

Cases in Adult Congenital Heart Disease

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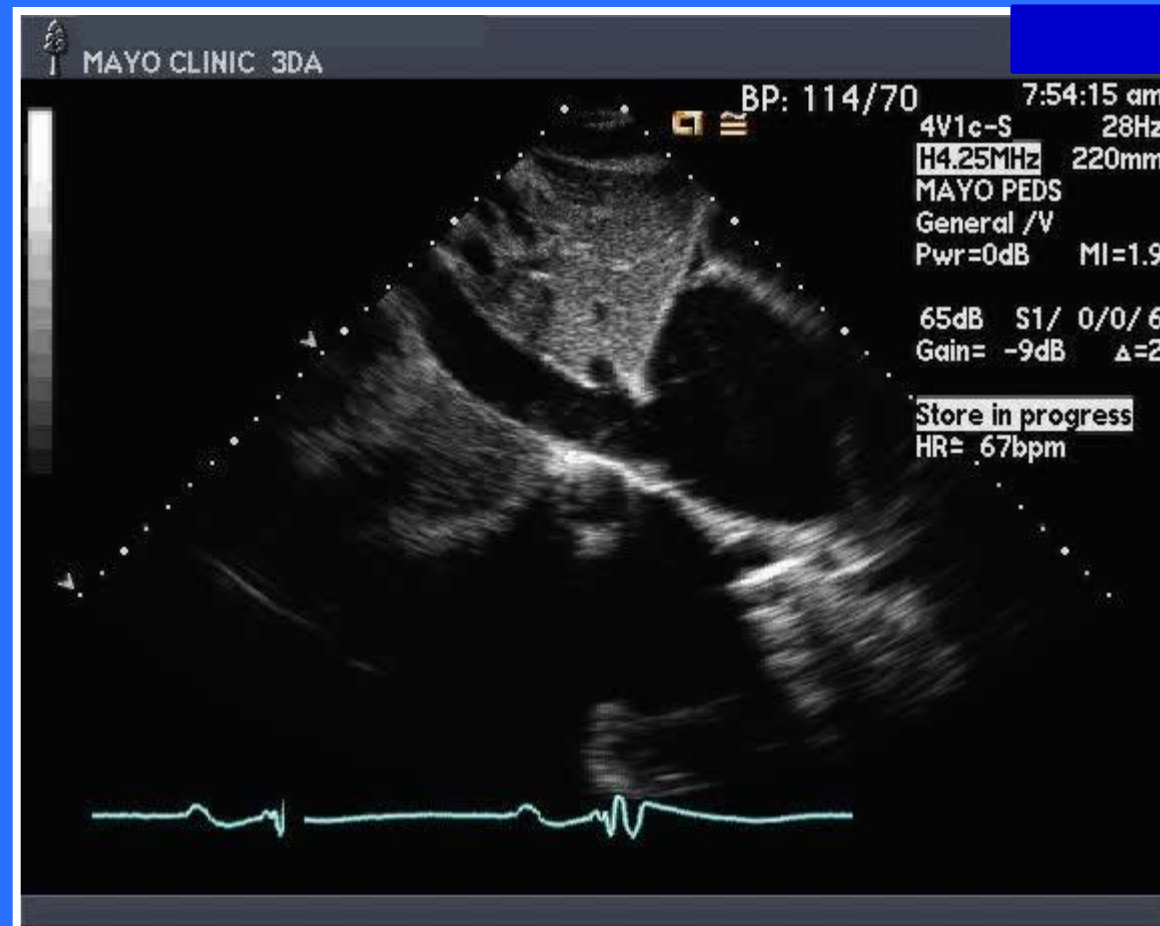
➤ **No Disclosures**

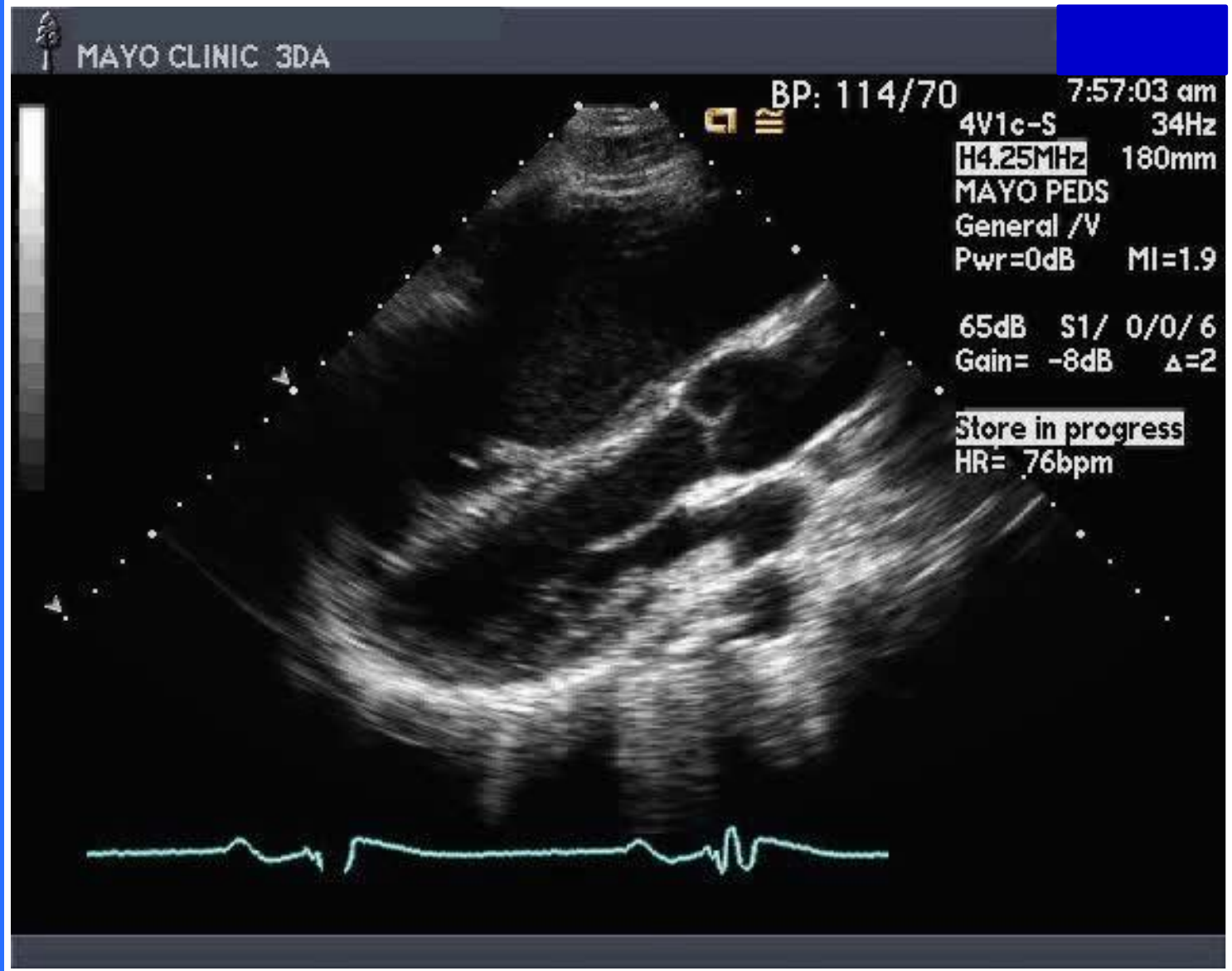
“I Have Palpitations”

