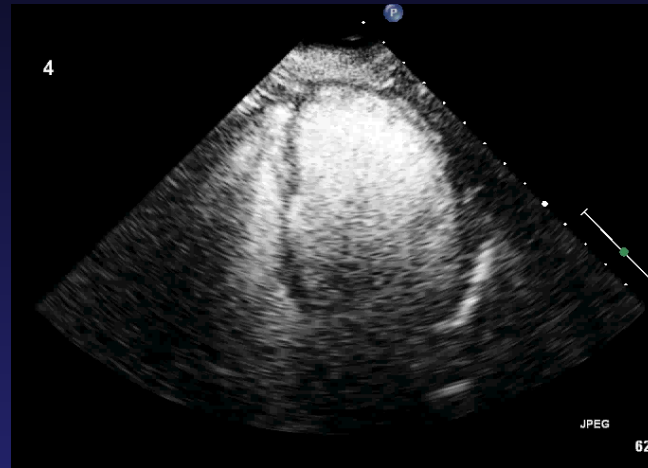


# Cardiac Sarcoidosis



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Professor of Medicine

Northwestern University

Bluhm Cardiovascular Institute

Medical Director, Echocardiography Laboratory

Northwestern Memorial Hospital

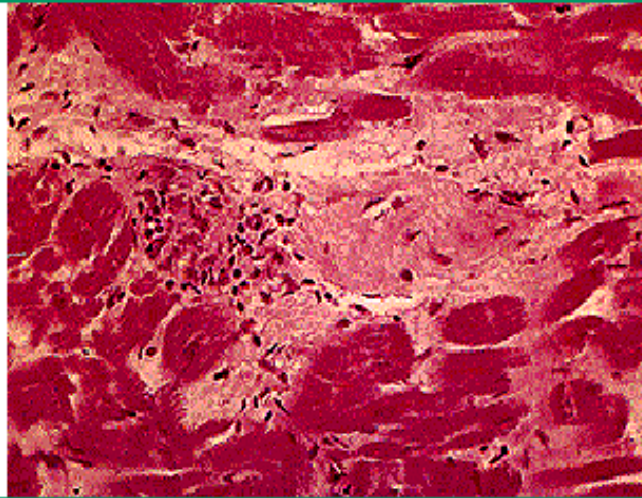
**Disclosures: None**

# Sarcoidosis

- Sarcoidosis is a heterogeneous, non-caseating, granulomatous disorder of unknown etiology that can involve any organ within the body.
- Myocardial involvement may be more frequent in patients with cardiac symptoms, but asymptomatic cardiac involvement appears to be common

## Myocardial sarcoidosis

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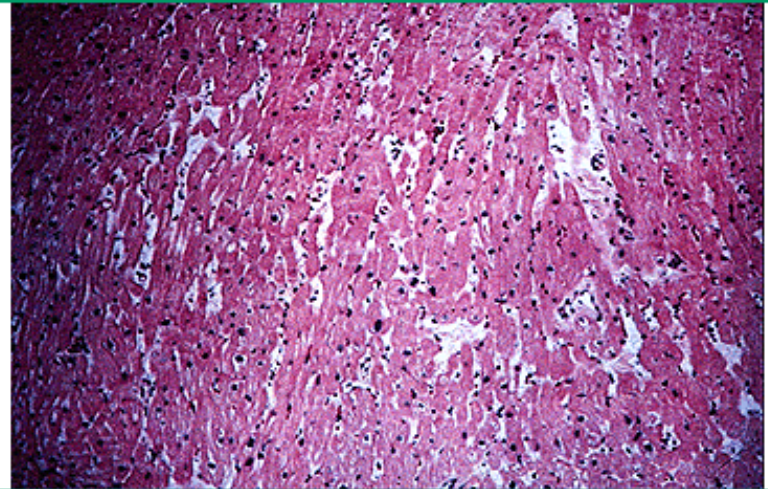


Photomicrograph demonstrating granulomatous inflammation within the myocardial wall.

*Courtesy of Om P Sharma, MD.*

## Normal endomyocardial biopsy

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Light microscopy from a normal endomyocardial biopsy.

*Courtesy of Helmut Rennke, MD.*

# Clinical Manifestations

- **Conduction abnormalities (atrioventricular block or bundle-branch block)**
- **Tachyarrhythmias**
- **Sudden cardiac death**
- **Coronary infiltration (leading to spasm or vasculitis)**
- **Cardiomyopathy**
- **Congestive heart failure**
  - **Granulomatous involvement of myocardium**
  - **Granulomatous involvement of valves, pap muscles**

# Prevalence of cardiac findings in cardiac sarcoidosis

- AV block: 26-62%
- BBB: 12-61%
- SVT: 0-15%
- Vtach: 2-42%
- HF: 10-30%
- SD: 12-65%

# Echo Findings

- Left ventricular dilatation
- Septal thinning
- Segmental or global hypokinesia of the left ventricle
- Ventricular aneurysms
- Valvular regurgitation
- Right ventricular dilatation and hypokinesia.

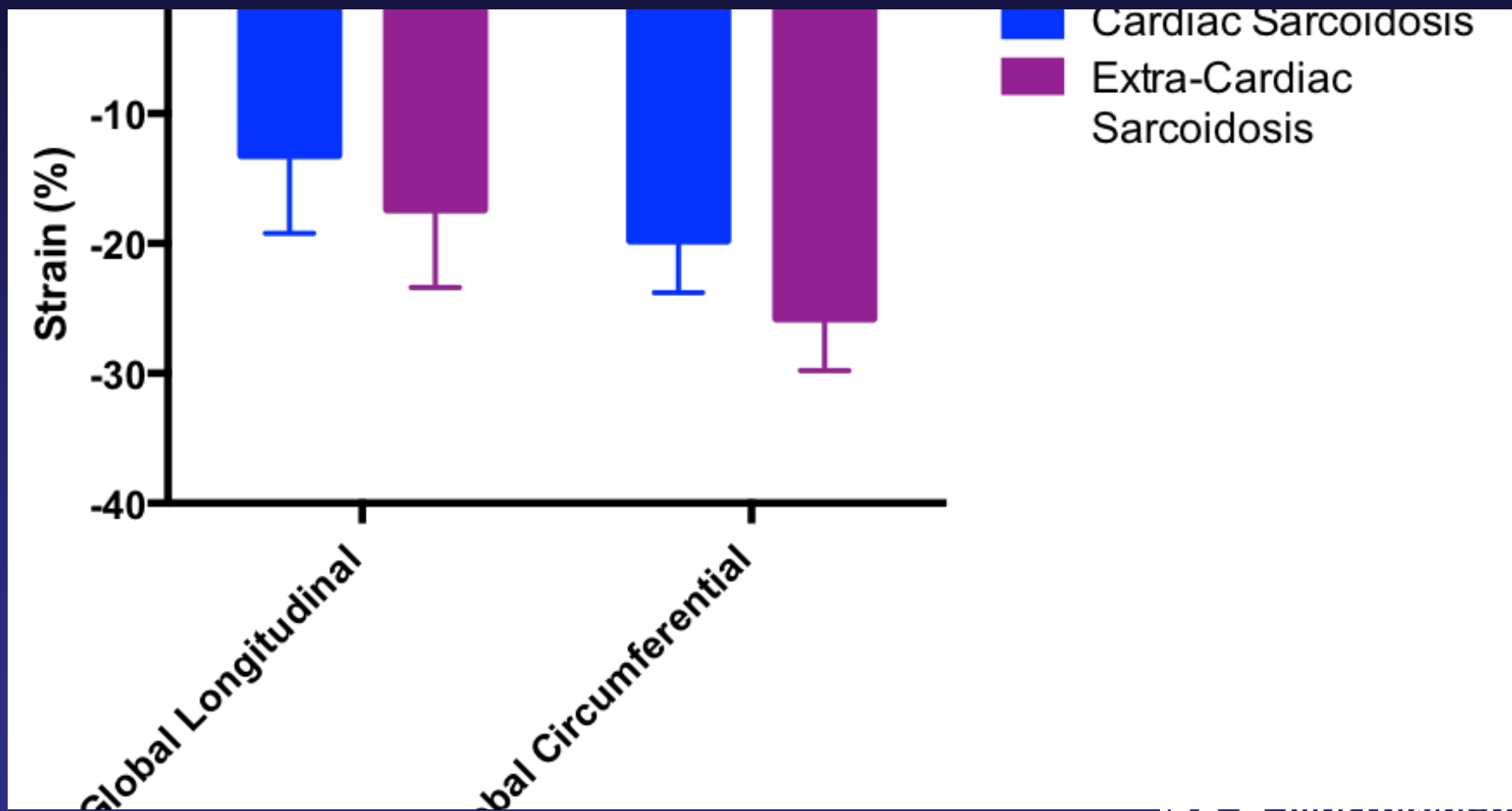
# Echo Findings

- Septum and LV free wall most commonly affected
- Increase in wall thickness simulating LVH or HCM
- Wall motion abnormalities in noncoronary distribution



# Speckle Tracking Echocardiography Identifies Patients with Cardiac Sarcoidosis

Sadiya S. Khan, MD; Jason Chodakowski, BS; Jyothy Puthumana, MD; Alex Chicos, MD

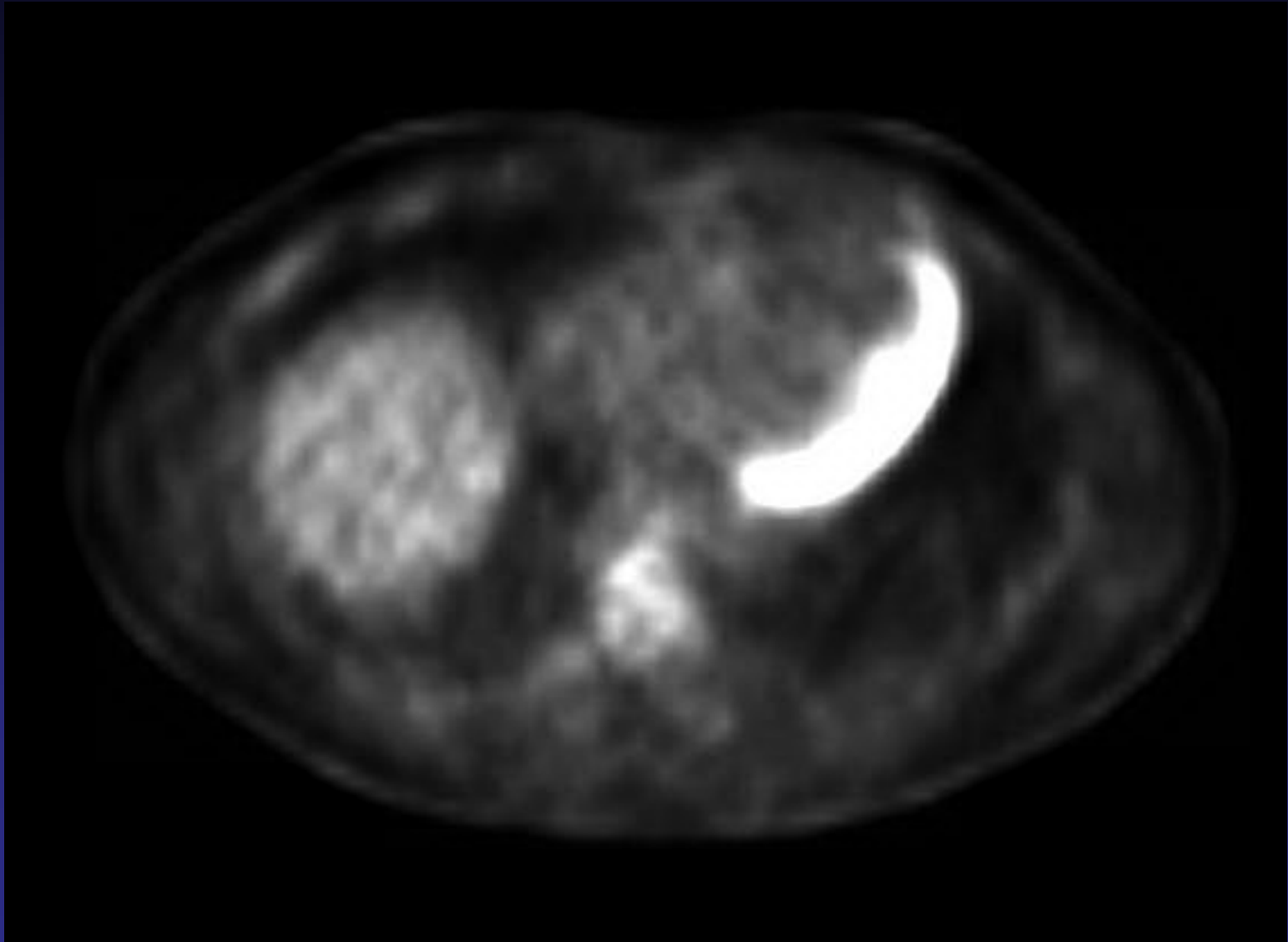


# Cardiac MRI

- T1 weighted images detect wall motion, hypertrophy, wall thinning
- T2 weighted images/early gad detect edema (inflammation)
- Late gad detects fibrosis/scar

# **$^{18}\text{F}$ -Fluorodeoxyglucose (FDG) PET**

- Detects active sarcoidosis with high sensitivity
- May be positive in other inflammatory cardiac conditions



# Radionuclide Imaging: Thallium-201

- Focal perfusion deficits may be seen at rest
- With exercise, “reverse redistribution” is seen
- Fixed defects may represent scar
- Gallium-67 can detect active inflammation

# Accuracy of Diagnostic Tests

<b>Diagnostic modality</b>	<b>Sensitivity</b>	<b>Specificity</b>
ECG	Low	Low
Echocardiography	Low to moderate	Low
$^{201}\text{Tl}$ or $^{99\text{m}}\text{Tc}$ scintigraphy	Moderate	Moderate
$^{67}\text{Ga}$ scintigraphy	Low	High
$^{18}\text{F}$ -FDG PET	High	Moderate to high
MRI	Moderate to high	High

Kim JS et al. Am Heart J 2009;157:9-21.

# Guidelines for Diagnosis of Cardiac Sarcoidosis

## Histologic diagnosis

Endomyocardial biopsy demonstrating noncaseating epithelioid granulomas

## Clinical diagnosis

Among patients with a histologic diagnosis of extracardiac sarcoidosis, cardiac sarcoidosis should be suspected when criterion "a" and at least one of criteria "b" to "e" is present in patients in whom other causes such as hypertension and coronary heart disease have been excluded:

- a. Complete right bundle branch block, AV block, ventricular tachycardia, ventricular premature beats or pathologic Q wave or ST-T changes on the electrocardiogram
- b. Abnormal wall motion, regional wall thickening, or left ventricular dilation
- c. Perfusion defect on myocardial perfusion imaging or abnormal accumulation of <sup>67</sup>Gallium citrate or <sup>99m</sup>Tc-PYP myocardial scintigraphy
- d. Abnormal intracardiac pressure, low cardiac, or abnormal wall motion or reduced ejection fraction of the left ventricle
- e. On endomyocardial biopsy, interstitial fibrosis or more than moderate cellular infiltration, even if the findings are nonspecific

*Japanese Ministry of Health and Welfare 1993*

# 2006 Revised Guidelines by the Japanese Society of Sarcoidosis and Other Granulomatous Disorders

## 1. Histological diagnosis

Cardiac sarcoidosis is confirmed when cardiac biopsy specimens demonstrate noncaseating epithelioid cell granuloma with histologic or clinical diagnosis of extracardiac sarcoidosis.

## 2. Clinical diagnosis group

Cardiac sarcoidosis is diagnosed in the absence of an endomyocardial biopsy specimen or in the absence of typical granulomas on cardiac biopsy when extracardiac sarcoidosis has been proven and a combination of major or minor diagnostic criteria has been satisfied as follows.

1. More than 2 of 4 major criteria are satisfied, OR
2. 1 of the 4 major criteria and 2 or more of the minor criteria are satisfied.

### Major criteria

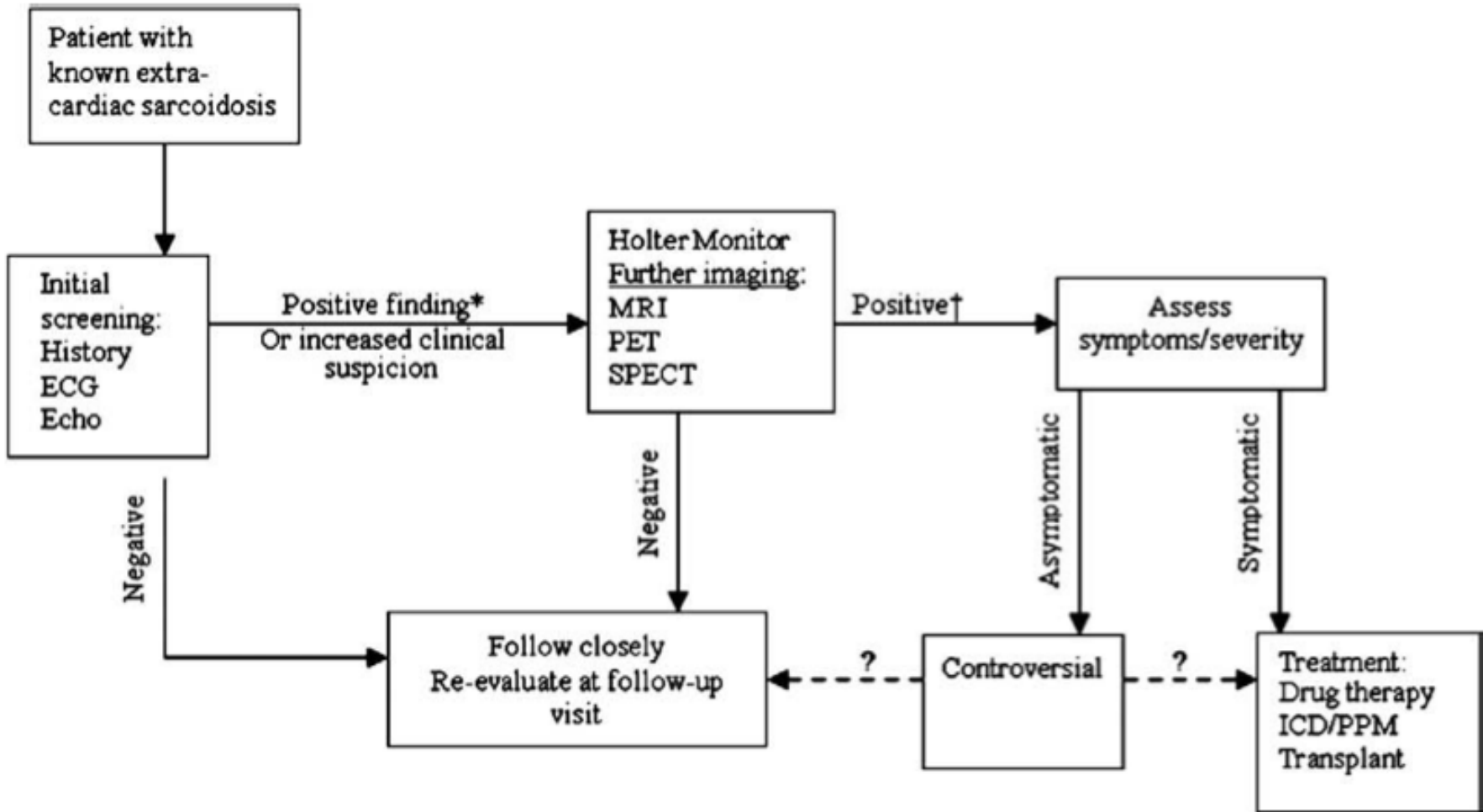
- (a) Advanced AV block
- (b) Basal thinning of the ventricular septum
- (c) Positive cardiac gallium uptake
- (d) Left ventricular ejection fraction less than 50%

### Minor criteria

- (a) Abnormal electrocardiogram findings including ventricular tachycardia, multifocal frequent premature ventricular contractions, complete right bundle branch block pathologic Q waves, or abnormal axis deviation
- (b) Abnormal echocardiogram demonstrating regional wall motion abnormalities, ventricular aneurysm, or unexplained increase in wall thickness
- (c) Perfusion defects detected by myocardial scintigraphy
- (d) Delayed gadolinium enhancement of the myocardium on cardiac MRI scanning
- (e) Interstitial fibrosis or monocyte infiltration greater than moderate grade by endomyocardial biopsy



