Pulmonary Hypertension

Common and Interesting Cases

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Here, it's different.

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No Disclosures To Report

Case #1

Newborn transitioning from fetal to extrauterine life

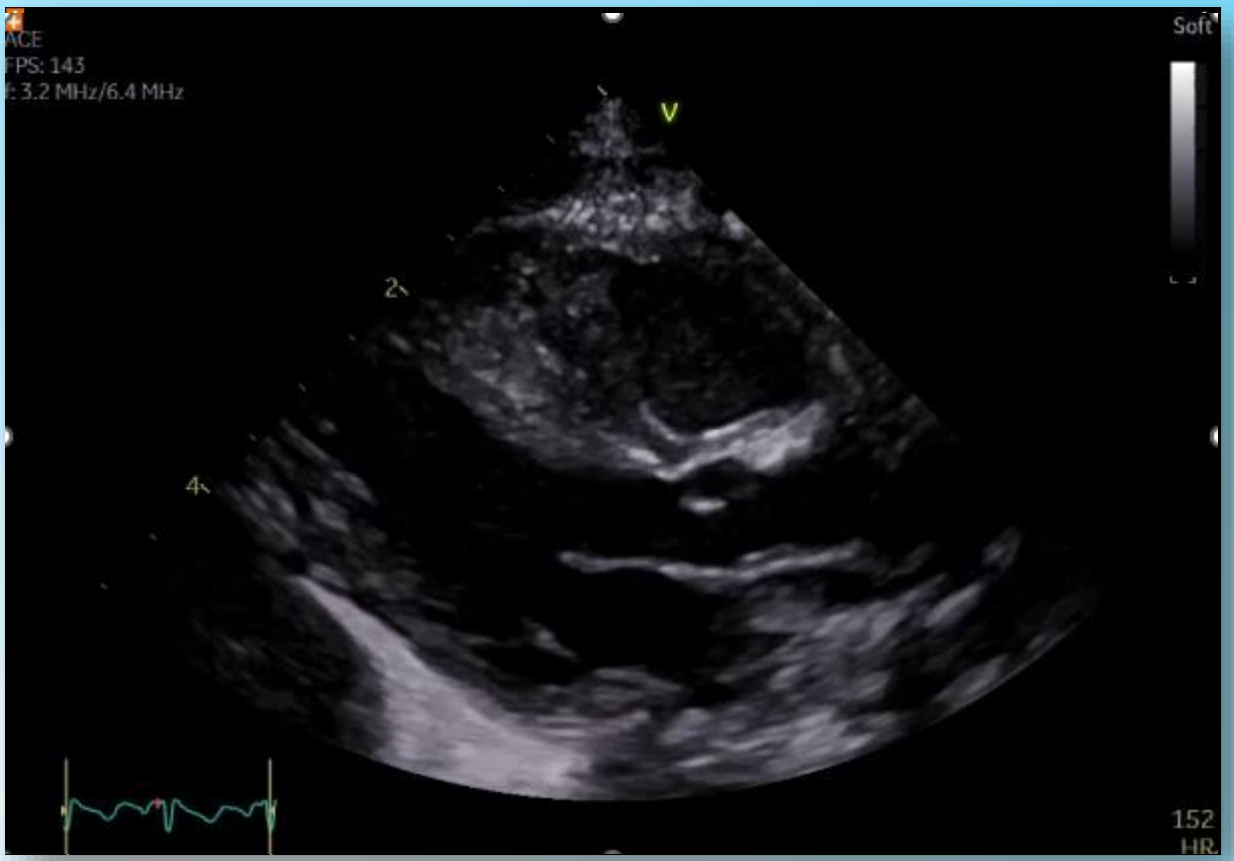
- 37 week infant on oxygen in the NICU

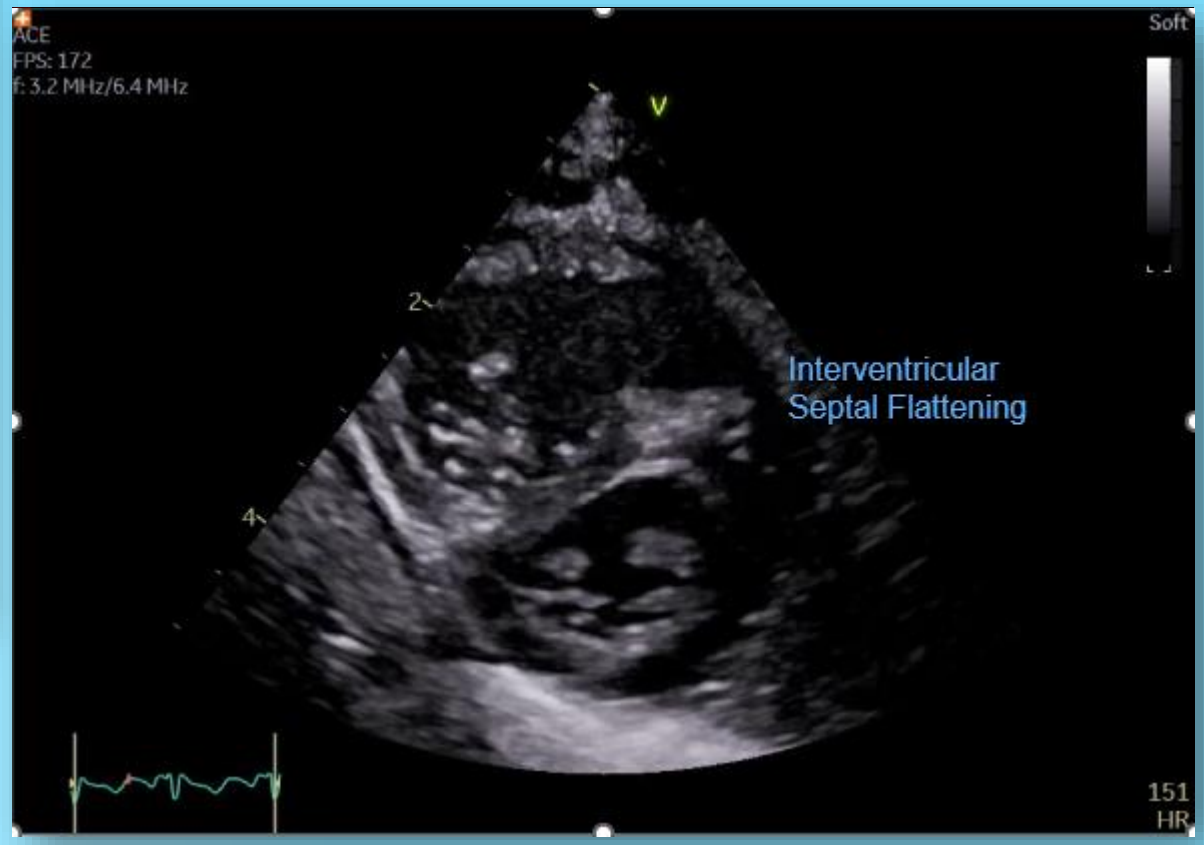
- Echo ordered for hypoxia on day-of-life #1

- Commonly seen in NICU

“And The Echo Says…”

