

Cases in Adult Congenital Heart Disease

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➤ No Disclosures



“I Have Palpitations”

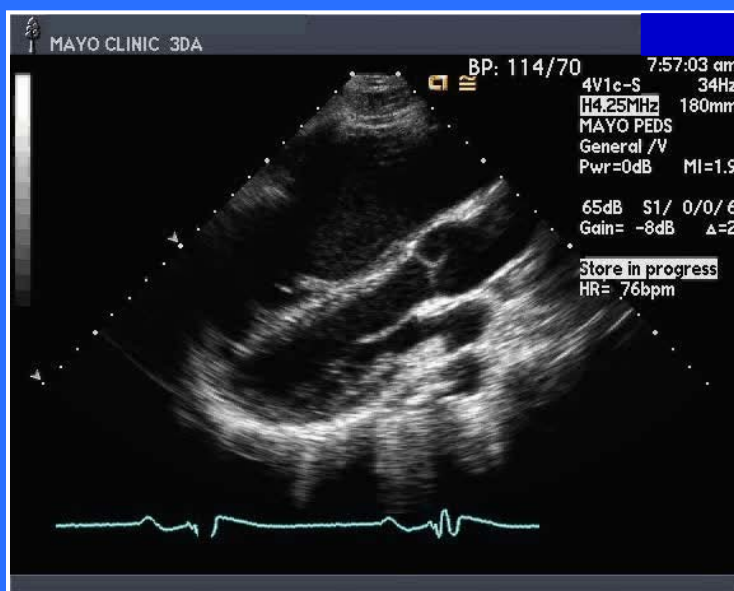
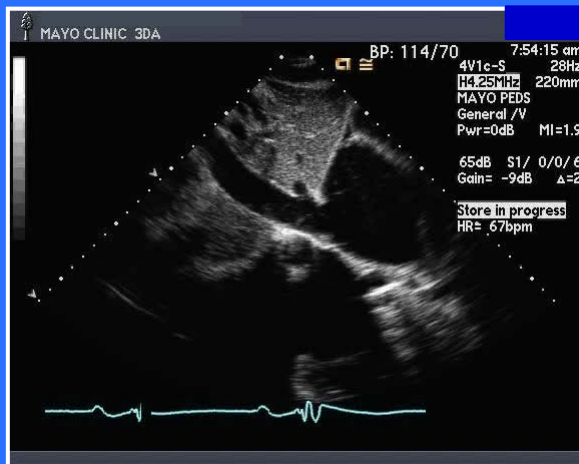


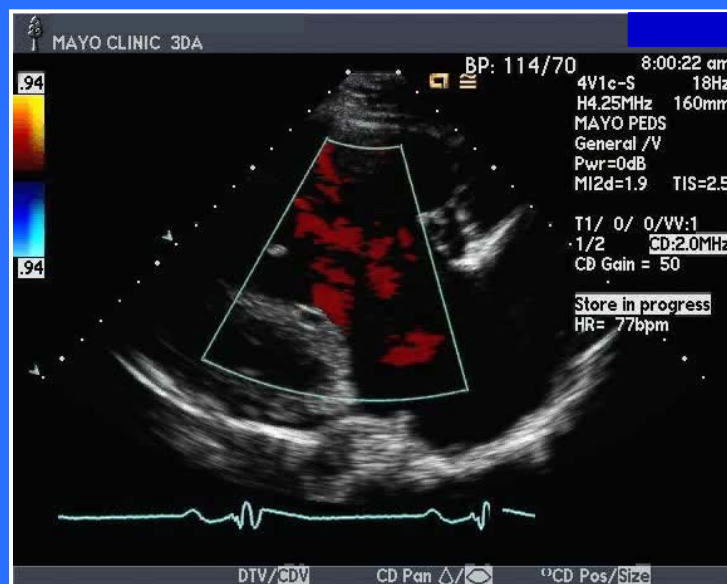
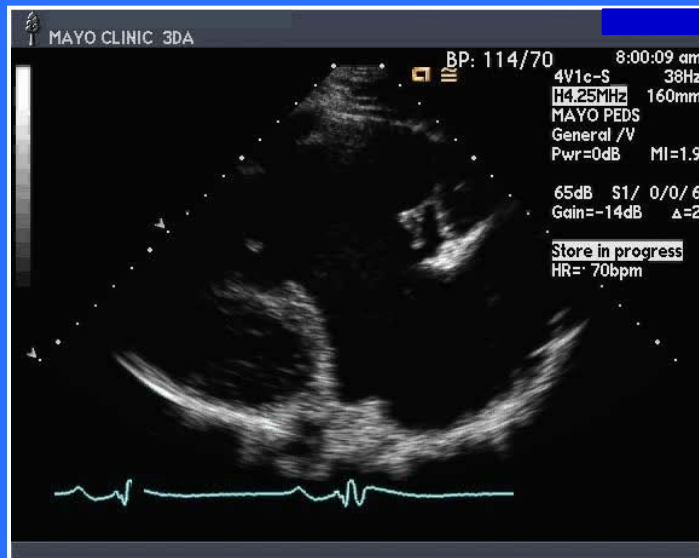
18 Year old Man

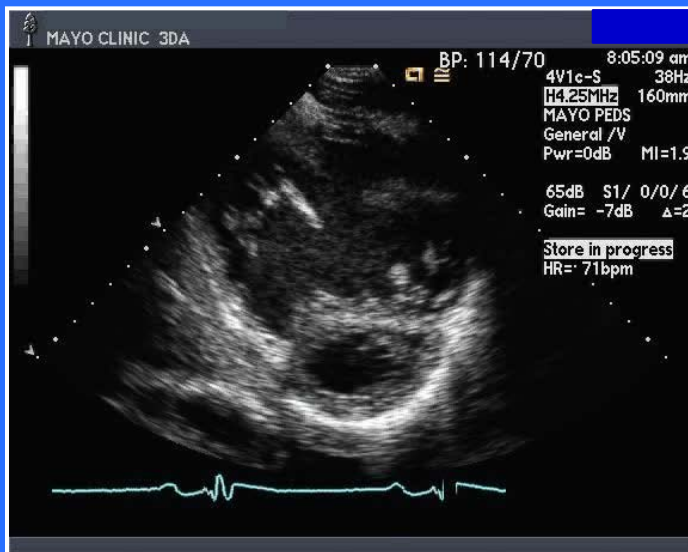
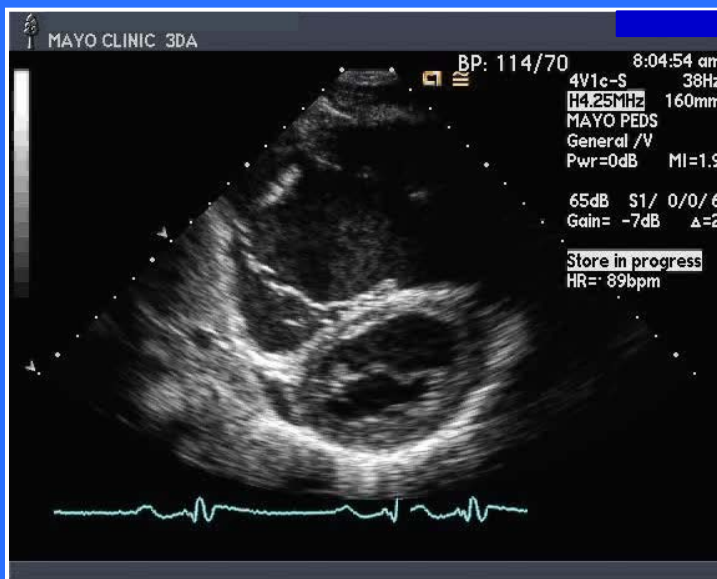
- Palpitations
- “abnormal” ecg and cxr