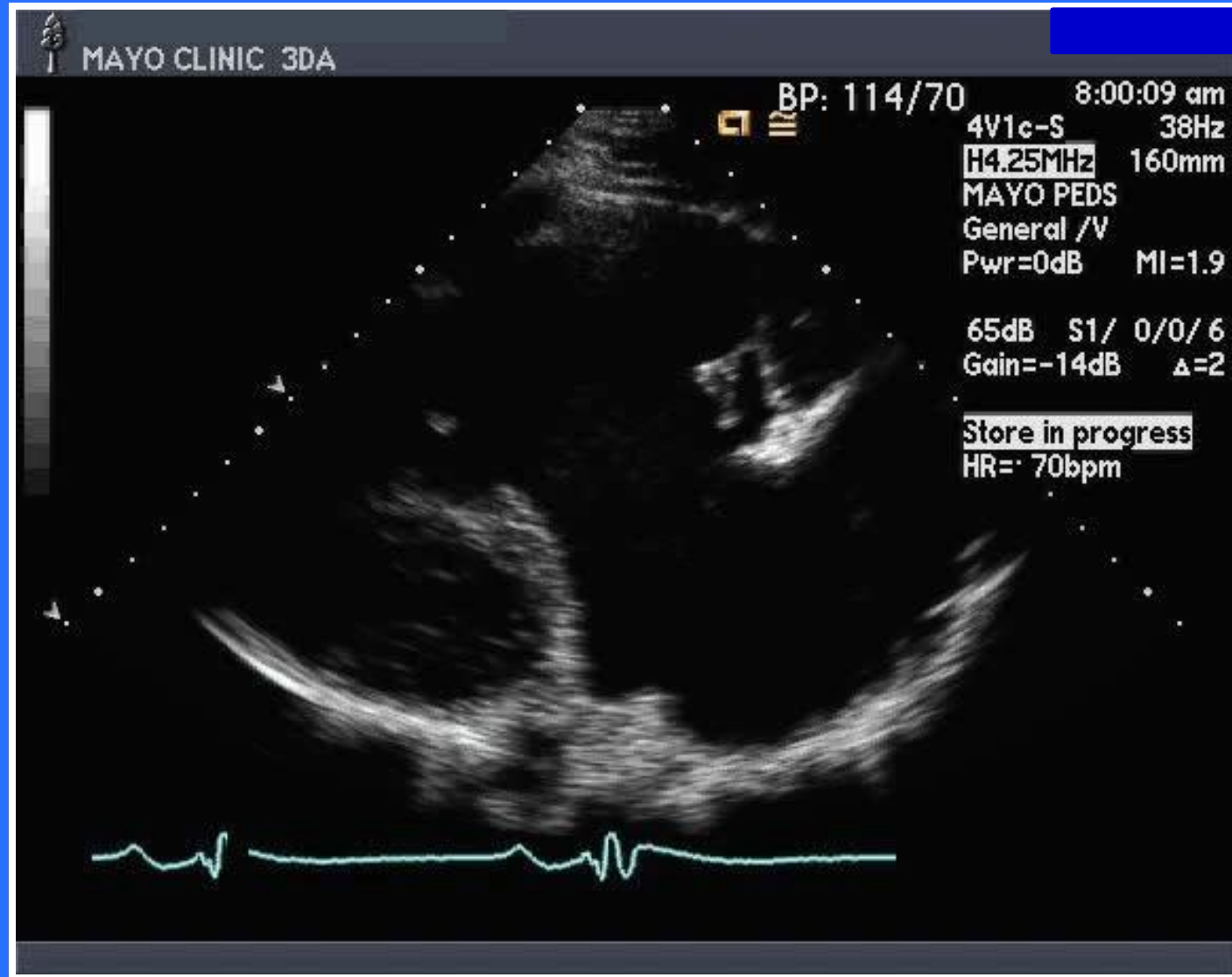
18 Year old Man

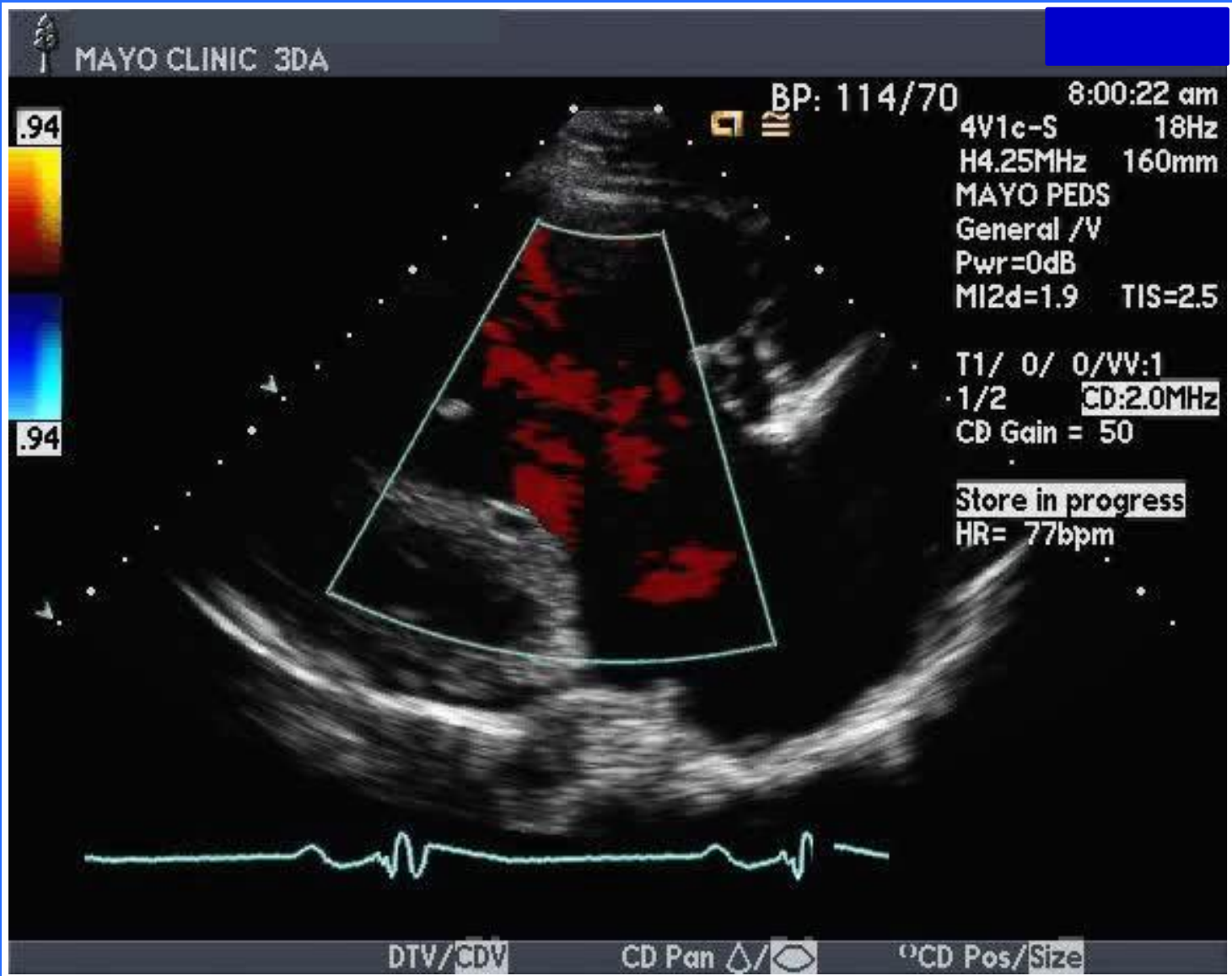
- Palpitations
- “abnormal” ecg and cxr

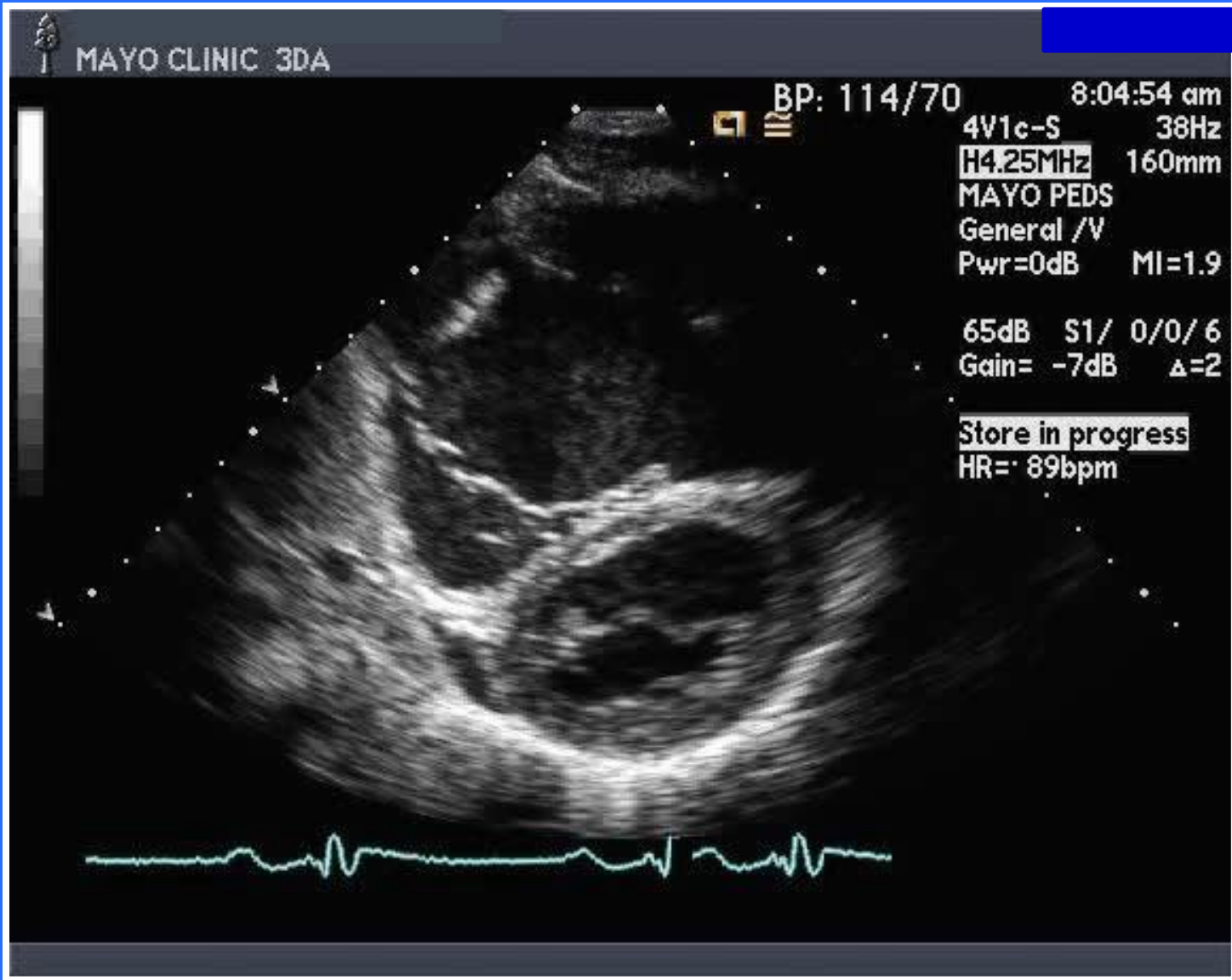
ECHO

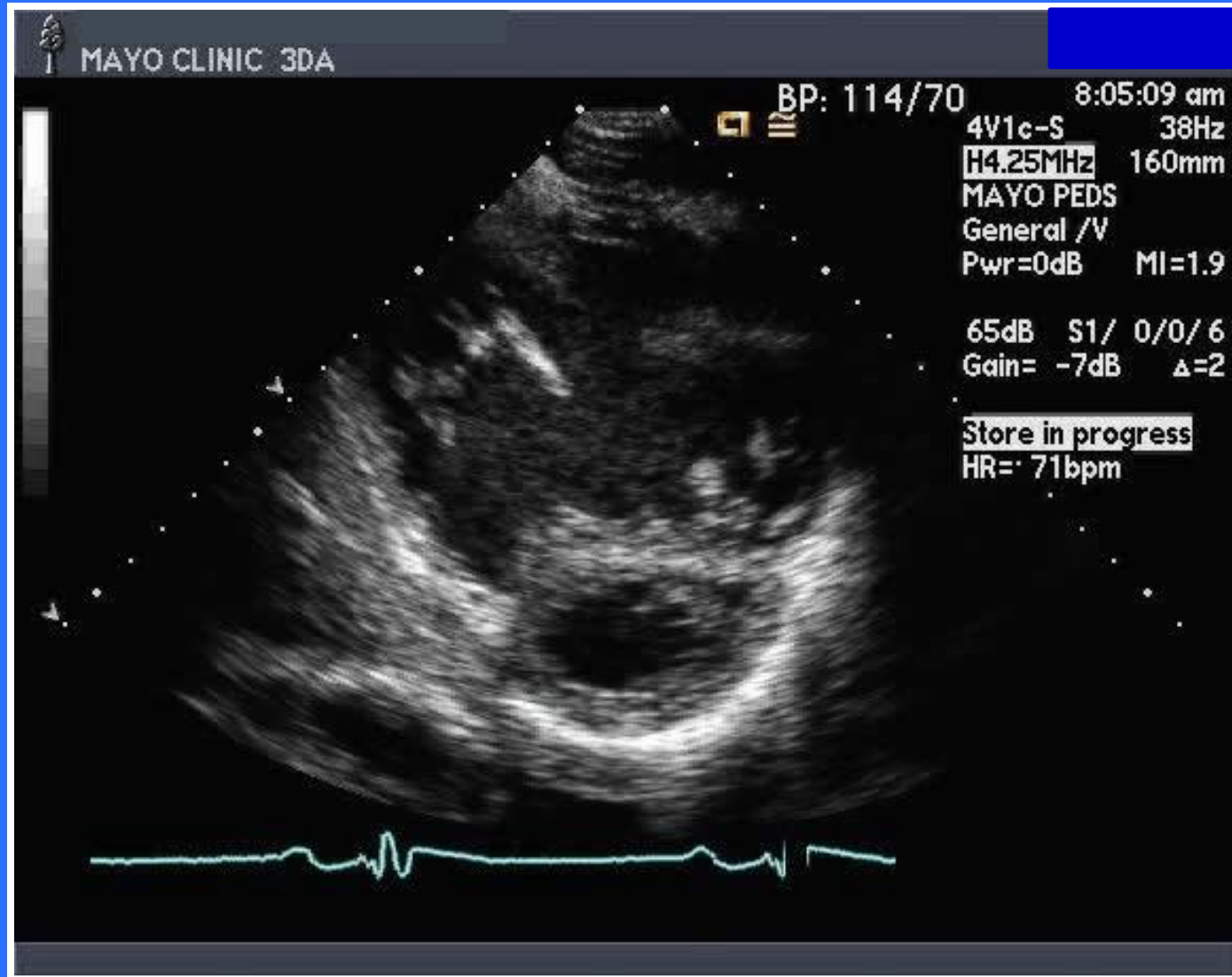


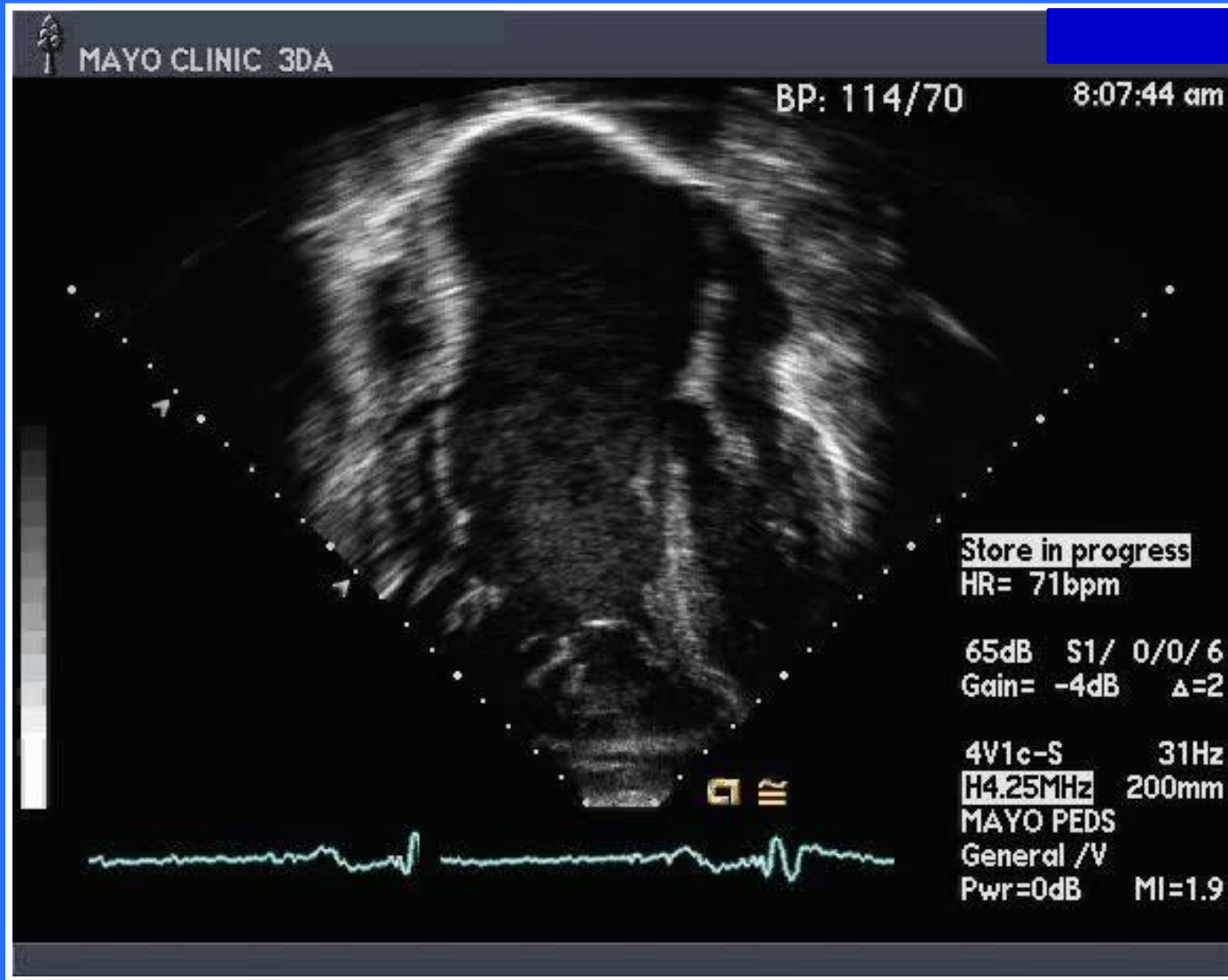








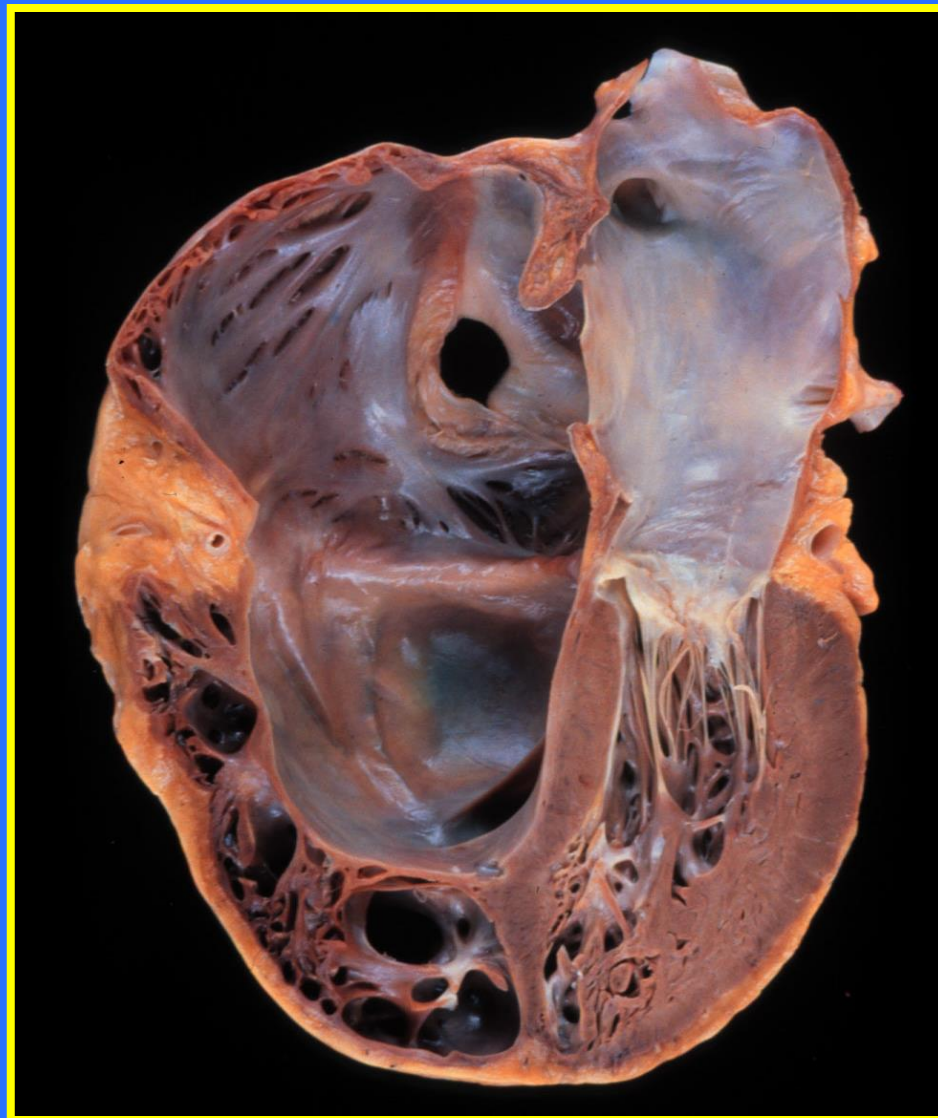




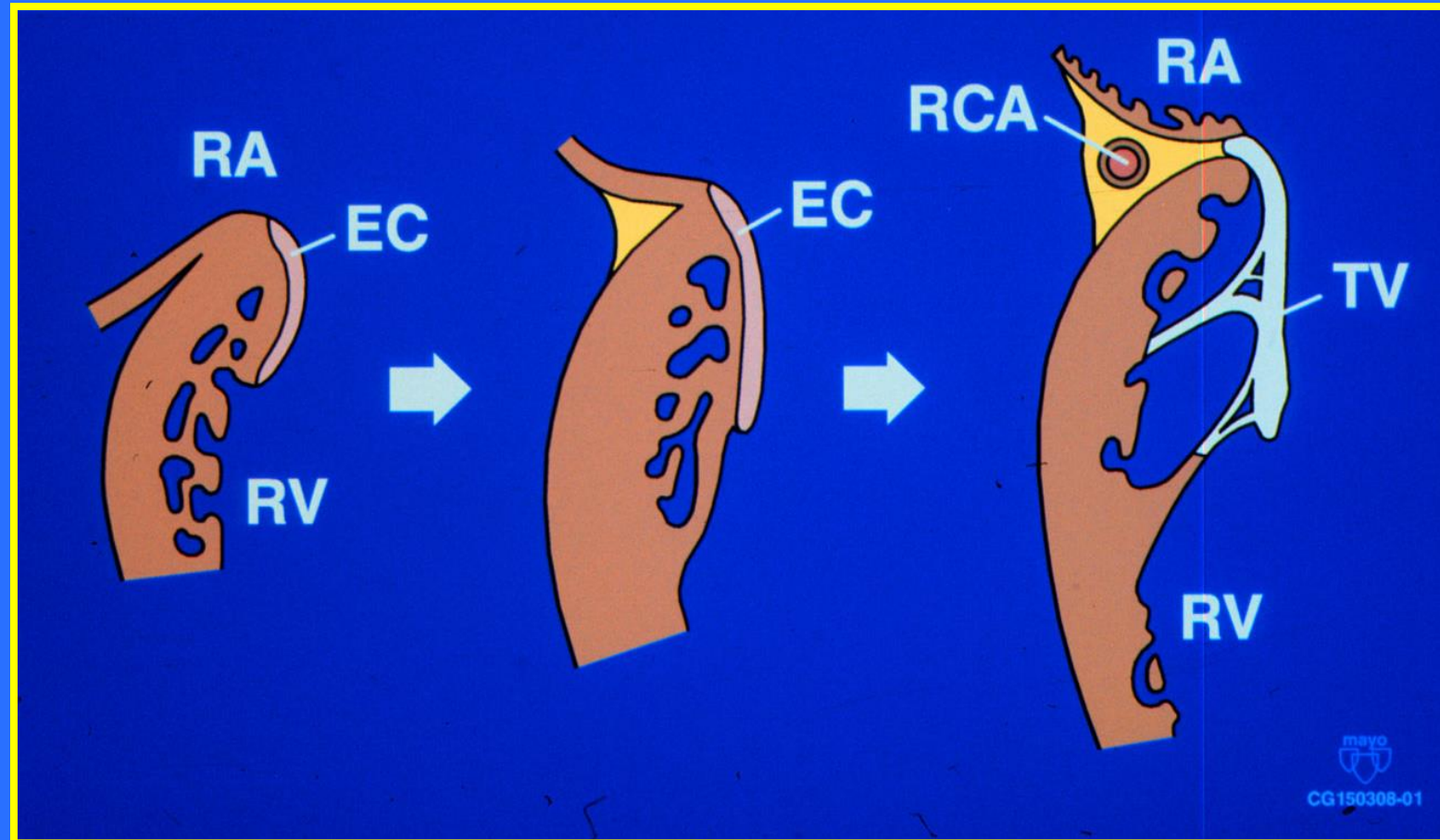




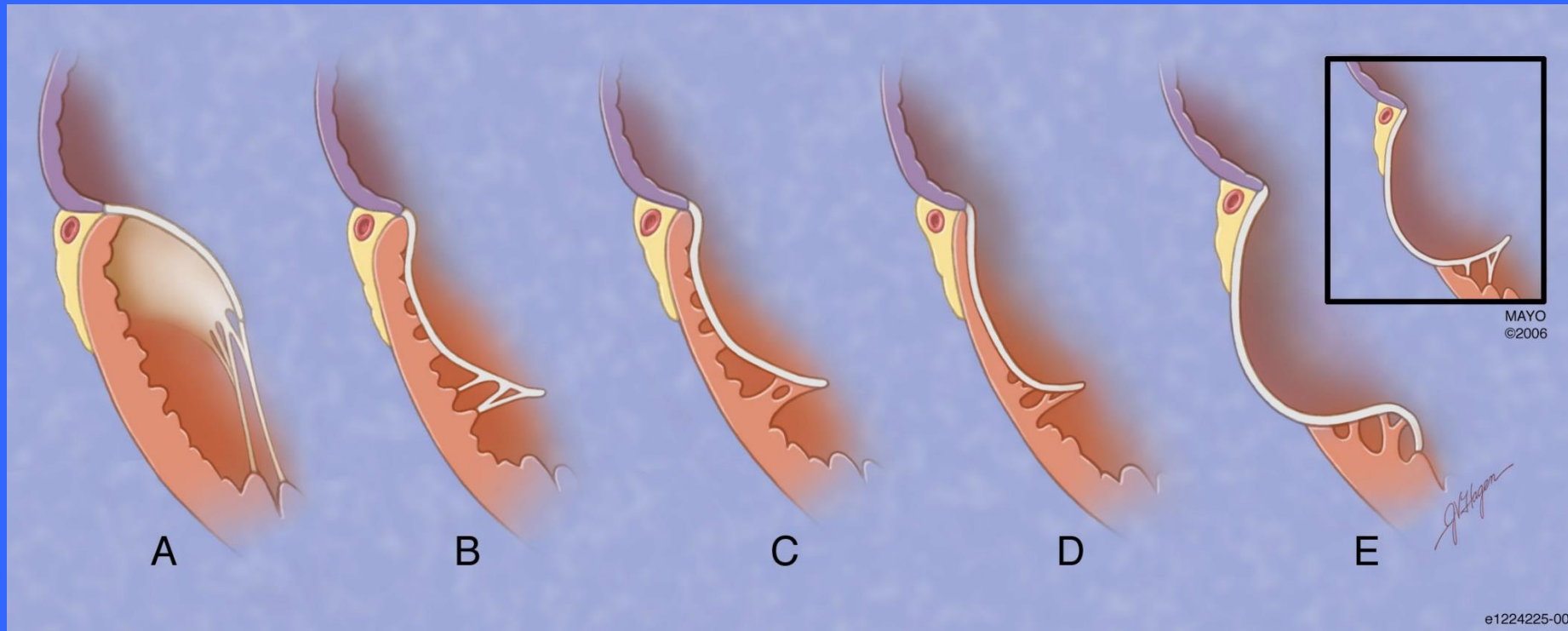
Ebstein Anomaly



Normal Delamination of the TV from the RV Myocardium

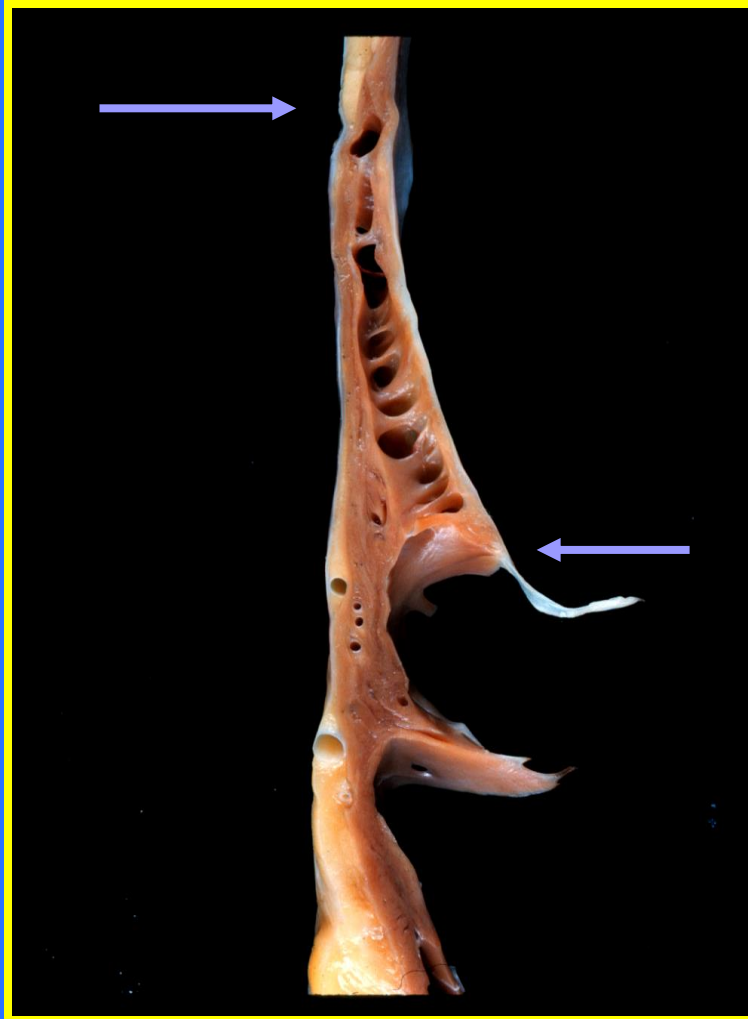


Failure of Delamination From the Myocardium



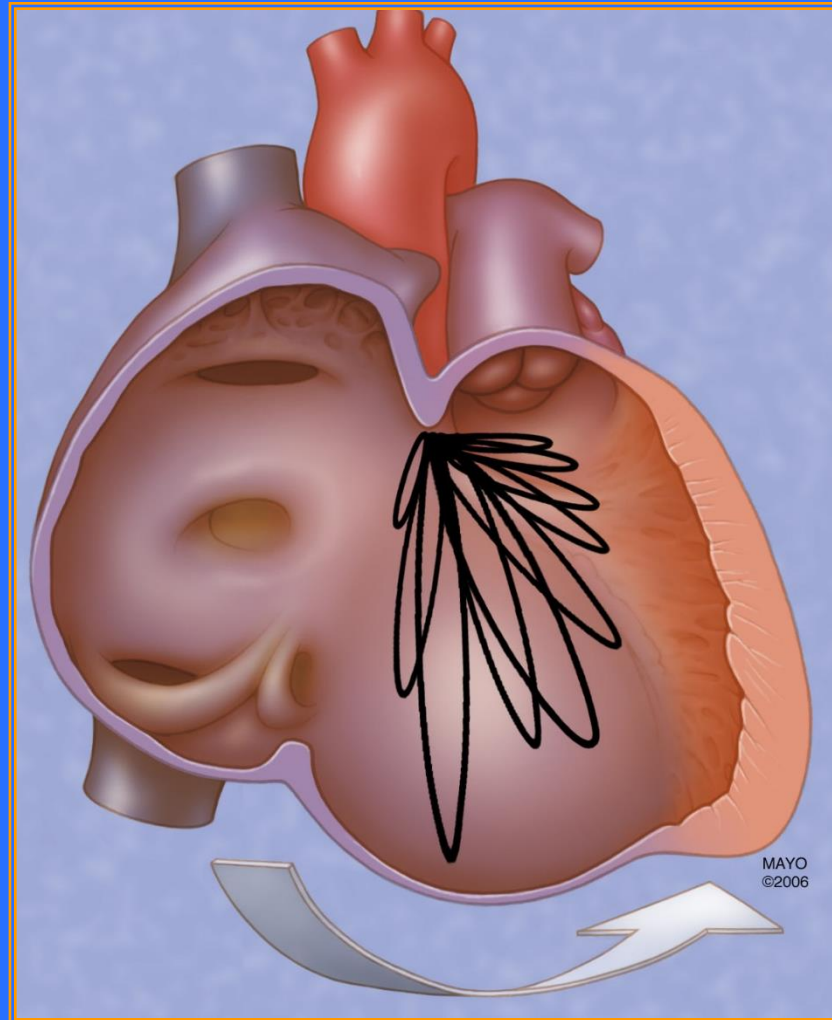
Spectrum with Infinite Variability

Failed Delamination results in ...



