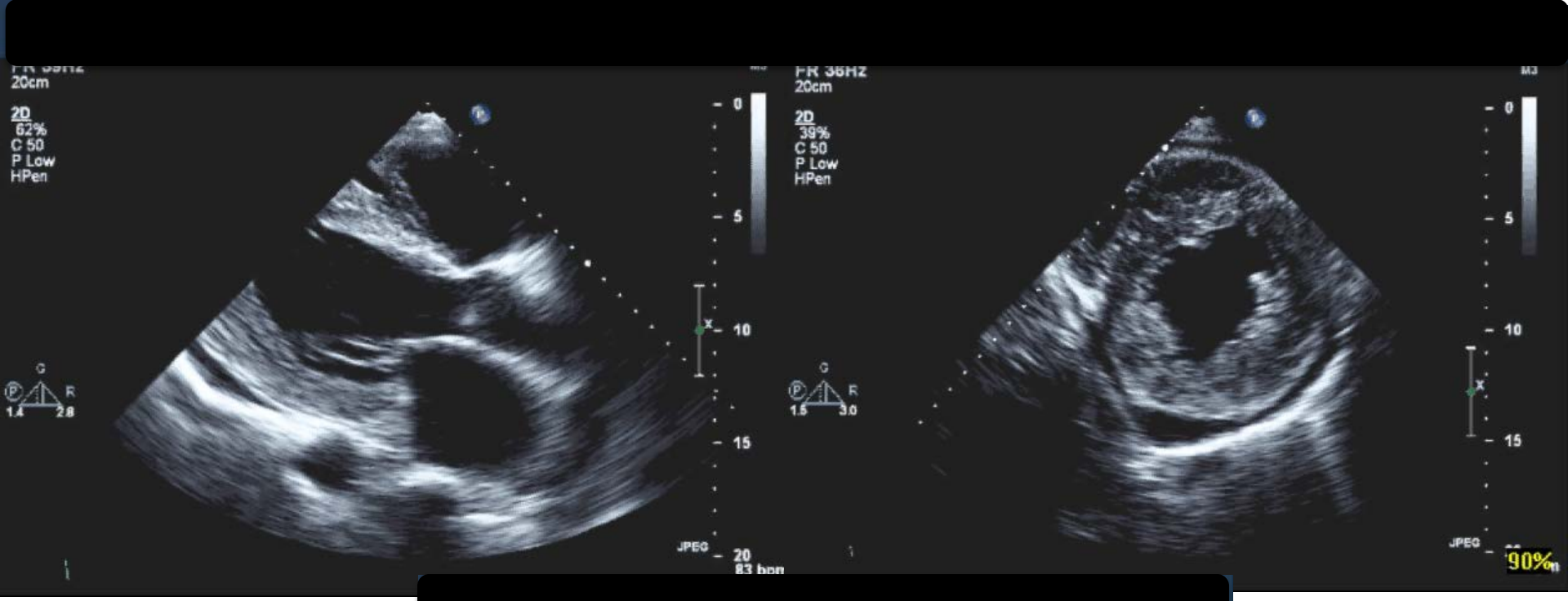
Case 3

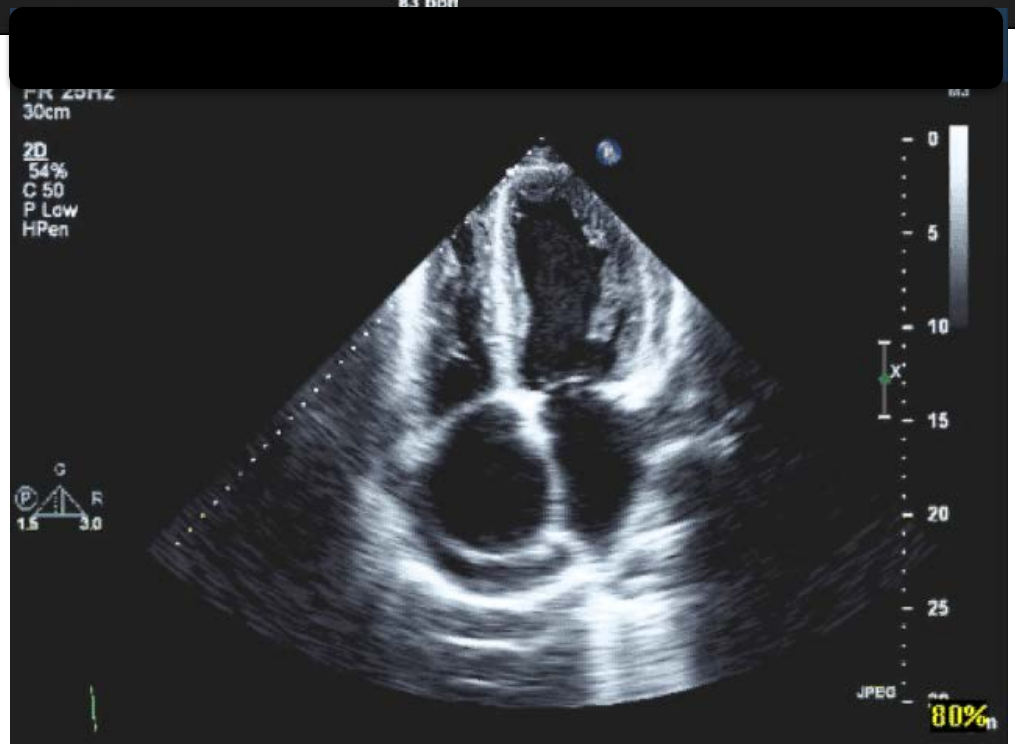
37 year old man with hypertension, end stage renal disease on dialysis, and Fabry's disease on RV biopsy

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


20cm
2D
62%
C 50
P Low
HPen
0
5
10
15
20
JPEG
FR 36Hz
20cm
2D
39%
C 50
P Low
HPen
0
5
10
15
JPEG
90%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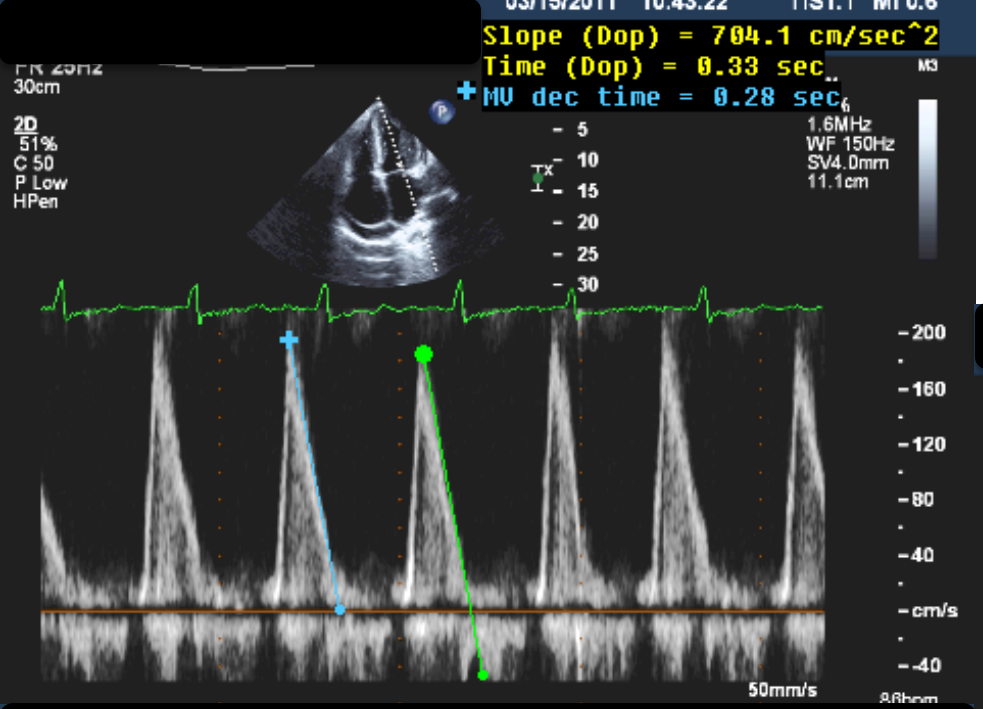
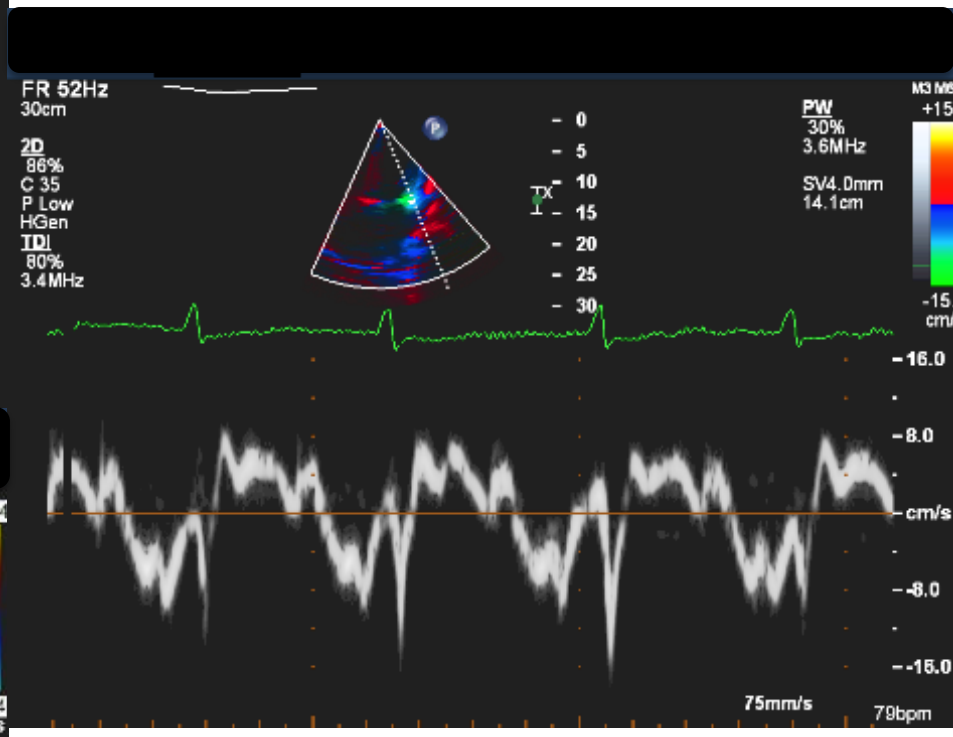
C 50
P Low
HPen
0
5
10
15
20
25
JPEG
80%


