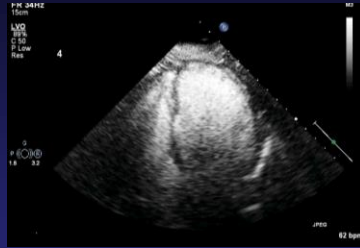


Echo in Systemic Disease



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 Northwestern
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No Disclosures

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Director, program for heart failure with preserved ejection fraction



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Introduction

- *A variety of systemic diseases can affect the heart*
- *Echo is usually the first imaging technique used to evaluate such patients*
- *Typical systemic diseases with cardiac effects:*
 - Infiltrative diseases (sarcoidosis, amyloidosis)
 - Autoimmune/collagen vascular
 - Endocrine
 - Malignancy
 - Radiation-induced injury
 - Drug-induced valvulopathy



Rheumatologic disease & CV disease

- *Pericardial disease: SLE, RA, SSc*
- *Epicardial: CAD: RA, SLE*
- *Cardiac microvascular disease: SSc*
- *Myocardial disease: SSc, SLE, (RA)*
- *Valvular disease: SLE, ankylosing spondylitis*
- *Conduction dz: SSc, ankylosing spondylitis*
- *Pulmonary arterial HTN: SSc, SLE*

*SLE=systemic lupus erythematosus, RA=rheumatoid arthritis,
SSc=systemic sclerosis (scleroderma)*



Systemic sclerosis (SSc)

- *Rare: US incidence 19 cases/million/year*
- *Female predominance (7.5:1 female:male)*
- *Complex disease:*
 - Extensive fibrosis, vascular alterations, autoantibodies against various cellular antigens
- *2 main types:*
 - Limited cutaneous SSc (lcSSc)
 - » “CREST” syndrome: usually associated with lcSSc
 - » Vasculopathy predominates, some fibrosis
 - Diffuse cutaneous SSc (dcSSc)
 - » Fibrosis predominates, some vasculopathy



Gabrieli A et al, NEJM 2009

Slide courtesy of Sanjiv Shah, MD



Cardiac Manifestations of Scleroderma

- *Diverse cardiovascular manifestations can occur in scleroderma*
 - Primary: myocardial fibrosis, microvascular ischemia, pericardial disease, conduction disease, arrhythmias
 - Secondary: pulmonary hypertension
- *Both PAH and PVH can occur in scleroderma*



Slide courtesy of Sanjiv Shah, MD



Case Presentation

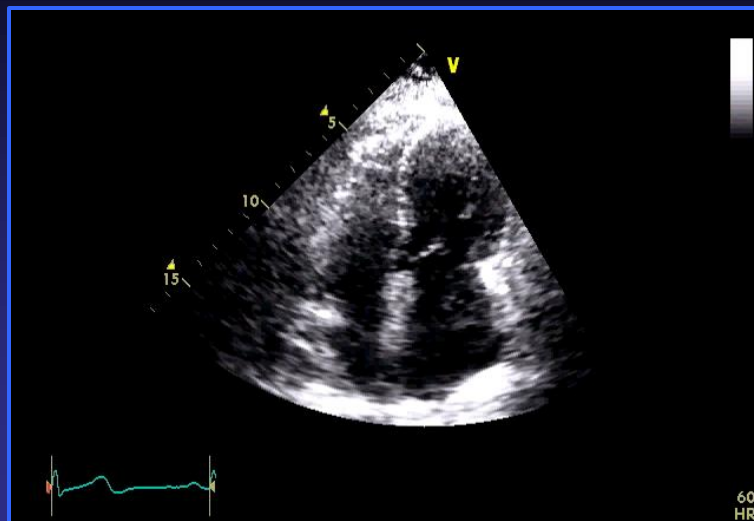
- *62-year-old female nurse with history of scleroderma and systemic HTN*
- *Progressive dyspnea, leg swelling*
- *Worsening exercise tolerance*
- *NYHA class III symptoms*
- *Scleroderma diagnosed 5 years earlier*
 - Raynaud's, skin tightening, GERD
- *HTN well-controlled*



Case provided courtesy of Sanjiv Shah, MD

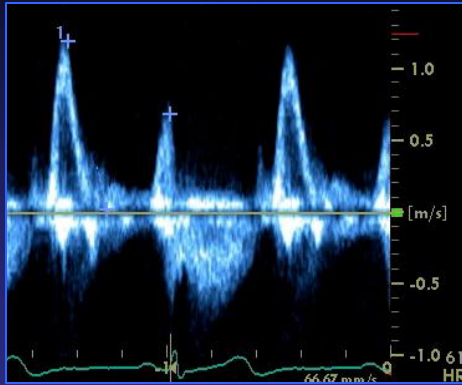
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Echocardiography



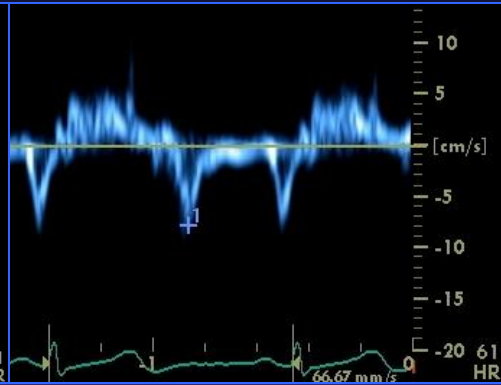
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Echocardiography



MITRAL INFLOW

E = 120 cm/s, DT = 170 ms
A = 72 cm/s, E/A ratio = 1.7

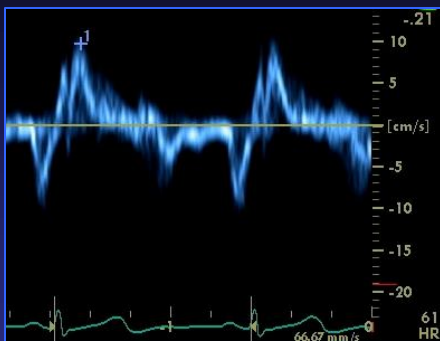


LATERAL TDI

e' = 7 cm/s
E/e' ratio = 17.1

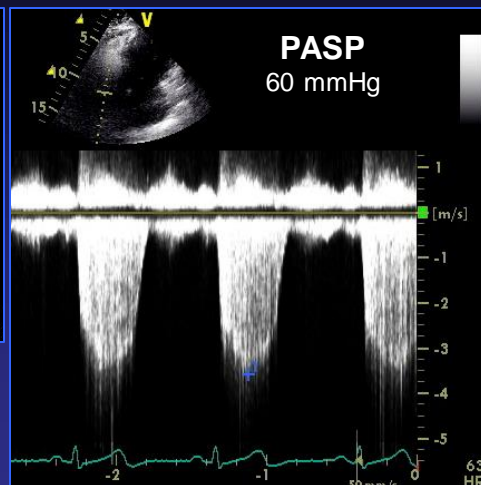


Echocardiography



RV FREE WALL TDI

RV s' = 10.3 cm/s



PASP
60 mmHg



Question

- *What is the most likely cause of this patient's symptoms?*
 - A. Pulmonary arterial hypertension
 - B. Pulmonary venous hypertension
 - C. Primary RV failure
 - D. Constrictive pericarditis
 - E. Amlodipine-induced LE edema



