

# Commonly Encountered Congenital Heart Disease in Adults

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**No Disclosures**



A 24 year old woman presents with dyspnea on exertion. On echocardiogram is found to have right heart dilatation with normal estimated RV systolic pressure. You should assess for which shunt lesions?

- A. Unroofed coronary sinus**
- B. Ventricular septal defect**
- C. Patent ductus arteriosus**
- D. None of the above**
- E. Both A and B**

A 30 year old man is known to have an unrestrictive membranous ventricular septal defect. On echocardiogram you would expect to find which of the following?

- A. Systolic velocity across the VSD  $> 4$  m/s**
- B. Pulmonary valve cusp prolapse into the defect**
- C. Laminar color Doppler flow across the defect**
- D. Left ventricular dilatation**
- E. AV valves at the same level – no apical displacement of the tricuspid valve**

# Atrial Septal Defect

- Second most common congenital defect recognized in adulthood
- Symptoms progressive
- Physical exam findings subtle

# Atrial Septal Defects

- Secundum
- Primum
- Sinus Venosus
- Unroofed Coronary Sinus

# Atrial Septal Defect

## ➤ Echo Diagnosis and Evaluation

- ◎ Location of Defect
- ◎ Right sided chamber size and function
- ◎ Estimation of PA pressure
- ◎ Tricuspid Regurgitation
- ◎ Other Lesions
- ◎ Repair Options

# Secundum Atrial Septal Defect

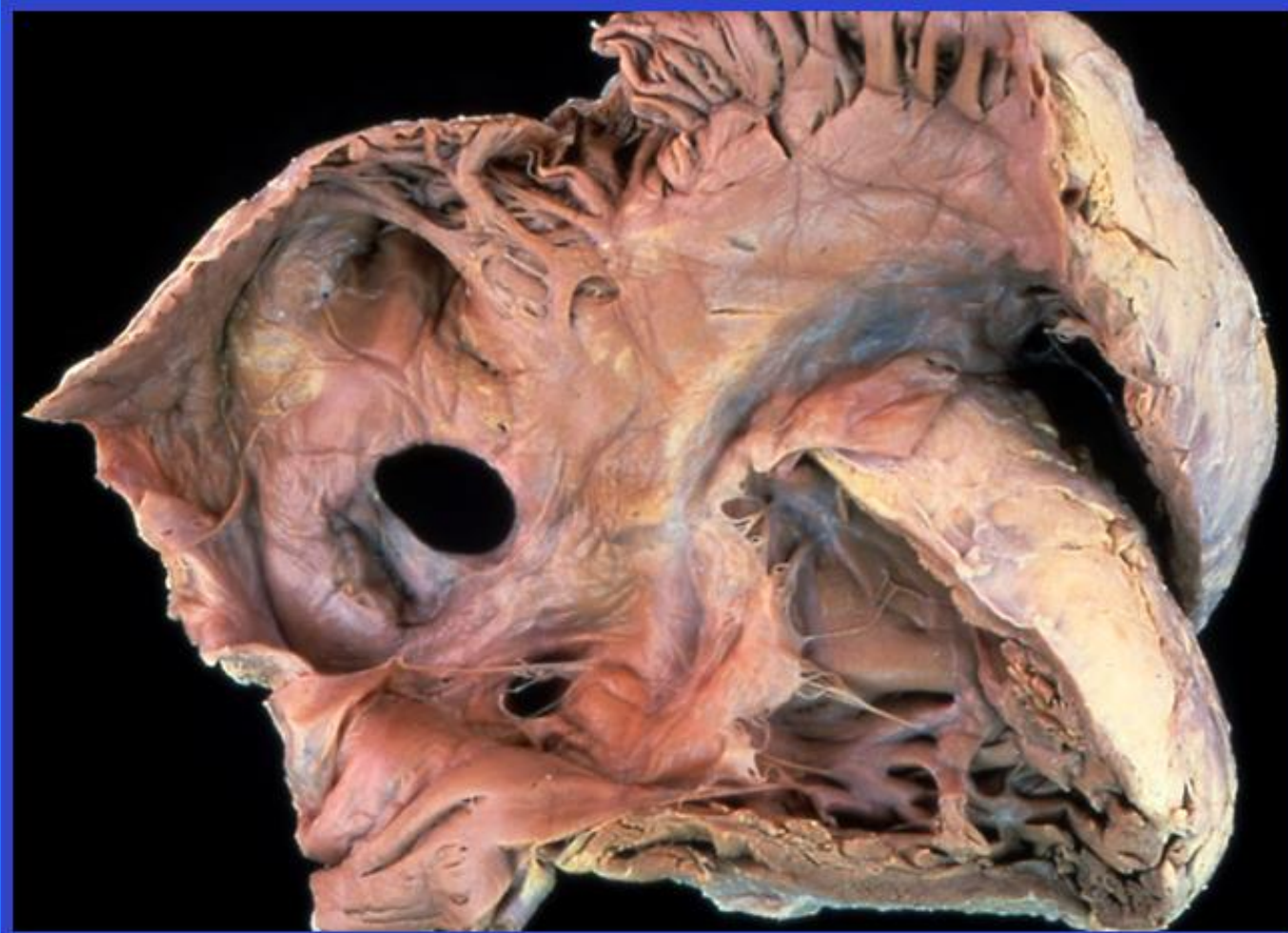
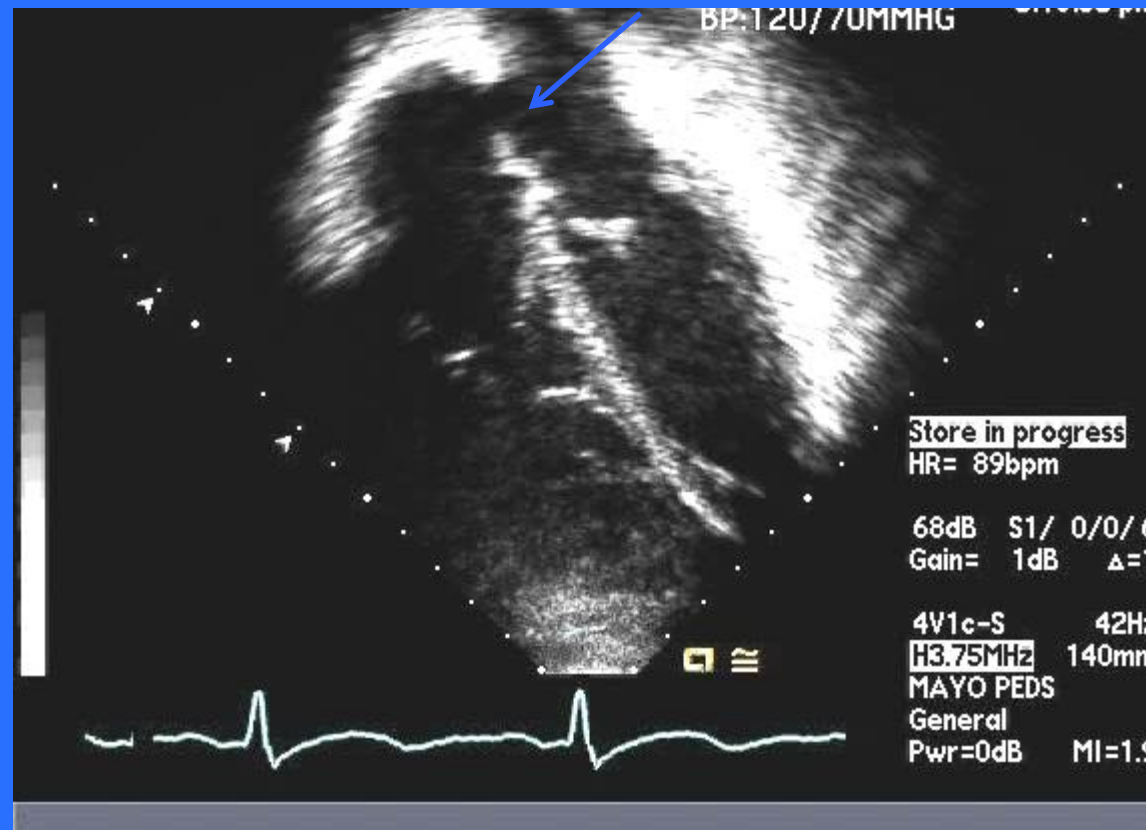


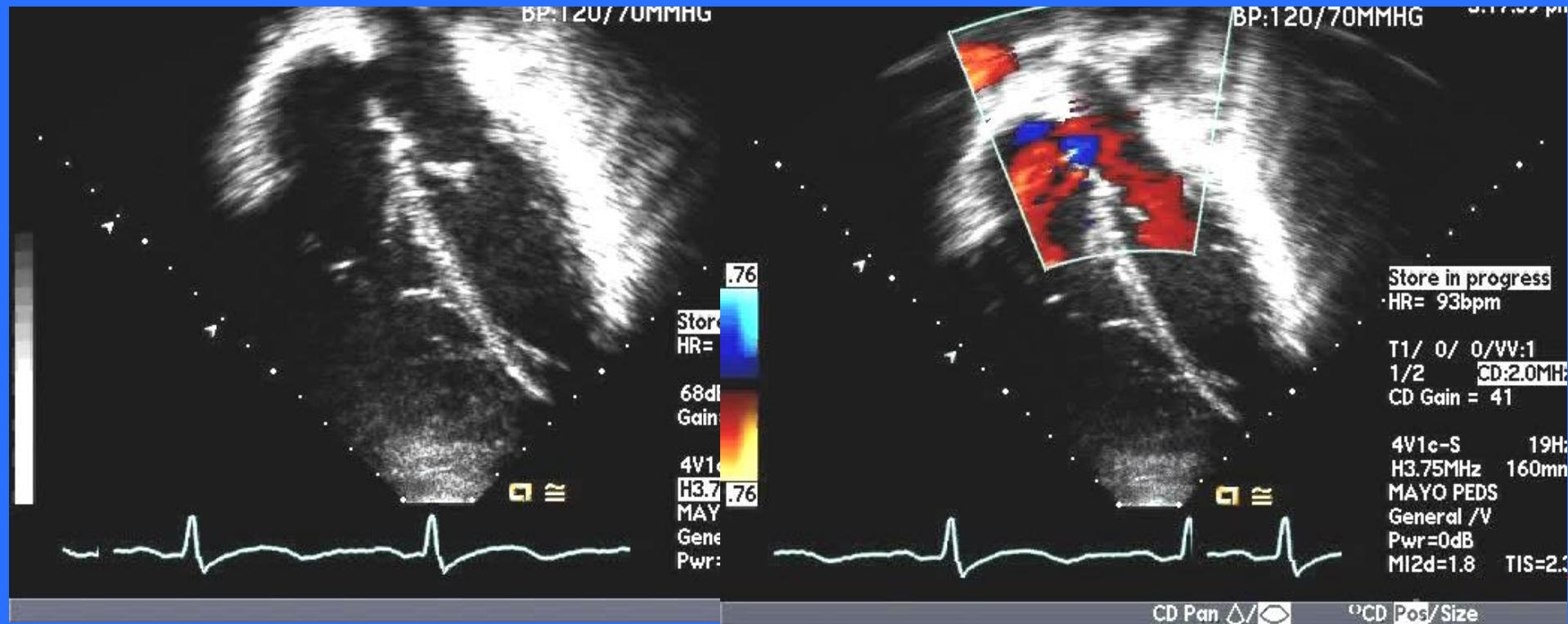
Image Courtesy of Dr. Bill Edwards



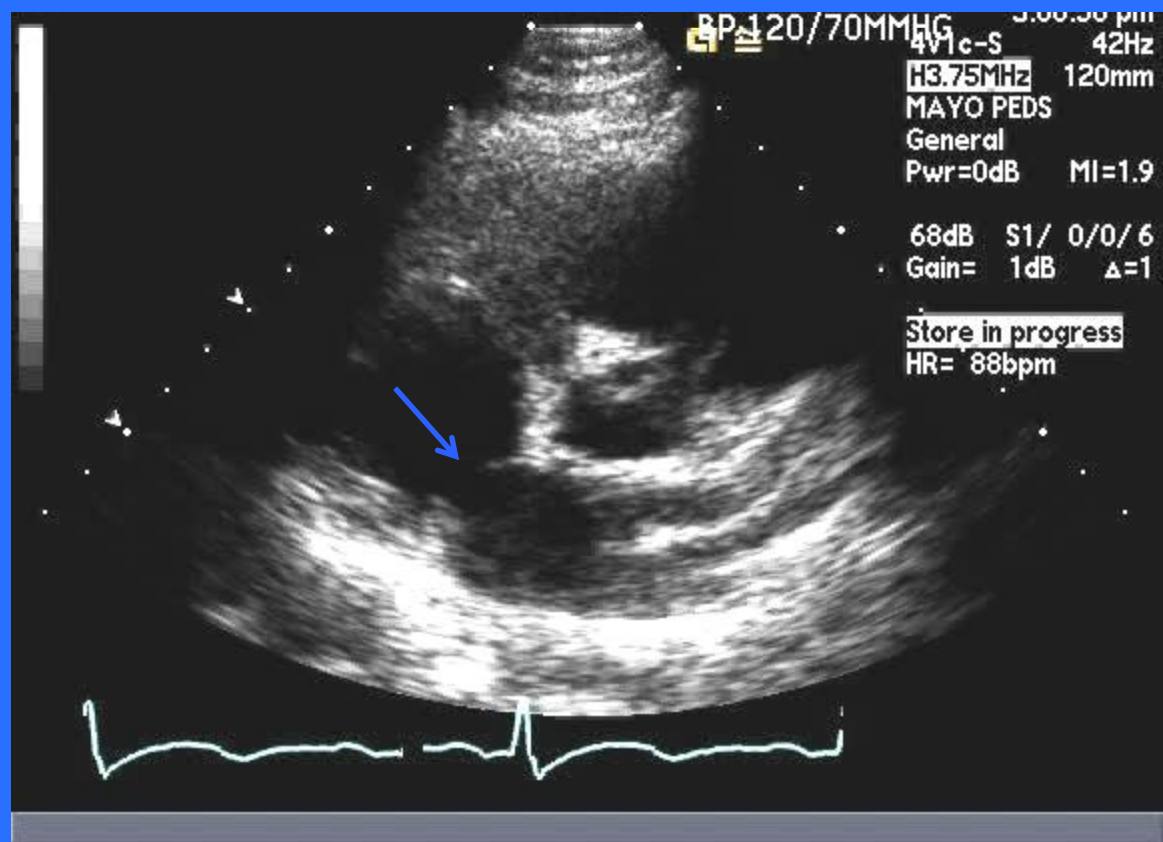
# Apical 4 Chamber Imaging



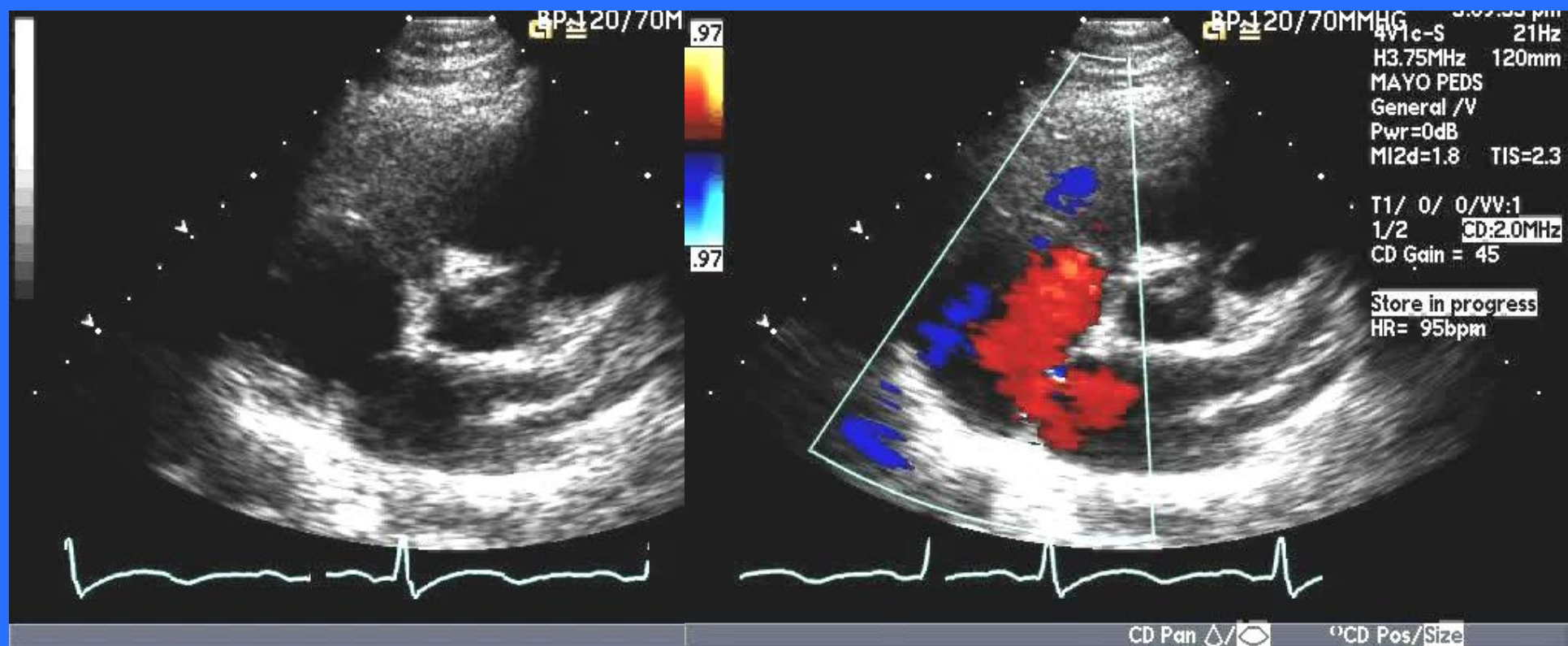
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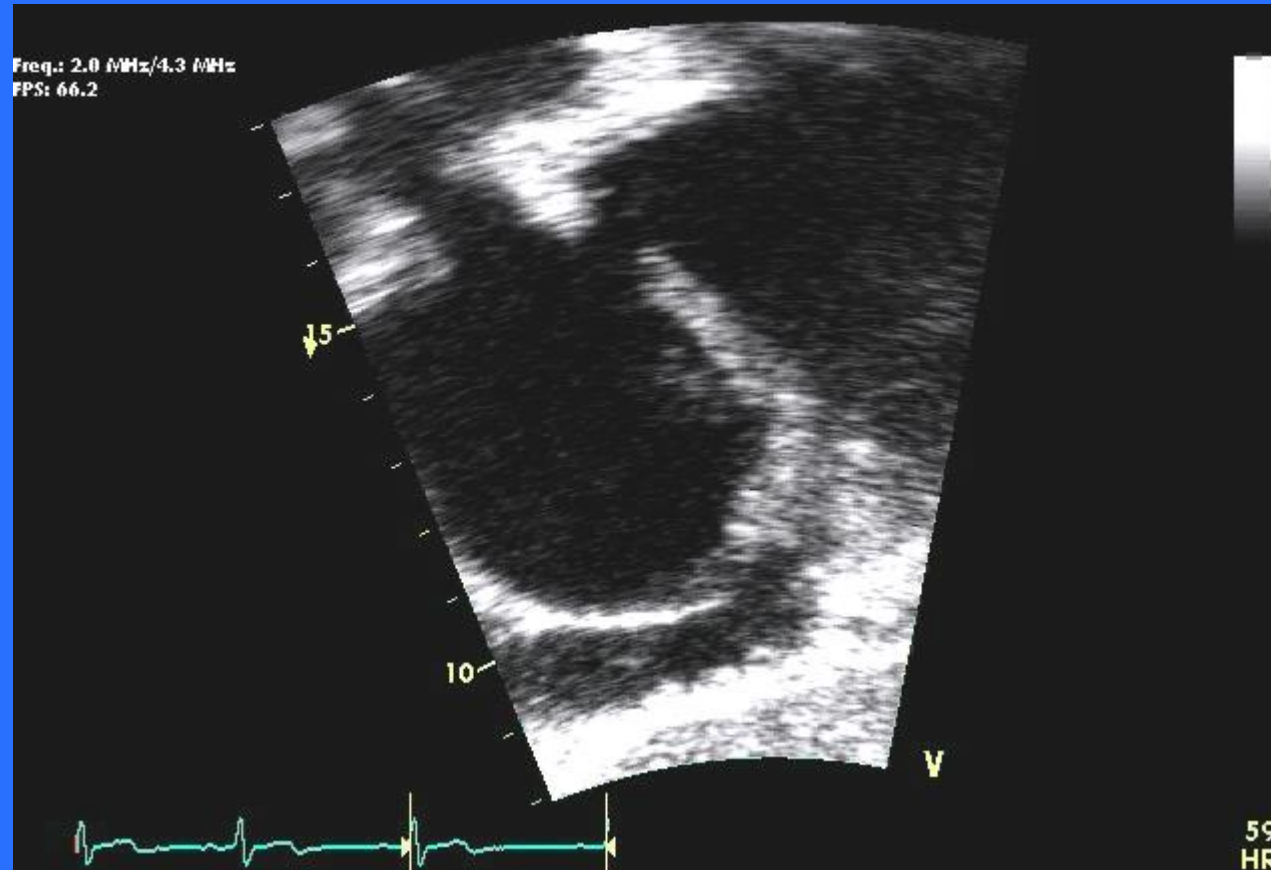
# Parasternal Short Axis Imaging