- adherence of leaflets to underlying RV myocardium
- displacement of the annular hinge points

Displacement Apically AND Toward the Right Ventricular Outflow Tract



Echocardiographic Diagnosis

- Apical displacement of the septal leaflet of the tricuspid valve $> 8\text{mm/m}^2$
- Right sided chamber enlargement with “atrialized” RV
- Tricuspid valve regurgitation – often appears laminar
- Elongated, tethered anterior TV leaflet

Ebstein Anomaly Associated Lesions

- Secundum ASD
- RV outflow tract obstruction
- LV non-compaction
- Accessory pathways

Ebstein Anomaly

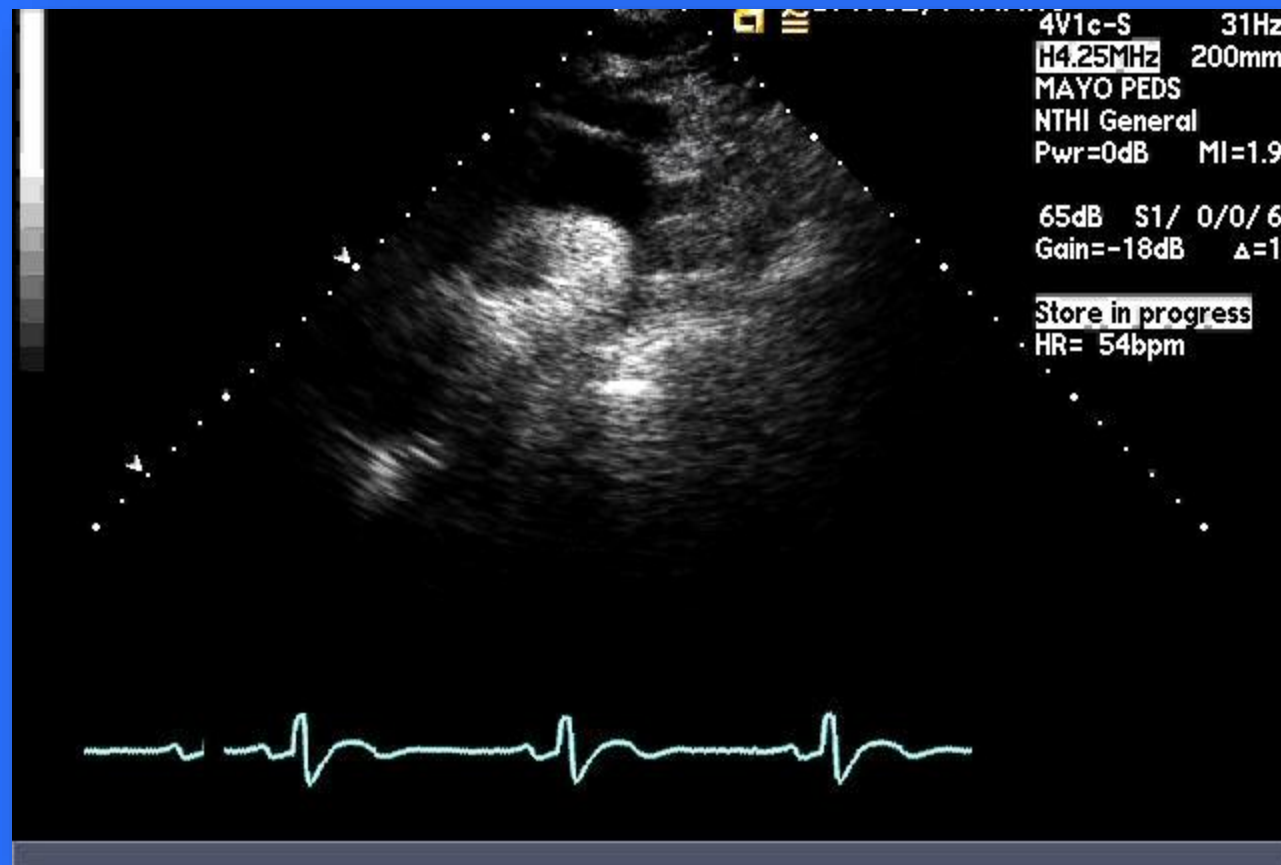
Indications for Operation

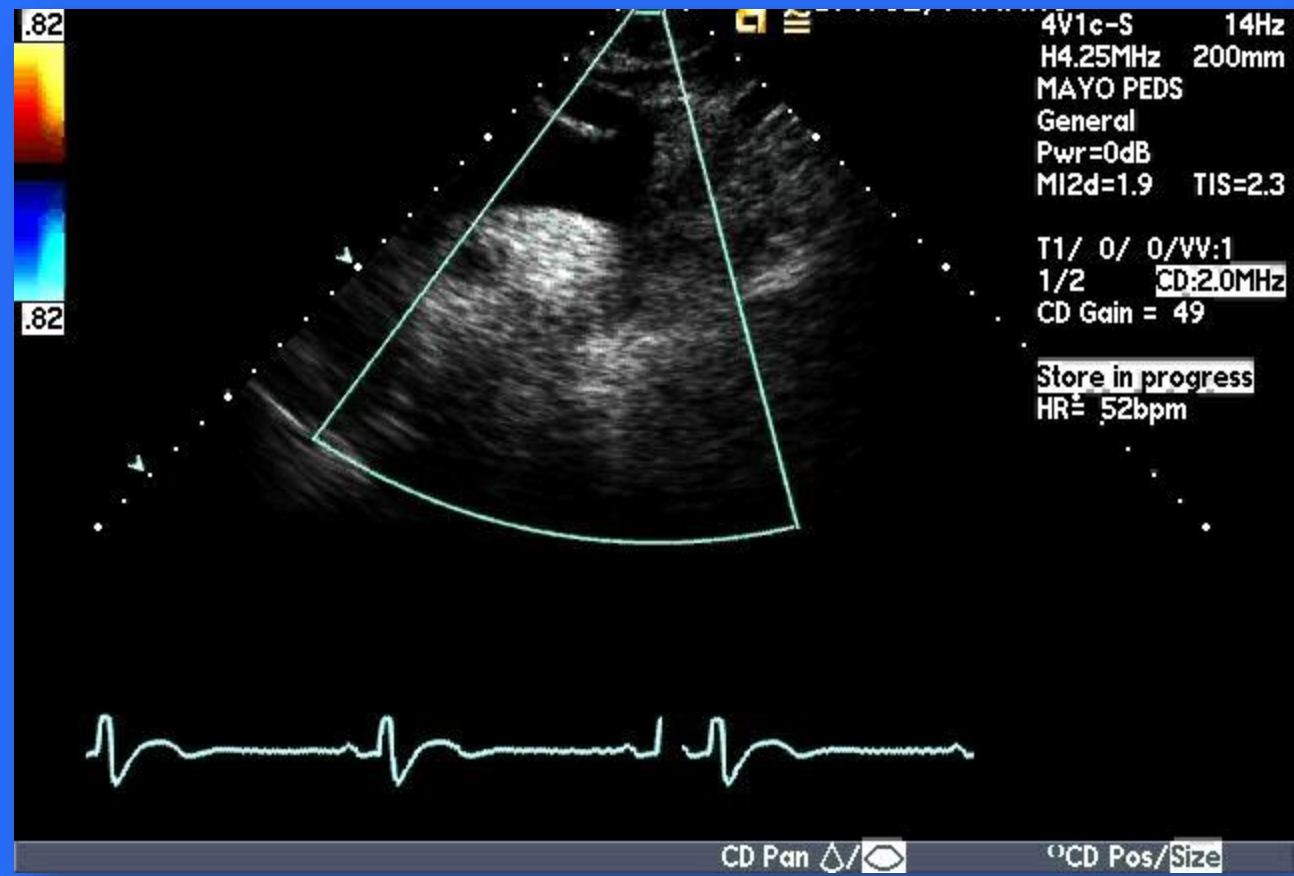
- symptoms, ↓ exercise tolerance, cyanosis
- progressive RV dilatation
- *before* significant RV dysfunction
- onset, progression of atrial arrhythmias
- ? earlier operation if TV repair is likely
- prior to LV dysfunction

“I Have a Headache”

36 Year Old Man

- Undergoing evaluation in neuro for headache
- Found to be hypertensive





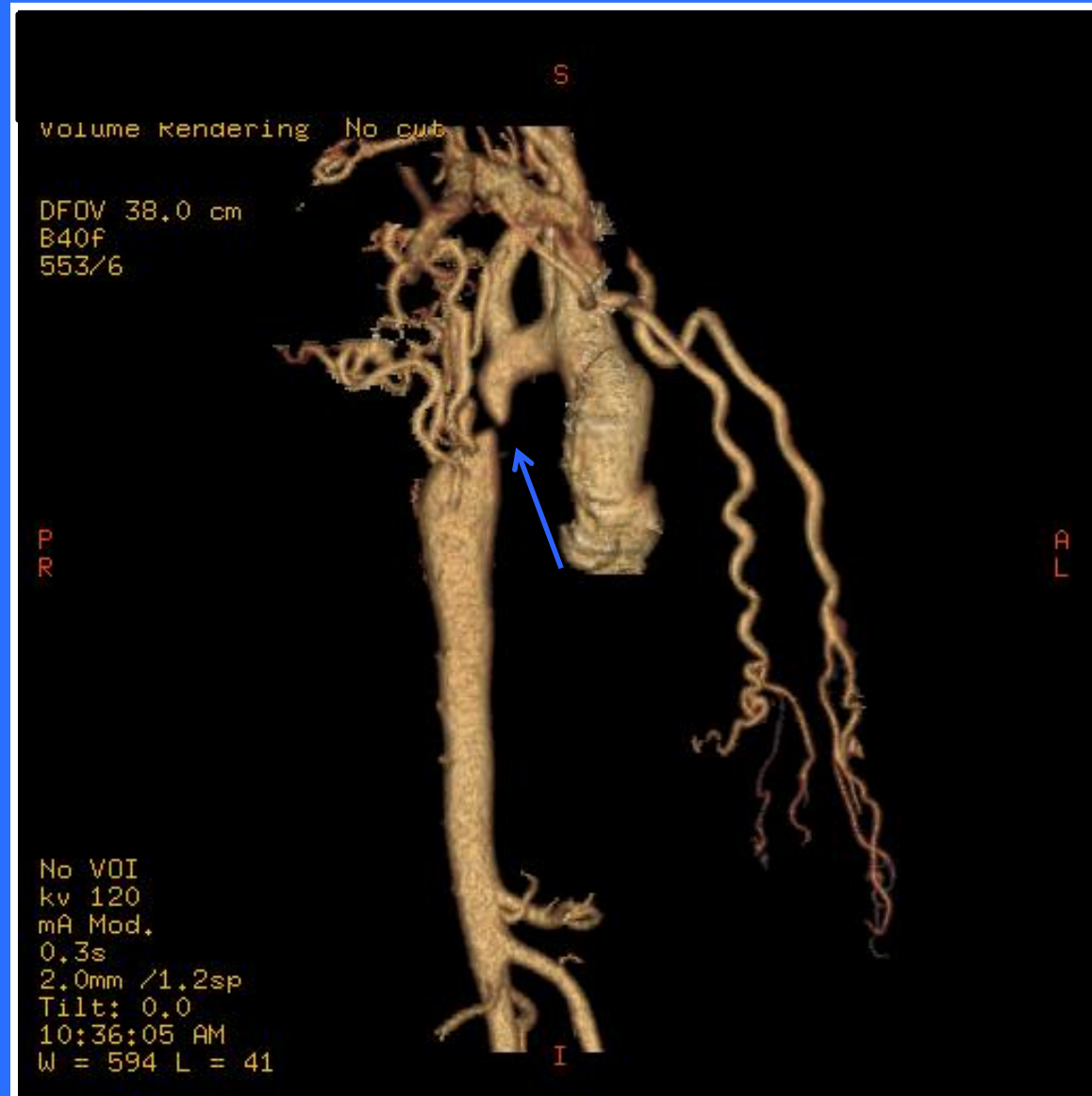


Is This Coarctation?

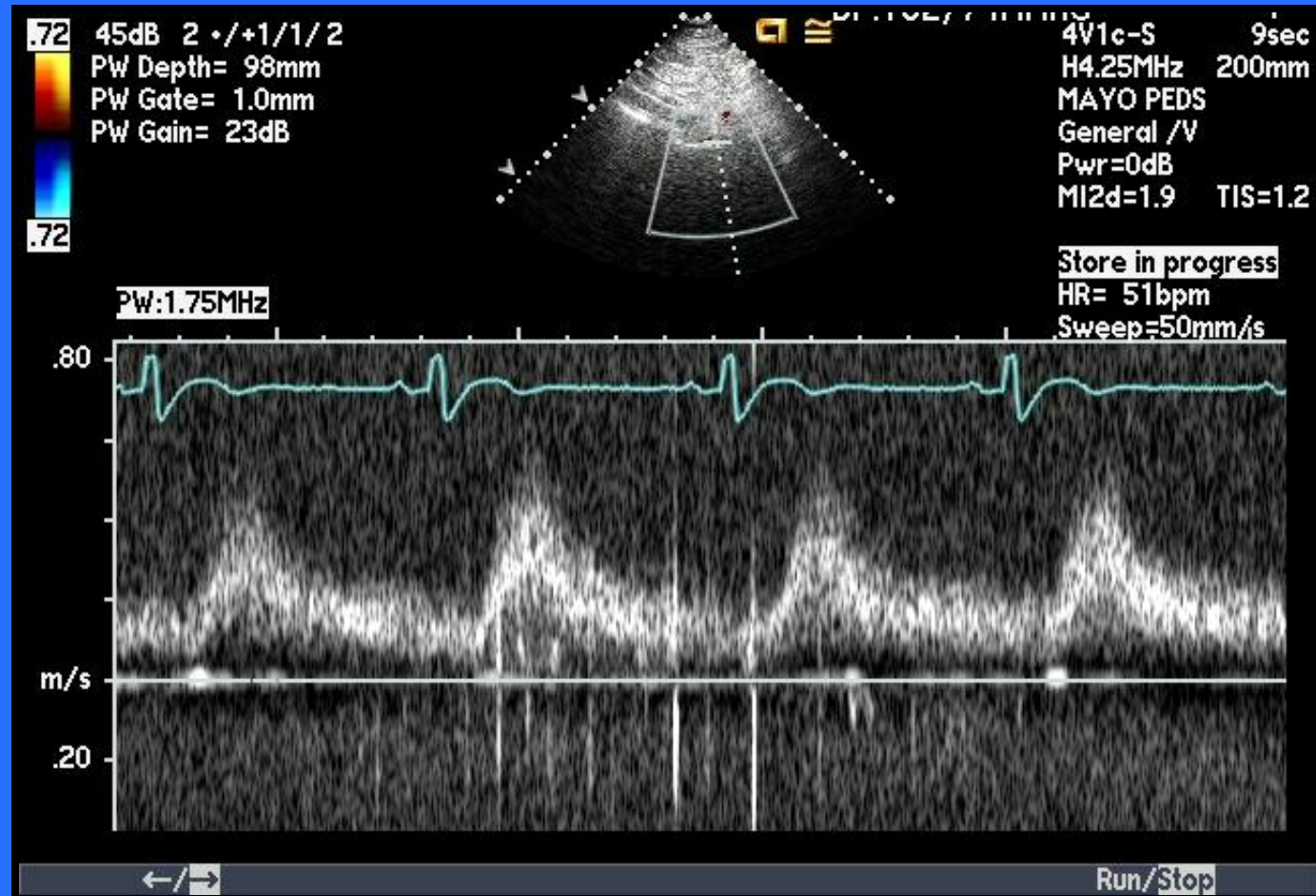
- A. Yes
- B. No
- C. Not Sure

Is This Coarctation?