Slope (Dop) = 704.1 cm/sec^2
Time (Dop) = 0.33 sec
MV dec time = 0.28 sec
30cm
2D
51%
C 50
P Low
HPen
1.6MHz
WF 150Hz
SV4.0mm
11.1cm
200
160
120
80
40
cm/s
-40
50mm/s


FR 52Hz
30cm
2D
86%
C 35
P Low
HGen
TDI
80%
3.4MHz
PW
30%
3.6MHz
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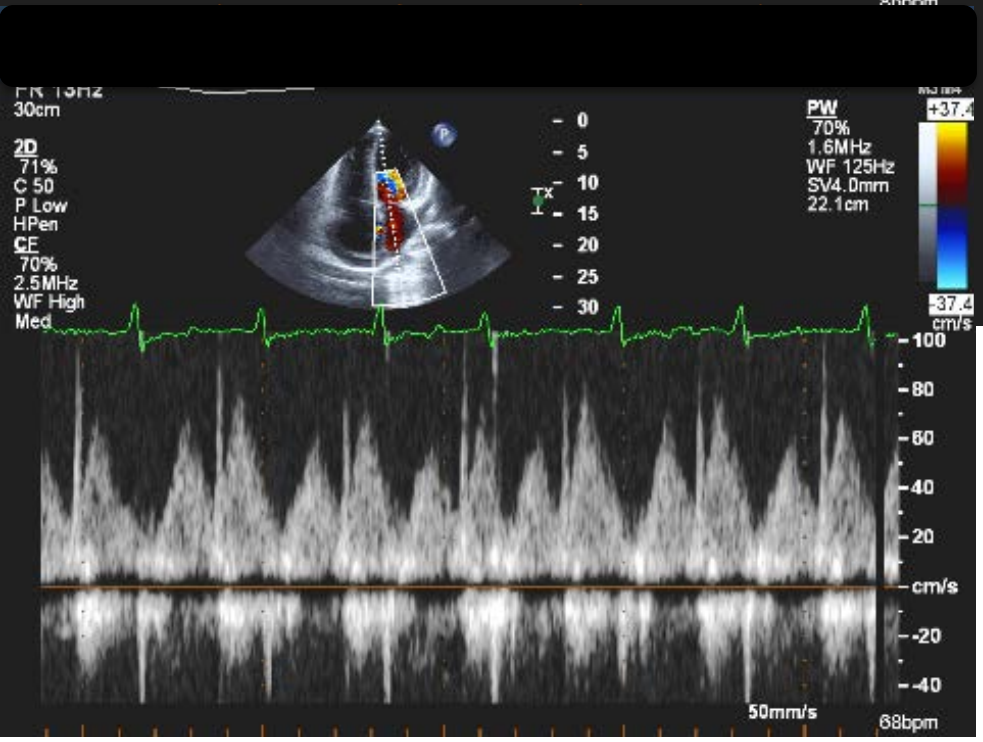
14.1cm
16.0
8.0
cm/s
-8.0
-16.0
75mm/s
79bpm


30cm
2D
71%
C 50
P Low
HPen
CE
70%
2.5MHz
WF High
Med
PW
70%
1.6MHz
WF 125Hz
SV4.0mm
22.1cm
+37.4
-37.4
cm/s
100
80
60
40
20
cm/s
-20
-40
50mm/s
68bpm