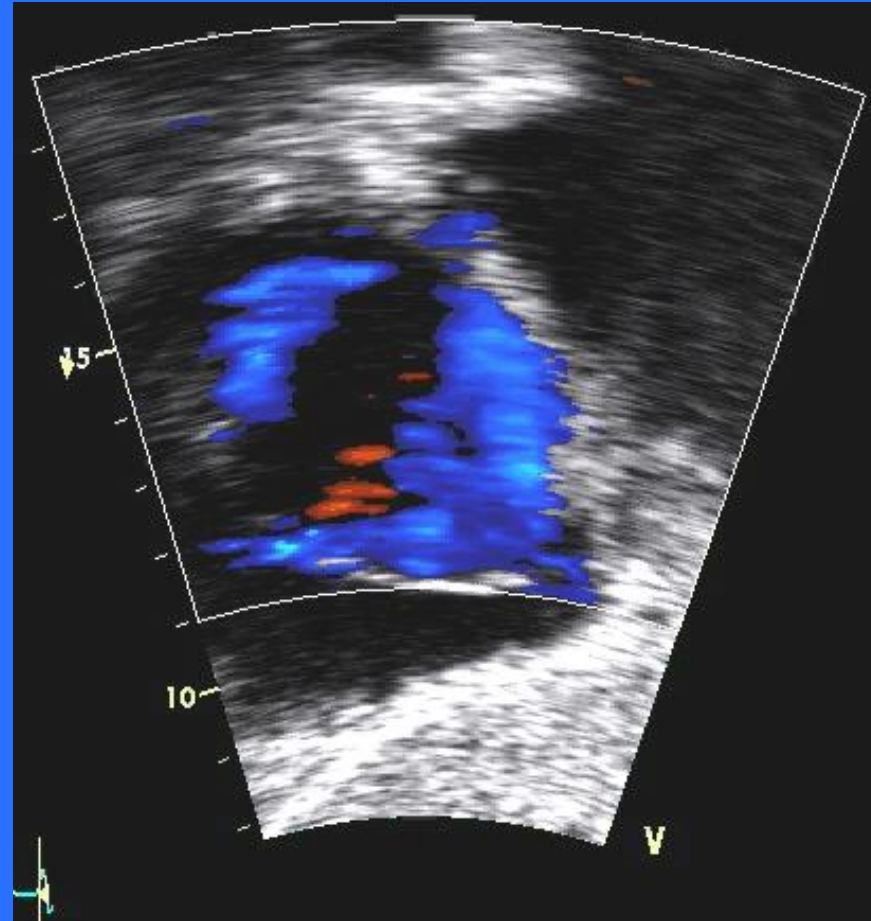
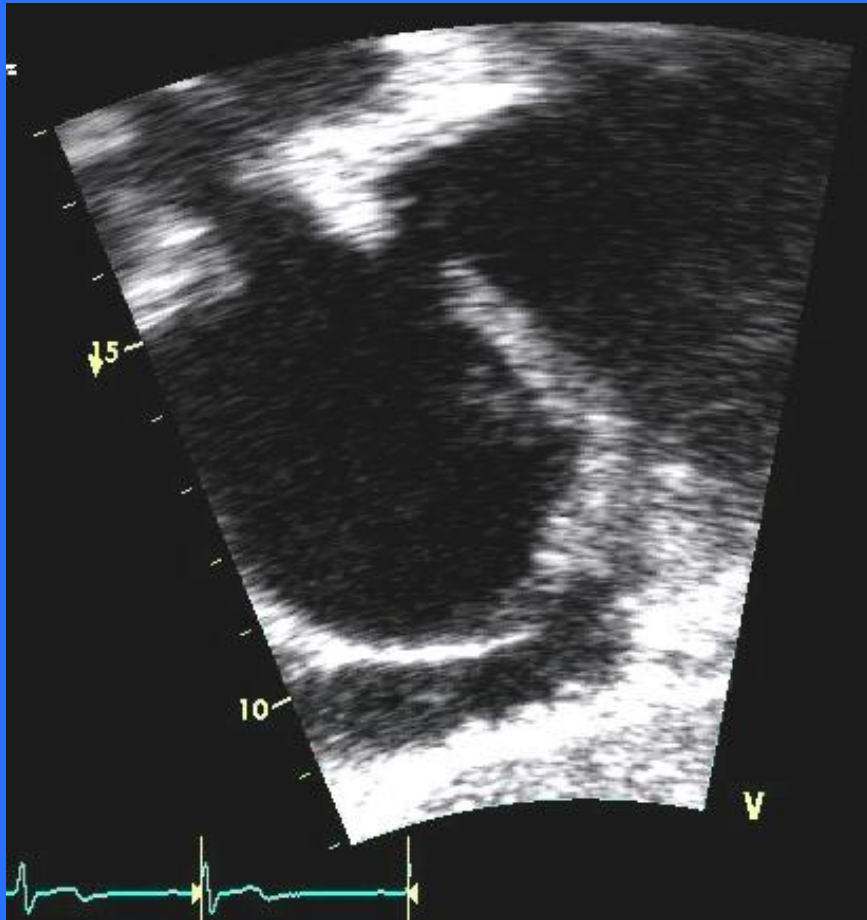


# Parasternal Short Axis Imaging



# Subcostal Imaging





# Primum Atrial Septal Defect

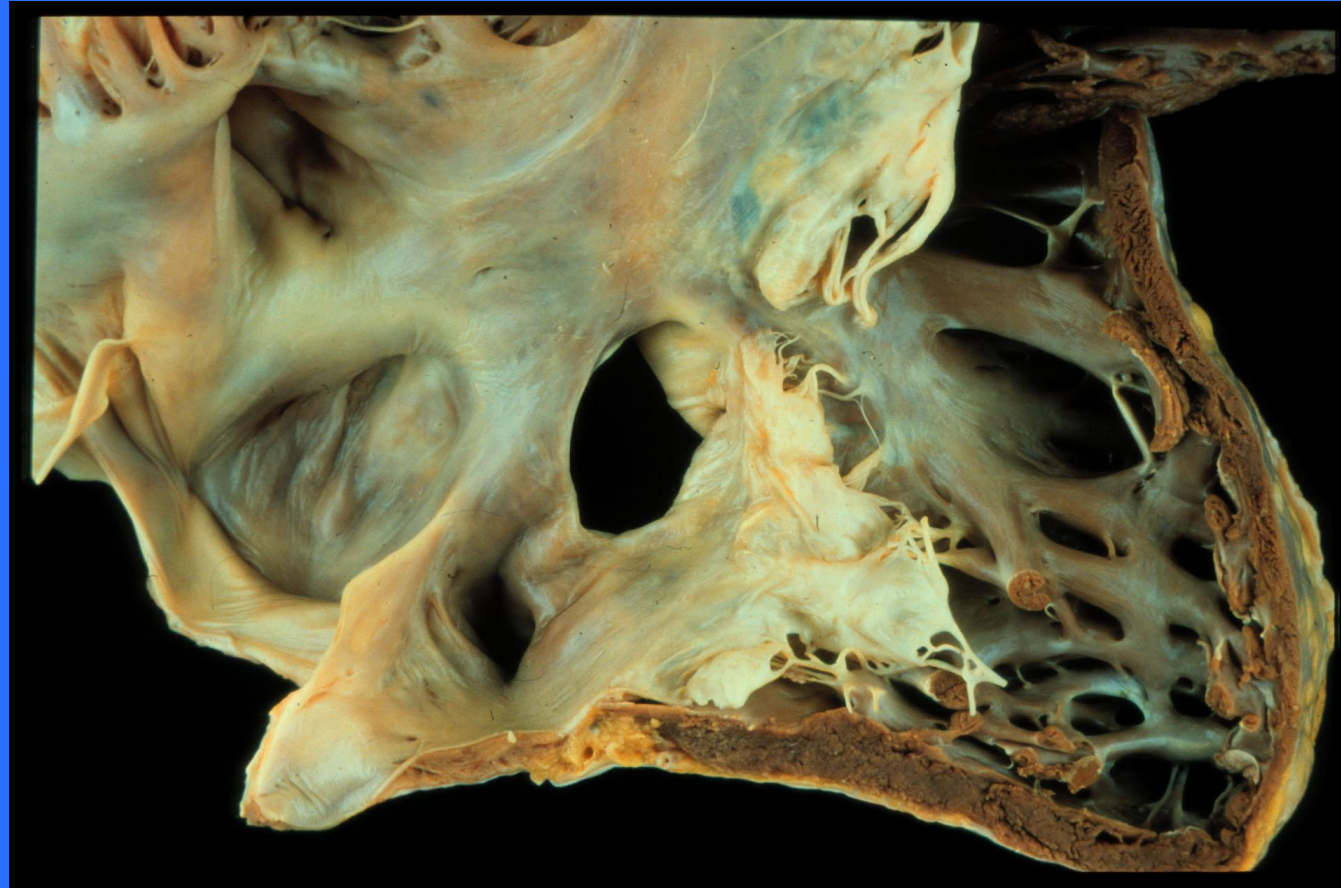
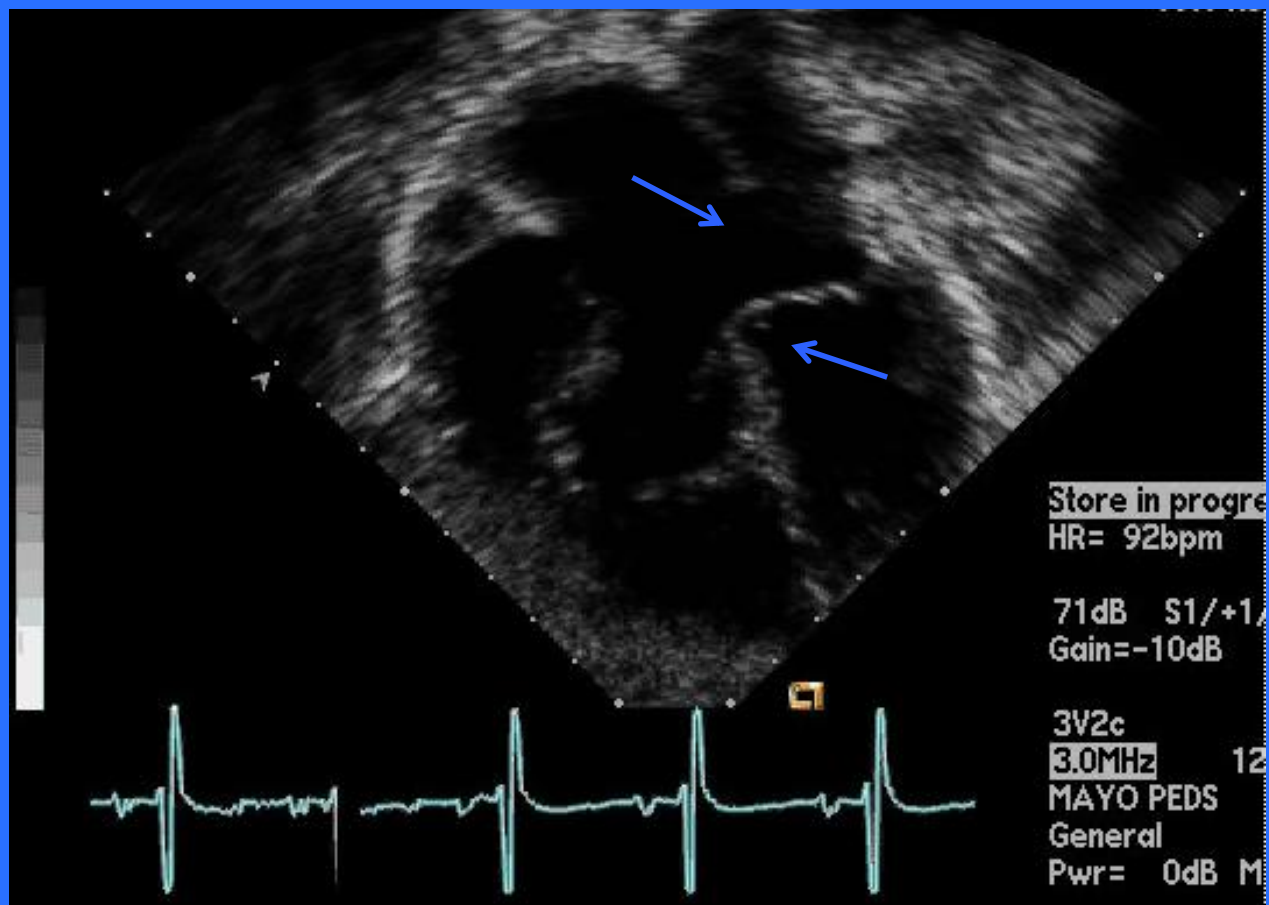