DDx of right heart failure

- *Pulmonary arterial hypertension*
 - $E/A < 1$, lateral E/e' normal, $RA > LA$
- *Pulmonary venous hypertension*
 - $E/A \gg 1$, lateral E/e' high, $LA \gg RA$
- *Primary RV failure*
 - Low PASP, reduced RV s' velocity (< 10 cm/s)
- *Constrictive pericarditis*
 - Diastolic septal bounce, preserved lateral e'
 - Respiratory variation in mitral inflow



PAH vs PVH: Practical tips on echo

- *Think PVH until proven otherwise*
- *Signs which favor PVH:*
 - Left atrial enlargement
 - Interatrial septum bowing L→R
 - Grade 2 or worse diastolic dysfunction
 - » *NOTE: severe PAH usually causes grade 1 DD!*
 - Reduced tissue Doppler lateral e' velocity
 - High E/e' (lateral) ratio

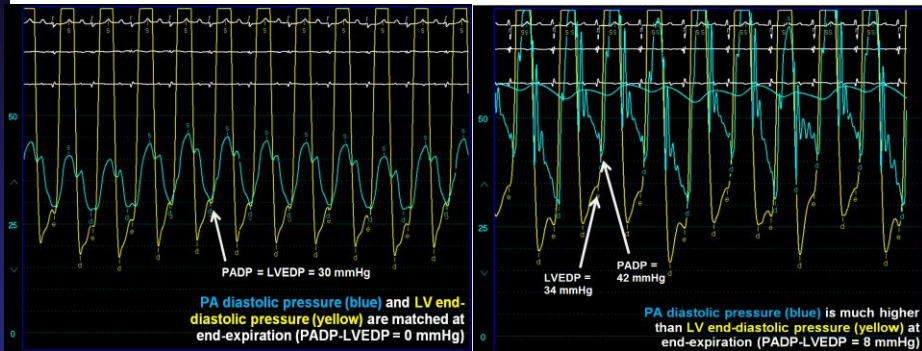


Invasive Hemodynamics

- *RA pressure:*
 - 10 mmHg (normal < 6 mmHg)
- *PA pressure:*
 - 62/22 mmHg, mean 35 mmHg
 - (normal 30/12, mean < 20 mmHg)
- *PCWP:*
 - 22 mmHg (normal < 12 mmHg)
- *CO: 6 L/min (normal 4-8 L/min)*
- *PVR: 2.2 Wood units (normal < 1.5 WU)*



PADP-LVEDP (or PADP-PCWP): key to diagnosis of "reactive" PH in left heart disease



PASSIVE PH

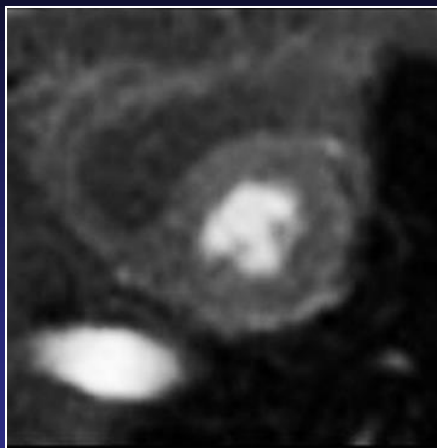
PADP-LVEDP = 0 mmHg
Pulmonary venous hypertension only

REACTIVE PH

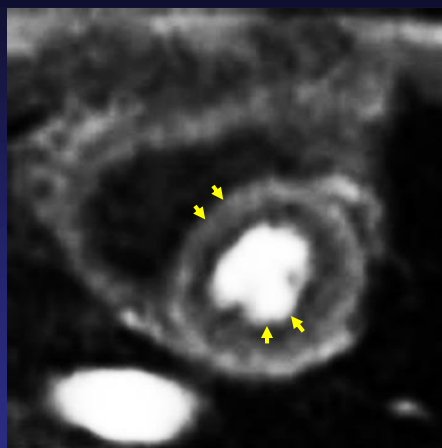
PADP-LVEDP = 8 mmHg
Pulmonary venous hypertension
w/ superimposed PAH



Adenosine perfusion cardiac MRI results



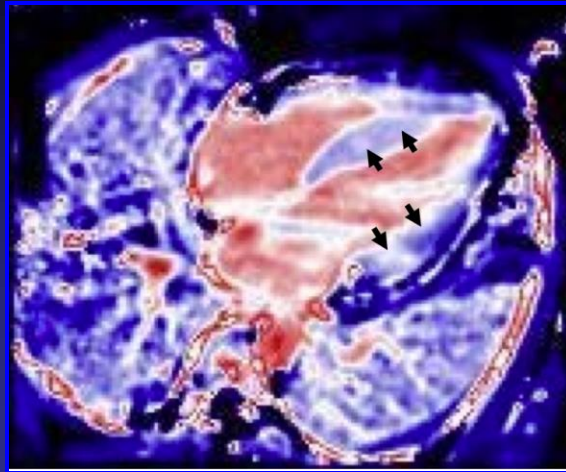
Rest



Stress



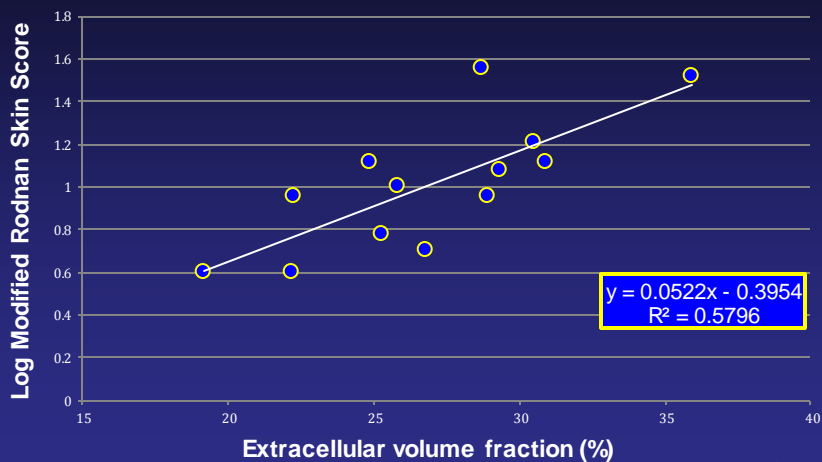
T1 mapping for diffuse fibrosis



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Diffuse cardiac fibrosis on MRI in SSc