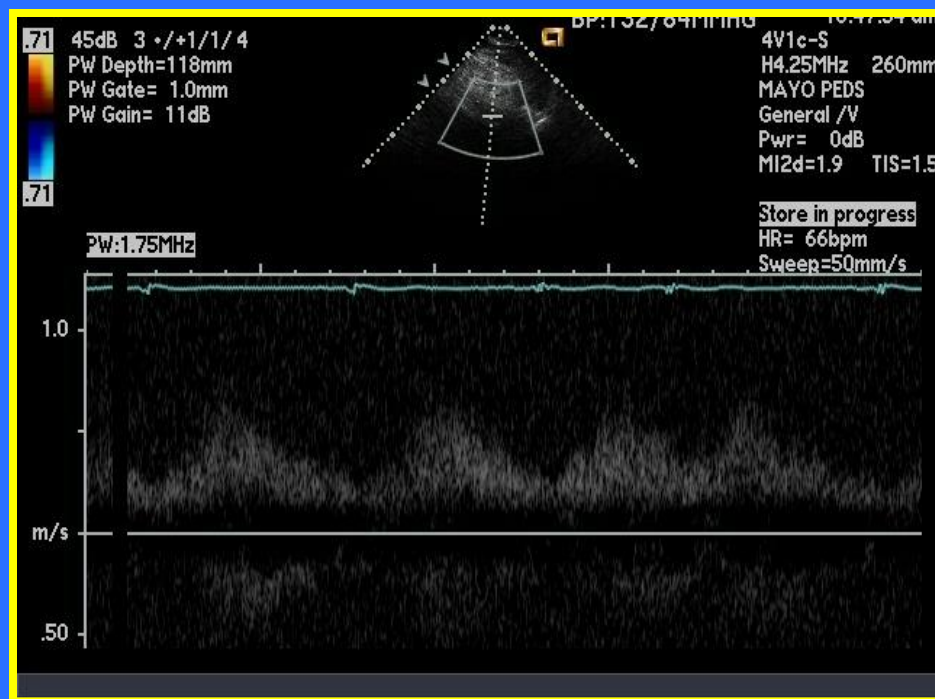
- A. Yes**
- B. No**
- C. Not Sure**



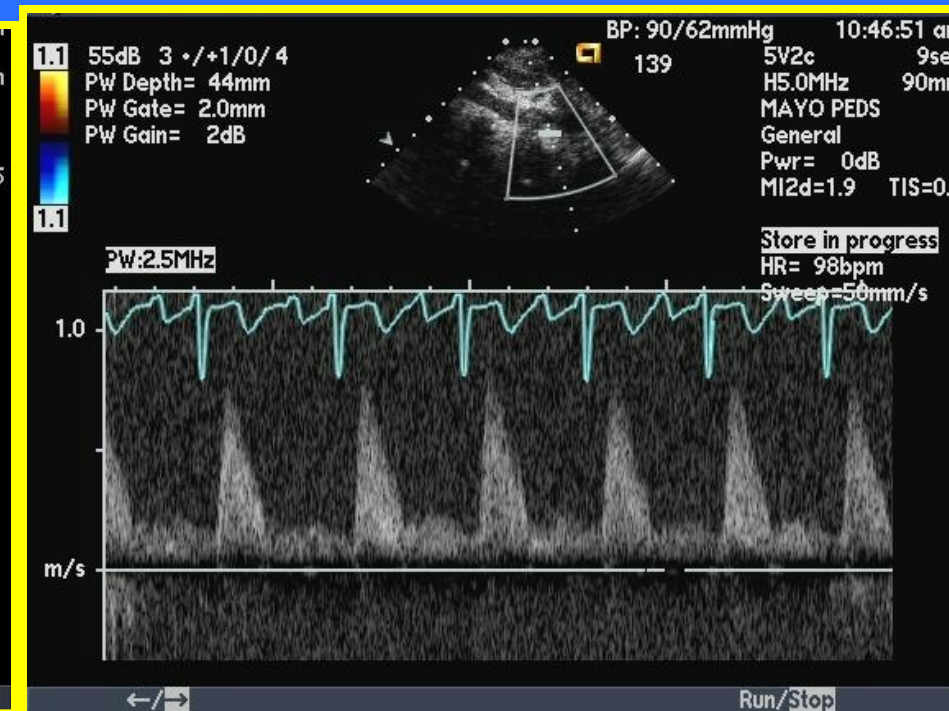
Abdominal Aorta Doppler



Abdominal Aortic Doppler



Significant Coarctation

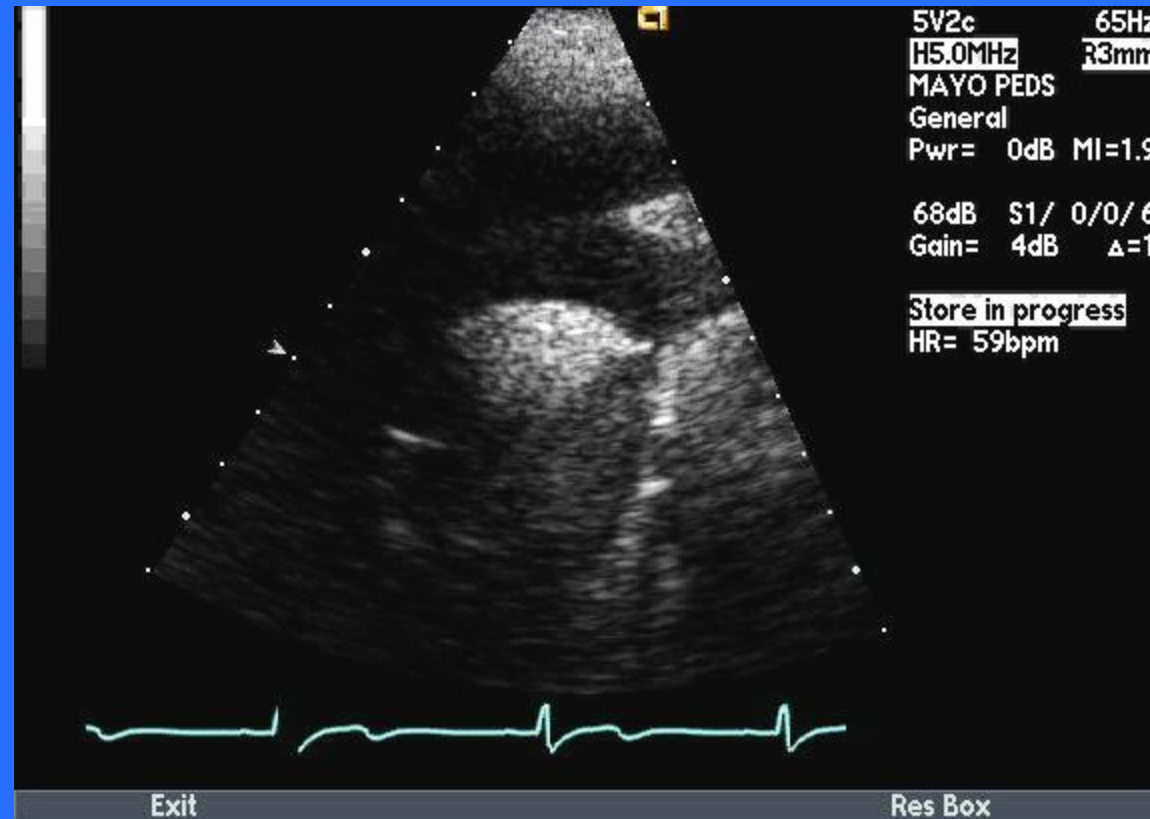


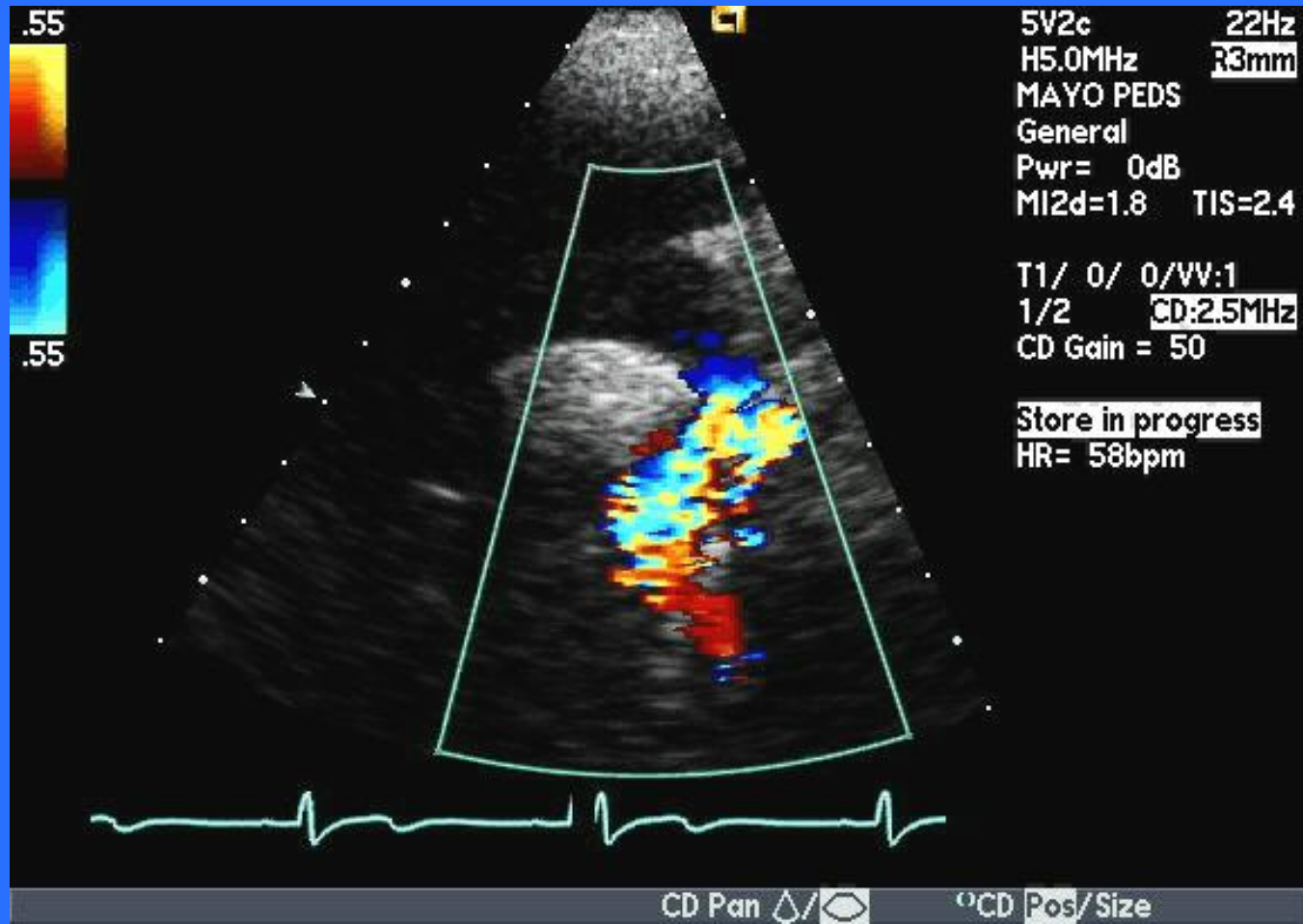
Normal

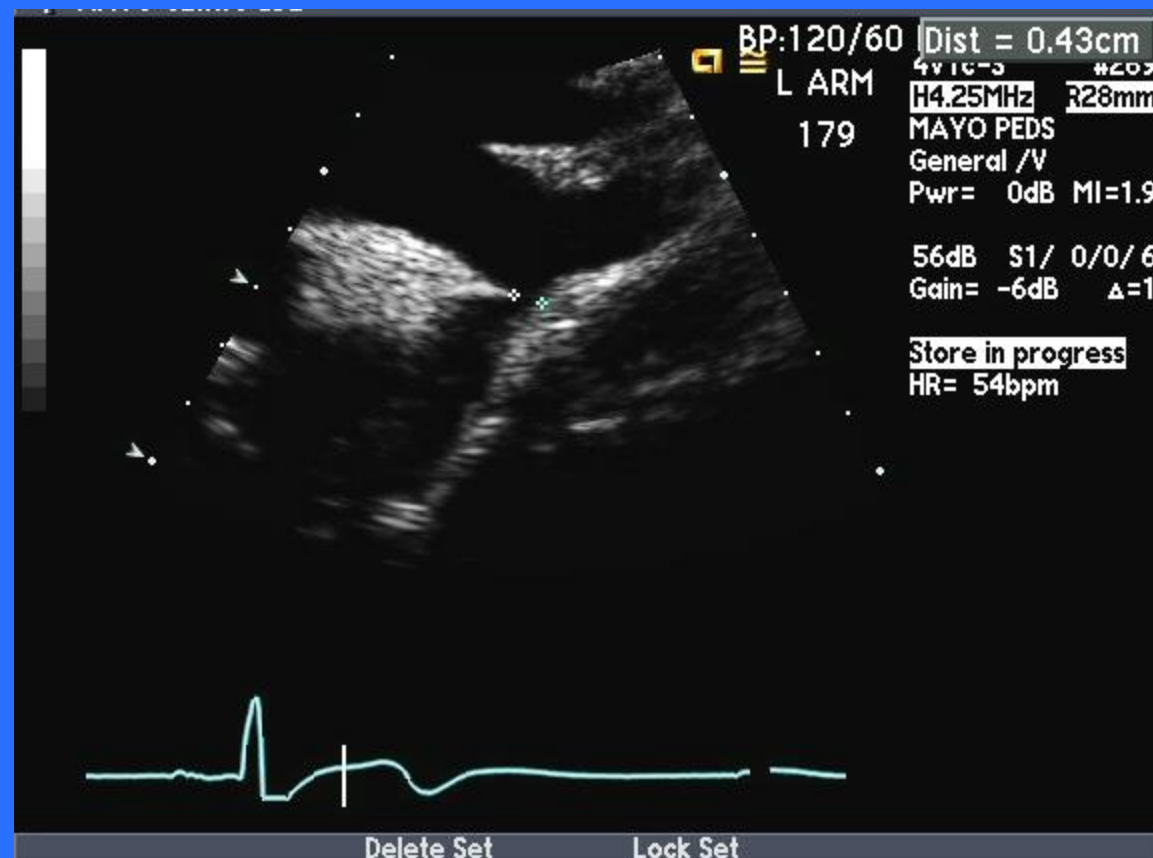
Imaging of Coarctation of the Aorta

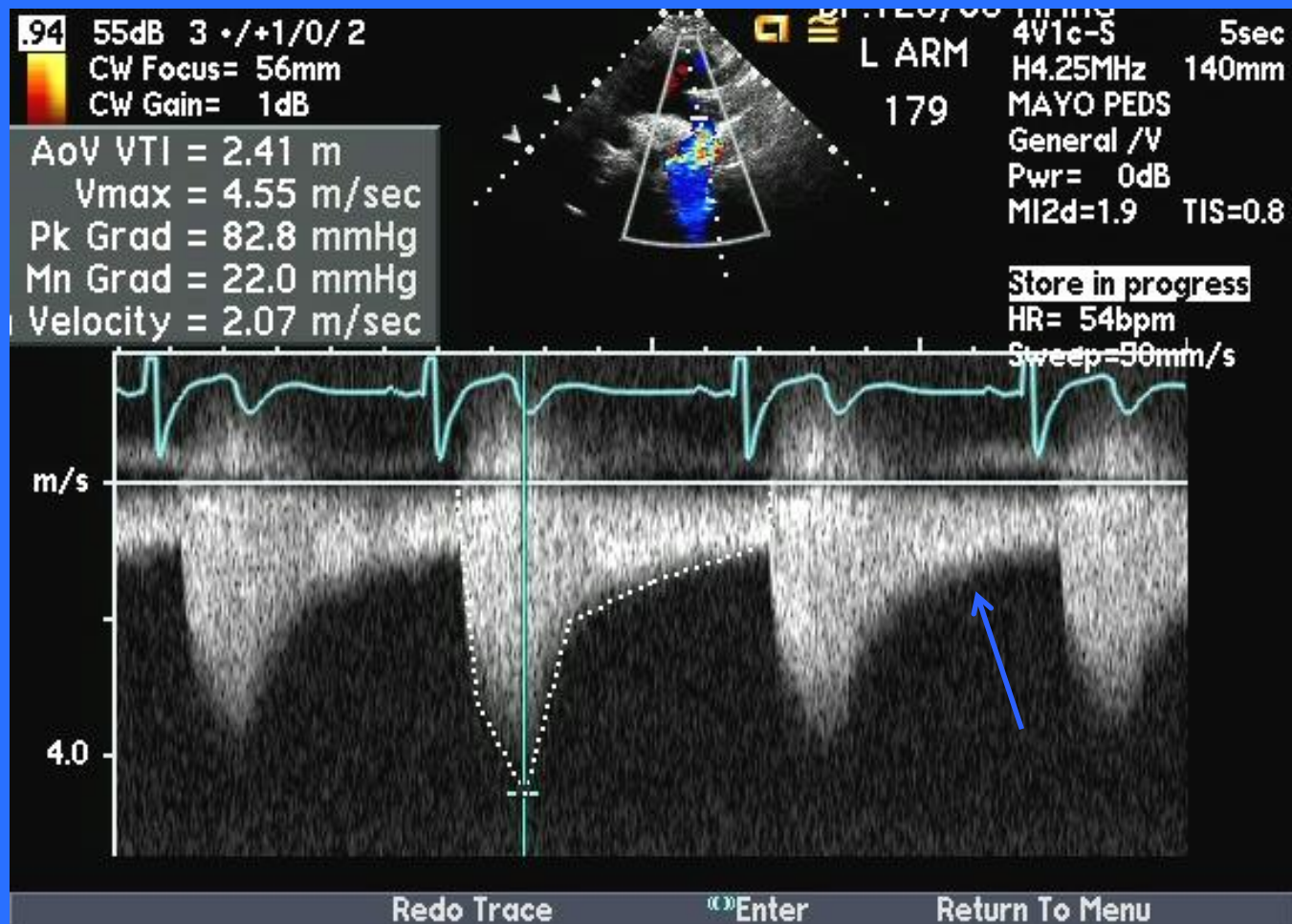
- Abdominal aorta Doppler
- Suprasternal notch imaging
- Parasternal short axis - ?BAV
- Parasternal long axis – ascending aortic dimension

Discrete Coarctation









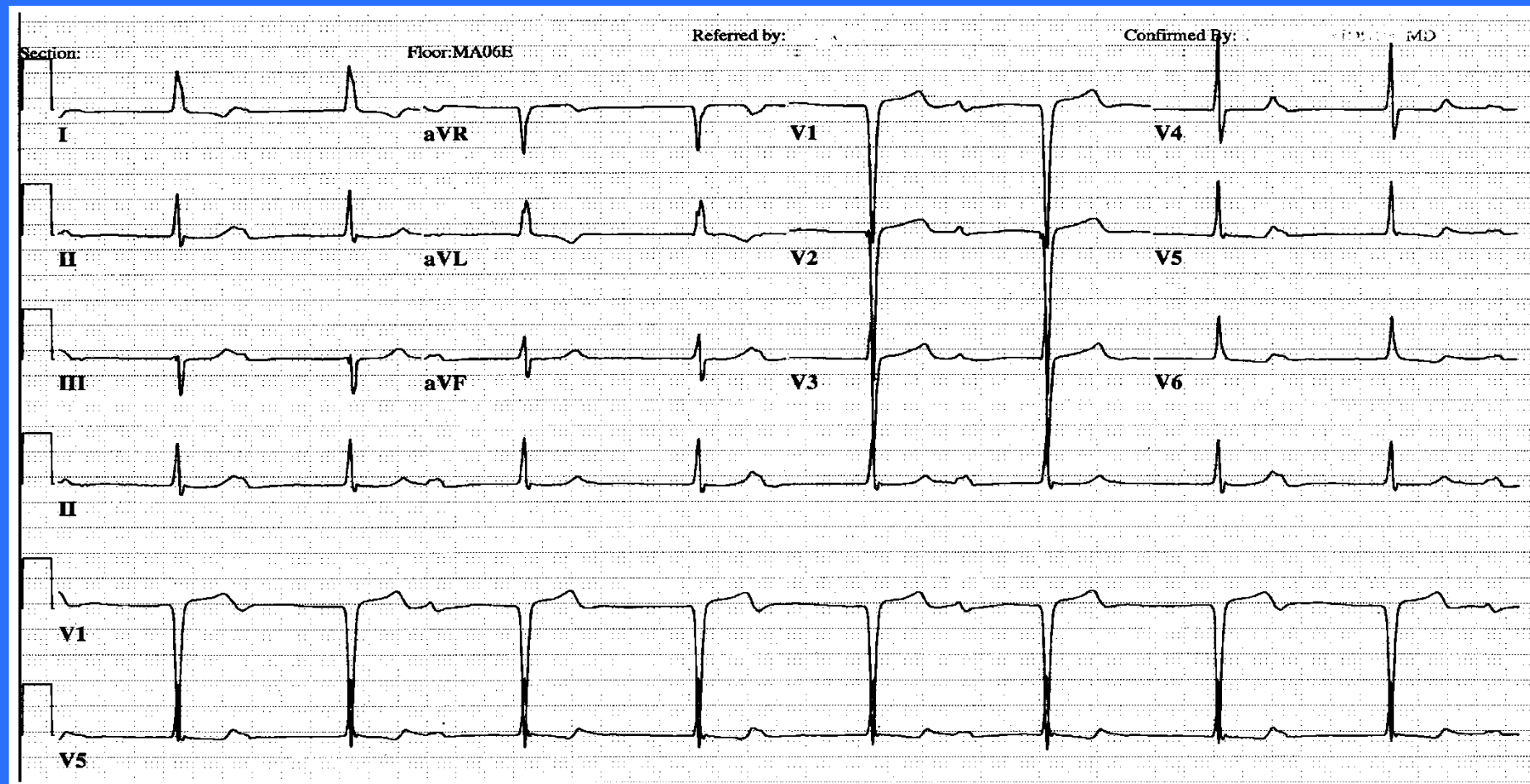
Coarctation Caveats

- Doppler gradient through the coarctation may be low 2° collaterals
- Abdominal Doppler pattern is critical
- Continuous flow in the thoracic aorta is helpful
- Don't forget association to BAV

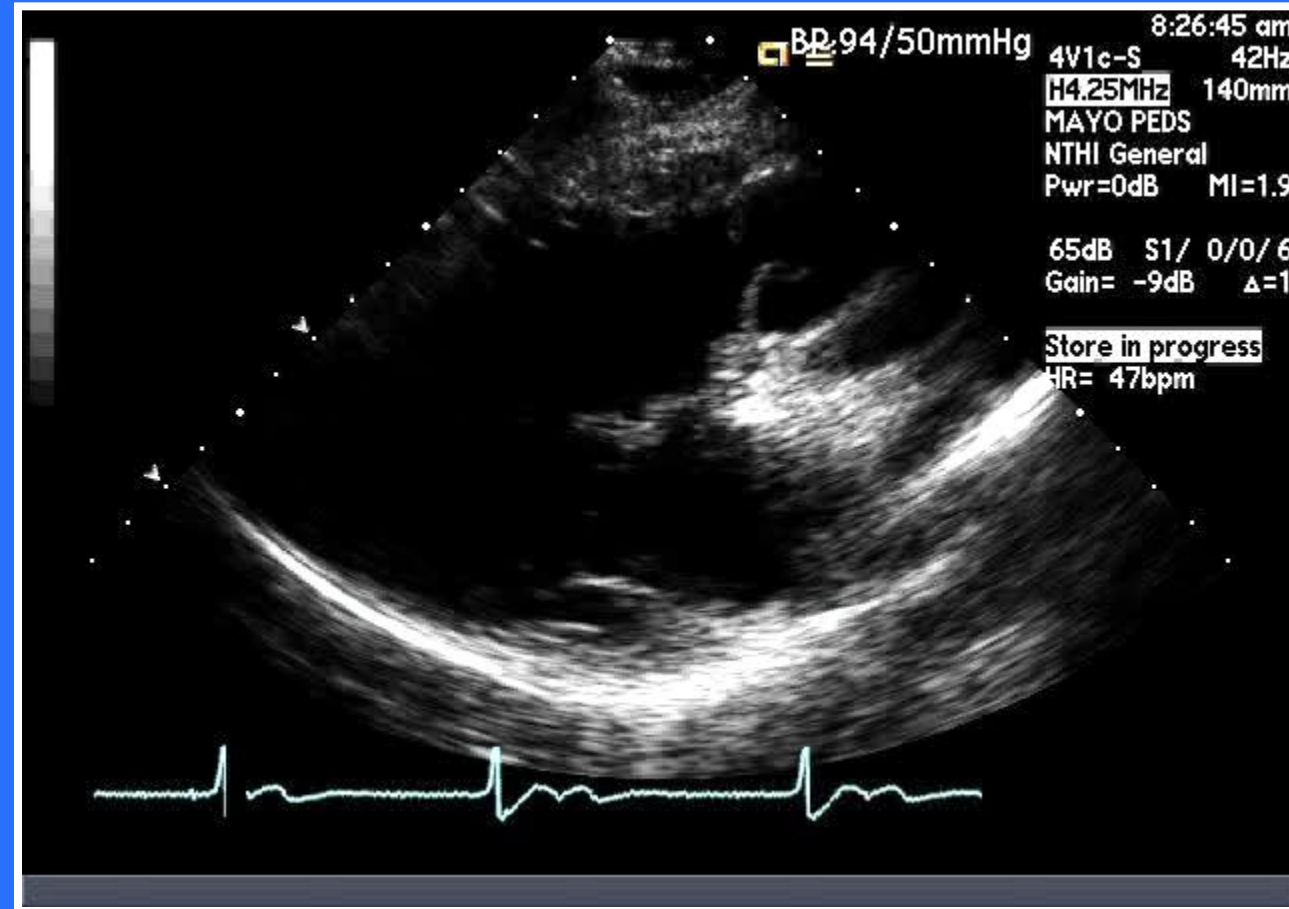
“Abnormal ECG on Life Insurance Exam”

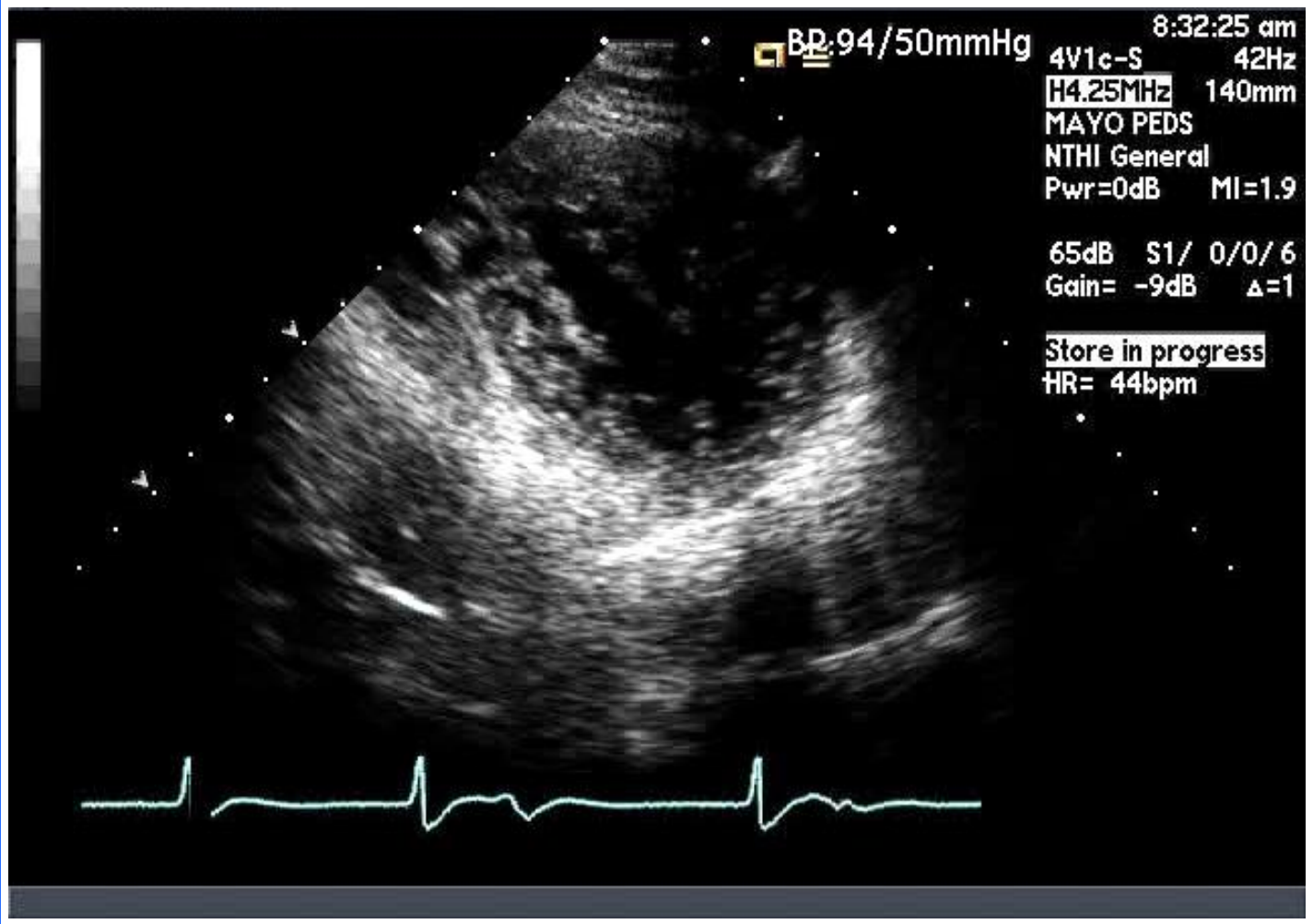
- **31 year old man**
- **Told as a child he had a “hole in his heart” – no intervention**