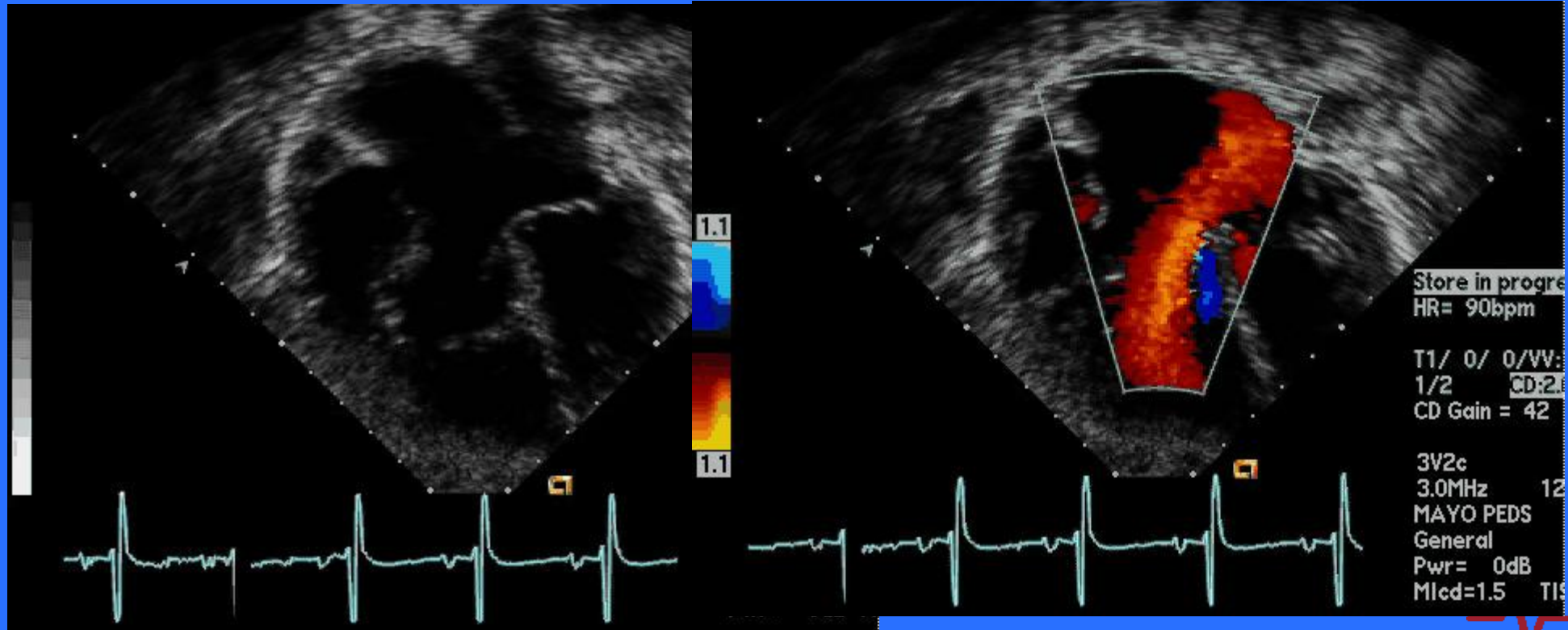
Image Courtesy of Dr. Bill Edwards

# Apical 4 Chamber Imaging





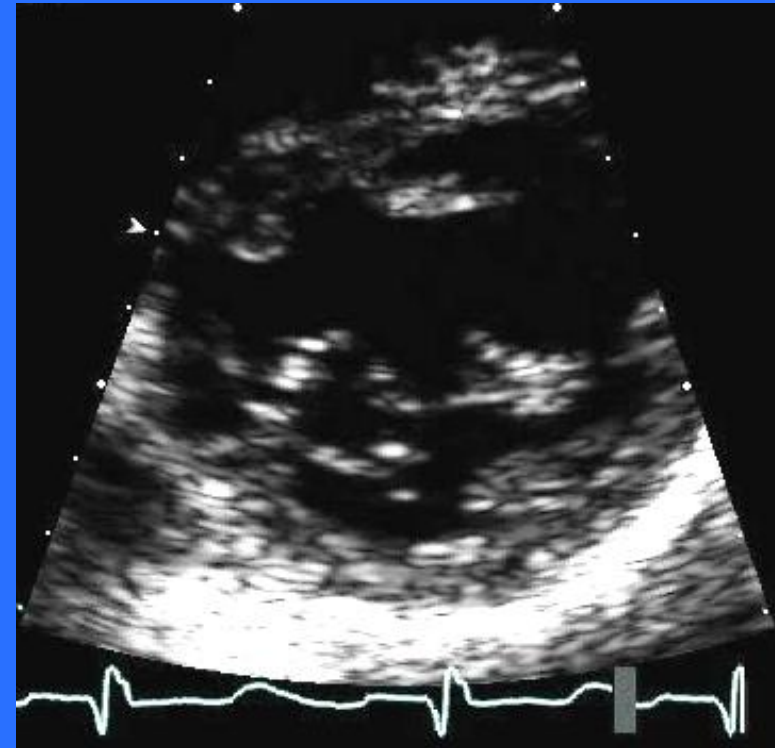
# Apical 4 Chamber Imaging



# Valvular Abnormalities Associated with Primum ASD

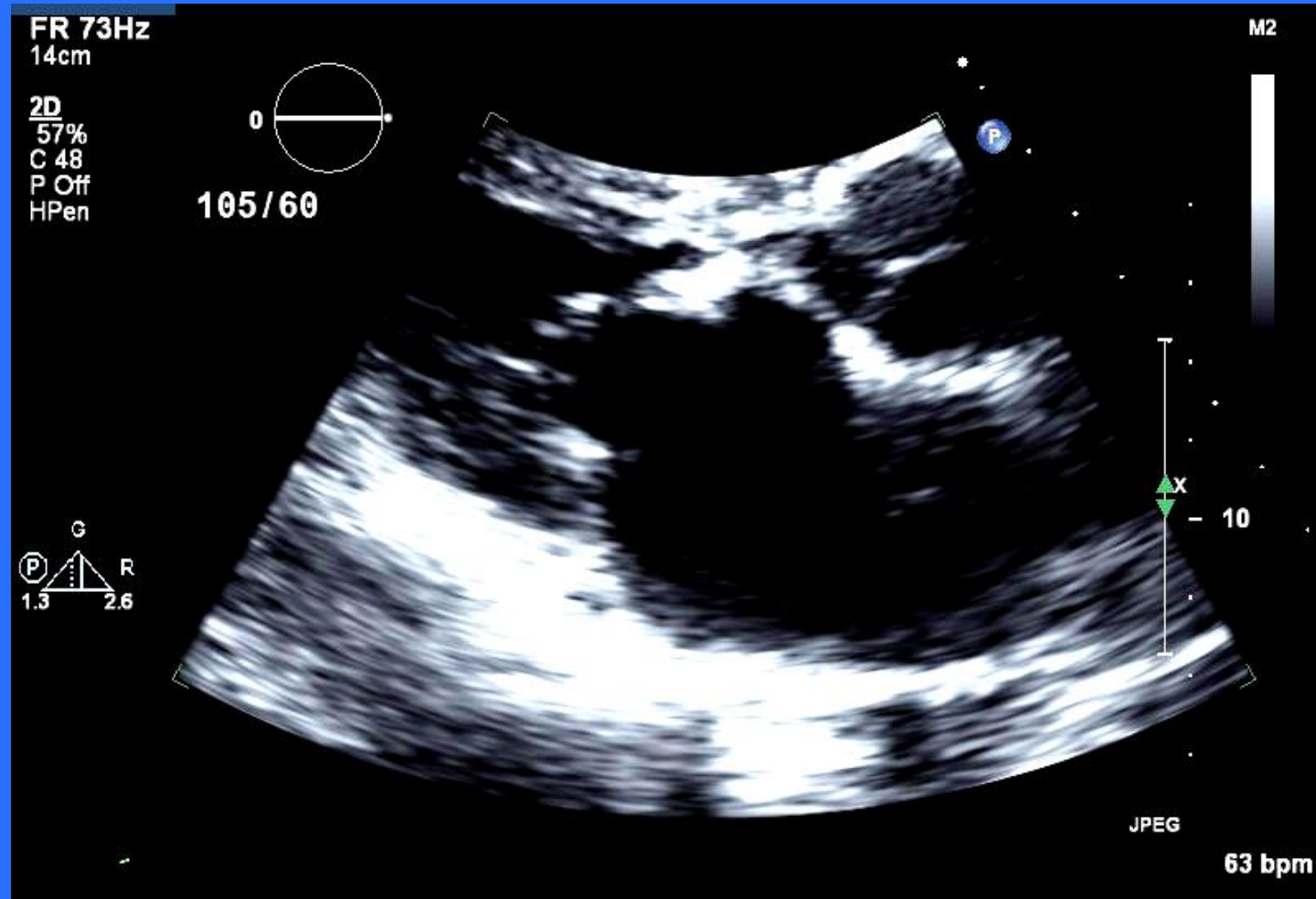


Cleft Mitral Valve



Double Orifice  
Mitral Valve

# LVOT Elongation, Narrowing, Anomalous Chords



# Sinus Venosus Atrial Septal Defect

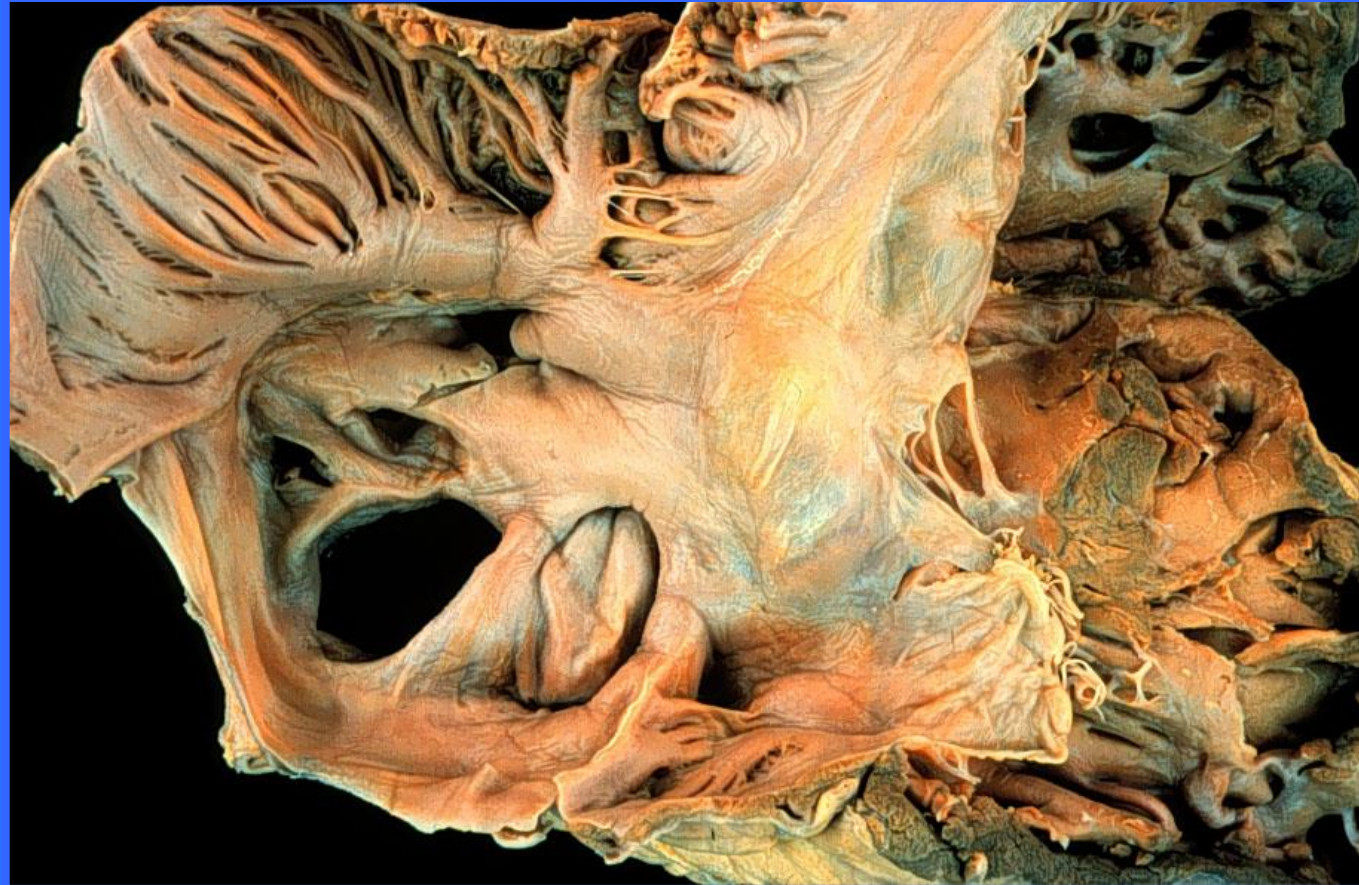
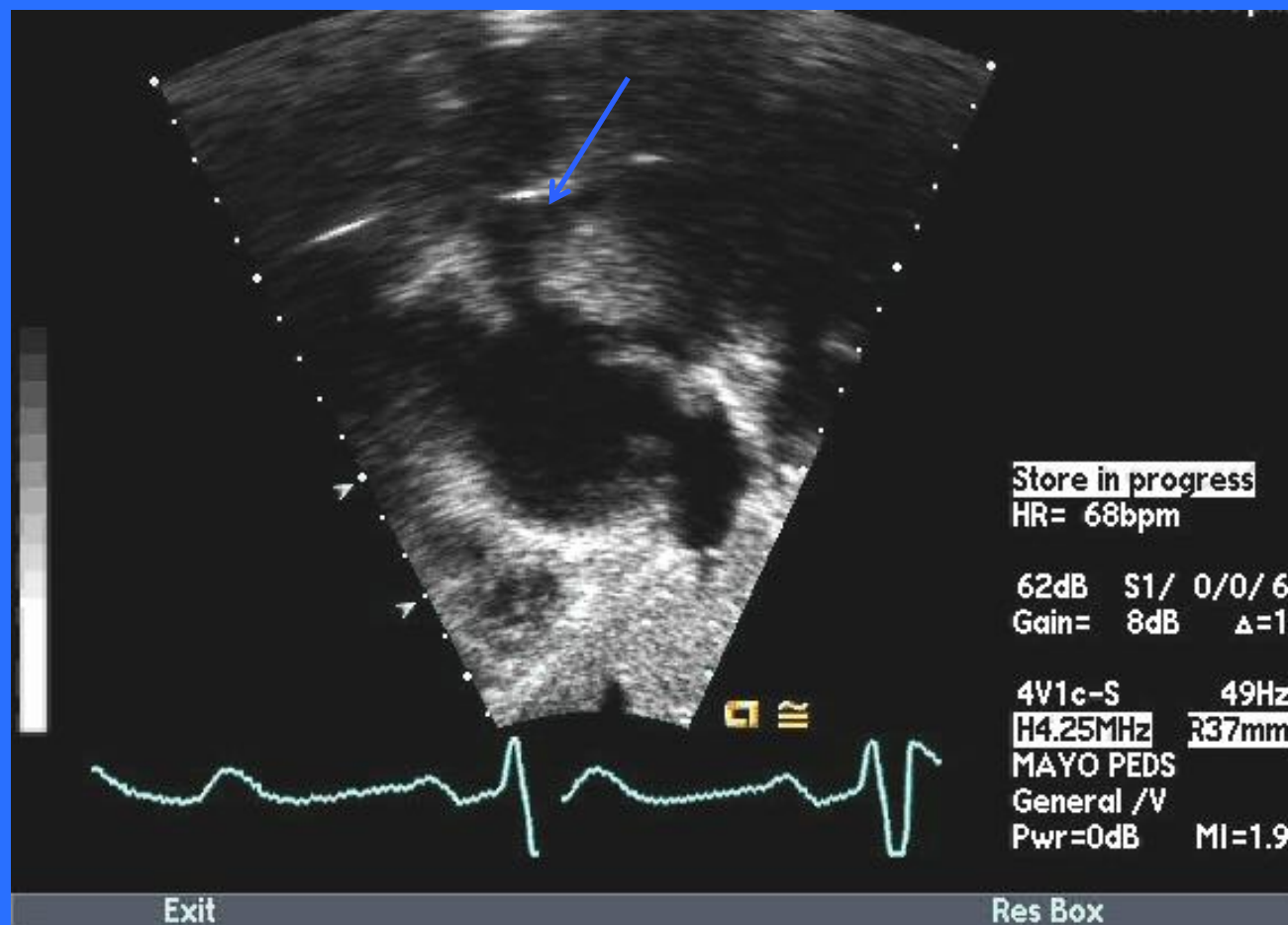
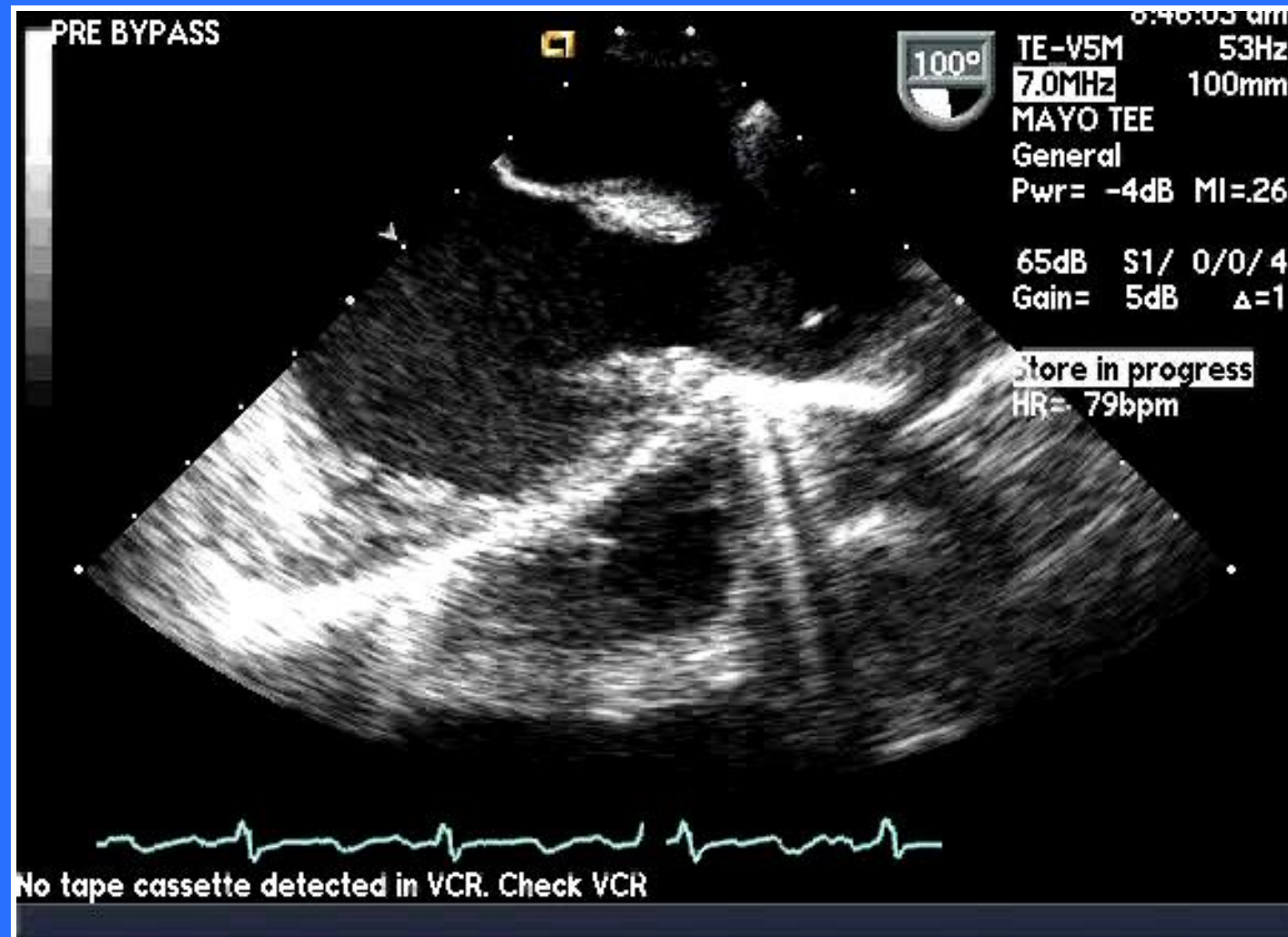


Image Courtesy of Dr. Bill Edwards

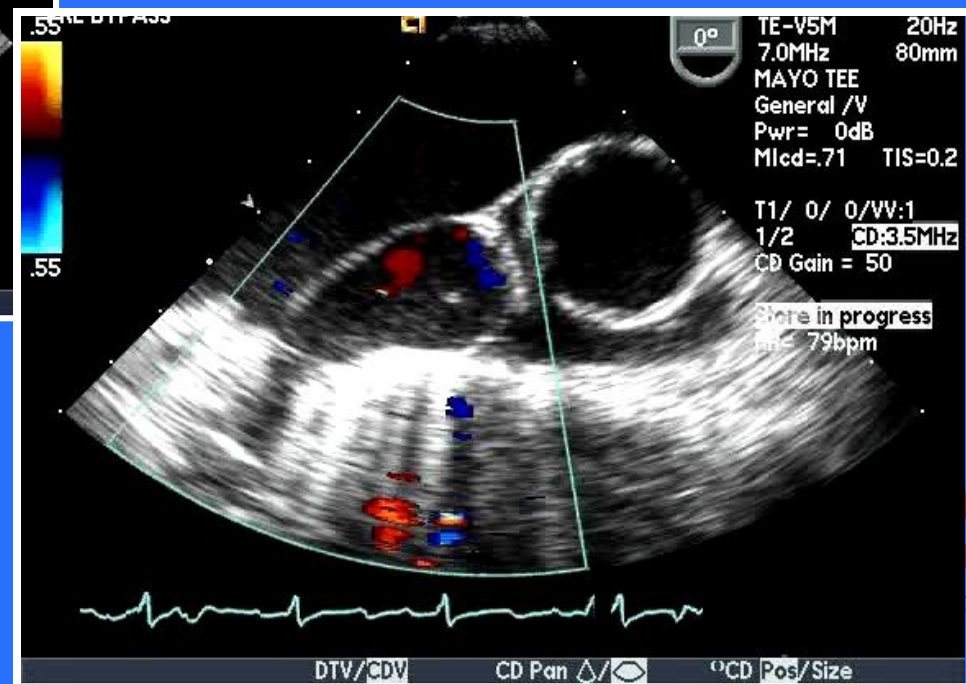
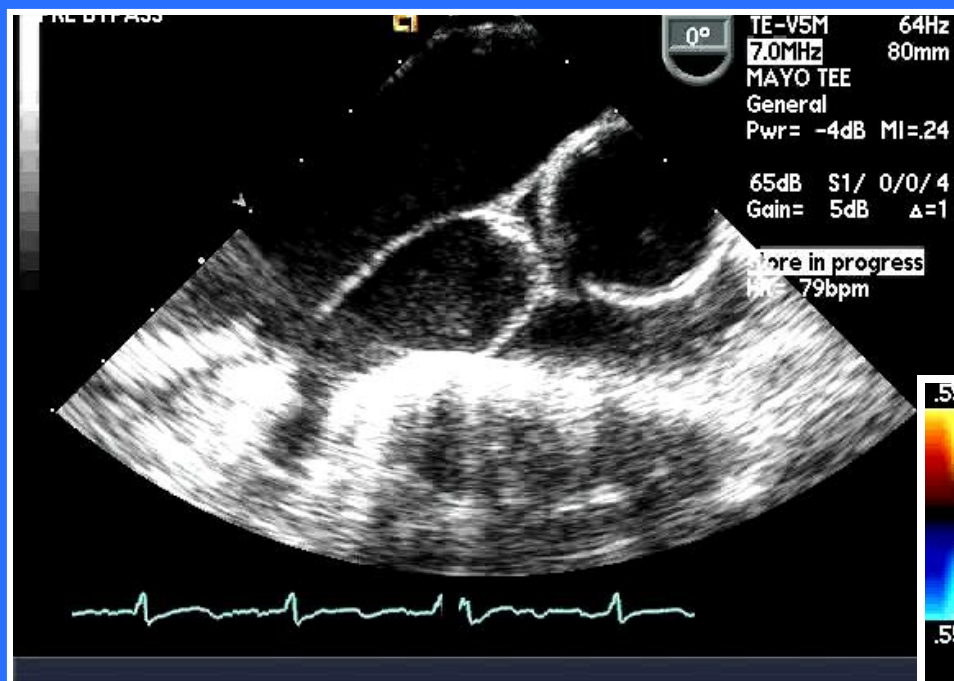
# Subcostal Imaging