Skin fibrosis vs. diffuse cardiac fibrosis



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Treatment

- *Rx with bumetanide and spironolactone*
- *Dramatic improvement in symptoms*
- *NYHA class I*

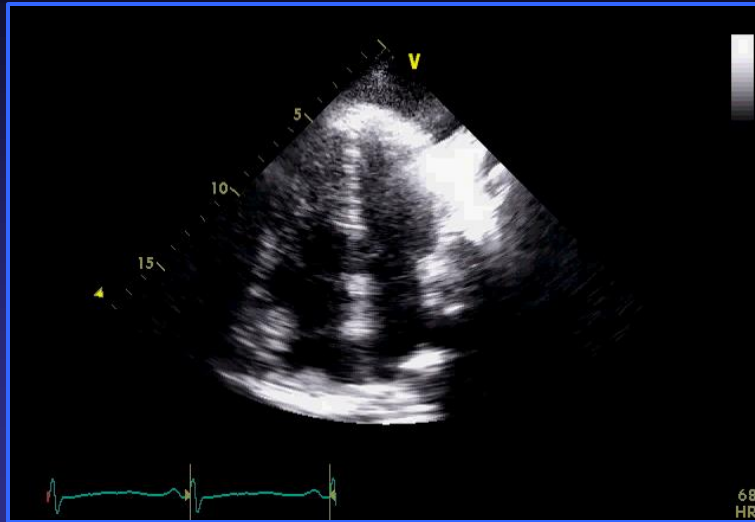


Follow-up

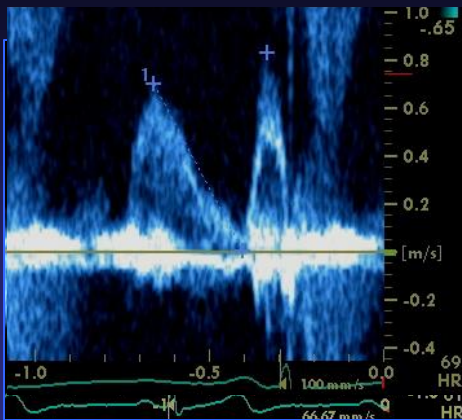
- *12 months after her initial evaluation → exertional lightheadedness and dizziness*
 - *Especially when climbing stairs*
- *Symptoms progressed → exertional syncope*
- *Hospitalized for further evaluation*



Echocardiography

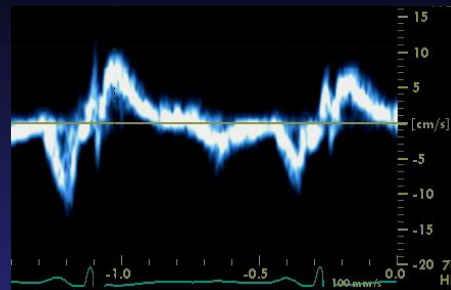


Echocardiography



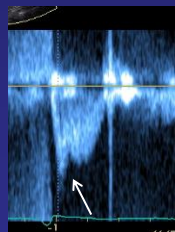
MITRAL INFLOW

E = 72 cm/s, DT = 250 ms
A = 81 cm/s, E/A ratio = 0.9



RV FREE WALL TDI

RV s' = 8.2 cm/s



Mid systolic notching
of RVOT PW

Treatment

- *Rx with sildenafil, bosentan*
- *Rapidly worsening symptoms*
- *Hypotension, recurrent syncope*
- *Admitted to CCU*



- Right heart catheterization in CCU
 - » RA: 15 mmHg
 - » RV: 73/15 mmHg
 - » PA: 73/30 (mean 46 mmHg)
 - » PCWP: 14 mmHg
 - » Cardiac output 2.5 L/min, PVR > 10 WU



Treatment

- *Initially treated with dobutamine*
 - Persistent hypotension
 - Urine output → dropped to zero
- *Treatment switched to:*
 - IV diuresis (furosemide gtt)
 - Inhaled nitric oxide 20 ppm
 - Phenylephrine
 - Urine output increased
 - Eventually transitioned to SQ treprostinil

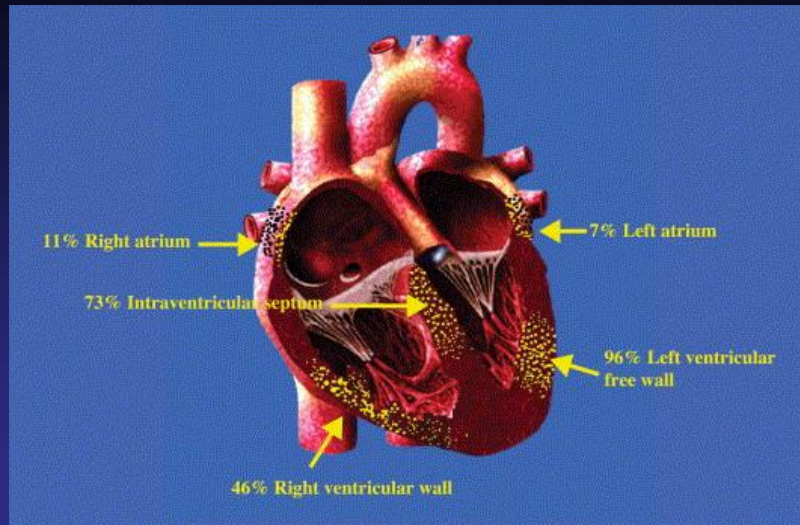


Sarcoidosis

- *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*



Common Sites of Sarcoid Granulomas in the Heart



Bargout R, Kelly RF. International Journal of Cardiology. 2004;97:173-182



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

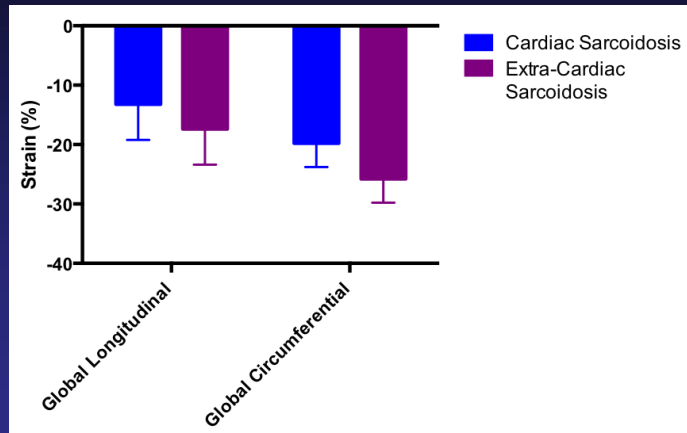
- *Left ventricular dilatation*
- *Increase in wall thickness simulating Hypertrophic Cardiomyopathy (HCM)*
- *Segmental or global hypokinesia of the left ventricle*
- *Wall motion abnormalities in noncoronary distribution*
- *Wall thickening and ventricular aneurysms in late stages*
- *Mitral regurgitation*
- *Right ventricular dilatation and hypokinesis.*

Echo can also be normal!!!



