Case Presentation

November 3, 2018

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Cleveland Clinic Children's

What’s That Murmur?

History

- 2 year old Amish child taken to pediatrician by his mother when she noticed chest asymmetry
- Murmur and failure to thrive noted by pediatrician
- Child referred to pediatric cardiology

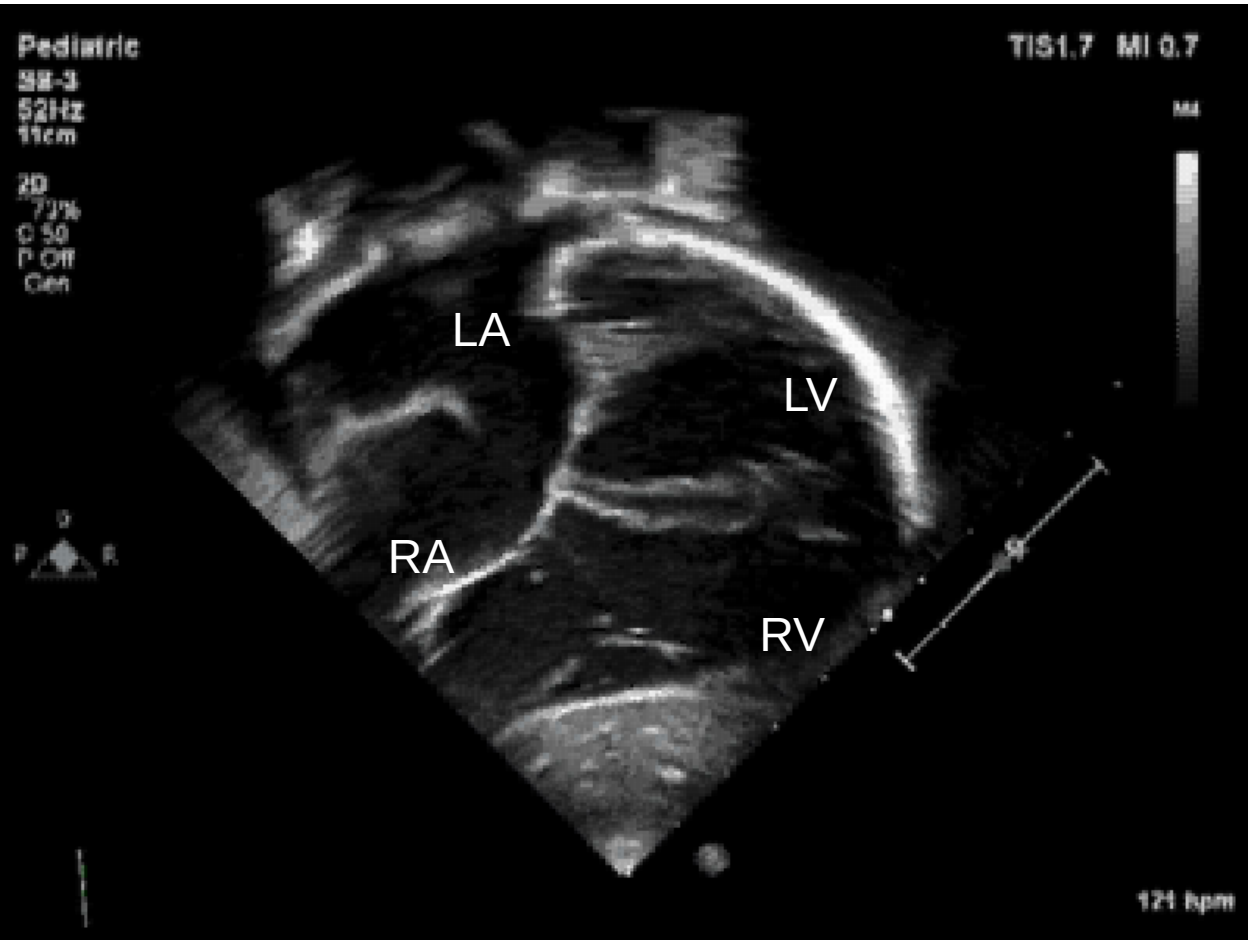
Exam

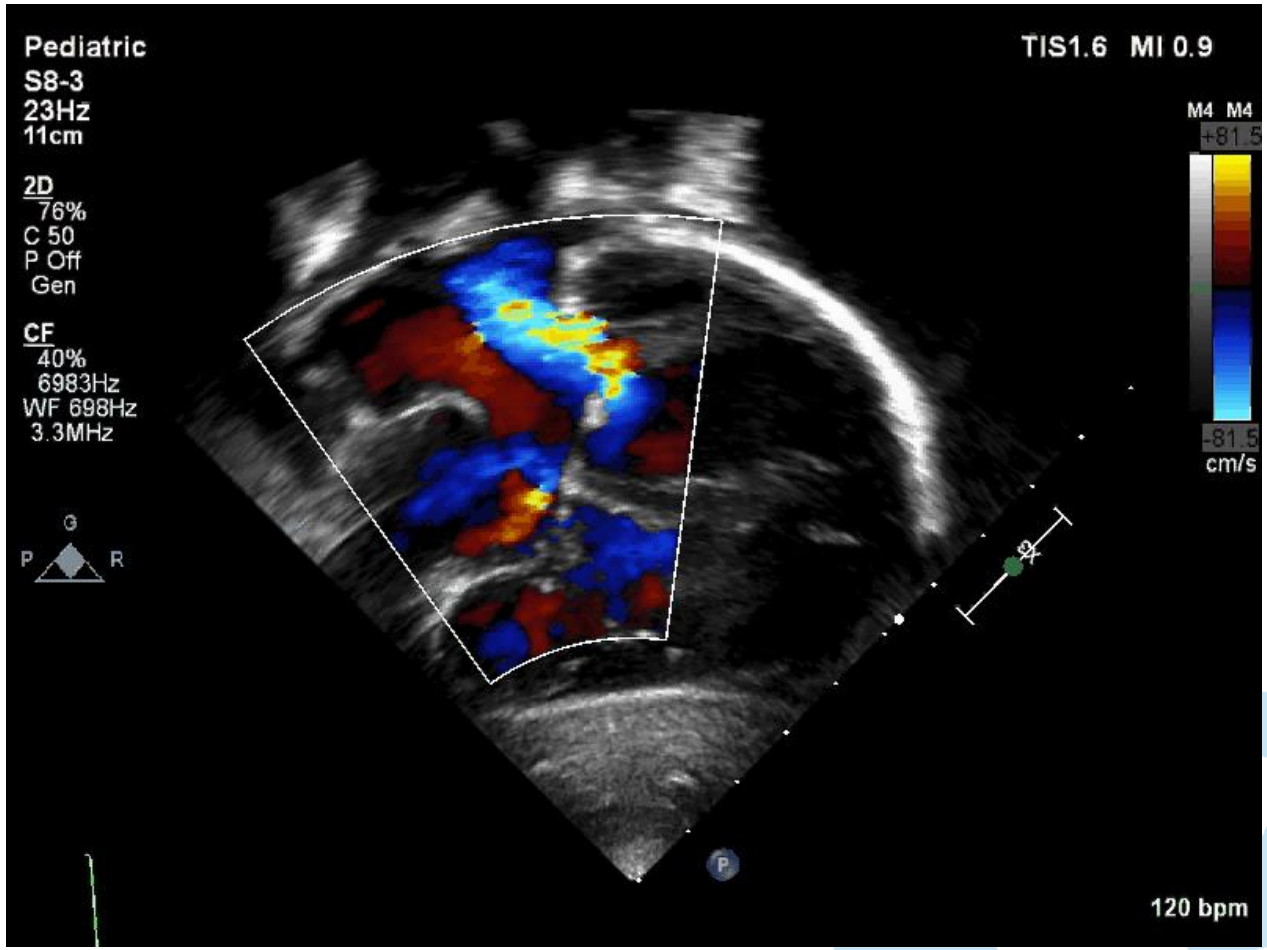
- 86cm (11th percentile), 11kg (2nd percentile), BP 88/60, O2 sat 99%
- Hyperdynamic precordium
- Normal S1, widely split S2, normal P2
- Harsh 3/6 vibratory, low pitched systolic ejection murmur best heard at left upper sternal border
- 2/6 medium pitched regurgitant murmur best heard at lower sternal border
- Soft diastolic murmur best heard at left mid sternal border