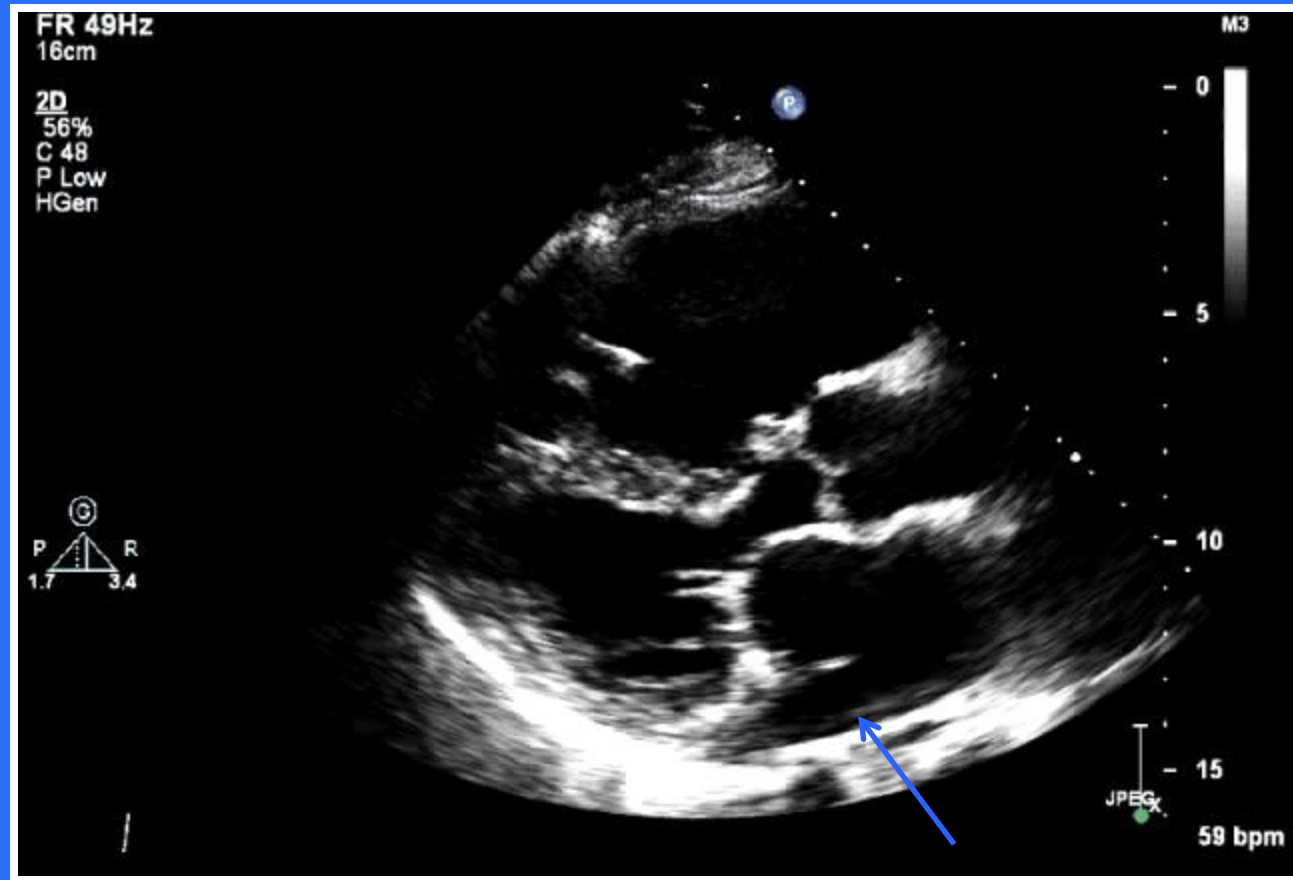
# TEE Imaging



# Anomalous Right Pulmonary Vein

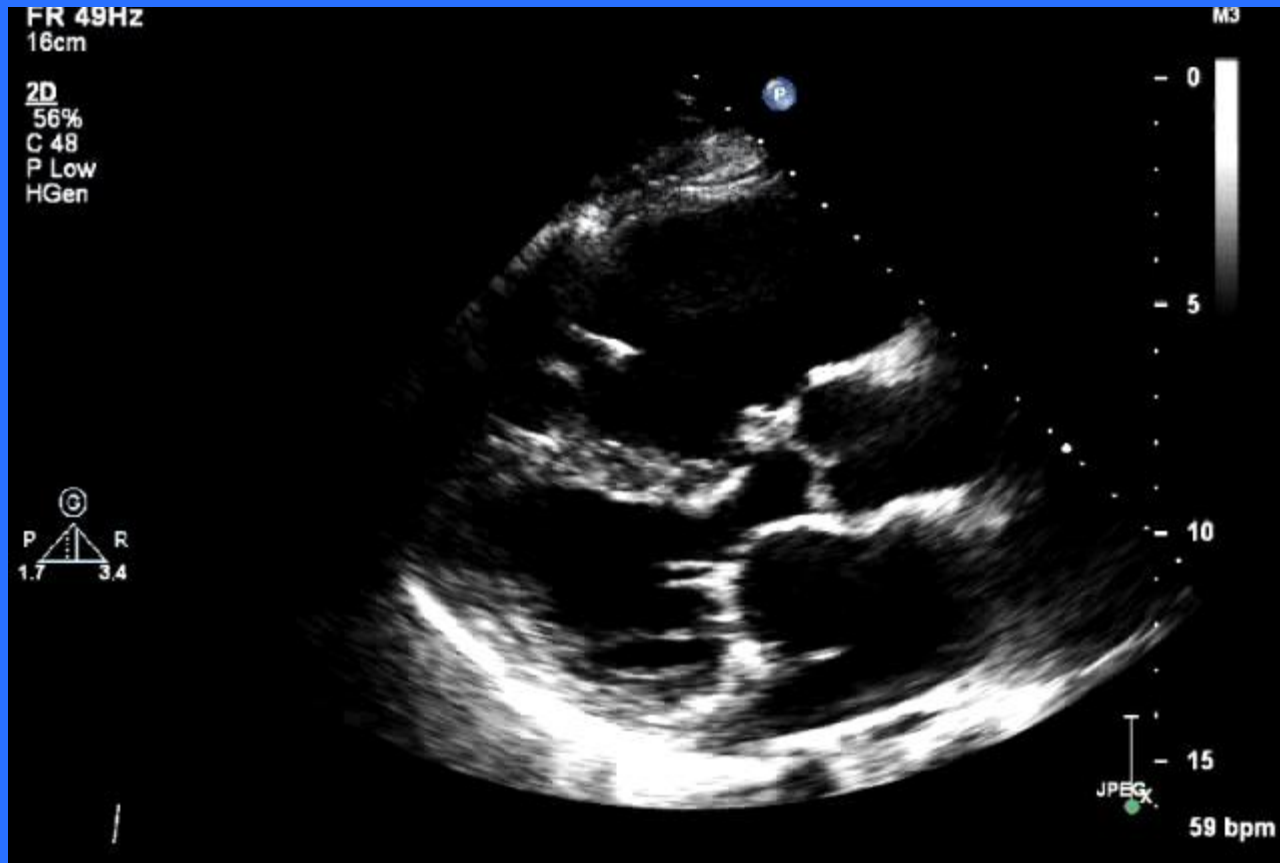


# Coronary Sinus Atrial Septal Defect

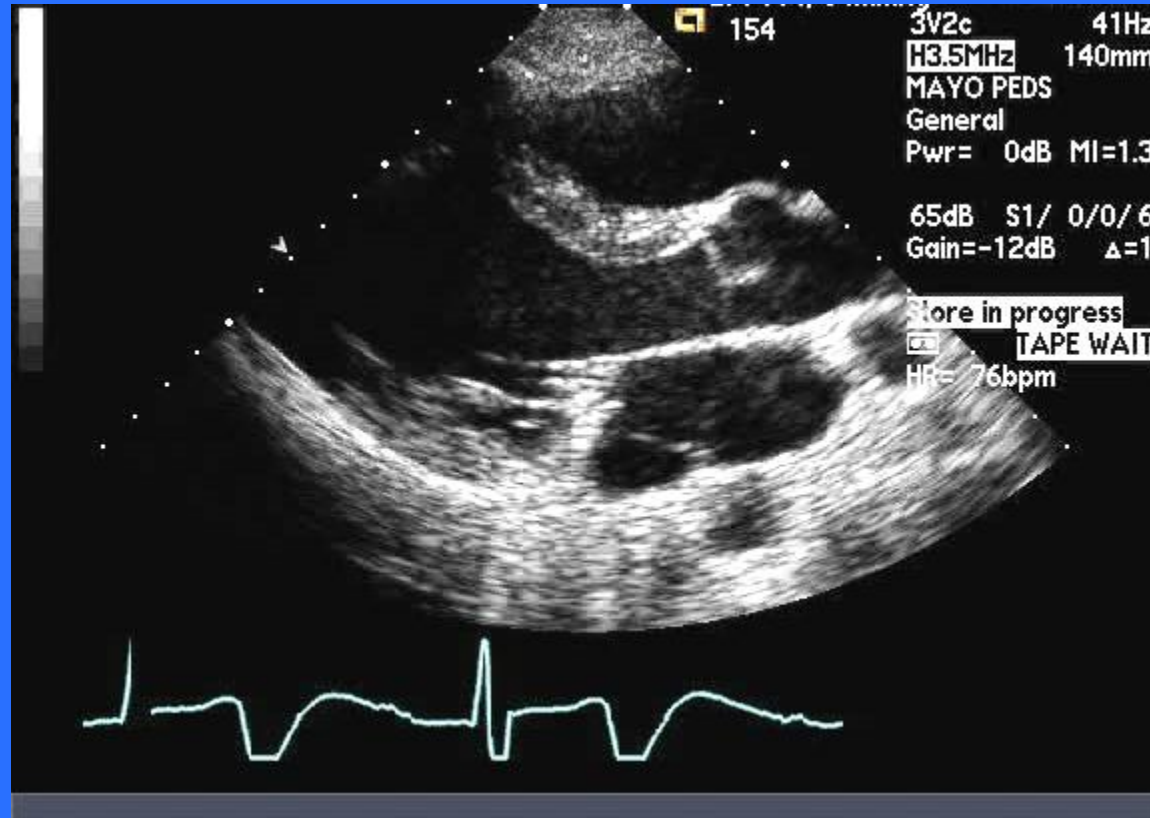


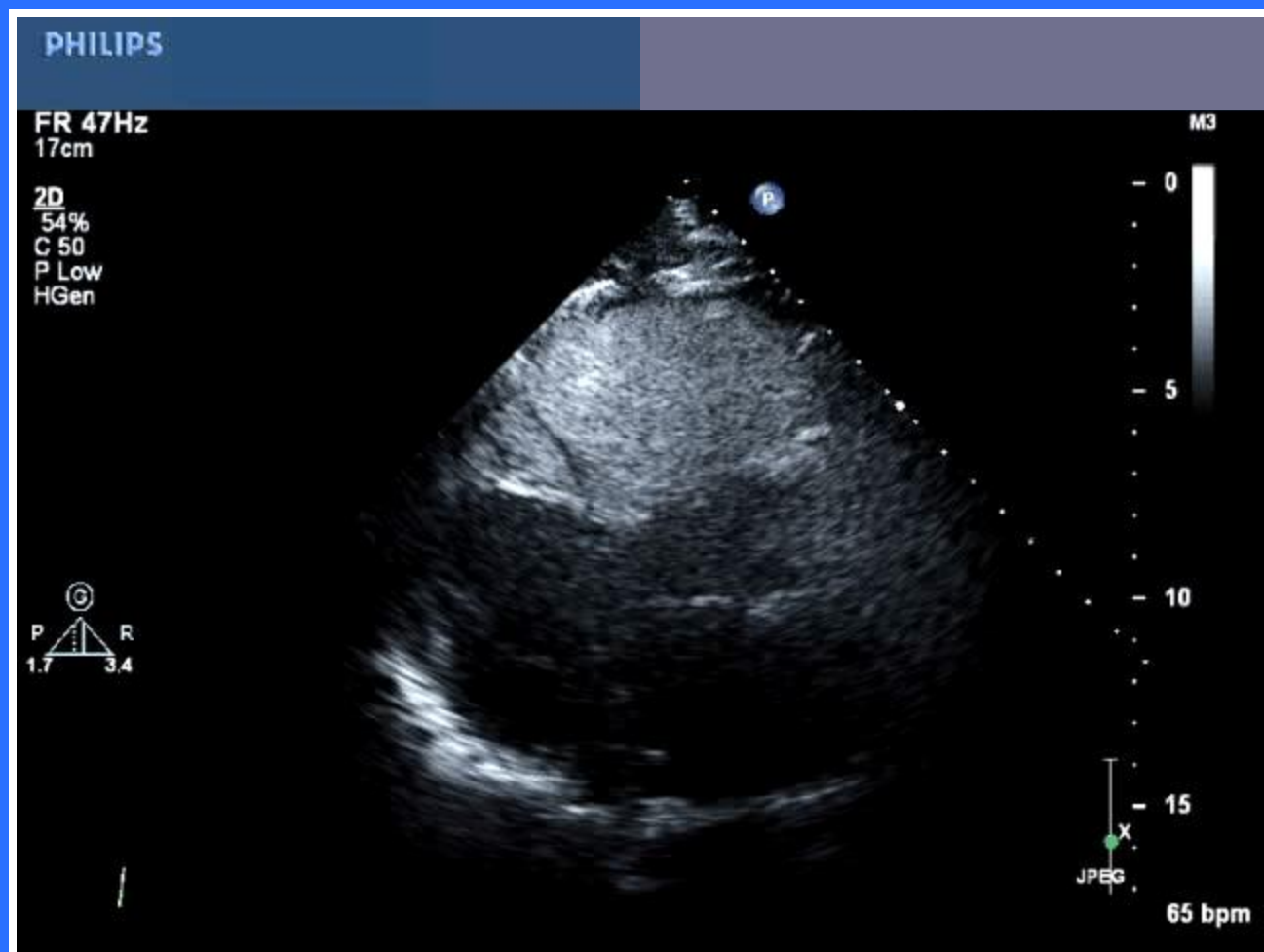


# Coronary Sinus Atrial Septal Defect









# Partial Anomalous Pulmonary Venous Return/Connection

# Variants of Partial Anomalous Pulmonary Venous Connection

- Right pulmonary venous anomalies are most common
- Left pulmonary venous anomalies only comprise 4% of PAPVC
- Scimitar syndrome 3% of PAPVC
- Connections to the CS exceedingly rare
- Bilateral PAPVC occurs, but rare

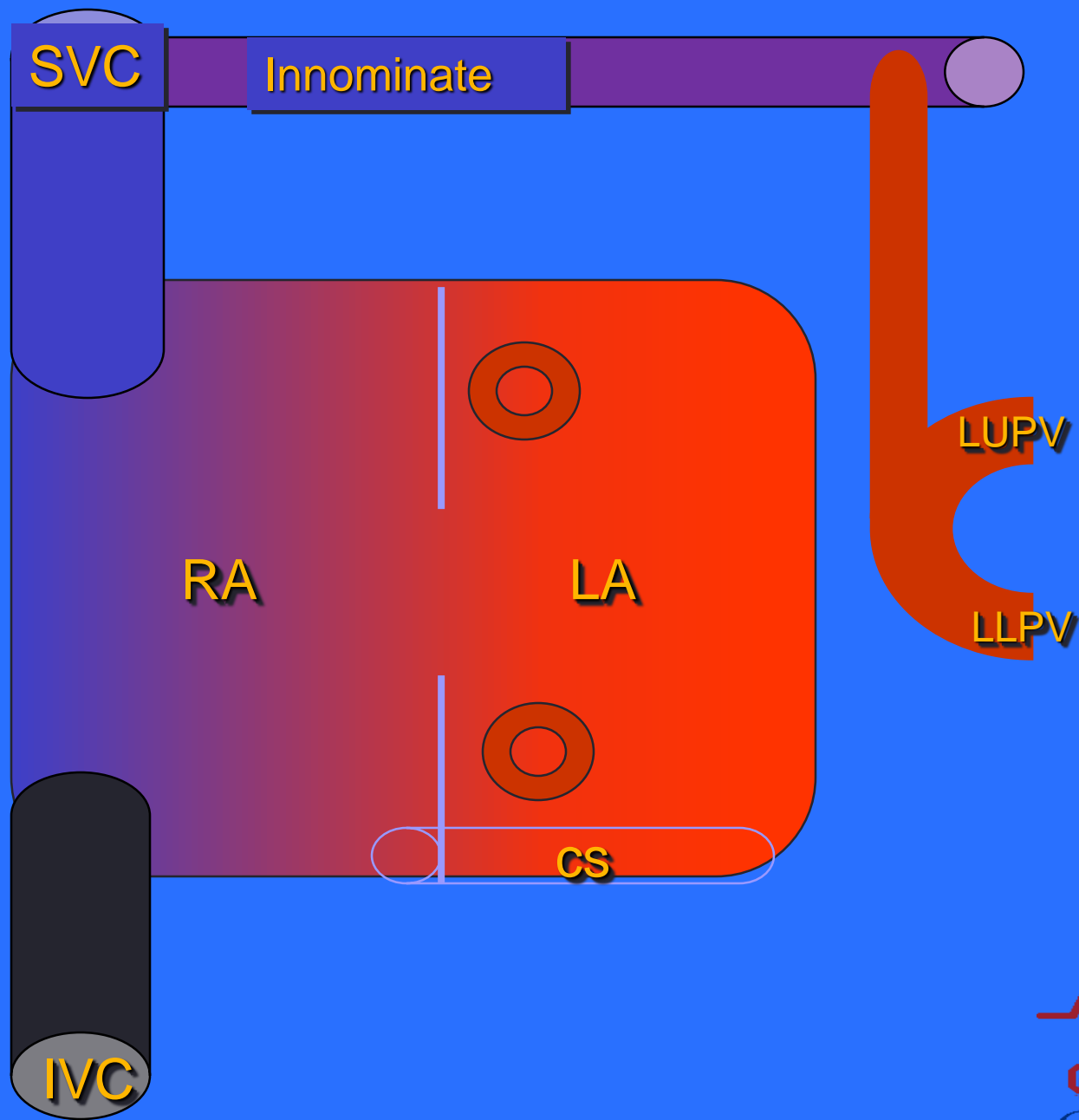
# PAPVC Physiology

- Left to right shunt
- Right chamber volume overload and dilatation
- Single anomalous veins – low risk of hemodynamic compromise
- Less than 50% shunt – rare to have symptoms in childhood

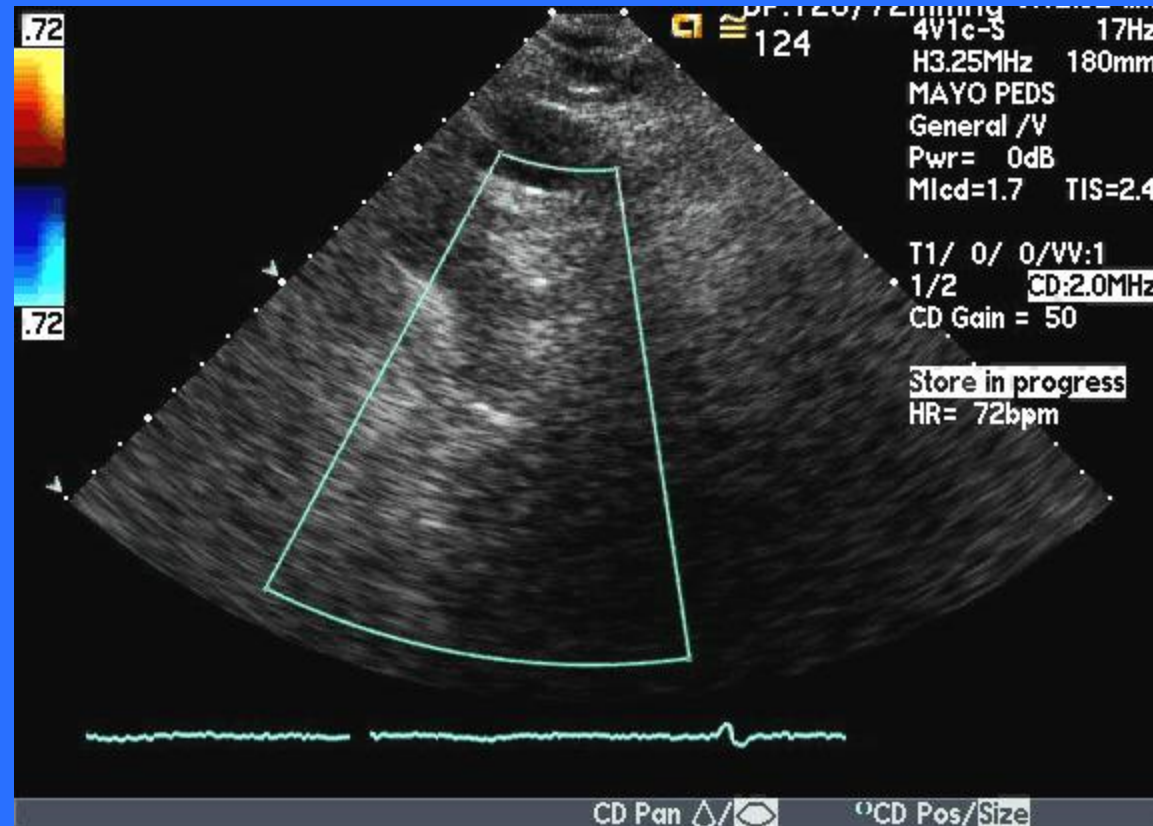
## **ECHO Evaluation of PAPVC**

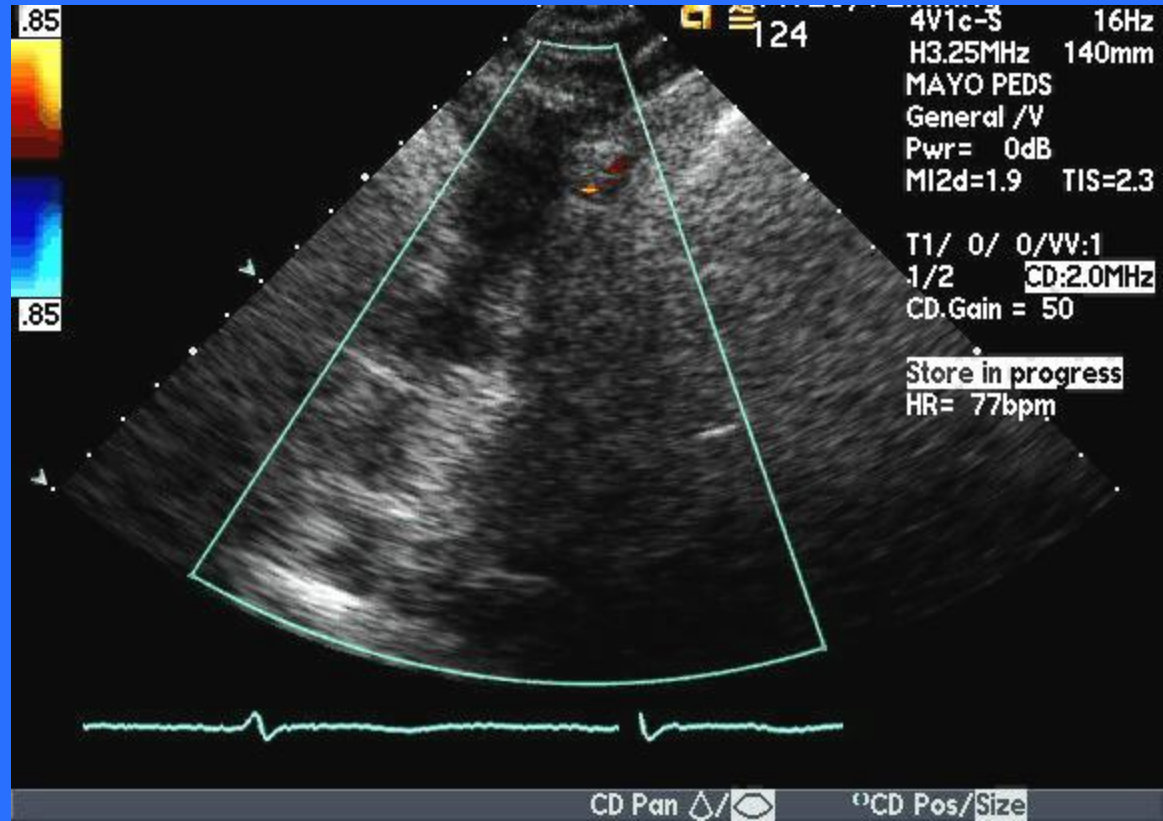
- **Type of connection**
- **Associated anomalies**
- **Right chamber size**
- **Right ventricular function**
- **Pulmonary artery pressure**