Speckle Tracking Echocardiography Identifies Patients with Cardiac Sarcoidosis

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

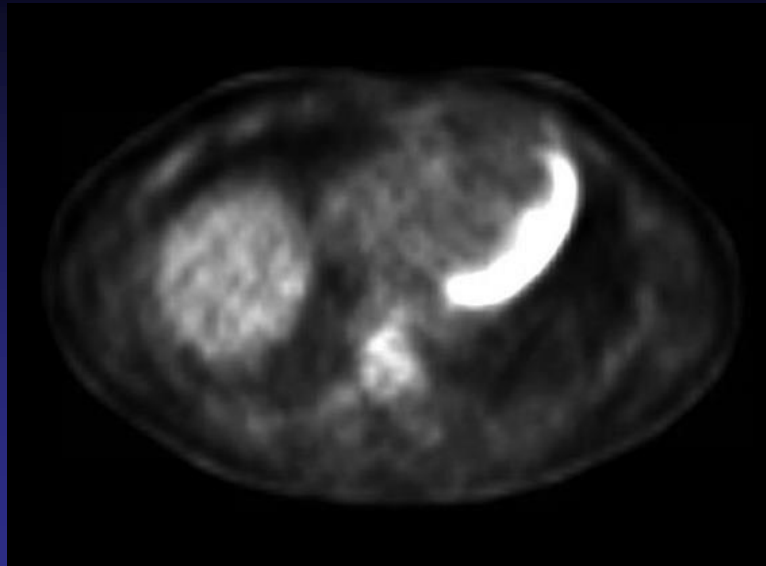


Cardiac MRI

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

^{18}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*



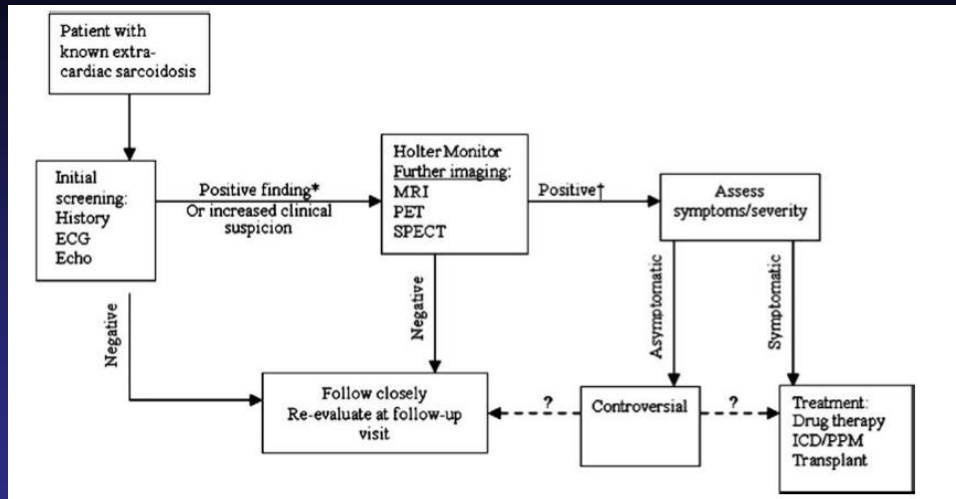
Accuracy of Diagnostic Tests

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

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



Suggested Clinical Algorithm



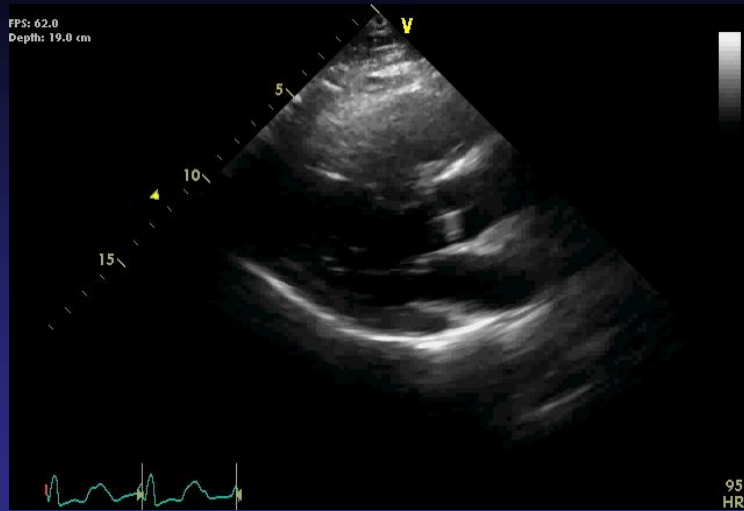
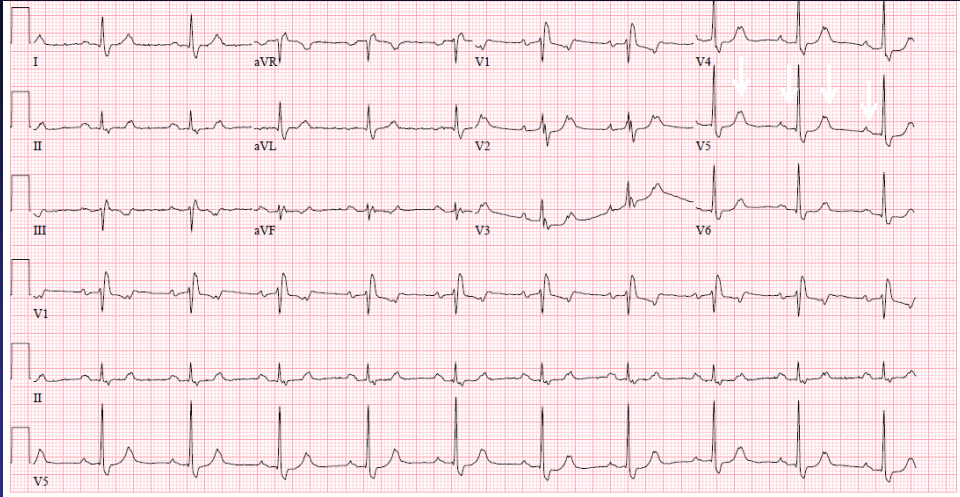
Kim JS et al. Am Heart J 2009;157:9-21.