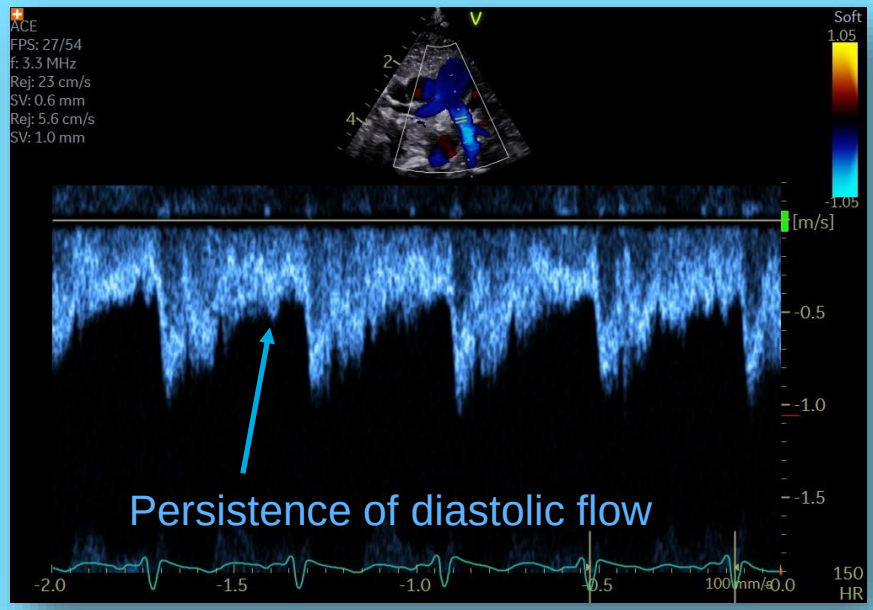
Parasteral Long and Short Axis

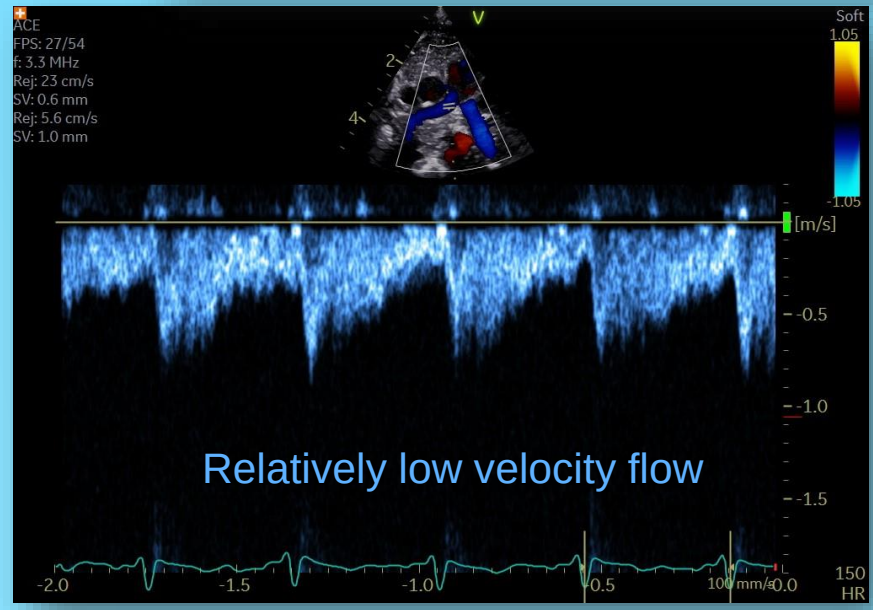




Branch Pulmonary Artery Flow

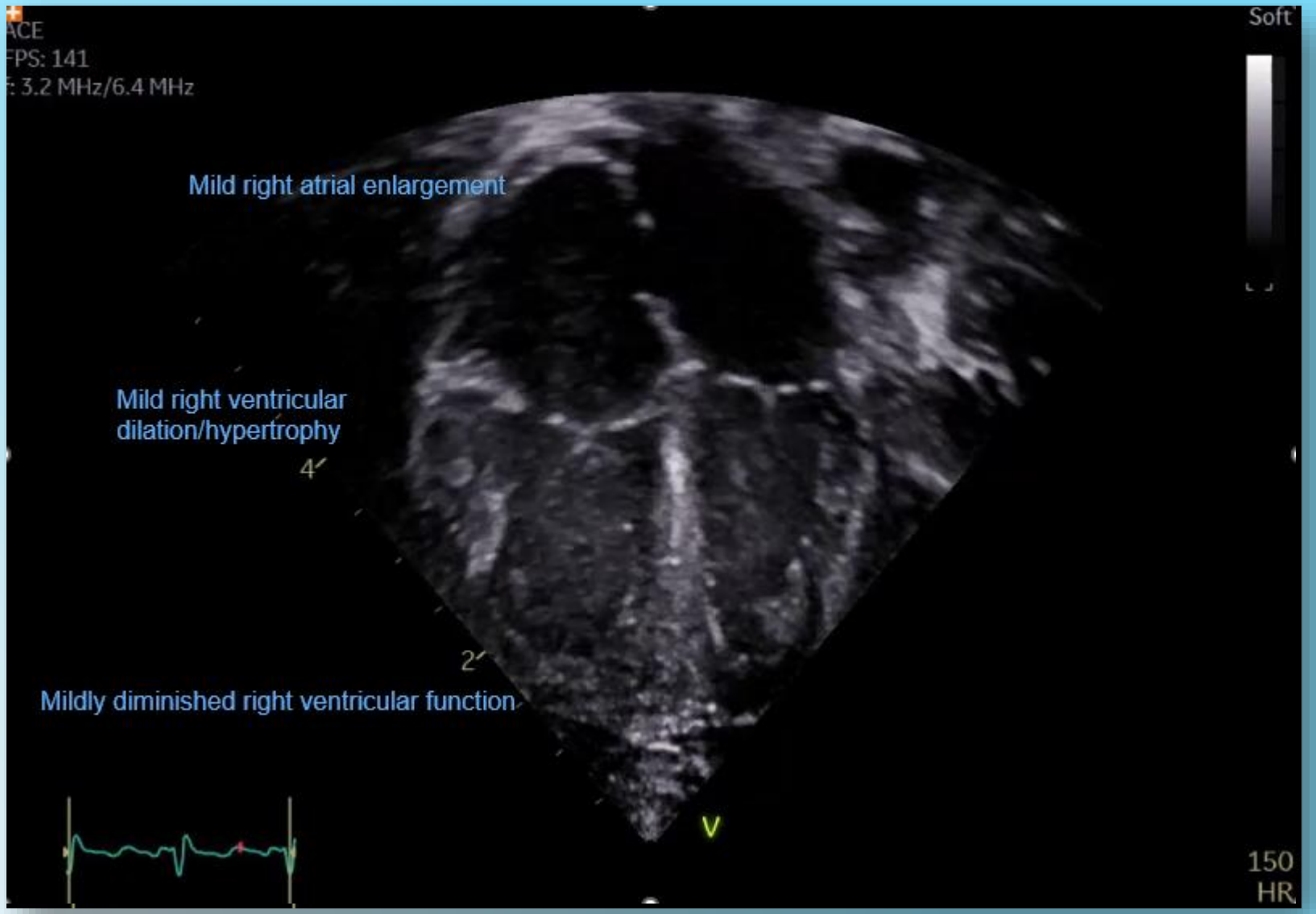
LPA



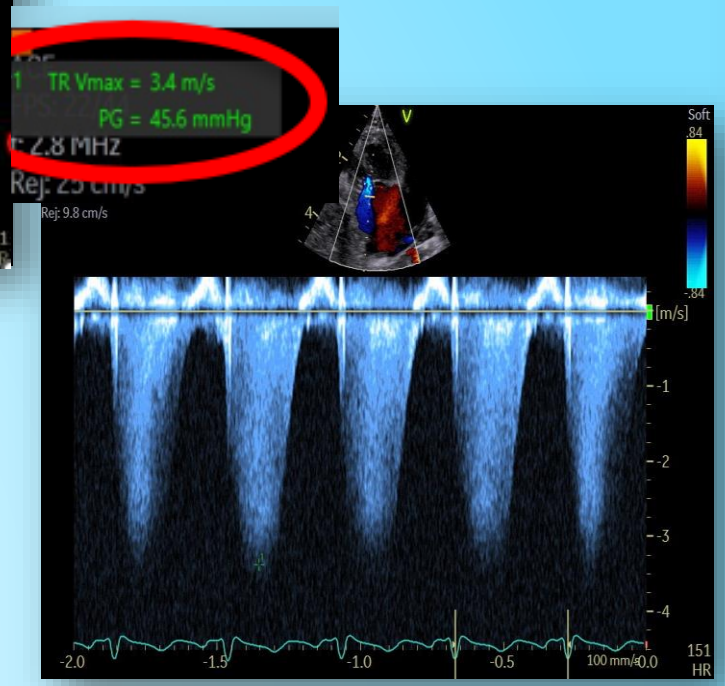


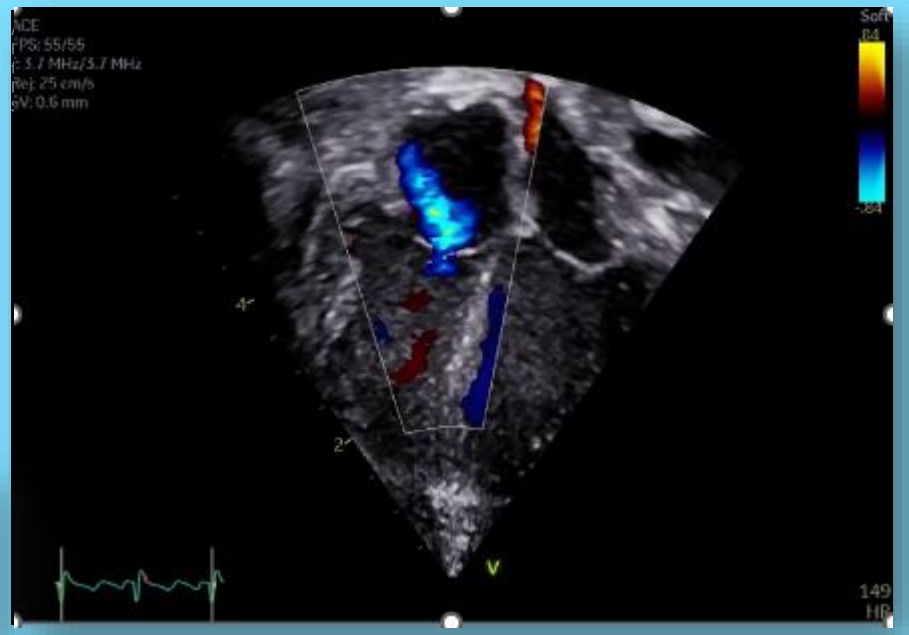
RPA

Apical Four Chamber



Tricuspid Regurgitation in Parasternal and Apical Windows

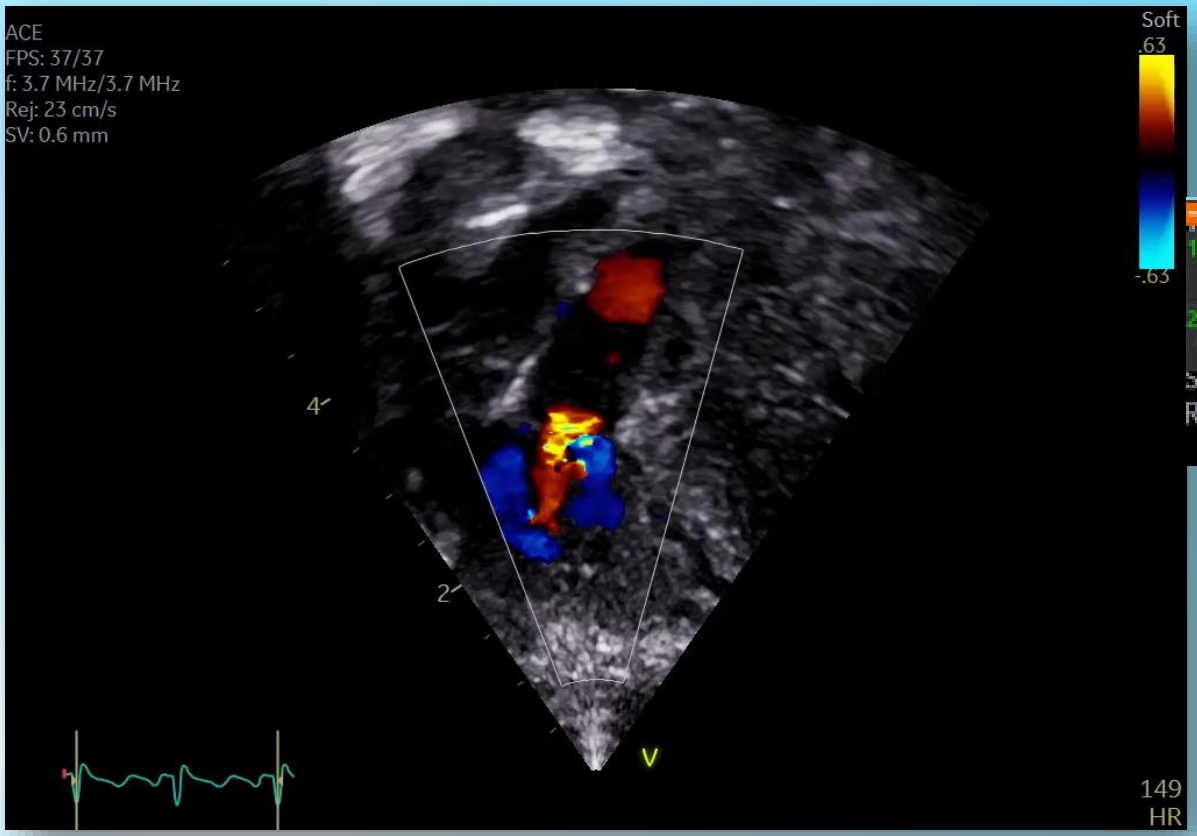


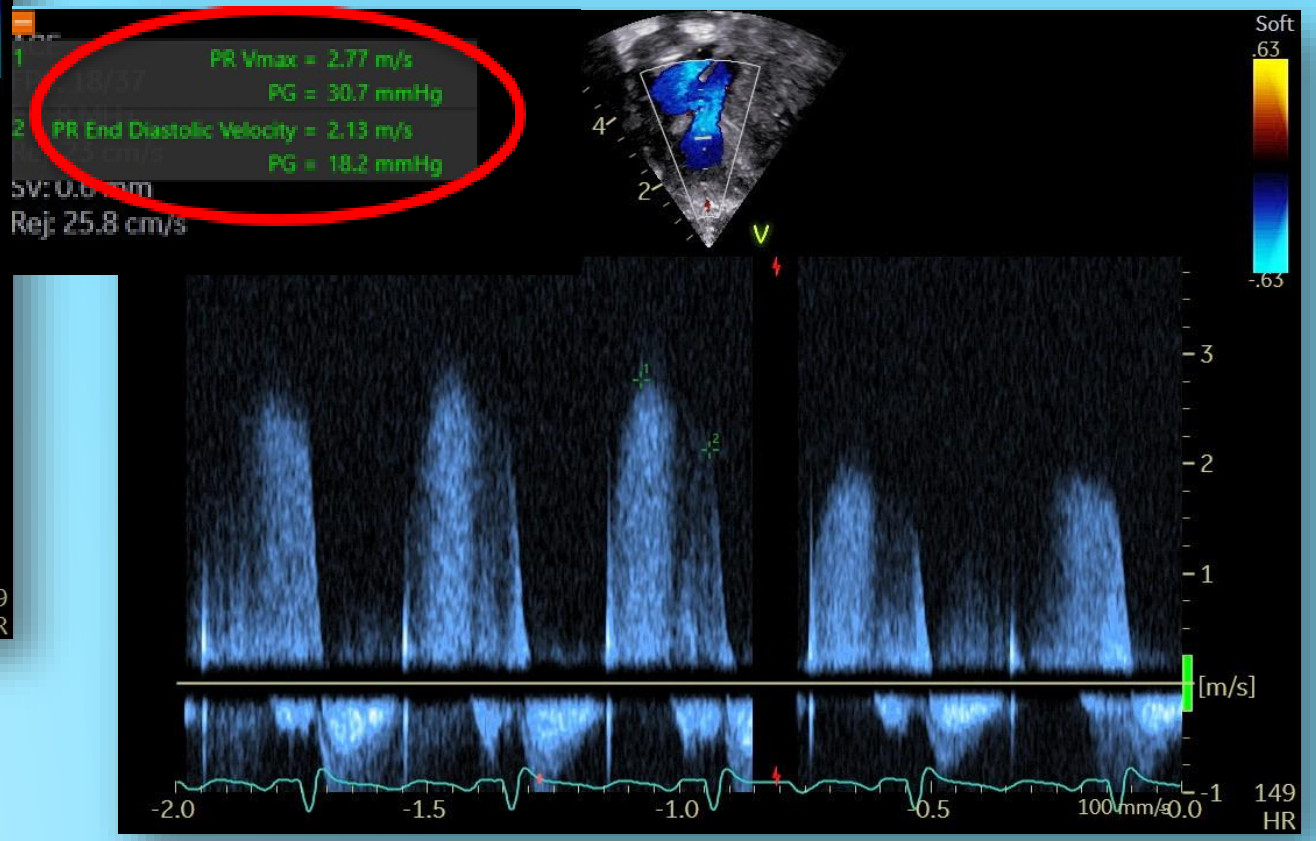


BP = 50/31

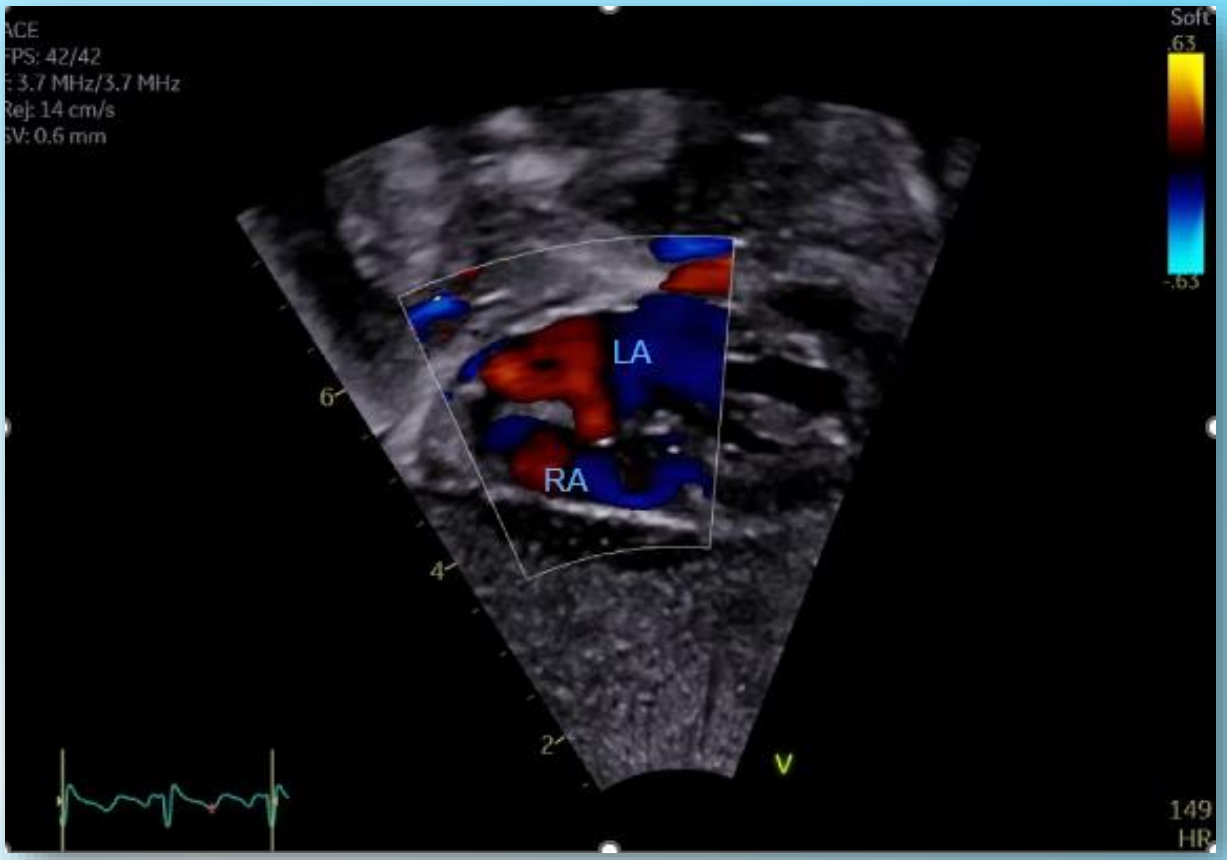
PAp = 46 mmHg +RAp = At least systemic PA pressure