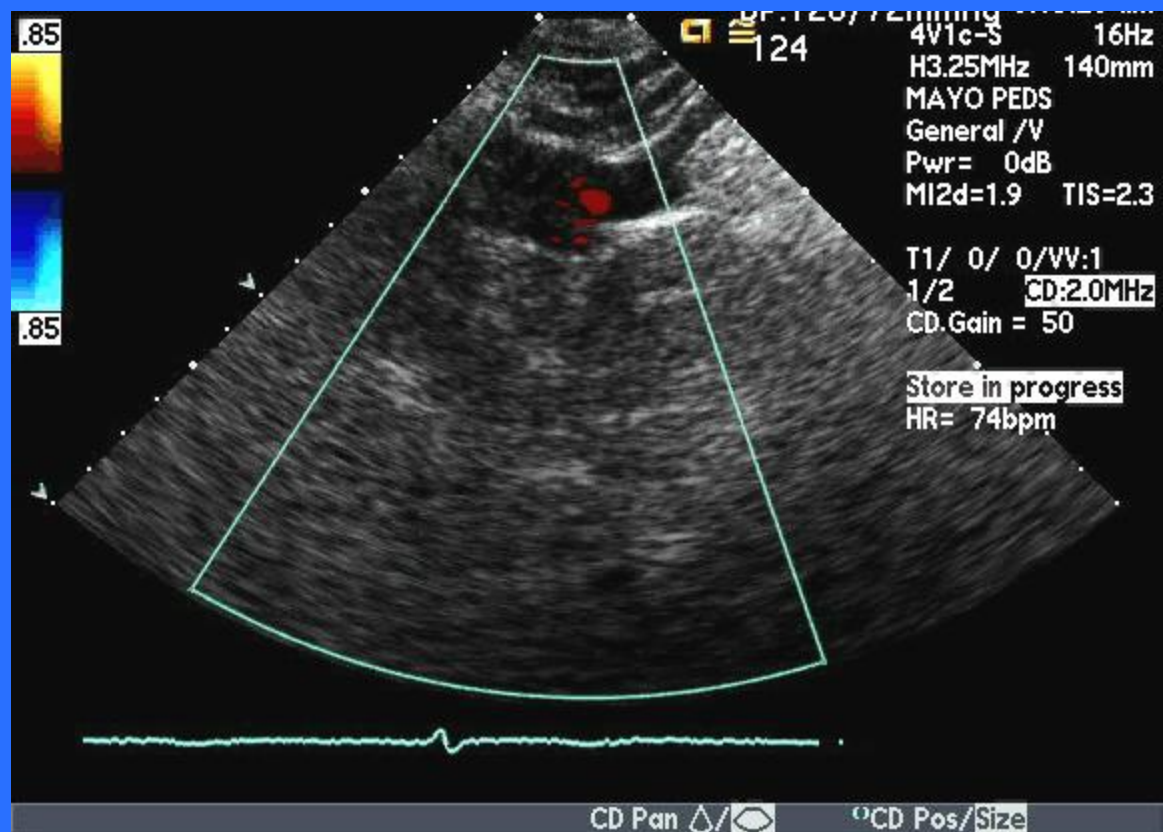


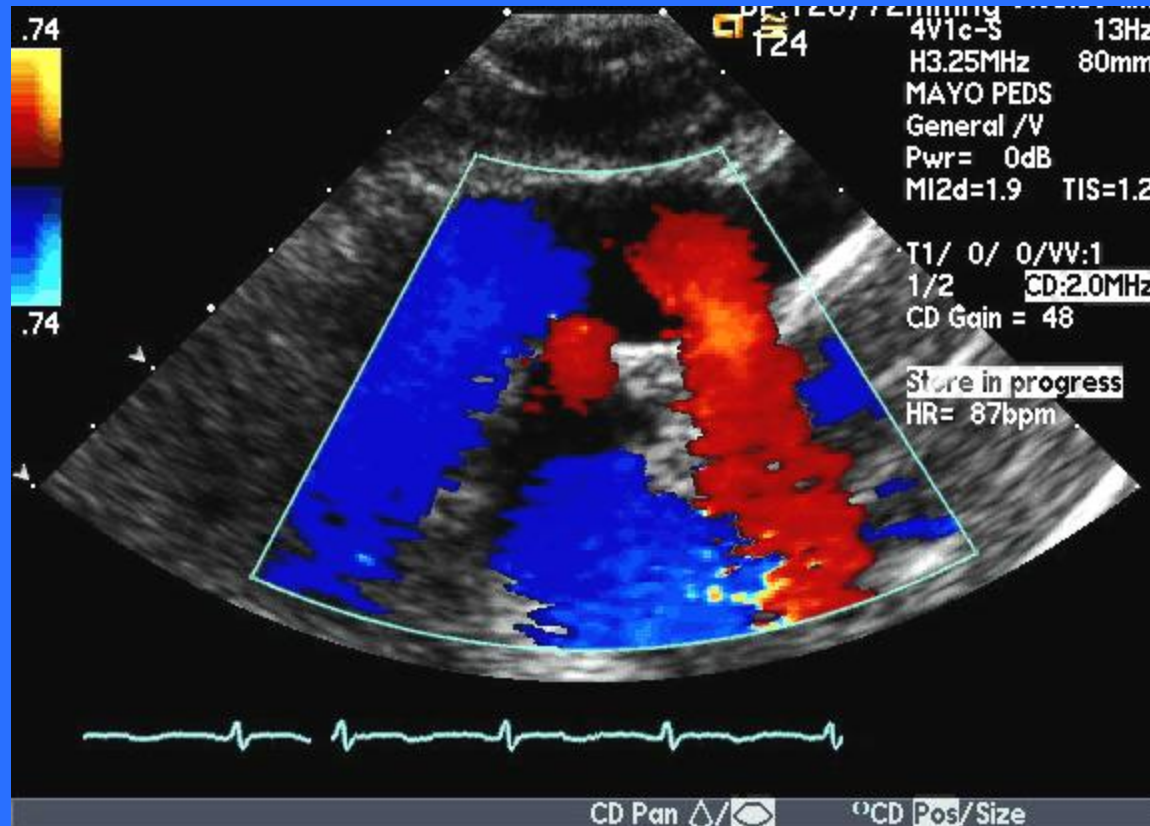


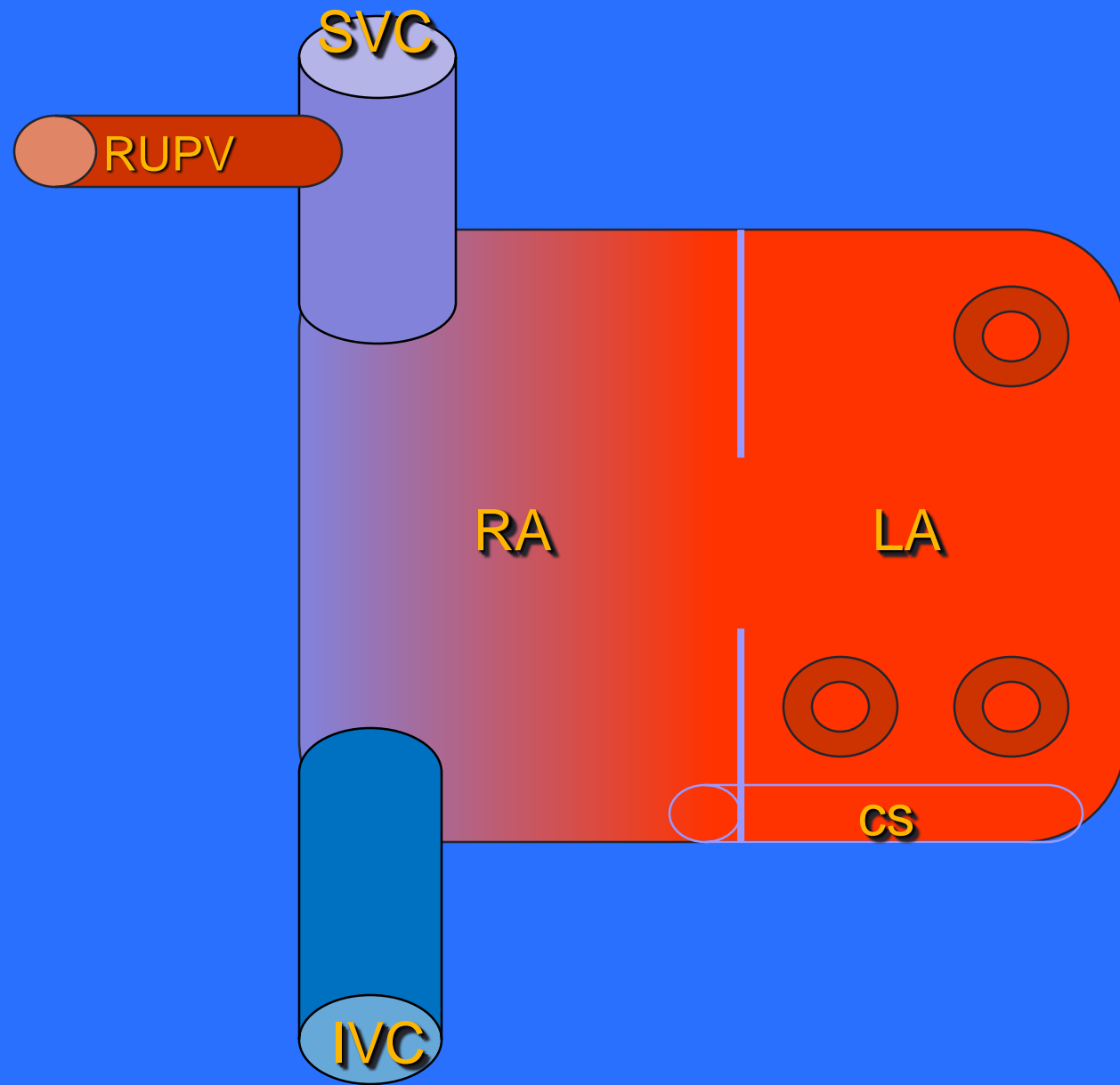
# Vertical Vein



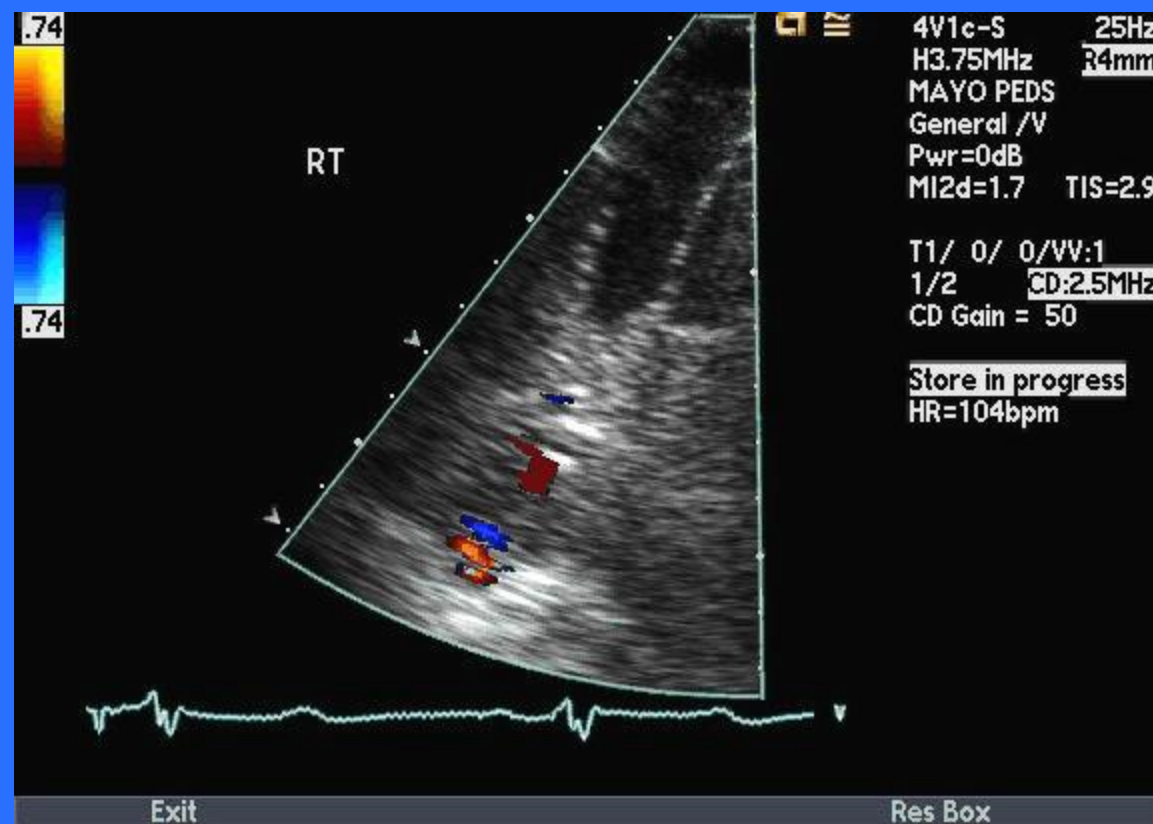


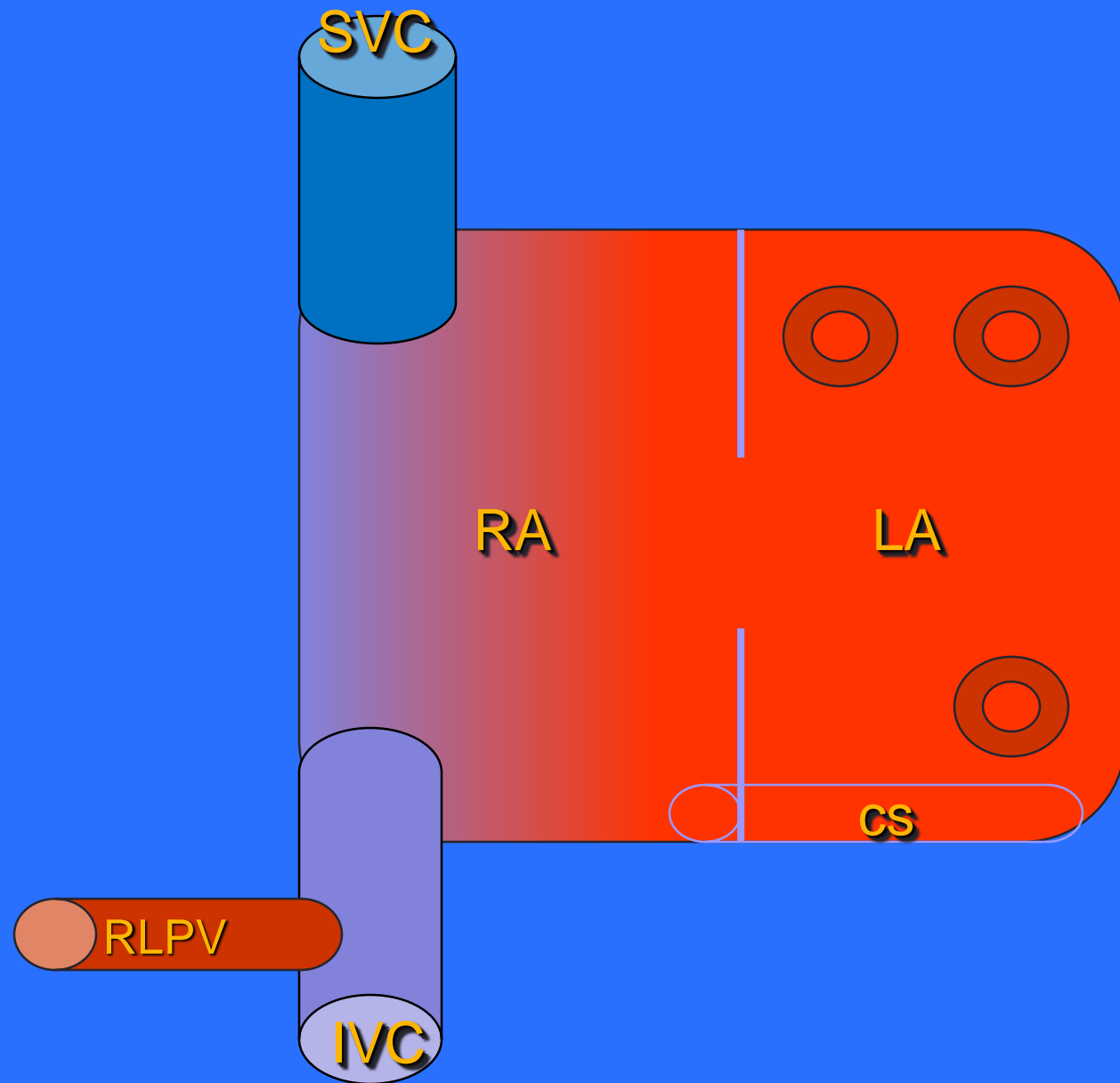






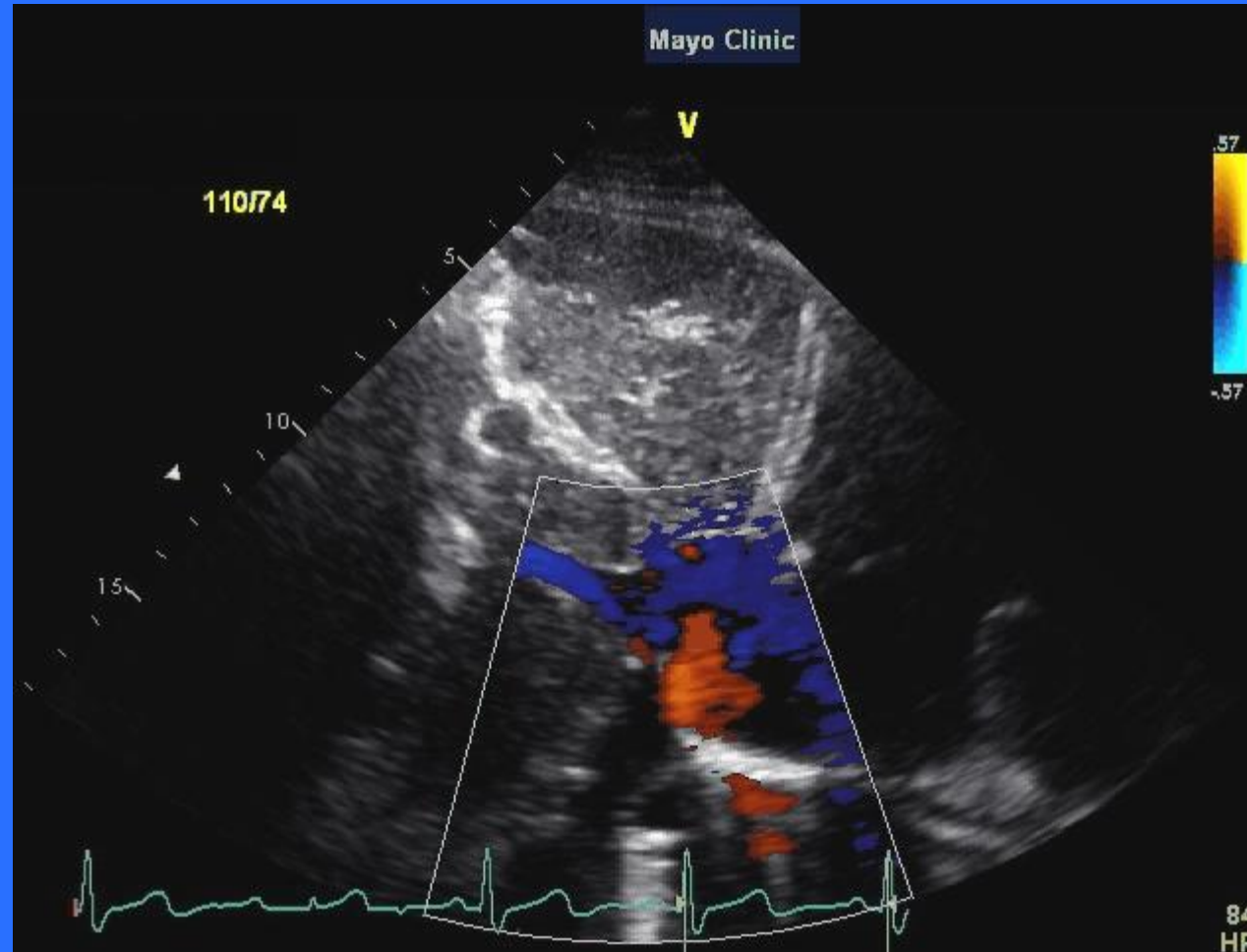
# Right Pulmonary Vein to SVC



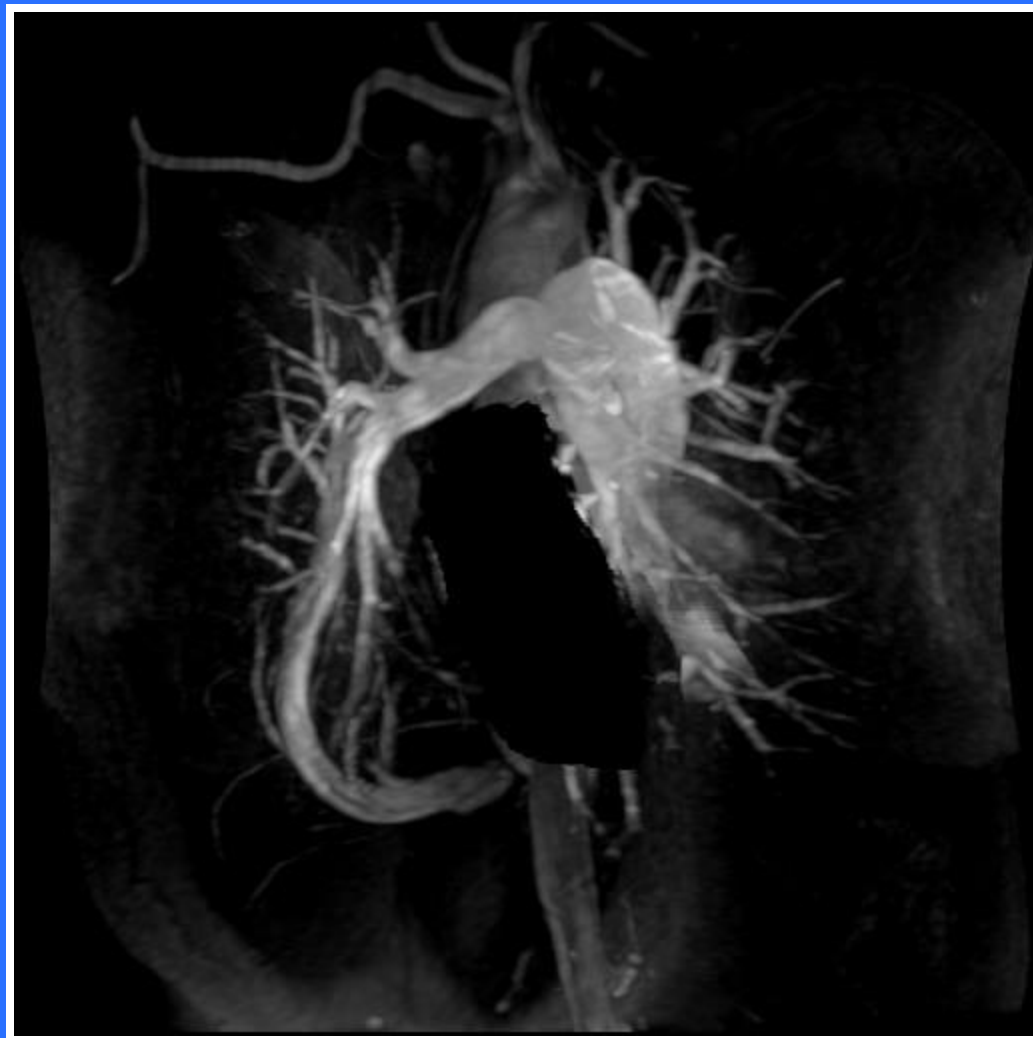




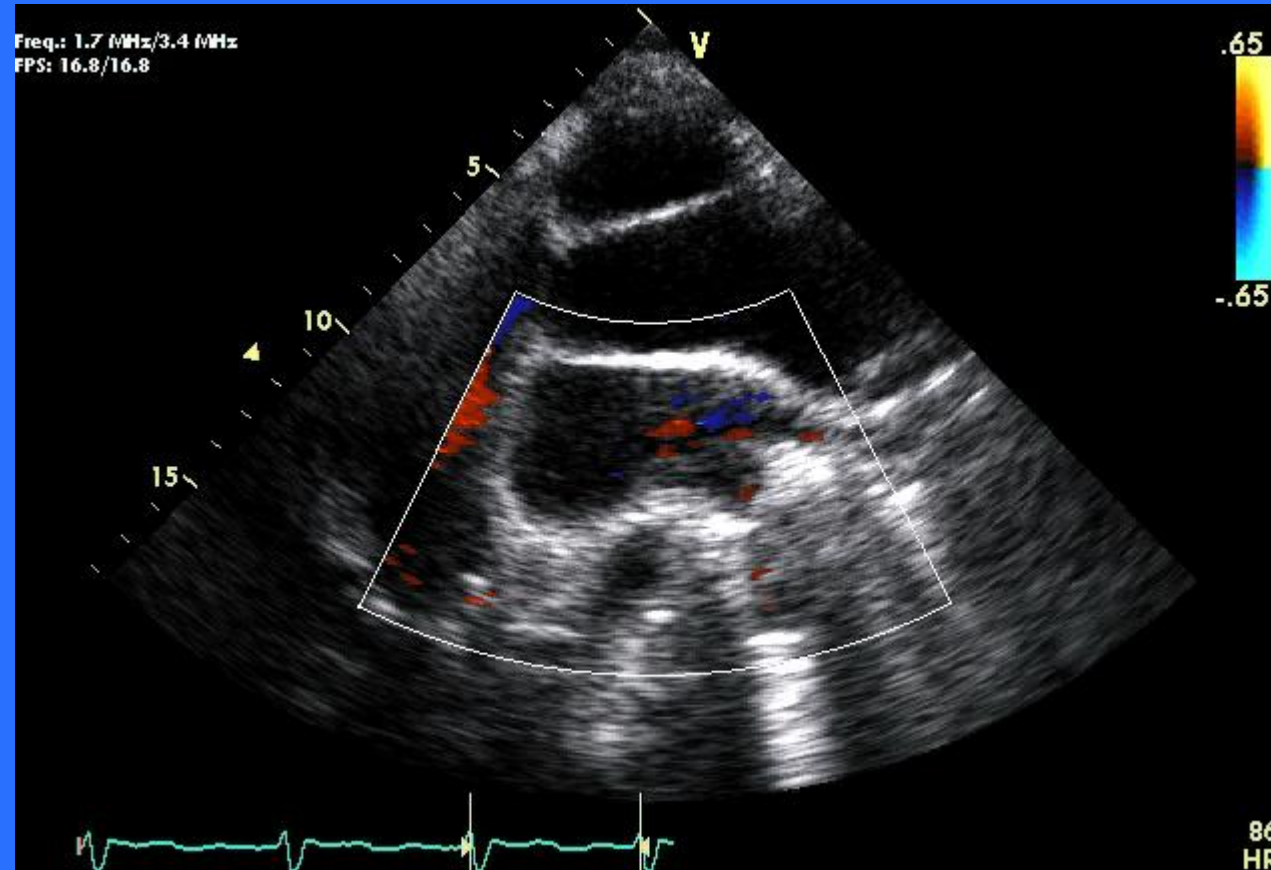
# Scimitar Syndrome



# MRA

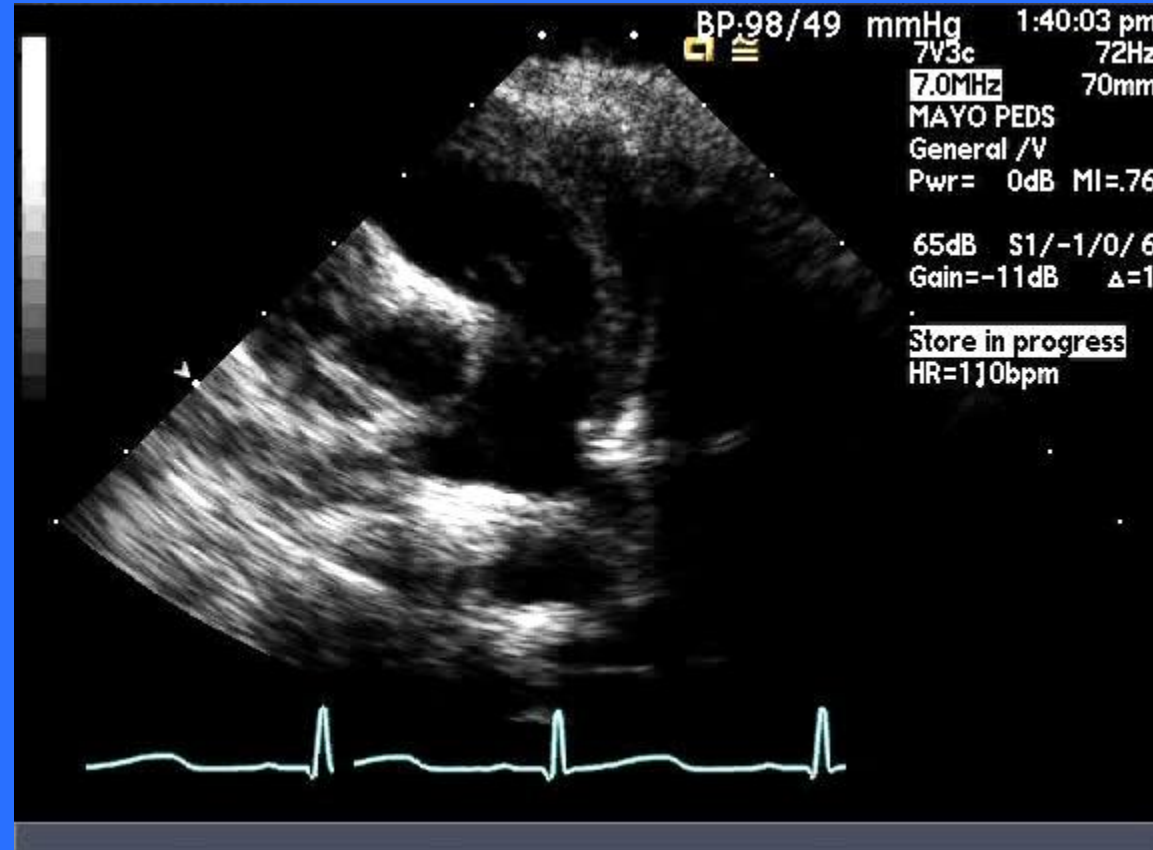


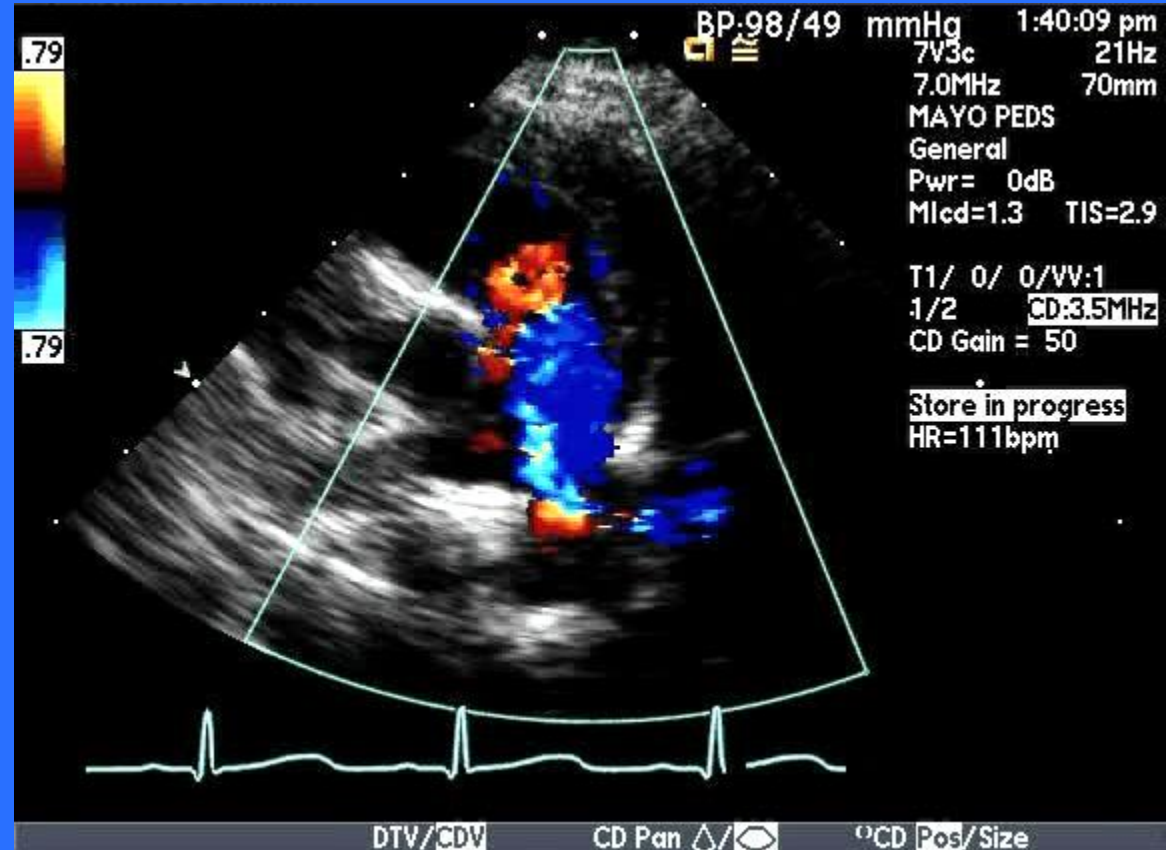
# Suprasternal Notch Coronal View (“Crab”)

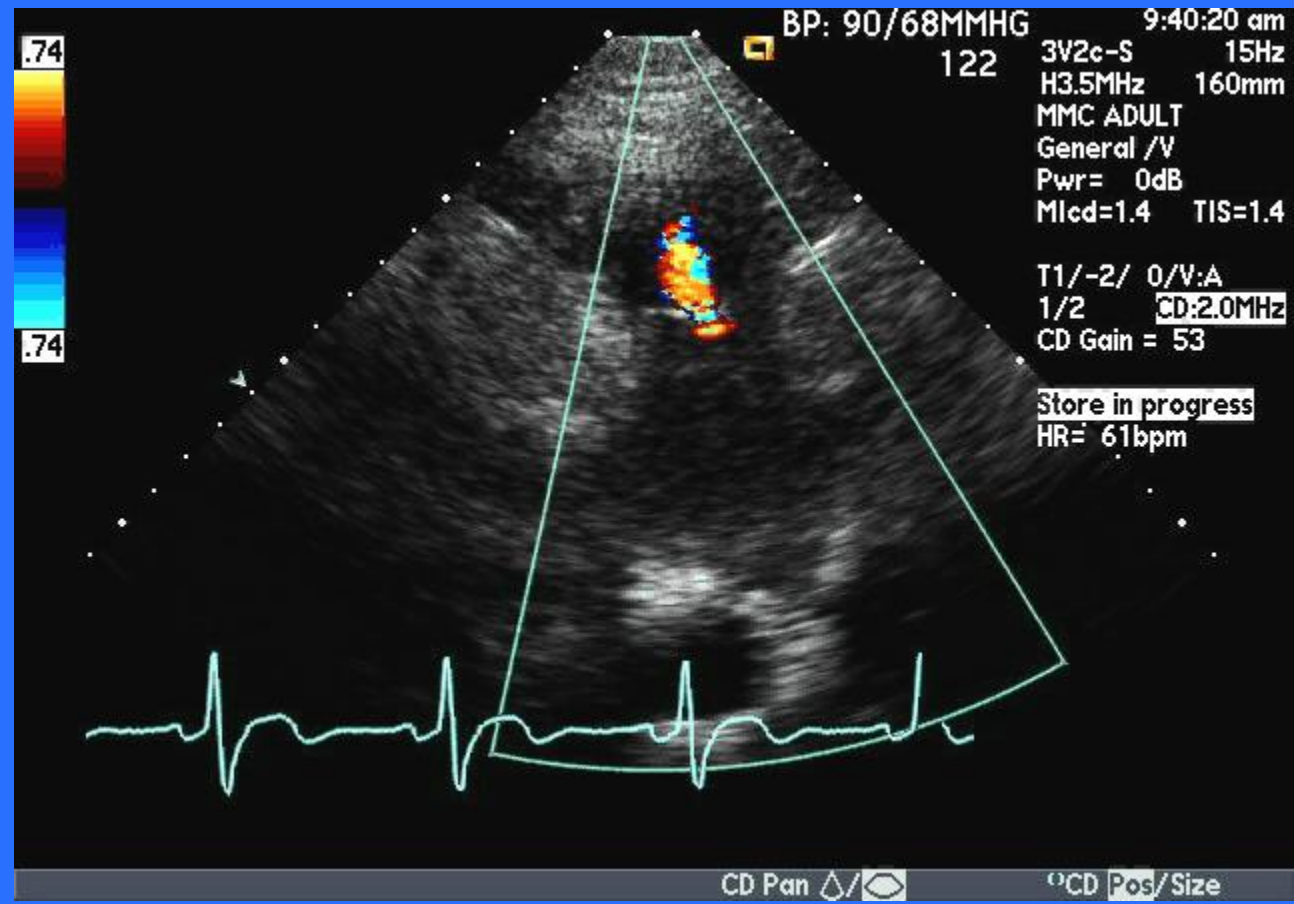