# Suggested Clinical Algorithm



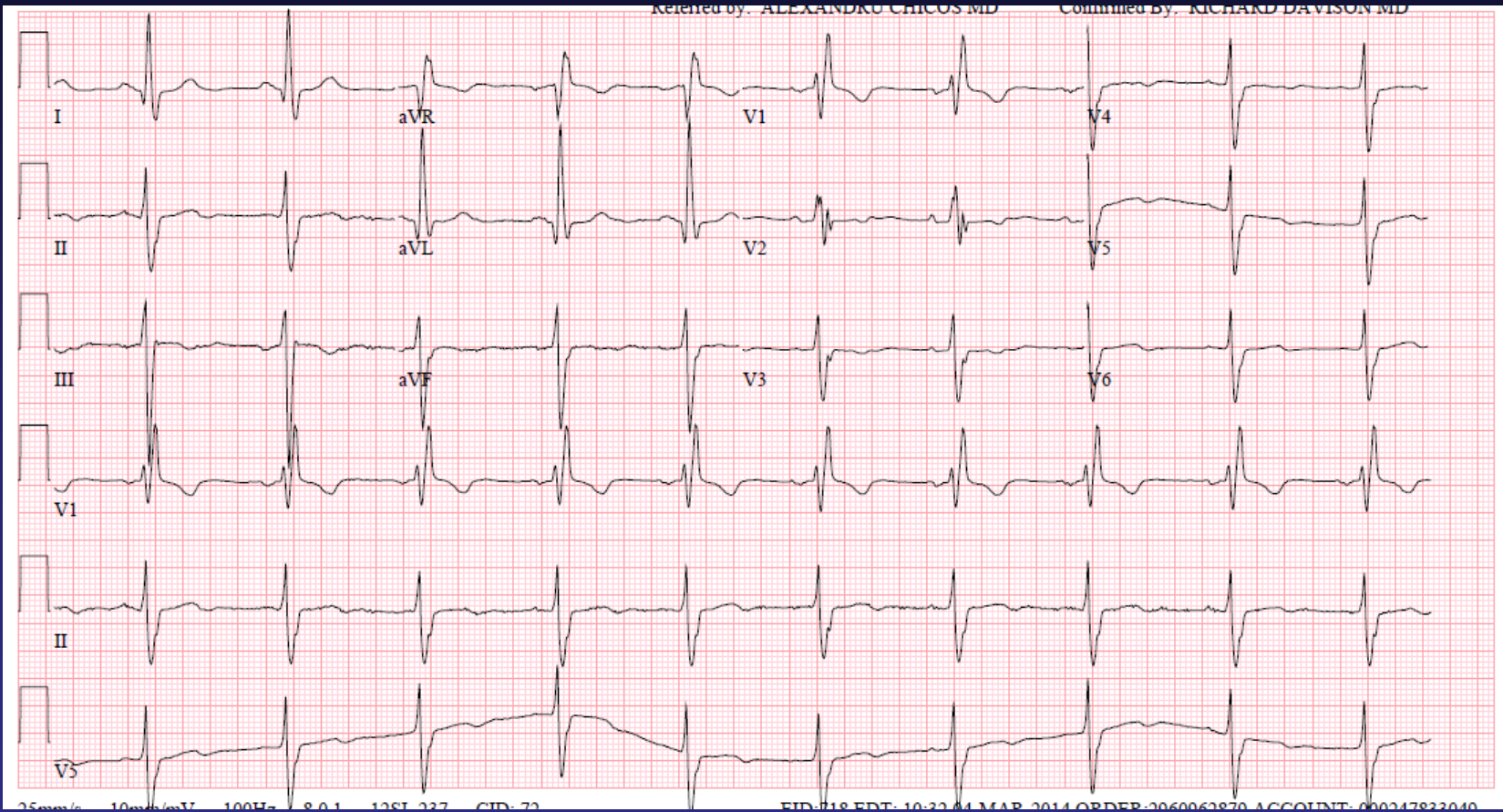
Kim JS et al. Am Heart J 2009;157:9-21.

# Case 1

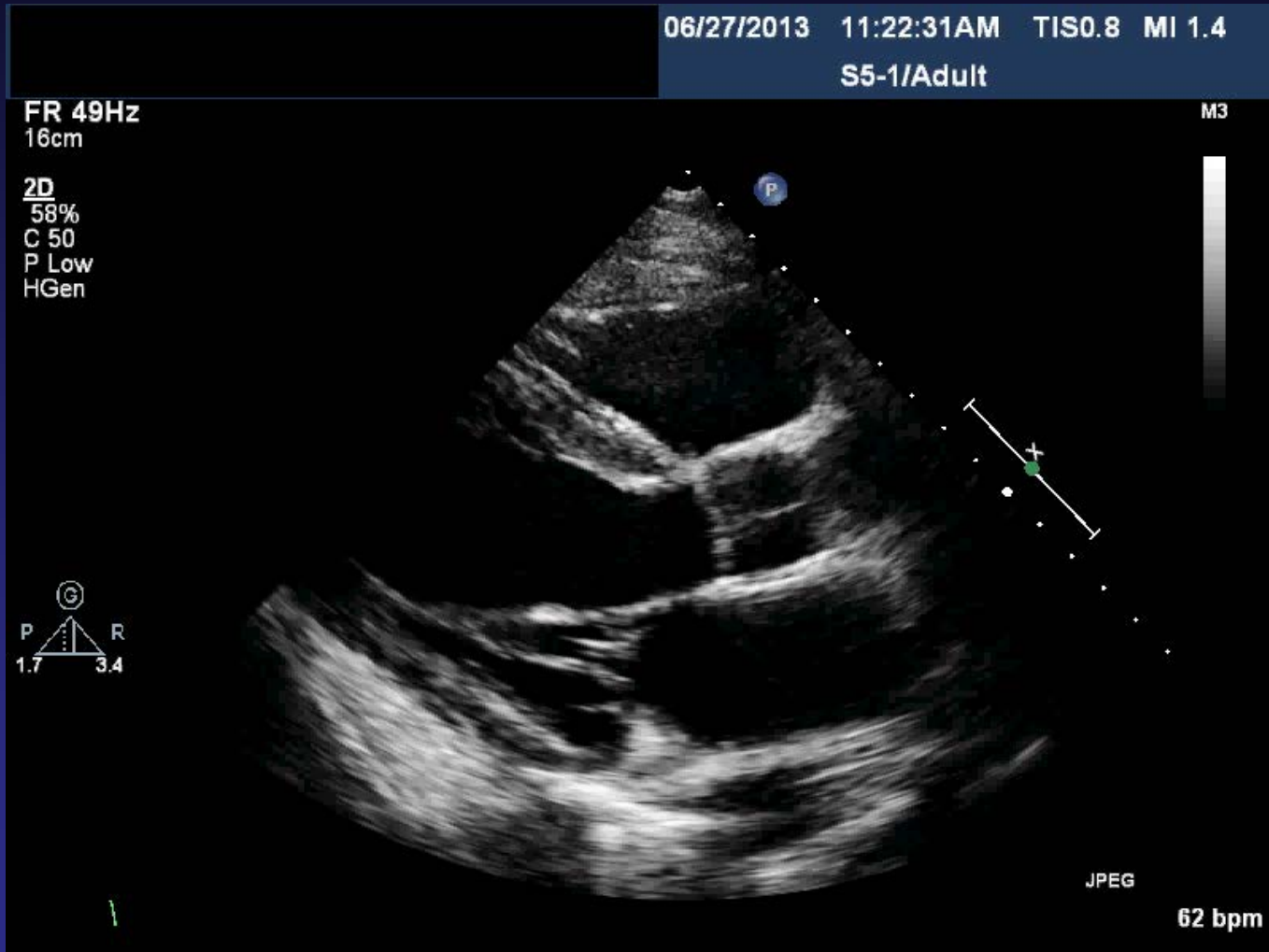
- 48 yr old female who complains of palpitations for 1 month
- She presented to the ER when palpitations occurred with lightheadedness
- Initial rhythm was Afib. She converted to SR. VT then noted
- CT ordered to look for PE: Lymphadenopathy noted

- Endobronchial biopsy: Sarcoidosis
- Endomyocardial biopsy: Negative
- Cardiac cath: No CAD

# ECG



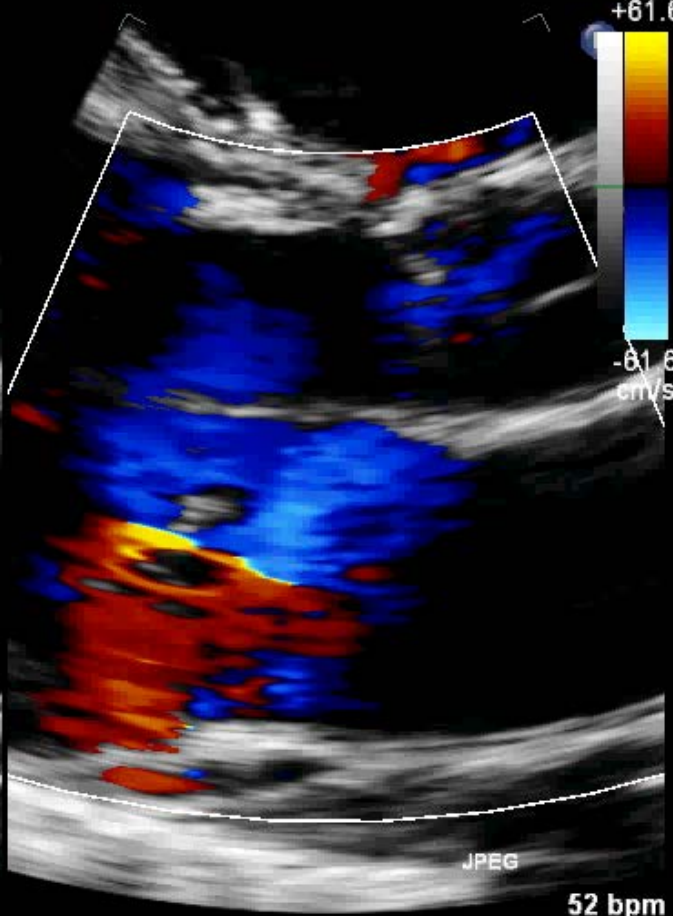
# Echo



06/27/2013 11:23:03AM TIS2.4 MI 1.2  
S5-1/Adult

FR 14Hz  
15cm

2D  
53%  
C 50  
P Low  
HGen  
CF  
66%  
2.5MHz  
WF High  
Med



M3 M4  
+61.6  
-61.6  
cm/s

52 bpm

06/27/2013 11:27:57AM TIS0.8 MI 1.4  
S5-1/Adult

FR 50Hz  
15cm

2D  
58%  
C 50  
P Low  
HGen

M3



JPEG

68 bpm

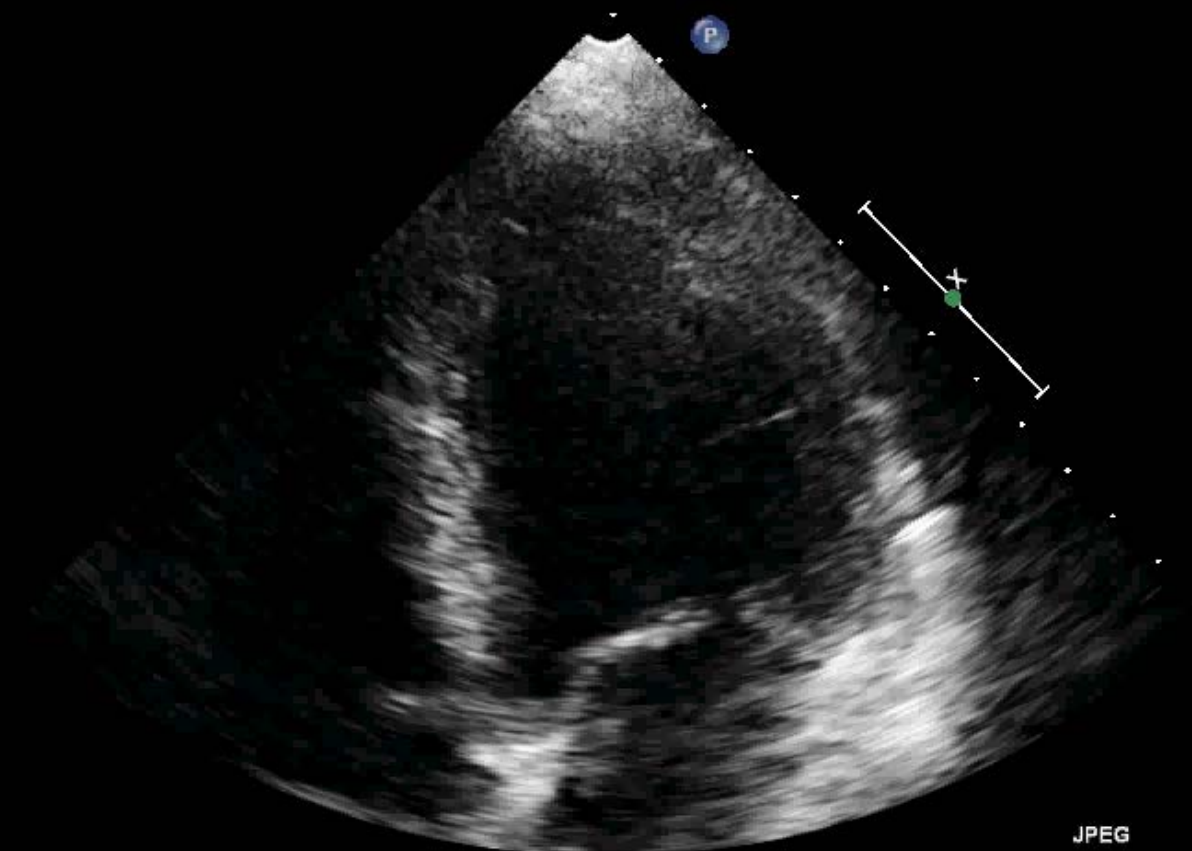
western

06/27/2013 11:31:59AM TIS0.7 MI 1.3  
S5-1/Adult

FR 55Hz  
13cm

M3

2D  
68%  
C 50  
P Low  
HGen



JPEG

63 bpm



FR 17Hz  
18cm

2D  
69%  
C 50  
P Low  
HGen

MV Decel Time 127 ms  
MV Peak A Vel  
Vel 28.6 cm/s  
PG 0 mmHg

MV Peak E Vel  
Vel 80.0 cm/s  
PG 3 mmHg

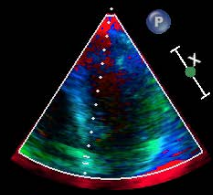


M3 M4  
+61.4  
FR 92Hz  
13cm

PW  
50%  
1.6MHz  
WF 125Hz  
SV4.0mm  
8.9cm

2D  
87%  
C 35  
P Low  
HGen

TDI  
89%  
-613.4MHz  
cm

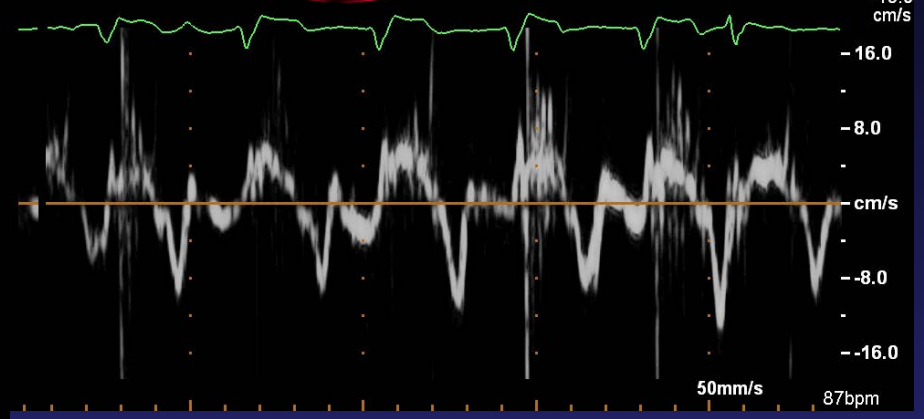
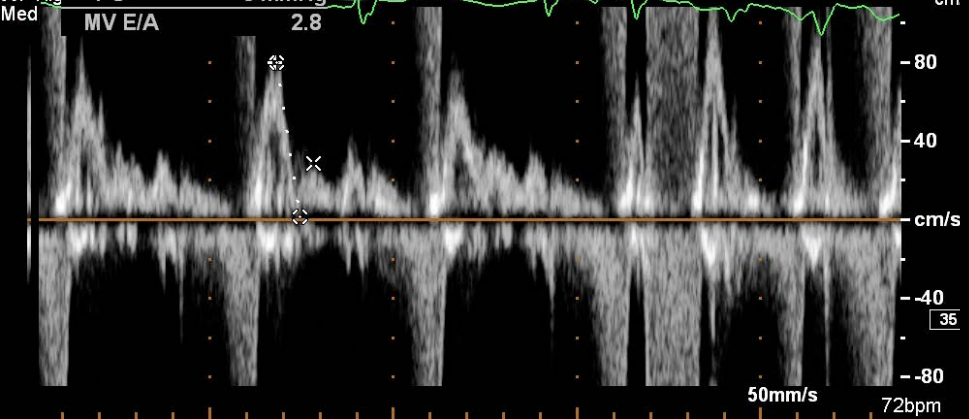