Pulmonary Insufficiency
(From apical window with anterior angulation)



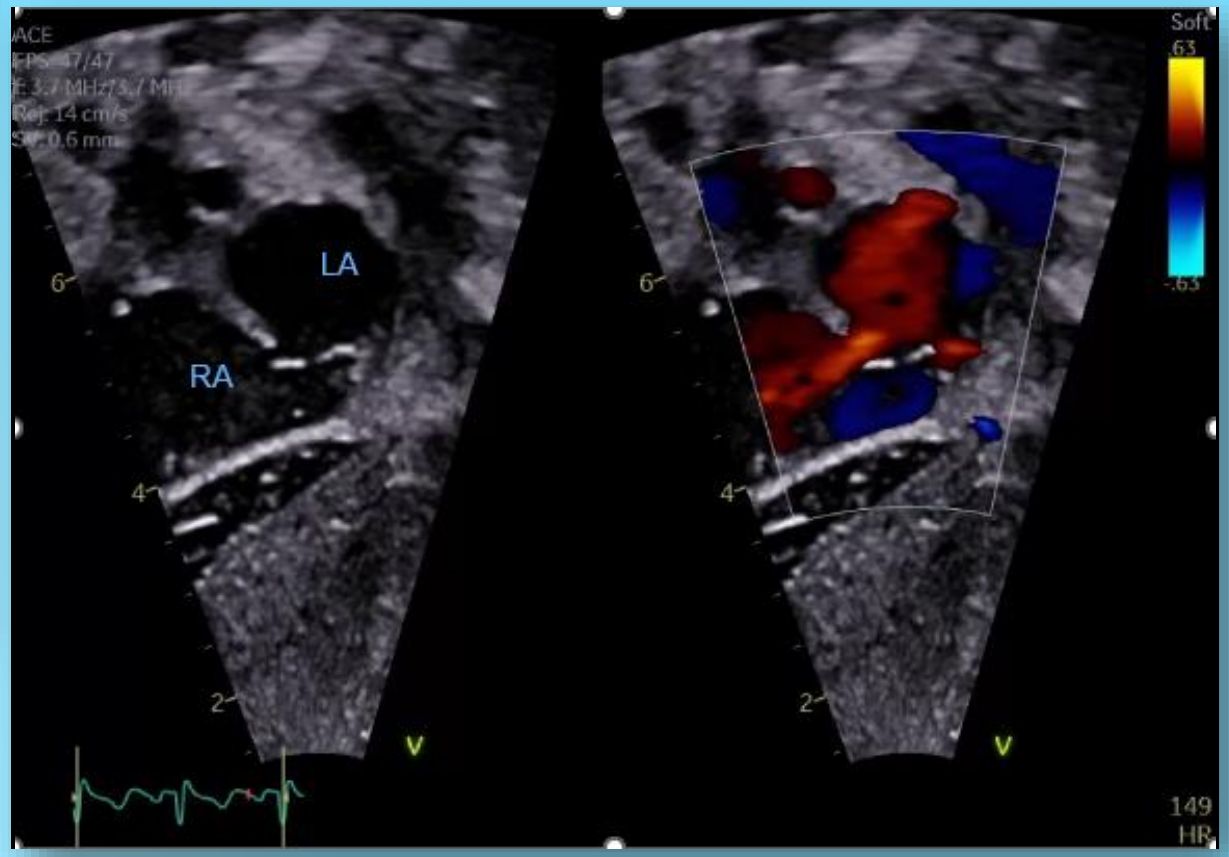


Atrial Level Shunt From The Subcostal Window

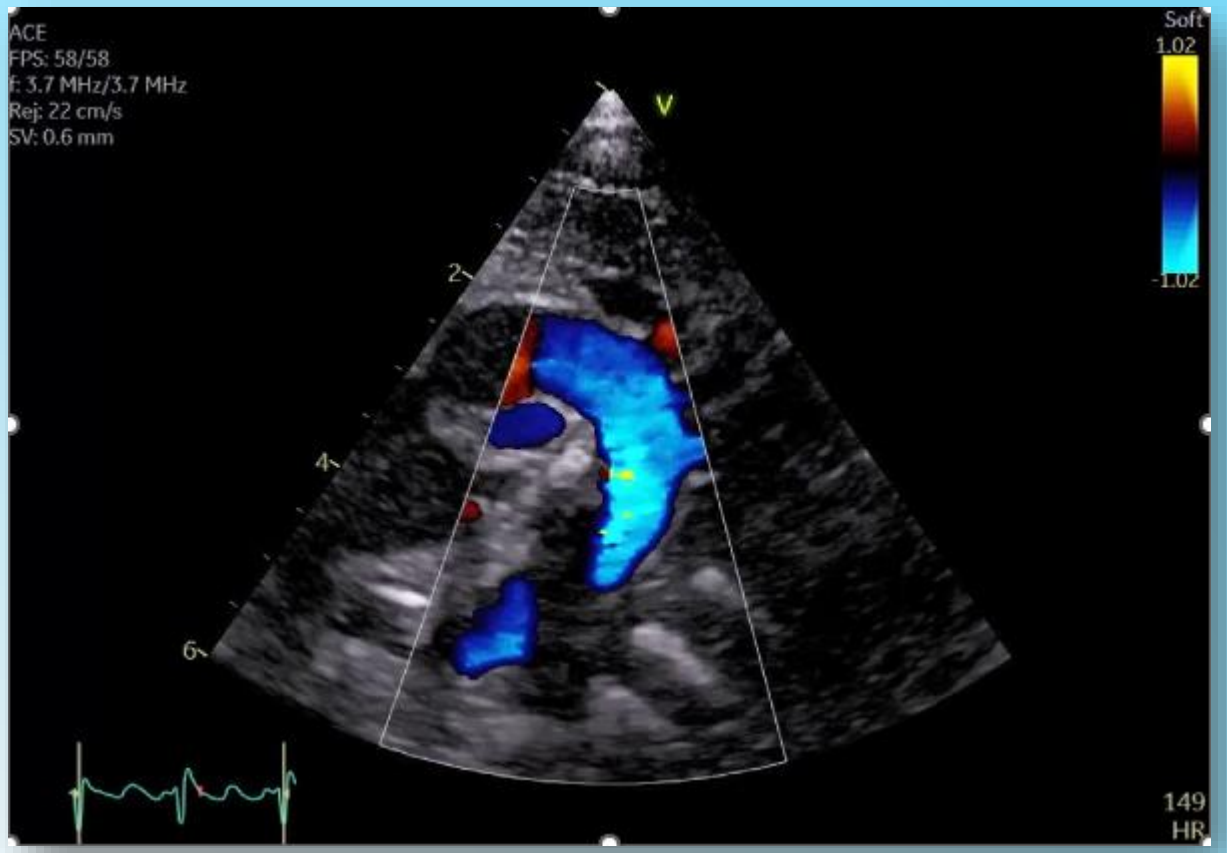


Subcostal Coronal (4-Chamber)

Modified Subcostal Saggital/Bicaval
(Angled leftward to profile shunt)