- Chest X-ray showed cardiomegaly and increased pulmonary vascularity

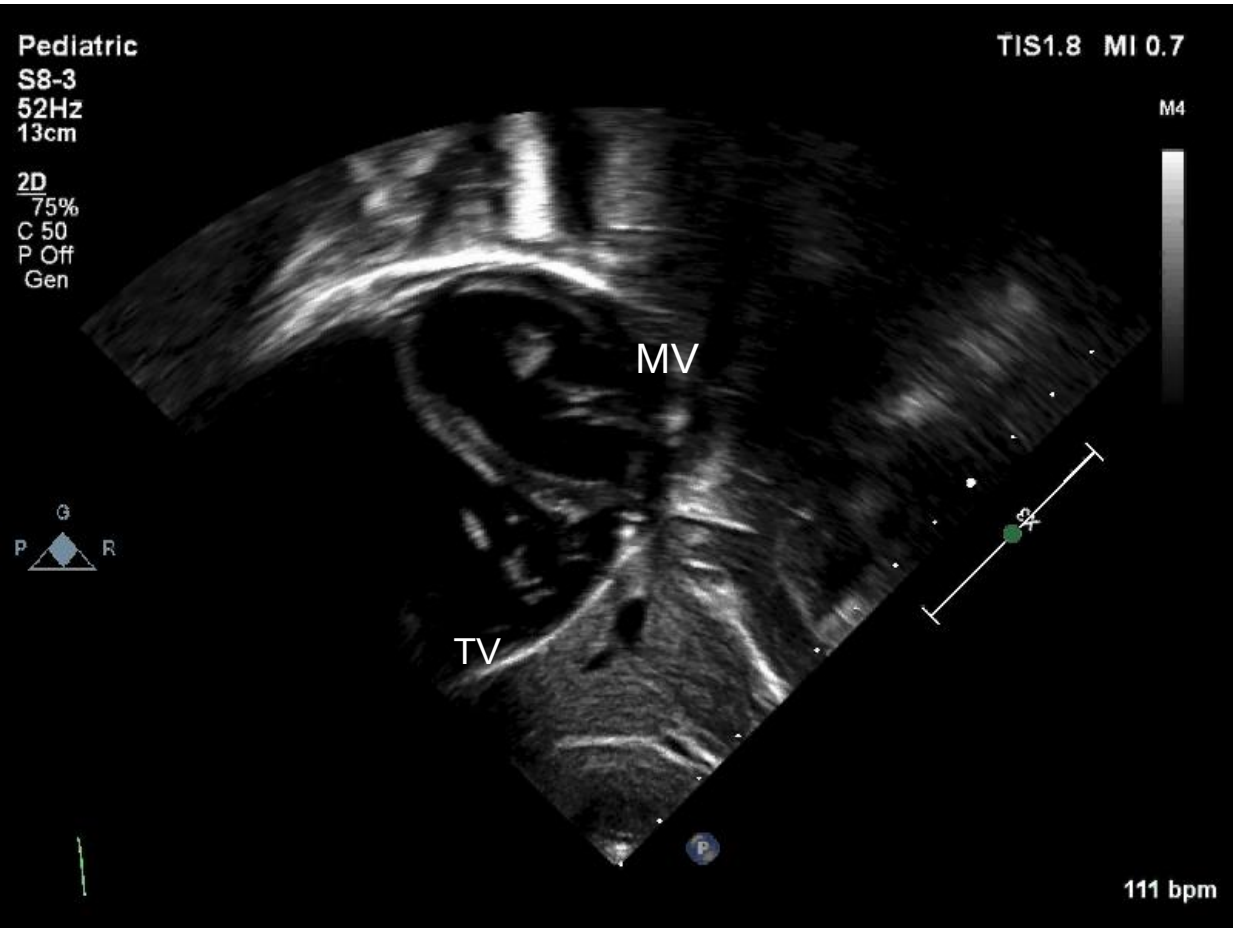
- An echocardiogram was ordered

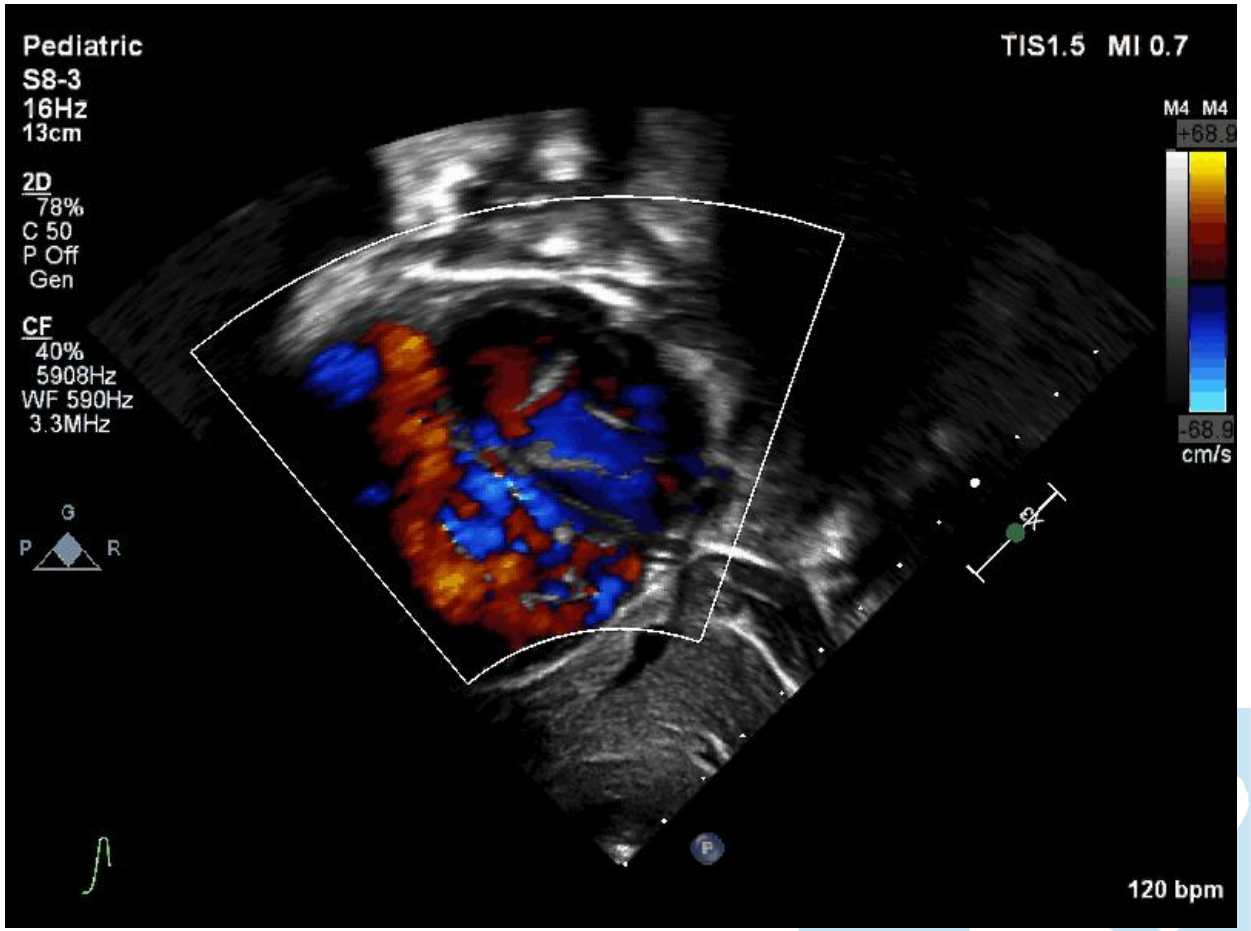
Subcostal Coronal View



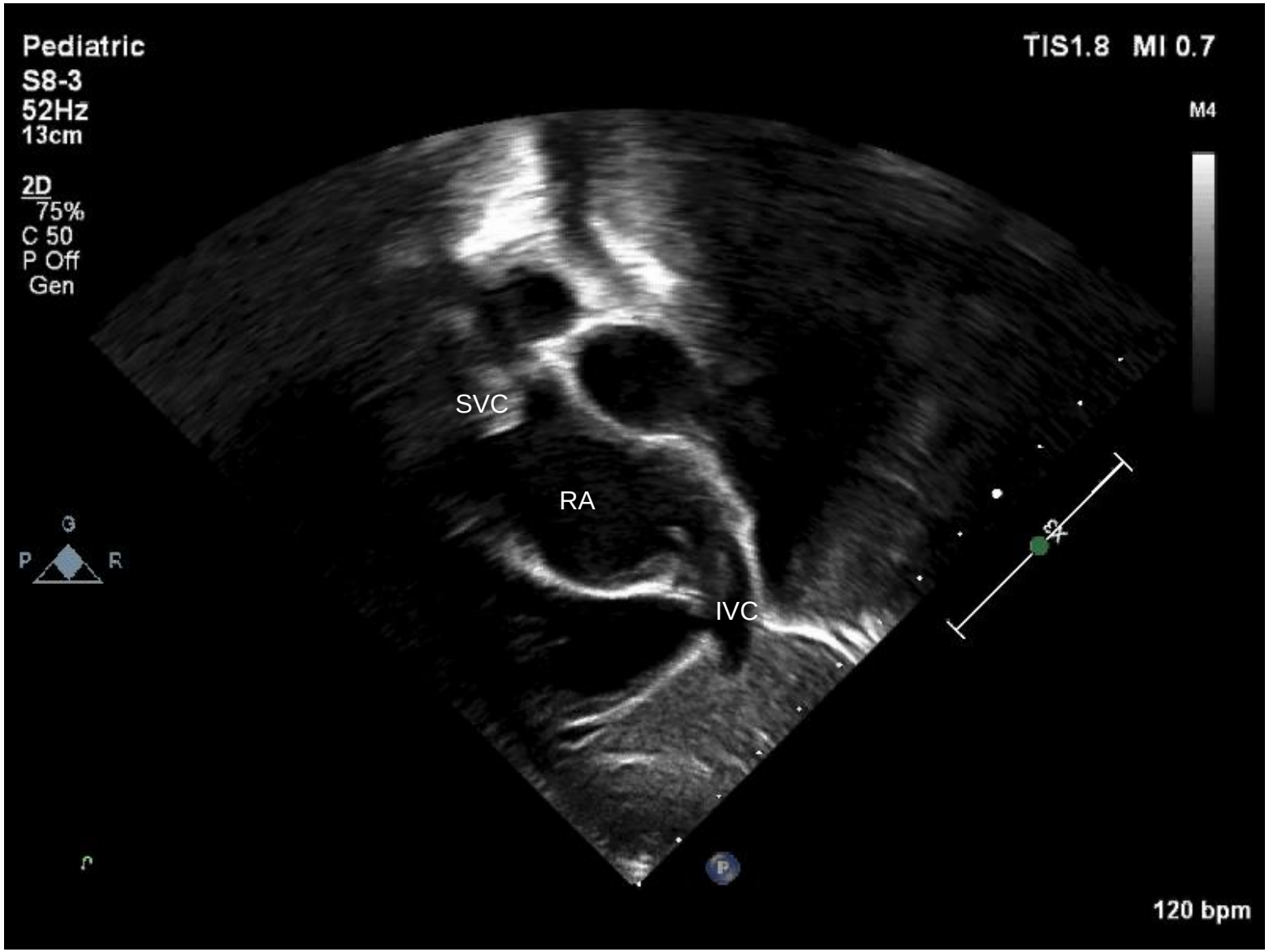