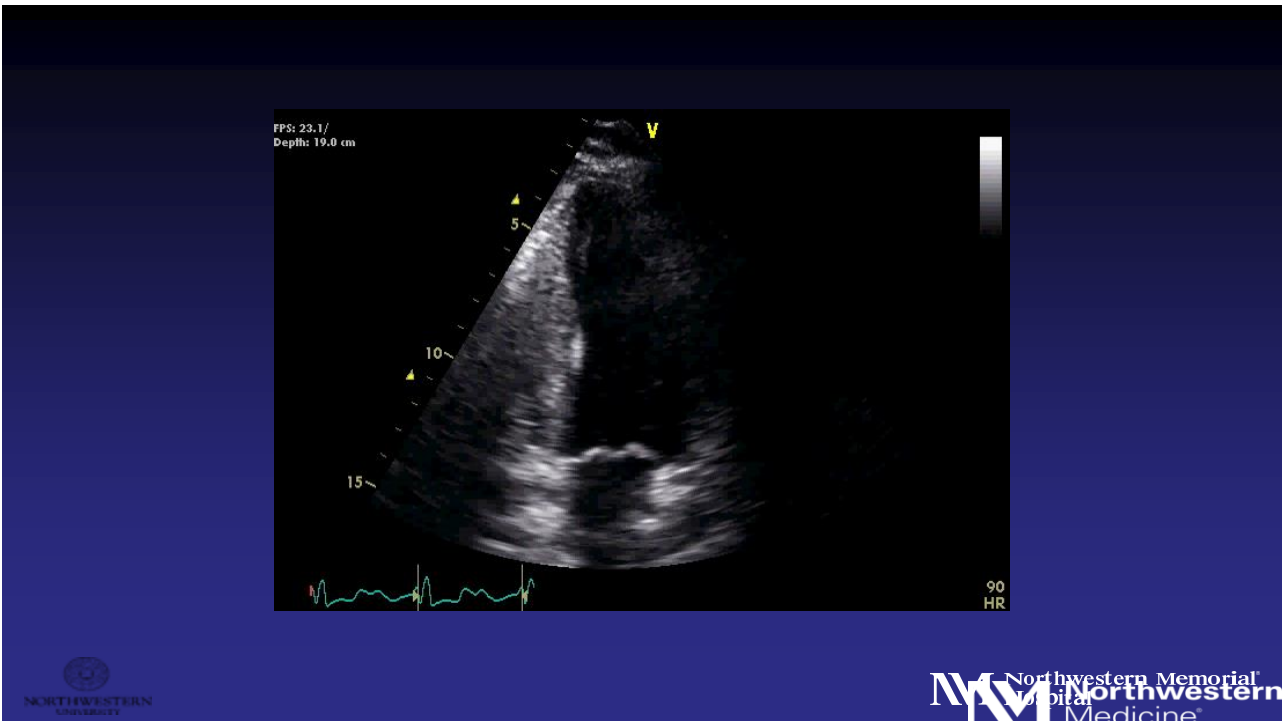
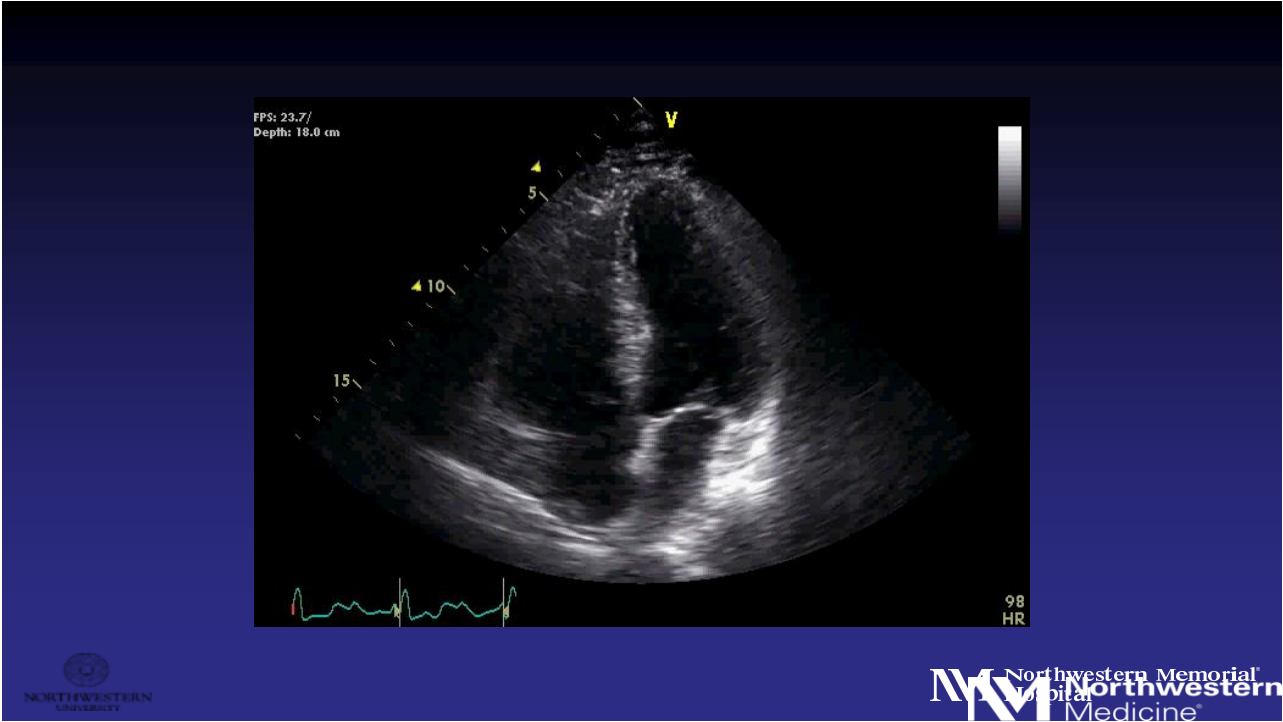
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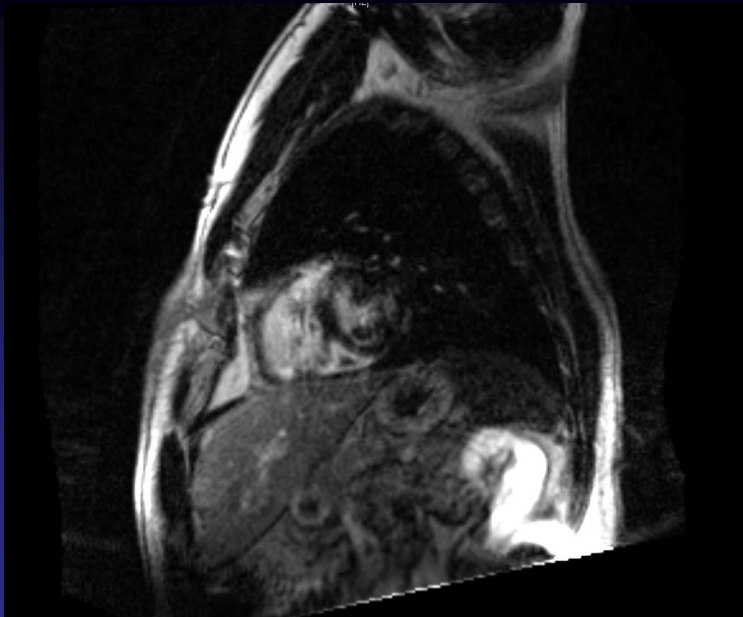
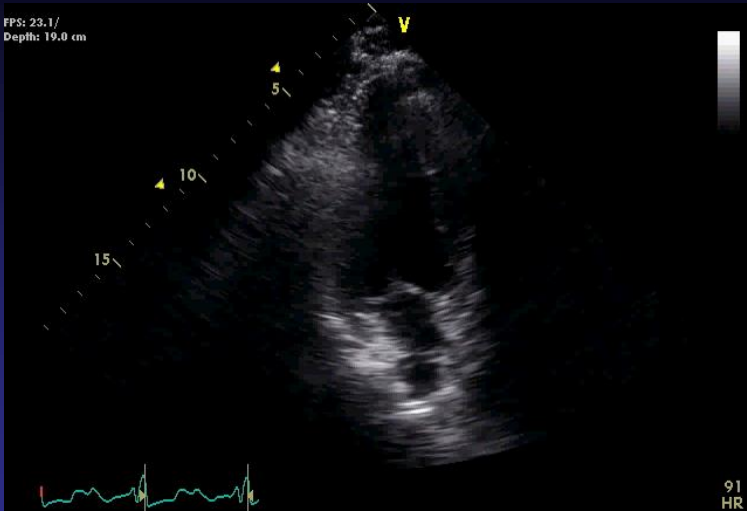
Case 1