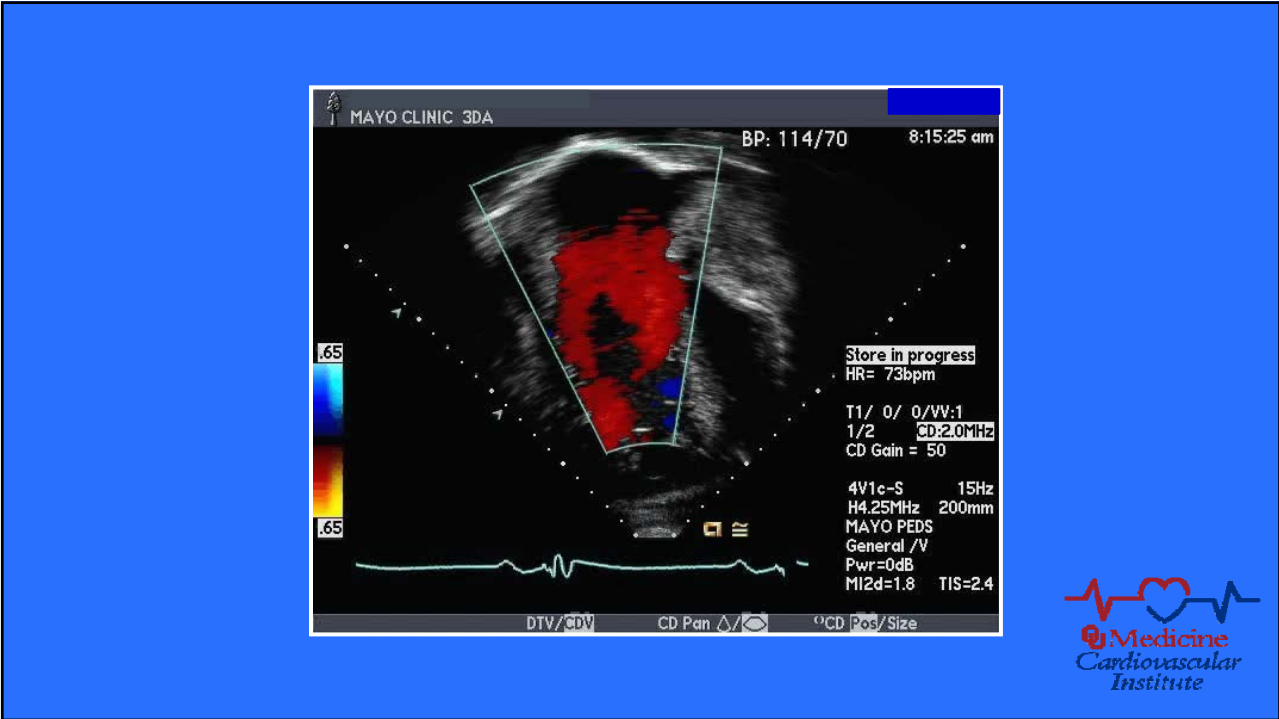
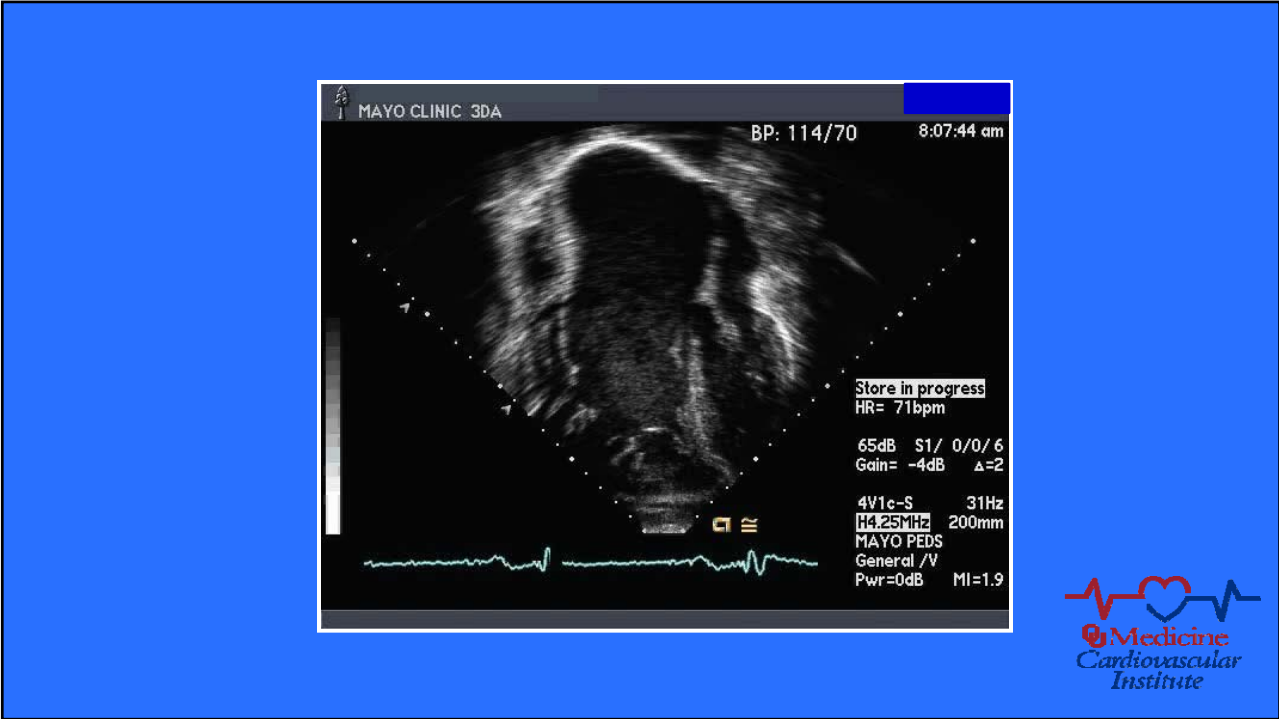


ECHO



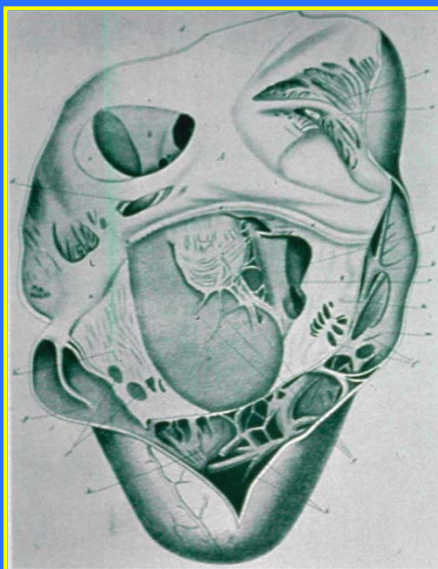




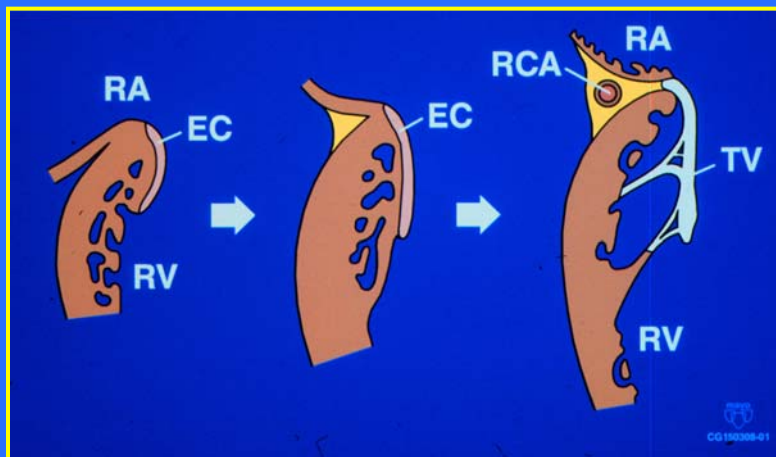




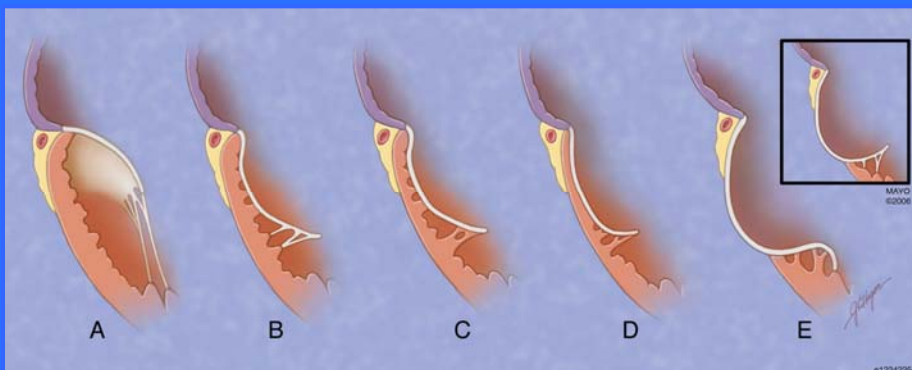
Ebstein Anomaly



Normal Delamination of the TV from the RV Myocardium



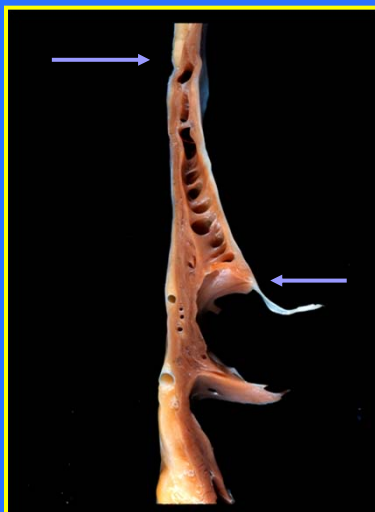
Failure of Delamination From the Myocardium



Spectrum with Infinite Variability



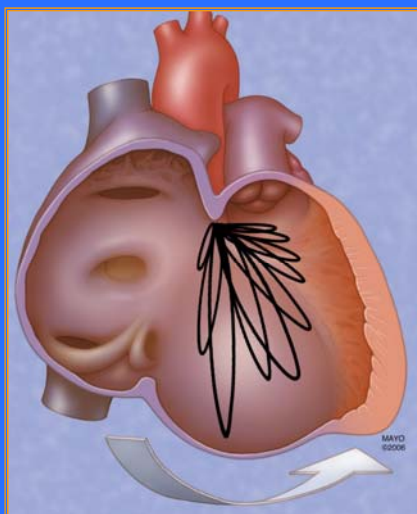
Failed Delamination results in ...