M3 M6  
+15.0

PW  
50%  
3.6MHz

SV5.0mm  
11.0cm

-15.0  
cm/s



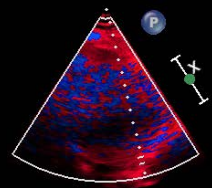
$E/e' = 20$

Vel 9.12 cm/s  
PG 0 mmHg

Vel 4.27 cm/s  
PG 0 mmHg

C 35  
P Low  
HGen

TDI  
89%  
3.4MHz



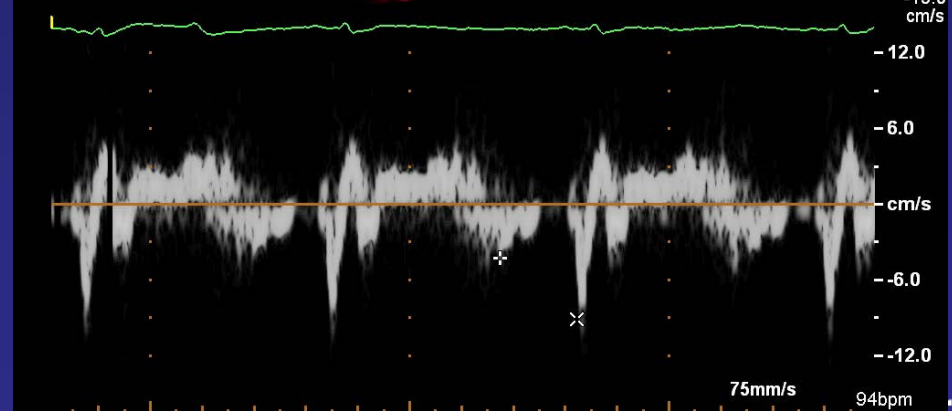
-0:36:01

M3 M6  
+15.0

PW  
55%  
3.6MHz

SV5.0mm  
12.0cm

-15.0  
cm/s

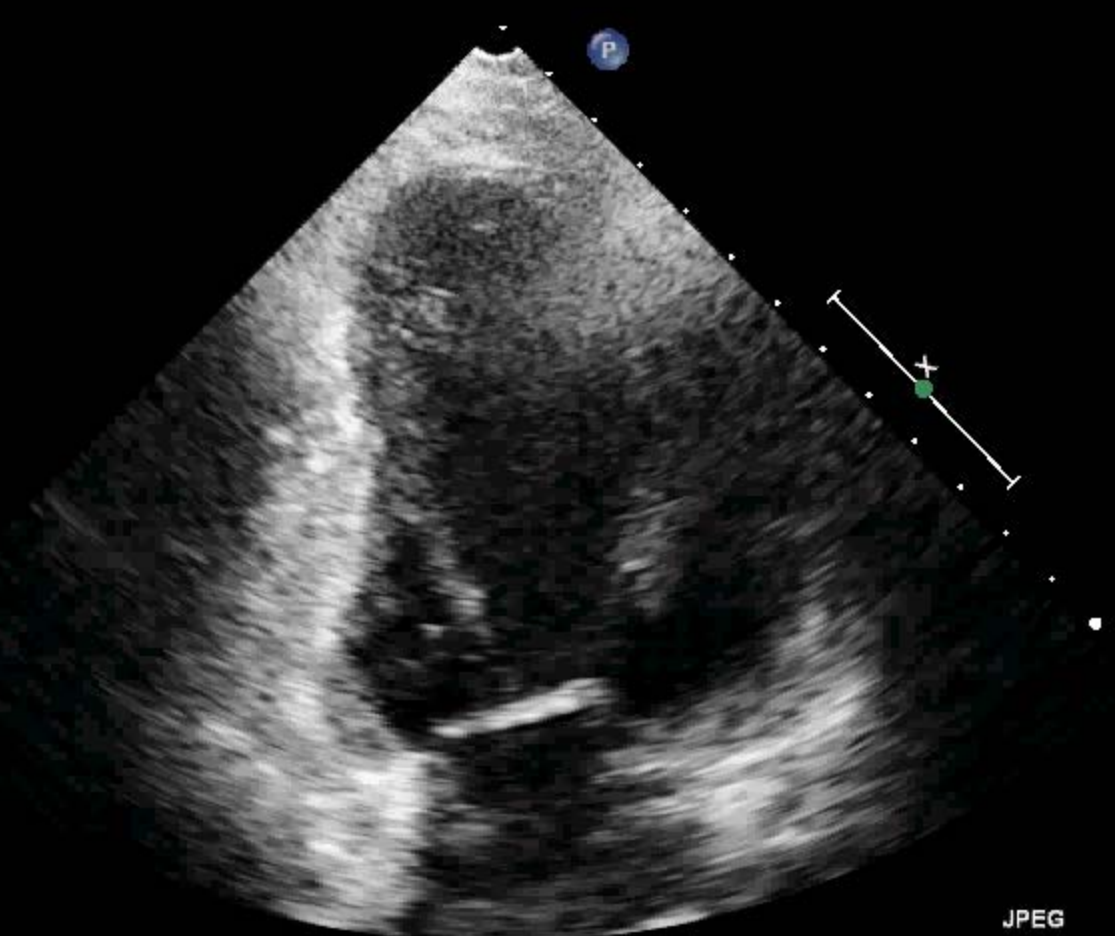


06/27/2013 11:36:24AM TISO.8 MI 1.4  
S5-1/Adult

FR 53Hz  
14cm

M3

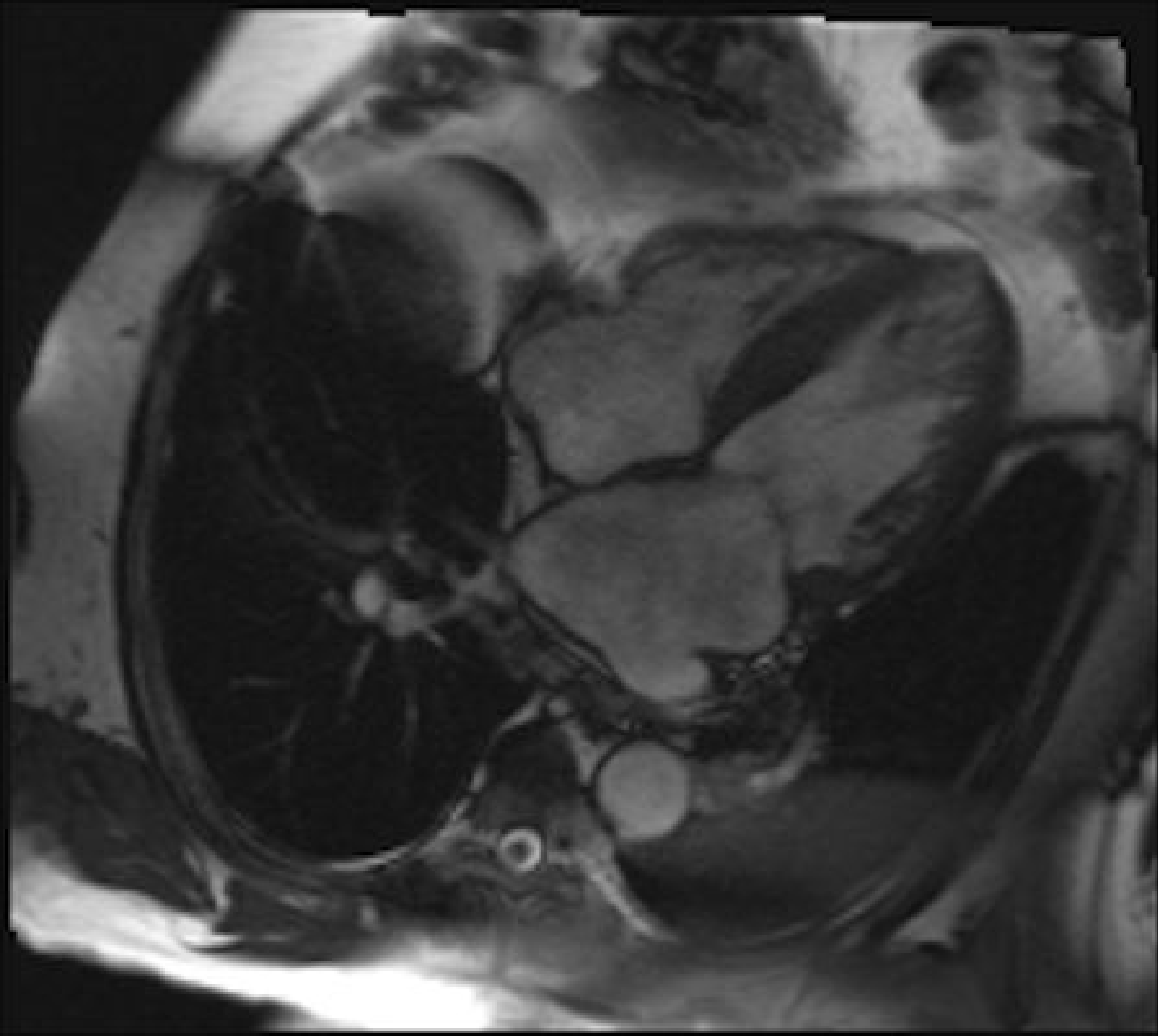
2D  
68%  
C 50  
P Low  
HGen

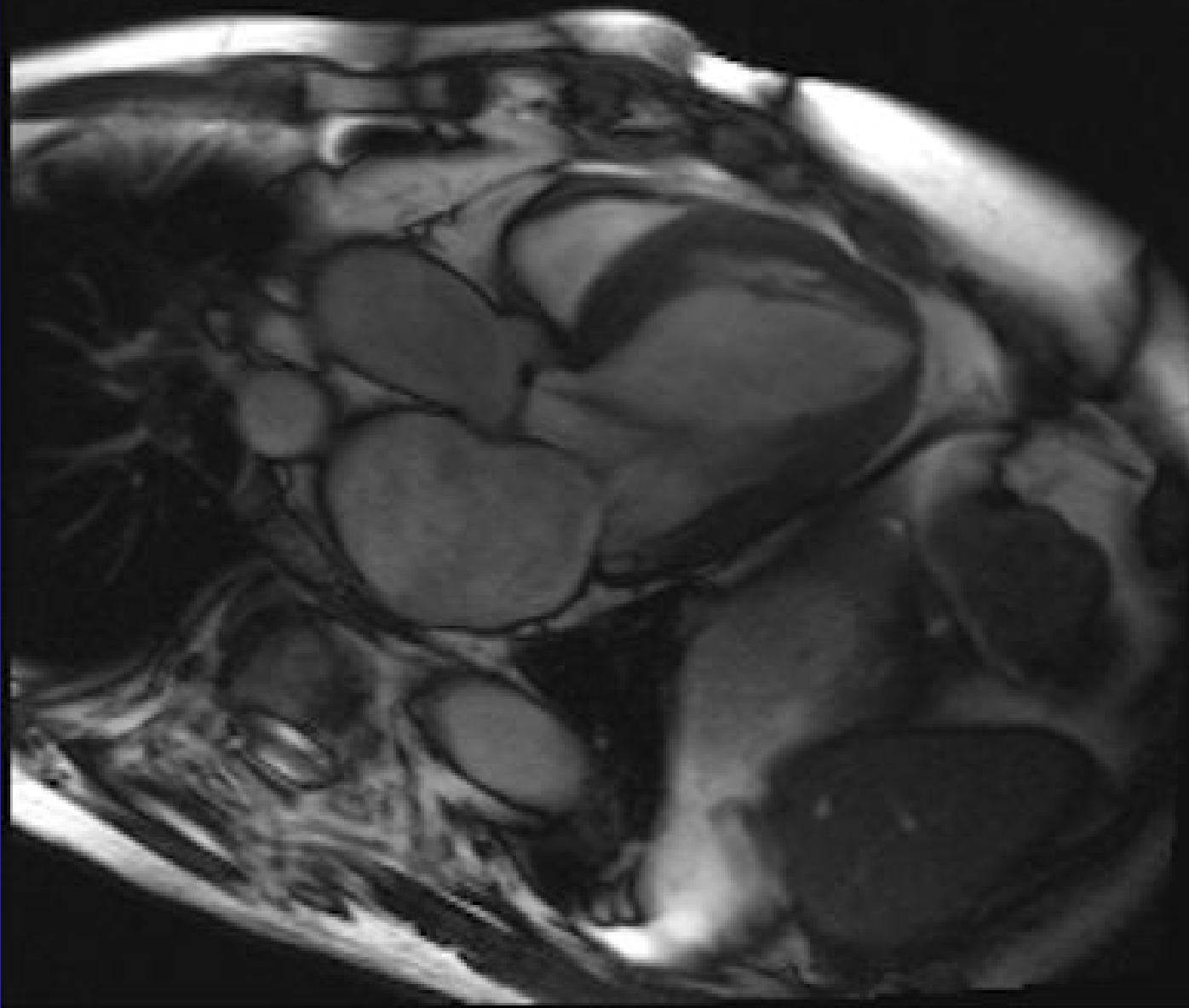


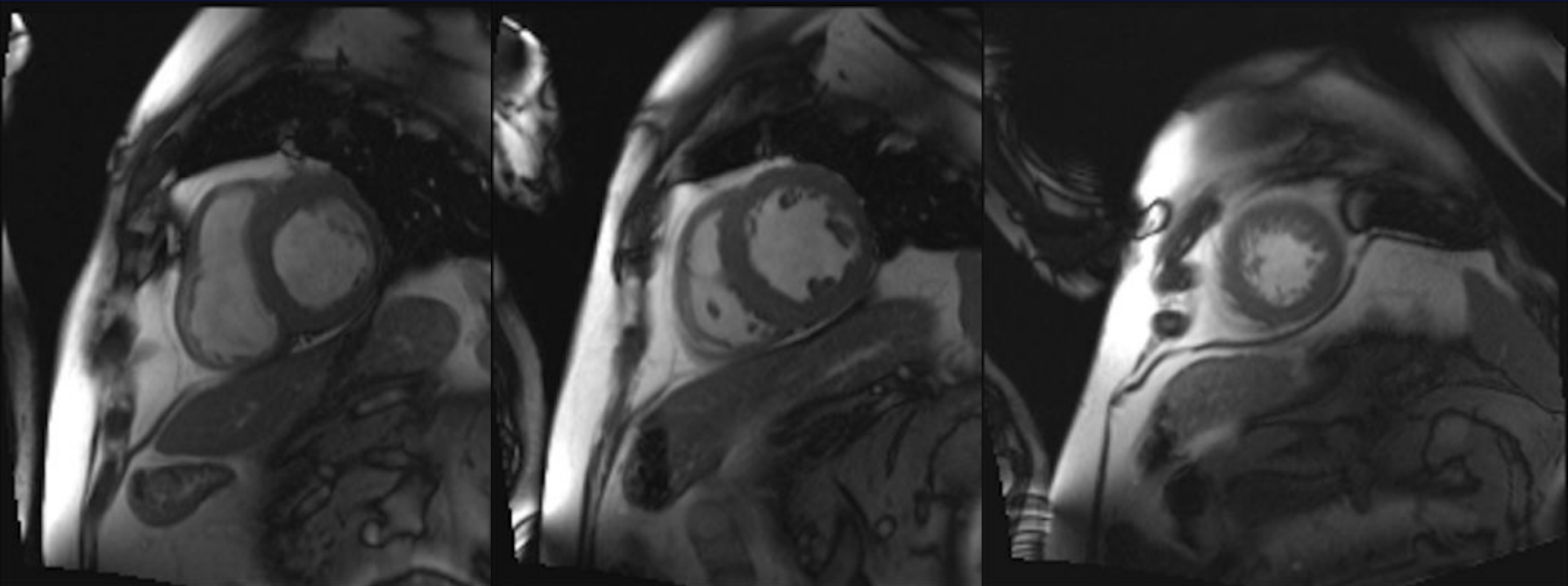
JPEG

117 bpm

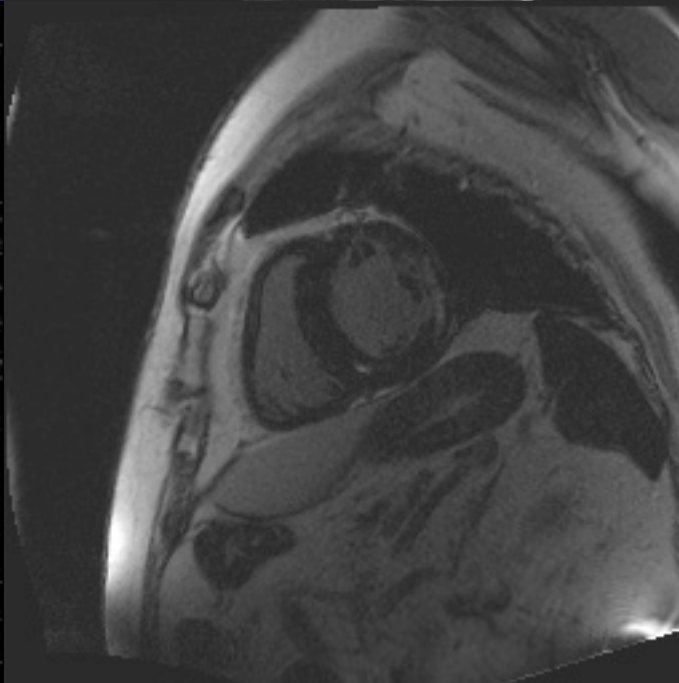
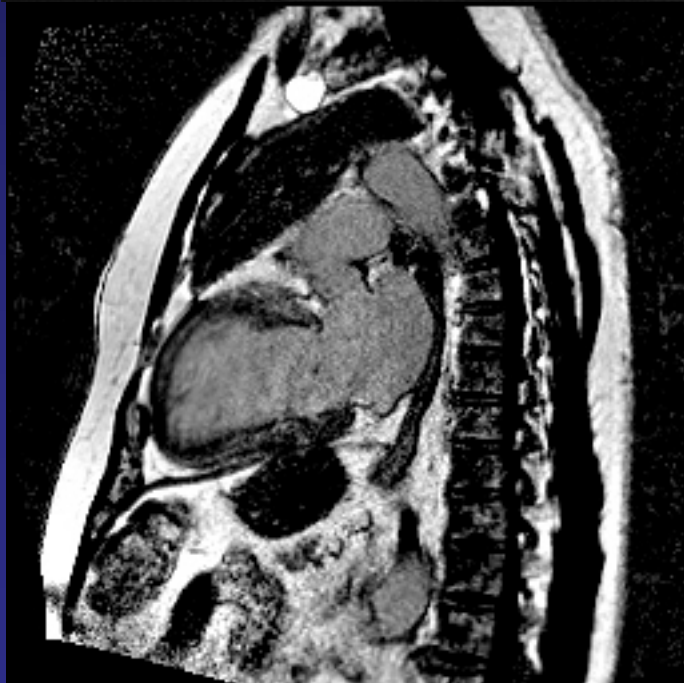
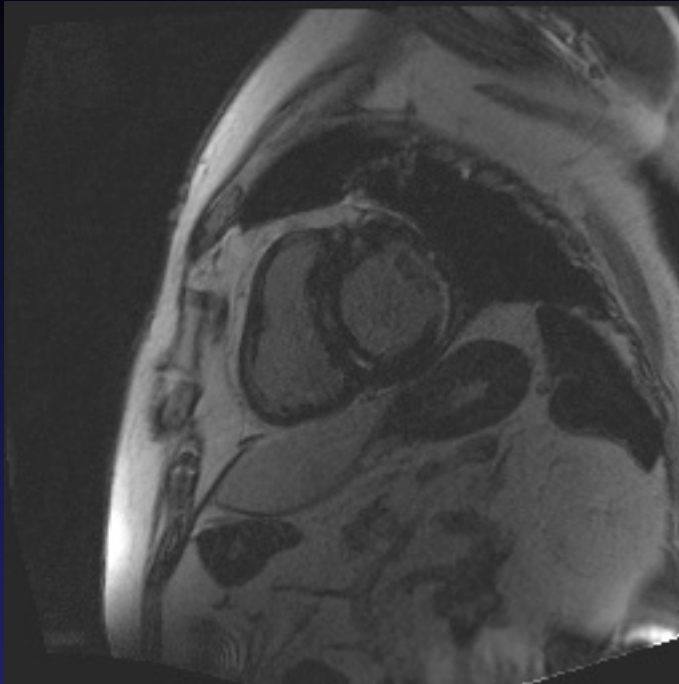












# Treatment

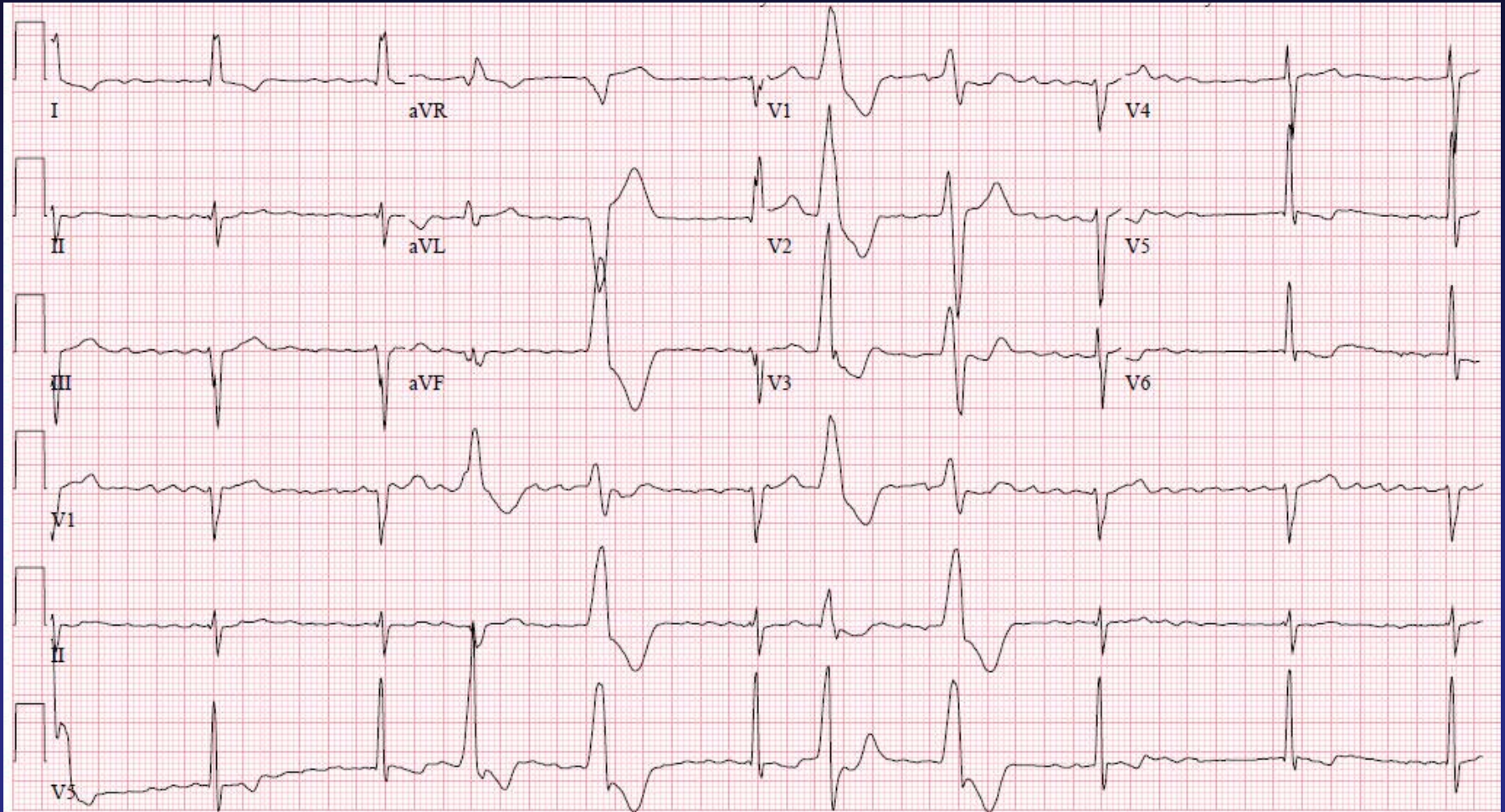
- Steroids
- Antiarrhythmic therapy
- ICD
- VT ablation (VT noted to come from multiple foci, including epicardium and paps)