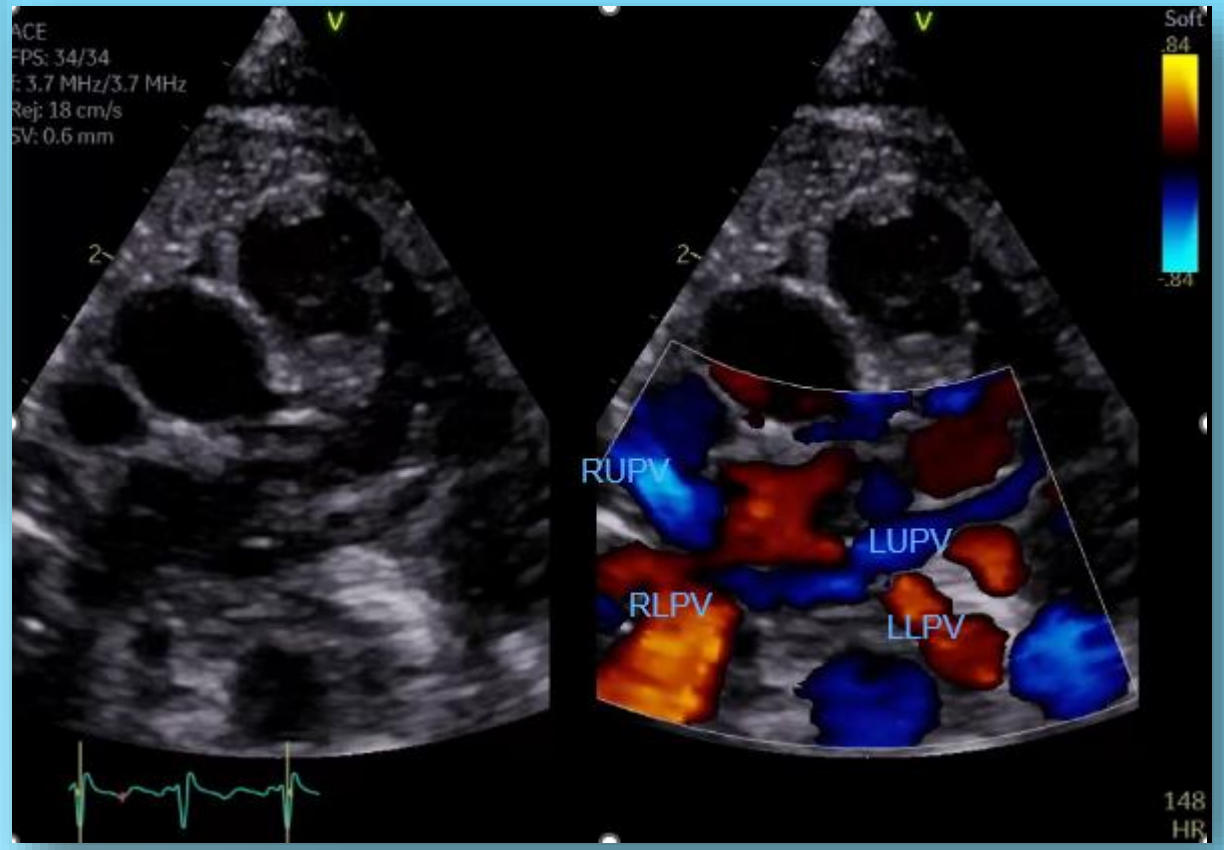
Suprasternal Window



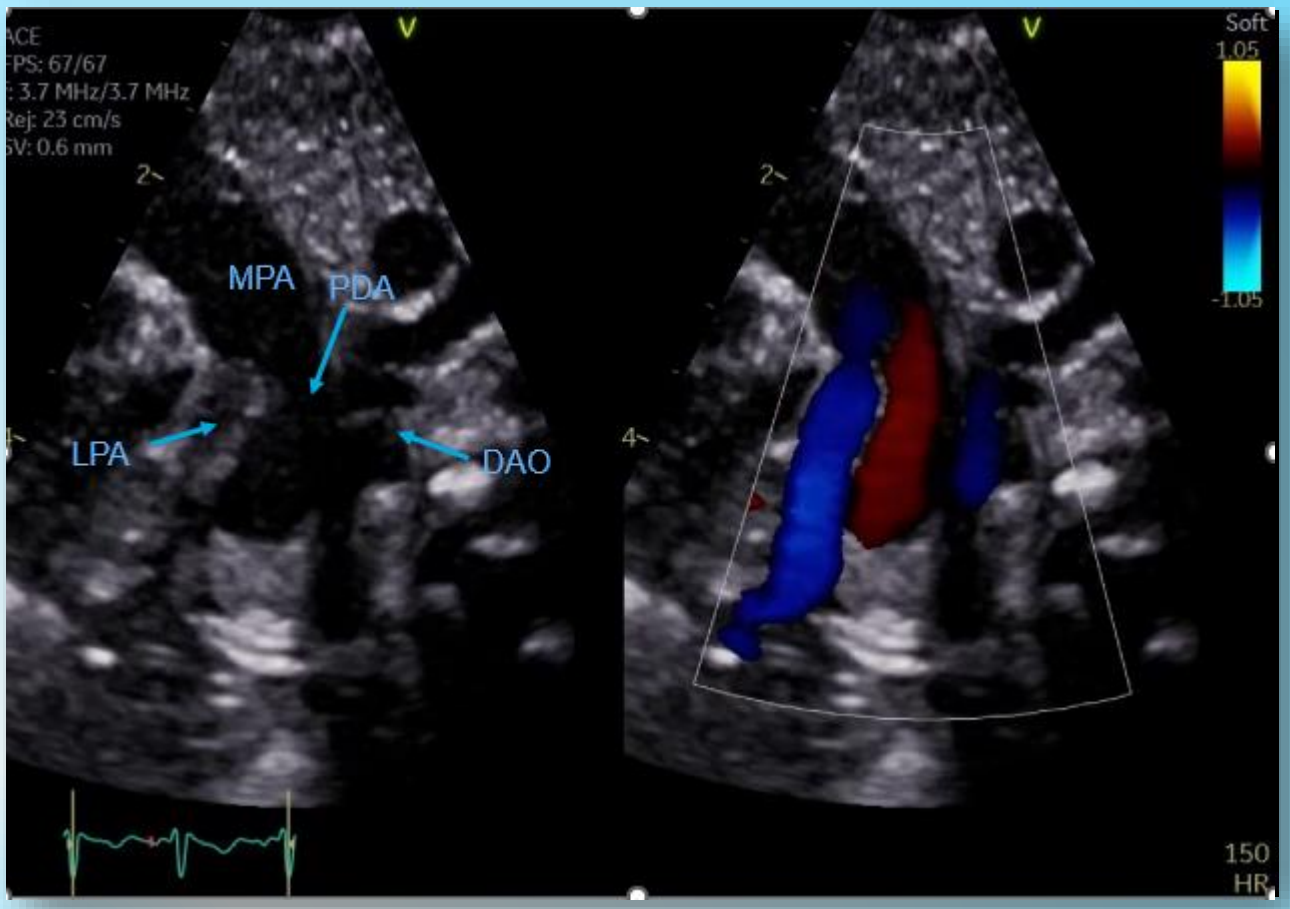
Normal aortic arch

Cannot rule out coarctation in the presence of PDA
(PDA not visualized in this image)

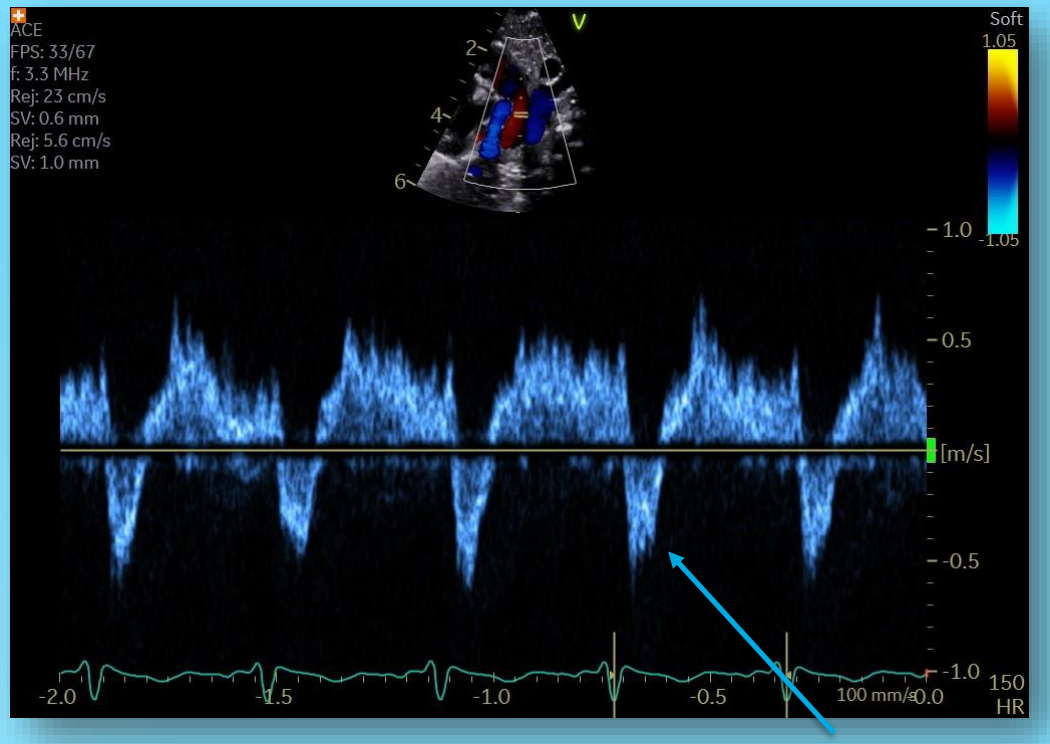
- At least 4 pulmonary veins entering the left atrium
- Spectral Doppler to insure no venous obstruction (normal in this patient)



Suprasternal ‘Ductal’ View



Ductal View



Bidirectional flow, right to left in systole
Indicating systemic pulmonary pressure

Information To Obtain For Pulmonary Hypertension

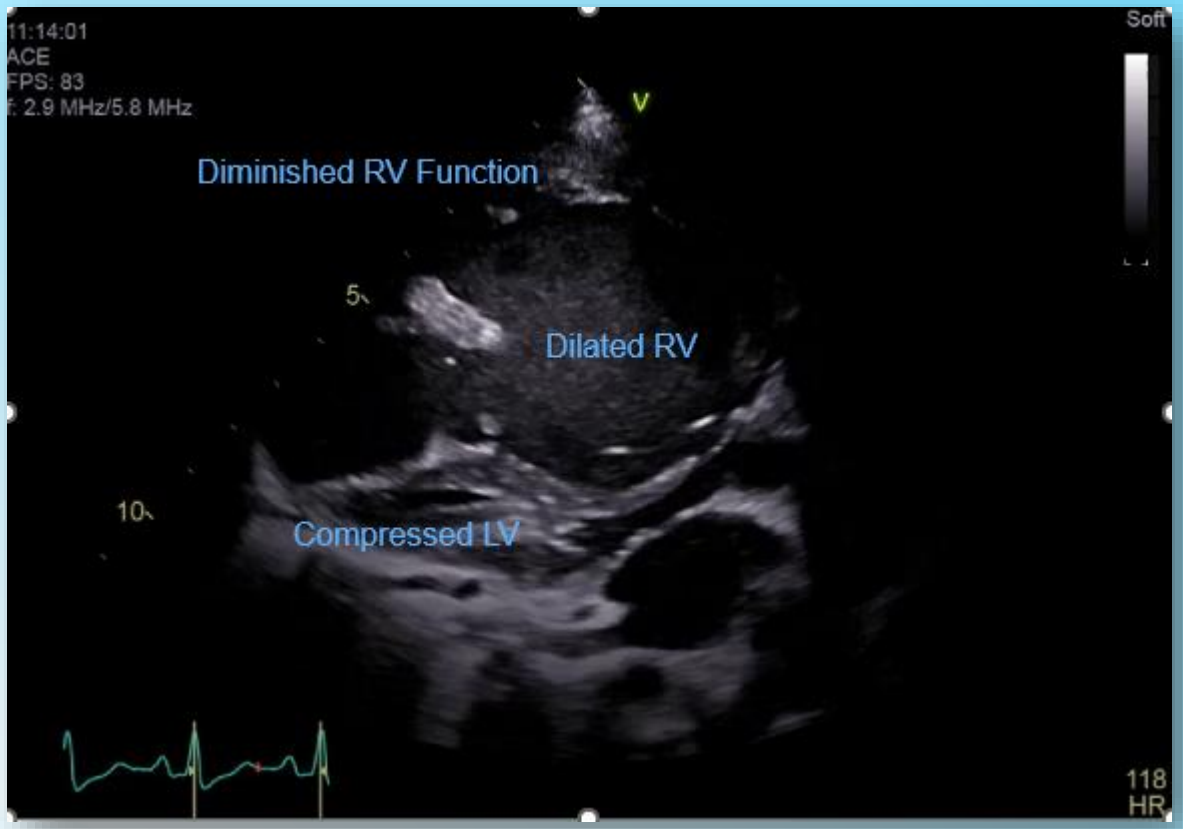
- Tricuspid Regurgitation – color and spectral Doppler
- Presence of interventricular septal flattening
- Pulmonary insufficiency – color and spectral Doppler
- Right ventricular dimension and function
- PDA size and shunt direction
- Presence of atrial level shunt – size and shunt direction
- Branch pulmonary artery size and flow
- Pulmonary veins entering unobstructed into the left atrium

Case #2, Potter Harry

- 29 week premie (2.2 lbs at birth), methamphetamine exposure; spent 3 months in an outside NICU
- Persistent asthma, hospitalized at least once per year at an outside hospital
- Age 6 – First Echo done at CHCO for increased oxygen demand during a hospital stay for asthma exacerbation

“Show Us The Echo!!”