Constrictive Pericarditis

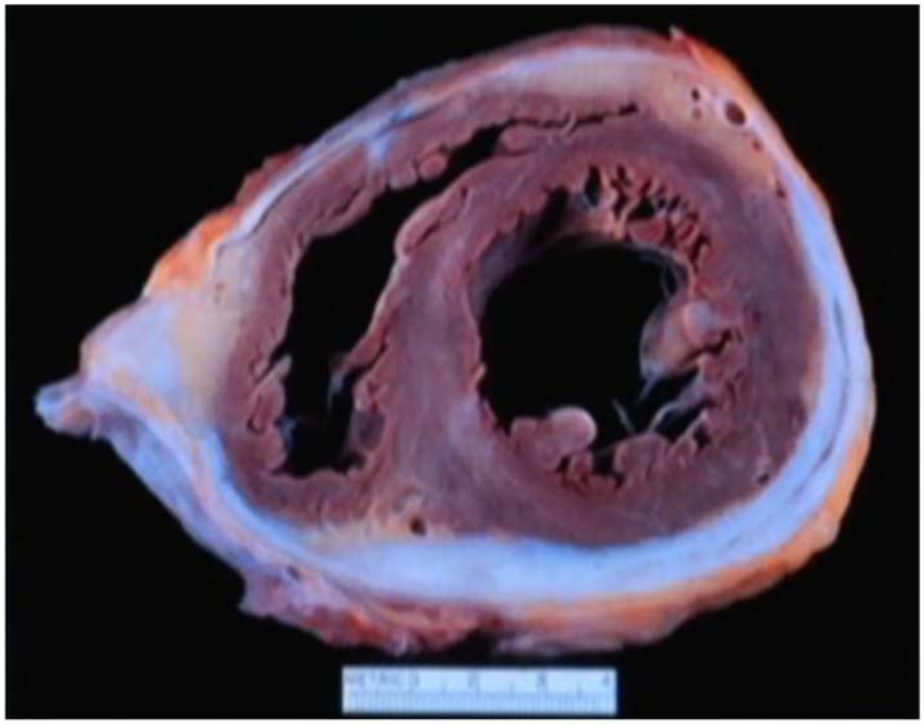
Constrictive Pericarditis

- **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**

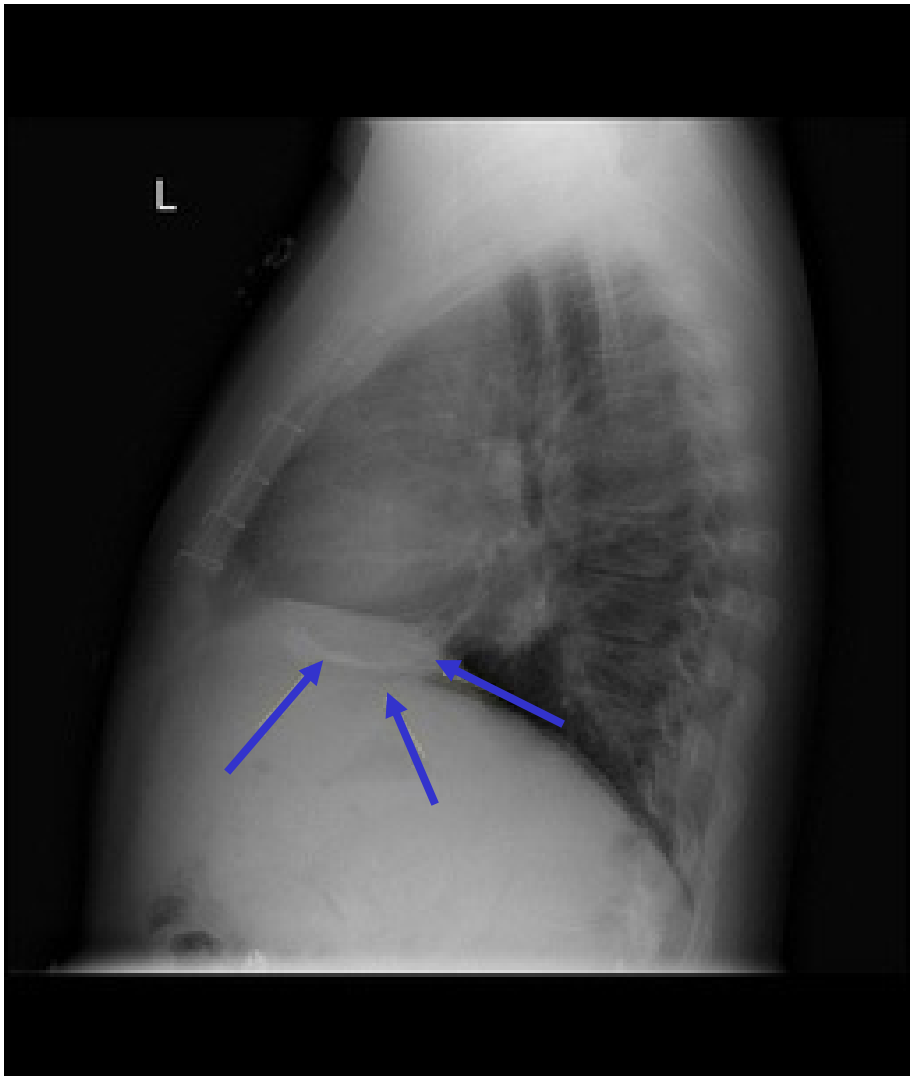
Constrictive Pericarditis

- **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



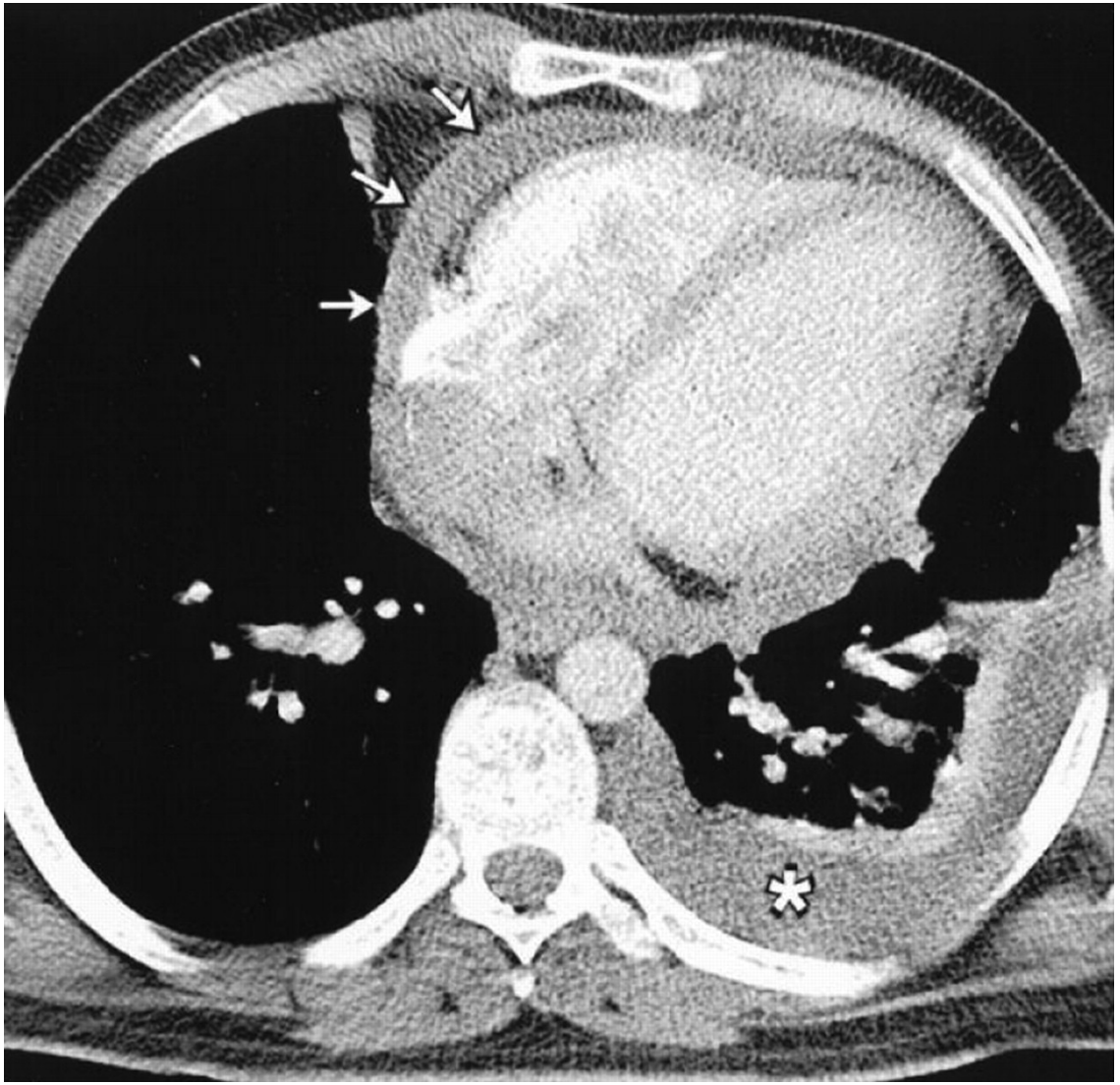
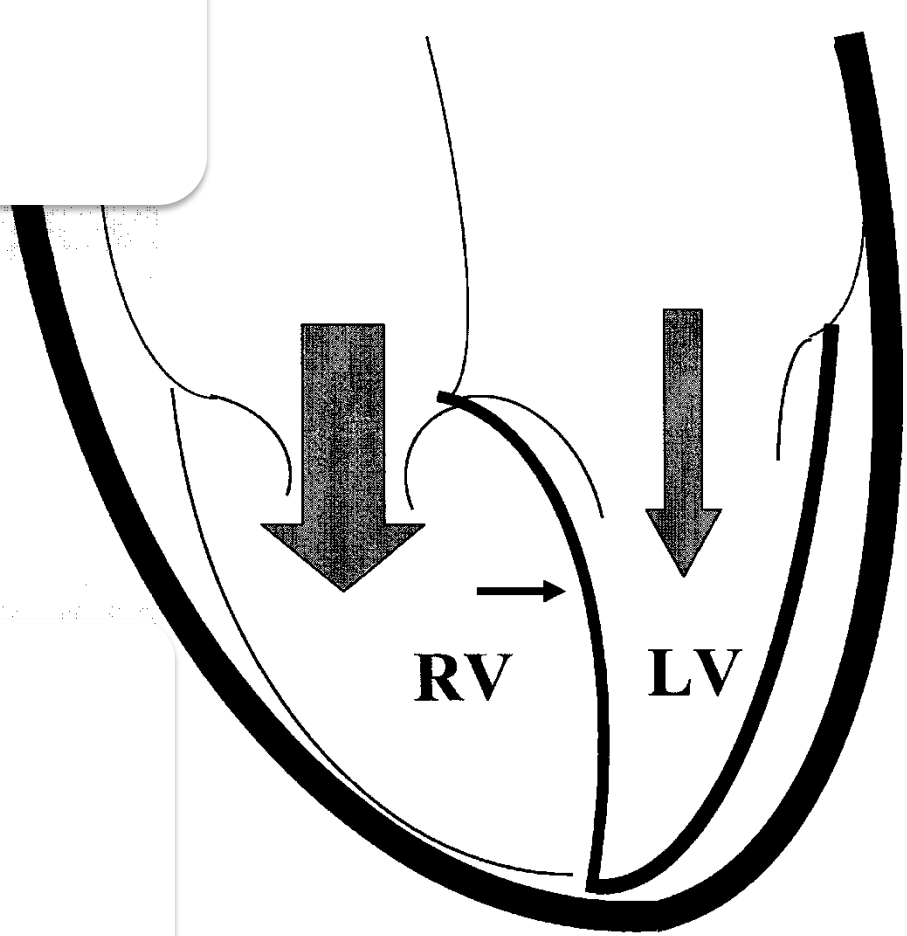