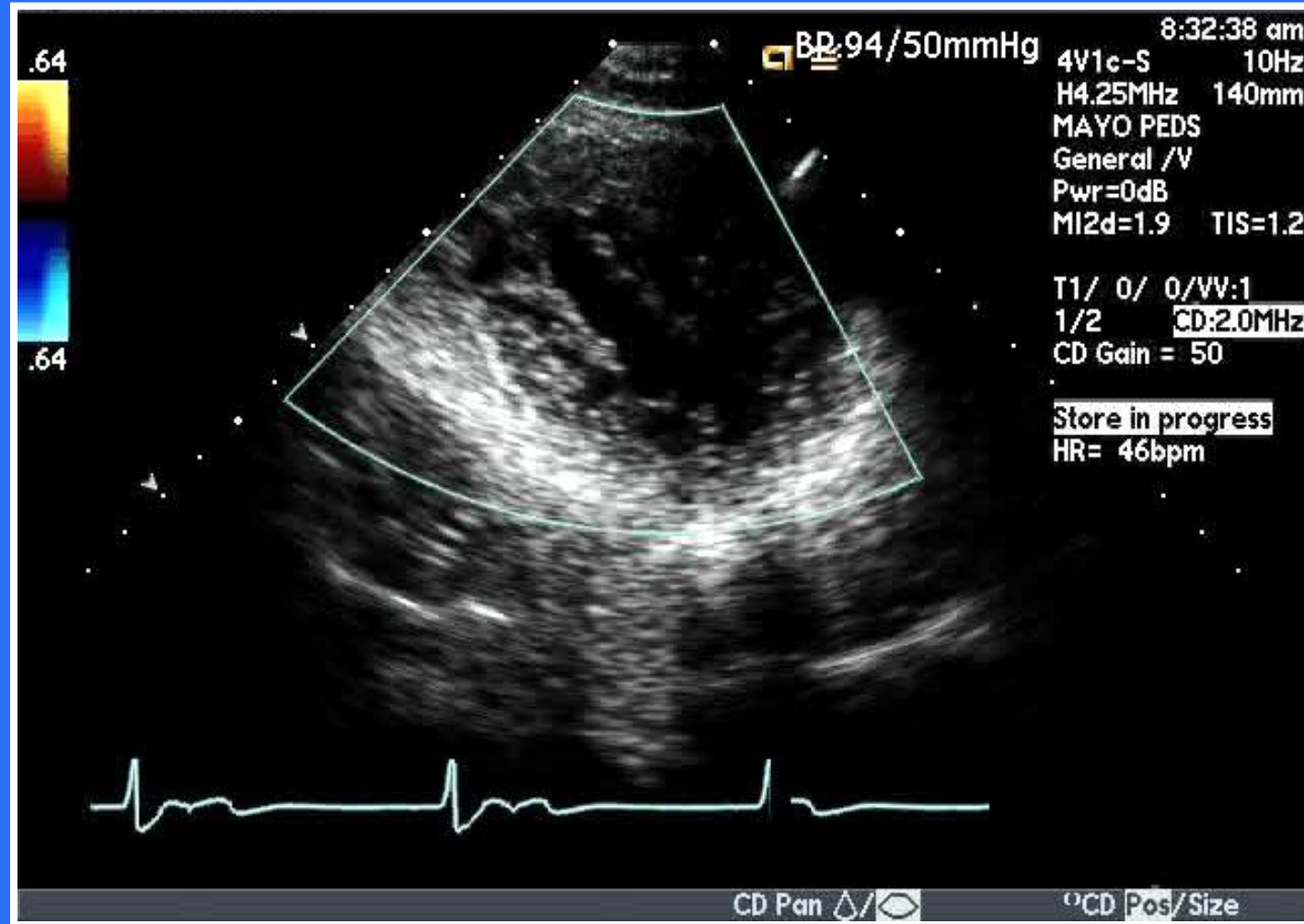
EKG

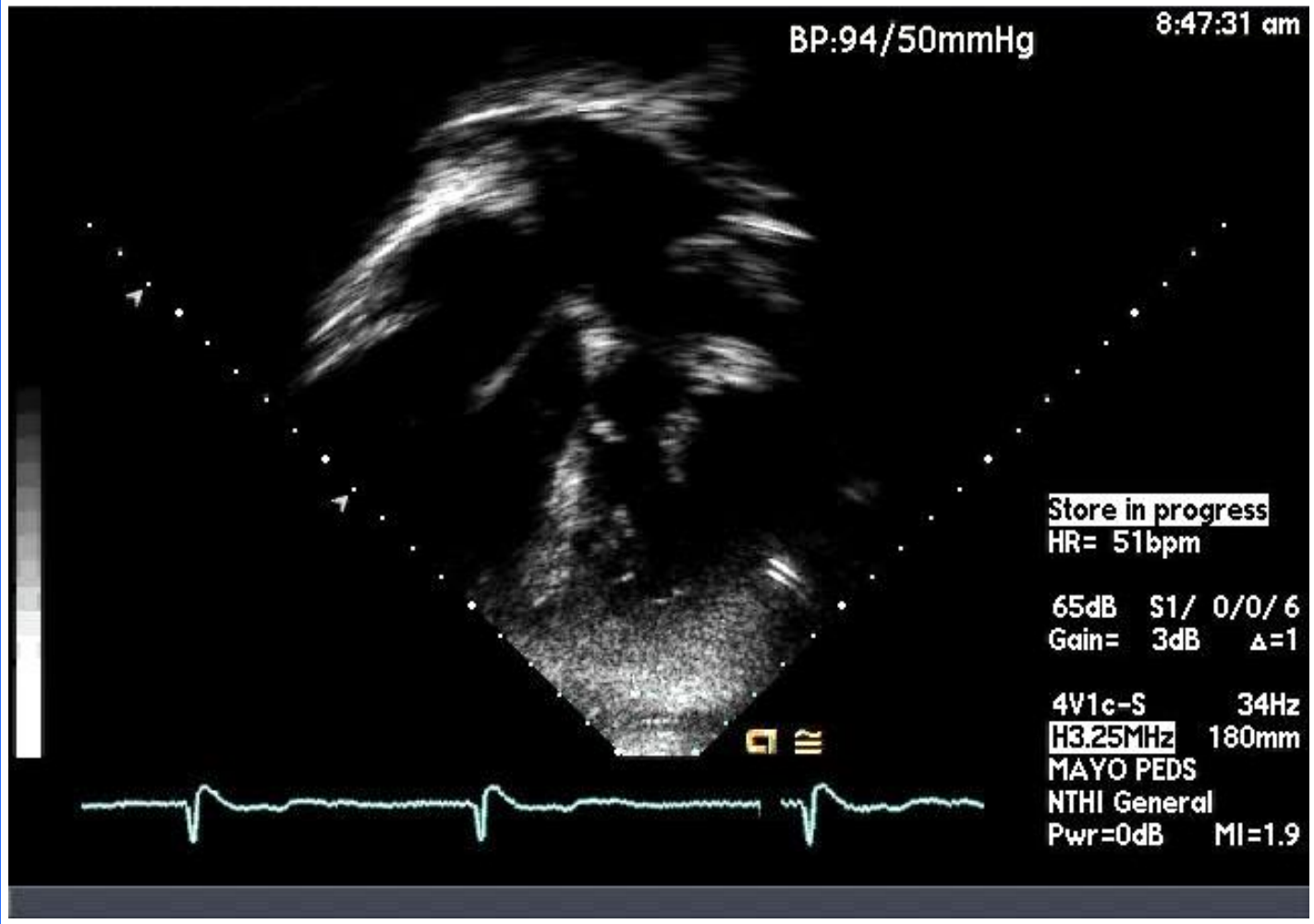


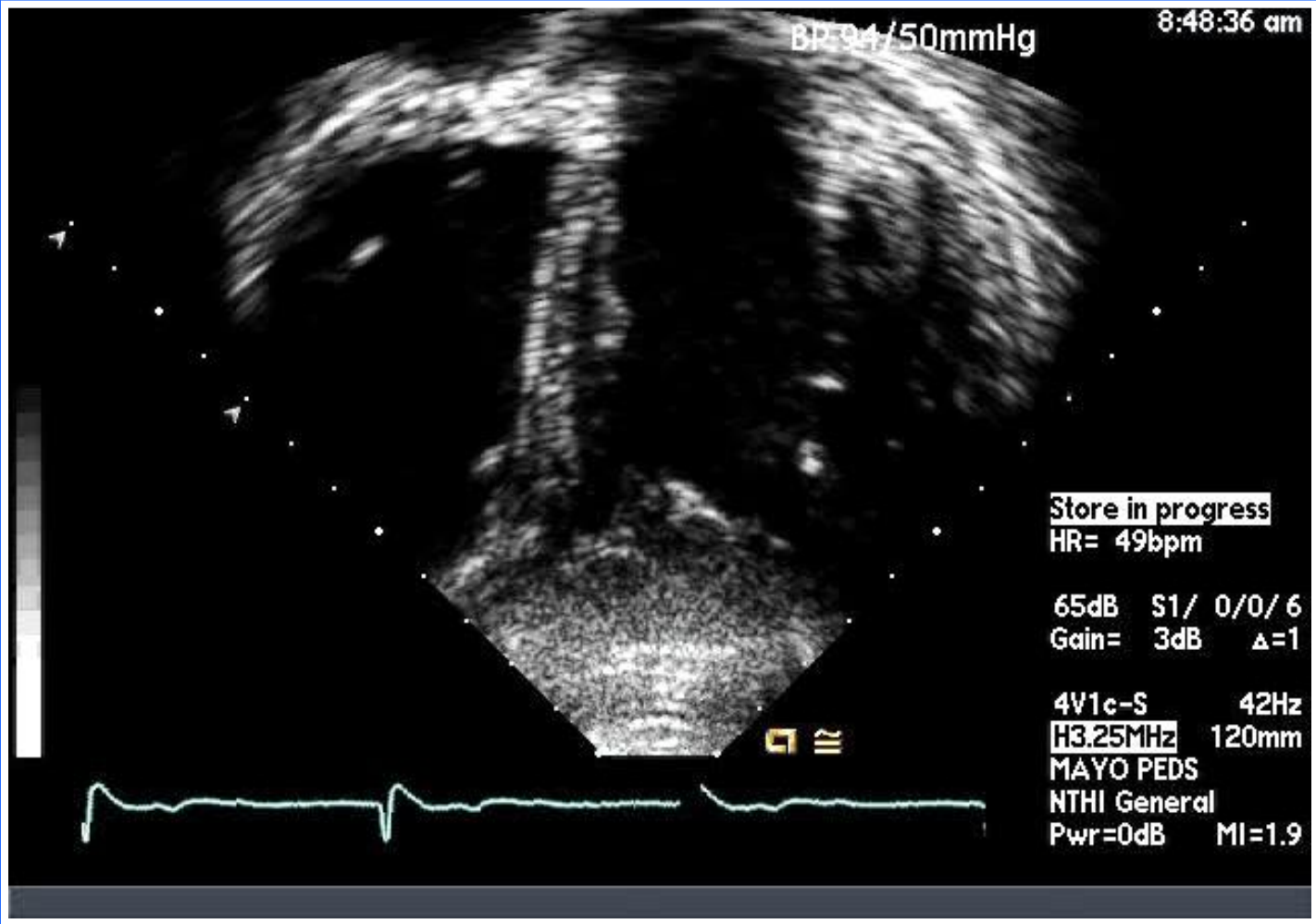
ECHO IMAGES

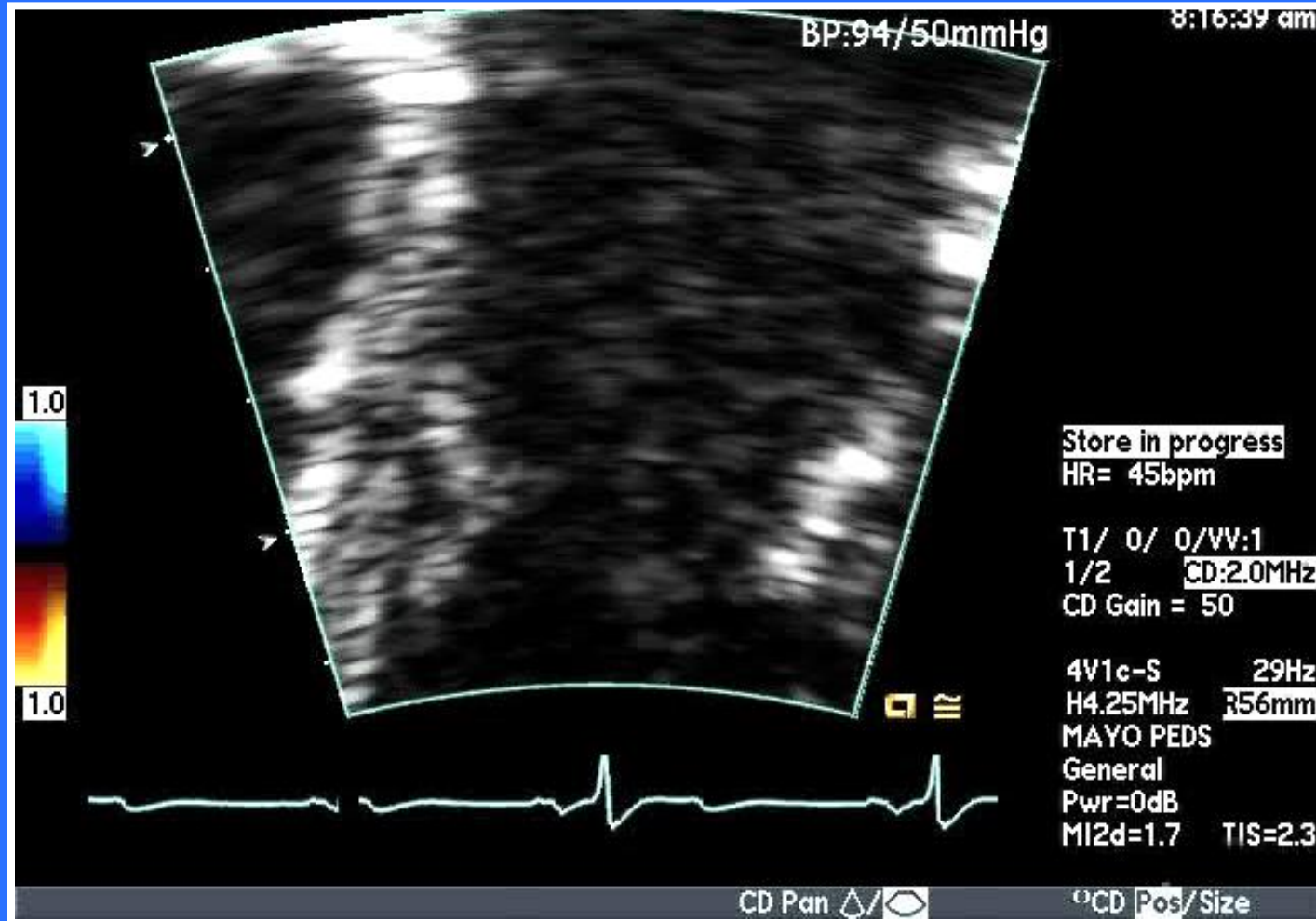


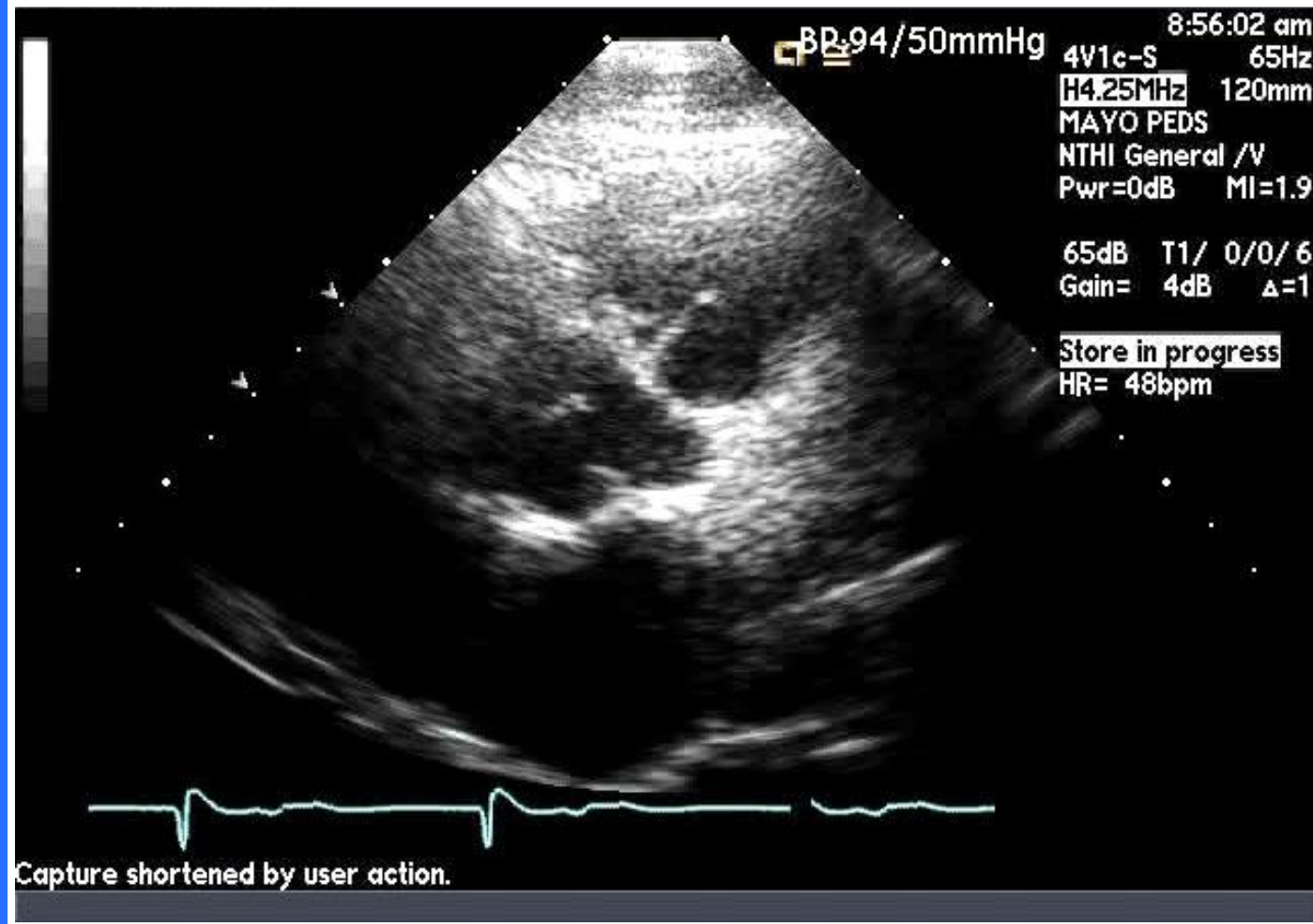












Diagnosis?

- A. Non-compaction cardiomyopathy**
- B. L-TGA**
- C. D-TGA**
- D. Ebstein anomaly**

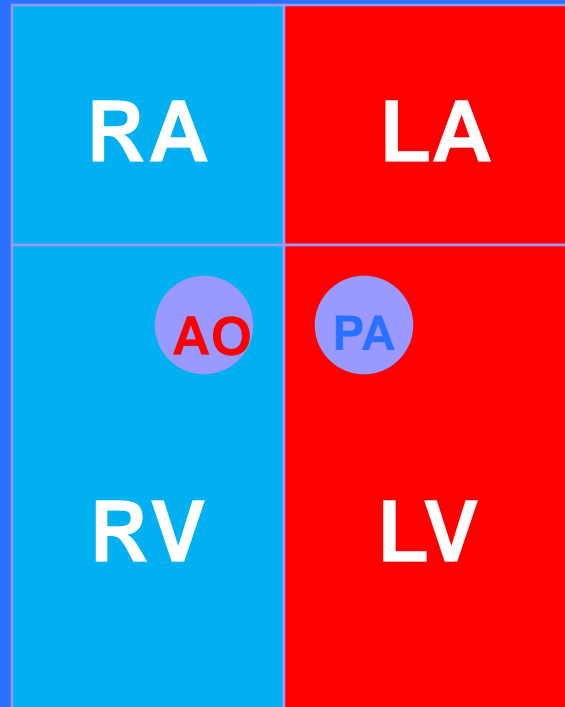
Diagnosis?