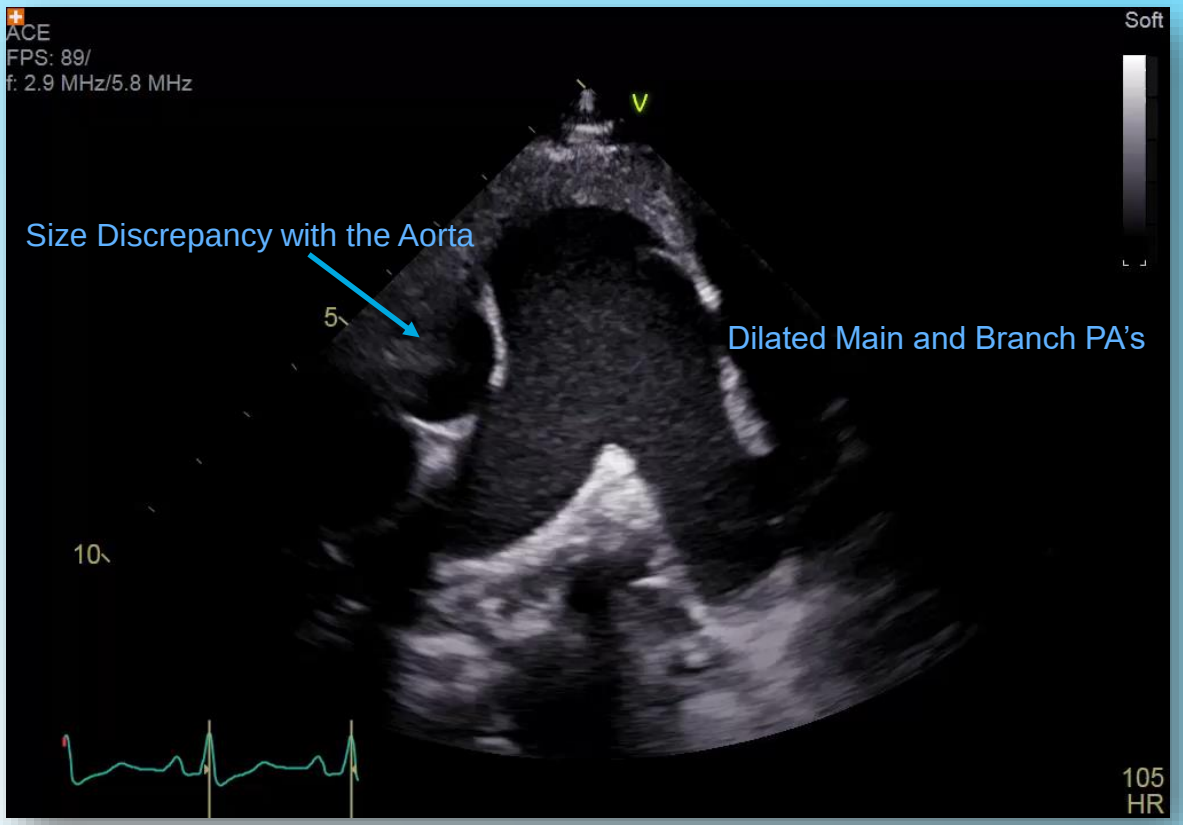
Parasternal Long Axis



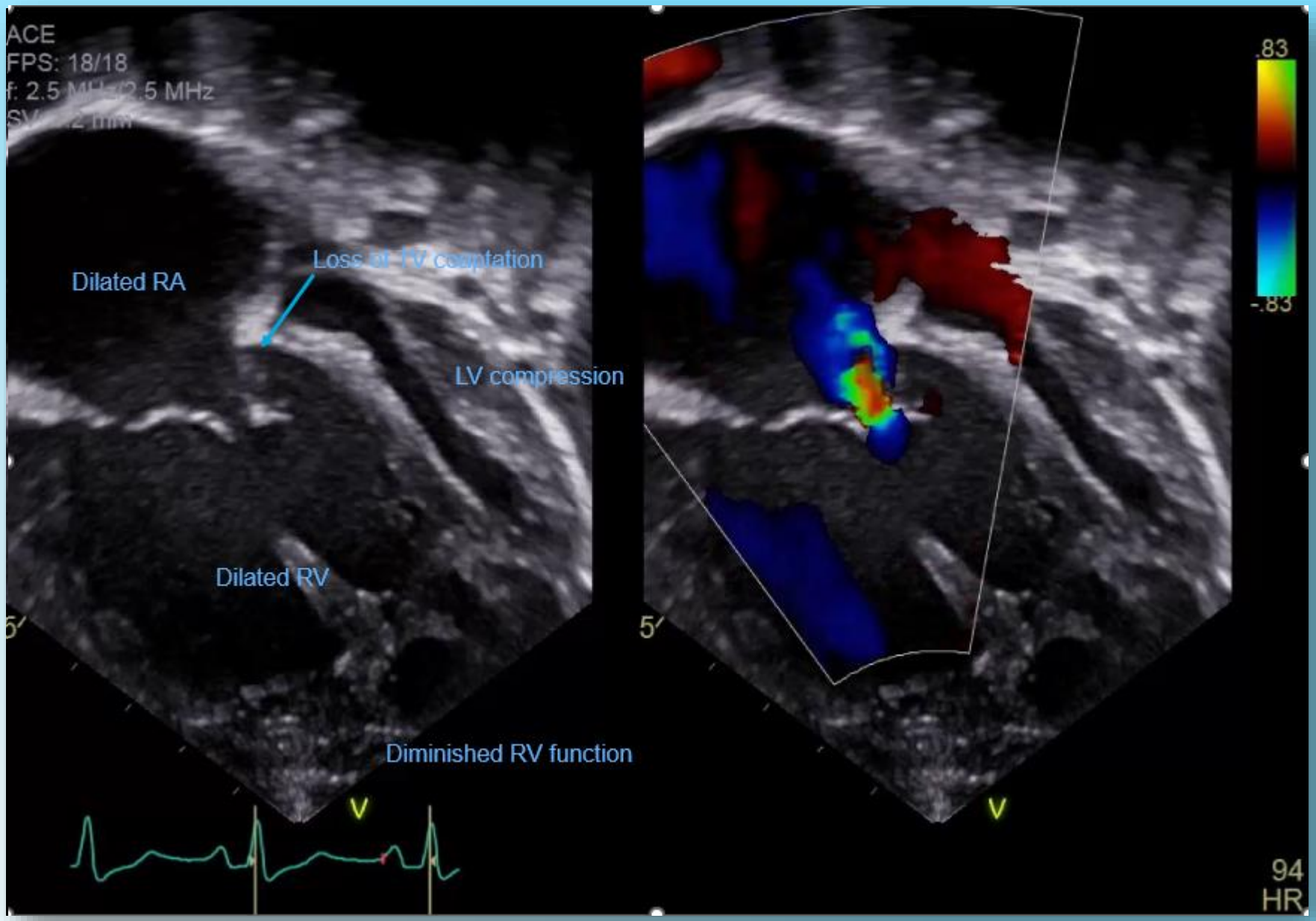
Parasternal Short Axis

The Abominable Snowmonster of the North | Christmas Specials Wiki | Fandom

Parasternal Short Axis
Branch Pulmonary Arteries



Apical Four Chamber

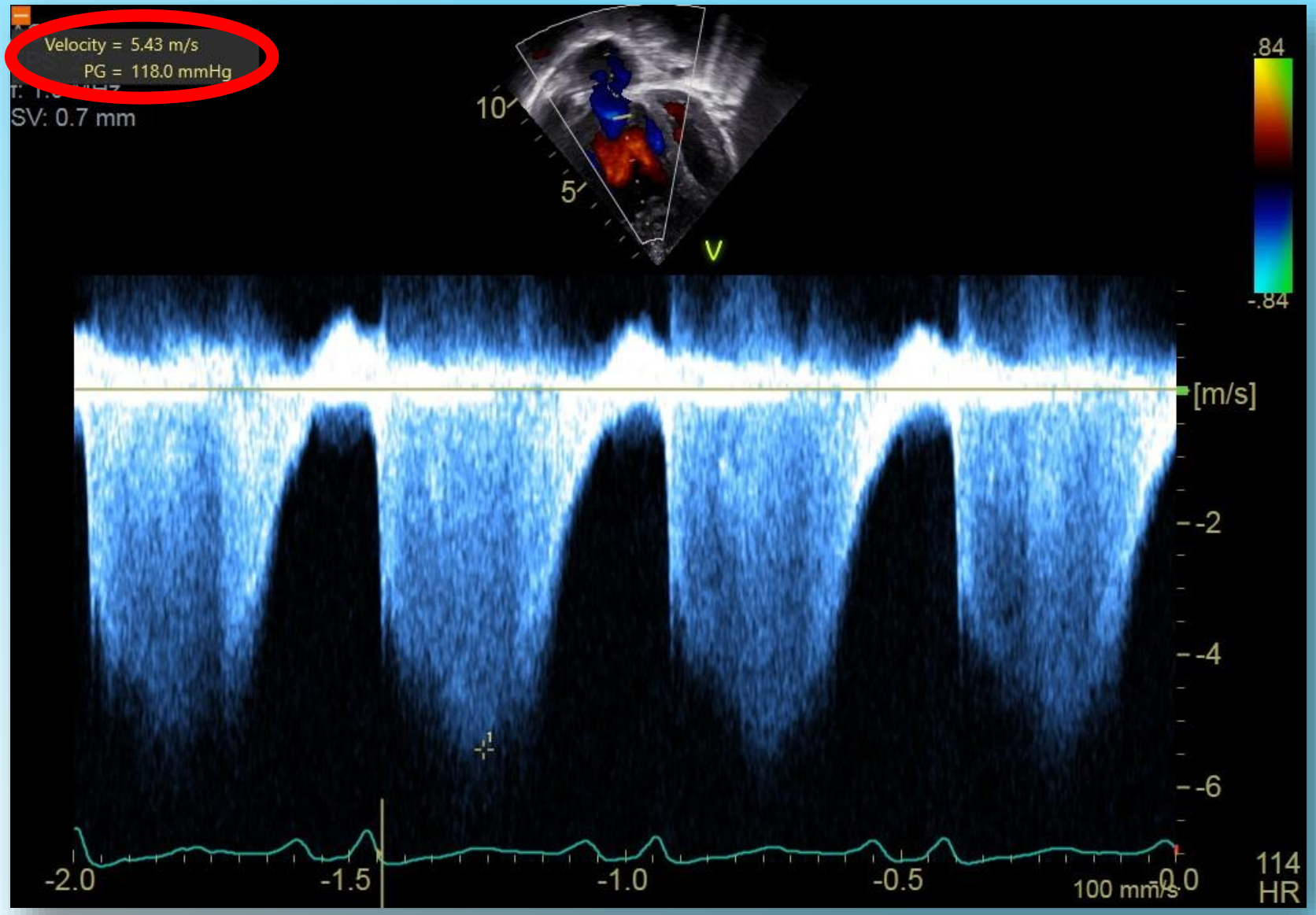


Spectral Doppler of Tricuspid Regurgitation

Calculated PAp:
118 mmHg
+ RAp (at least 5 mmHg) =
123 mmHg

Systemic Blood Pressure = 98/63

Suprasystemic RV Pressure / Severe Pulmonary Hypertension



Potter’s Diagnosis and Treatment

- Diagnosis: Severe pulmonary arterial hypertension
- Medications past and present – PH meds: Tadalafil, Ambrisentan, Sildenafil, Remodulin; Lasix
- Surgery – Potts procedure May 2019

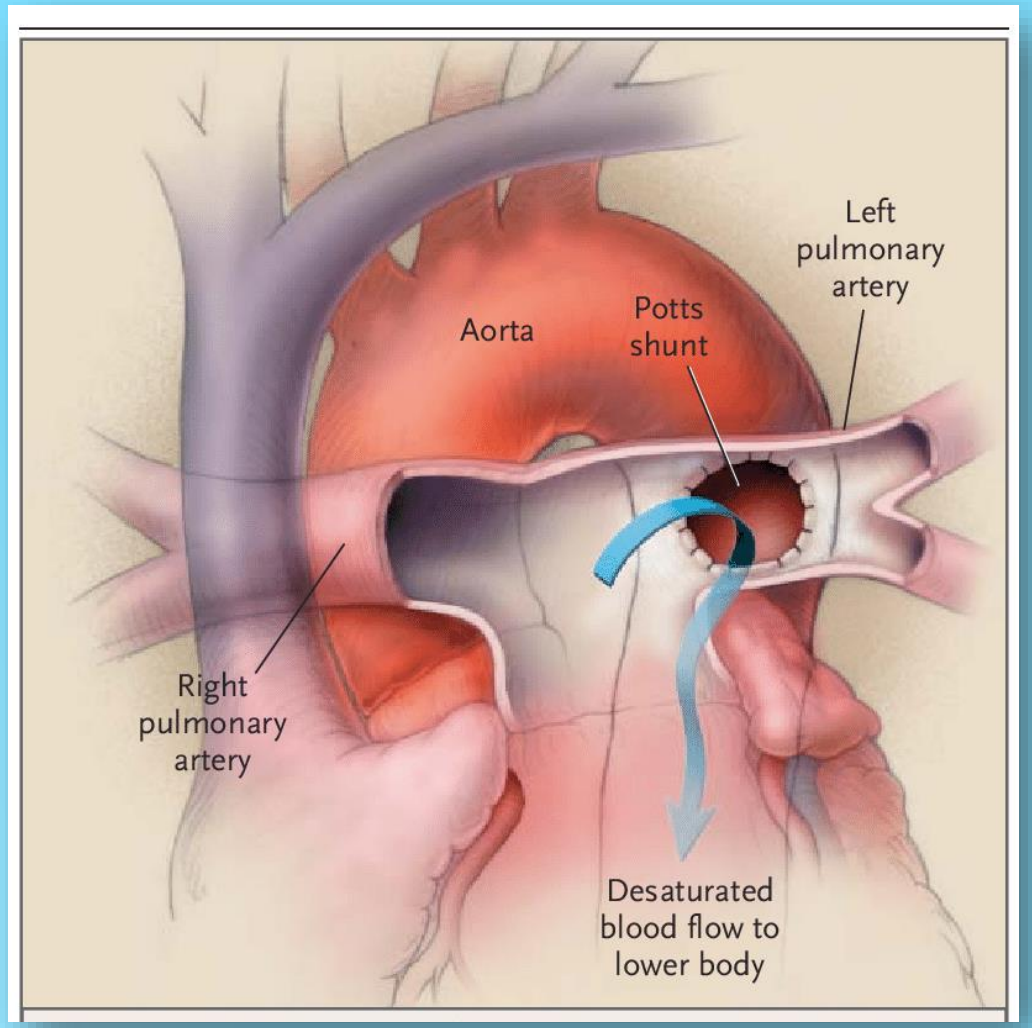
Potts Shunt

Definition

Side-by-side anastomosis of the left pulmonary artery to the descending aorta, acting as a ‘pop-off’ connection for the pulmonary artery during periods of suprasystemic RV pressure