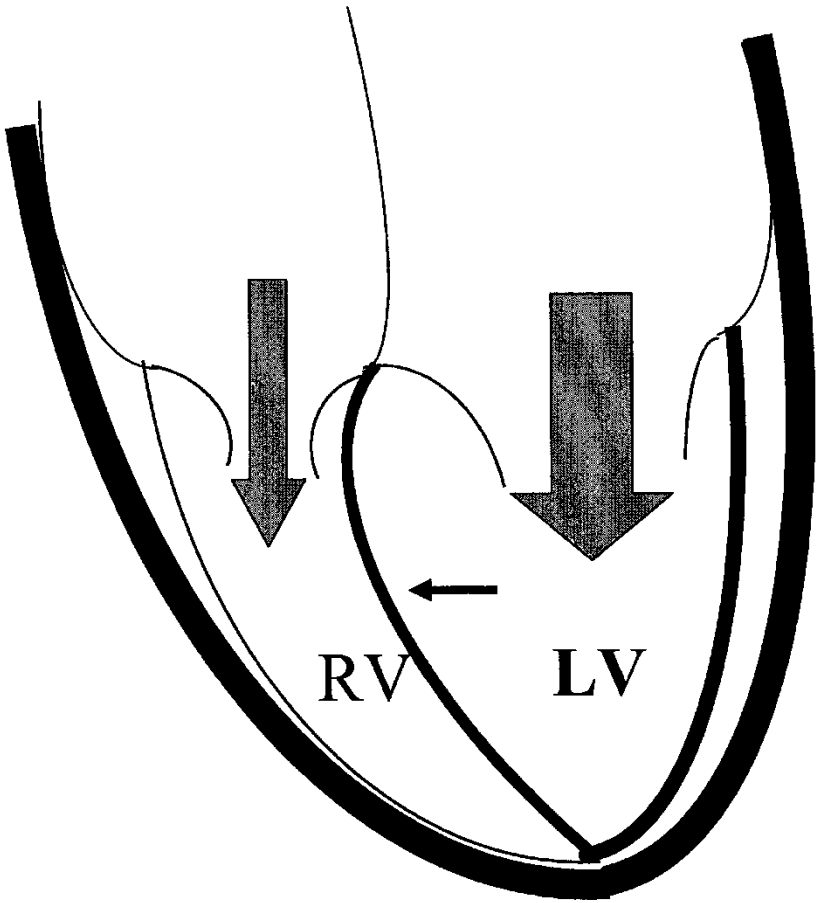
Figure 5. Chest CT from a patient with pericardial constriction showing thickened pericardium (arrows) and a left pleural effusion.

William C. Little, and Gregory L. Freeman Circulation. 2006;113:1622-1632

Constrictive Pericarditis



Inspiration



Expiration

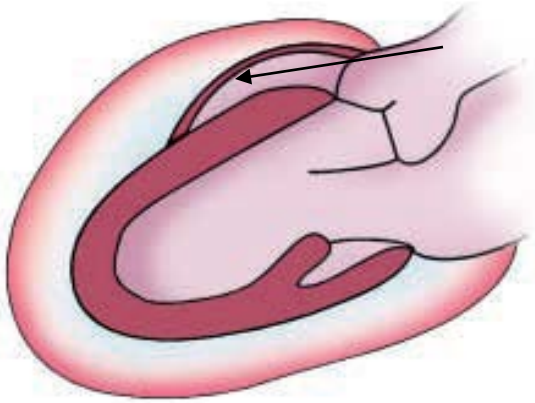
Constrictive Pericarditis: Pathophysiology

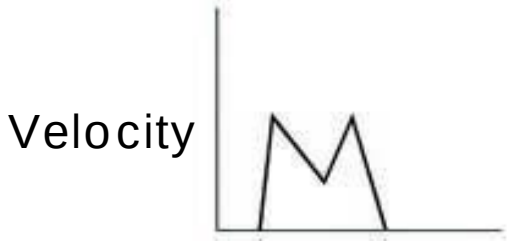
- **Marked restriction of filling**
- **Elevation and equilibration of filling pressures in all heart chambers**
  - **Early to mid diastole ventricular filling is abrupt and rapid**
  - **This filling abruptly ceases when the intracardiac limit reaches its set limit**
- **Systemic venous congestion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

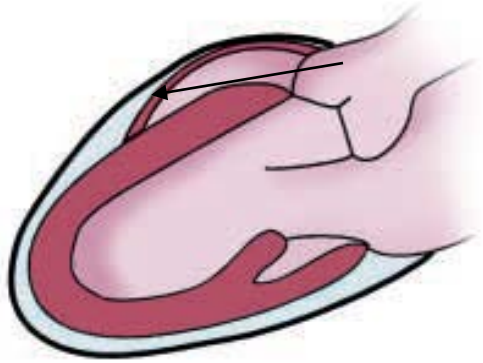
With constriction, early diastolic filling is rapid but ends abruptly when the volume limits of the rigid pericardial space are reached