- adherence of leaflets to underlying RV myocardium
- displacement of the anular 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

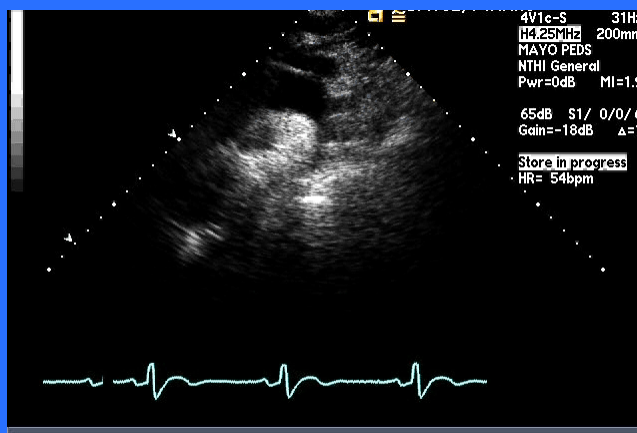


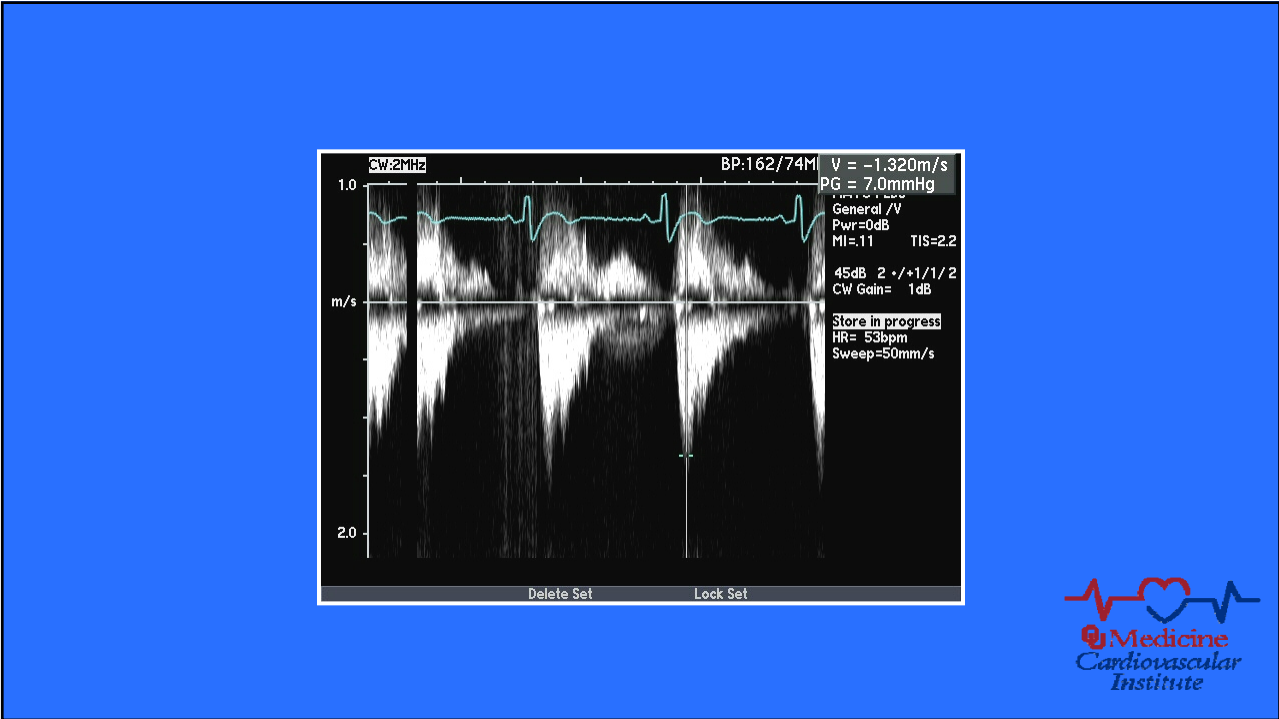
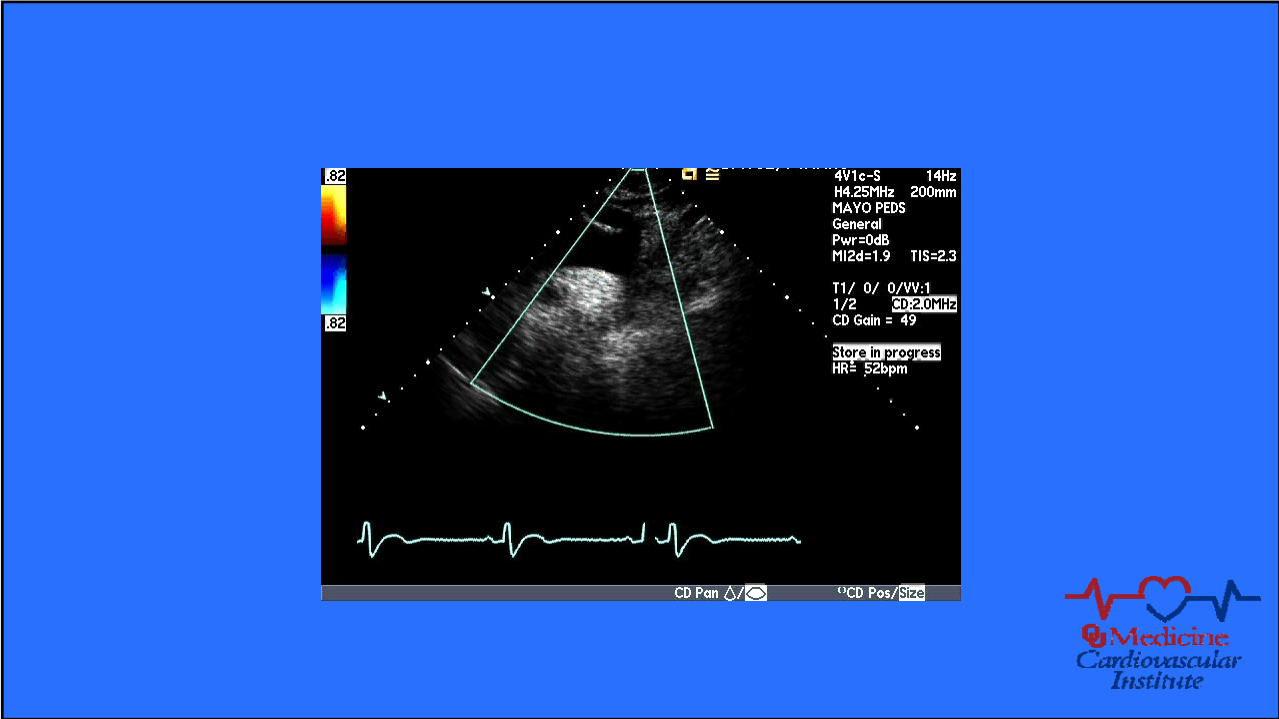
“I Have a Headache”



36 Year Old Man

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





Is This Coarctation?



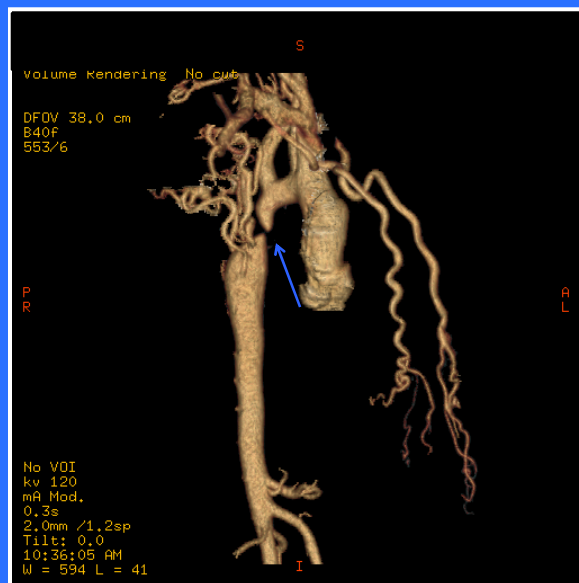
Is This Coarctation?

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

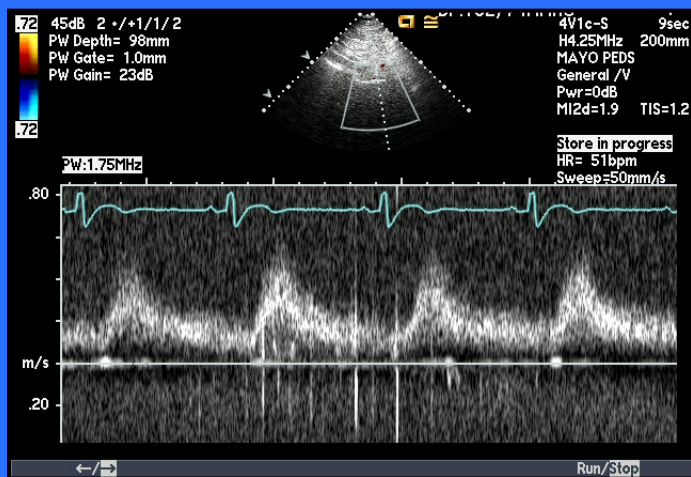


Is This Coarctation?