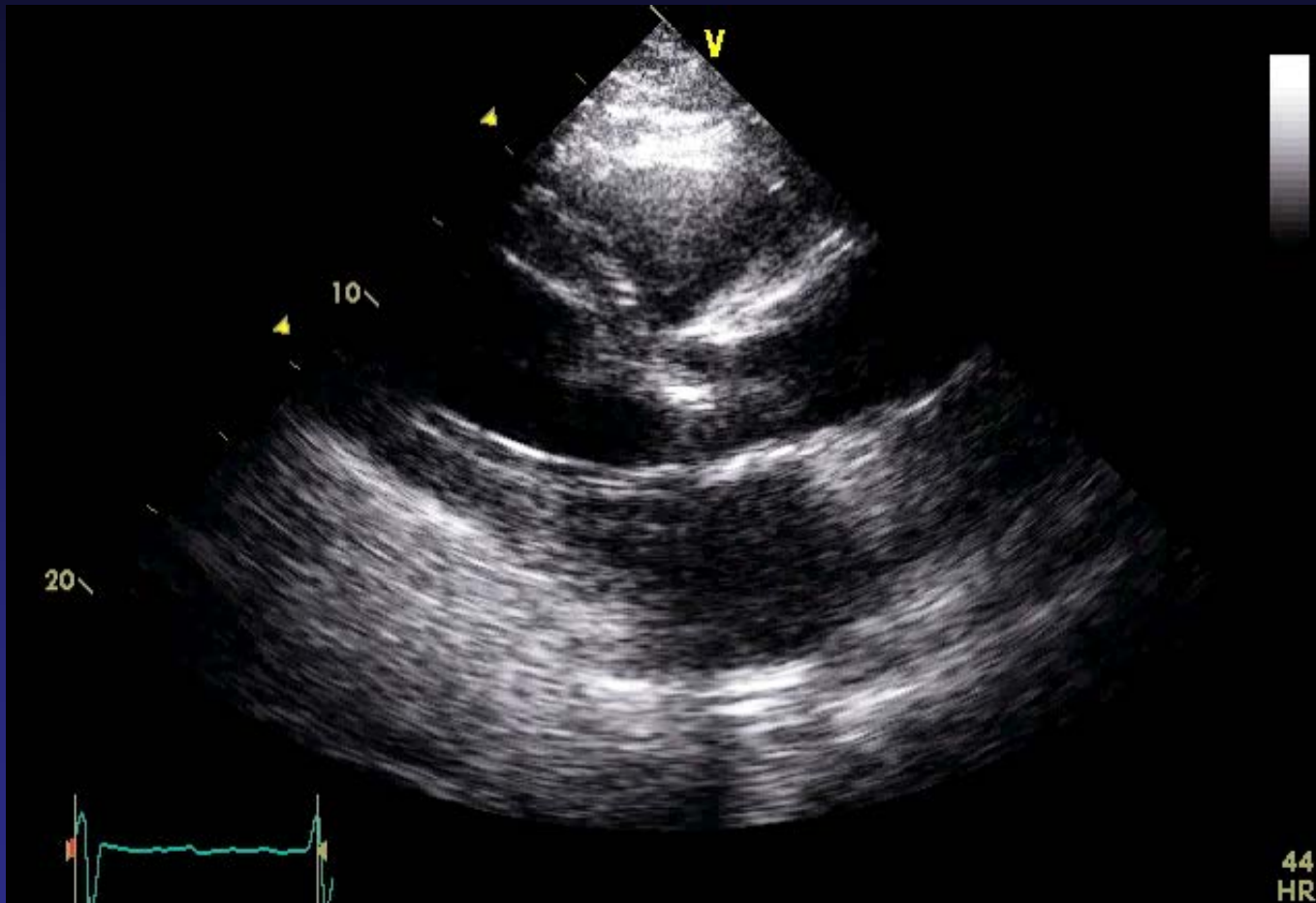
## Case 2

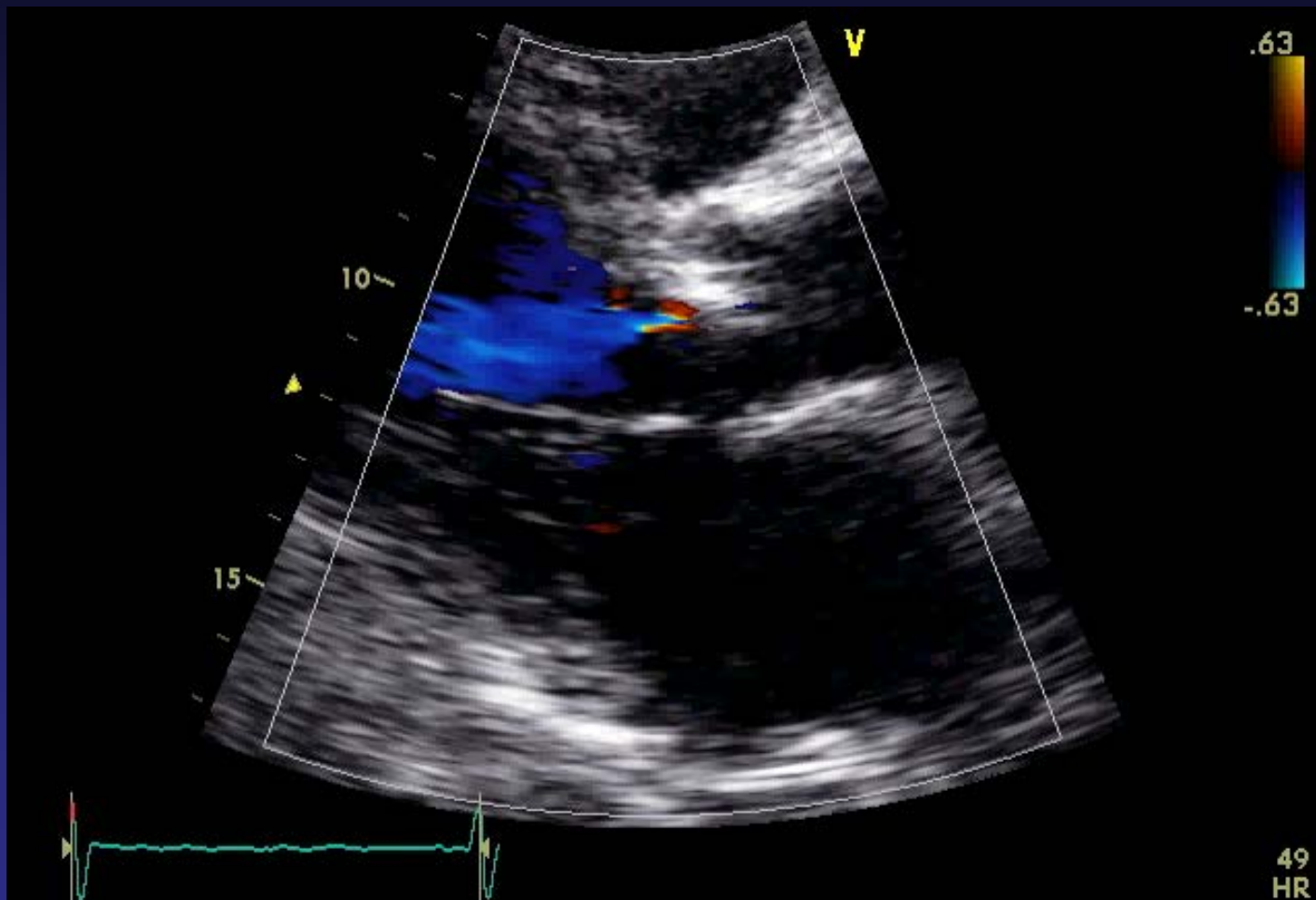
- 75 yr old male with a history of pulmonary sarcoidosis
- He complained of SOB and palpitations
- ECG showed Afib
- Cardiac cath: No obstructive CAD
- Endomyocardial biopsy: Negative

# ECG



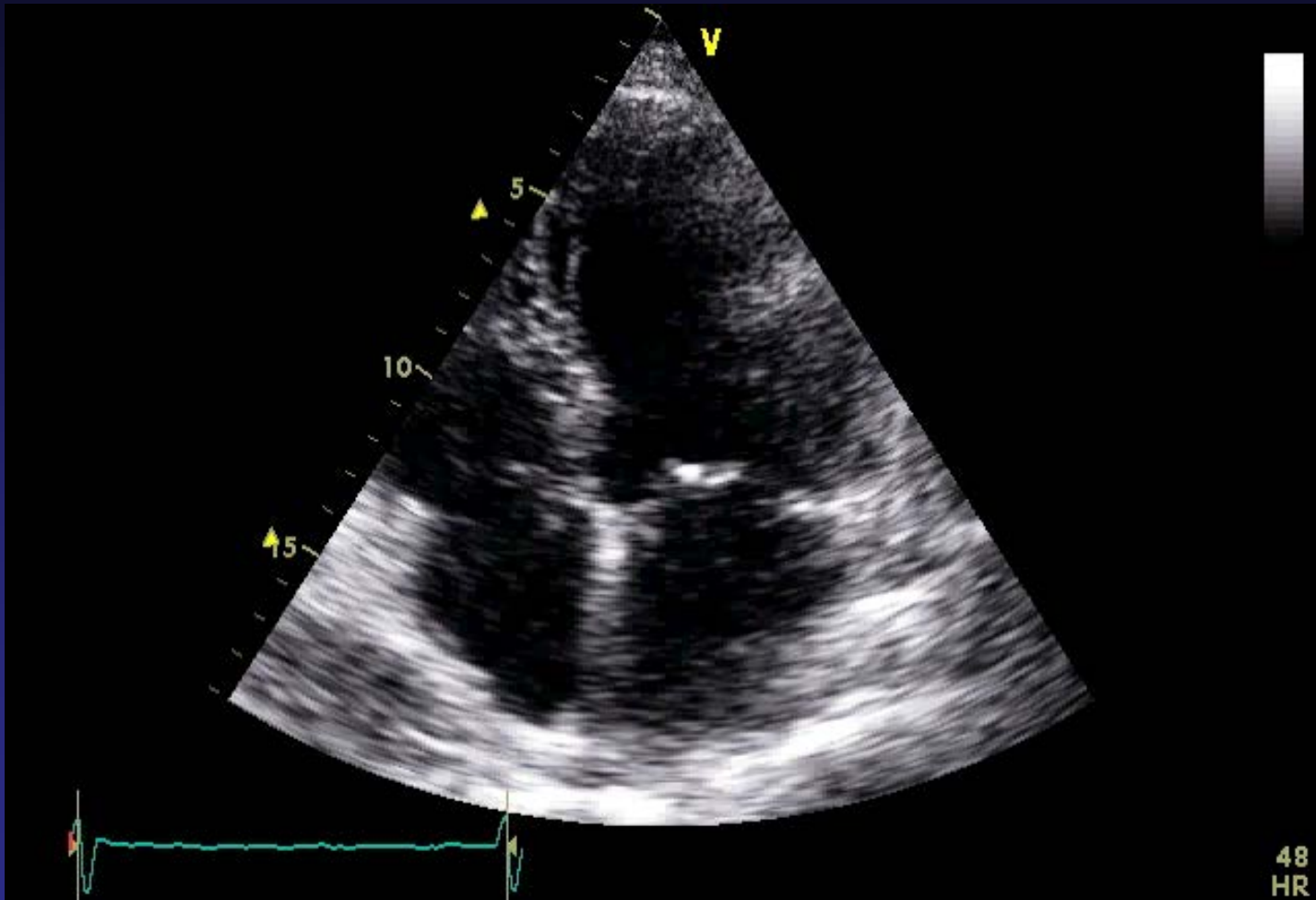
# Echo



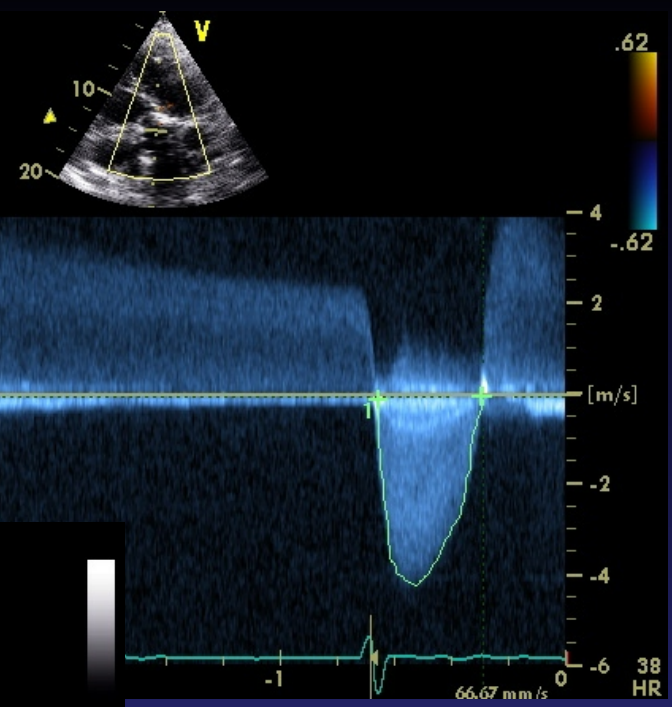




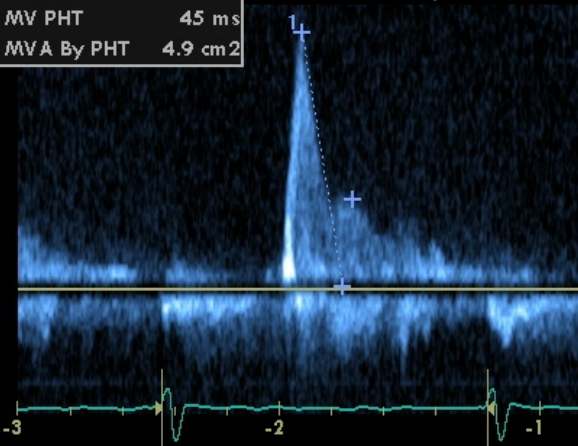
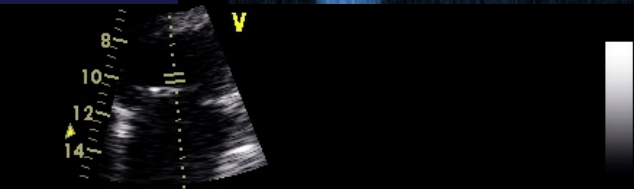




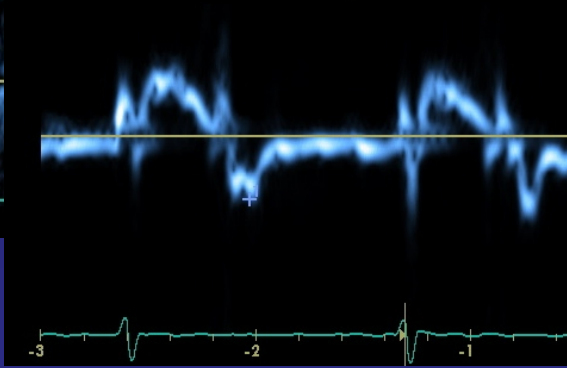
v	0.06 m/s
p	0.01 mmHg
AVA Vmax	0.3 cm <sup>2</sup>
AVA (VTI)	0.3 cm <sup>2</sup>
1 AV Vmax	4.23 m/s
AV Vmean	3.06 m/s
AV maxPG	71.53 mmHg
AV meanPG	42.51 mmHg
AV VTI	110.2 cm
AV Env.Ti	360 ms



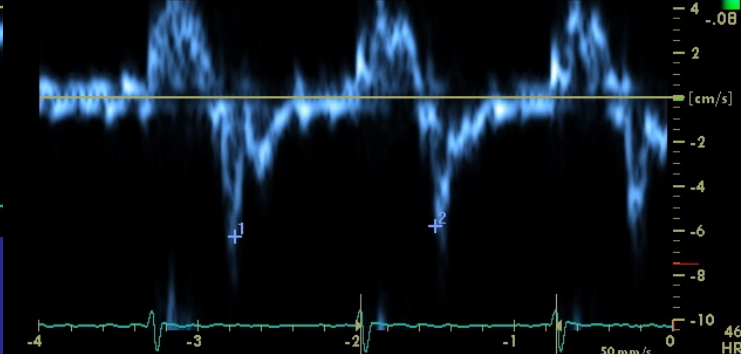
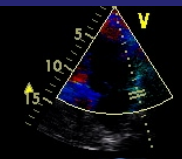
1 MV E Vel	1.47 m/s
MV DecT	156 ms
MV Dec Slope	9.4 m/s <sup>2</sup>
MV A Vel	0.51 m/s
MV E/A Ratio	2.88
MV PHT	45 ms
MVA By PHT	4.9 cm <sup>2</sup>