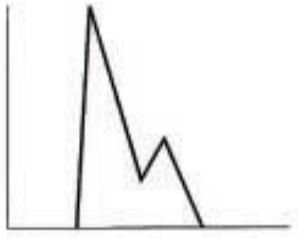
TAMPONADE

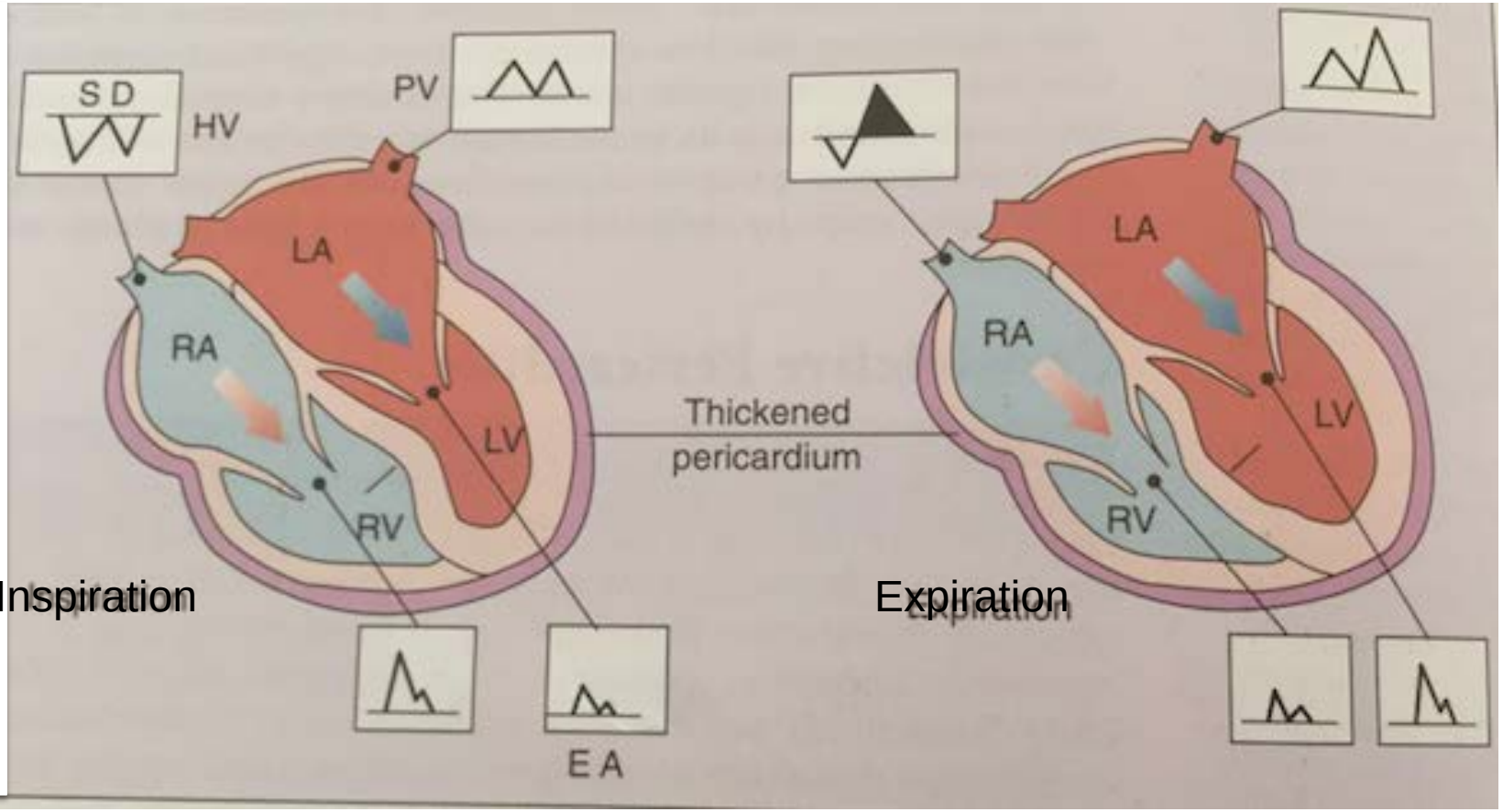




CONSTRICTION







Inspiration

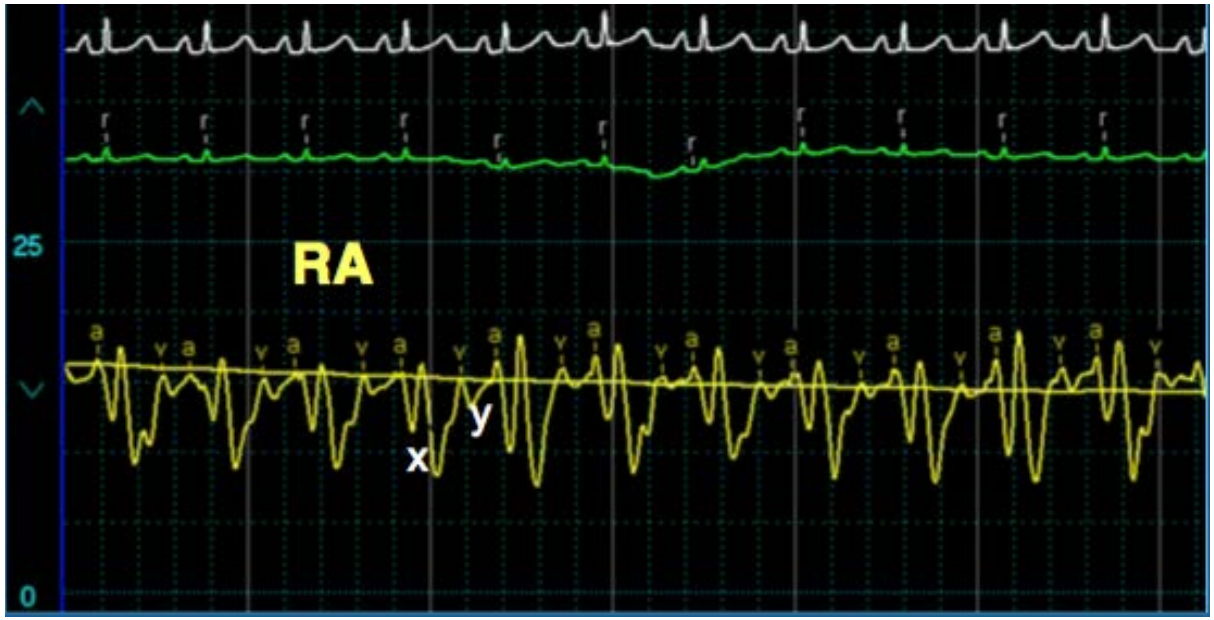
Expiration

Braunwald's Heart Disease.

Constriction: Hemodynamics

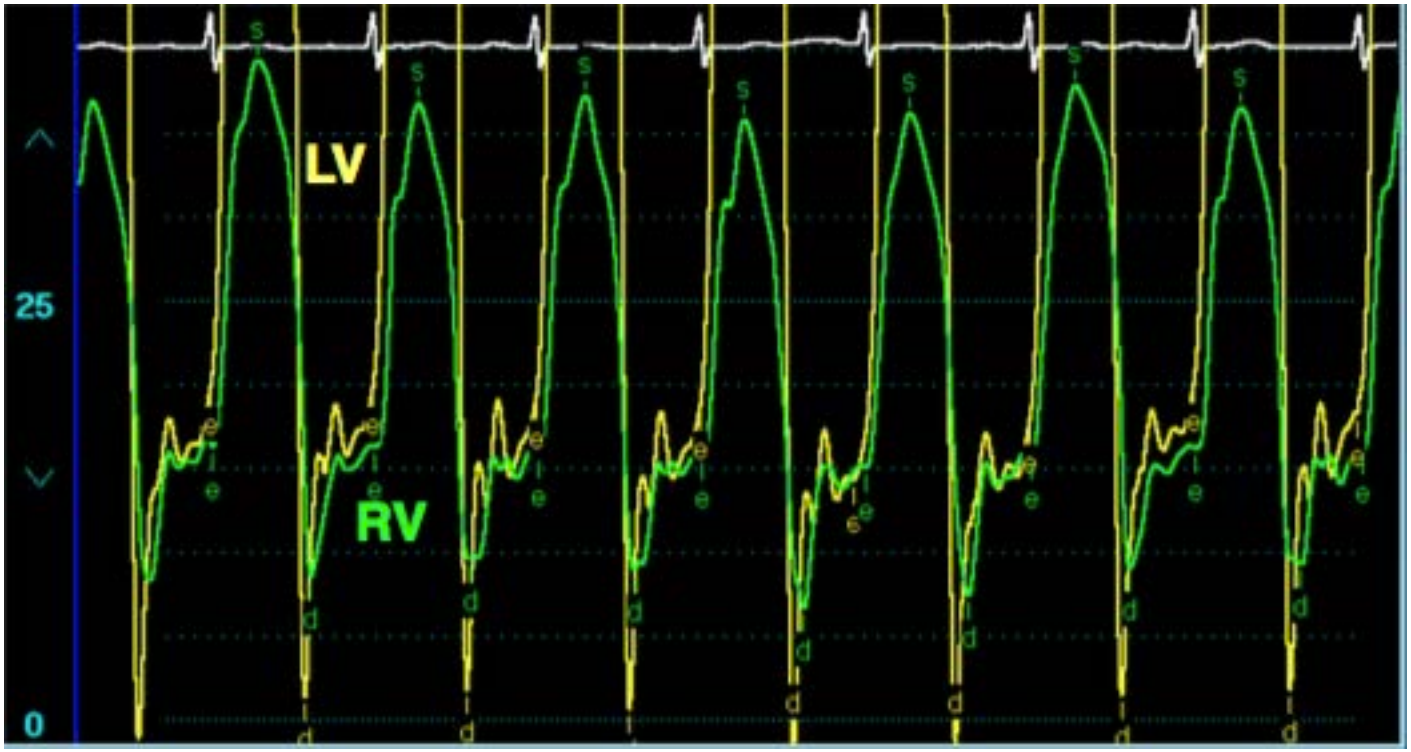
- **RA: elevated pressure with “M” or “W” pattern**
- **RV: “dip and plateau” or “square root” sign**
- **Diastolic equalization of LV and RV pressures**
  - **Volume loading**
- **Systolic discordance of LV and RV pressures**