Subcostal Sagittal View

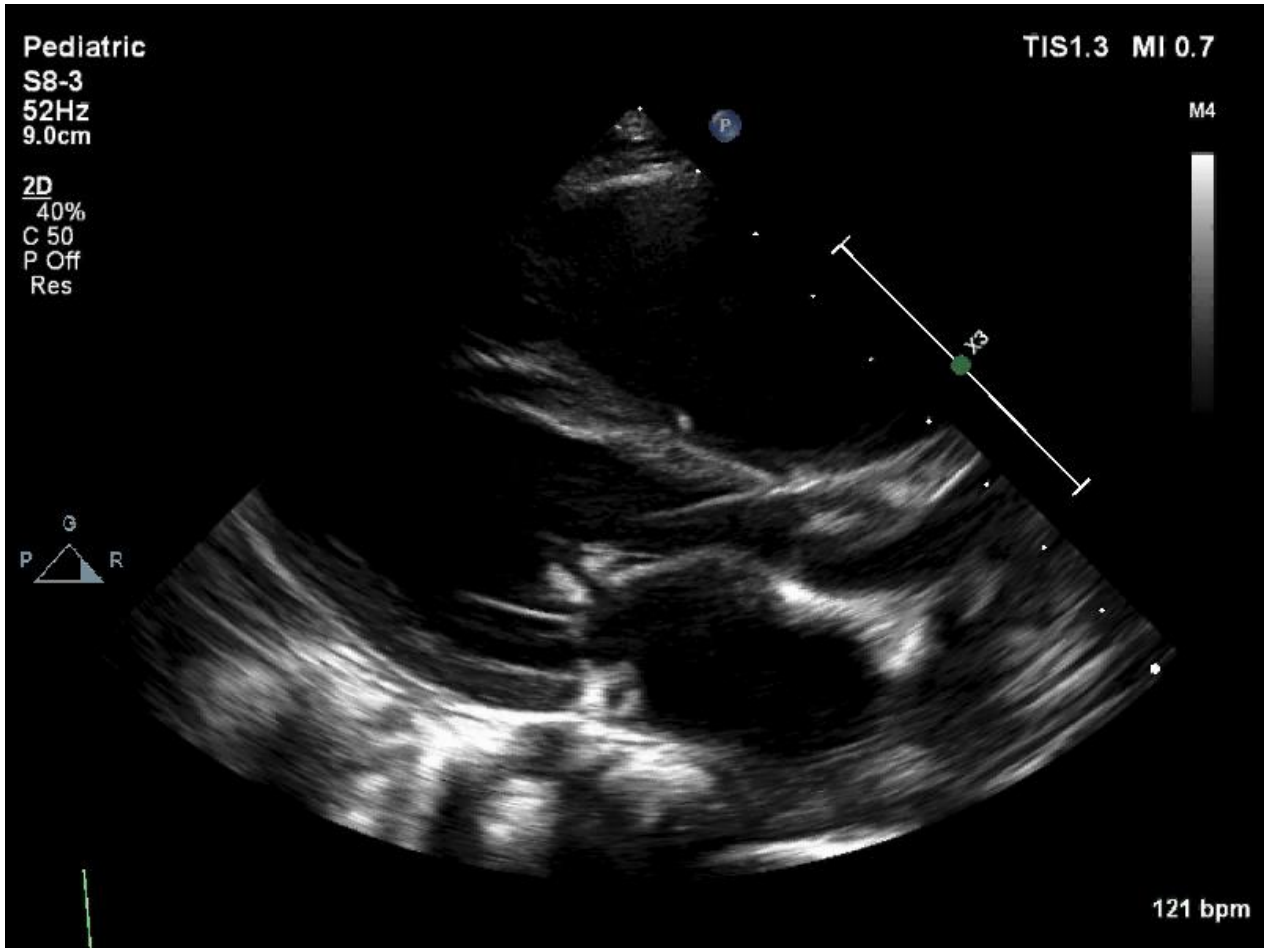


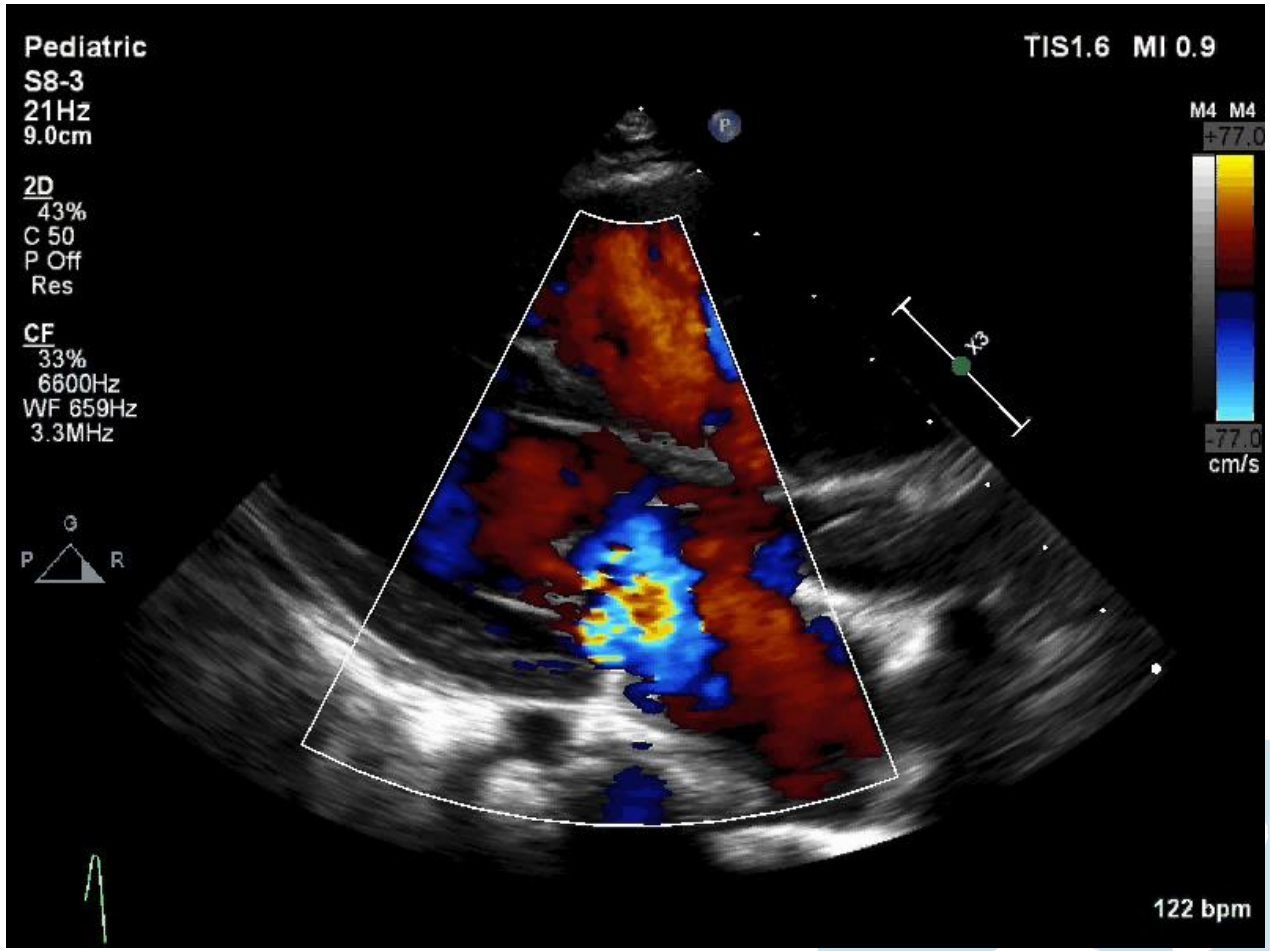


Subcostal Bicaval View

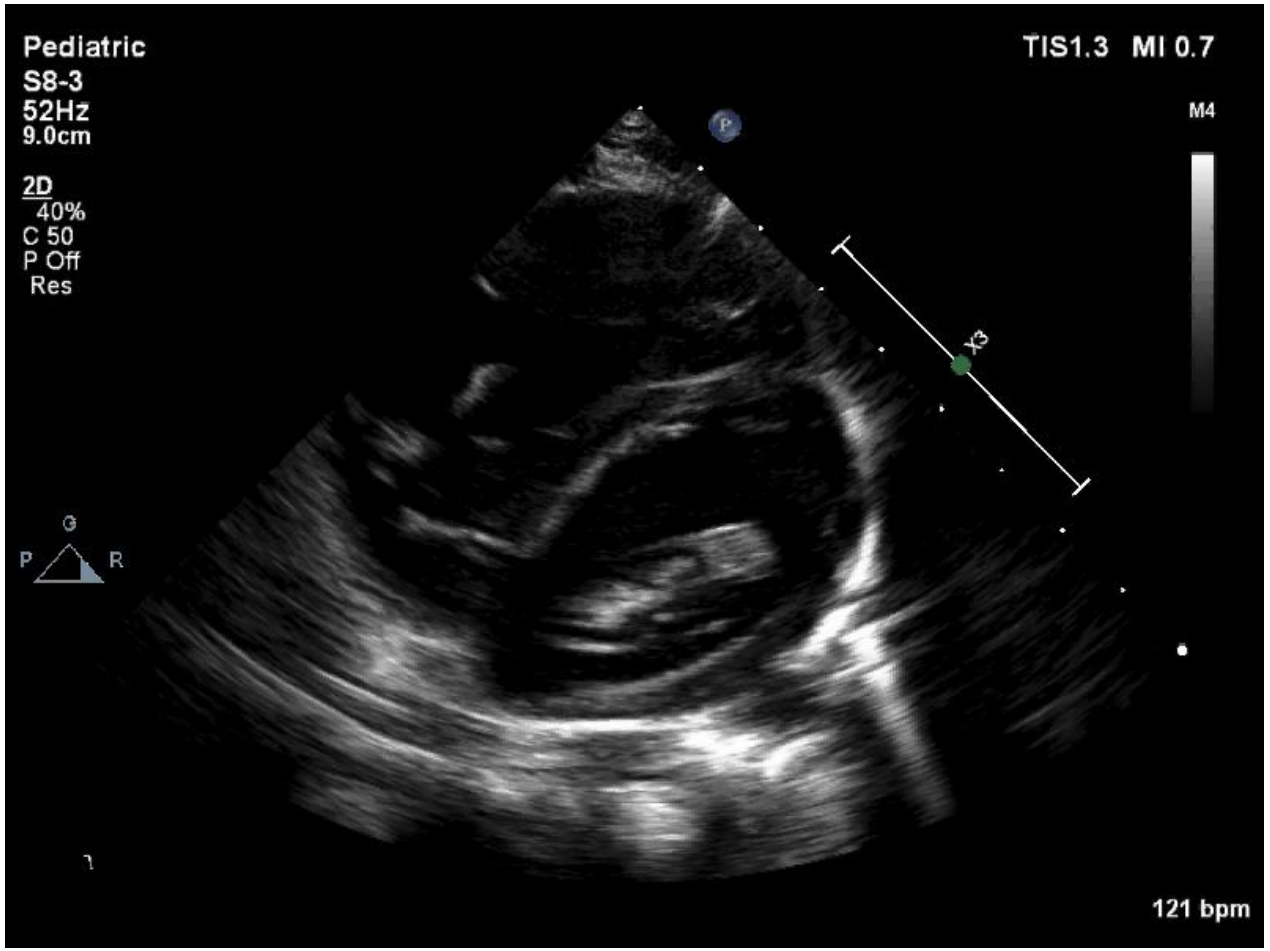


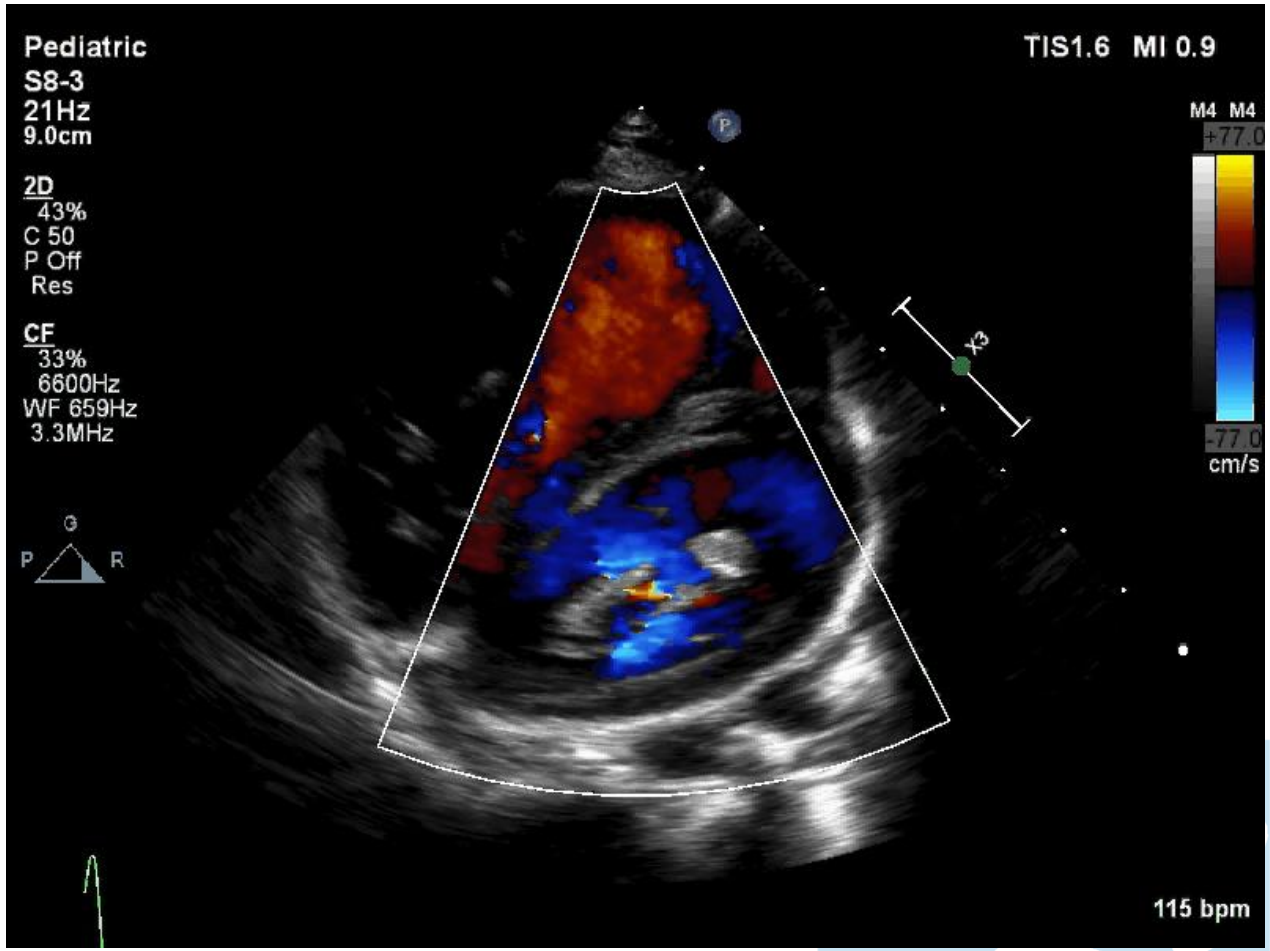
Parasternal Long Axis



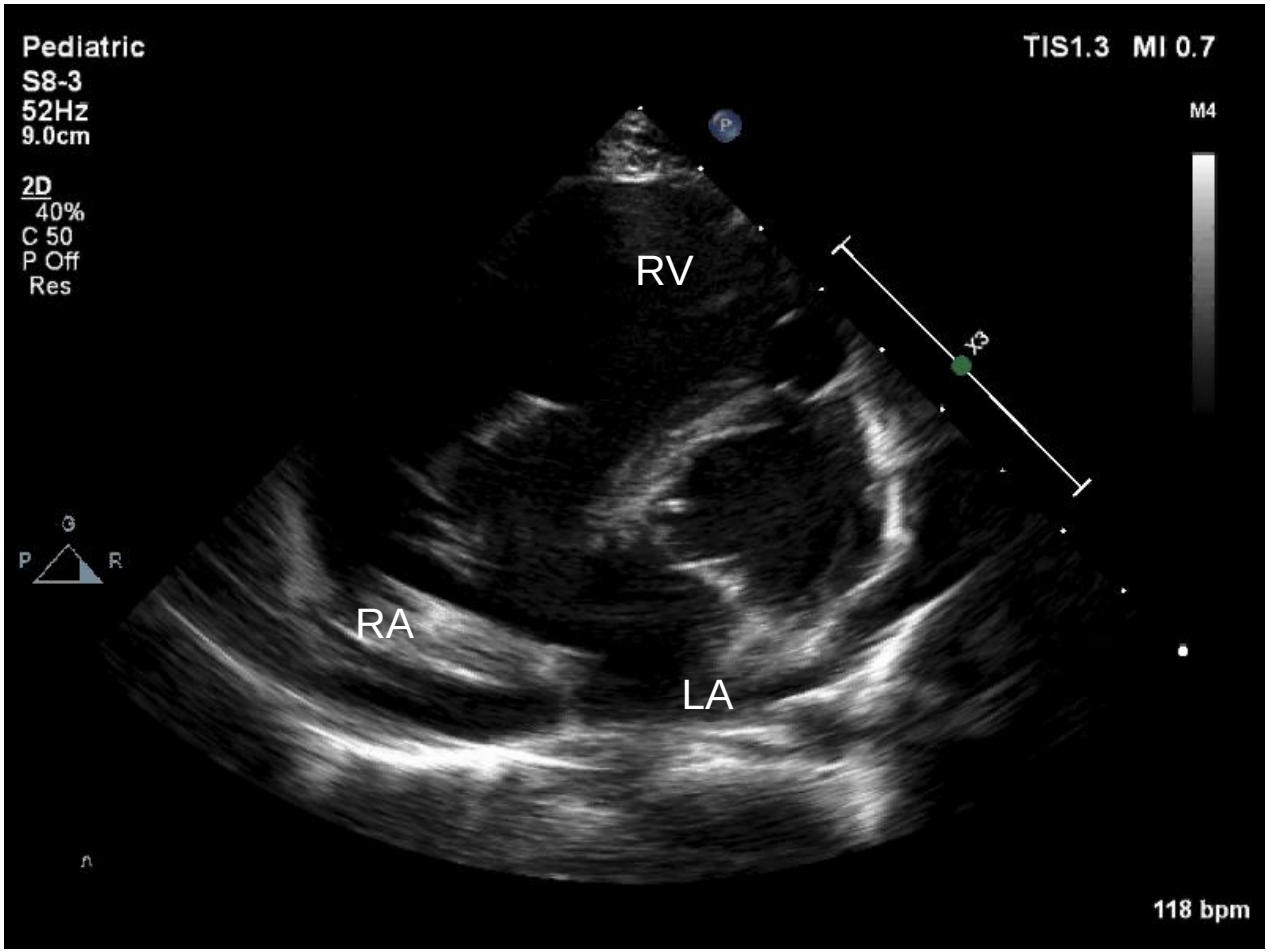