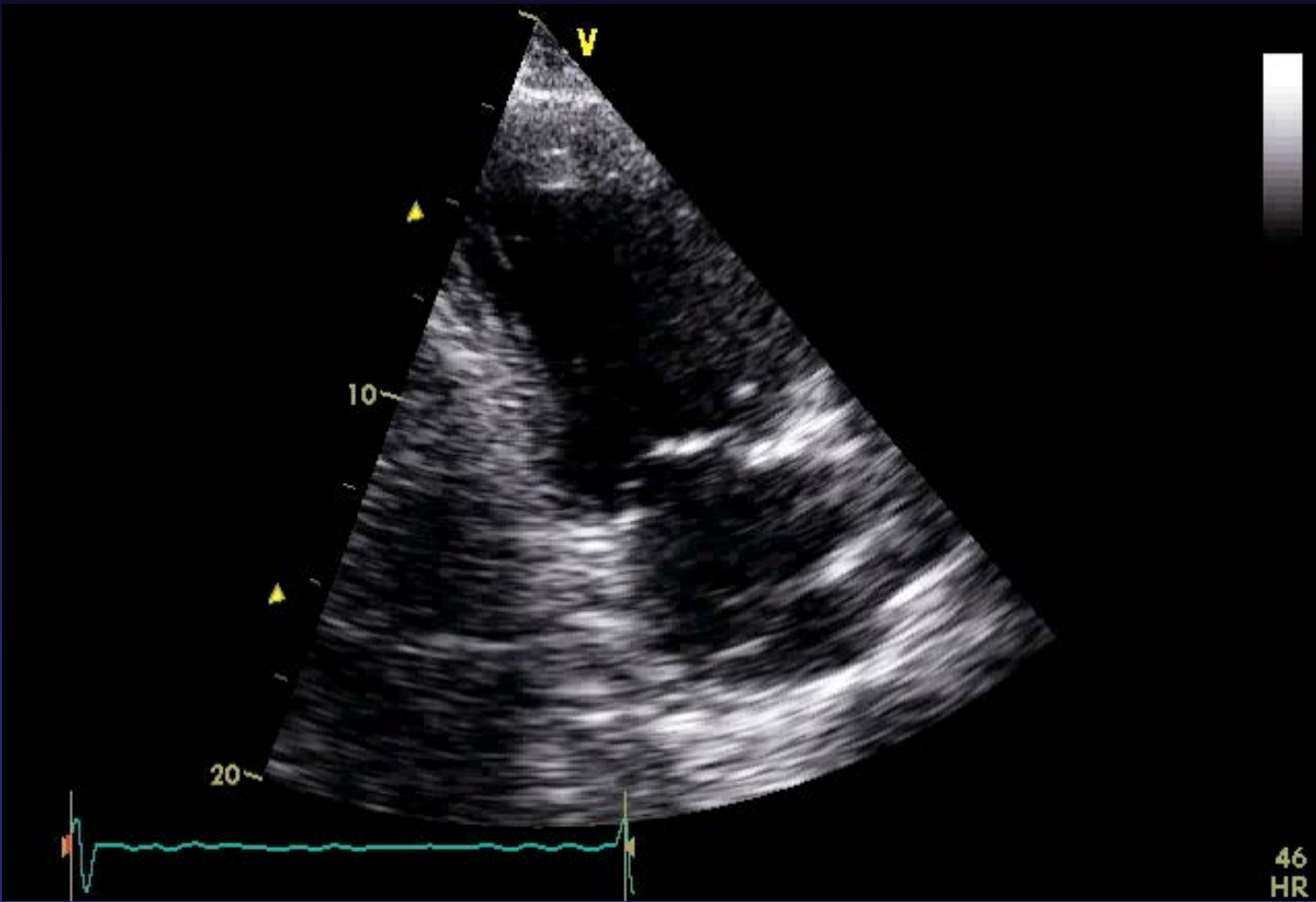
1 Sep E' 0.03 m/s
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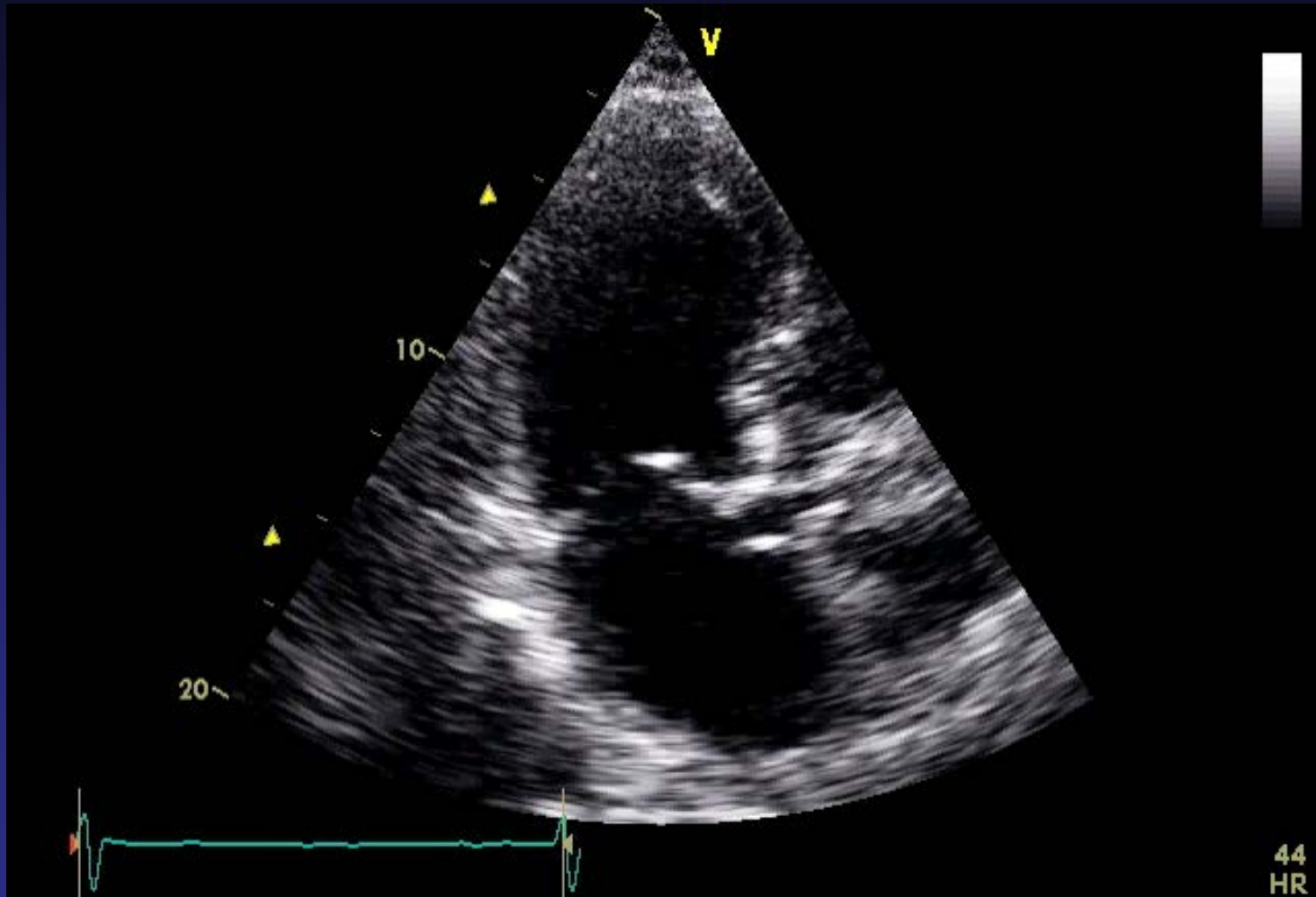
2 Lat E' 0.06 m/s
1 Lat E' 0.06 m/s

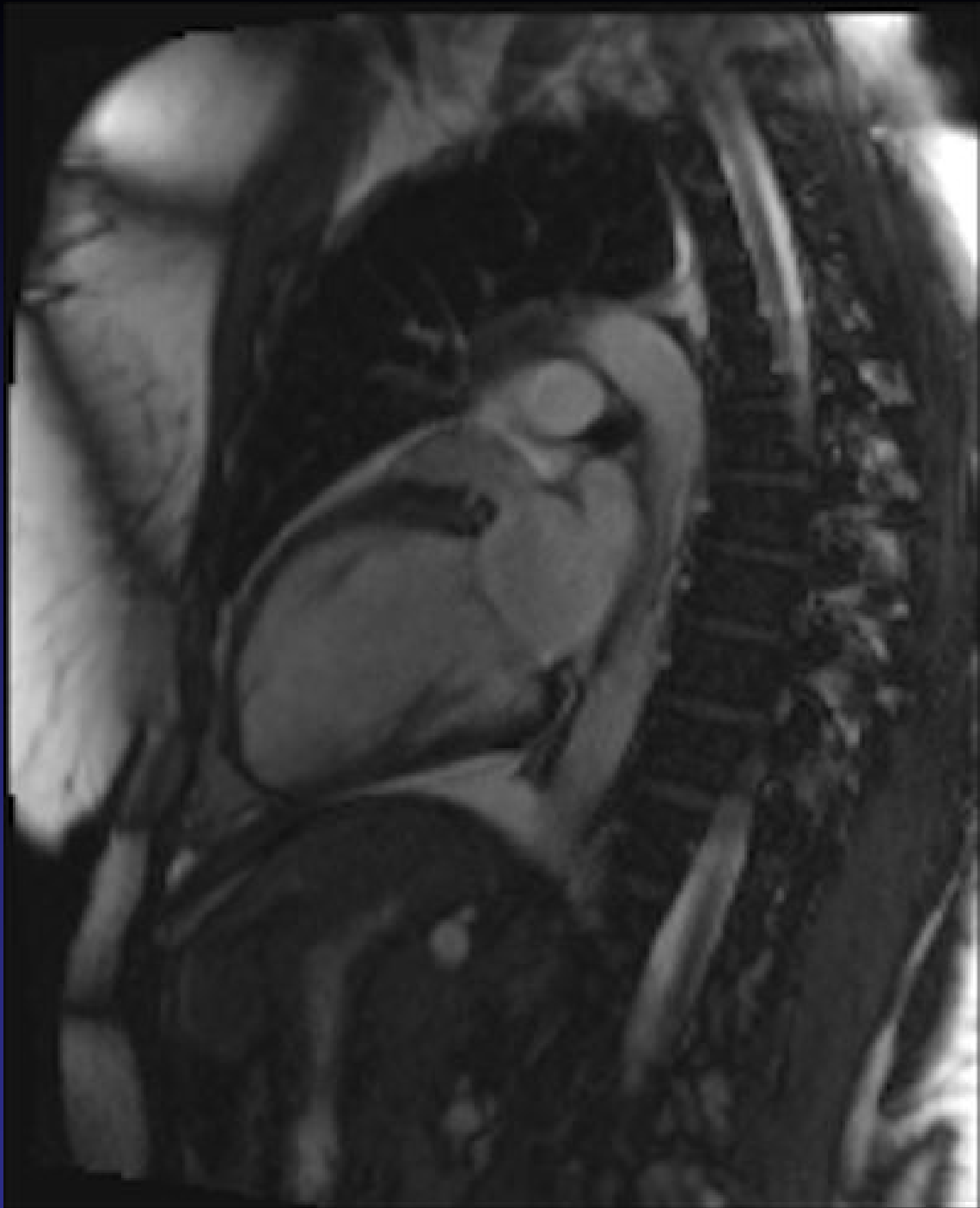


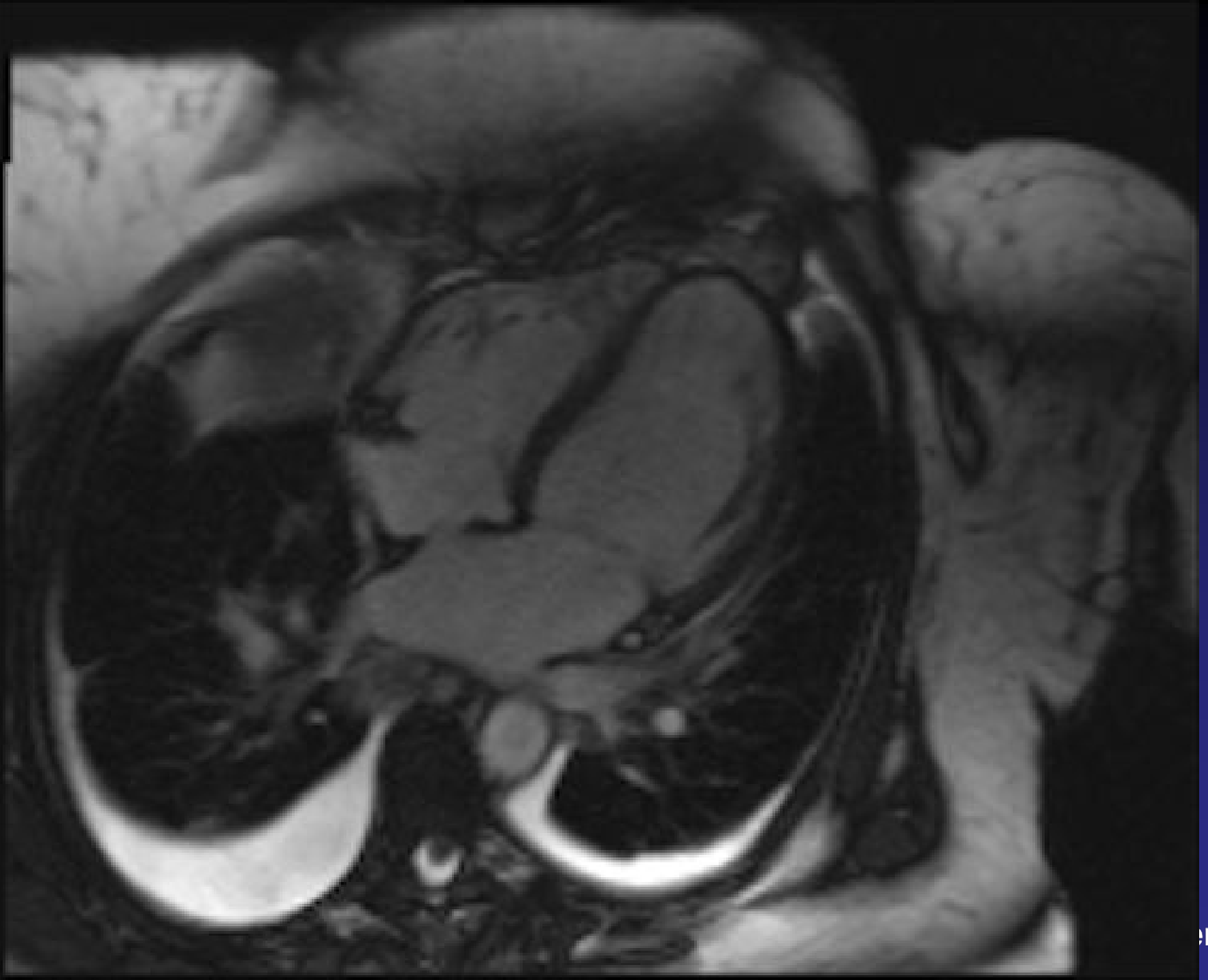
$E/e' = 33$

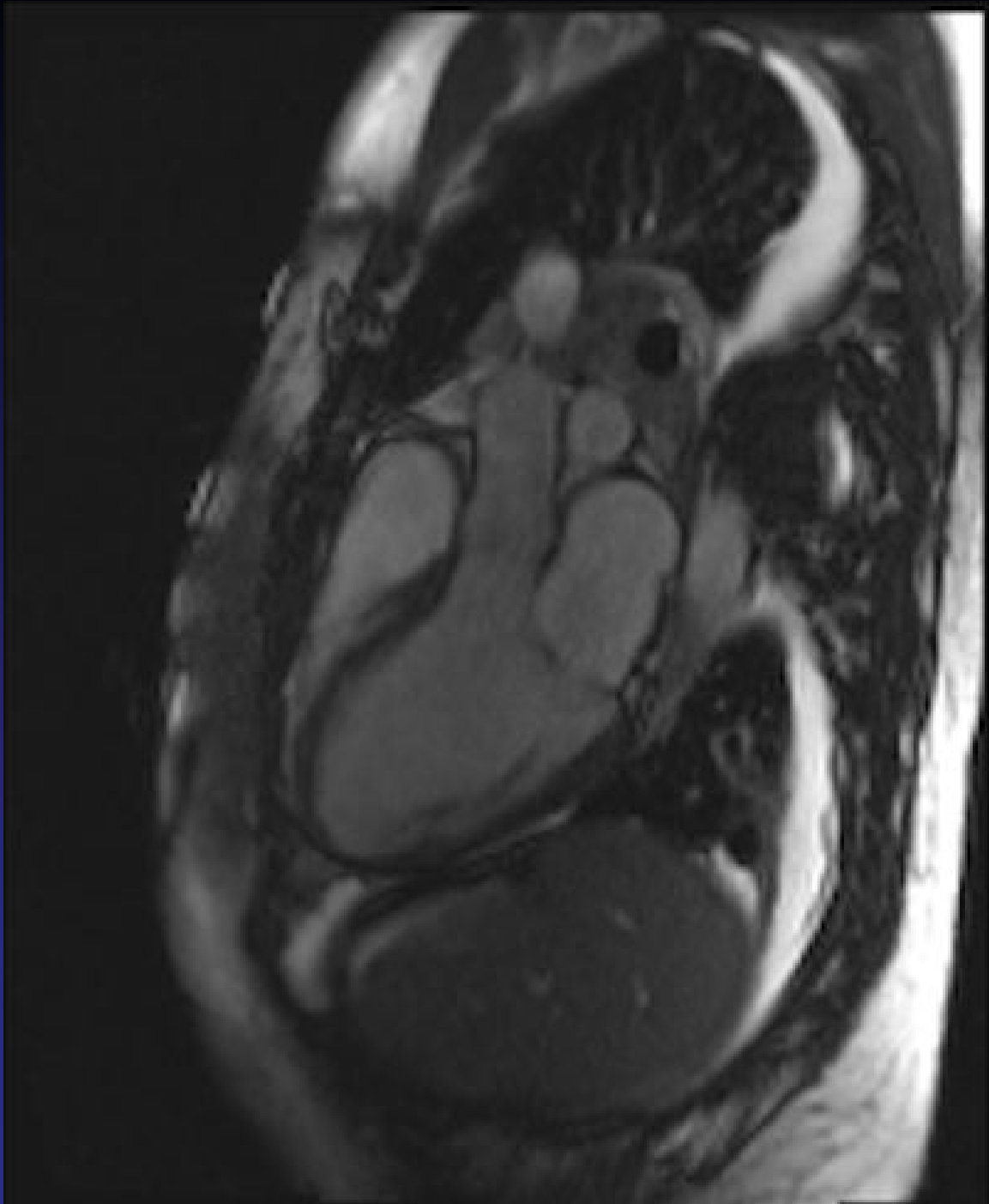


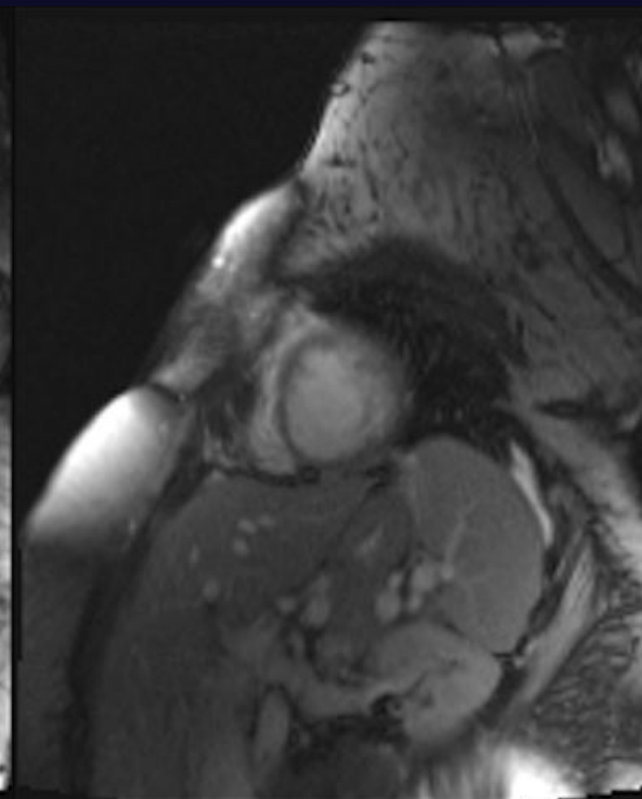
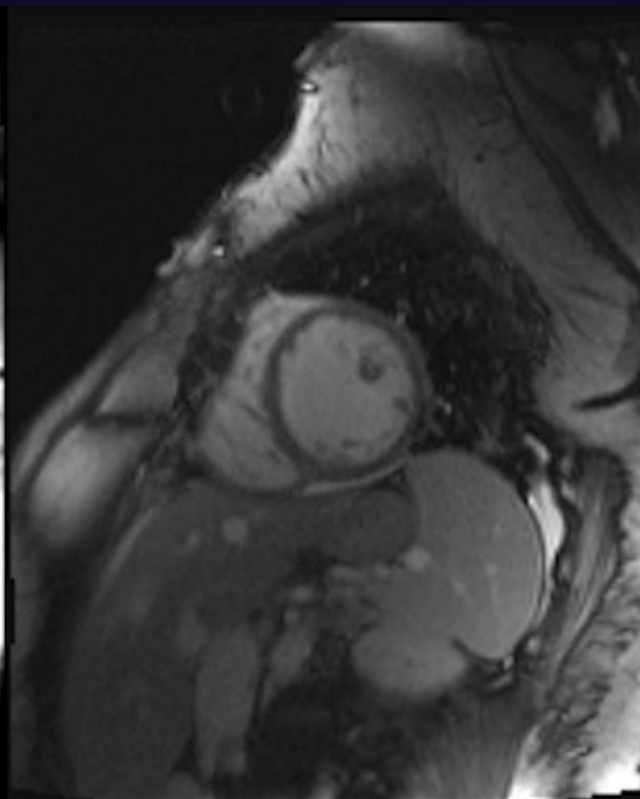
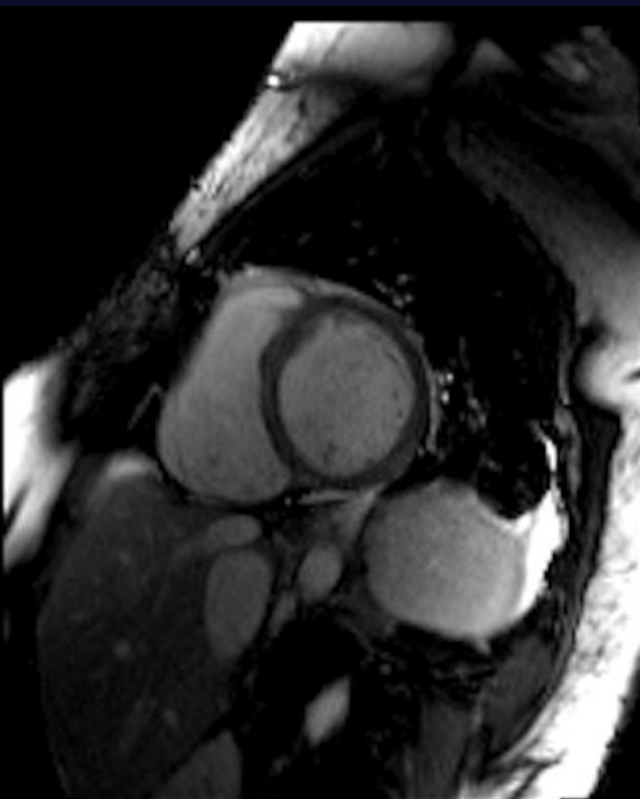