Tamponade

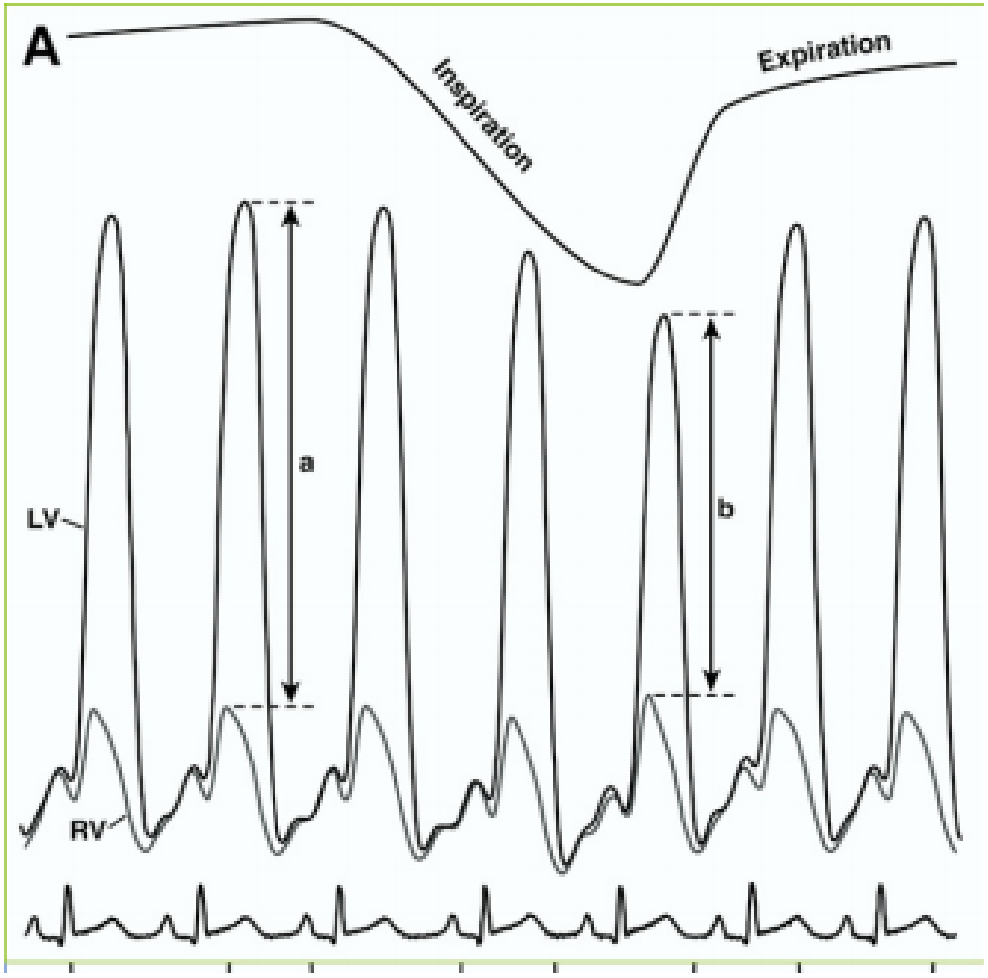


Constriction

Constriction: Diastolic Equalization



“Dip and plateau”

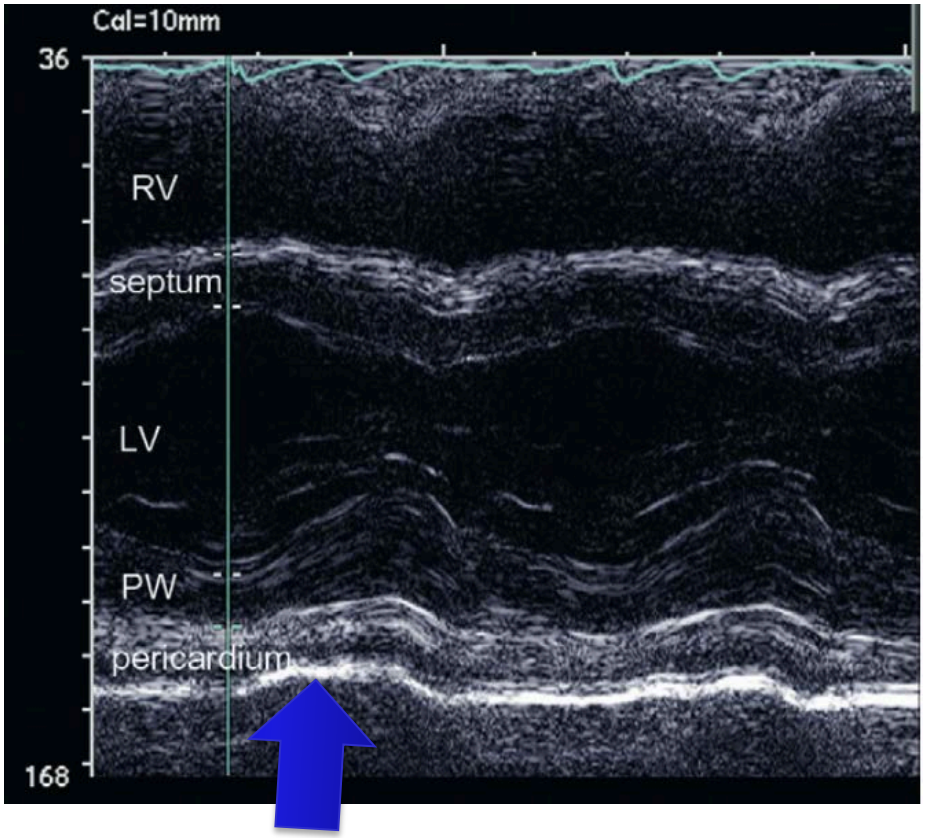


Echo findings in Constriction

- 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

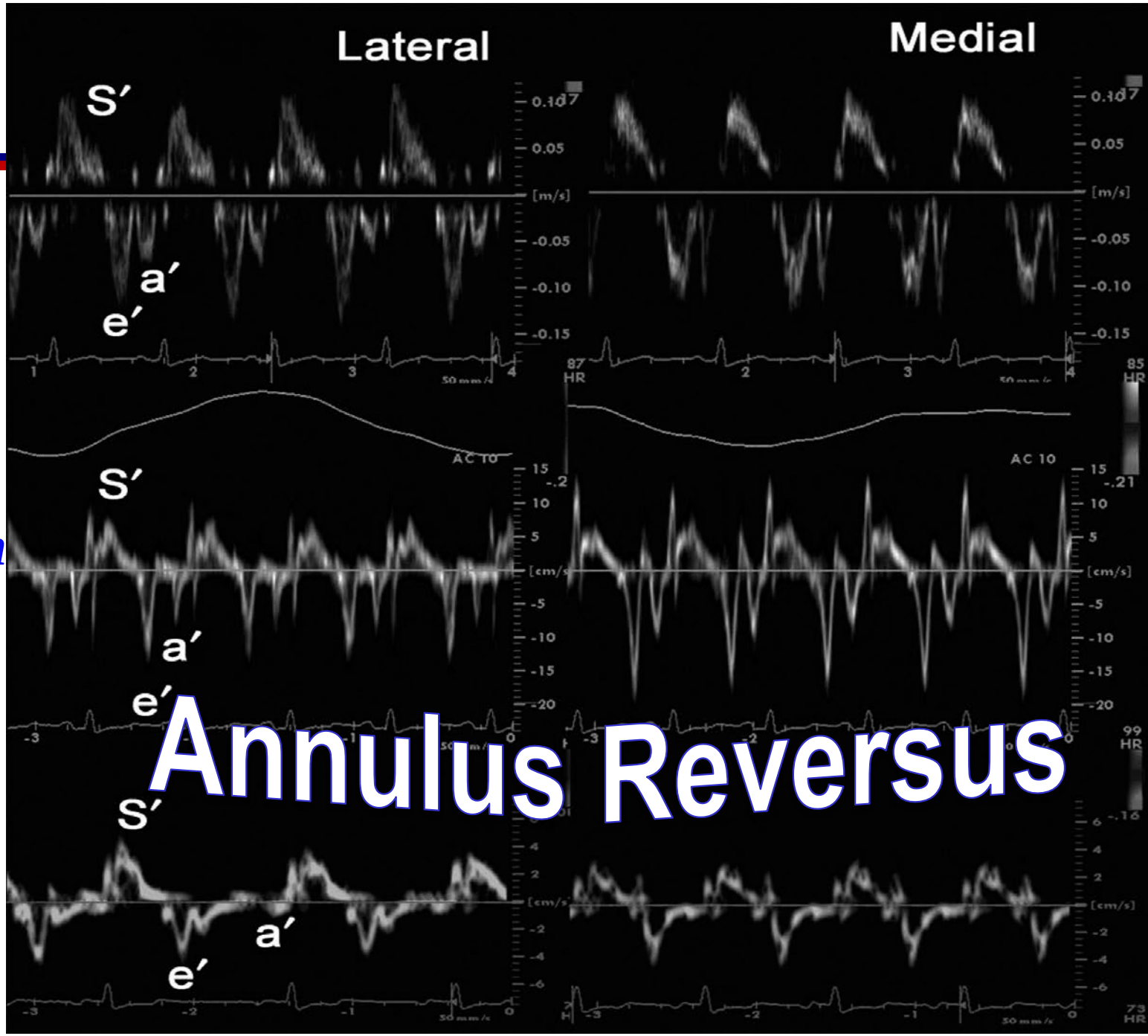
M-mode

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 ≥25%**
    - **TV inflow: increases ≥40%**
- **Hepatic venous flow reversal with expiration**