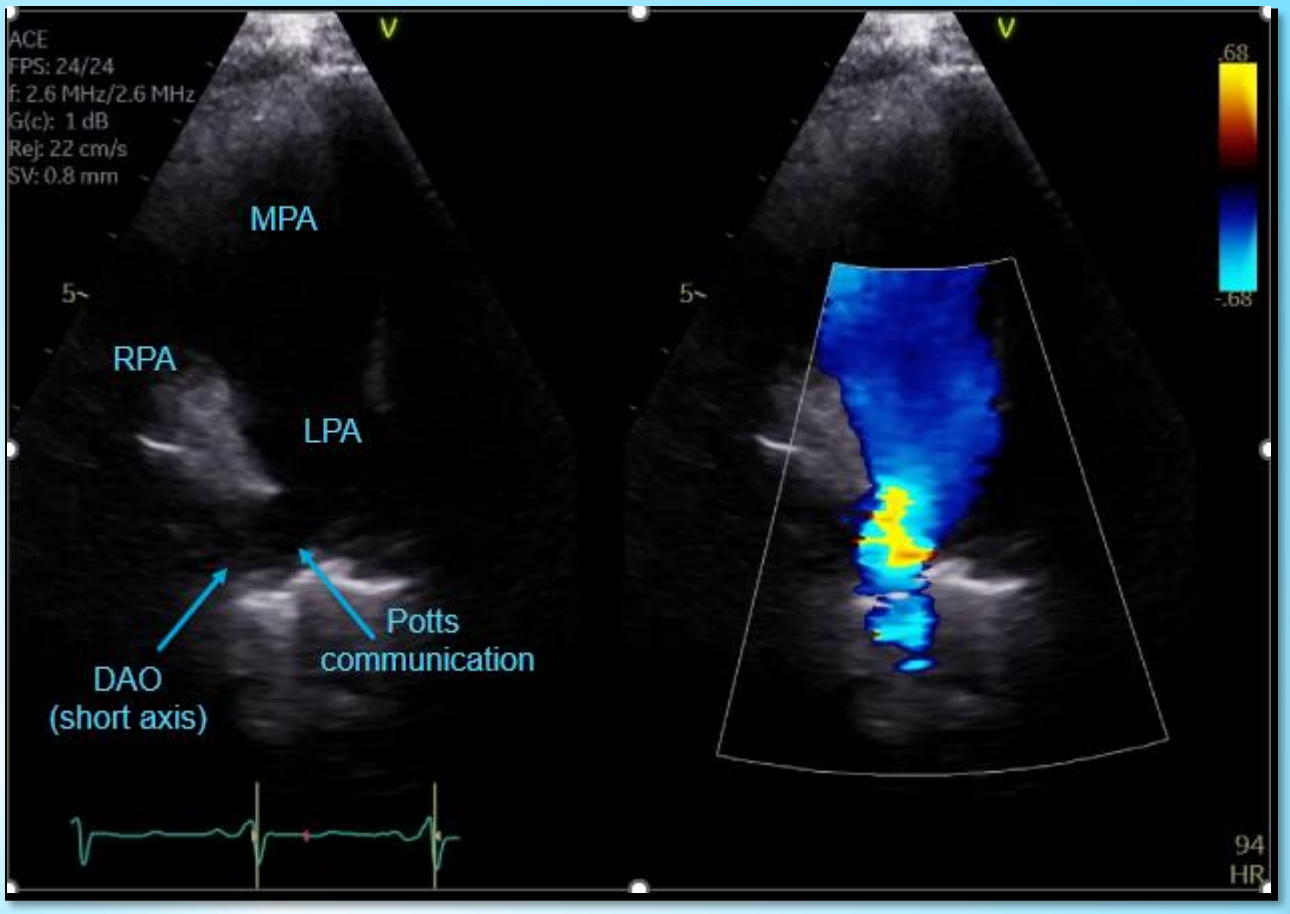
Benefits

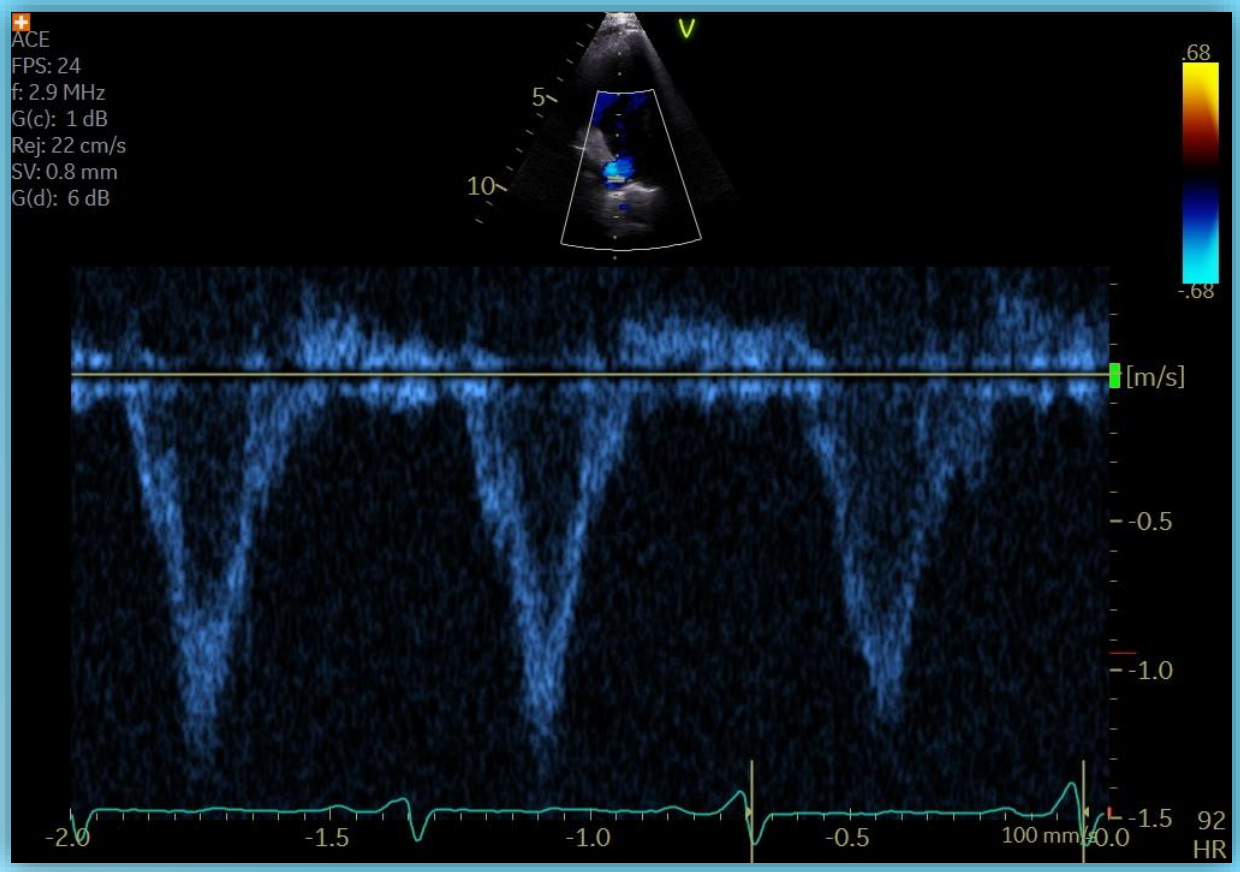
- Decreases RV pressure to prevent right heart failure
- May improve LV cardiac output by decreasing leftward bowing of the interventricular septum / compression of the LV
- Delays the need for lung transplantation



Source: The New England journal of Medicine
https://www.researchgate.net/figure/The-Potts-Shunt-Procedure_fig1_8887246

Potts Shunt Imaging
Parasternal Short Axis





Pulsatile systolic right to left shunting
(from the LPA into the descending aorta)

Potter’s Current Condition and Prognosis

- Potter Harry is now 14 years old (5 years post-op) - doing well with the Potts shunt; currently on oxygen, Tadalafil, bronchodilators for asthma

- Potter has been discussed for lung transplantation but currently does not meet criteria

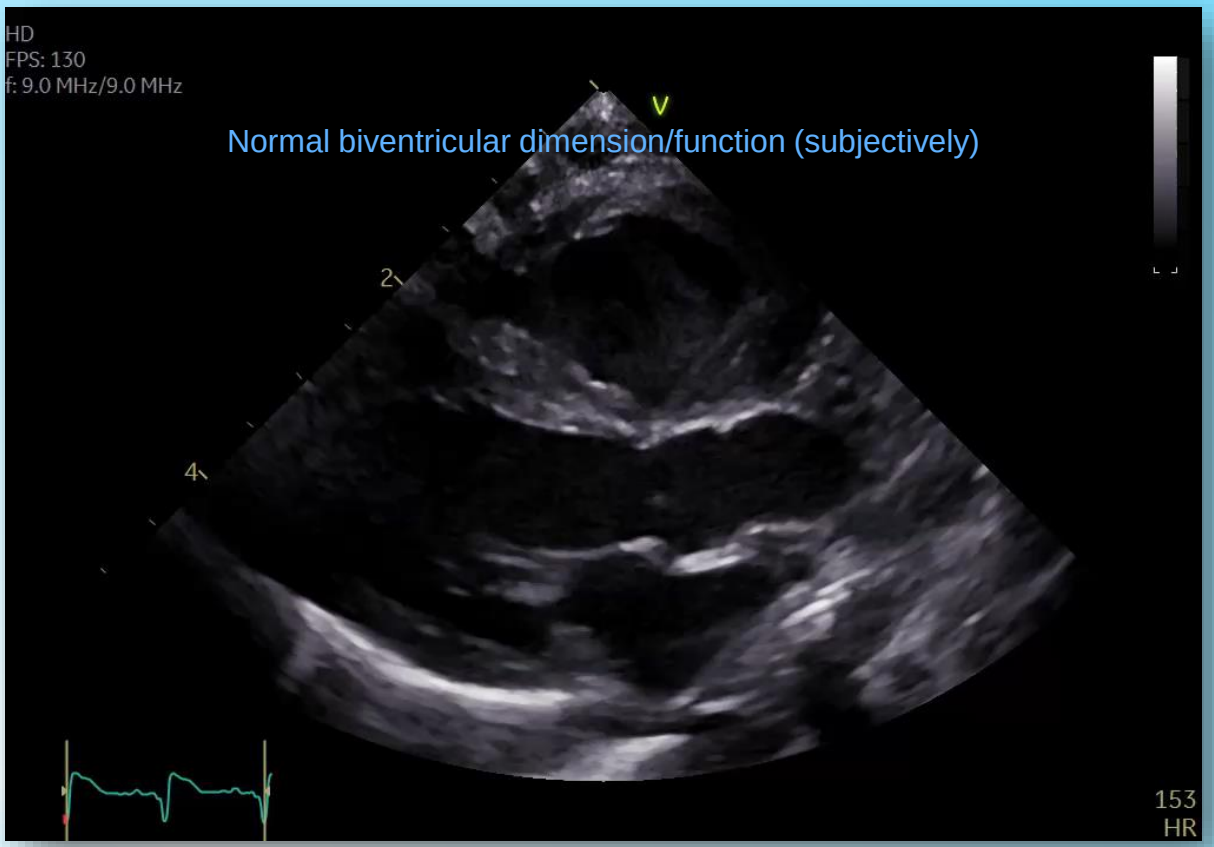
Case #3, Penelope Harper

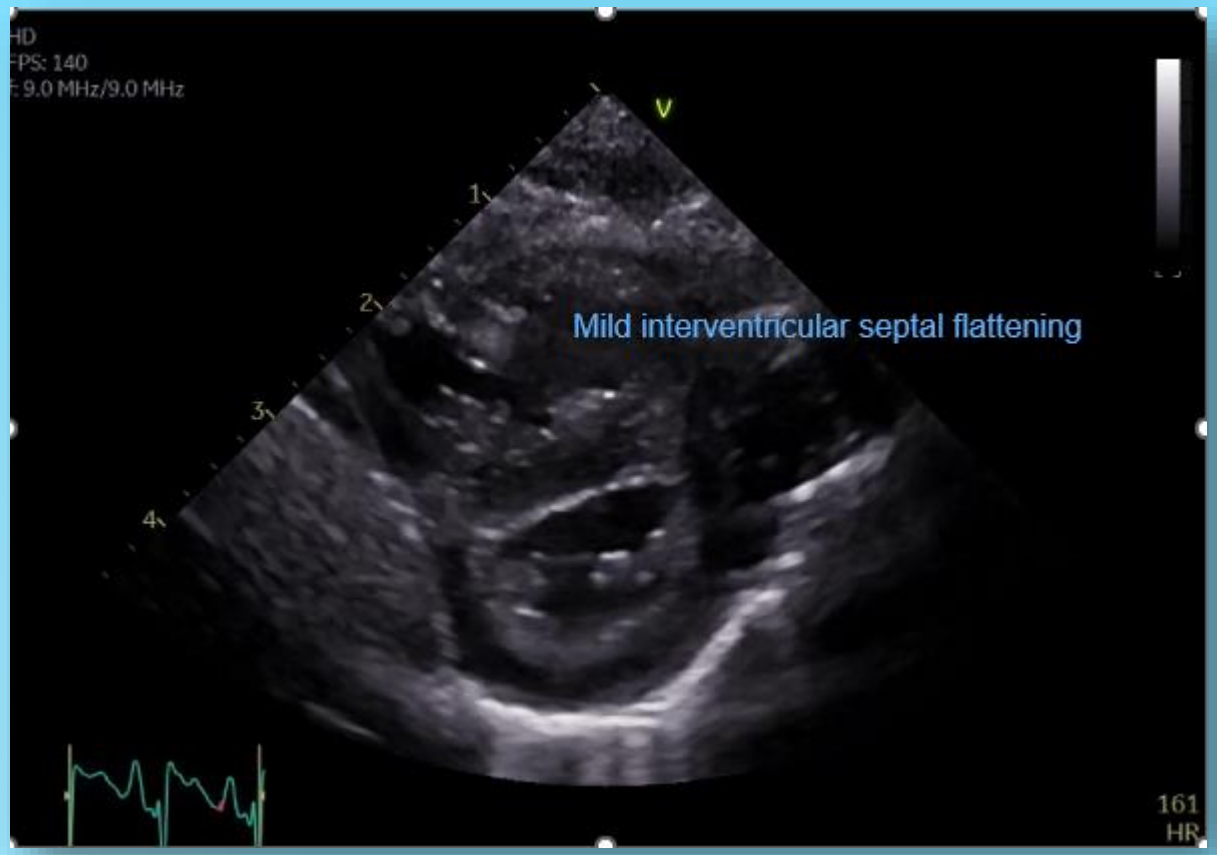
- 34 week premie with prenatal ultrasound concerning for multiple anatomic anomalies, including low right lung volume, duodenal atresia, dextrocardia

- Day-of-life #2, Penelope displayed increased work of breathing on CPAP and right sided heart sounds - echo ordered

“Enough Talk, More Echo!”