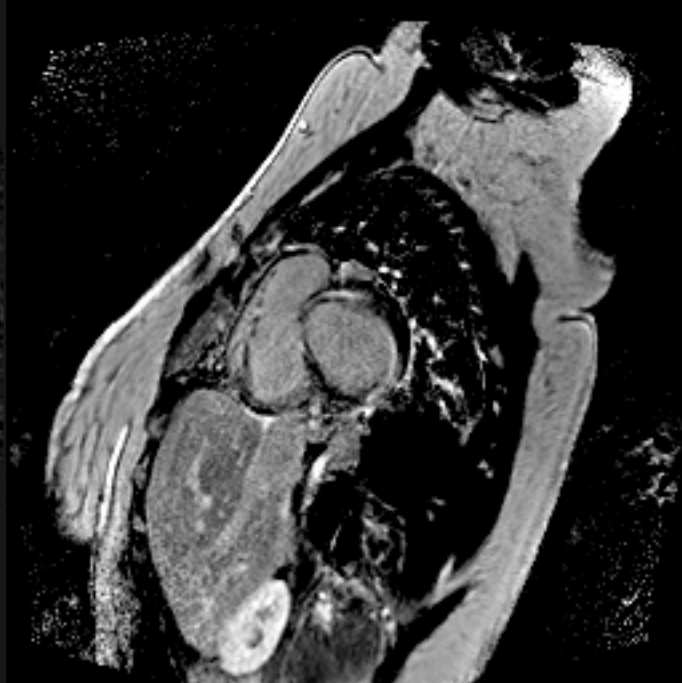












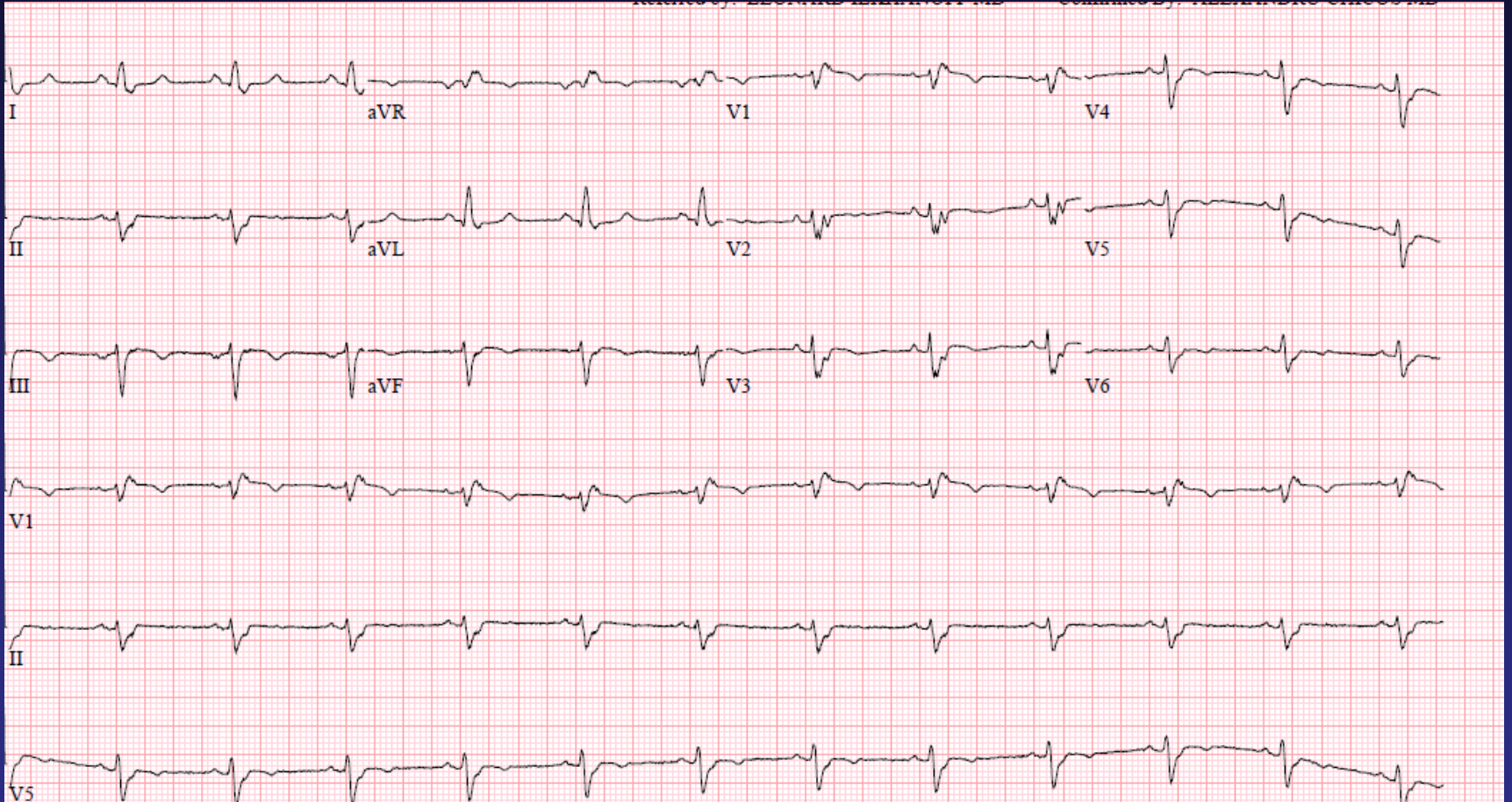
- Patient sent to the OR for AVR
- Myocardial biopsy performed in the OR:  
Positive for sarcoidosis

## Case 3

- 48 yr old female who developed progressive SOB, LE edema and increasing abdominal girth.
- Admitted to the hospital with decompensated HF



# ECG



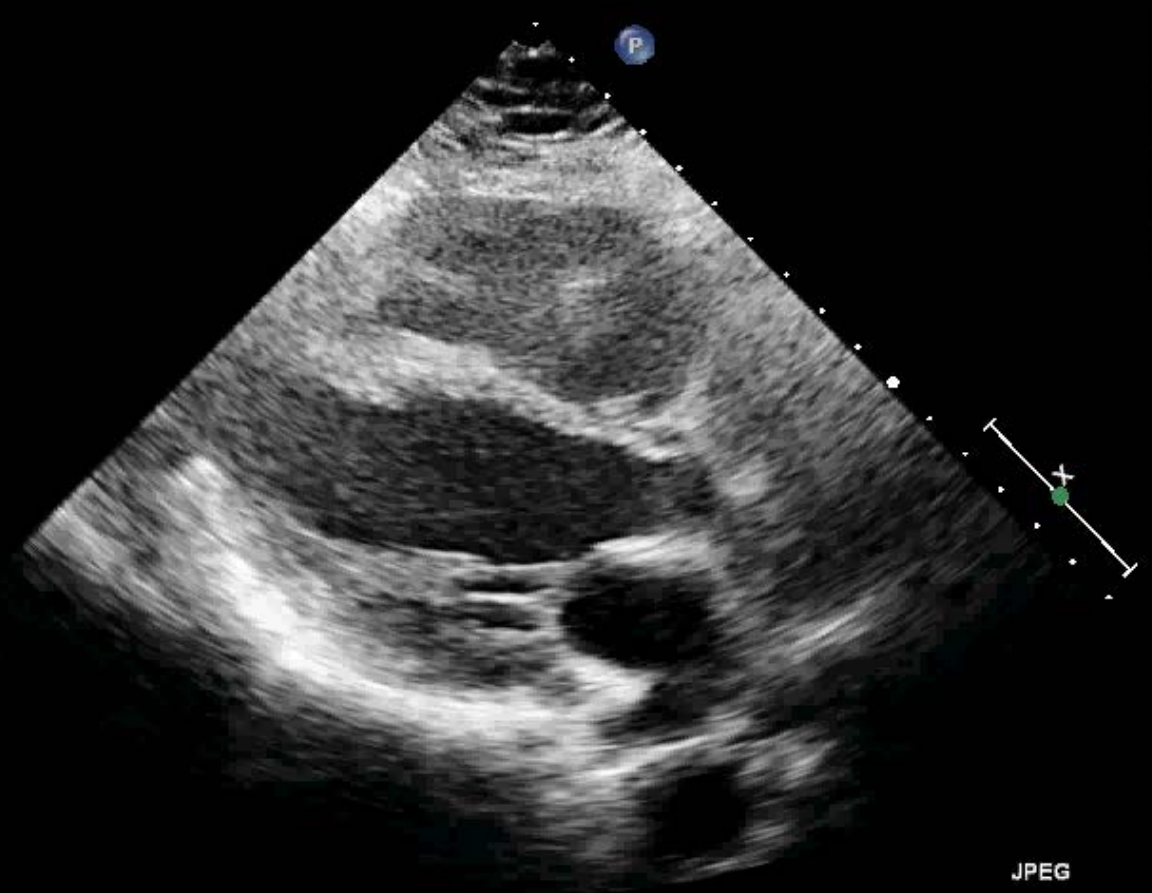
- Cardiac cath: No obstructive CAD
- Cardiac MRI: Myocardial infiltration suspicious for sarcoidosis
- Endomyocardial biopsy:  
+Sarcoidosis