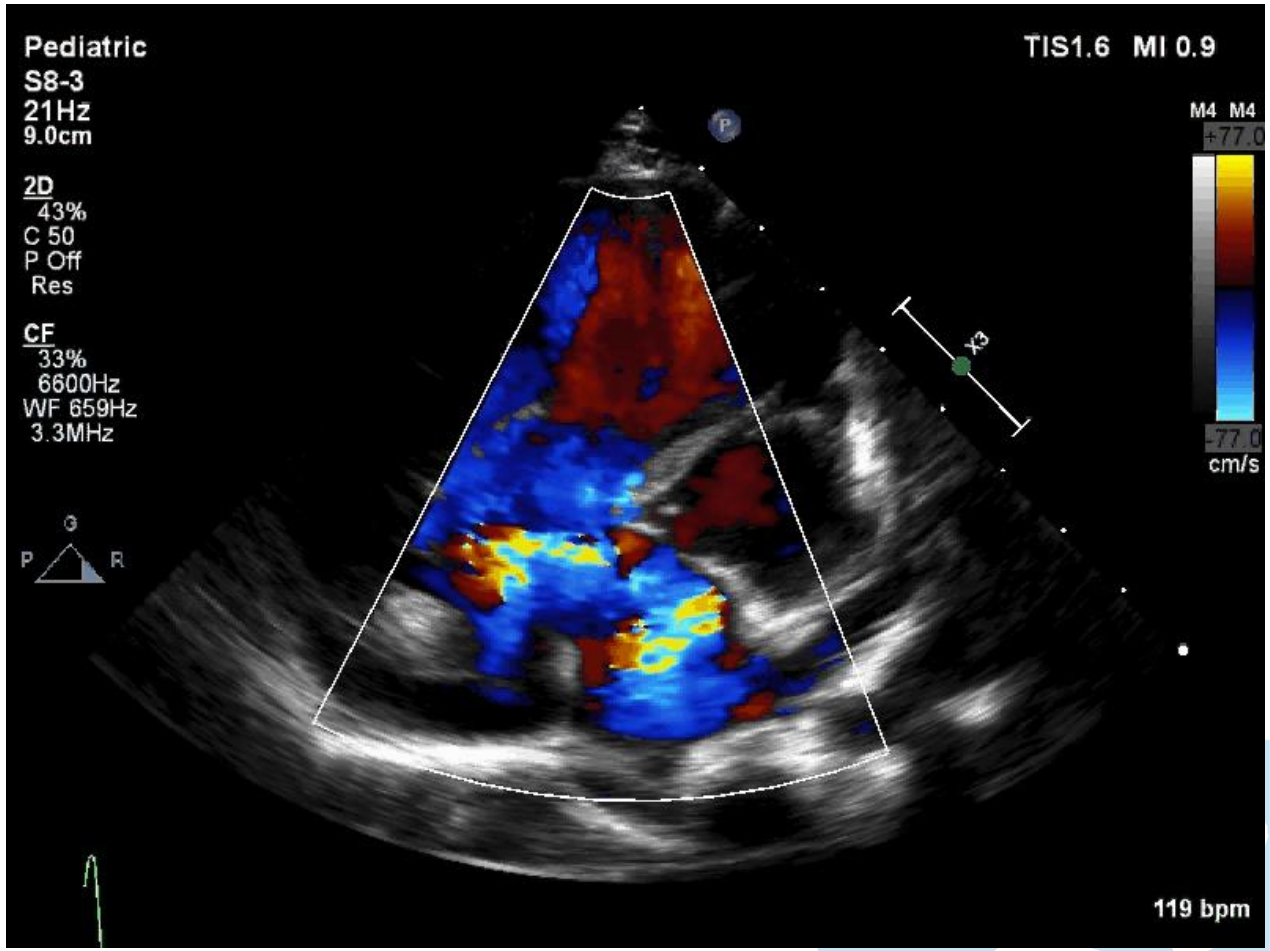
Parasternal Short Axis



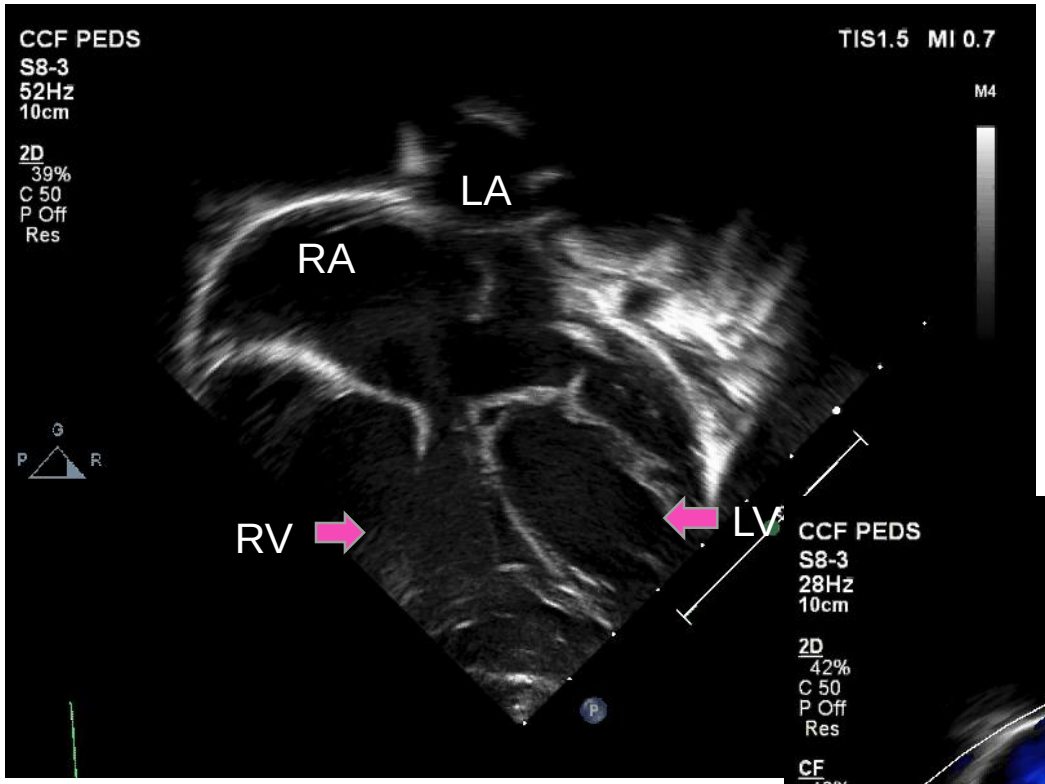


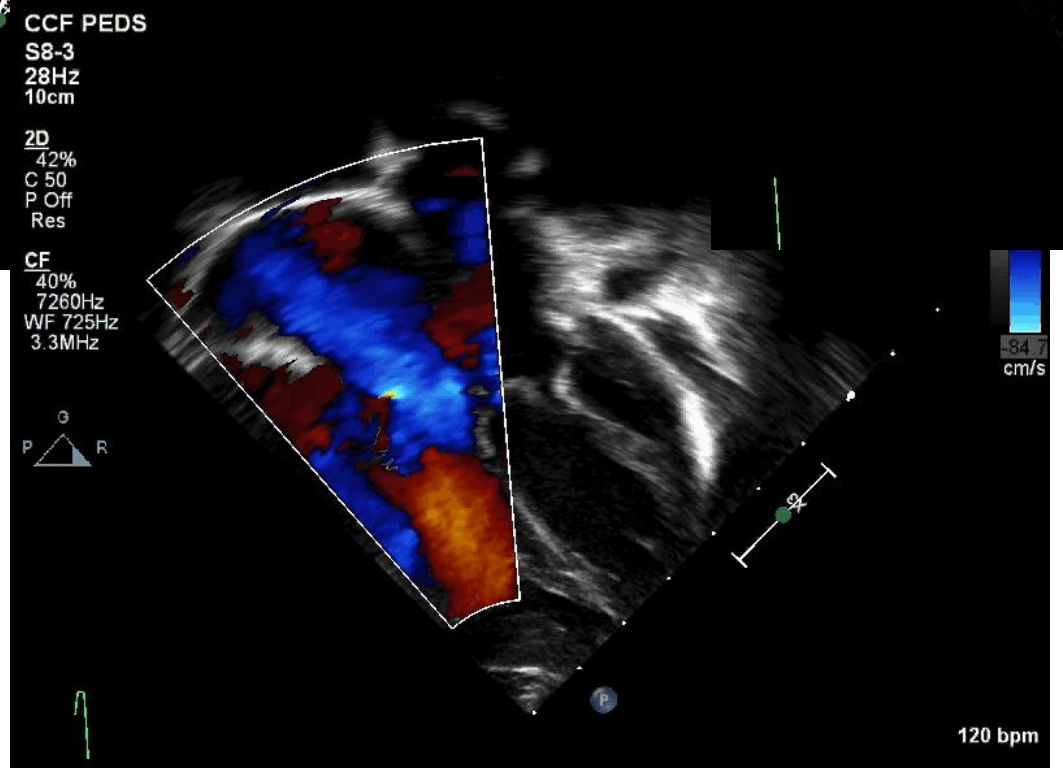
Parasternal “Off-Axis” View

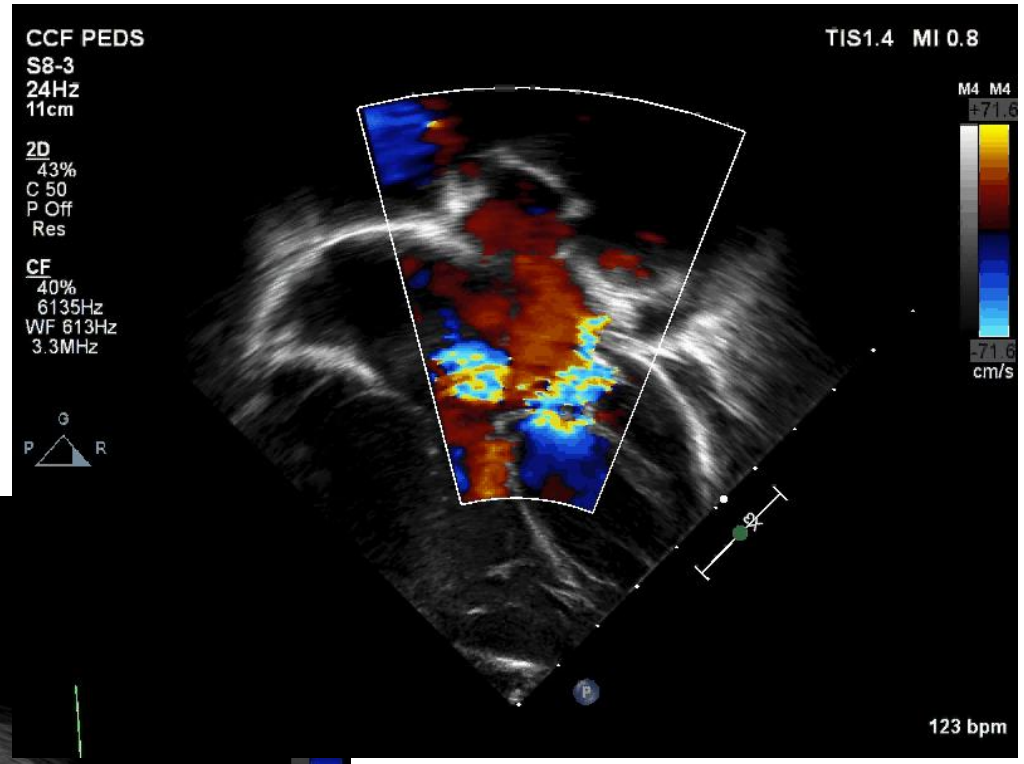




Apical Four Chamber View