Parasternal Long/Short Axis





Parasternal Short Axis

Size discrepancy of the branch PA's – mild RPA hypoplasia

“Apical Four Chamber”

Image obtained from between the apical and subcostal windows

Foreshortened

On the RV apex- unable to get directly on the LV apex

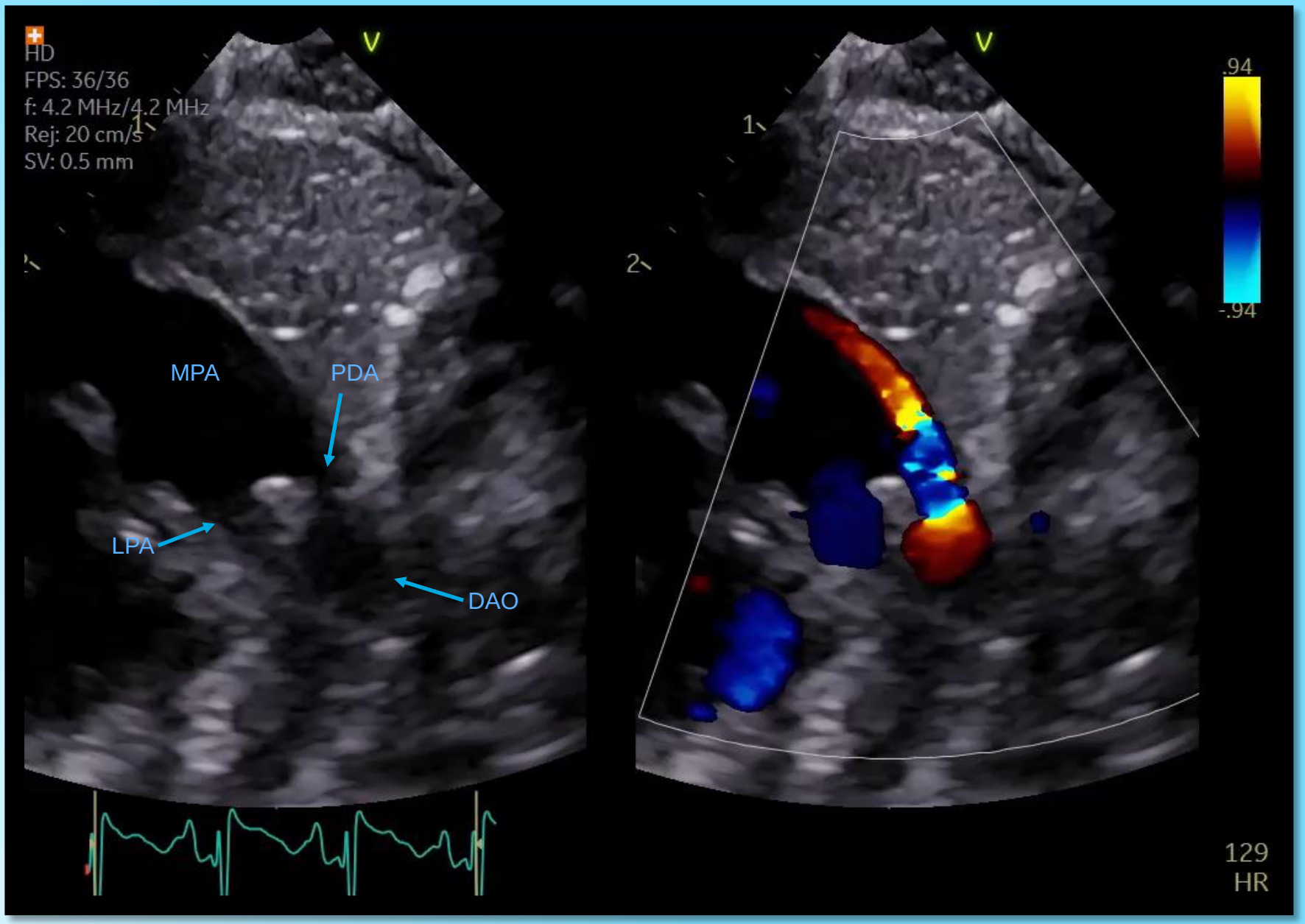
Heart is shifted more rightward than normal

Mesocardia

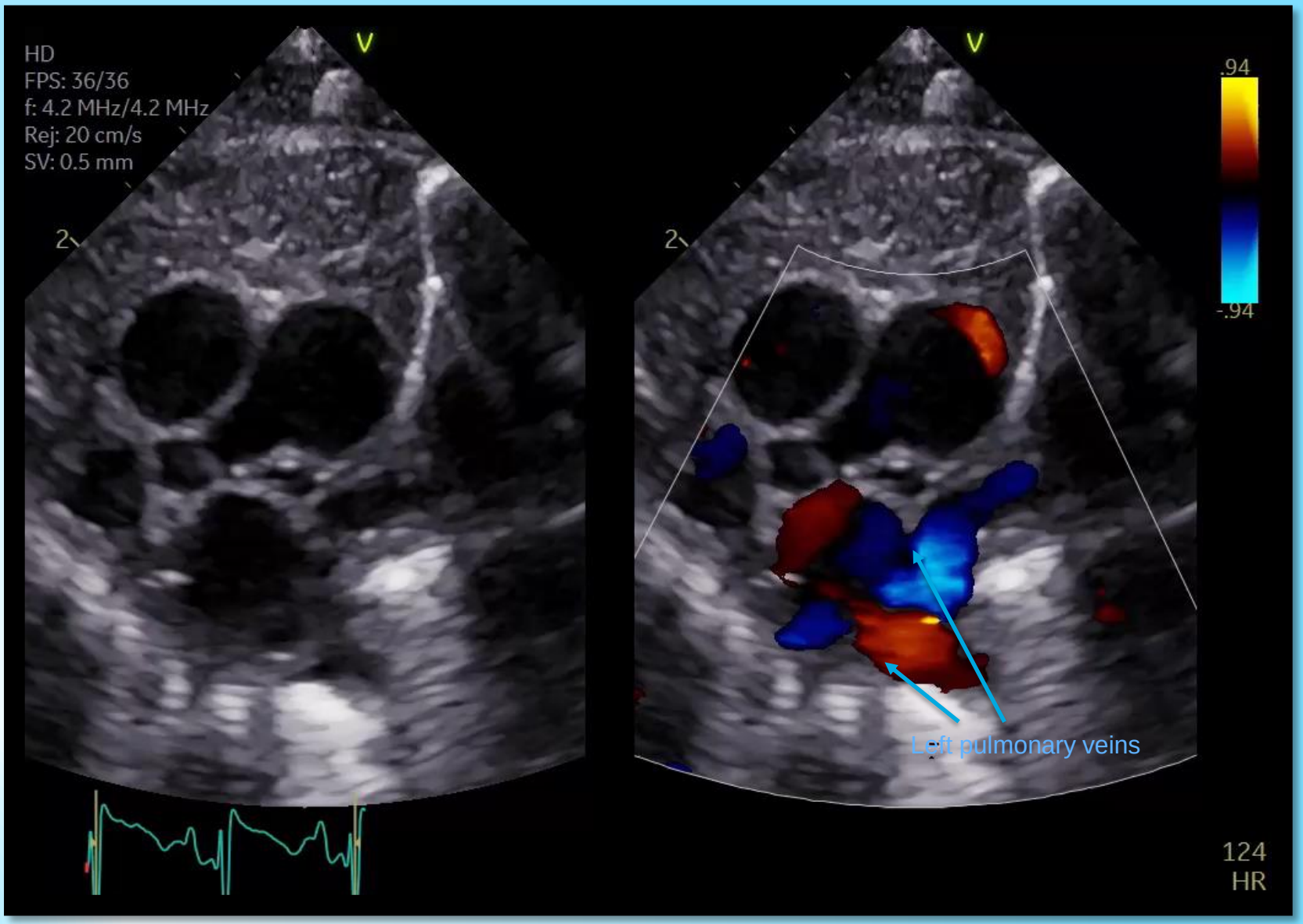
Suprasternal Ductal View

PDA is bidirectional, with right to left flow in systole

This indicates systemic pulmonary artery pressure



Suprasternal Window

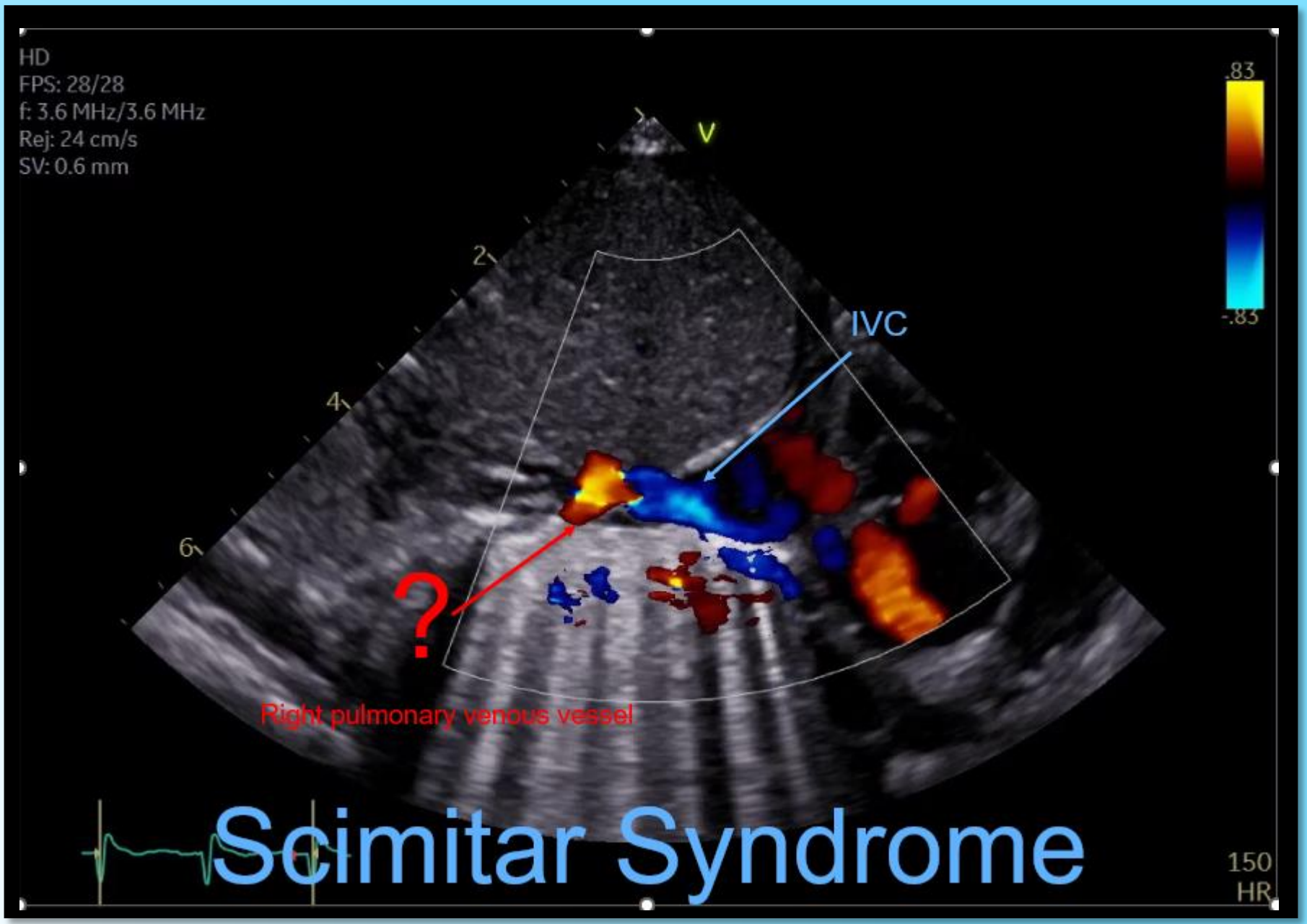


Unable to visualize the right pulmonary veins

Penelope’s Findings So Far…

- Mild interventricular septal flattening
- Hypoplastic right pulmonary artery
- Difficulty scanning the true LV apex - Mesocardia
- Bidirectional PDA
- Inability to image the right pulmonary veins