07/13/2011 11:18:20AM TISO.8 MI 1.4  
S5-1/Adult

FR 47Hz  
17cm

M3

2D  
61%  
C 50  
P Low  
HGen



JPEG

229 bpm

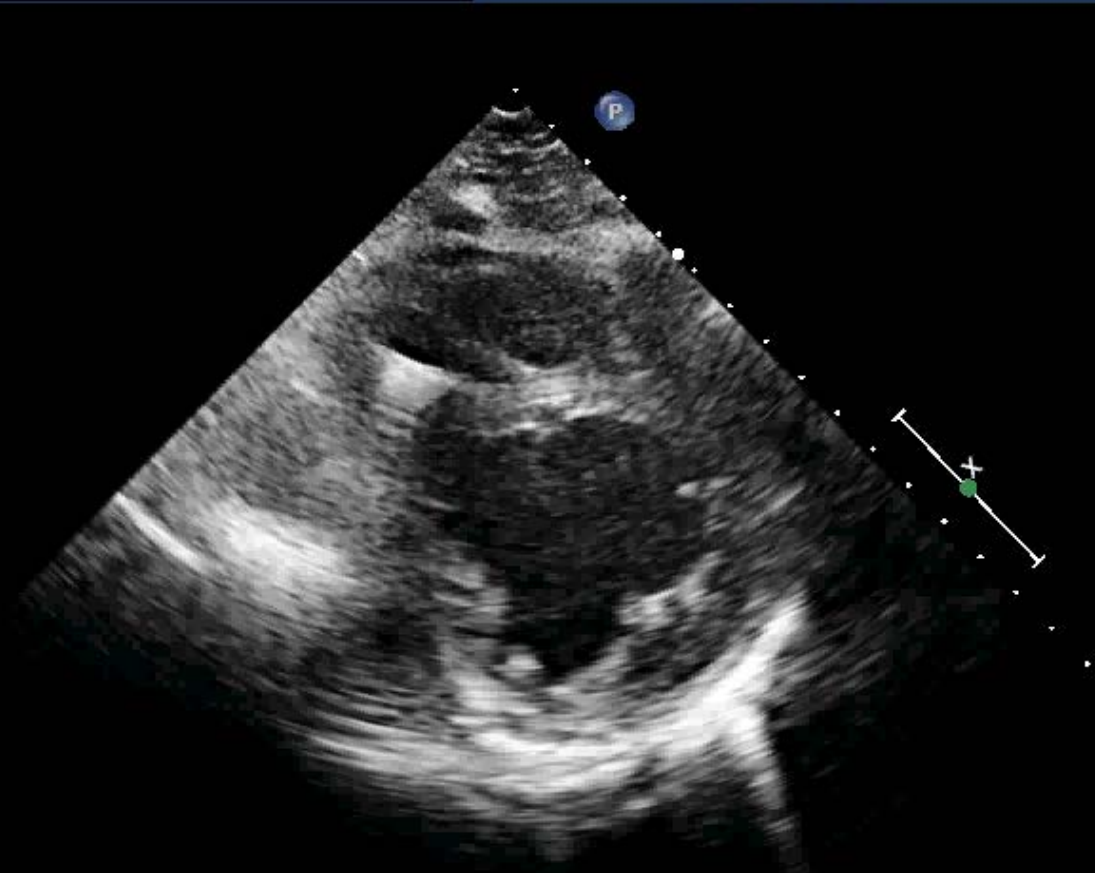
# Echo

07/13/2011 11:22:44AM TIS0.8 MI 1.4  
S5-1/Adult

FR 47Hz  
17cm

2D  
61%  
C 50  
P Low  
HGen

M3



JPEG

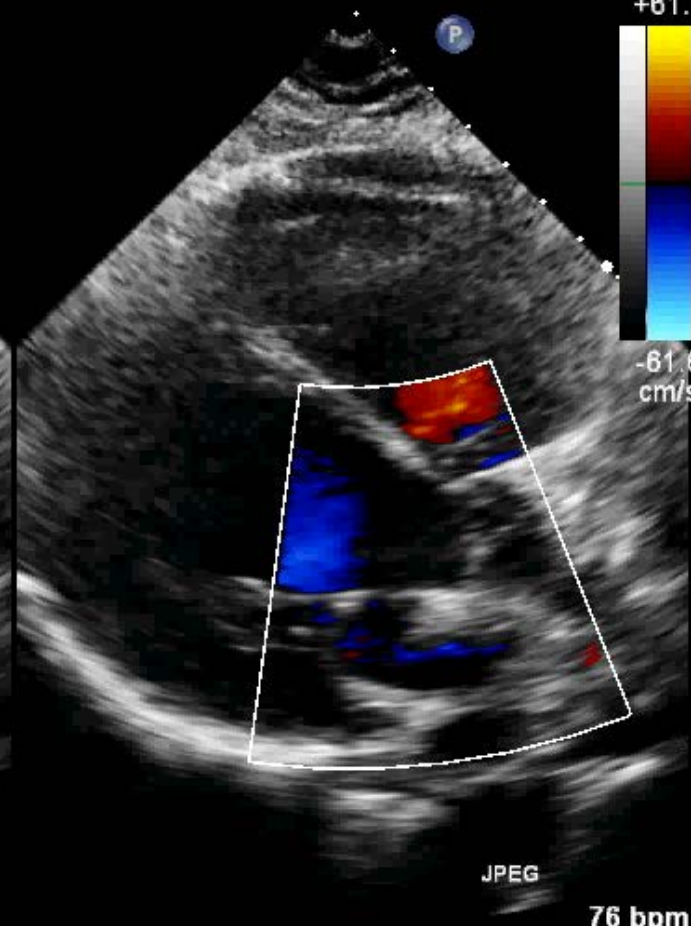
59 bpm

07/13/2011 11:19:53AM TIS2.2 MI 1.2

S5-1/Adult

FR 18Hz  
17cm

**2D**  
60%  
C 50  
P Low  
HGen  
**CF**  
66%  
2.5MHz  
WF High  
Med



M3 M4  
+61.6  
-61.6  
cm/s

JPEG

76 bpm

Western

07/13/2011 11:45:28AM TIS0.0 MI 0.23  
S5-1/Cont LVO

FR 34Hz  
15cm

LVO  
89%  
C 50  
P Low  
Res

4

M3

G  
P ((O))®  
1.6 3.2



JPEG

62 bpm

Western



07/13/2011 11:46:06AM TISO.0 MI 0.23  
S5-1/Cont LVO

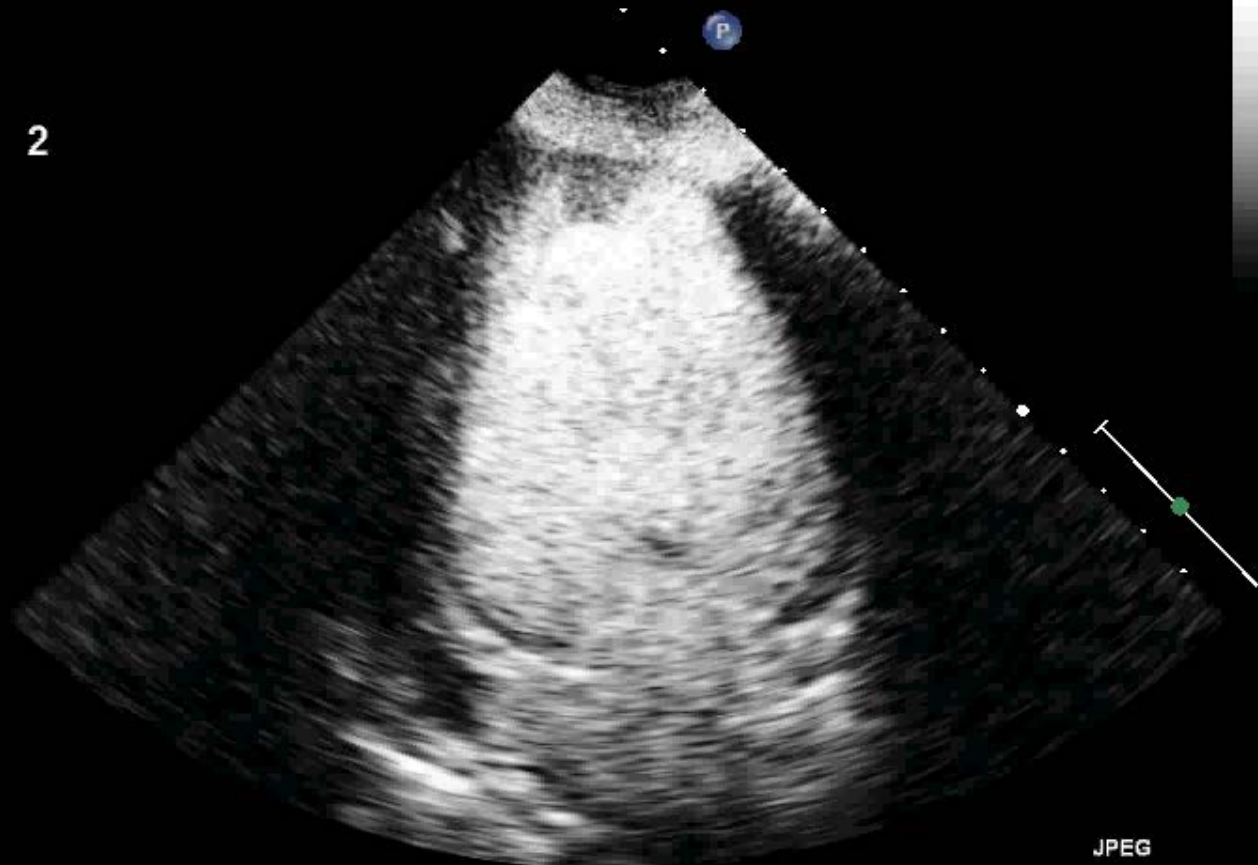
FR 34Hz  
15cm

LVO  
89%  
C 50  
P Low  
Res

2

M3

G  
P ((O))<sup>®</sup>  
1.6 3.2



JPEG

89 bpm



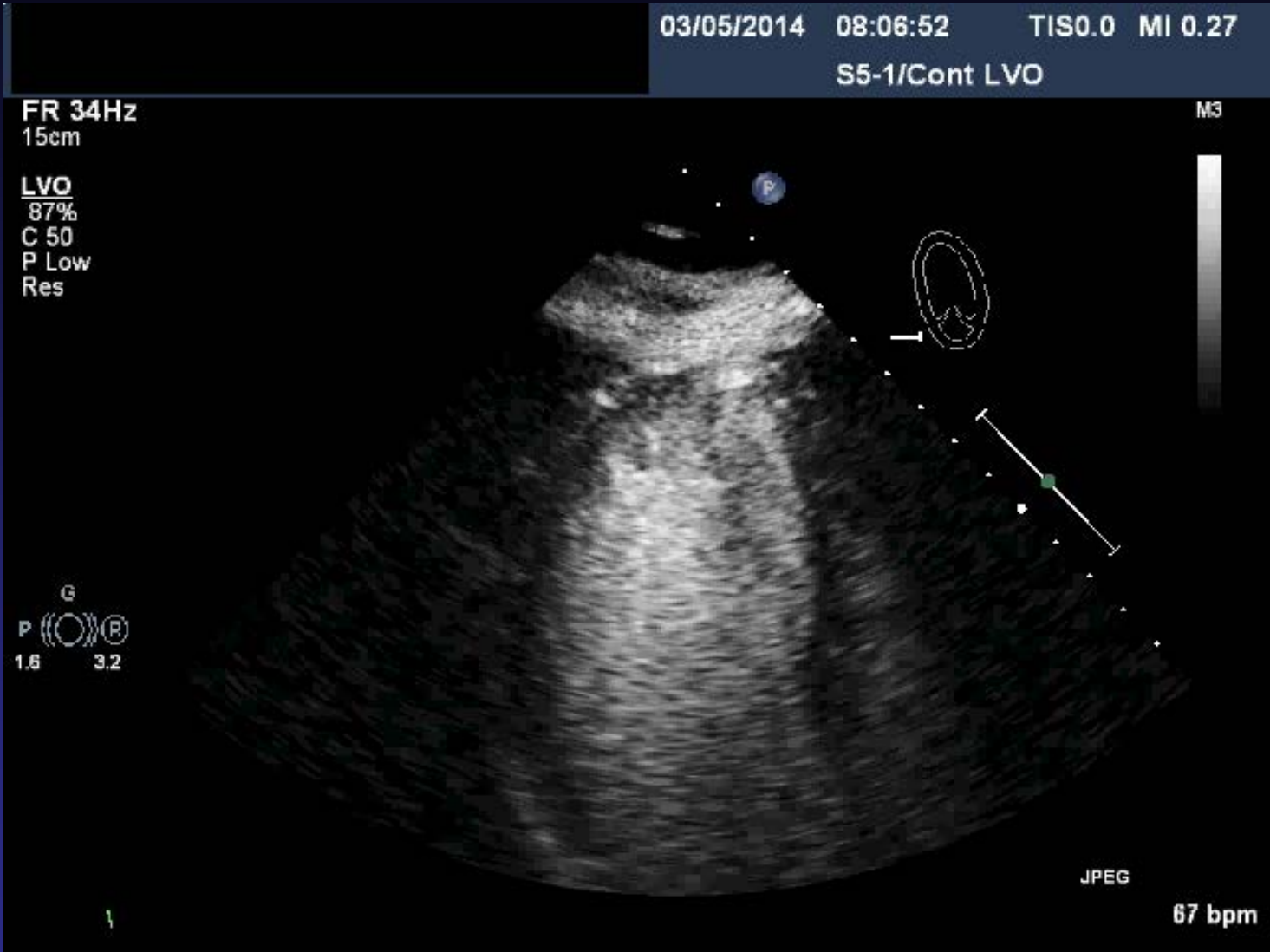
03/05/2014 08:06:52 TISO.0 MI 0.27  
S5-1/Cont LVO

FR 34Hz  
15cm

LVO  
87%  
C 50  
P Low  
Res

M3

G  
P ((O)) (B)  
1.6 3.2



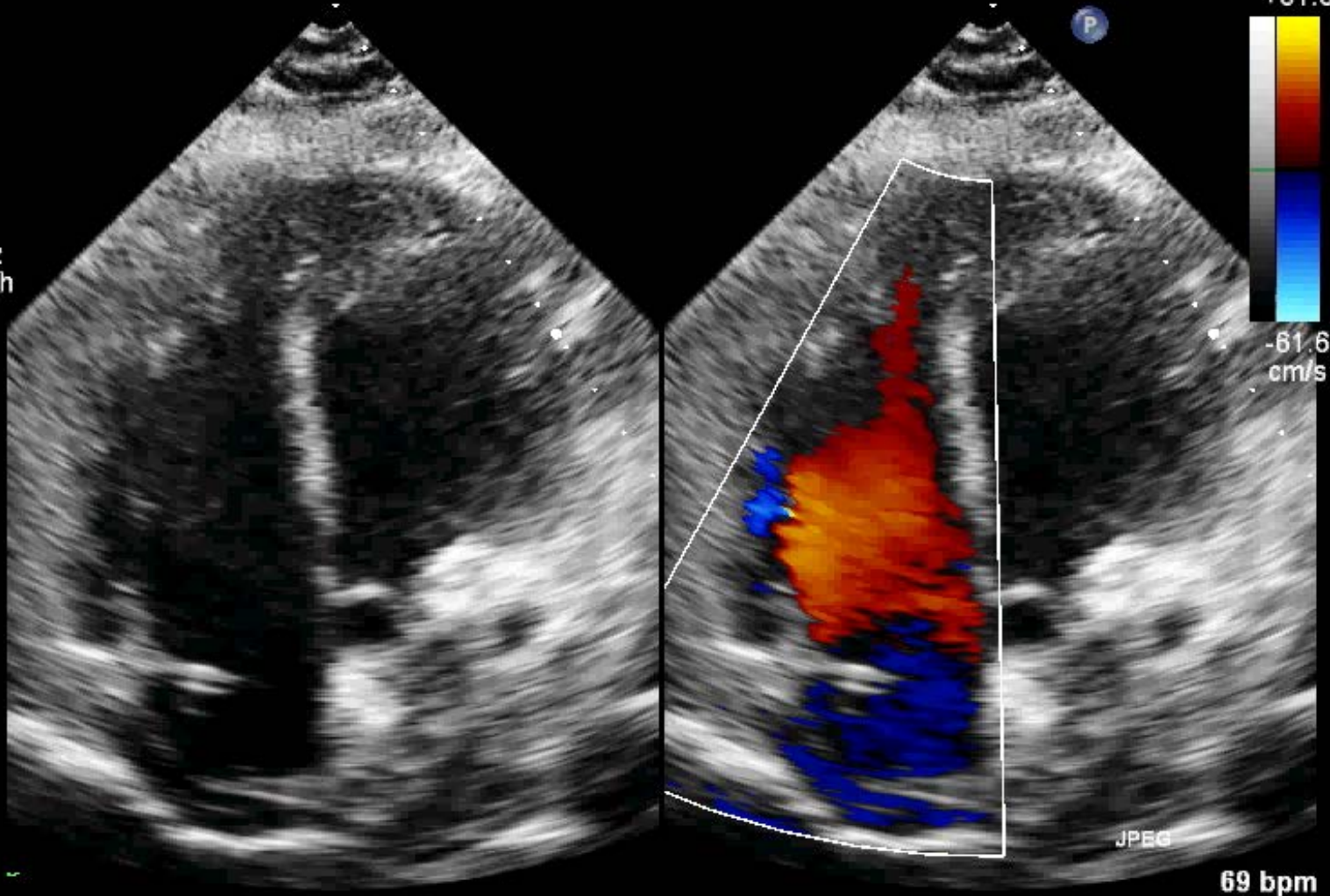
JPEG

67 bpm

07/13/2011 11:29:16AM TIS2.2 MI 1.2  
S5-1/Adult

FR 17Hz  
17cm

2D  
64%  
C 50  
P Low  
HPen  
CF  
66%  
2.5MHz  
WF High  
Med



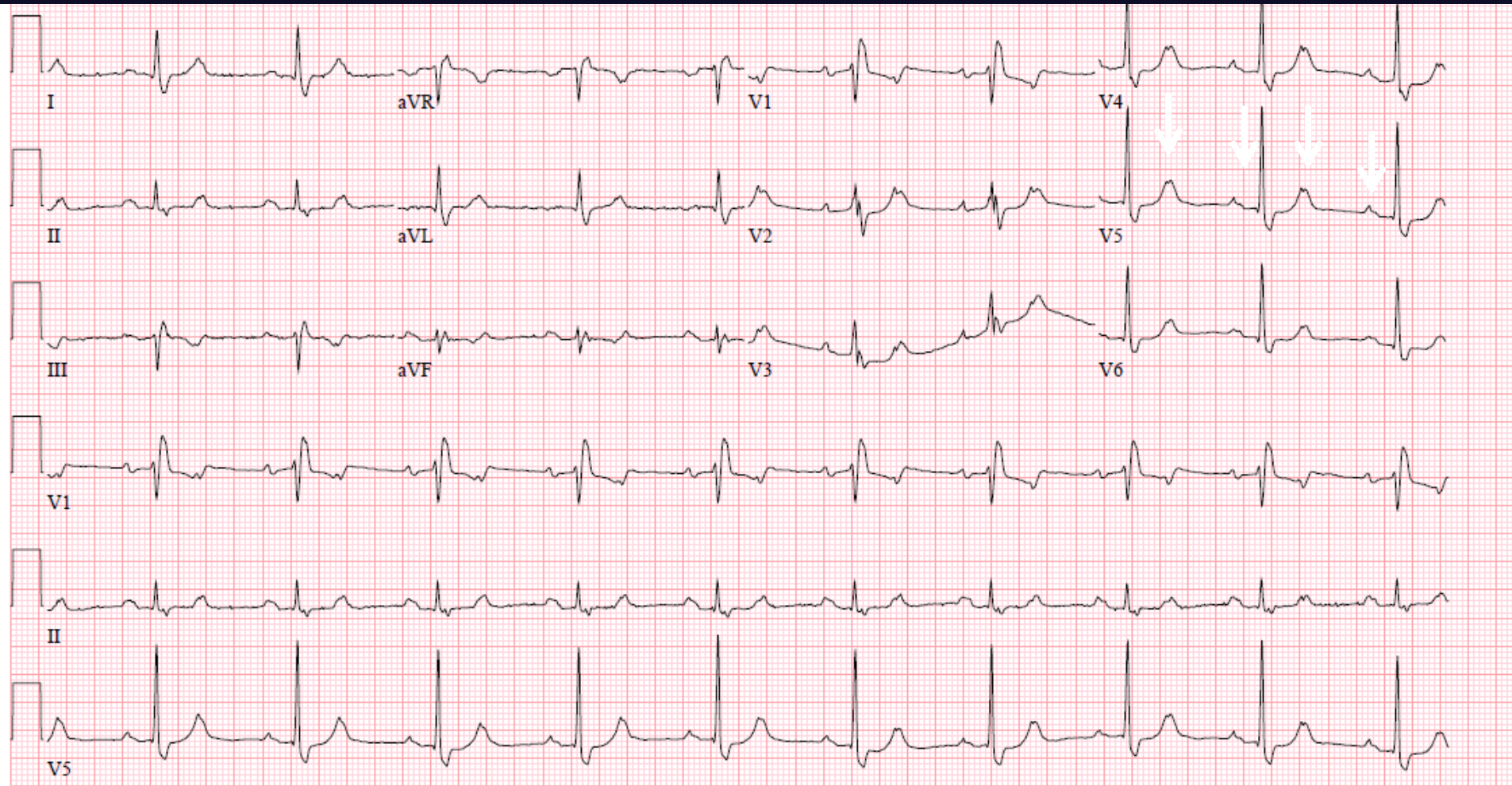




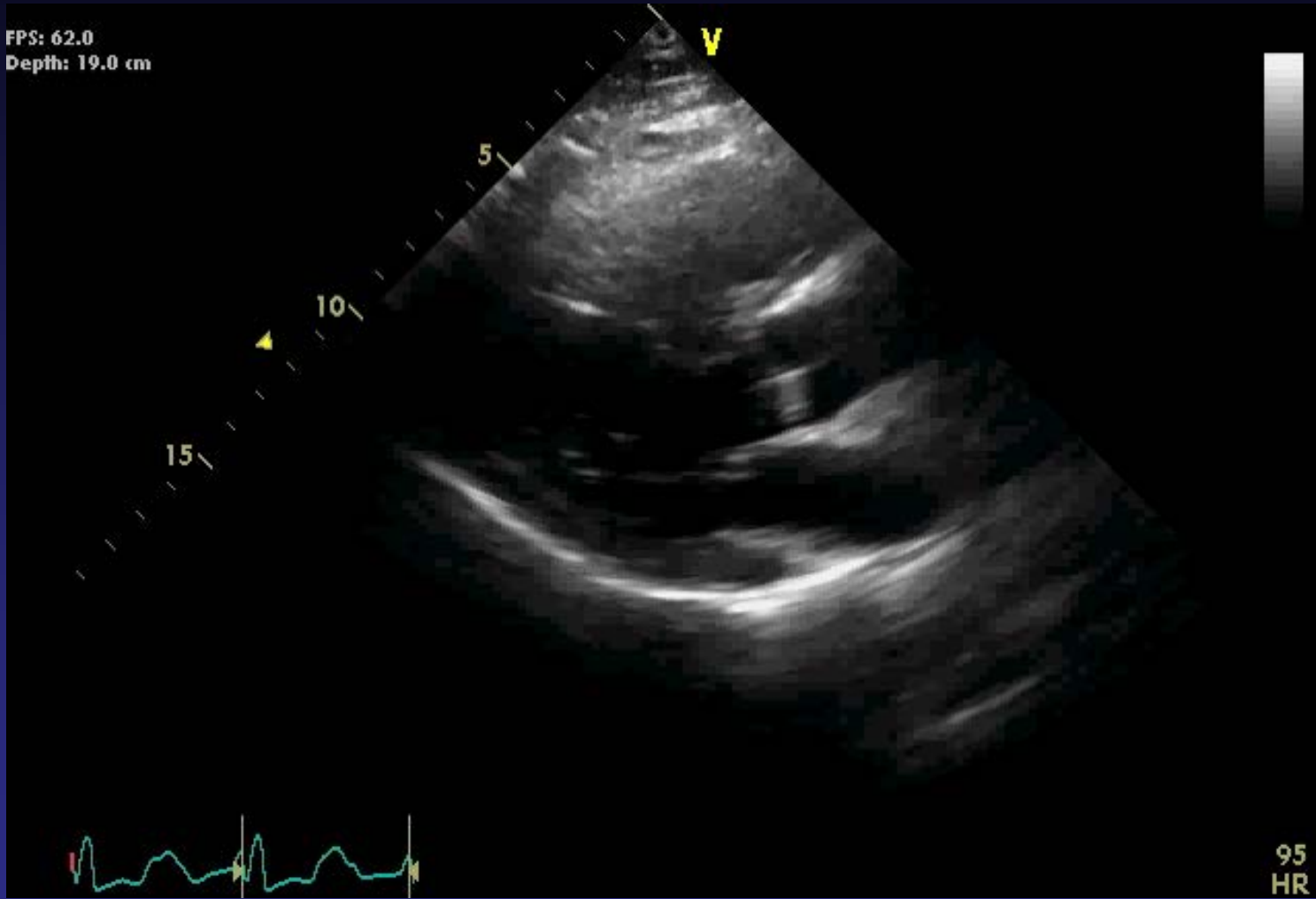
- Patient had progressive heart failure despite maximal meds
- She ultimately underwent heart transplantation

## Case 4

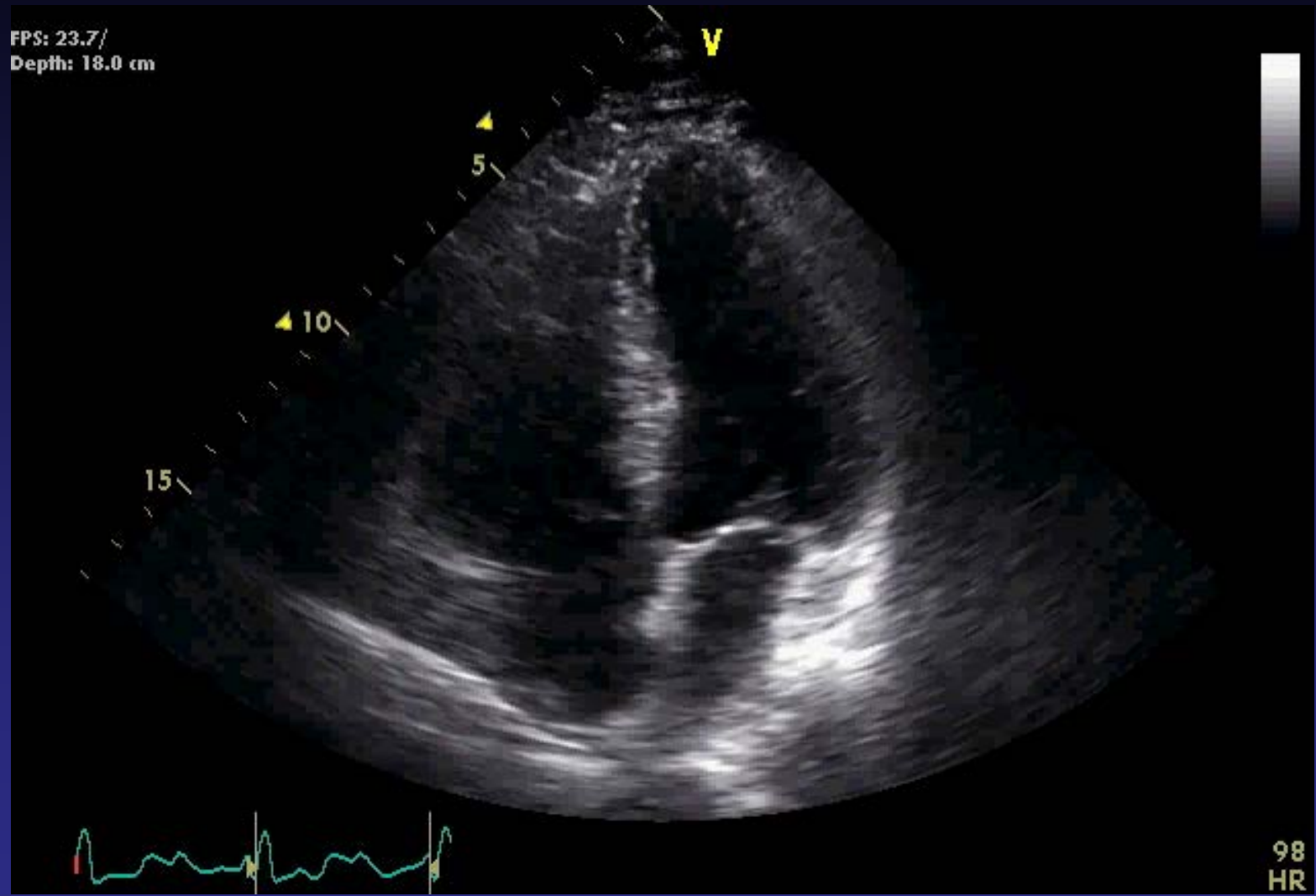
- 56 yr old ER physician with no past medical history who presented with dyspnea and palpitations