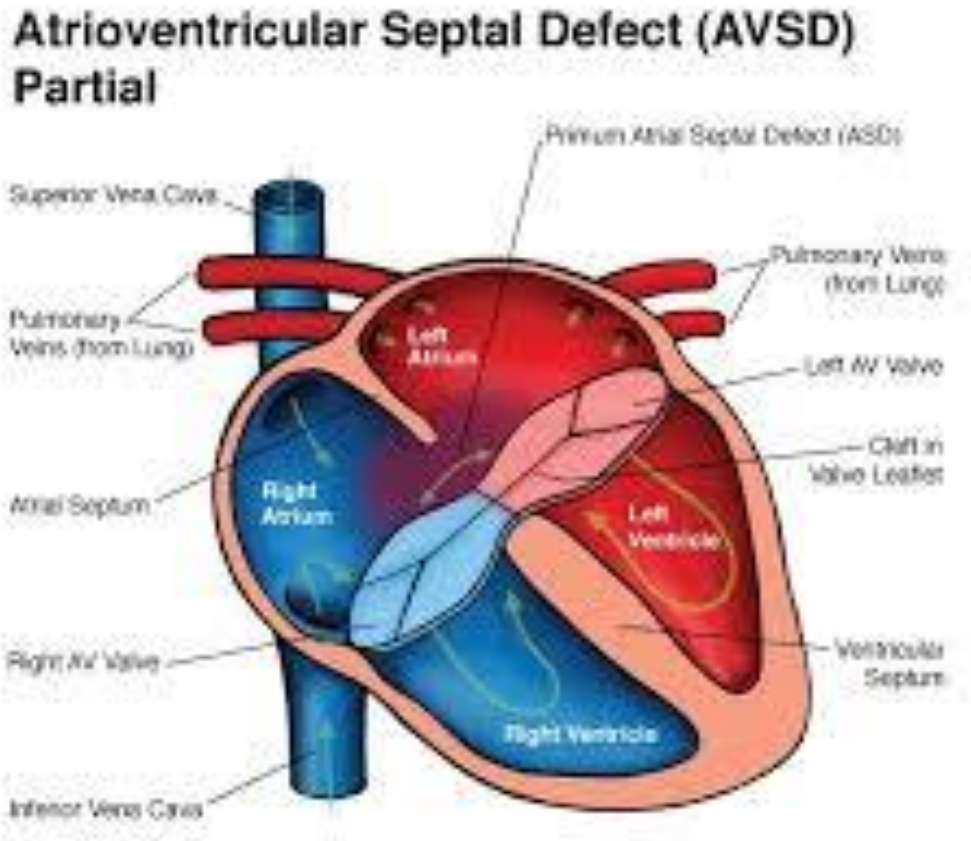






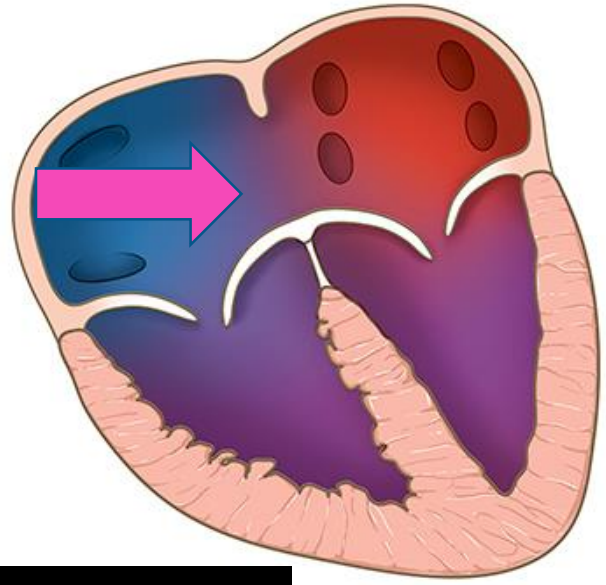
So what did we find??

Partial AV Septal Defect (or Partial AV Canal)

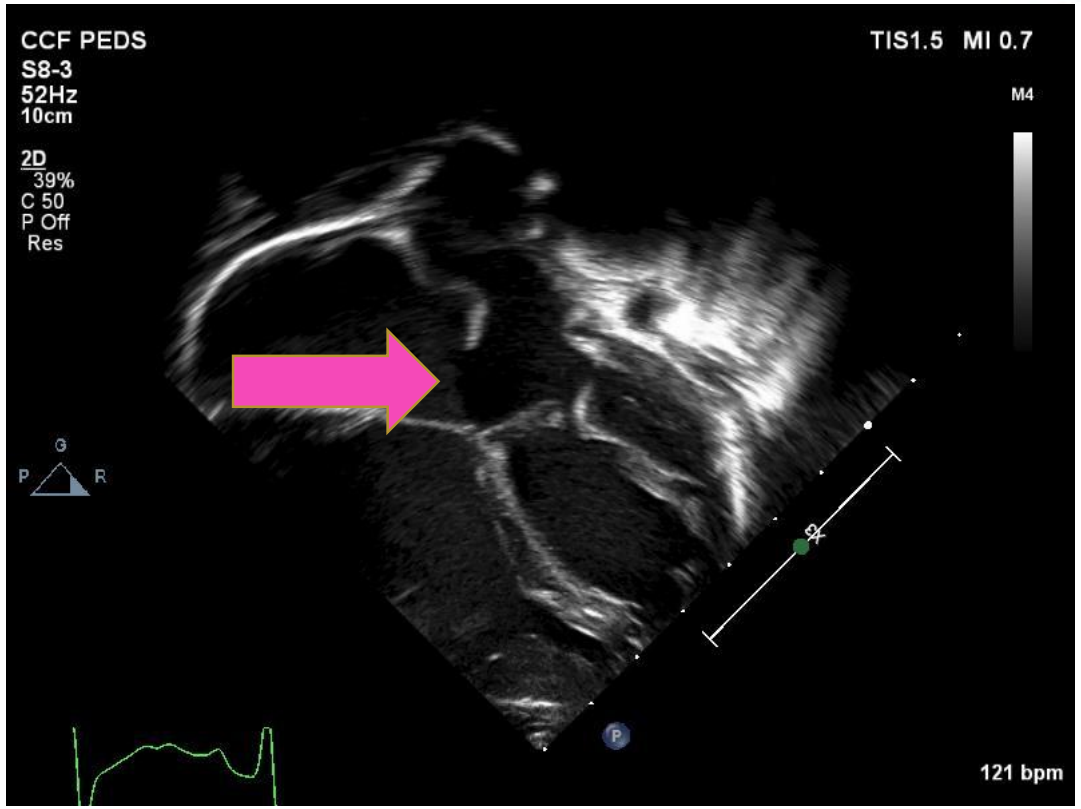


- Primum ASD
- Cleft mitral valve

Primum Atrial Septal Defect