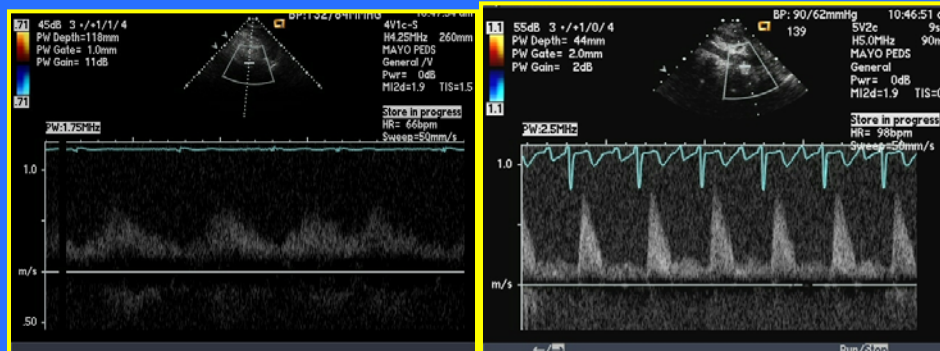
- A. Yes
- B. No
- C. Not Sure



Abdominal Aorta Doppler



Abdominal Aortic Doppler



Significant Coarctation

Normal

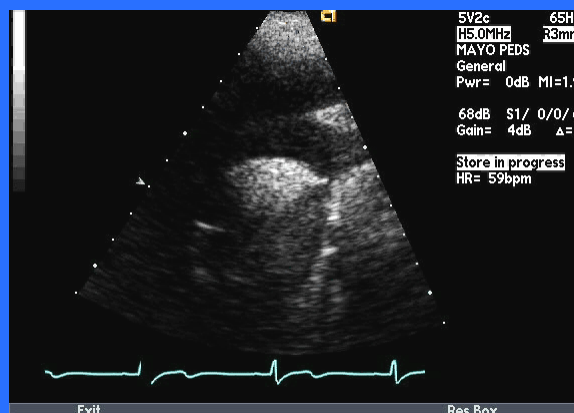


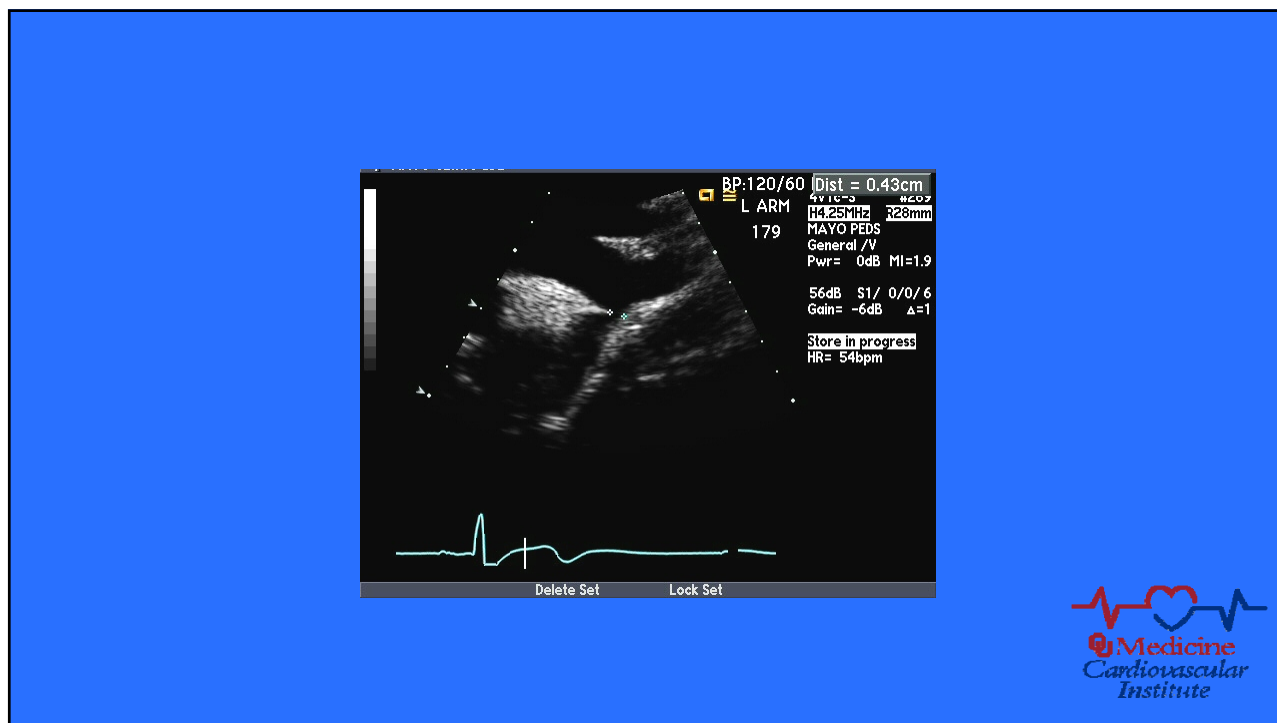
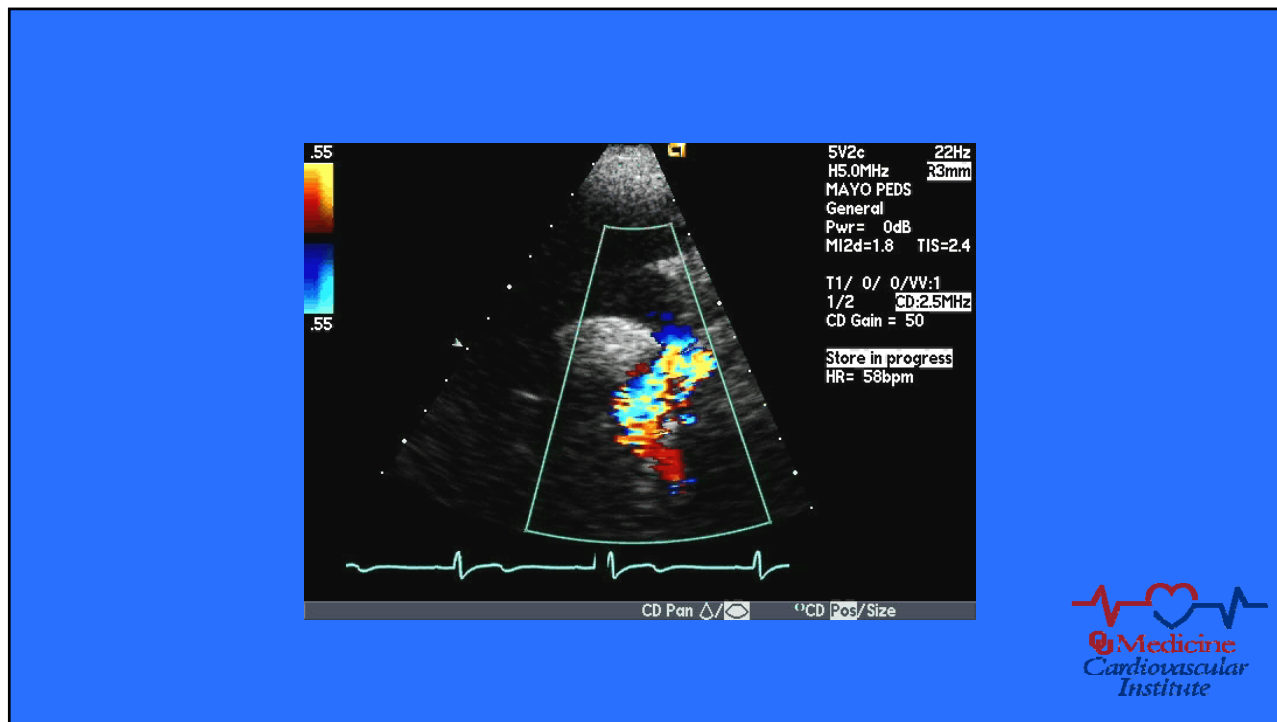
Imaging of Coarctation of the Aorta