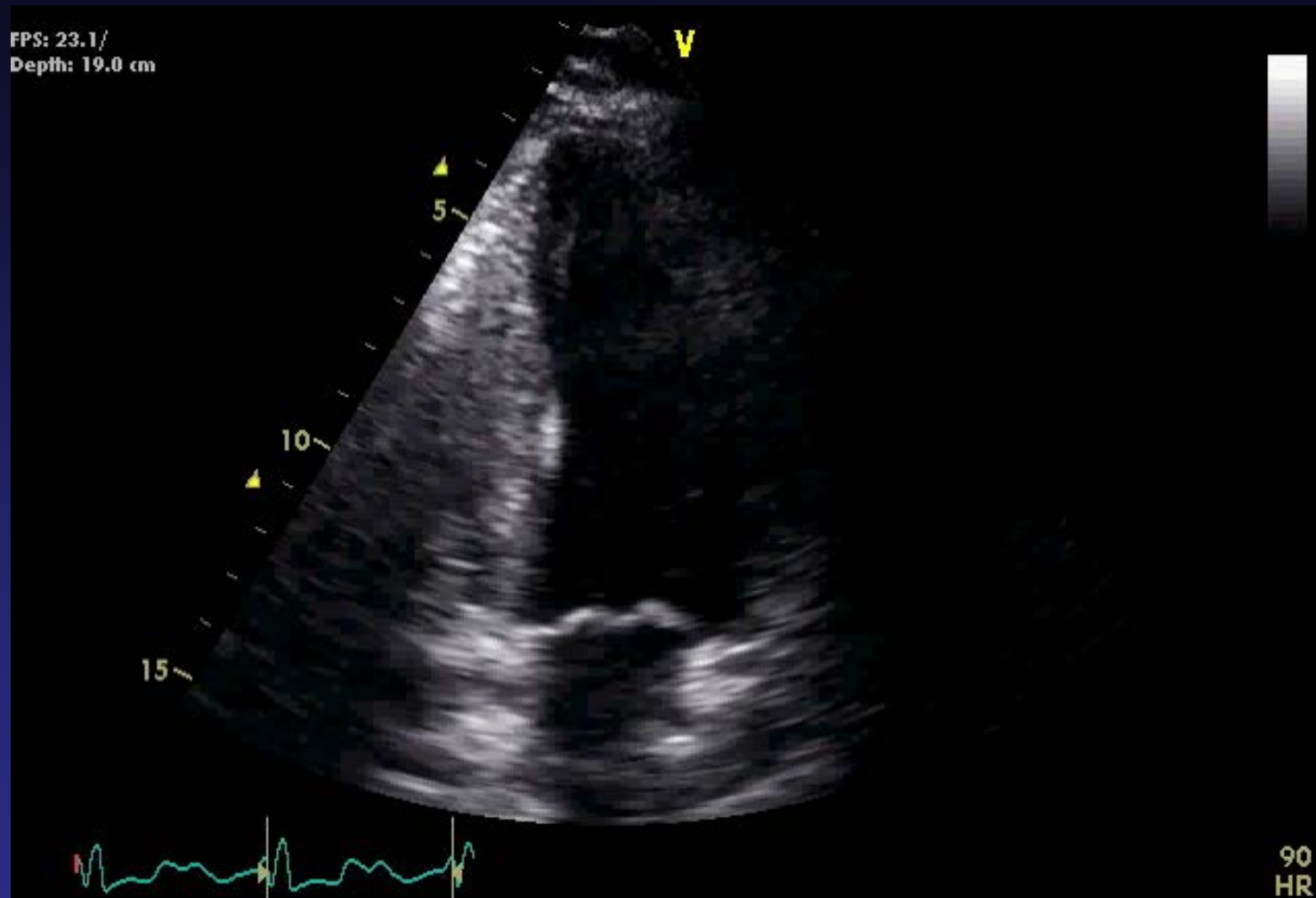


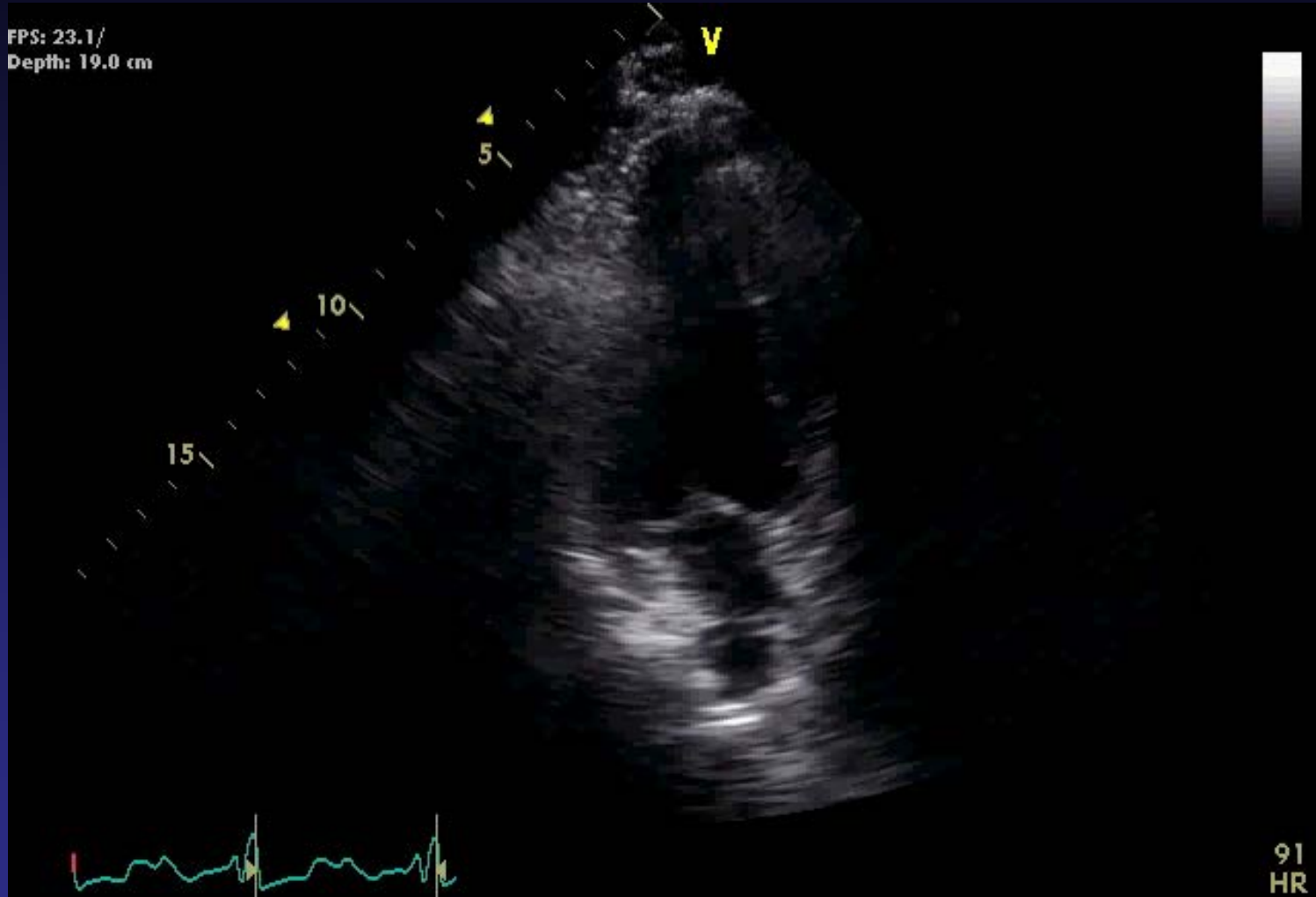


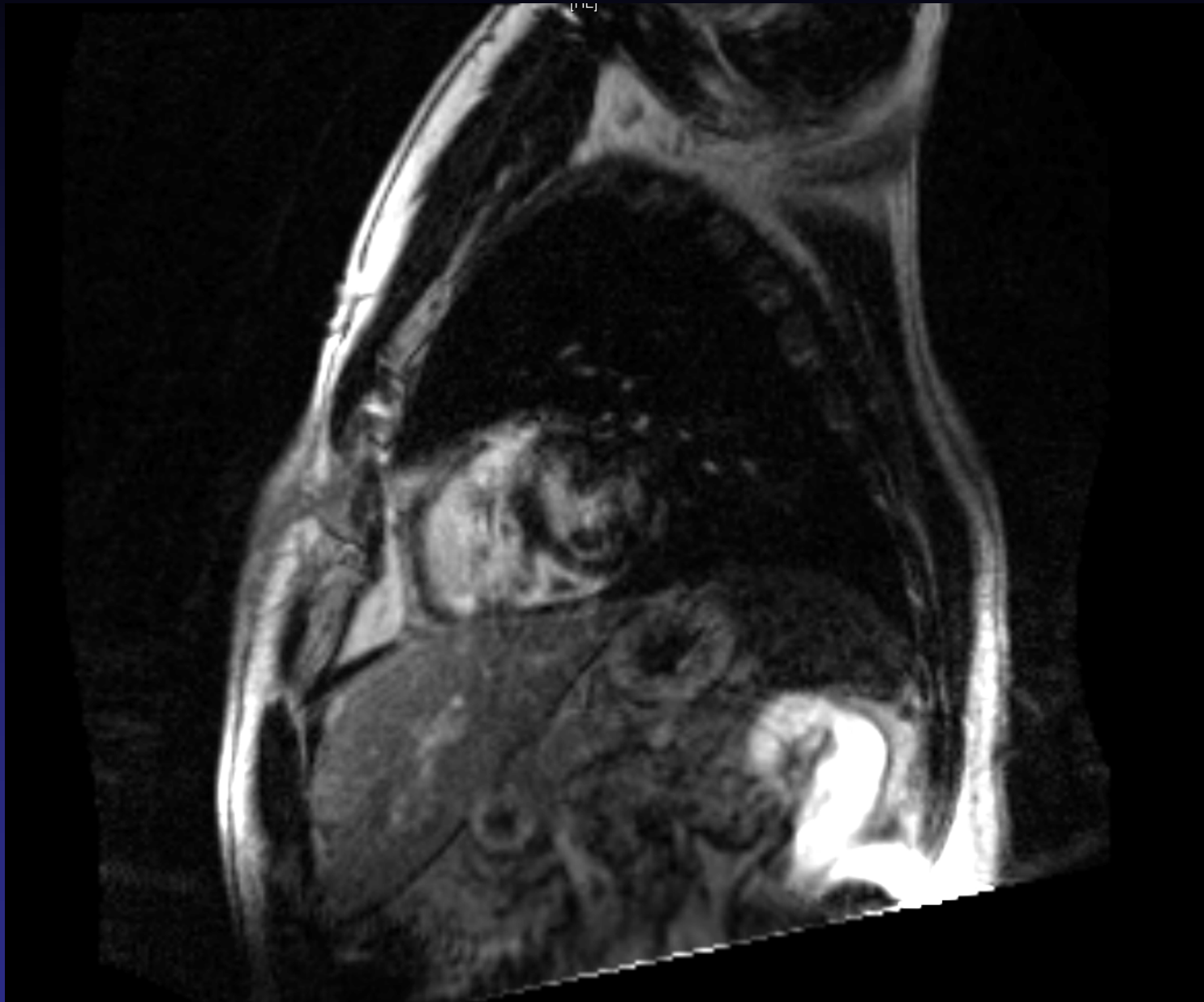
FPS: 23.7/  
Depth: 18.0 cm

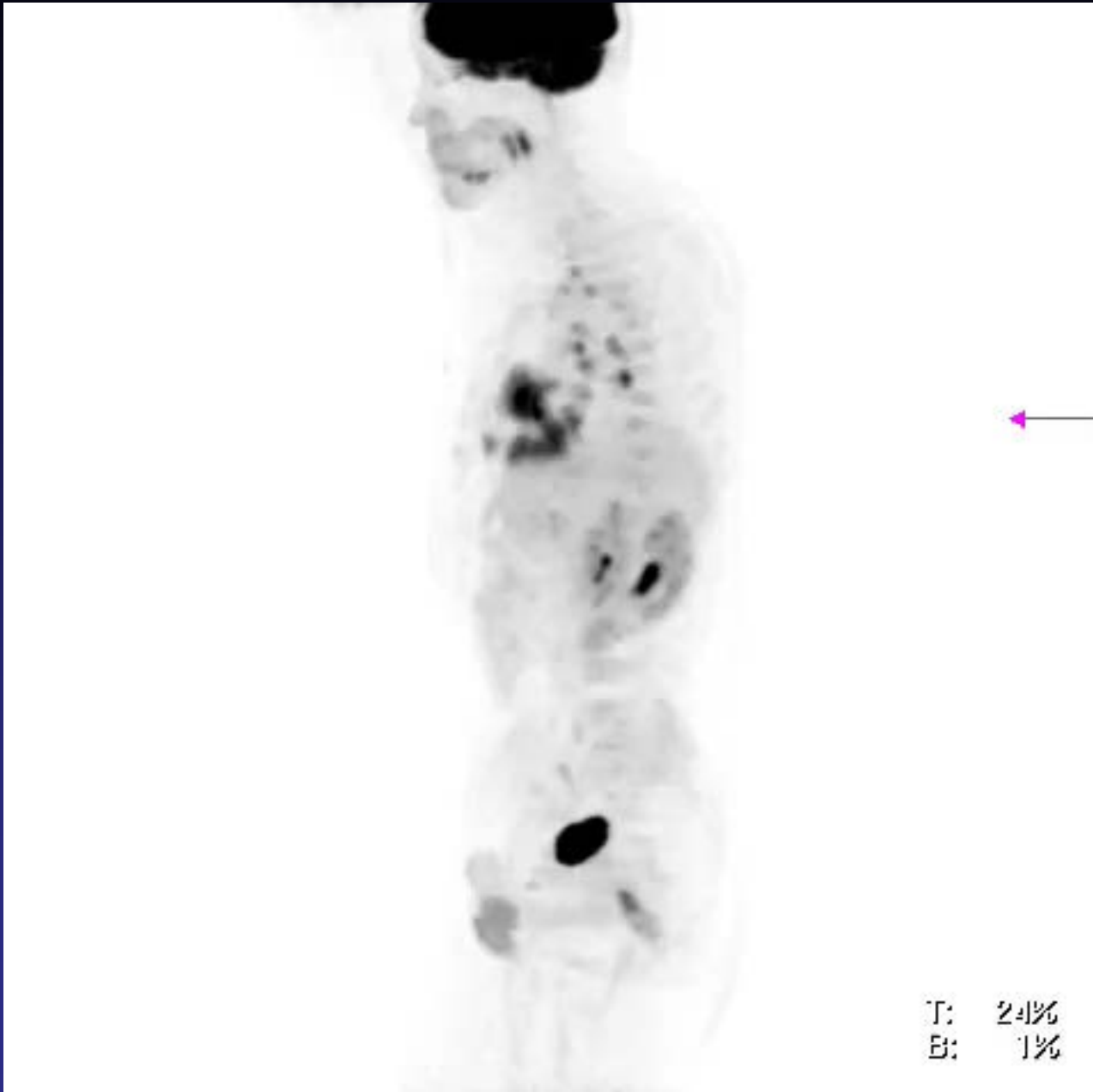


FPS: 23.1/  
Depth: 19.0 cm











HU 3 %

85

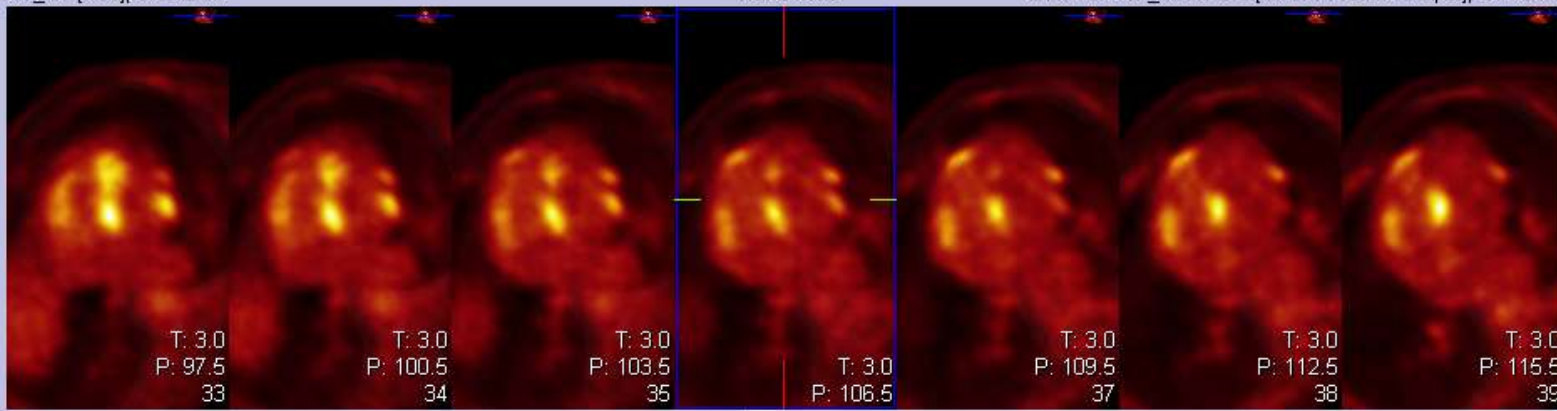
Right

Anterior

Posterior

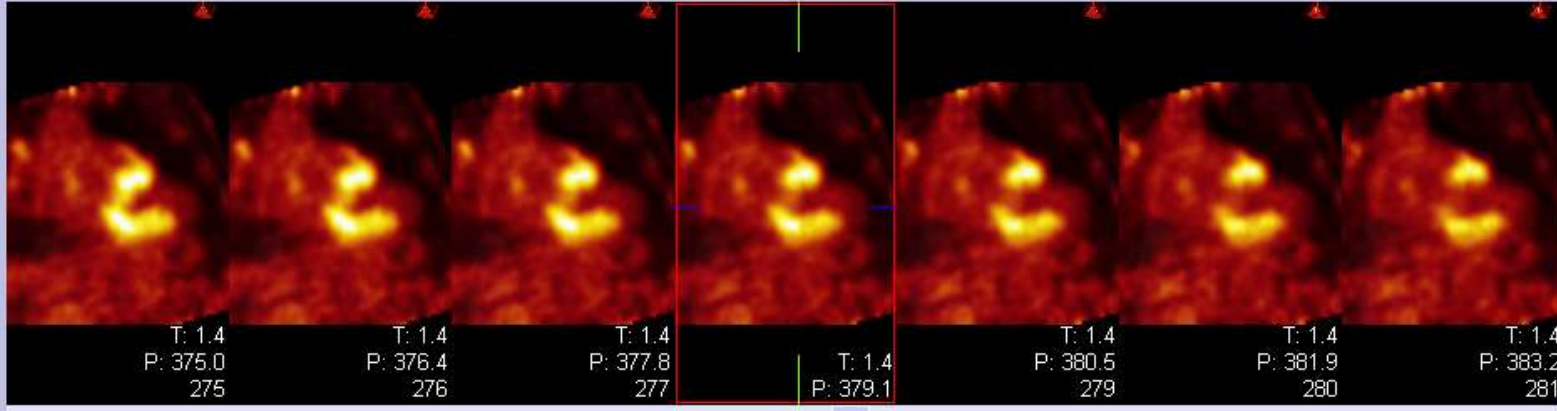
Bottom to Top

A



Sagittal

A



Posterior

Top

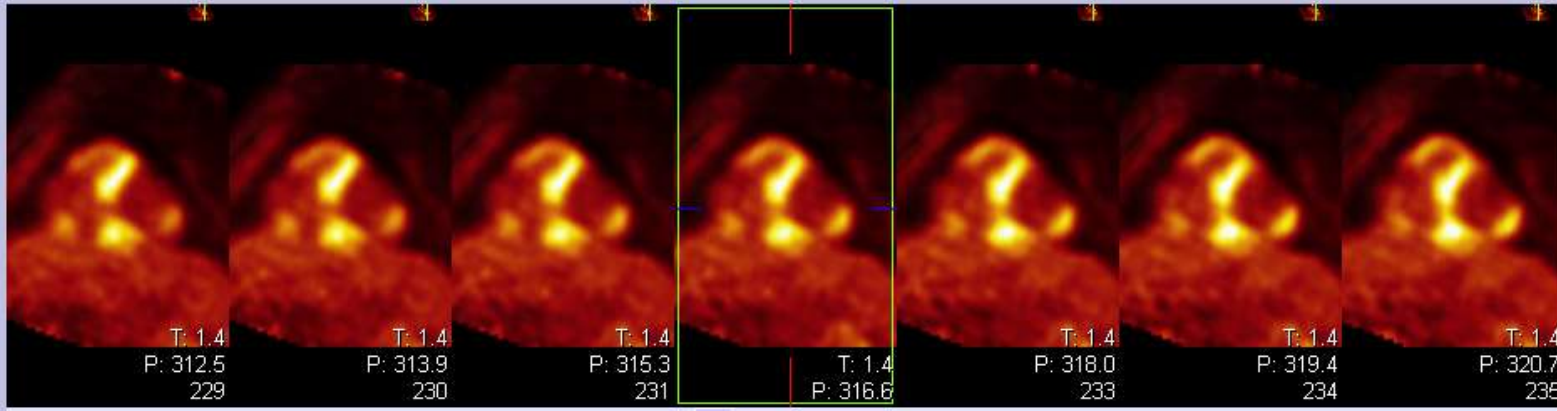
Anterior

Bottom

Right to Left

Coronal

A



Right

Top

Left

Bottom

Anterior to Posterior

# Hospital Course

- Endobronchial biopsy pending
- ICD/pacer implanted





Thank You