- A. Non-compaction cardiomyopathy
- B. L-TGA**
- C. D-TGA
- D. Ebstein anomaly

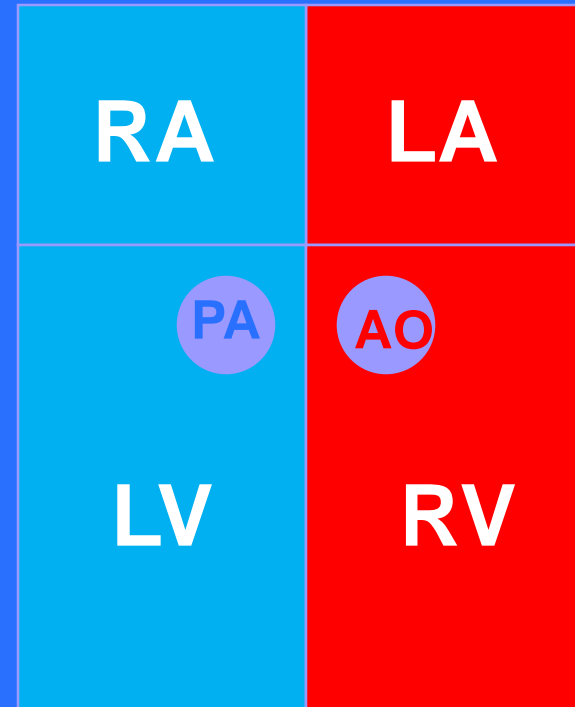
Congenitally Corrected Transposition



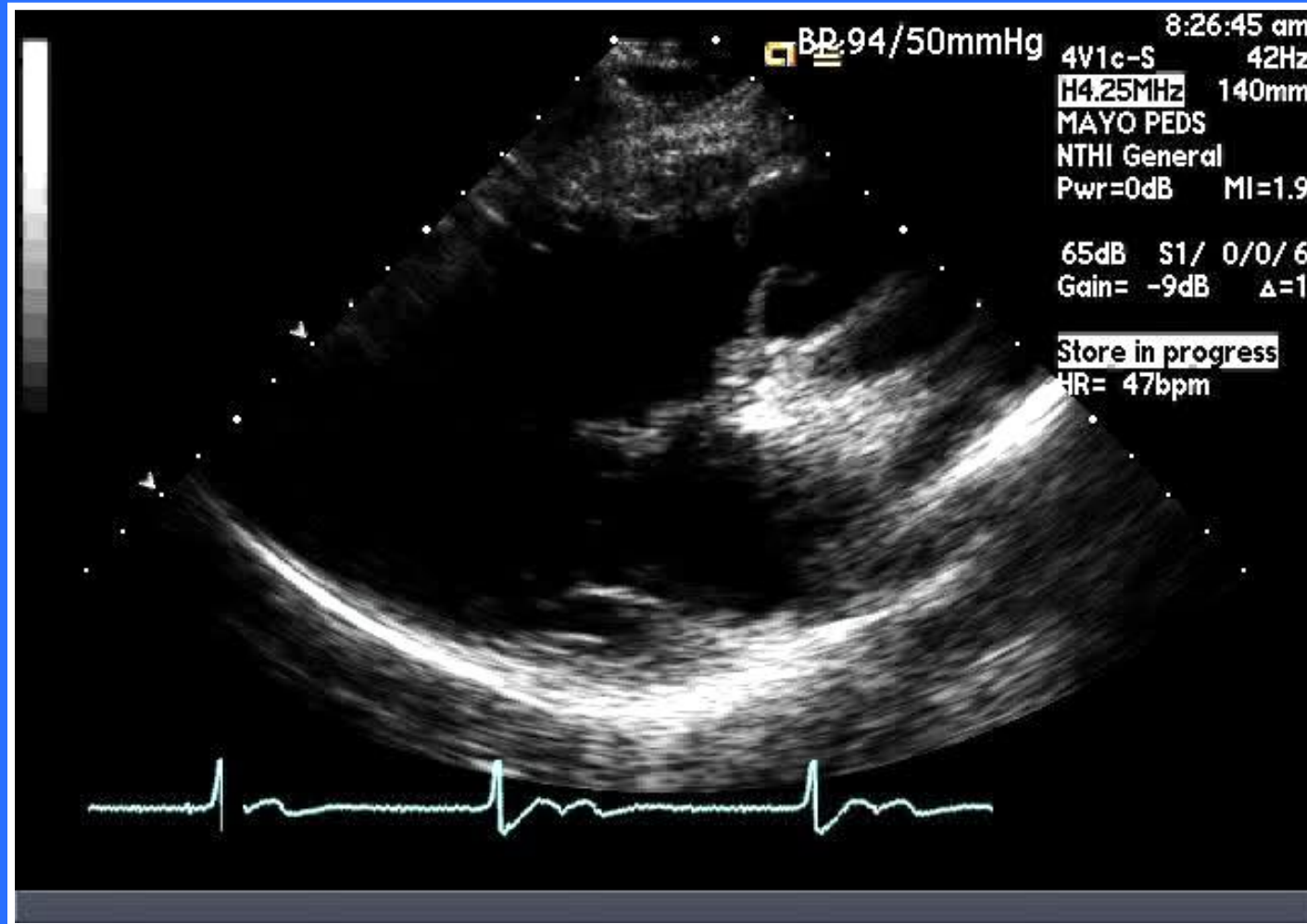
Complete Transposition (D-TGA)



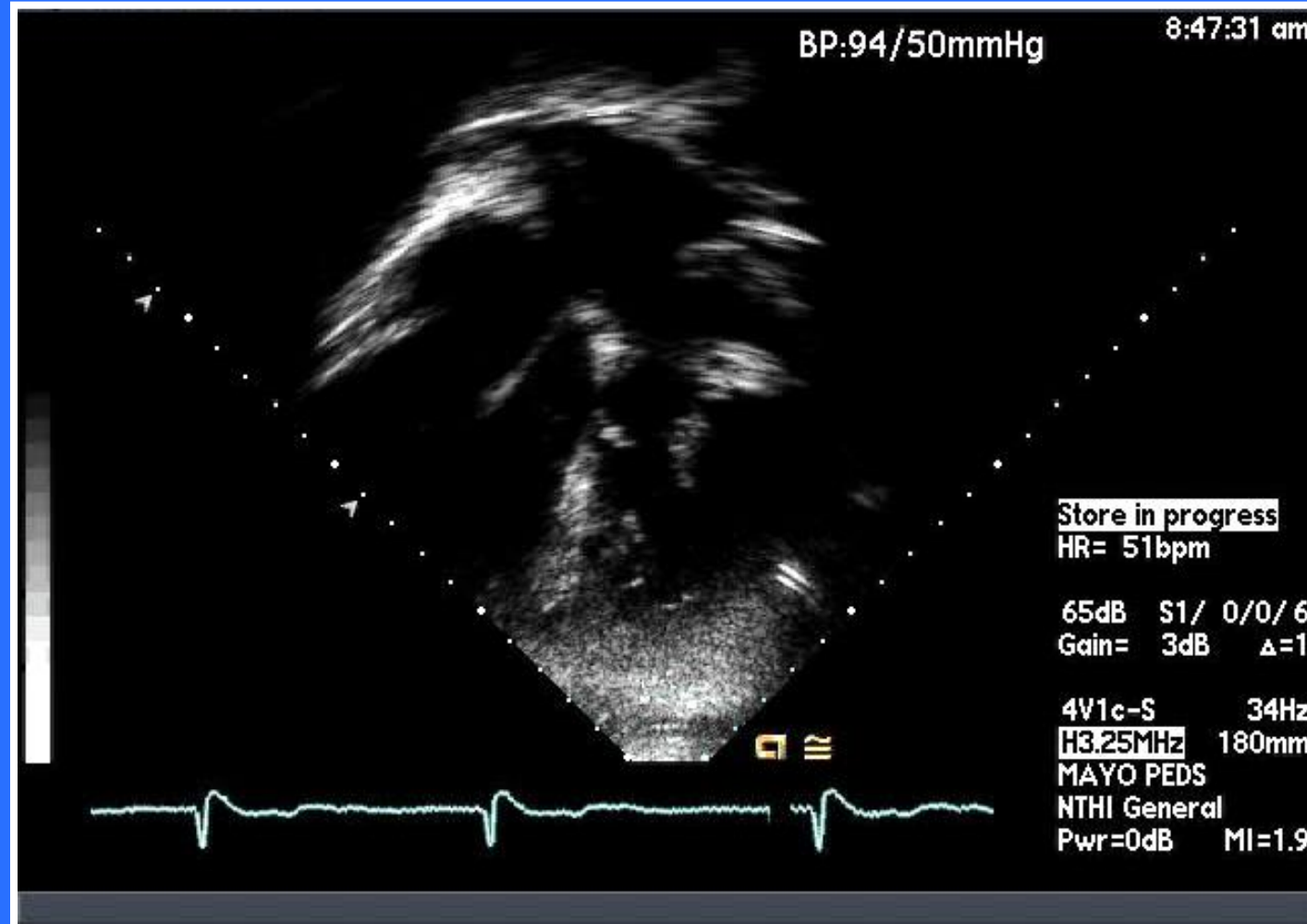
Congenitally Corrected Transposition (L-TGA)



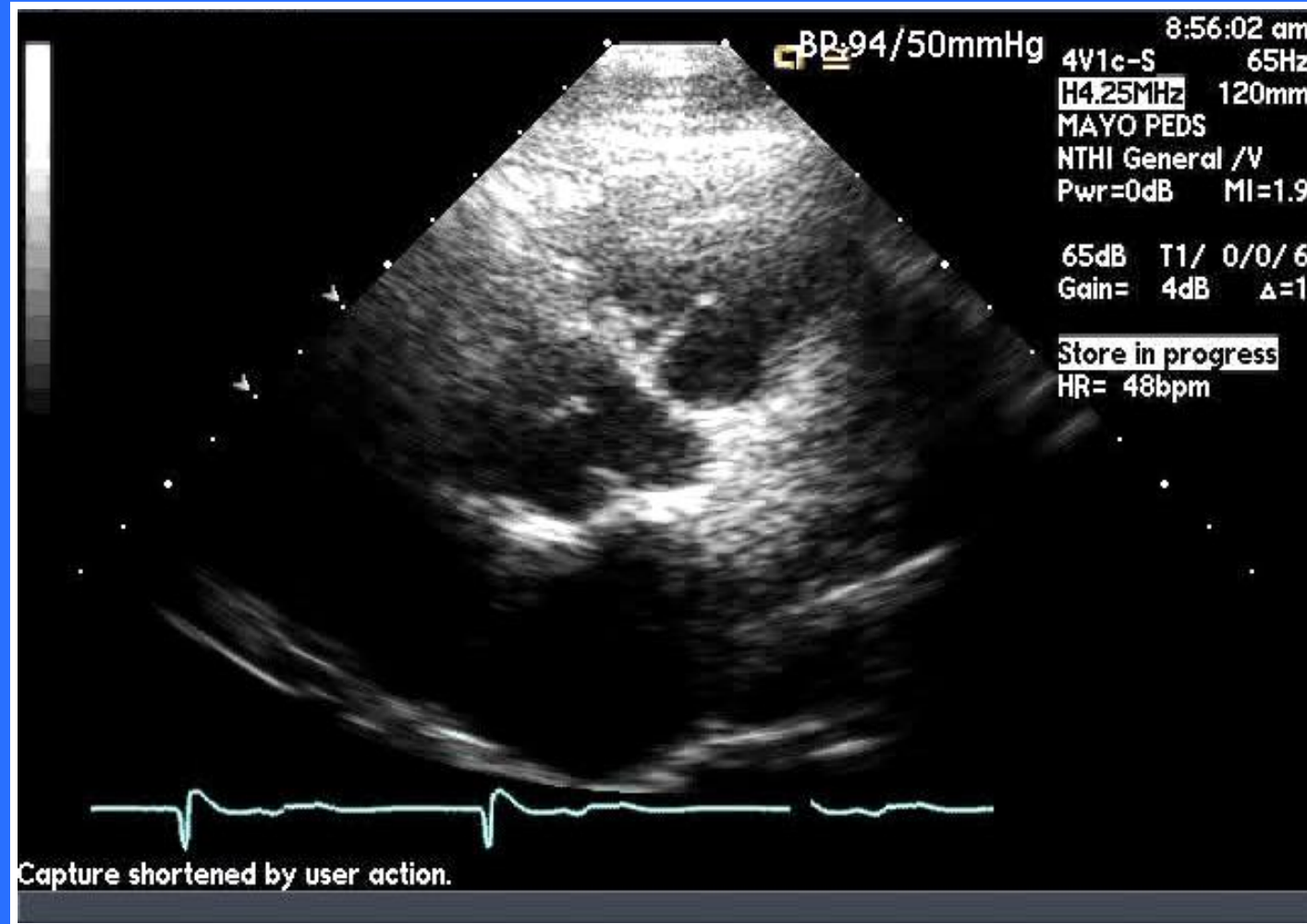
Conus present in the “LVOT”



Left A-V valve displaced apically



Side-by-side semi-lunar valves



Lesions Associated with ccTGA

- Ventricular Septal Defect (70%)
- Subpulmonary ventricular outflow tract obstruction (40%)
- Tricuspid valve dysplasia/Ebstein malformation (90%)
- Situs Inversus
- Dextrocardia

“Second Opinion”

38 Year Old Woman

- Present for second opinion re: treatment of pulmonary hypertension
- Significantly limited
- Marked cyanosis

Past Medical History

- Evaluated at 3 months of age for pneumonia
- Diagnosed with VSD, PDA, coarctation
- PA banding, PDA ligation and coarctation repair performed

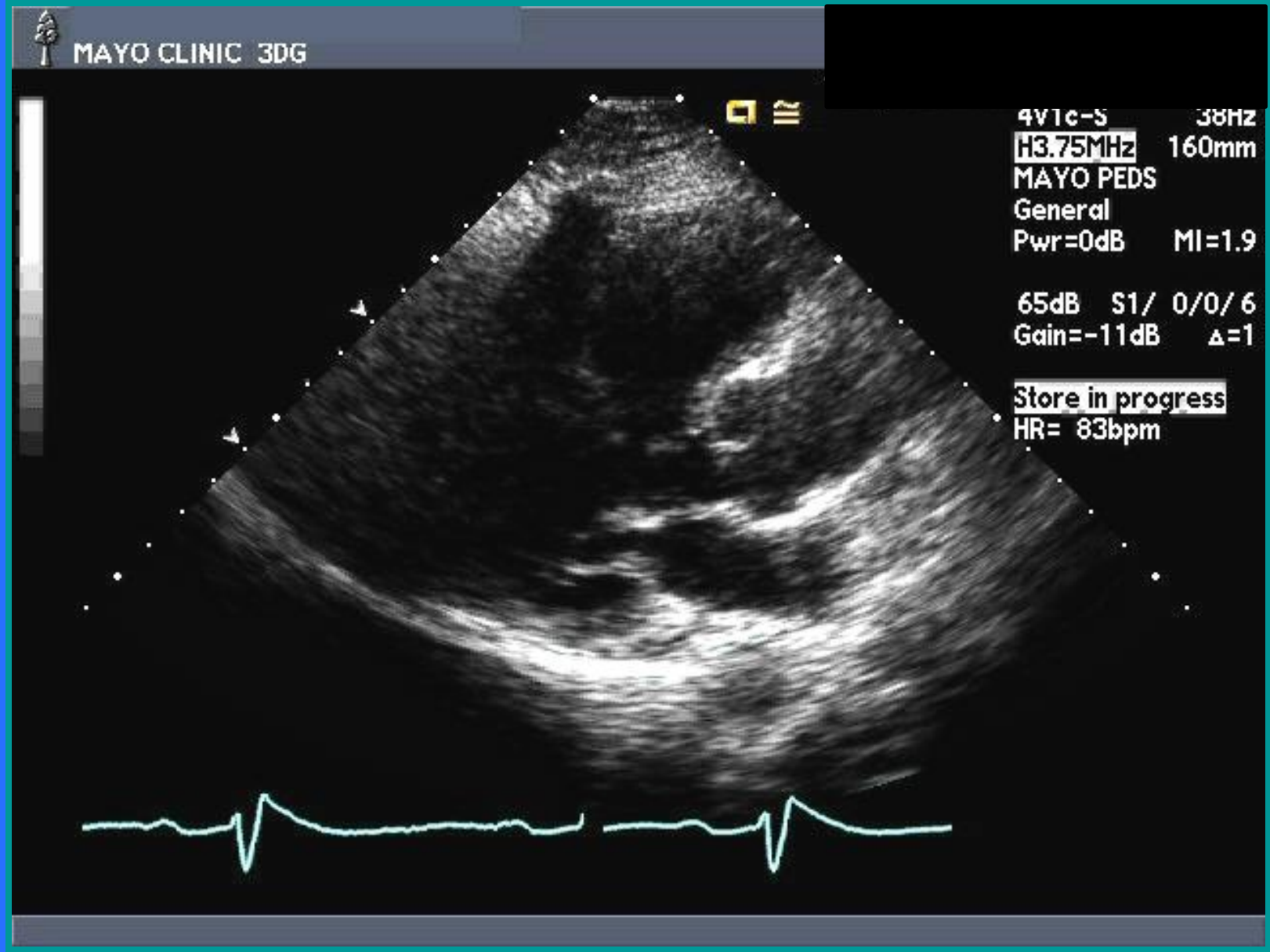
Past Medical History

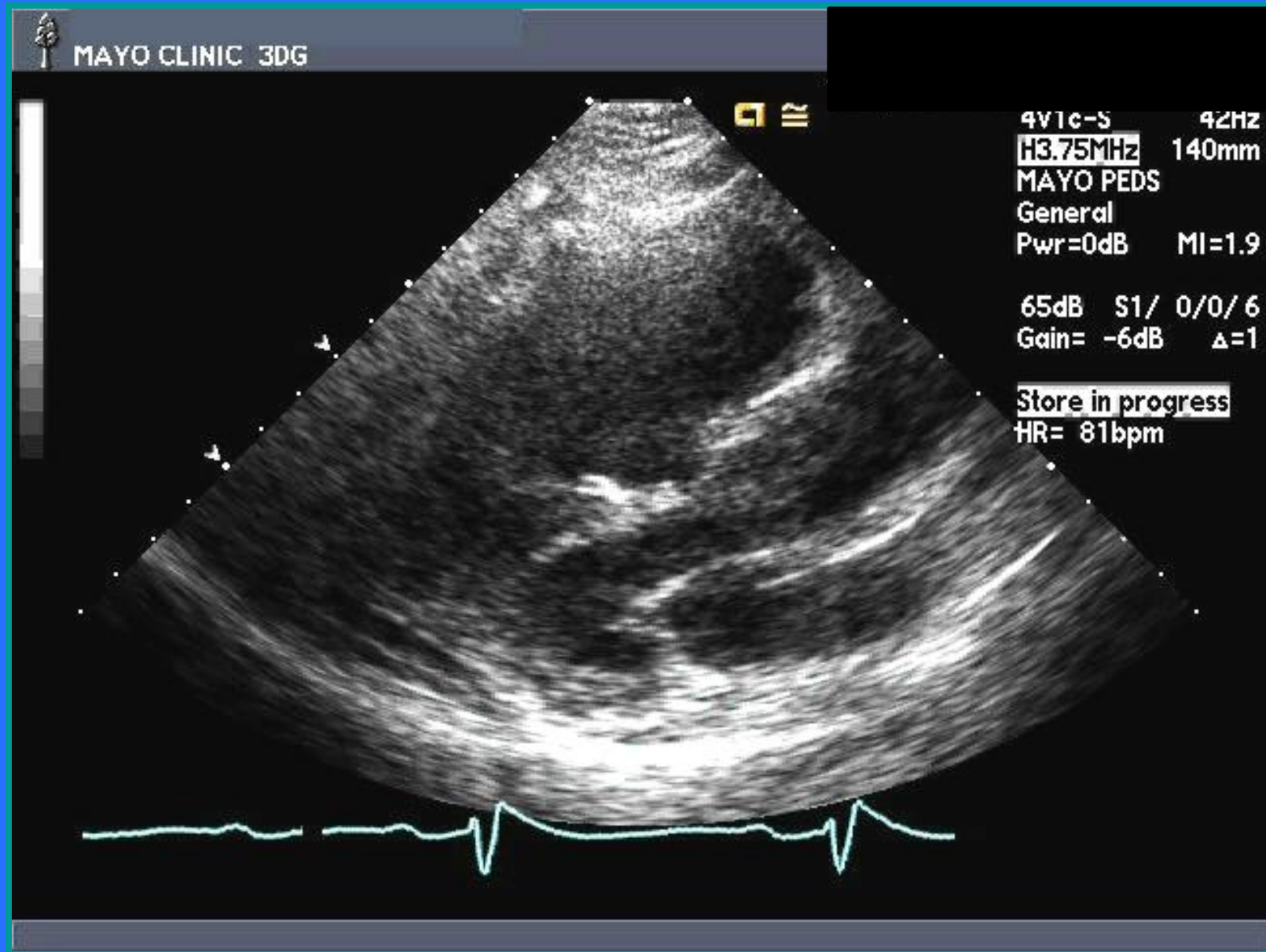
- 6 years: diagnosed with Eisenmenger syndrome
- Treated with frequent phlebotomy
- Placed on Coumadin in adulthood
- Placed on the heart/lung transplant list 5 years (elsewhere)
- No birth control being used