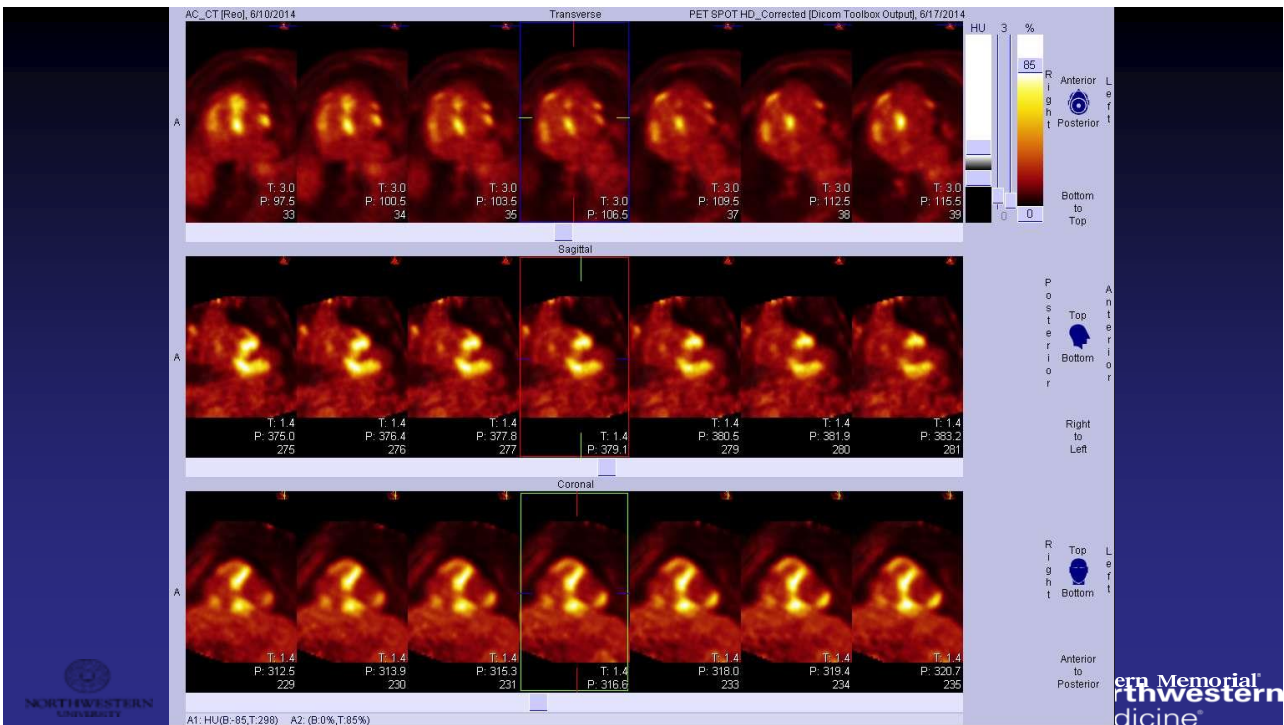
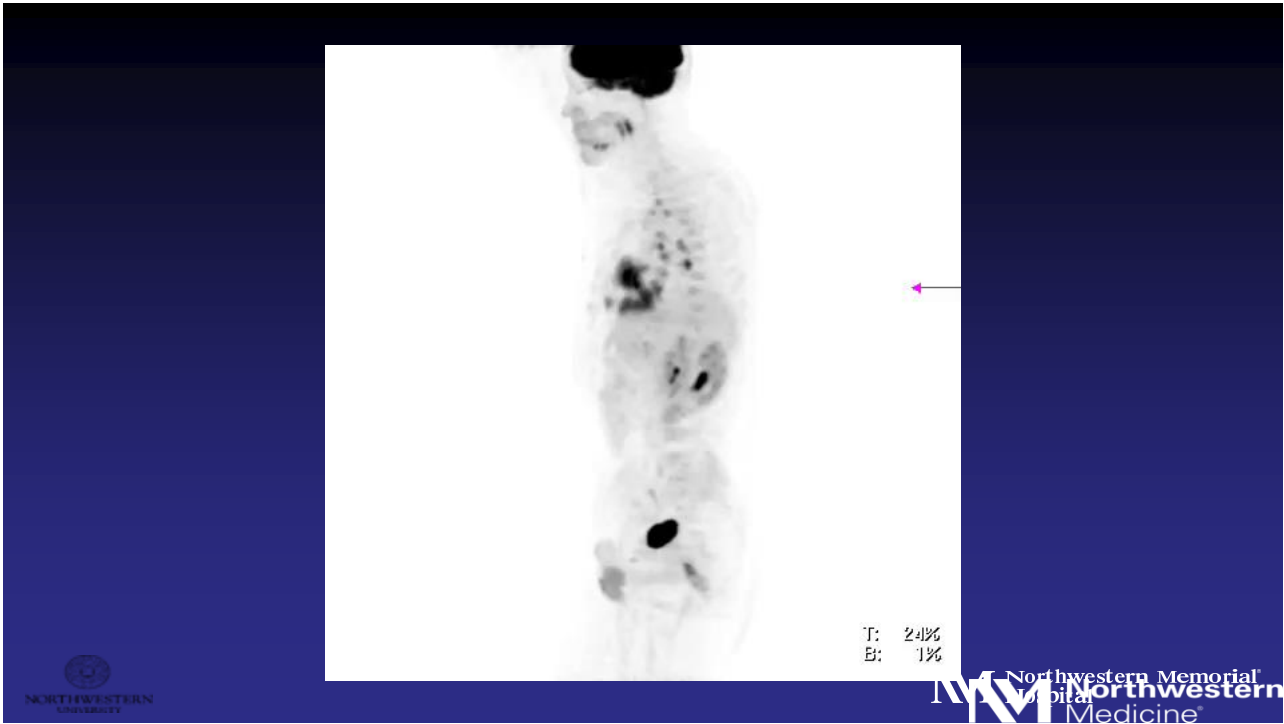
- *56 yr old male with no past medical history who presented with dyspnea and palpitations*