# Patent Ductus Arteriosus

- Left Heart Enlargement
- Pulmonary hypertension common if the PDA is large – may not see a shunt on echo (equal pressures)







# Ventricular Septal Defects



# Ventricular Septal Anatomy

➤ **Membranous**

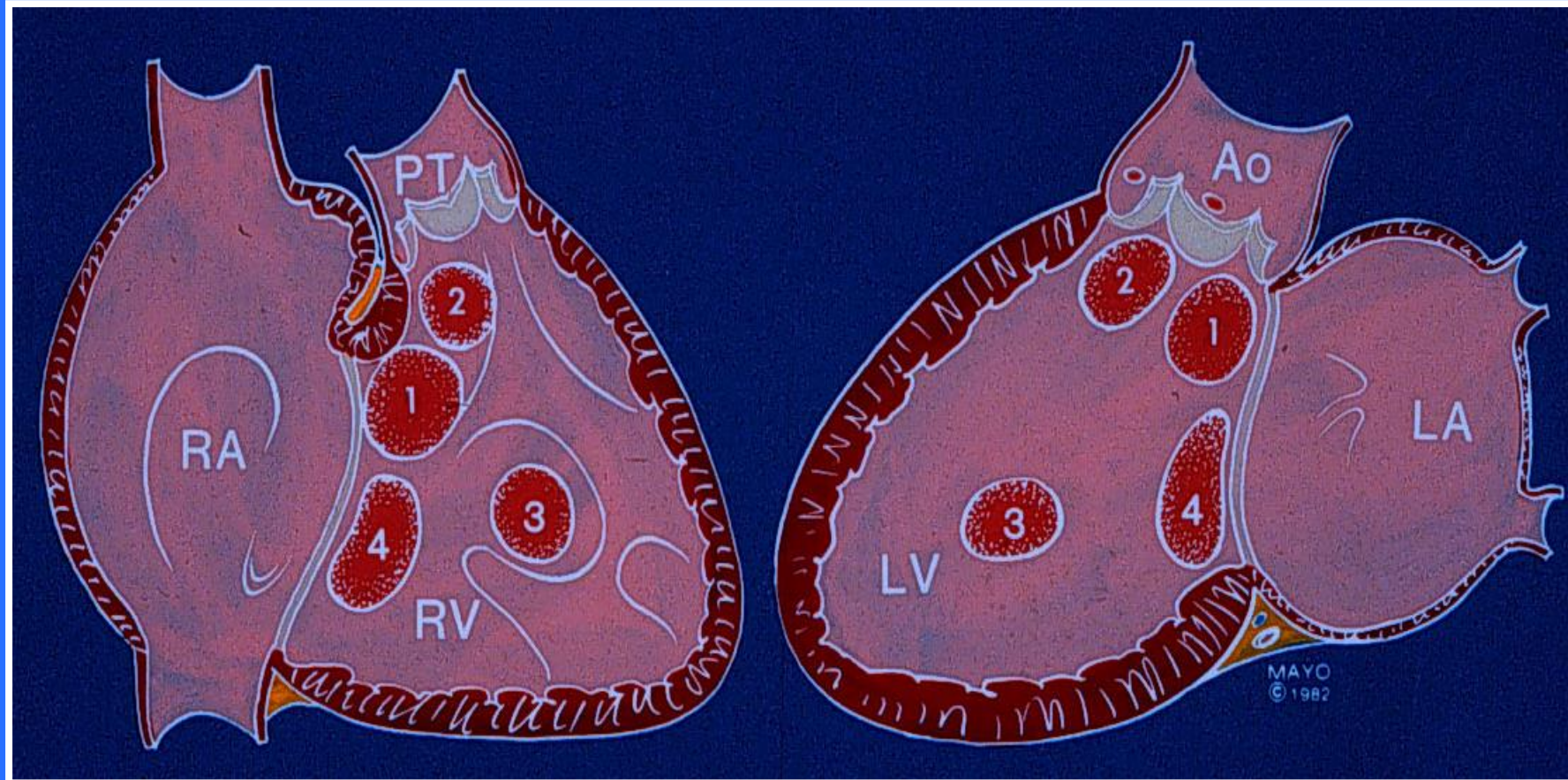
➤ **Muscular**

- **Inlet: Separates ventricular inflow**
- **Trabecular**
- **Outlet: Separates outflow tracts**

# Ventricular Septal Defects

- Membranous (80%)
- Muscular (trabecular septum)
- Inlet
- Outlet
  - Infundibular
  - Supracristal/Subarterial (5%)
- Post-MI