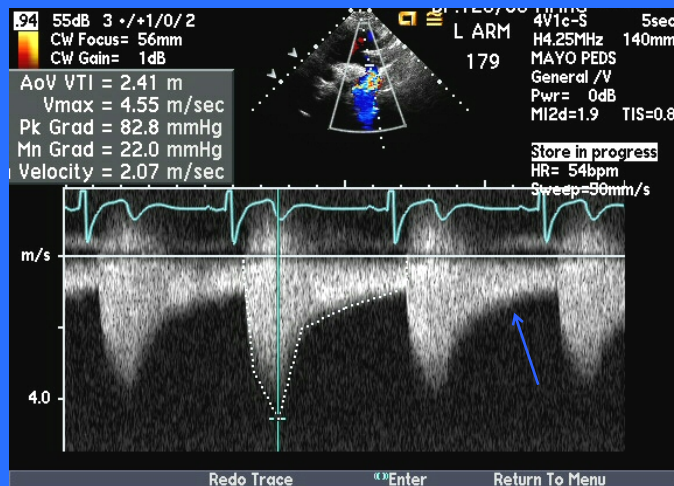
- Abdominal aorta Doppler
- Suprasternal notch imaging
- Parasternal short axis - ?BAV
- Parasternal long axis – ascending aortic dimension



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



“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

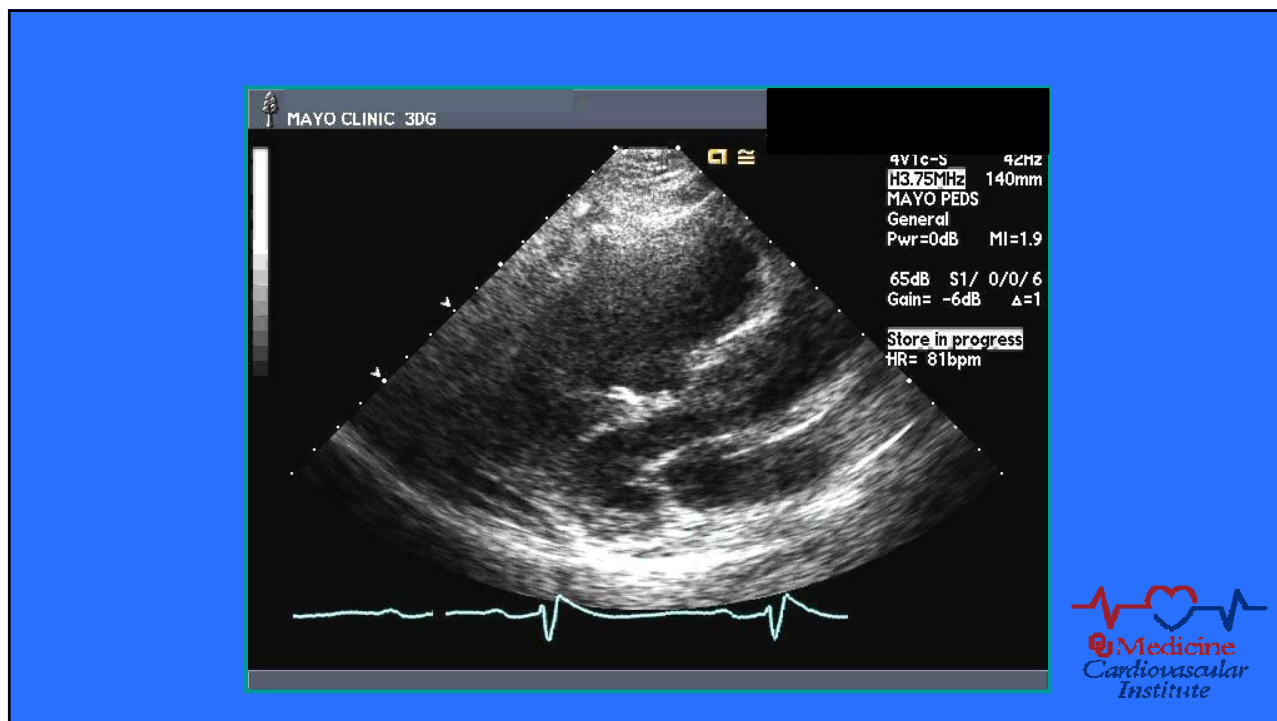
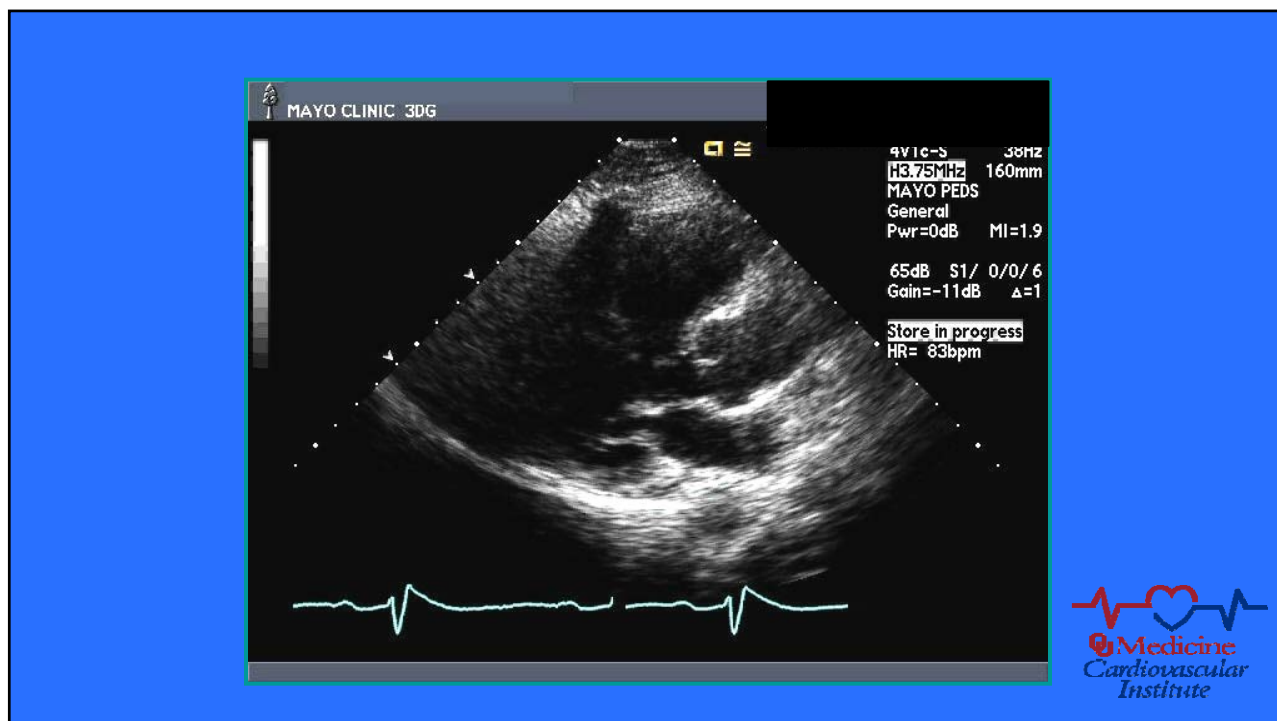
- 6 years: diagnosed with Eisenmenger syndrome
- Treated with frequent phlebotomy
- Placed on Coumadin in adulthood
- Placed on the heart/lung transplant list 5 years (elsewhere)
- No birth control being used