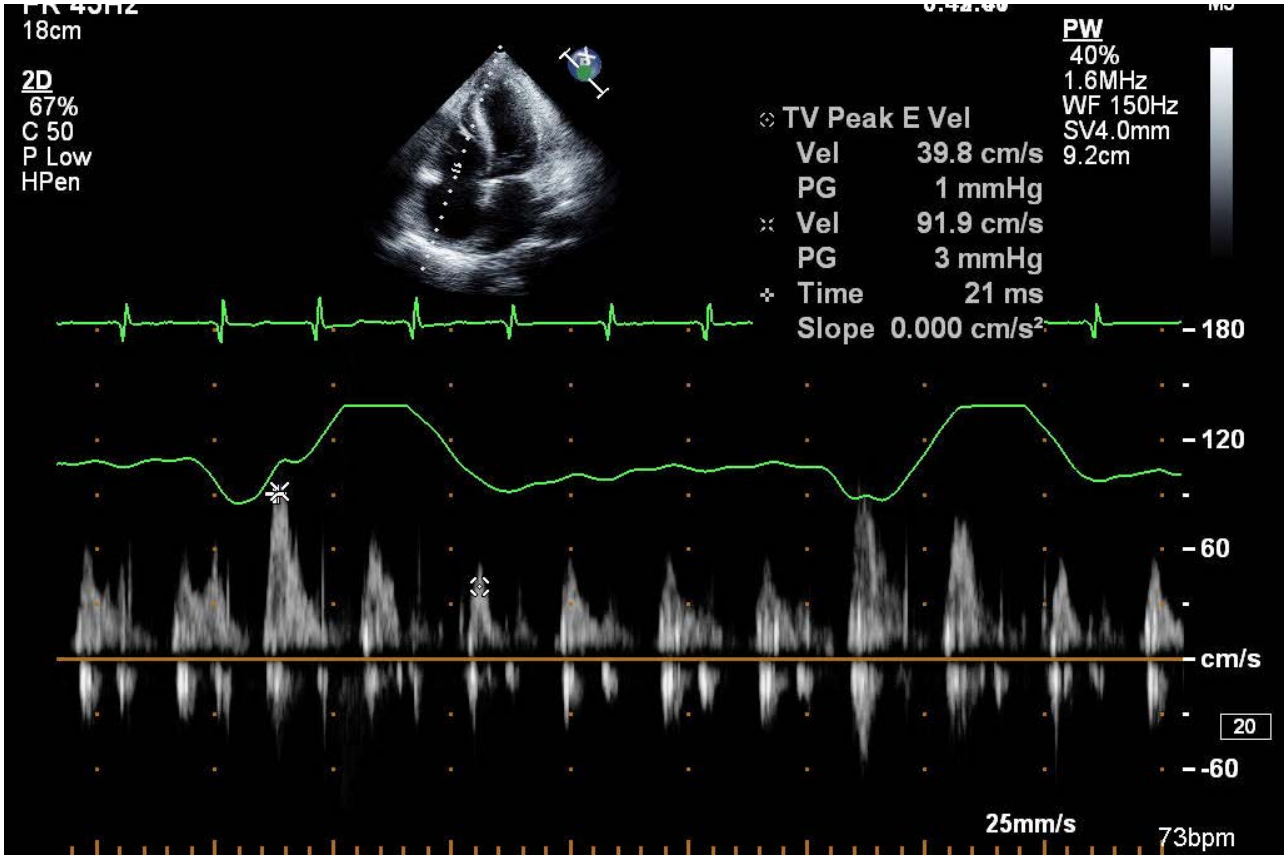
Normal

Constriction

Restriction

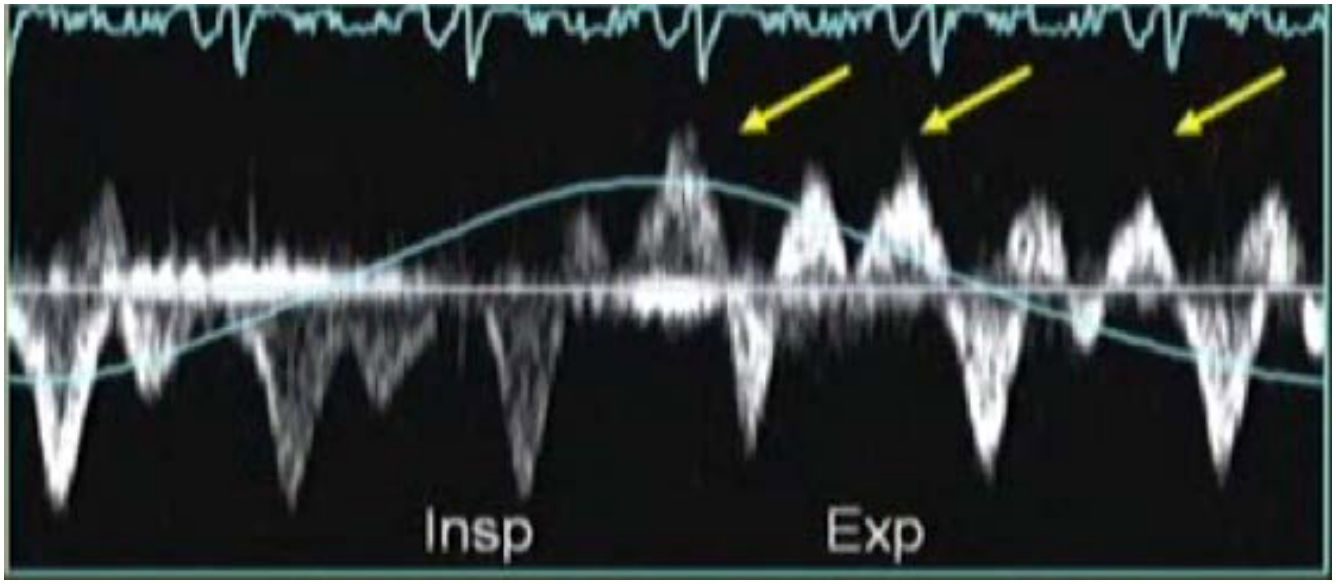
Tricuspid Valve In-Flow

- **Tricuspid inflow**
  - **E wave increase ≥ 40% with inspiration**



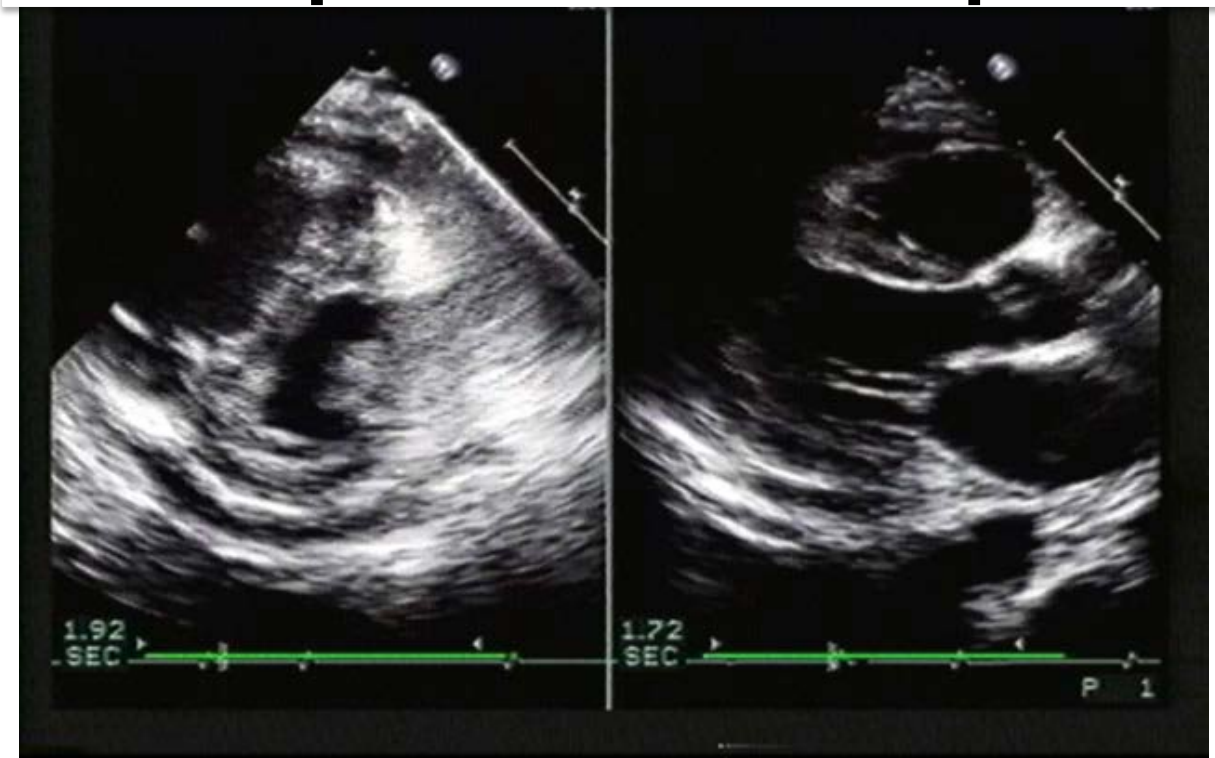
Doppler Findings: Constrictive Pericarditis