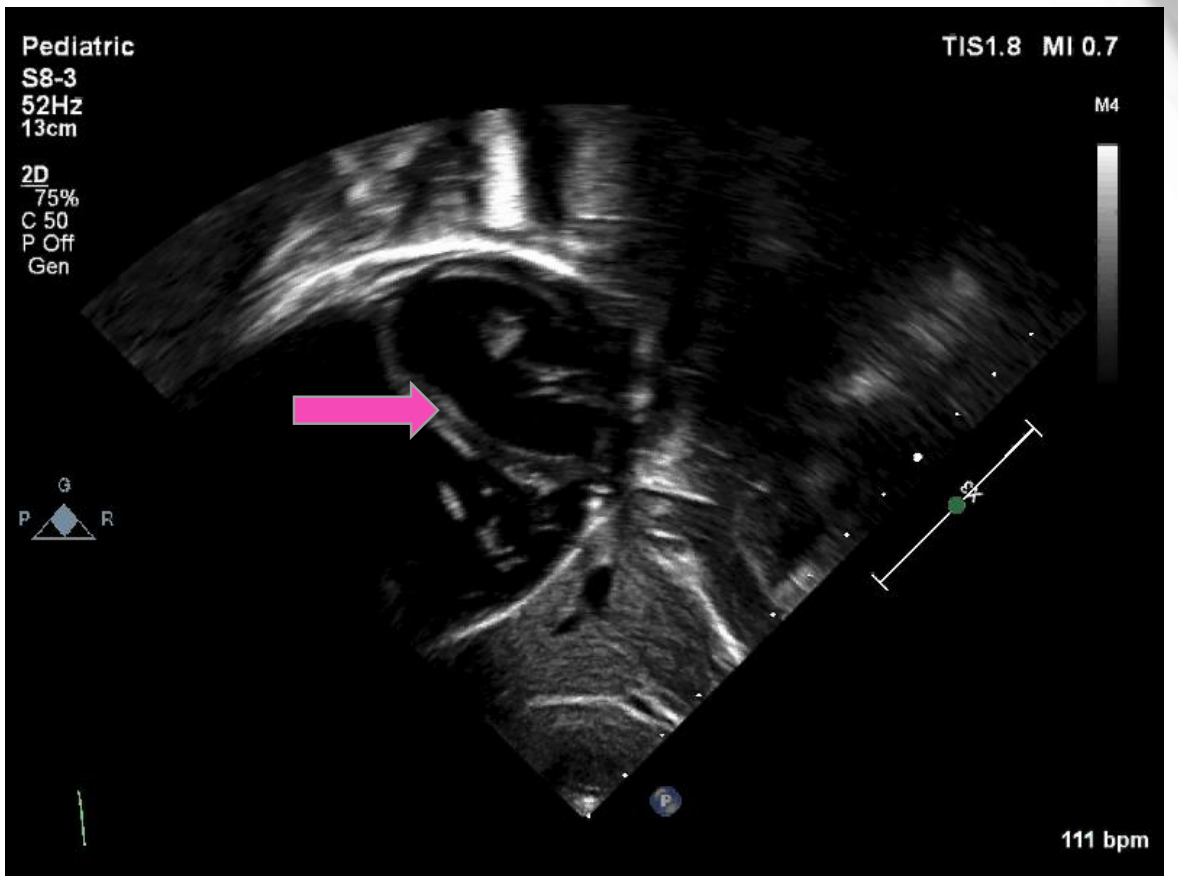


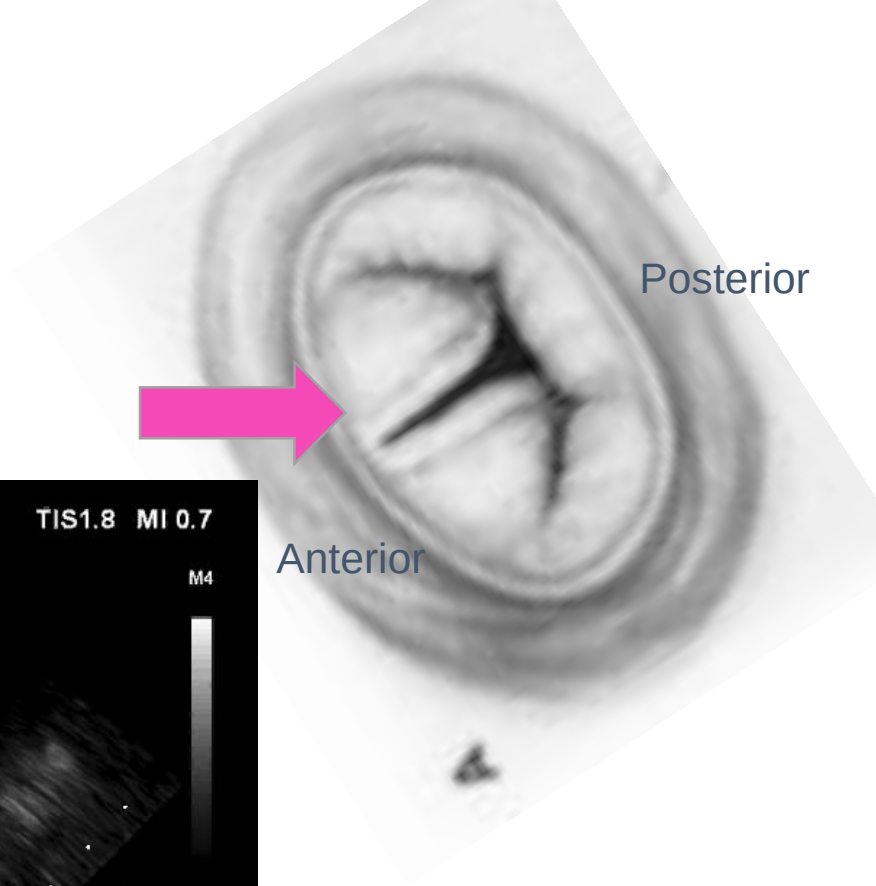
- Located in the lower portion of the atrial septum
- AV valves are at the same level in an apical 4 chamber view
- Usually associated with a cleft in the anterior mitral leaflet