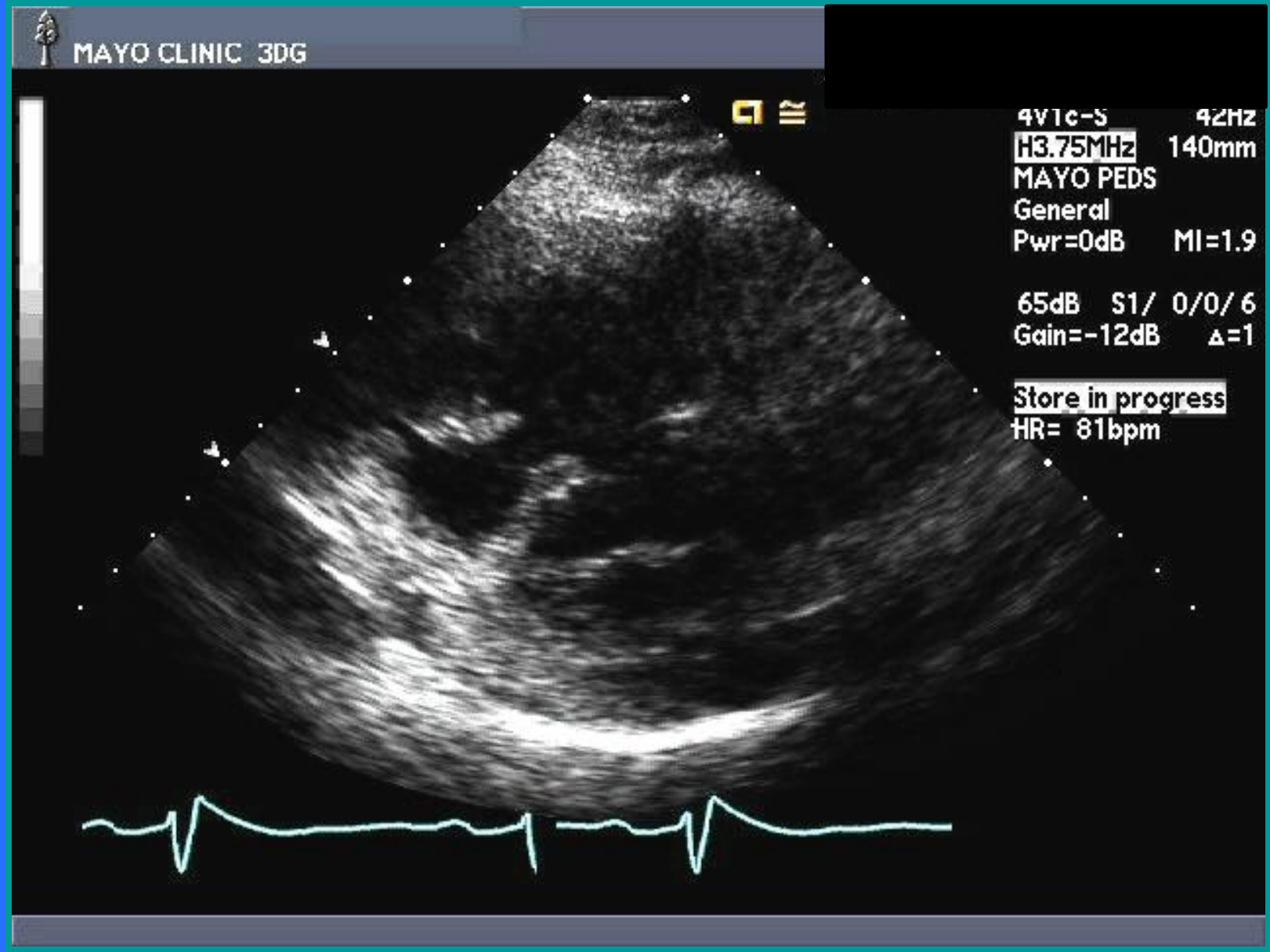
Current Exam

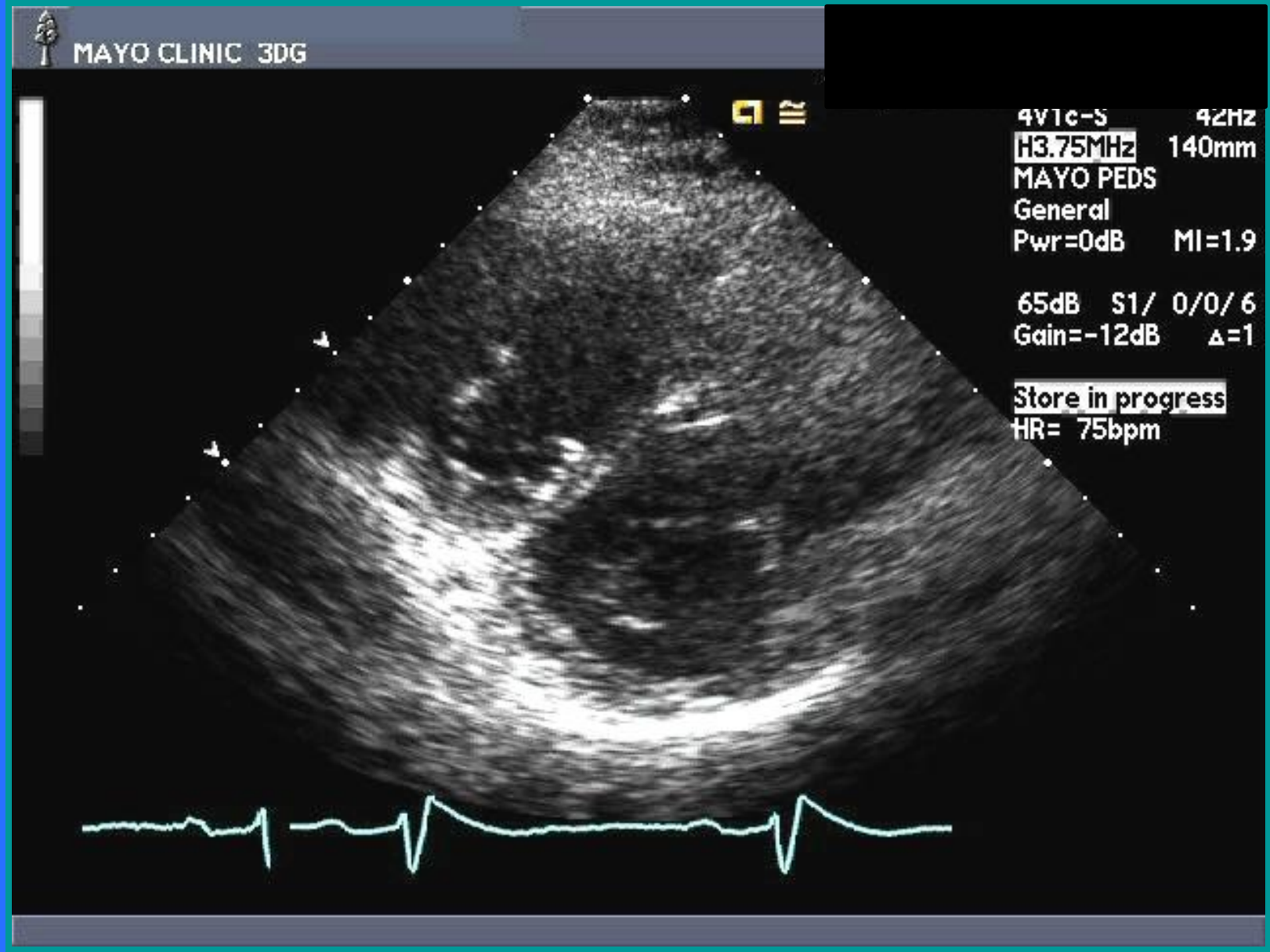
- Significant cyanosis
- Conjunctival injection
- 2+ RV impulse, normal LV impulse
- 3/6 systolic crescendo-decrescendo murmur left upper sternal border
- No diastolic murmur





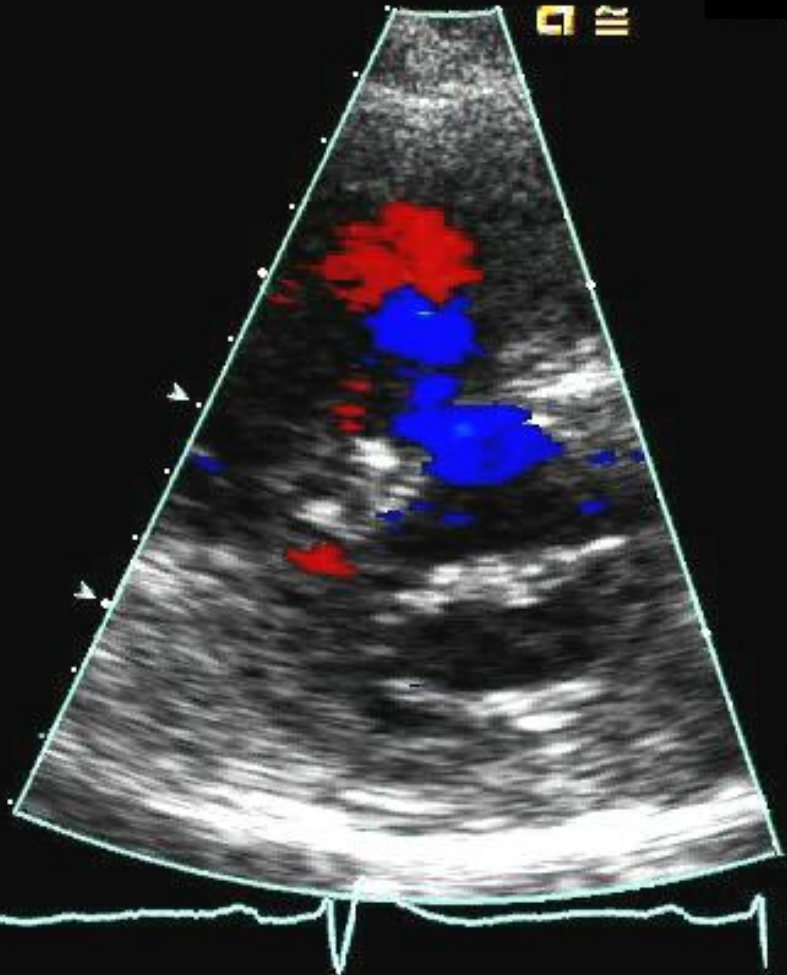






MAYO CLINIC 3DG

.80
Color scale bar
.80



4VIC-5 19MHz
H3.75MHz 310mm
MAYO PEDS
General /V
Pwr=0dB
MI2d=1.8 TIS=2.2

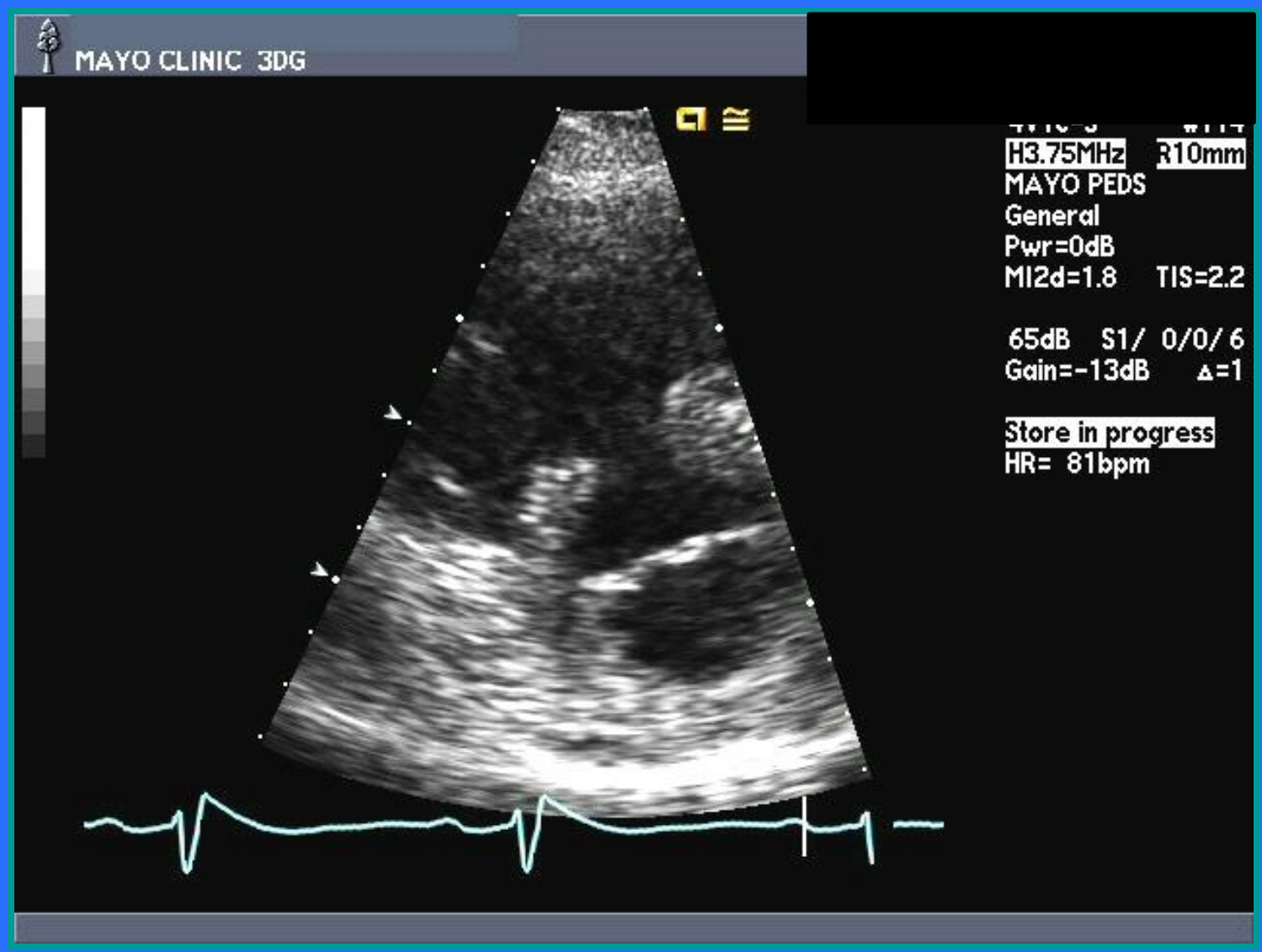
T1/ 0/ 0/VV:1
1/2 CD:2.0MHz
CD Gain = 48