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Hospital Course

- *Endobronchial biopsy: +Sarcoidosis*
- *ICD/pacer implanted*

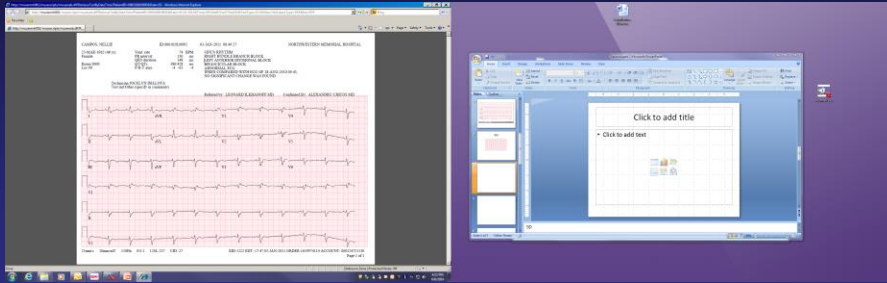


Case 2

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

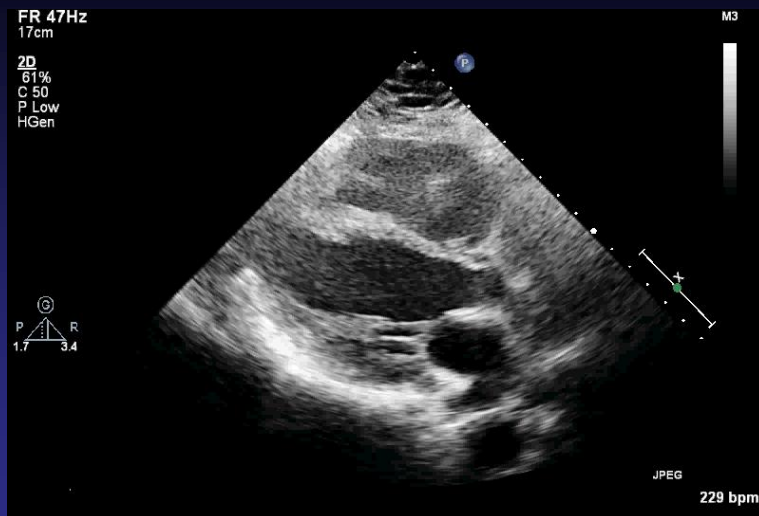
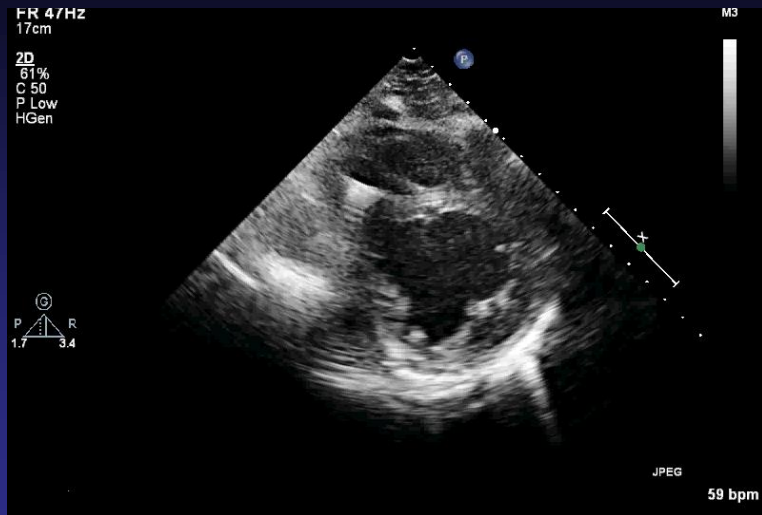