Cleft Mitral Valve

- Defined as a division or slit in the anterior mitral valve leaflet
- Results in mitral regurgitation

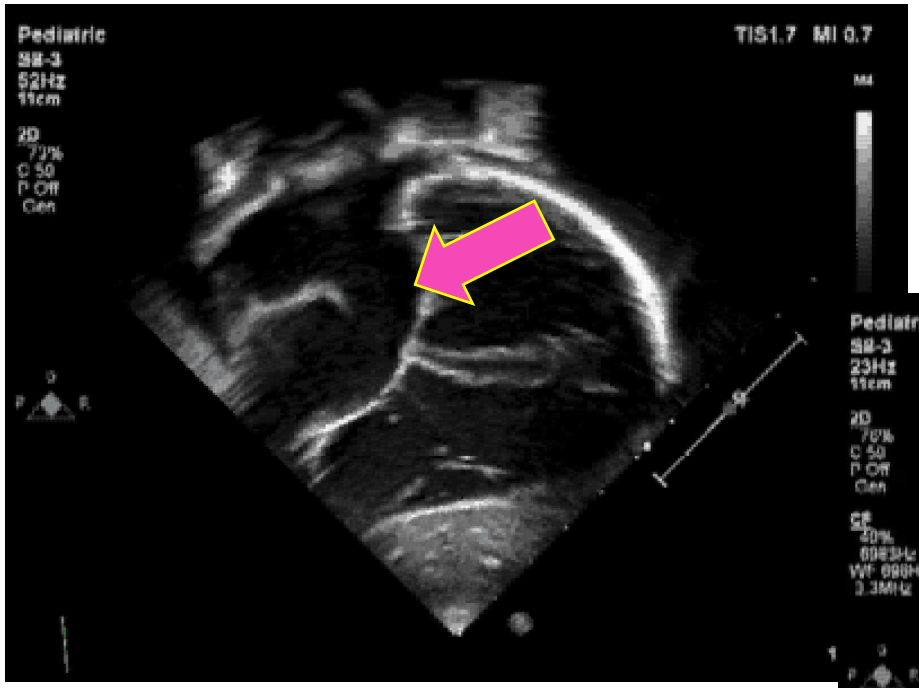


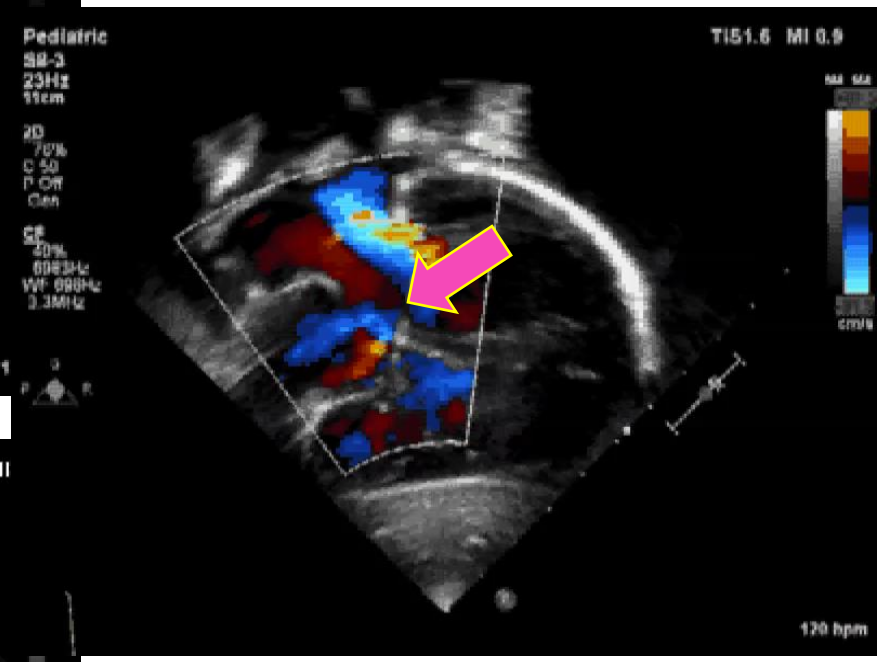


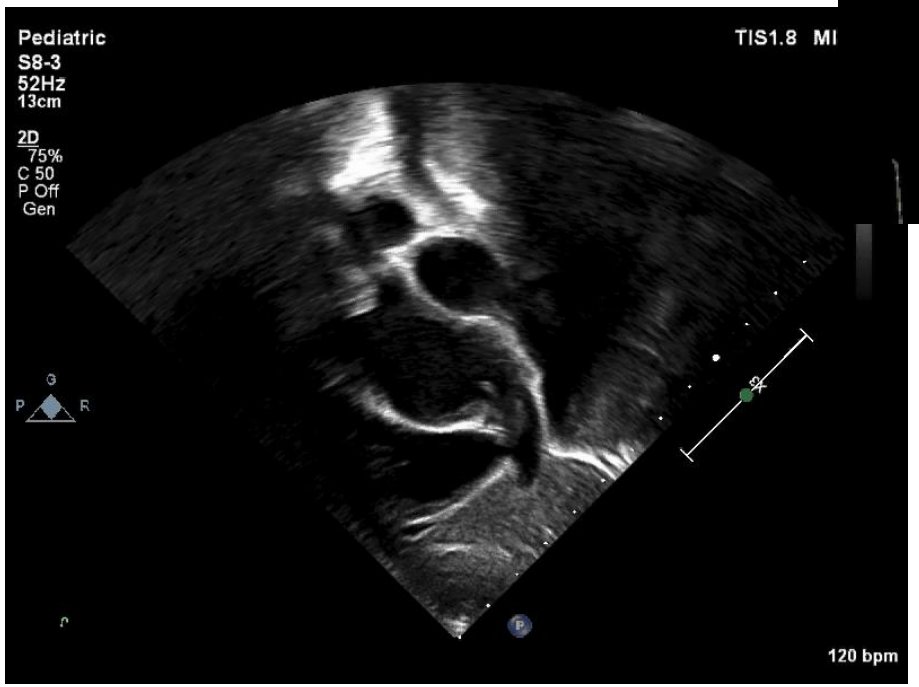
Echocardiographic Assessment of Partial AV Canal

Evaluate Atrial Septum

- Determine the size of the primum ASD
- Assess the direction of shunt with color Doppler
  - Spectral Doppler can be helpful in determining if shunting is bidirectional
- Assess for additional defects