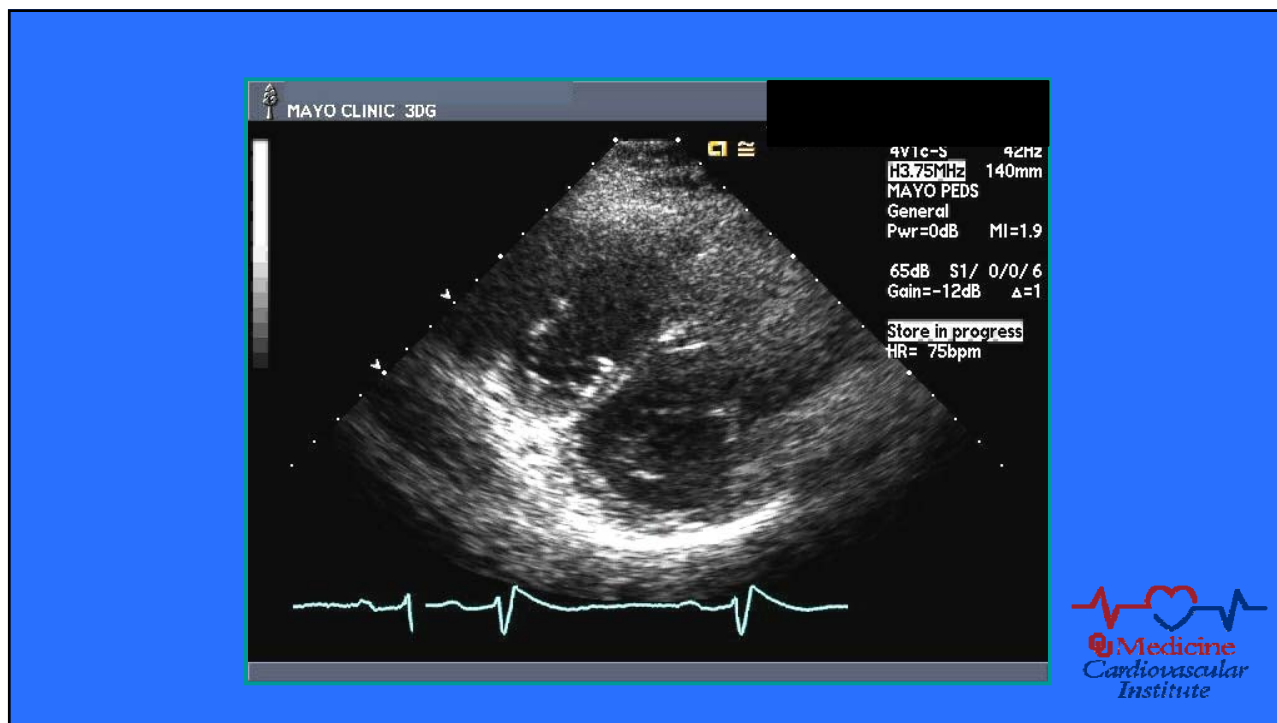
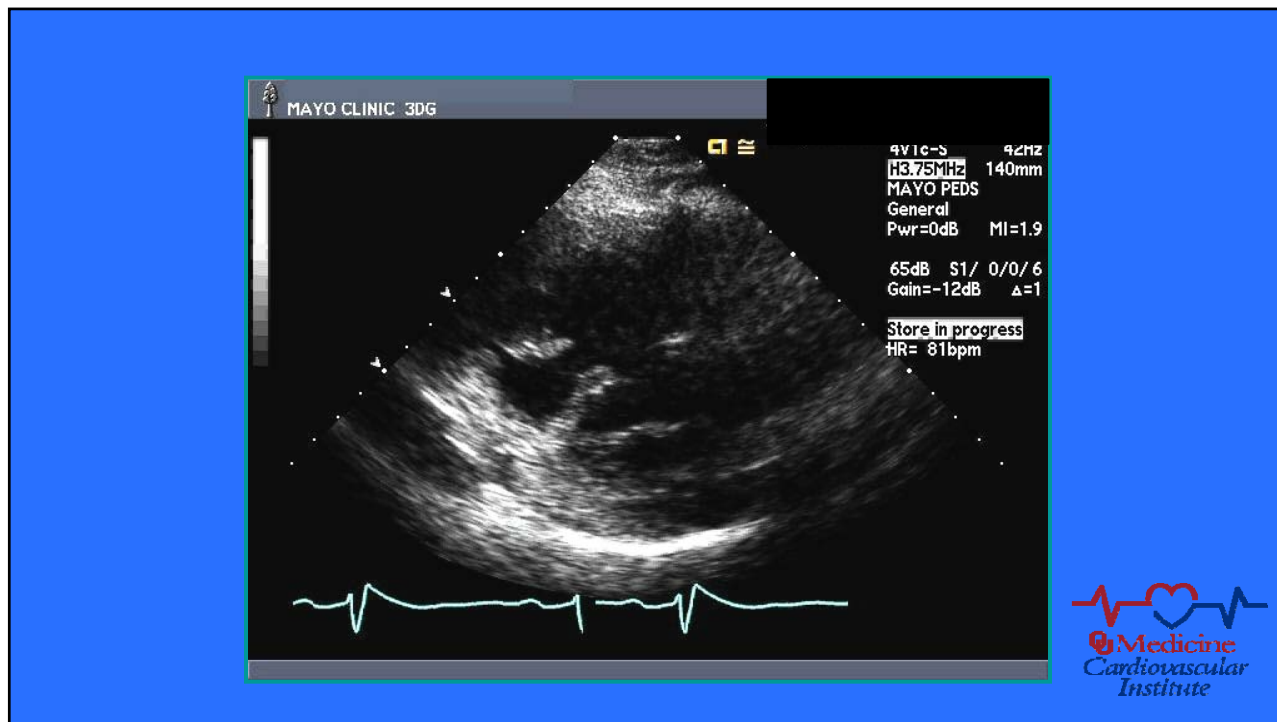


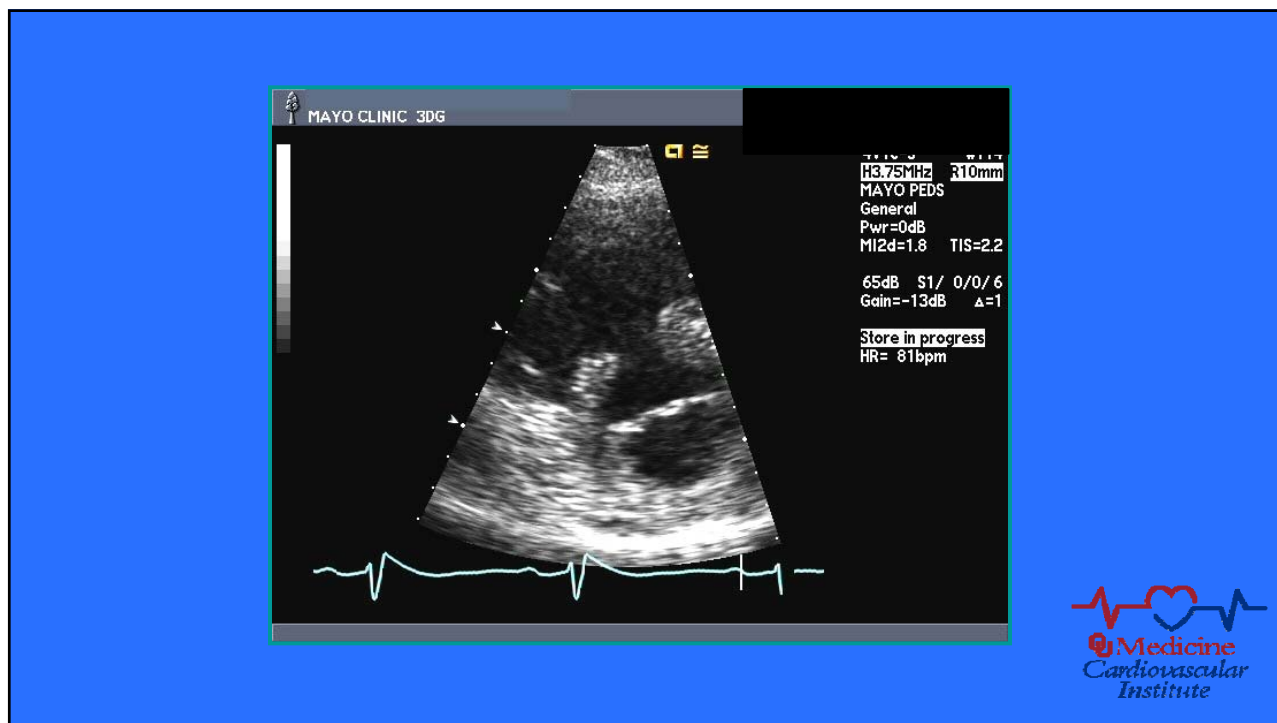
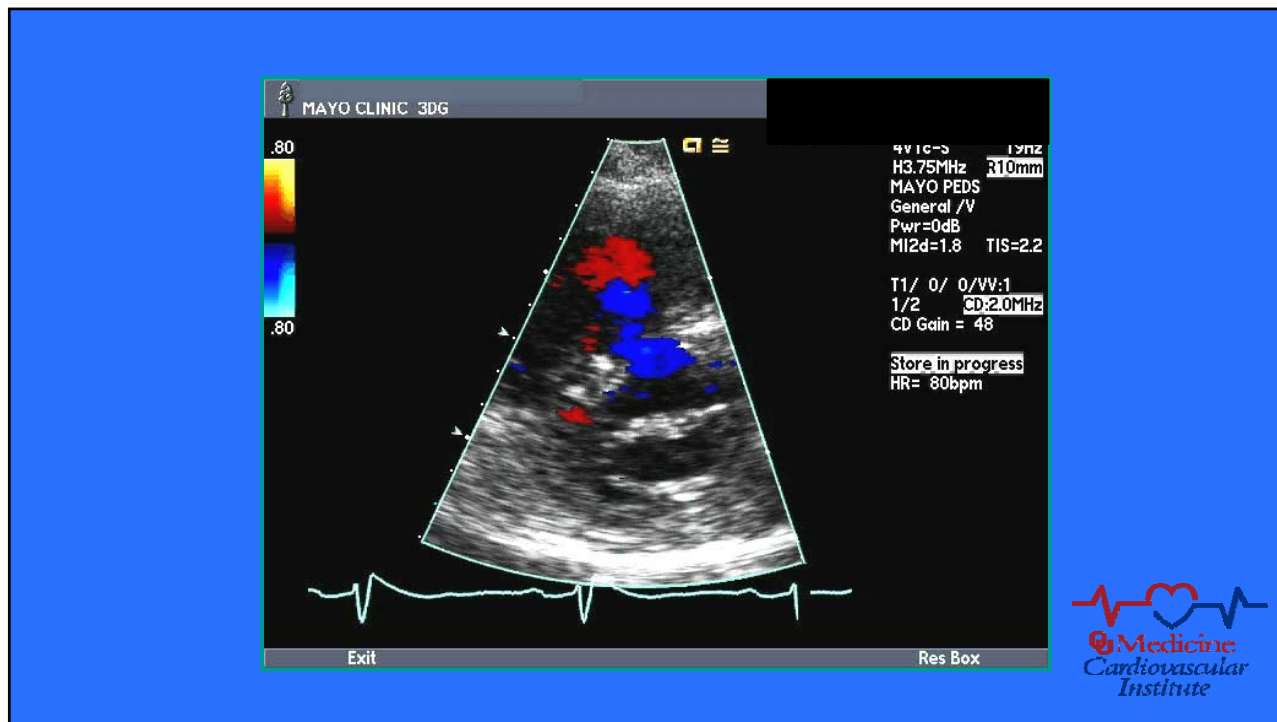
Current Exam