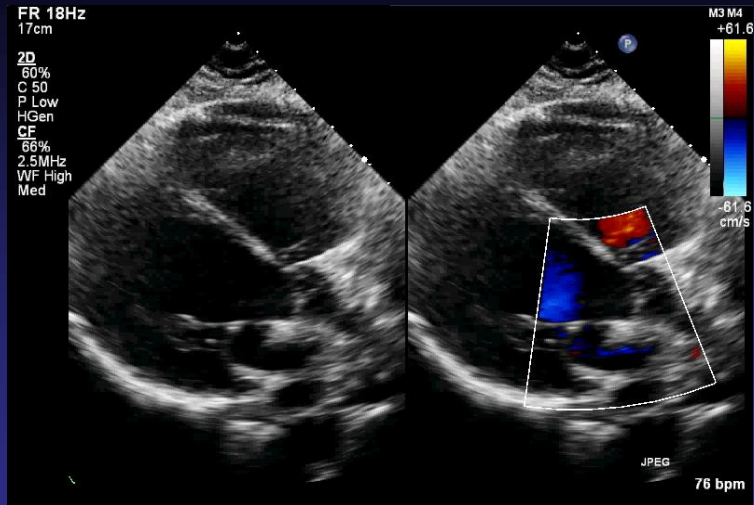
ECG

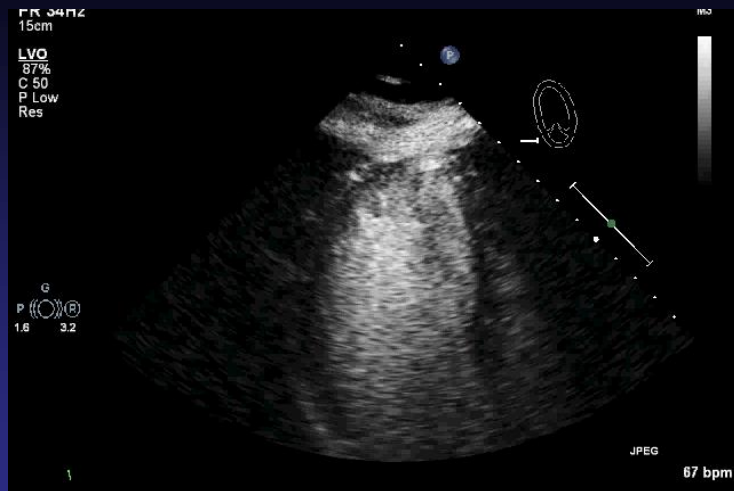
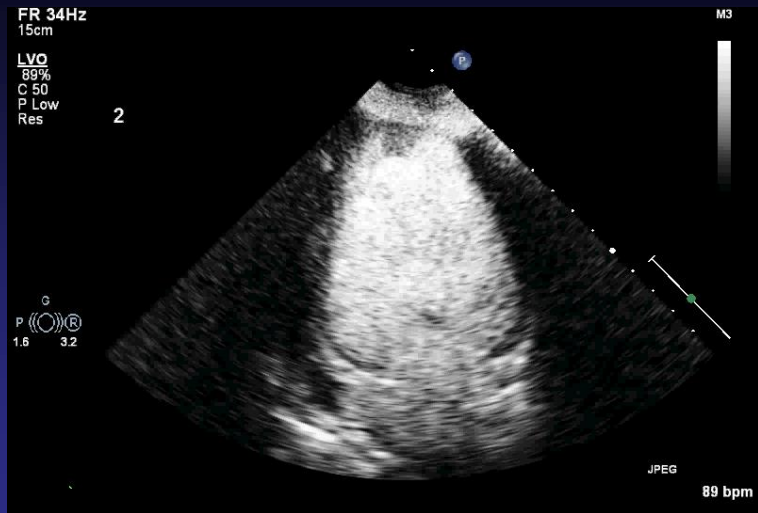


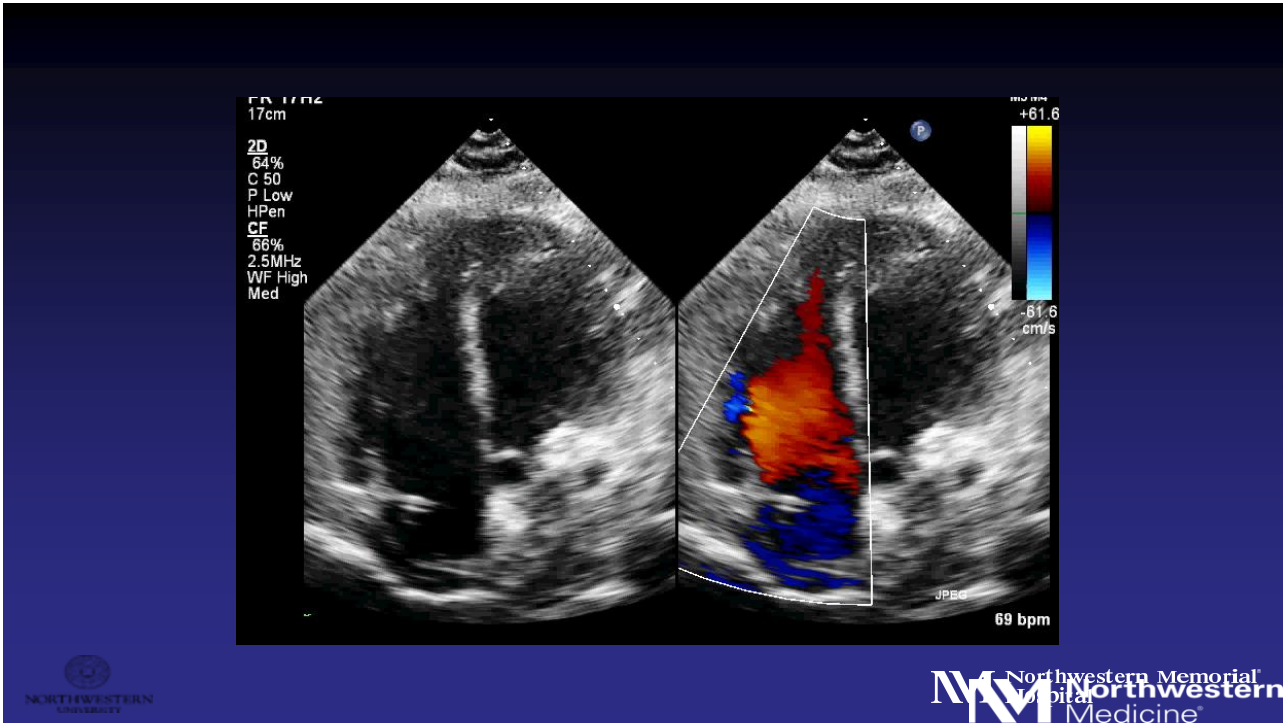
- *Cardiac cath: No obstructive CAD*
- *Cardiac MRI: Myocardial infiltration suspicious for sarcoidosis*
- *Endomyocardial biopsy: +Sarcoidosis*

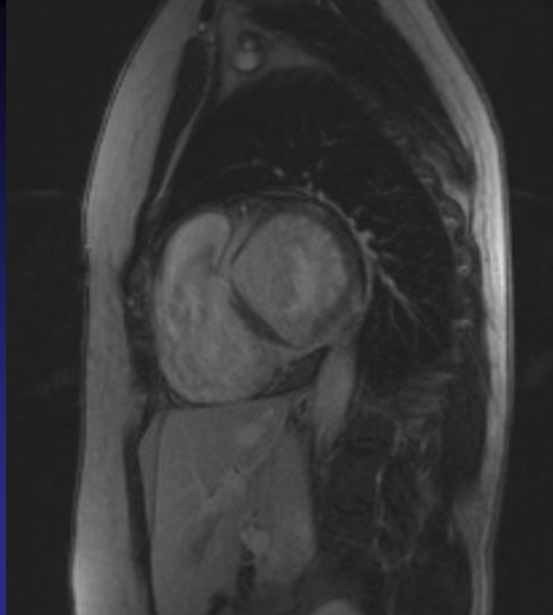
Echo











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

Summary

- *Echo is usually the first imaging modality used to image the heart in pts with systemic diseases*
- *Thorough knowledge of the cardiac manifestations of systemic diseases is necessary*
- *Scleroderma can have primary and secondary effects on the heart – know how to differentiate!*
- *Cardiac sarcoidosis can range from subtle to extreme cardiac abnormalities. Have a high index of suspicion, especially when arrhythmias and/or conduction disease present*



Thank You