Subcostal Long Axis



Scimitar Syndrome

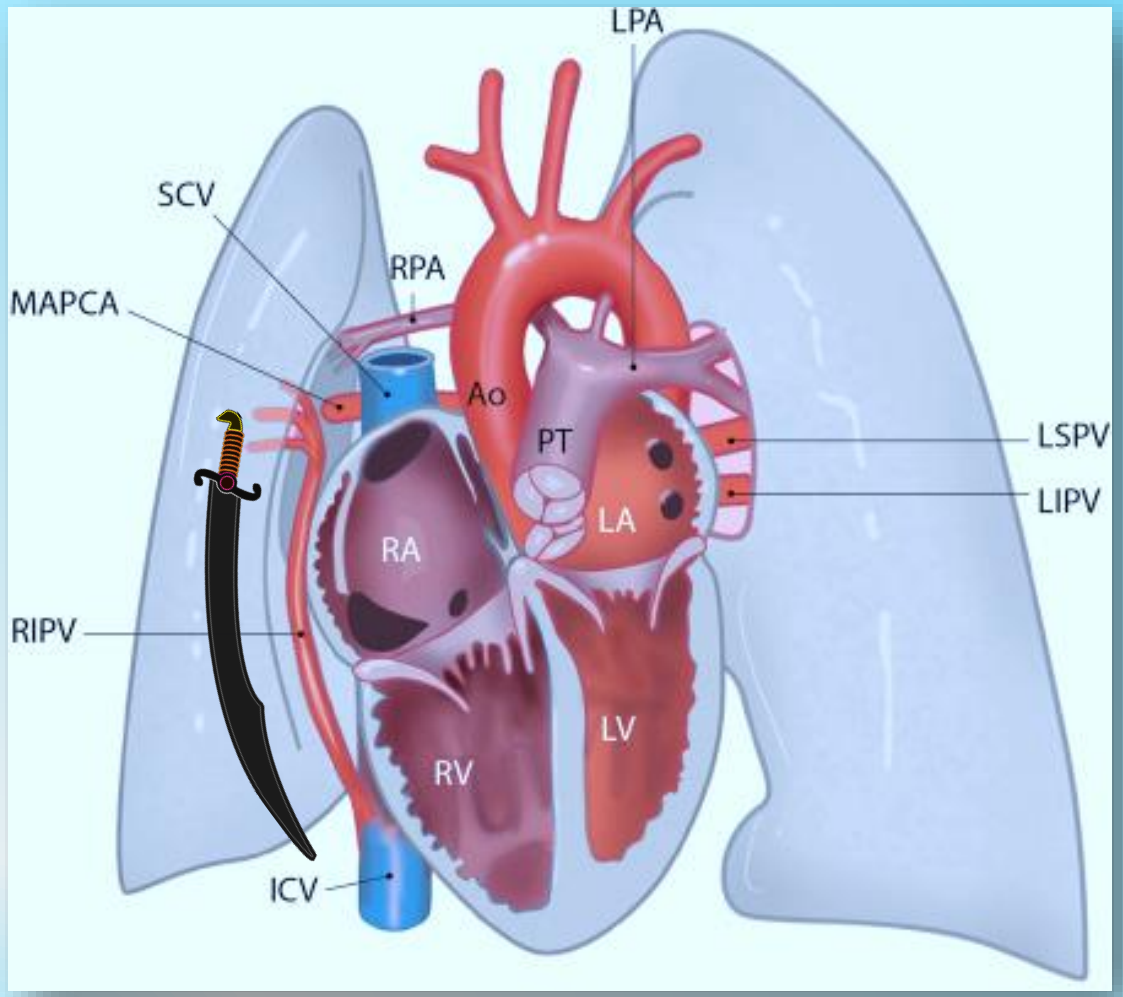
1. Rare congenital heart defect, occurs in ~ 2 of 100,000 live births
2. Variant of partial anomalous pulmonary venous return (PAPVR)
3. Etiology is unknown, thought to be an error in the development of the right lung bud

Characteristics seen via imaging:

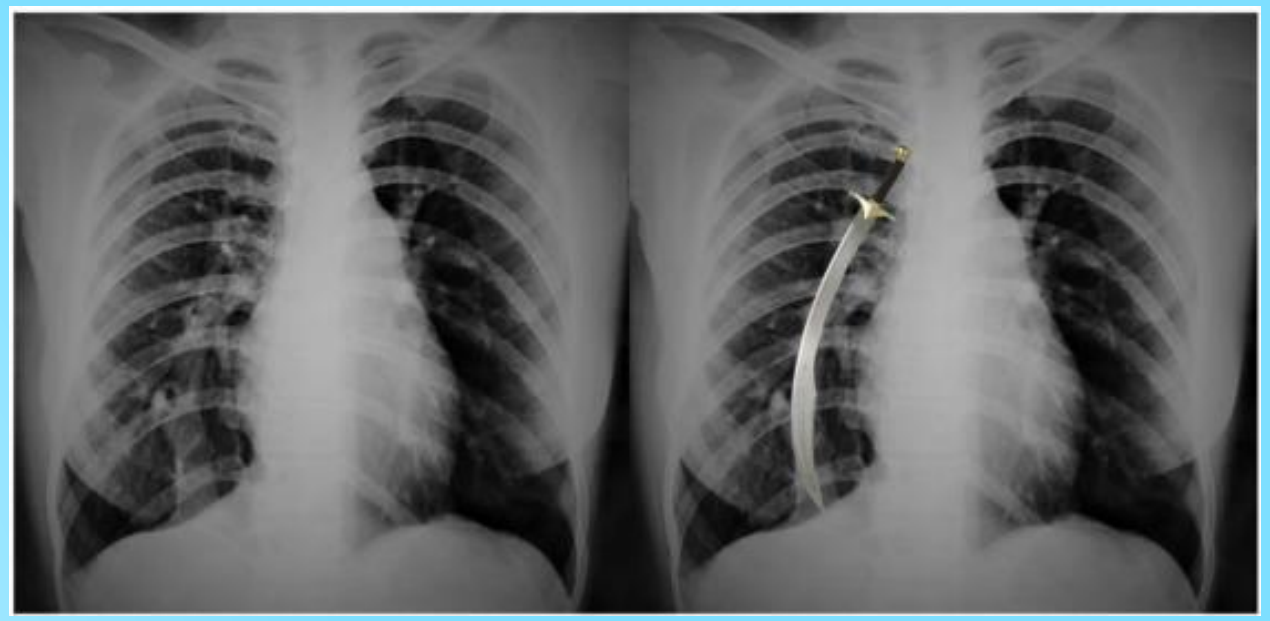
1. Partial or complete venous drainage of the right lung to the Inferior Vena Cava via a single curved vessel, shaped like a scimitar sword on CXR ★
2. Varying degrees of right lung and right pulmonary artery hypoplasia ★
3. Mesocardia or dextrocardia ★
4. Often associated with other heart defects (ASD, persistent PDA, VSD) ★

NIH National Library of Medicine; StatPearls, Josue Diaz-Frias, Jason Widrich

Scimitar Syndrome

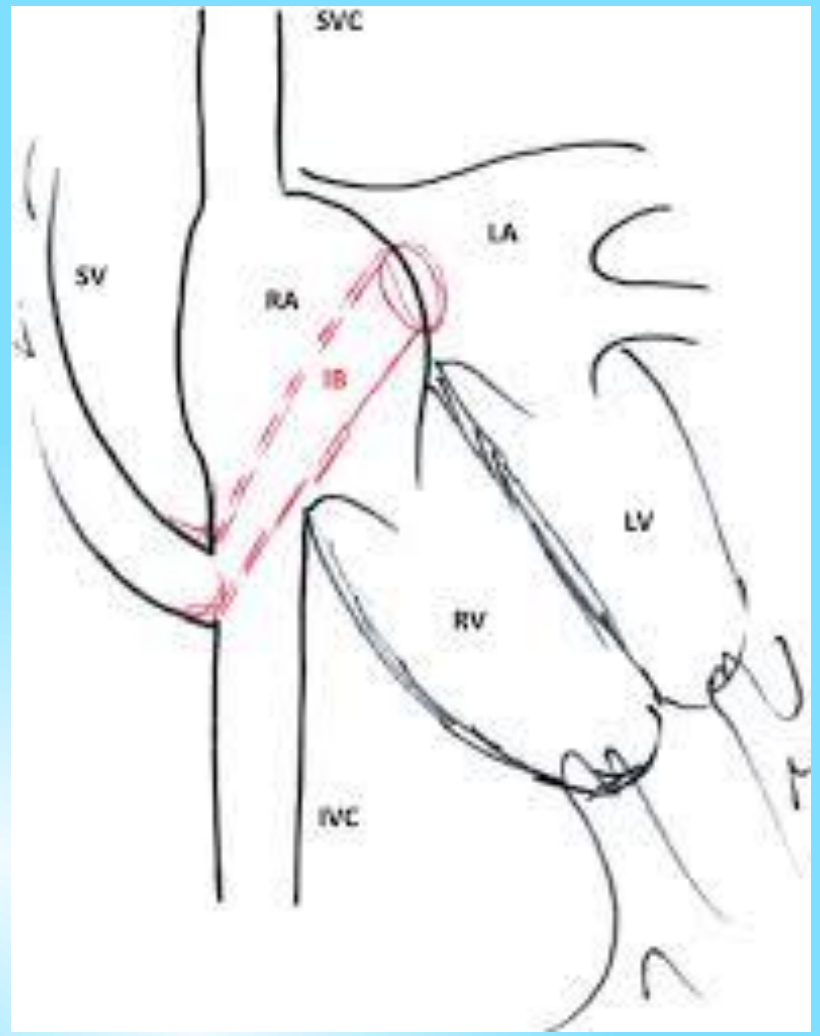


Science Direct: https://ars.els-cdn.com/content/image/3-s2.0-B9780128104064000035-f03-01-9780128104064.jpg

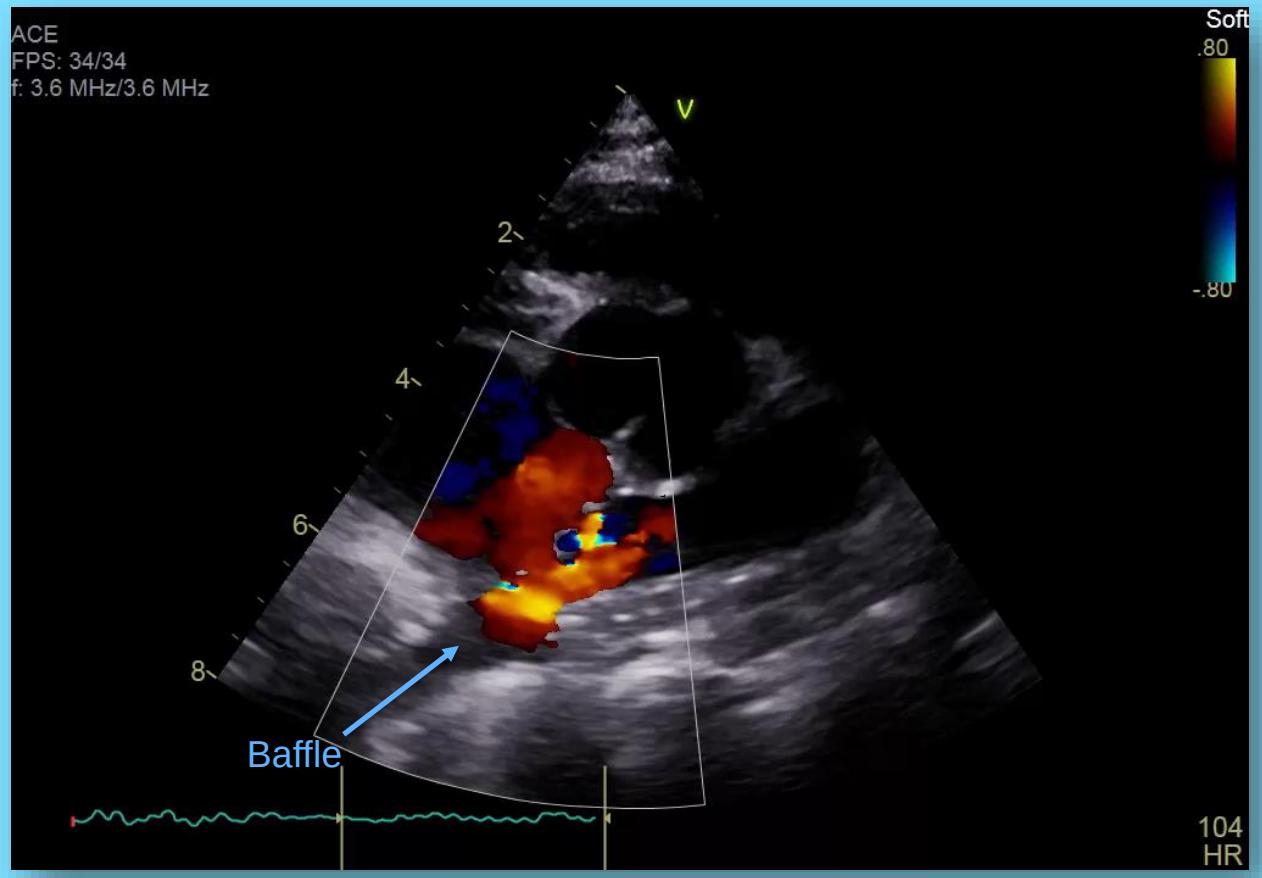


Science Direct: https://ars.els-cdn.com/content/image/1-s2.0-S1930043322002515-gr2.jpg

Scimitar Surgical Repair (PSAX)



Wiley Online Library- Werner Budts, MD https://encrypted tbn0.gstatic.com/images?q=tbn:ANd9GcRDhbh8Z1uS5HI_I-Bd_IEqpzTzYWzajwXX6A&s



- Surgical repair at ~2.5 years of age
- Currently 4 years old with no post-operative issues
- Will follow up to evaluate for baffle stenosis/leak

Recap of Important Information To Obtain For Pulmonary Hypertension

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- Presence of interventricular septal flattening
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