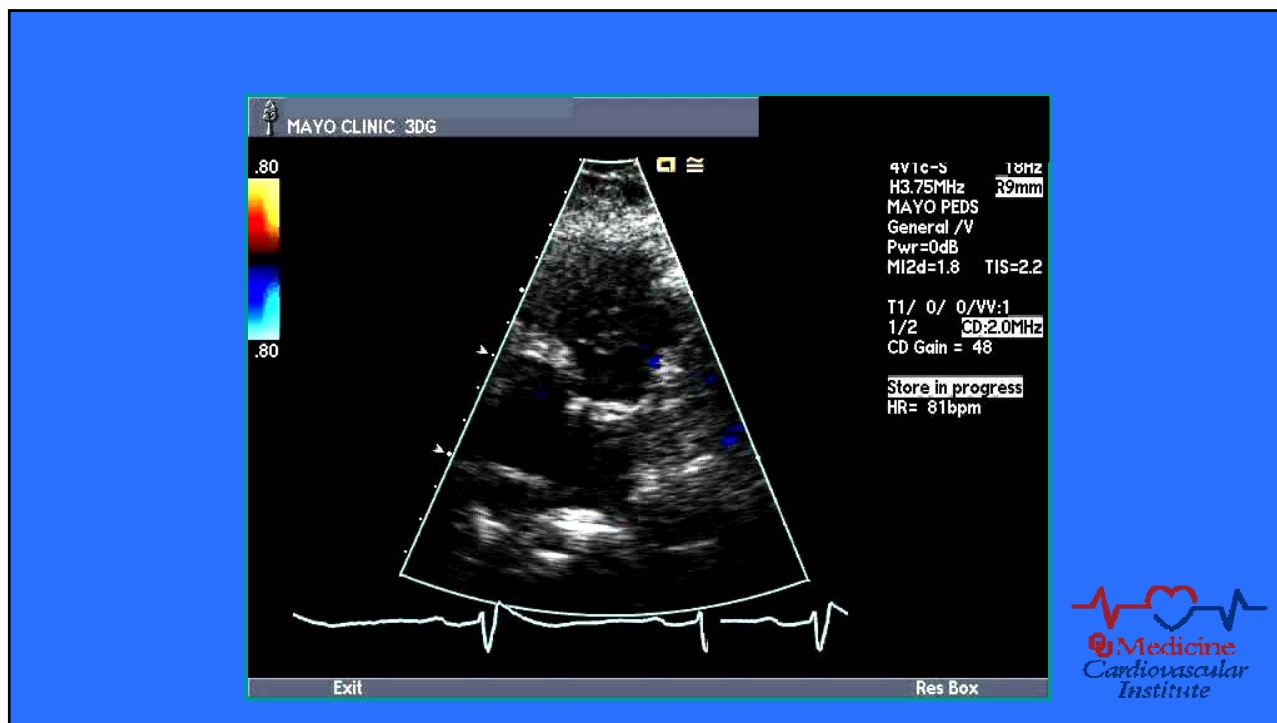
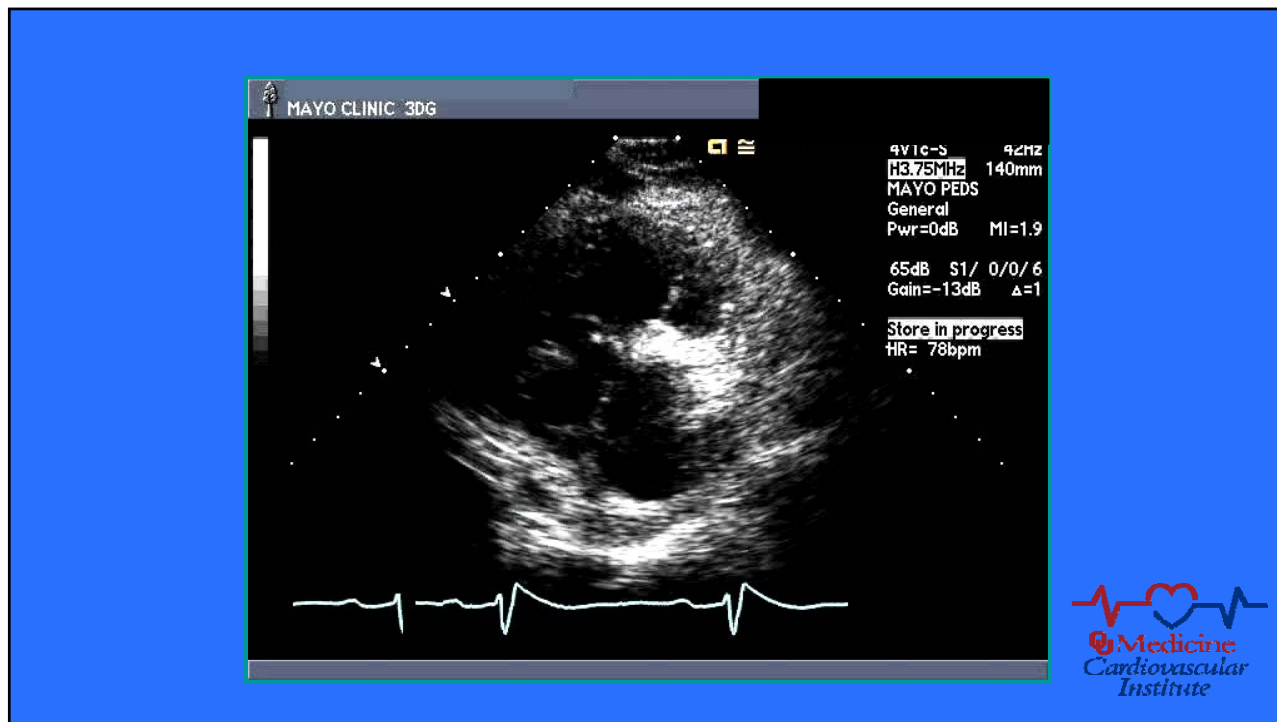
- Significant cyanosis
- Conjunctival injection
- 2+ RV impulse, normal LV impulse
- 3/6 systolic crescendo-decrescendo murmur left upper sternal border
- No diastolic murmur