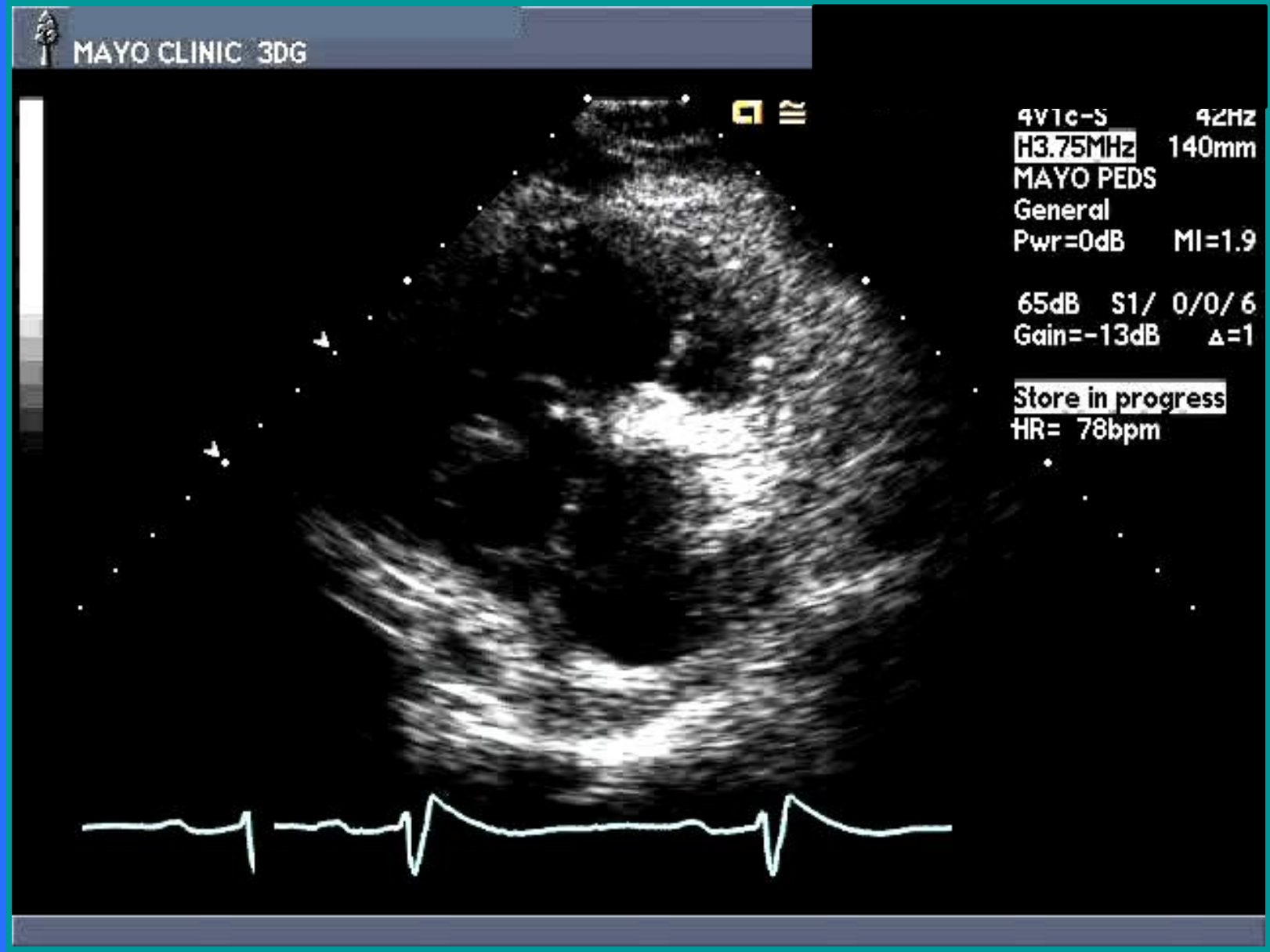
Store in progress
HR= 80bpm

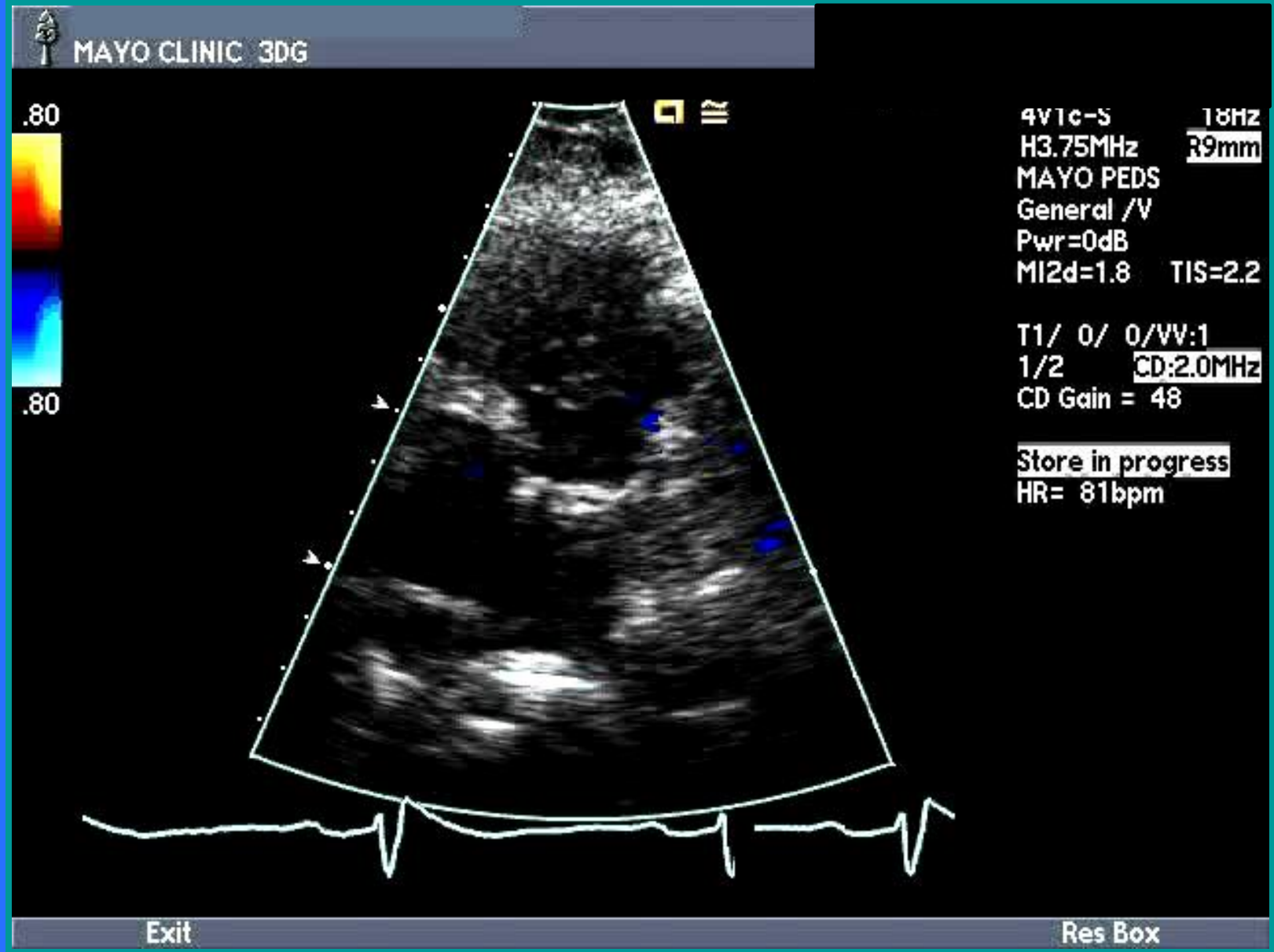
Exit

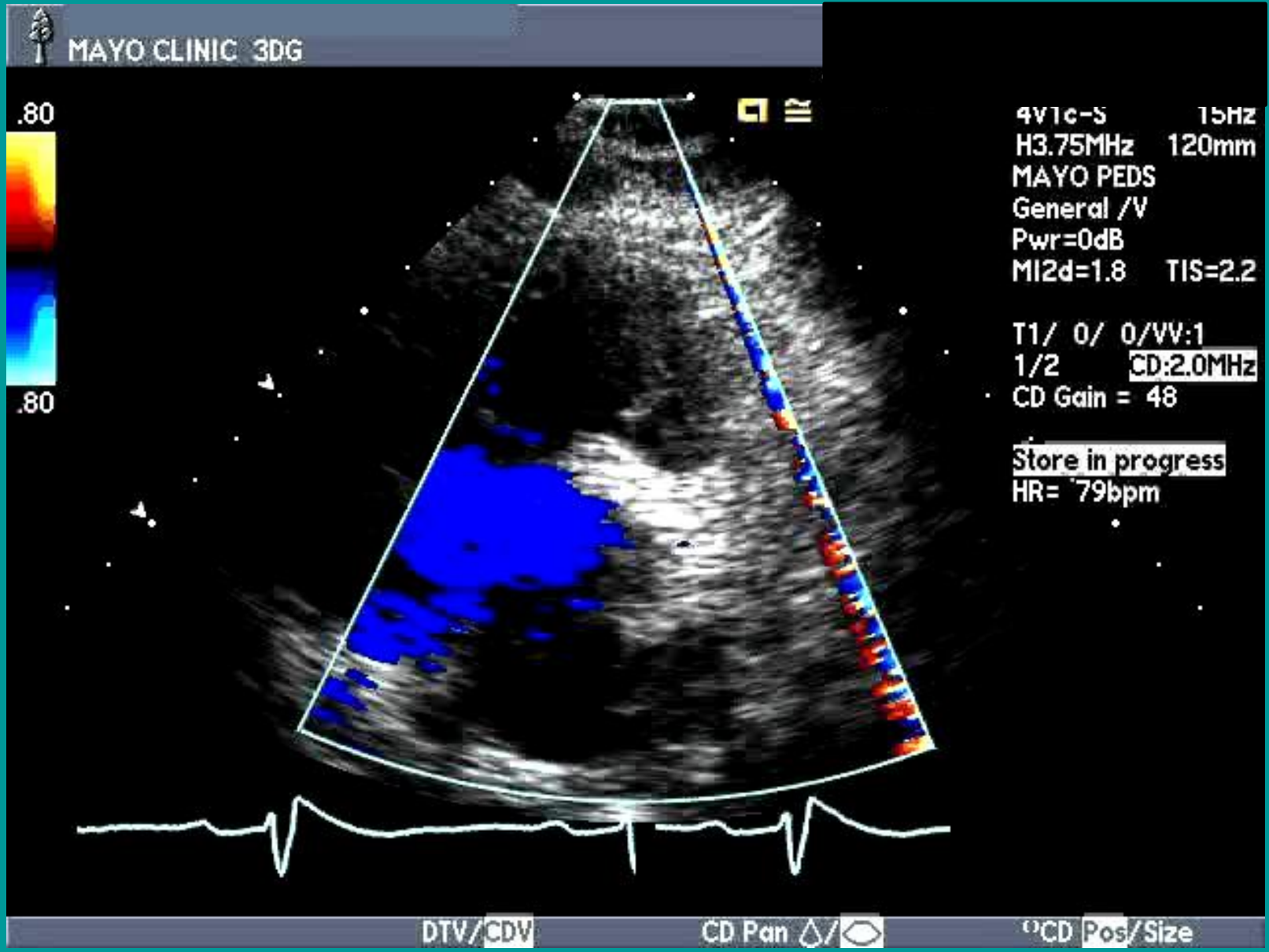
Res Box

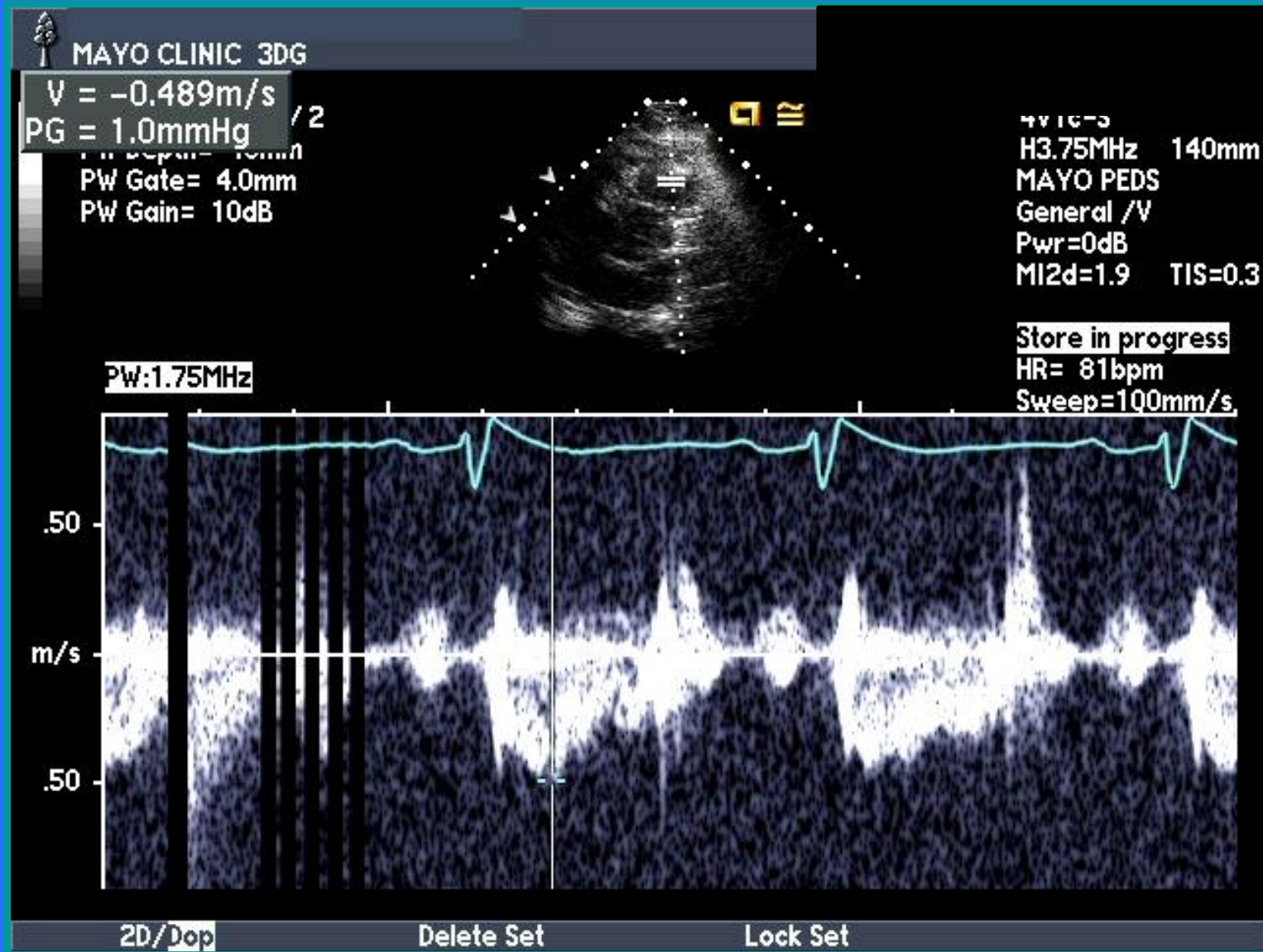




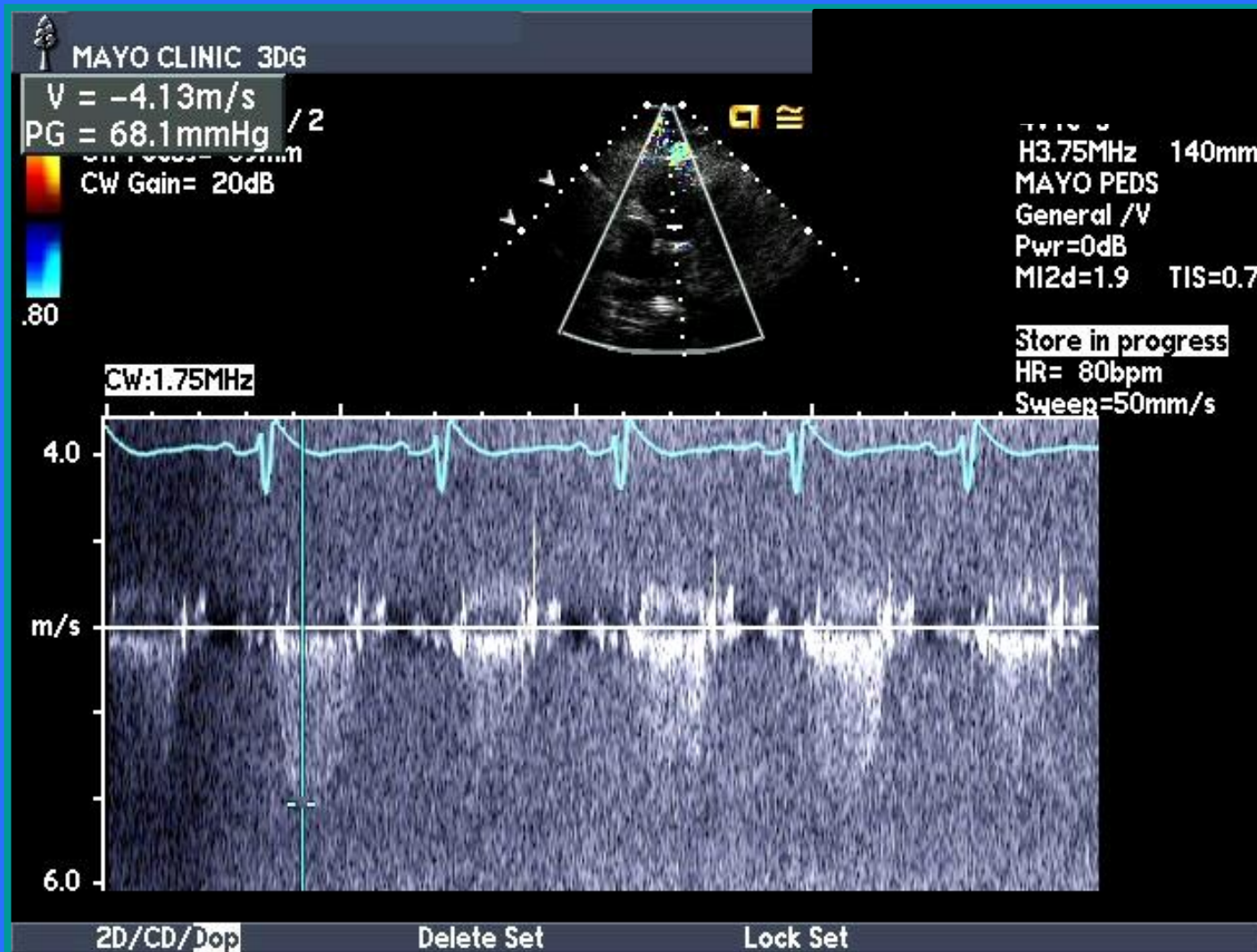




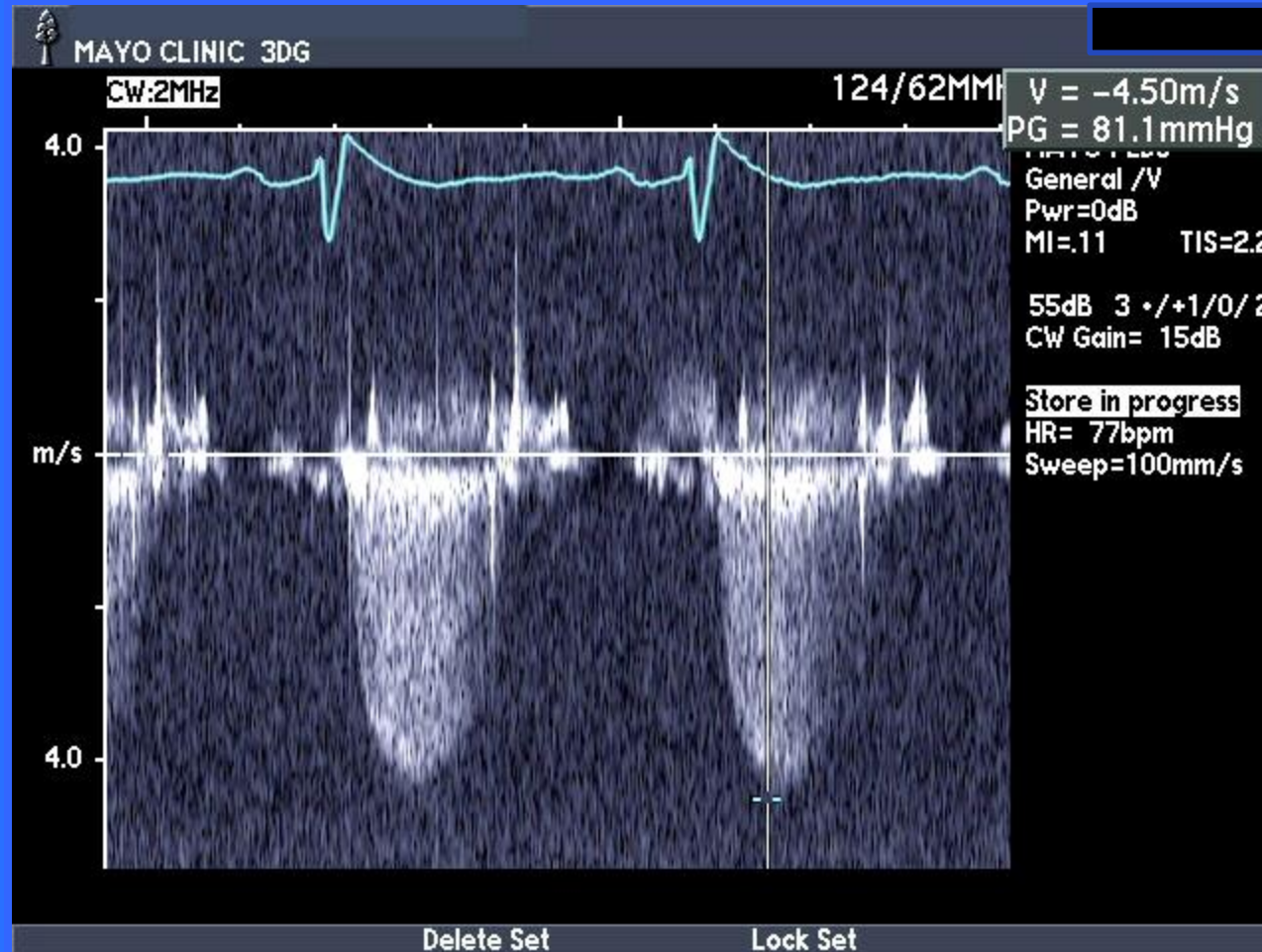




PWD velocity = 0.5 m/s



CWD velocity = 4.1 m/s



Non-imaging Doppler velocity = 4.5 m/s

Does This Patient Have Eisenmenger's Syndrome?

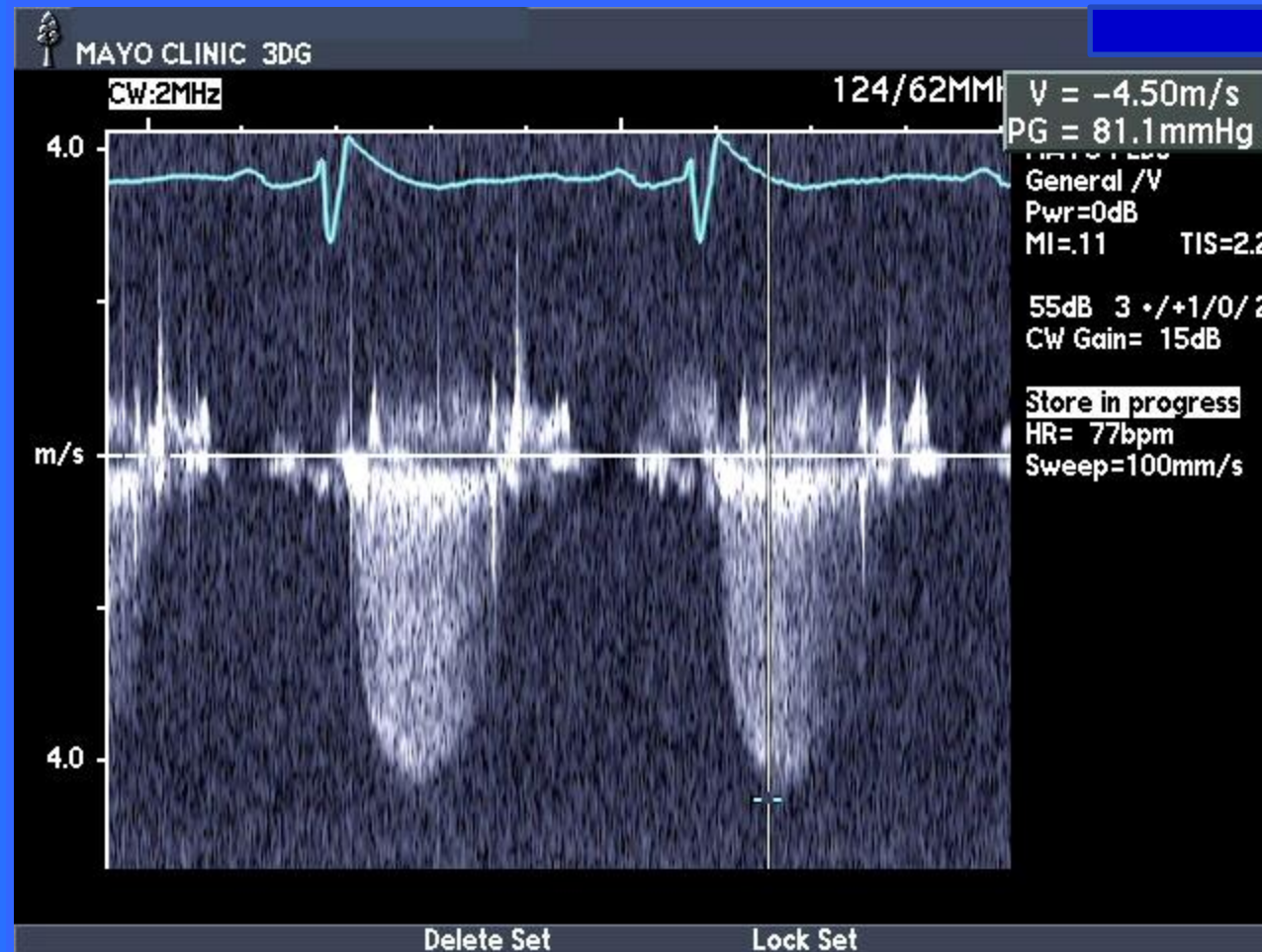
A. Yes

B. No

Does This Patient Have Eisenmenger's Syndrome?

A. Yes

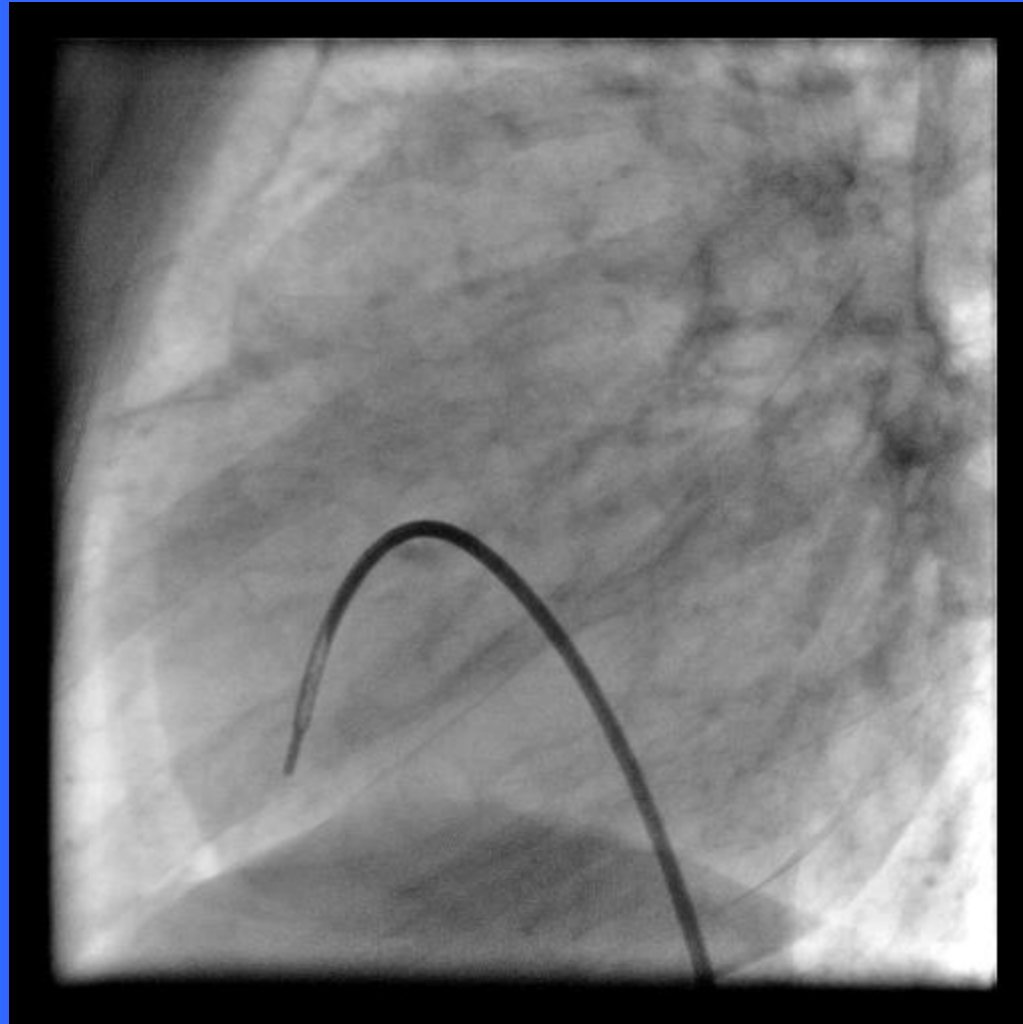
B. No



Non-imaging Doppler velocity = 4.5 m/s

Cath

- Tight PA band in appropriate location without distortion of the pulmonary valve
- Distal PA pressure 35/11
- Band gradient: 80 mmHg
- Pulmonary blood flow < 1 L/min/m²
- No residual coarctation
- No PDA



Outcome

- Successful PA debanding and VSD closure
- Transient post-op reperfusion lung injury
- Returned for 6 month follow-up: room air sat 95%. Normal 6 minute walk. RVSP: 51 mmHg
- Discontinued disability and began a new job

Teaching Points

- A VSD with a bidirectional shunt \neq Eisenmenger syndrome – look for obstruction to RV outflow causing RV hypertension
- Patients with Eisenmenger VSD do not have loud systolic ejection murmurs
- Review cath reports carefully with your interventionalist – communication between the care team is essential