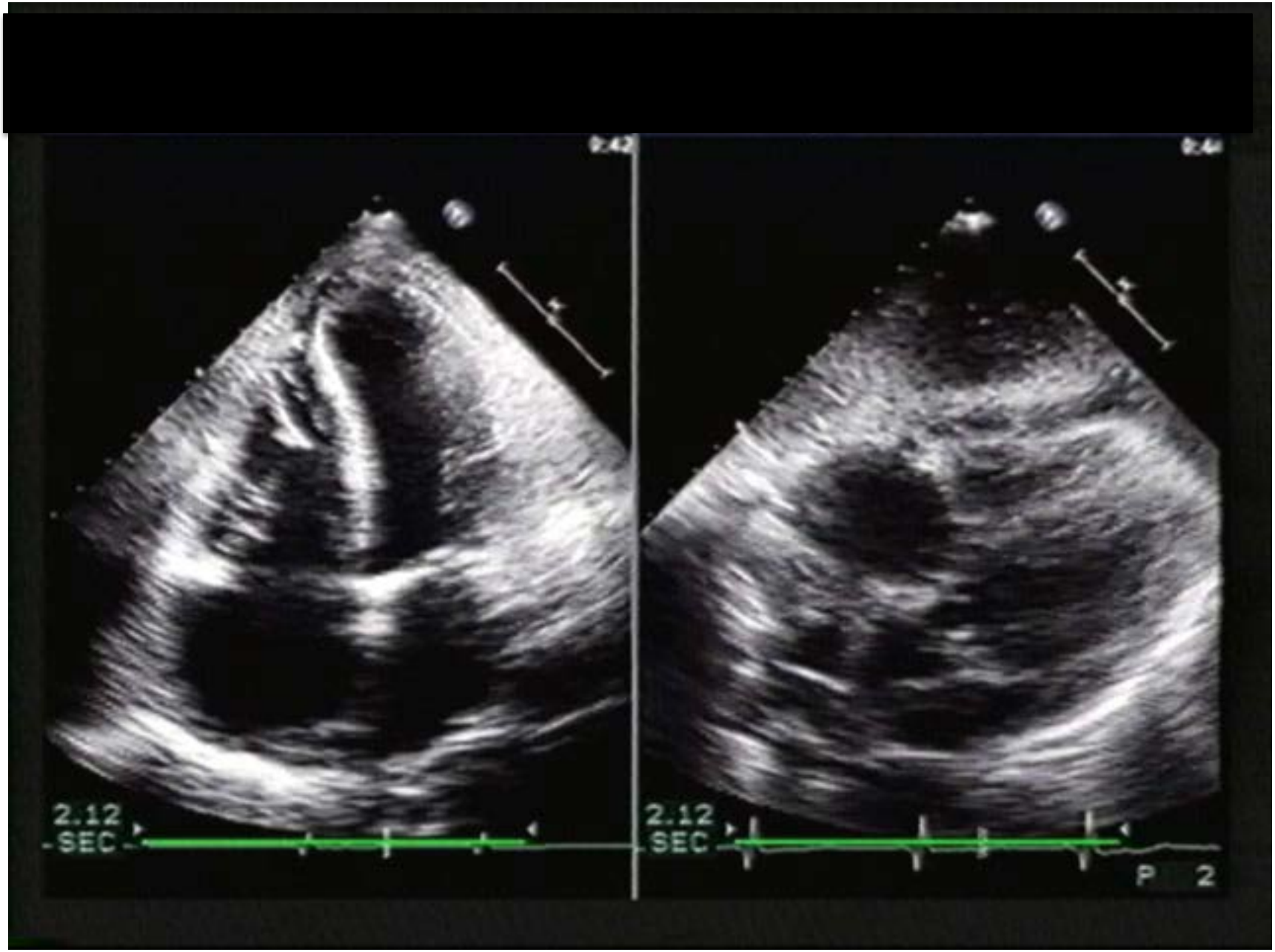
- **Hepatic vein**
  - ***Expiration*: Enhanced diastolic flow reversal**



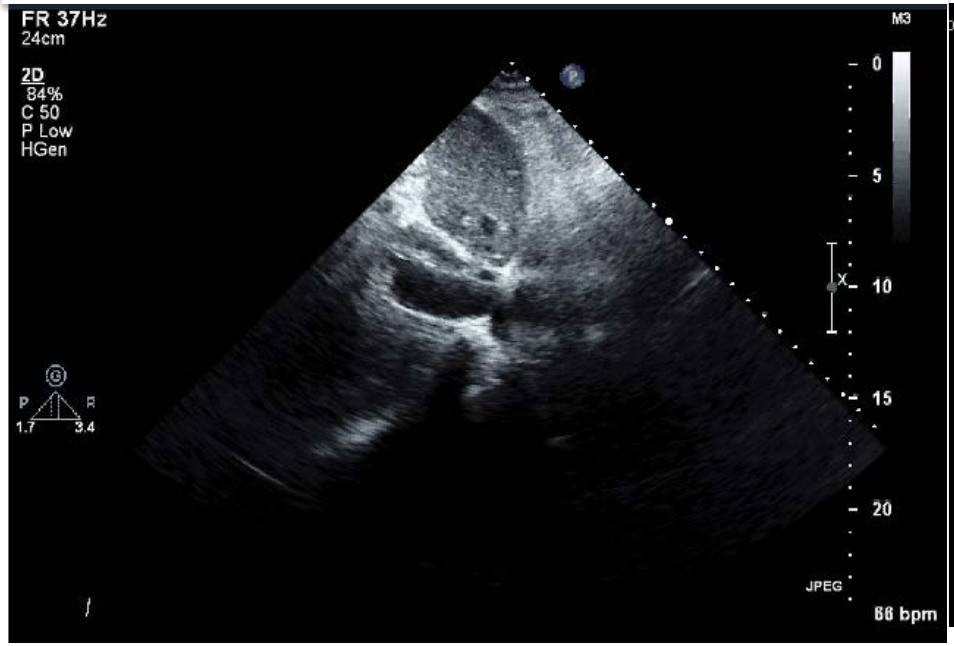
Diastolic Septal Bounce

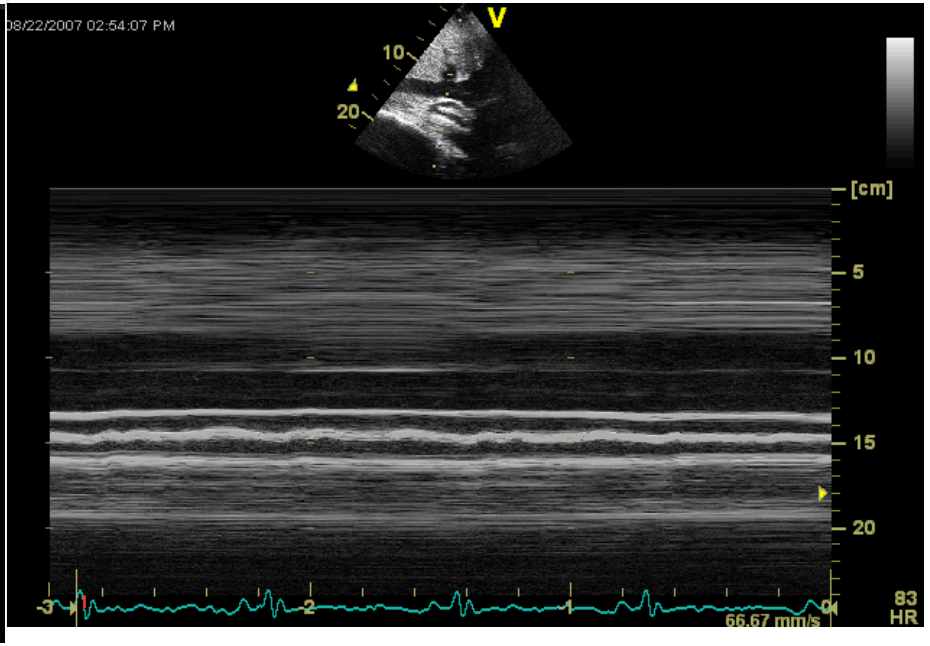
Rapid filling during early diastole leads to asymmetrical filling of the RV and LV creating a fluctuating pressure gradient and an abrupt shift of the septum




2.12
SEC
2.12
SEC
P 2

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
Physiological 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	≥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 noninflitrative, 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!