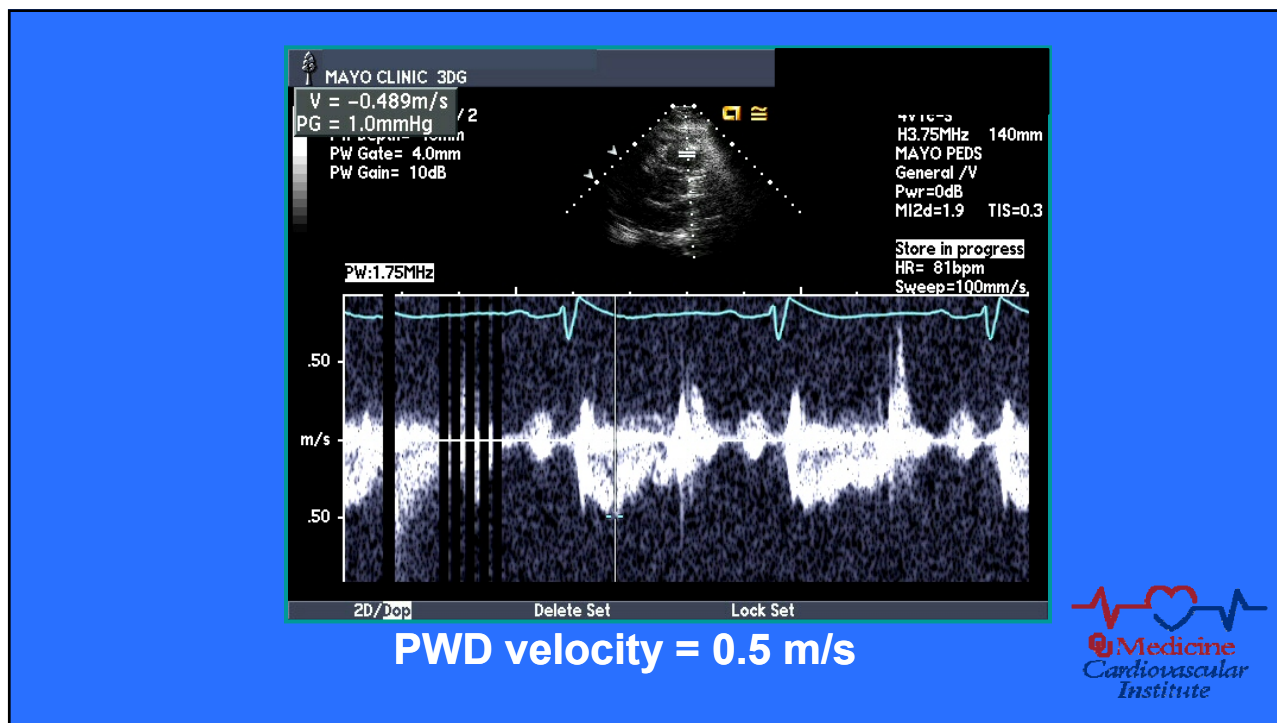
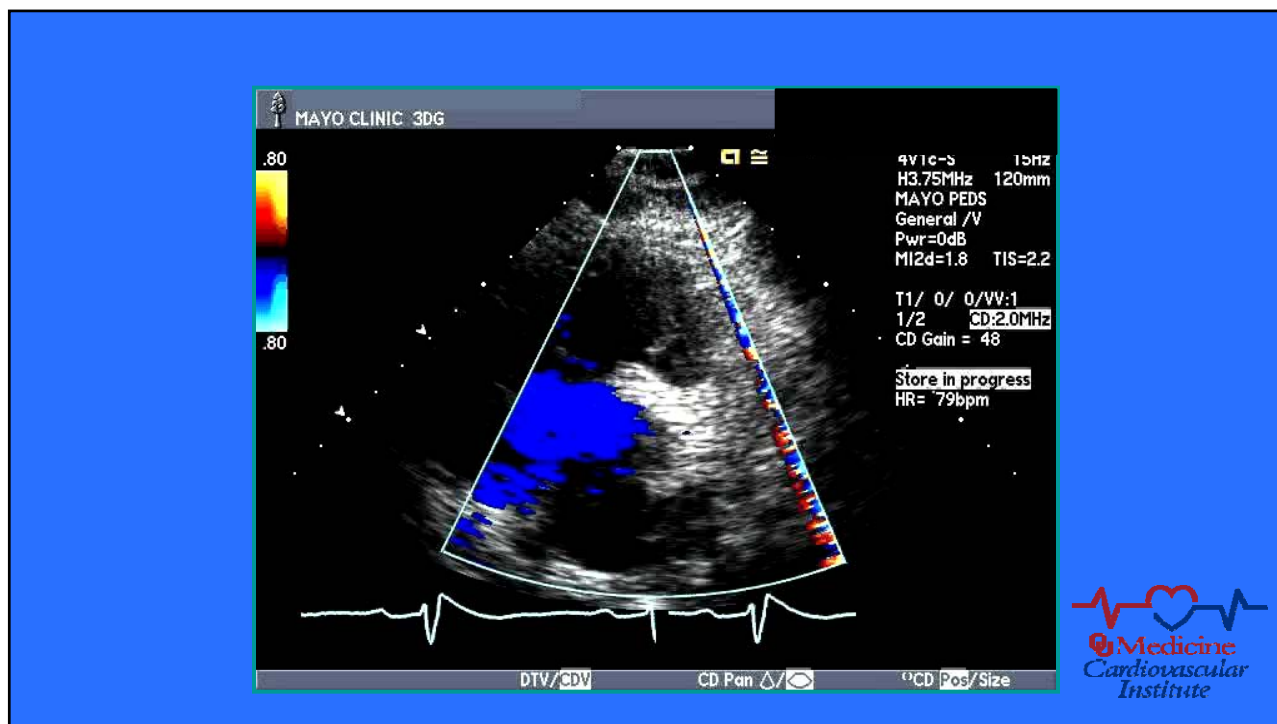






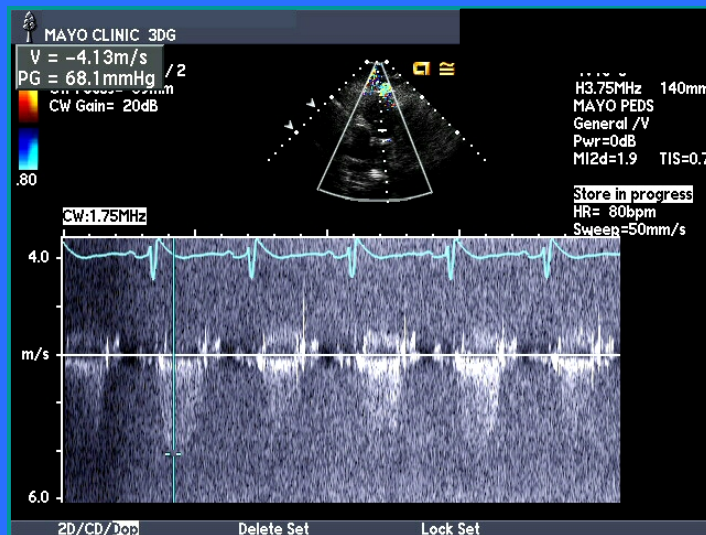




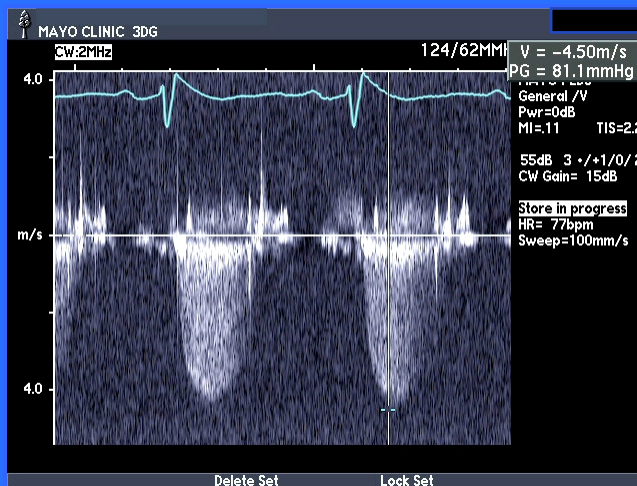


PWD velocity = 0.5 m/s





CWD velocity = 4.1 m/s



Non-imaging Doppler velocity = 4.5 m/s



Does This Patient Have Eisenmenger's Syndrome?

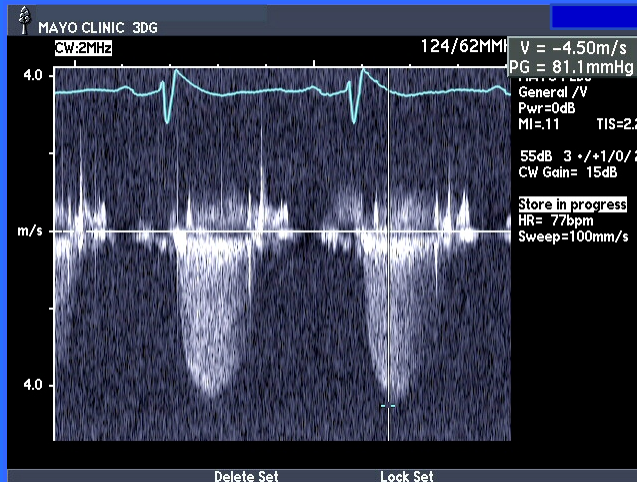
- A. Yes
- B. No**



Does This Patient Have Eisenmenger's Syndrome?

- A. Yes
- B. No**





Non-imaging Doppler velocity = 4.5 m/s



What Next ?

- A. Cath
- B. MRI
- C. Sildenafil
- D. Bosentan
- E. Flolan



What Next ?

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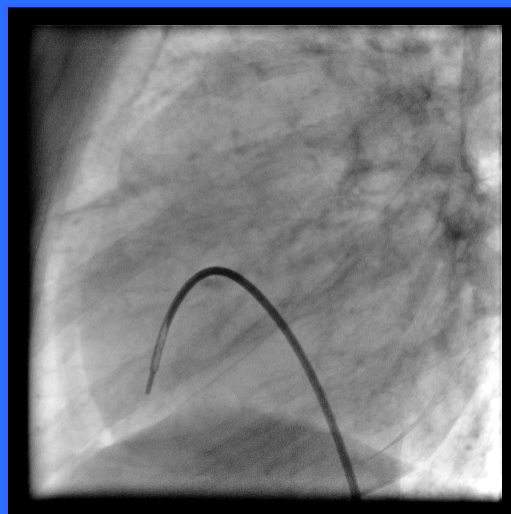
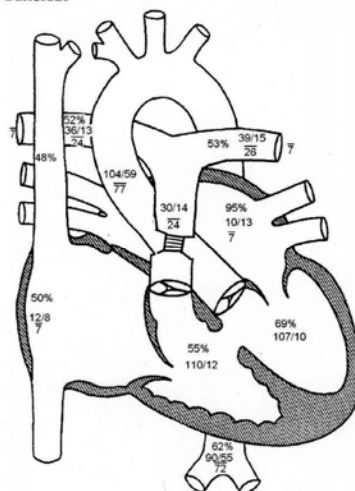


Cath

- 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



CONGENITAL ANATOMY DIAGRAM



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