# VSD Anatomy

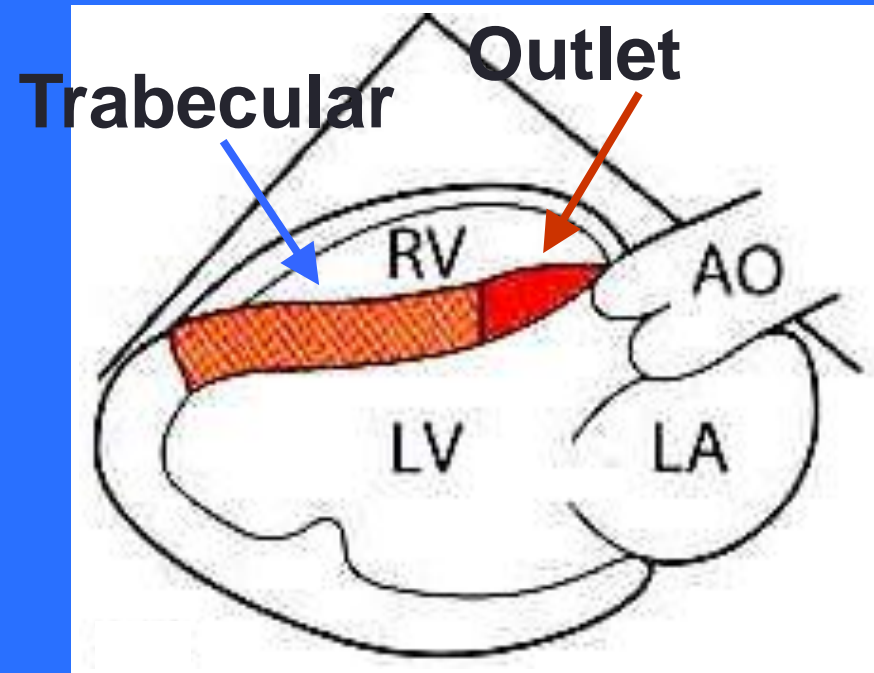


# Echo Evaluation of VSDs

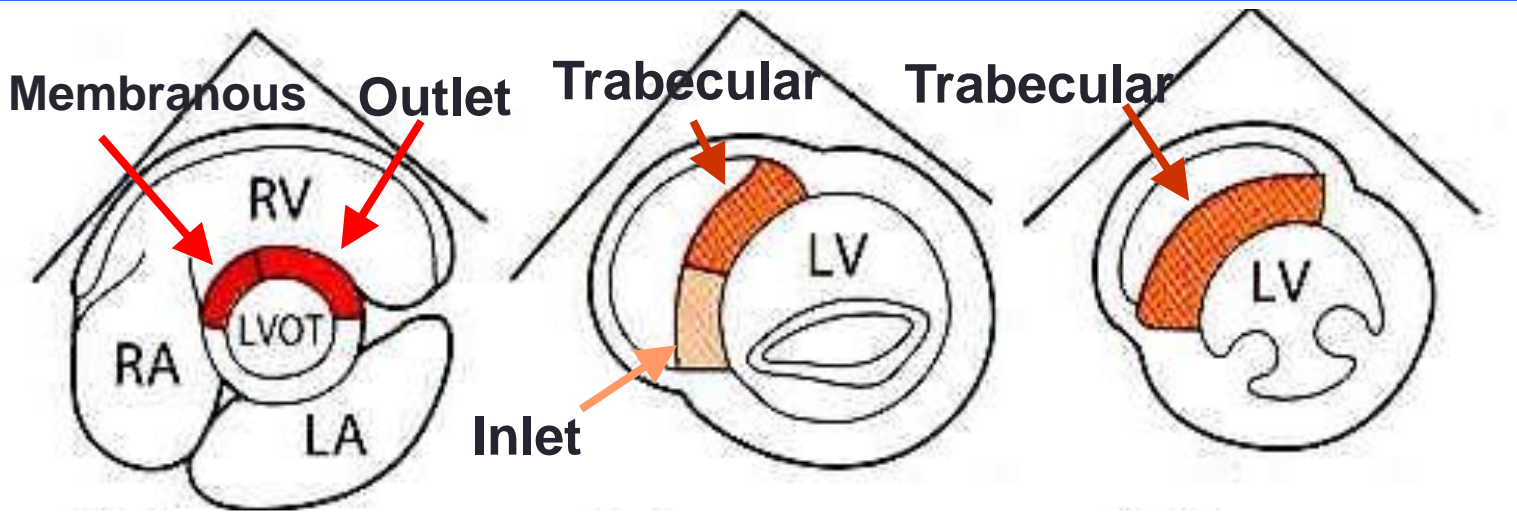
- Location
- Size
- Involvement of other structures
- **Left** ventricular and **left** atrial size
- Estimated right ventricular systolic pressure
- Associated anomalies

# Location by Echocardiogram

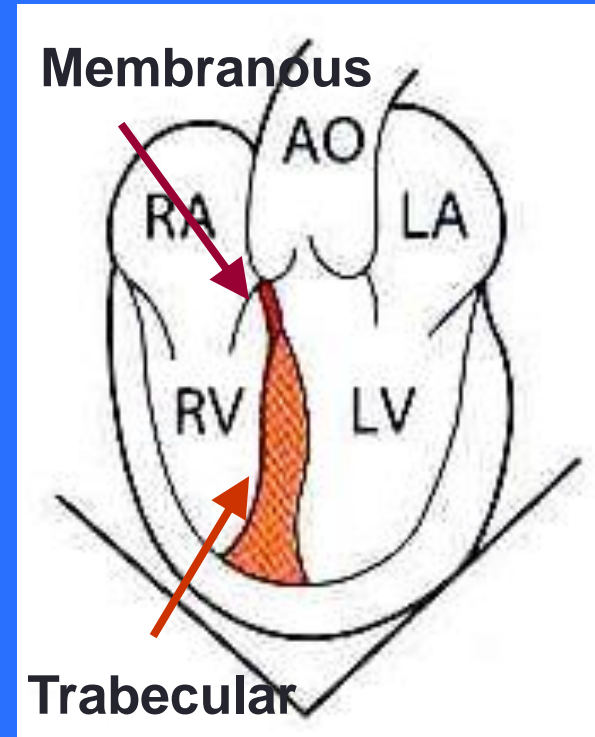
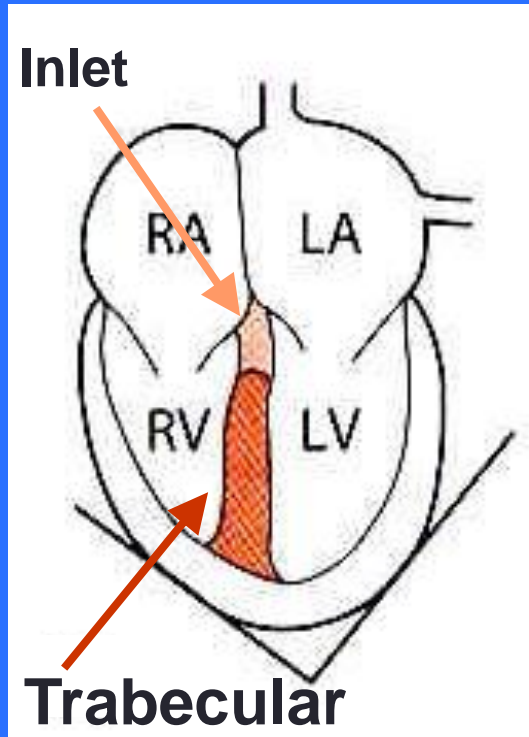
# Parasternal Long Axis



# Parasternal Short Axis

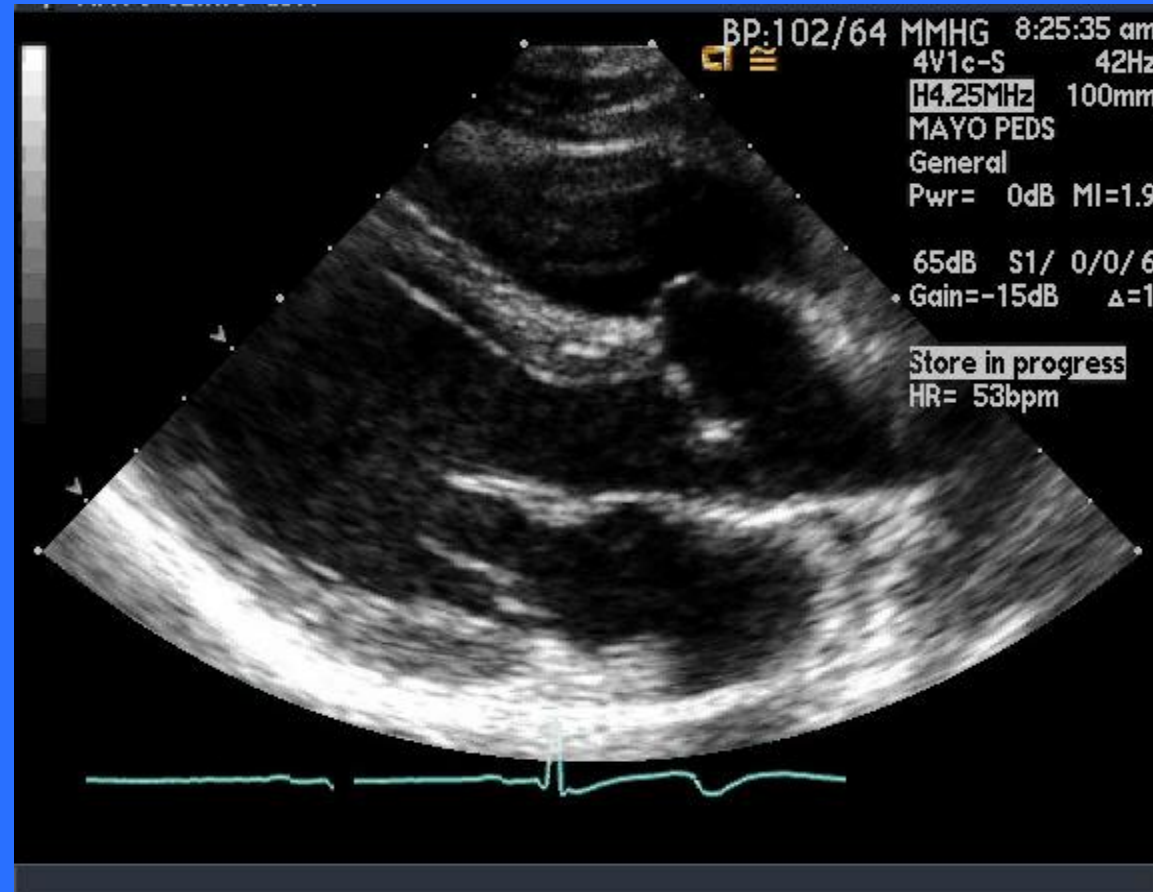


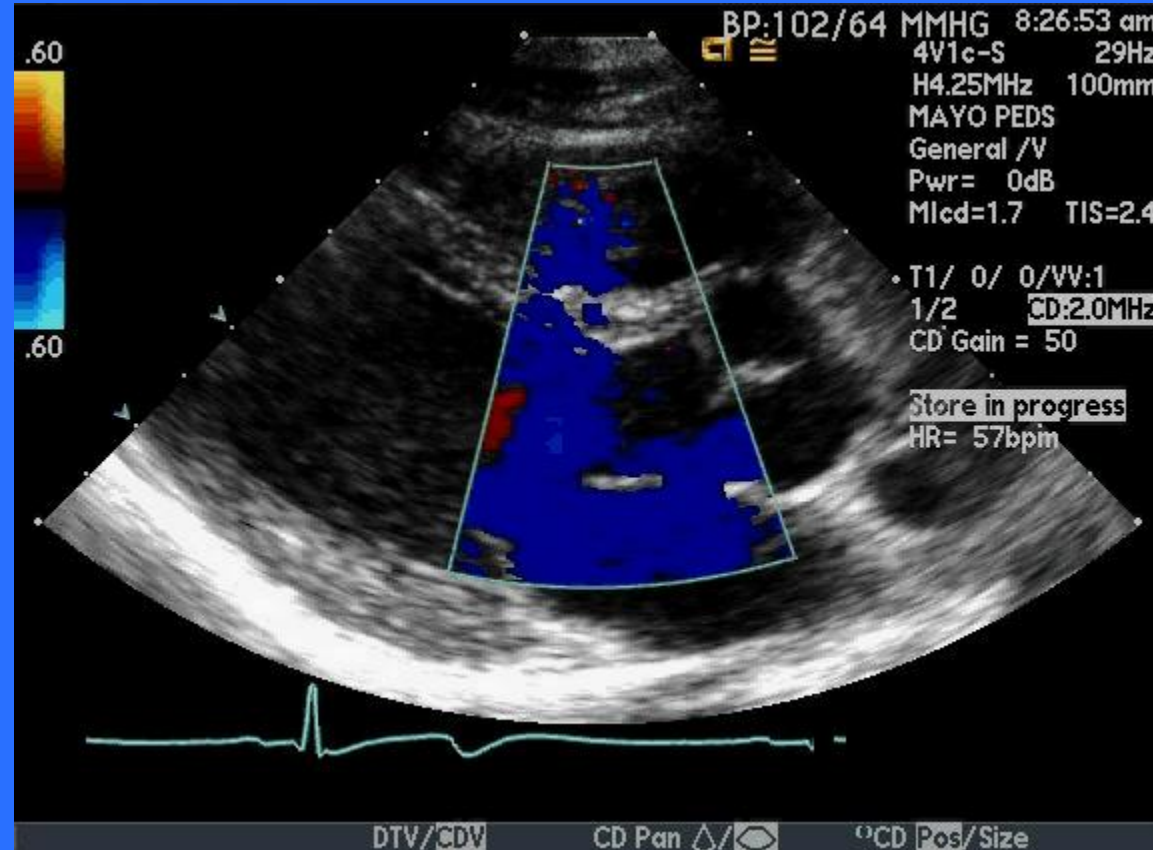
# Apical

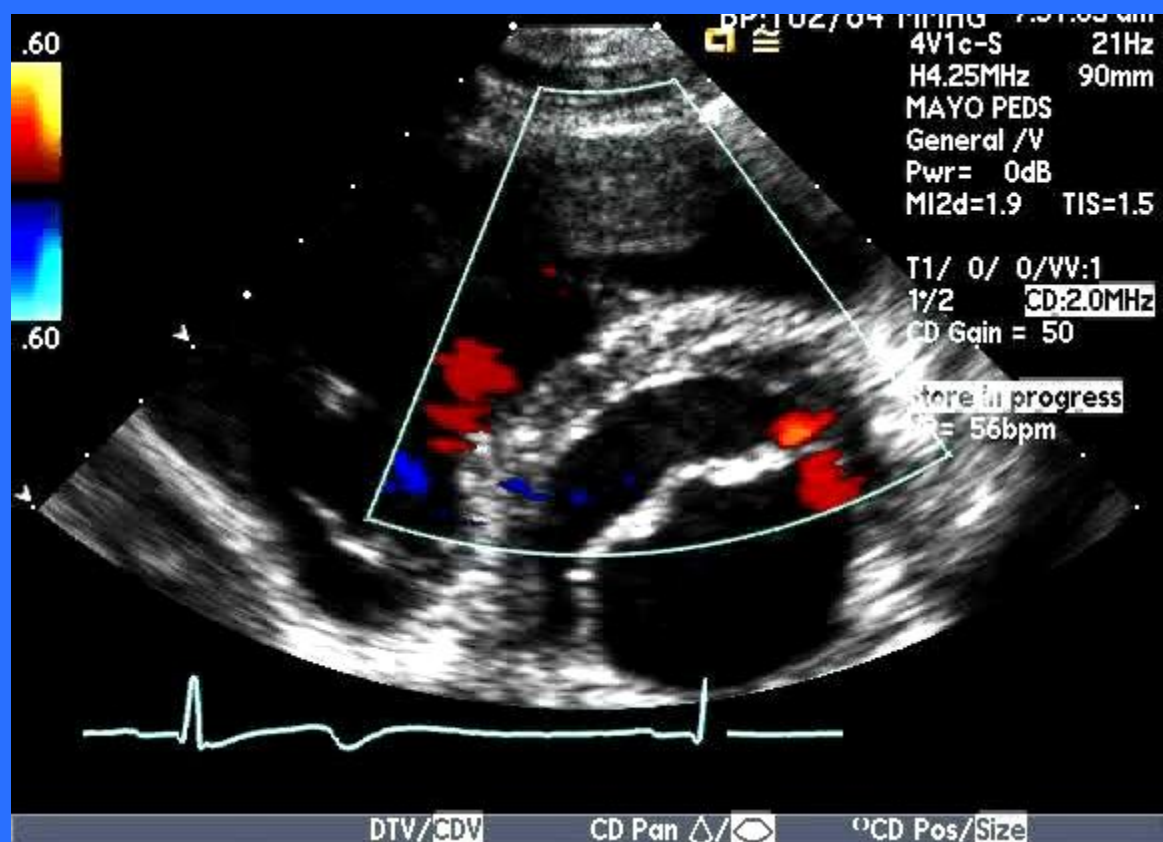




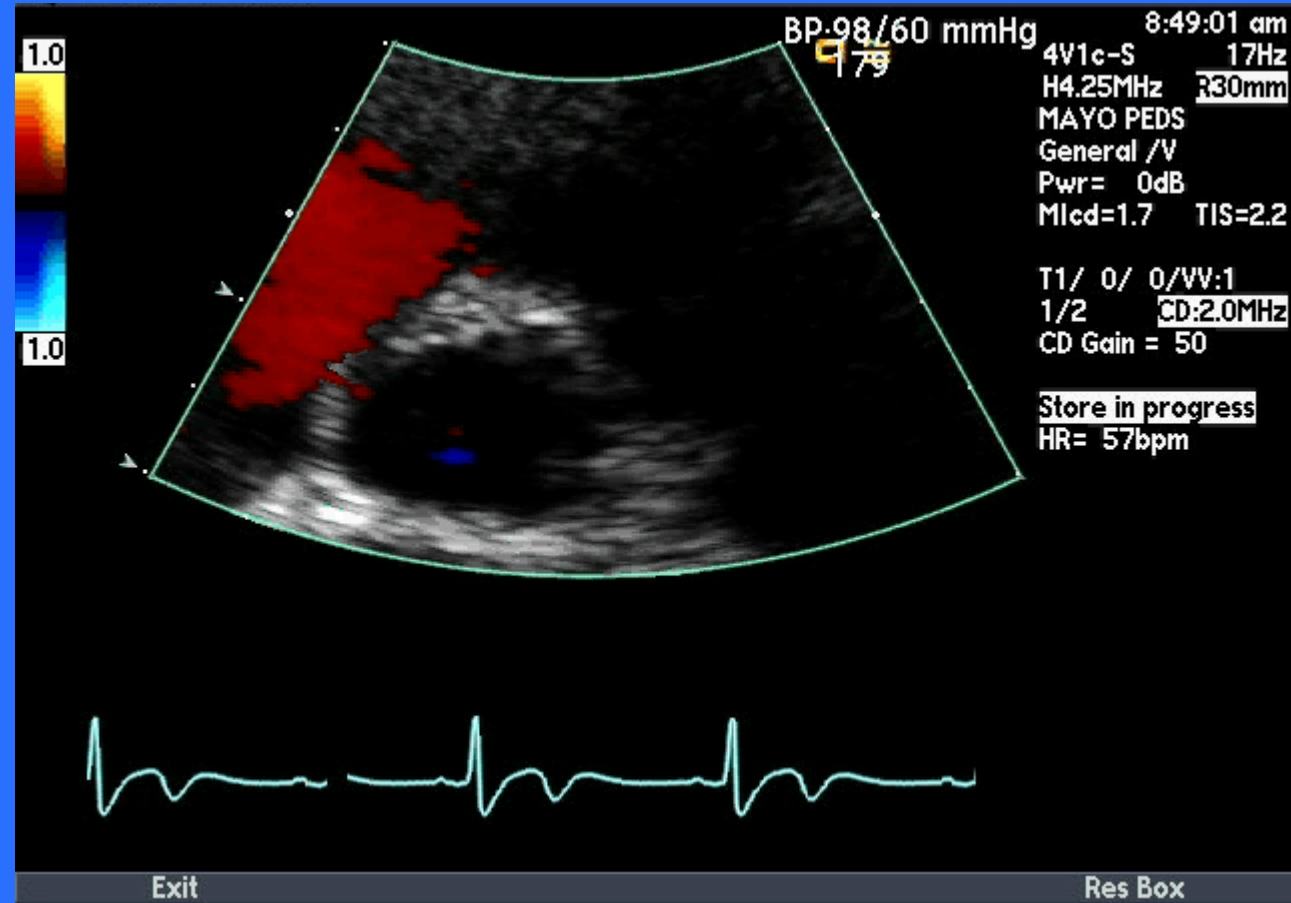
# Outlet/Infundibular Septal Defect







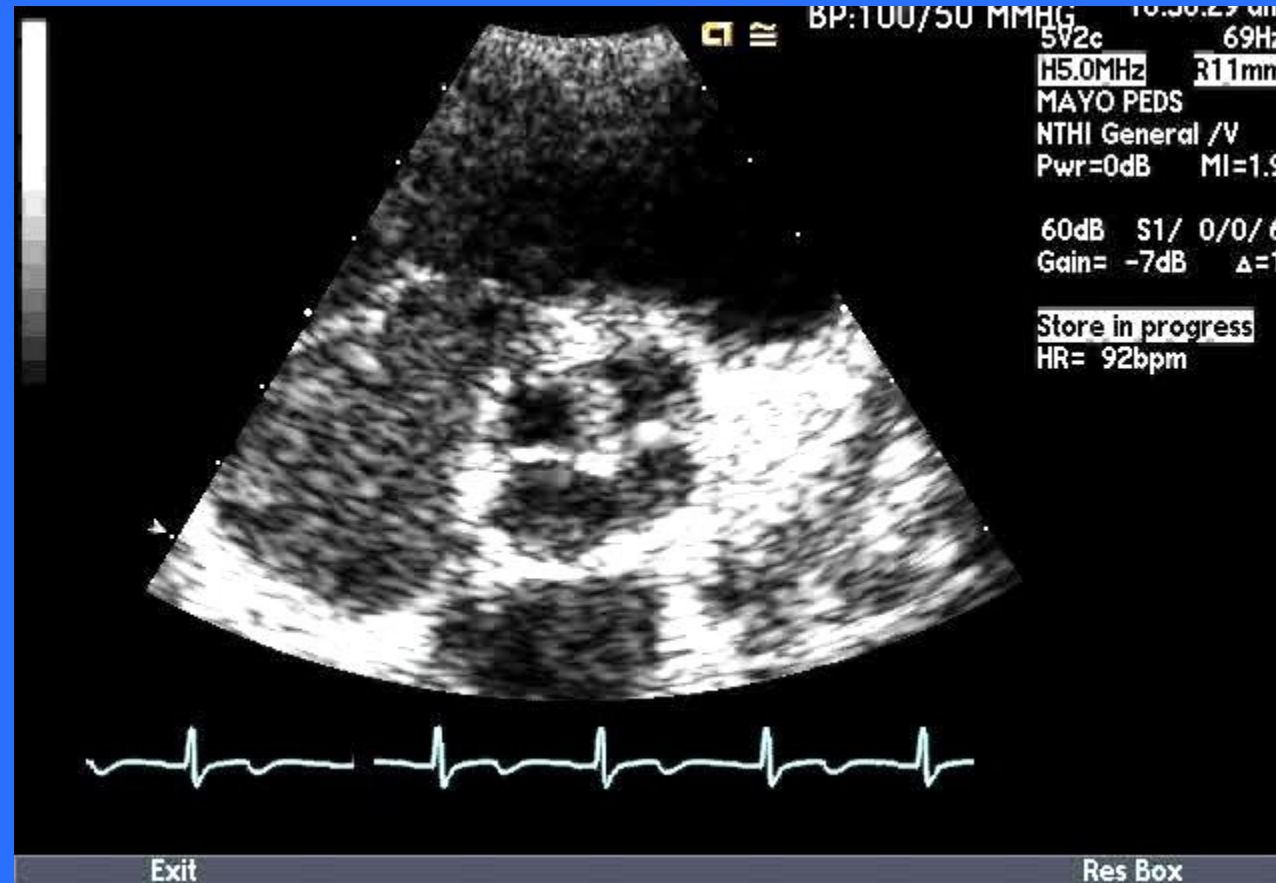
# Supracristal VSD

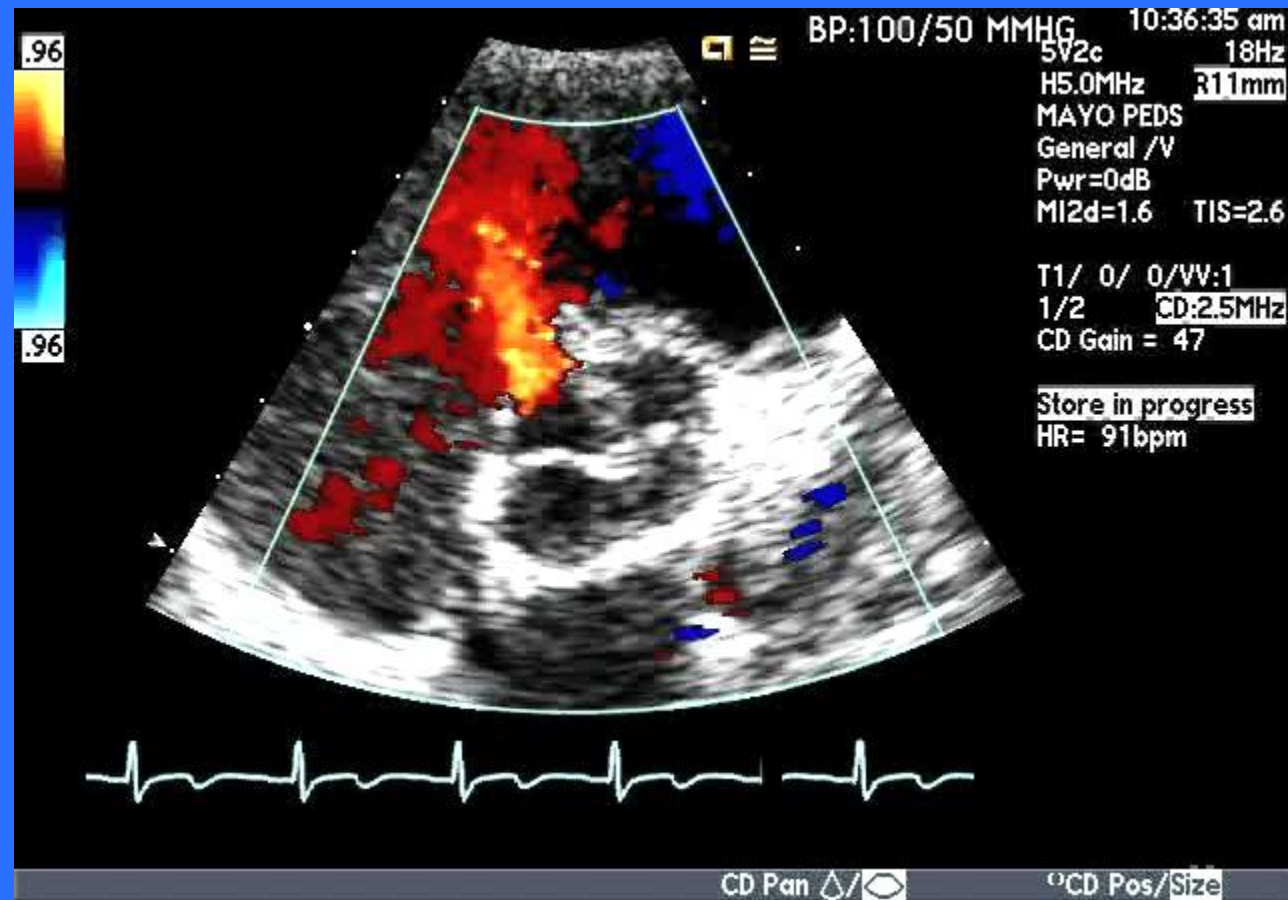


# Don't Get Confused!

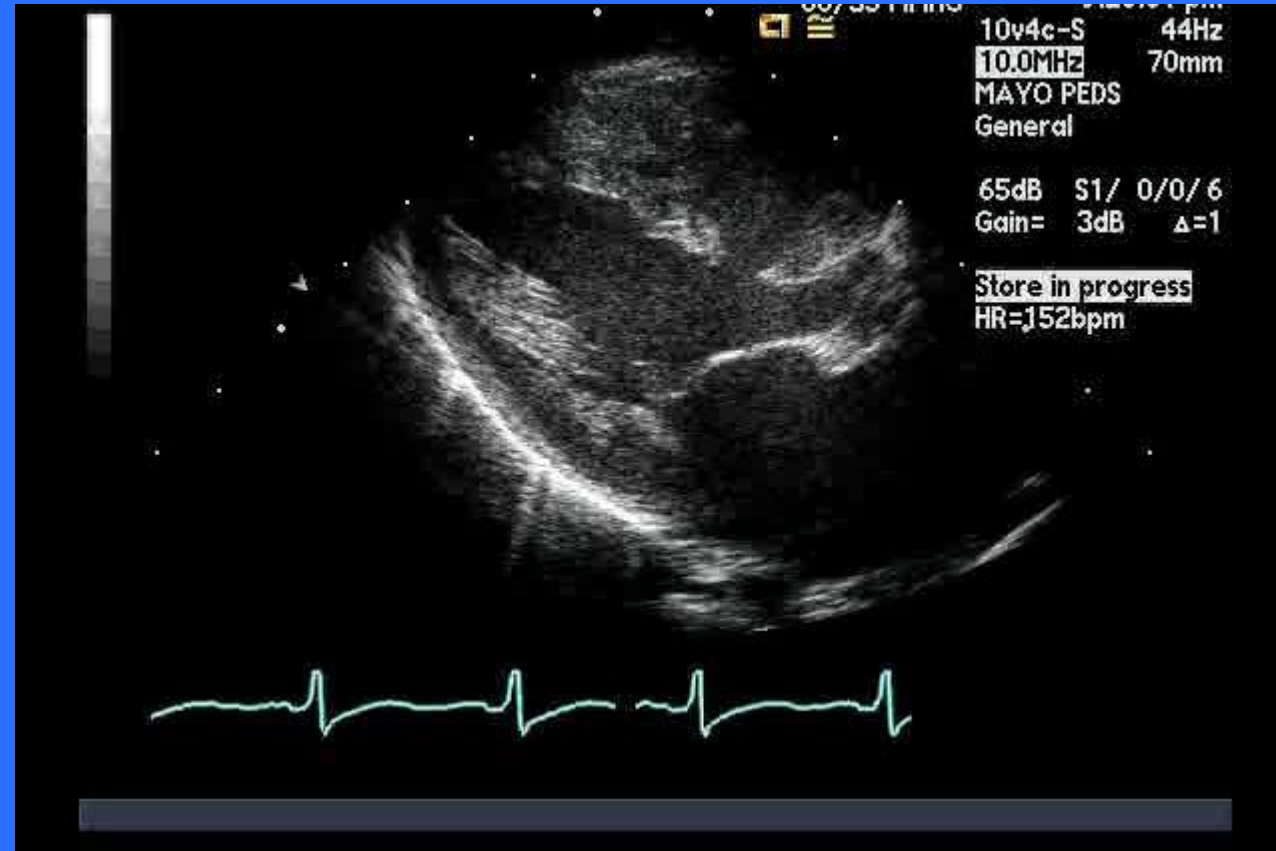


# Perimembranous Defect

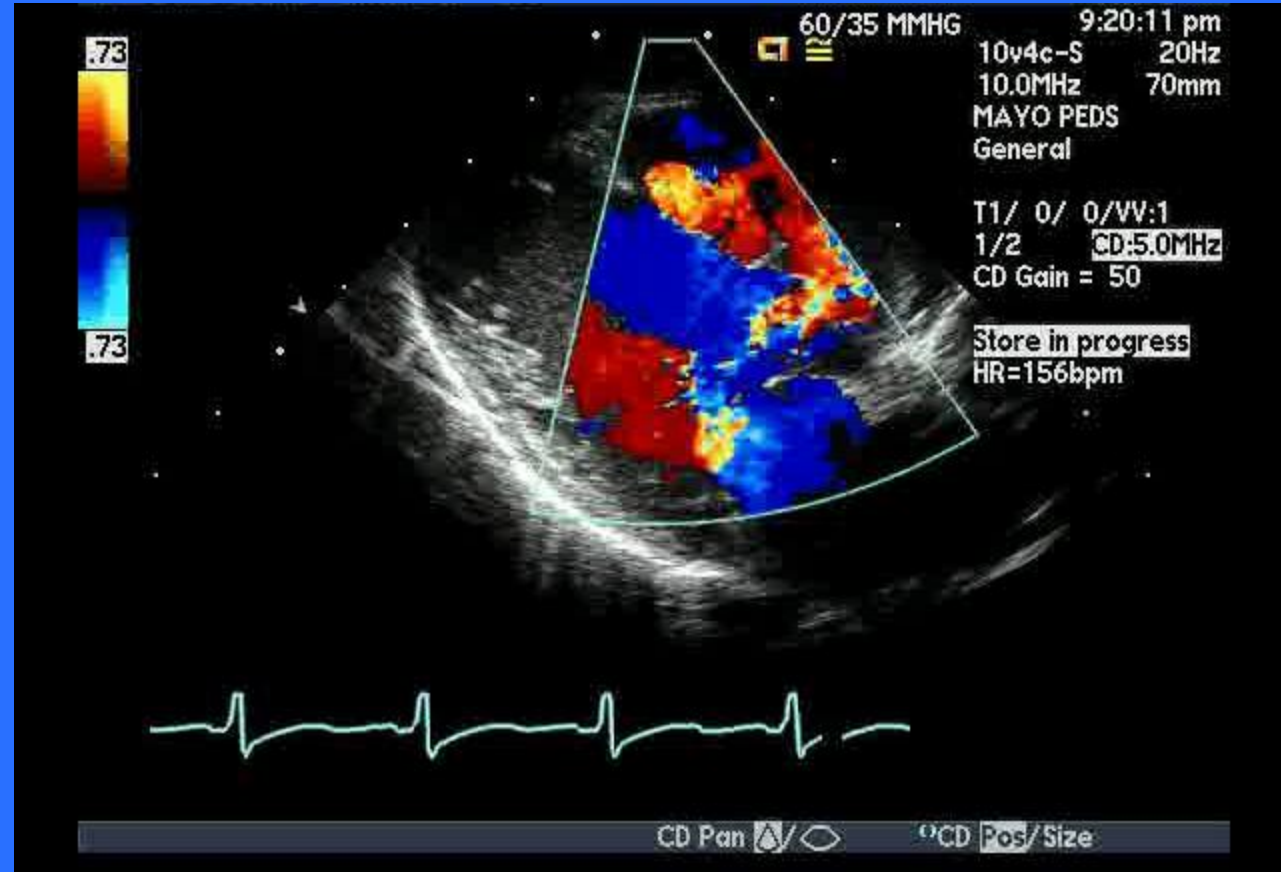




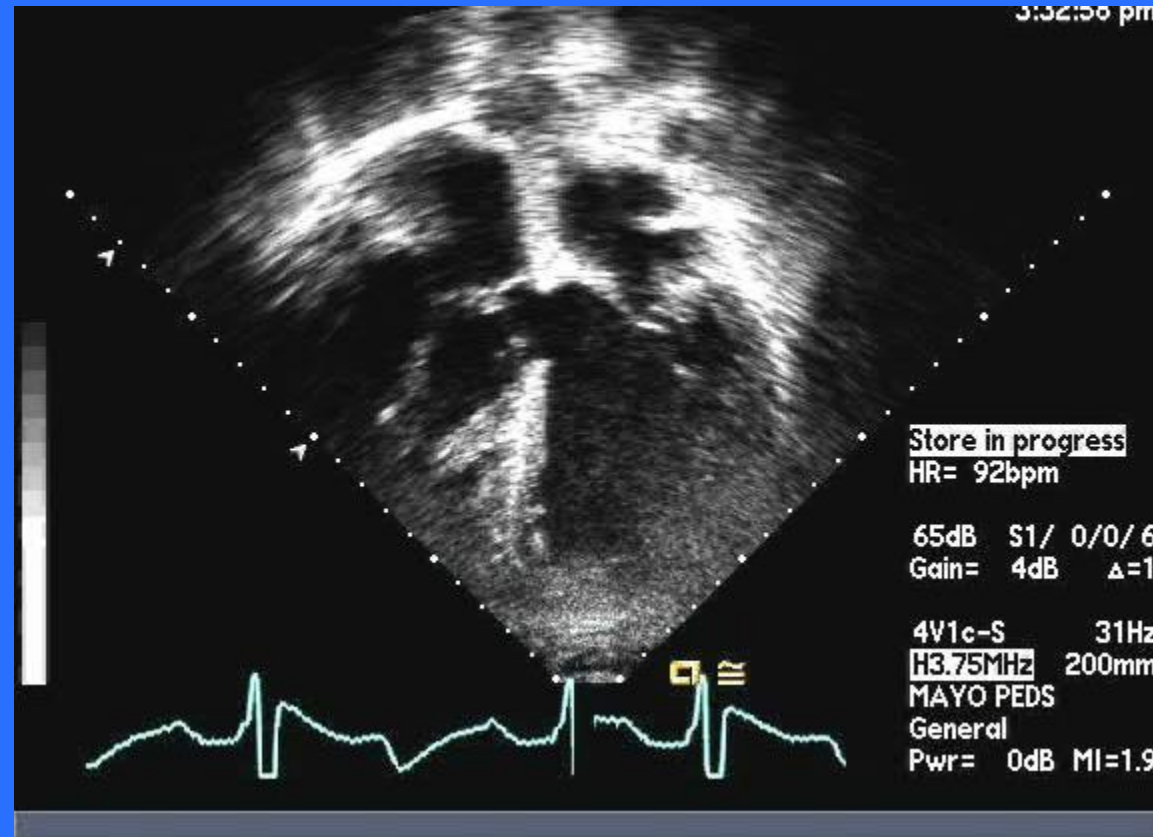
# Trabecular (Muscular) Defect







# Inlet VSD



## VSD Size

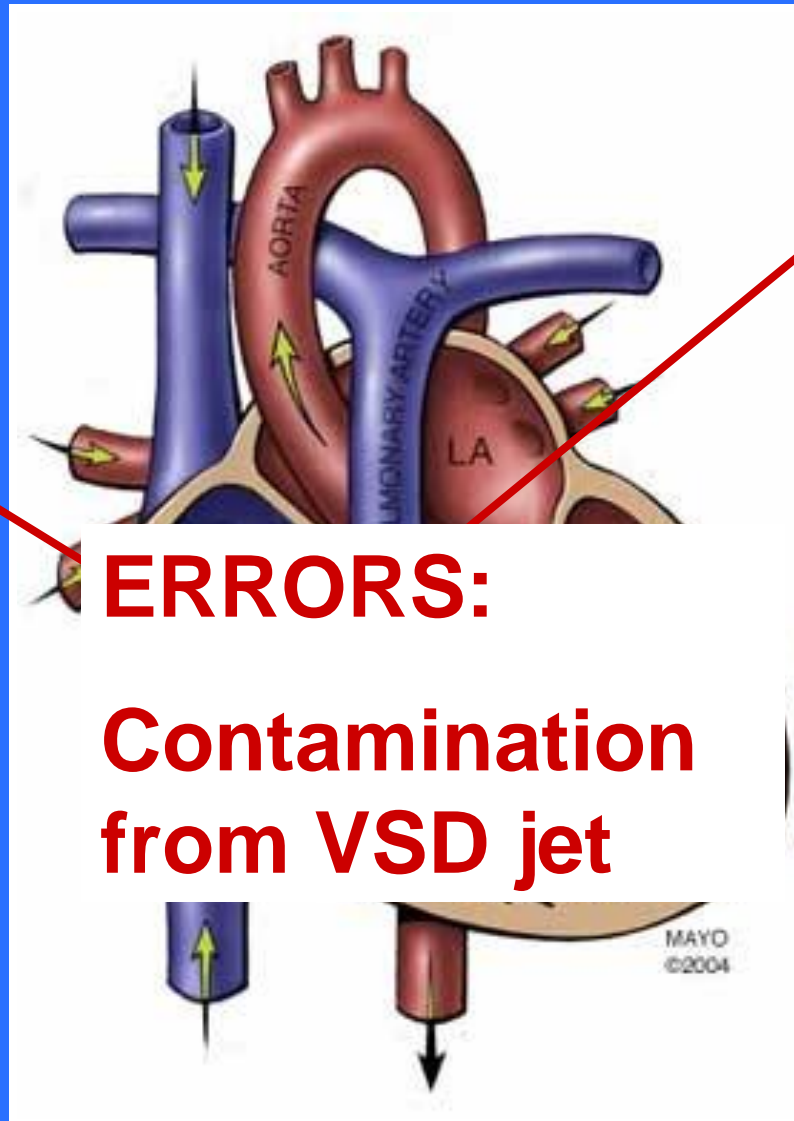
- **Small (restrictive):** Defect size  $< 1/3$  aortic root; velocity  $> 4$  m/s
- **Moderate:** Defect size  $1/2$  aortic root; velocity 3 m/s
- **Large (non-restrictive):** right and left ventricular systolic pressure near equal

## VSD Caveats

- The VSD jet may contaminate the TR signal
- Patients with high RV pressures may not have much color flow

# Pulmonary Hypertension?

TR estimates  
RV pressure



Outflow  
Obstructions:

1. PS
2. Double chamber RV

**ERRORS:**

**Contamination  
from VSD jet**

$$\text{PAP} = \text{QP} \times \text{PVR}$$

**PAP: Pulmonary artery pressure**

**QP: Pulmonary blood flow**

**PVR: Pulmonary vascular resistance**

## Indications for Closure

- Large VSD (left heart enlargement, QP/QS > 1.5) without irreversible pulmonary vascular disease
- Aortic valve prolapse with progressive regurgitation
- RV outflow tract obstruction
- Recurrent endocarditis

# Atrial Septal Defect vs. Ventricular Septal Defect

## ASD

- Increased pulmonary blood flow
- Primarily volume load
- Low incidence of pulmonary hypertension in adulthood

## VSD

- Increased pulmonary blood flow
- Primarily pressure load
- High incidence of pulmonary hypertension in adulthood



## Conclusion

- ASD and PAPVC = Right heart enlargement
- VSD and PDA = Left heart enlargement
- Patients with pulmonary HTN may not have significant shunt visible

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