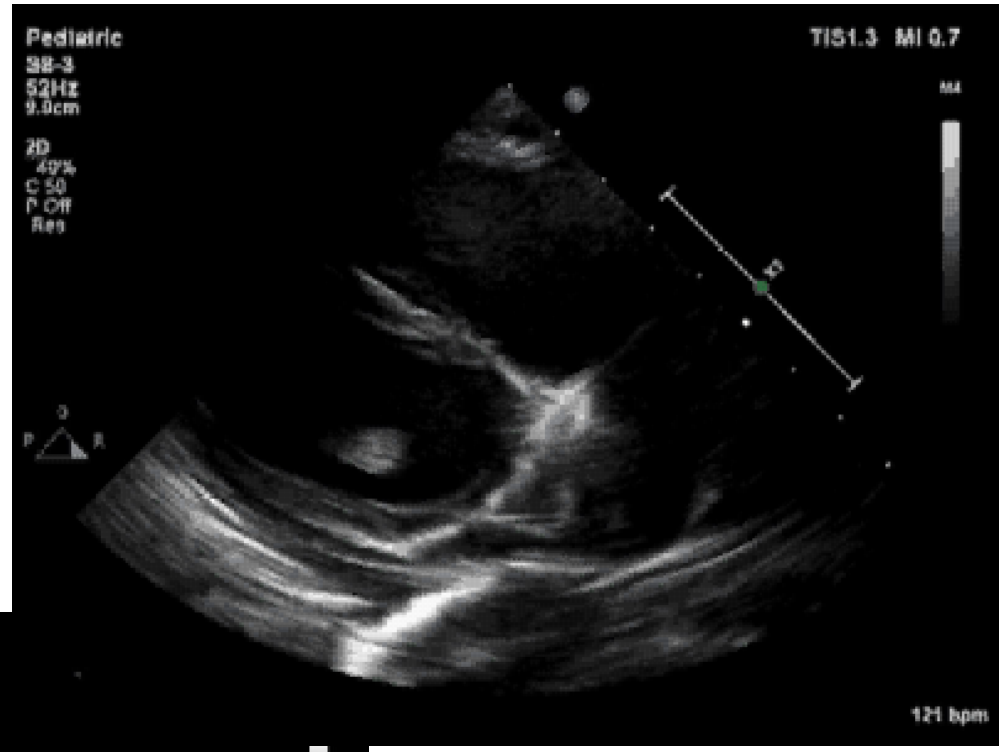


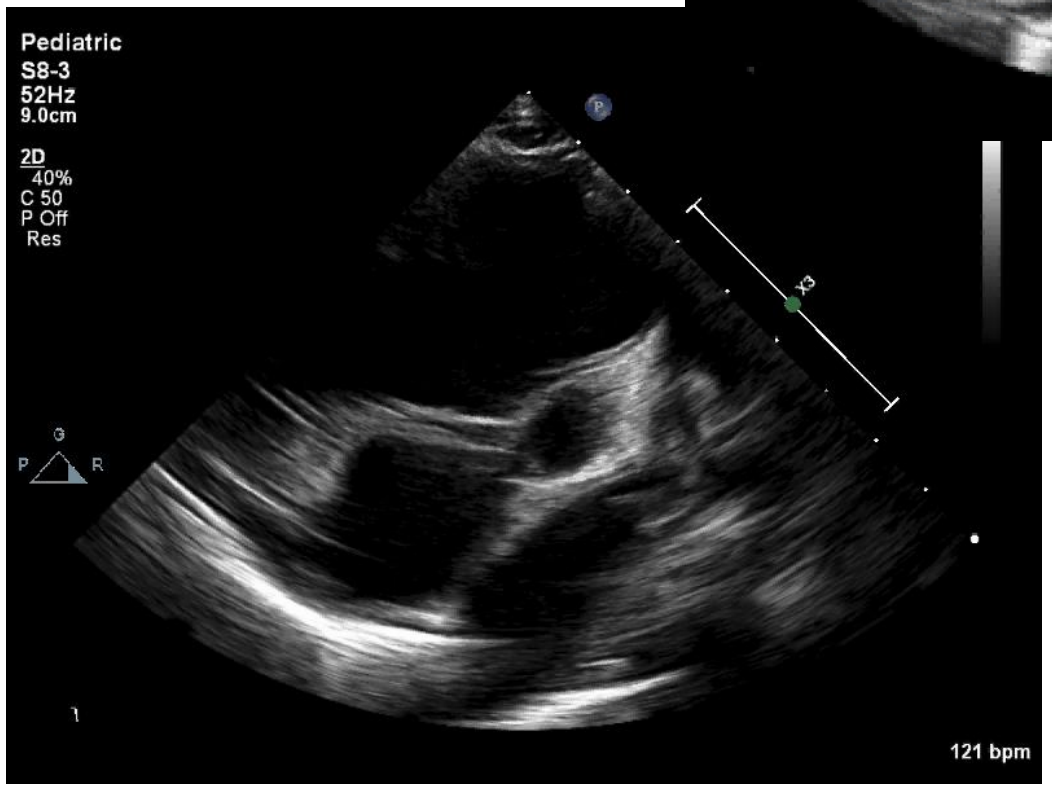




Evaluate Ventricular Septum

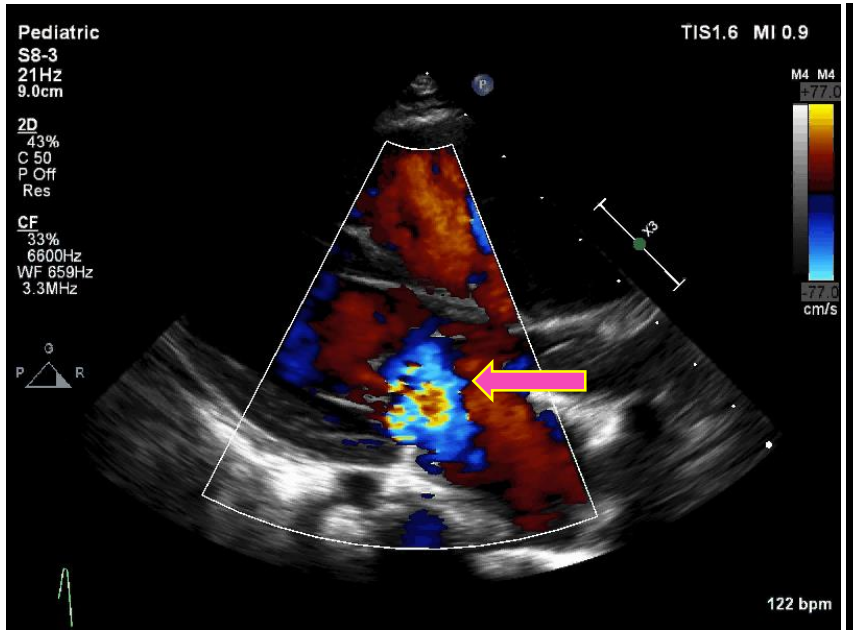
- Assess for ventricular level shunting
- Parasternal long and short axis sweeps
- “Partial” denotes the absence of a VSD

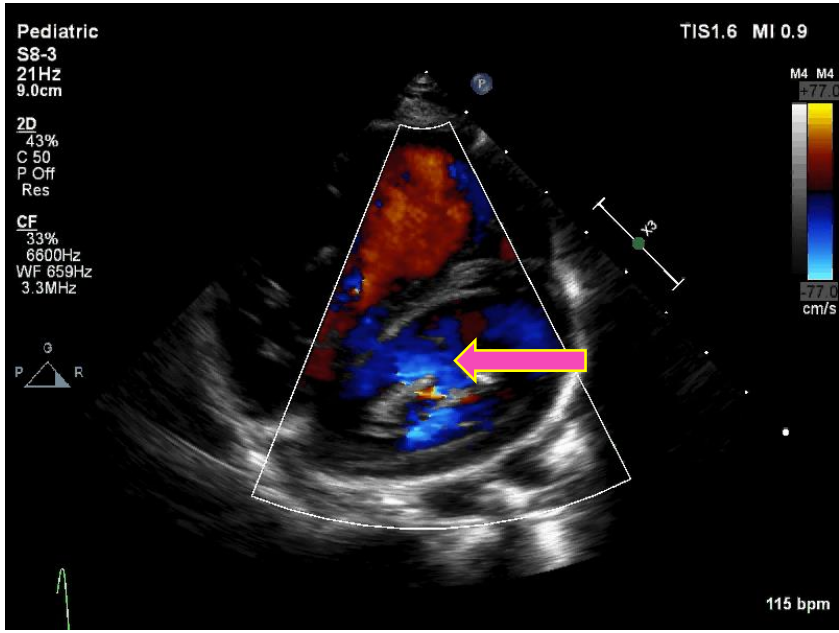


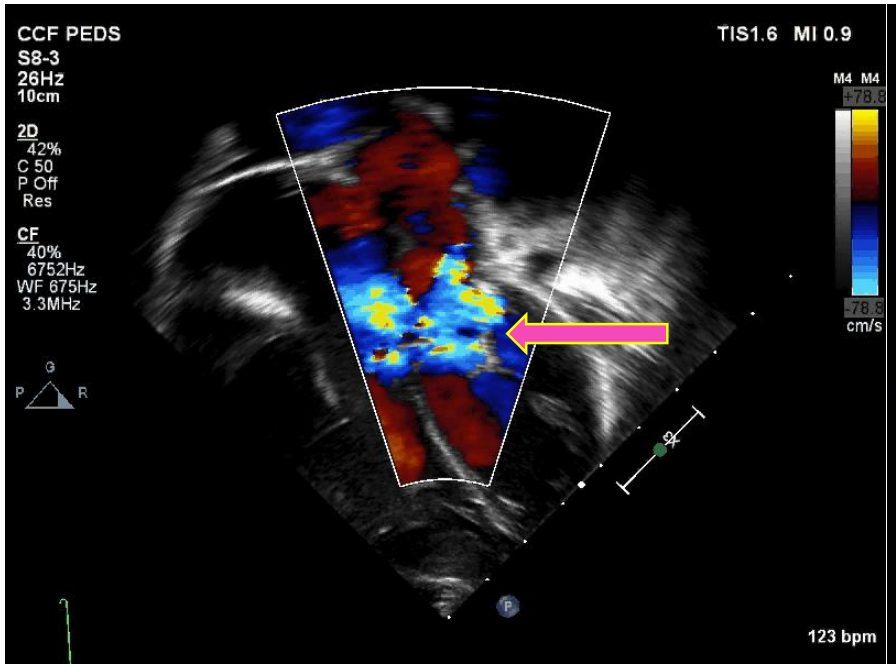


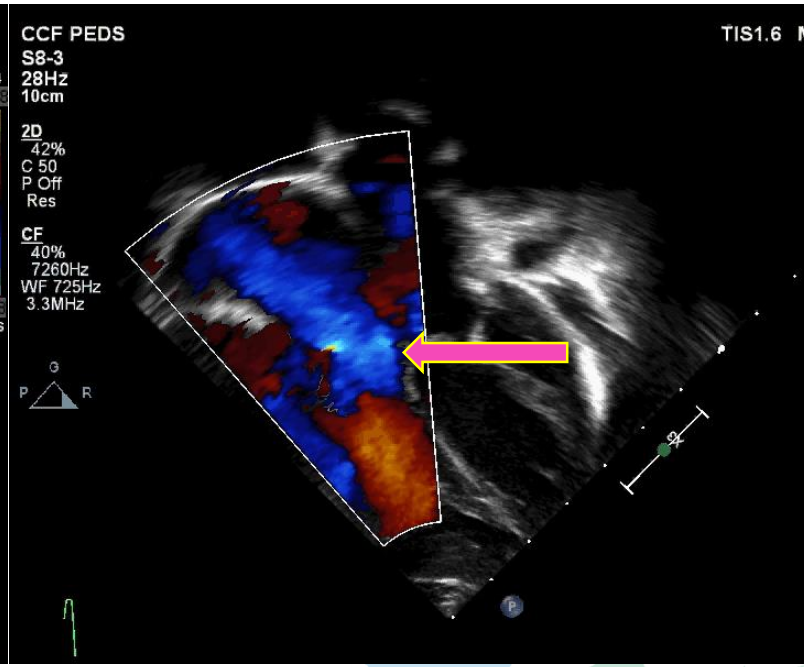
Evaluate AV valves

- Color flow and spectral Doppler to determine presence of regurgitation
- Quantify degree of regurgitation
- Tricuspid regurgitation velocity for RV pressure



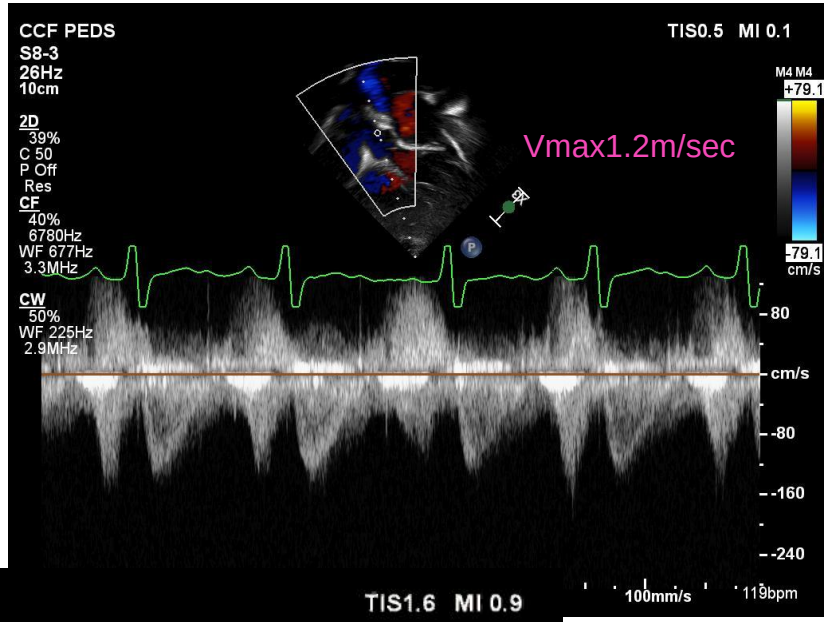


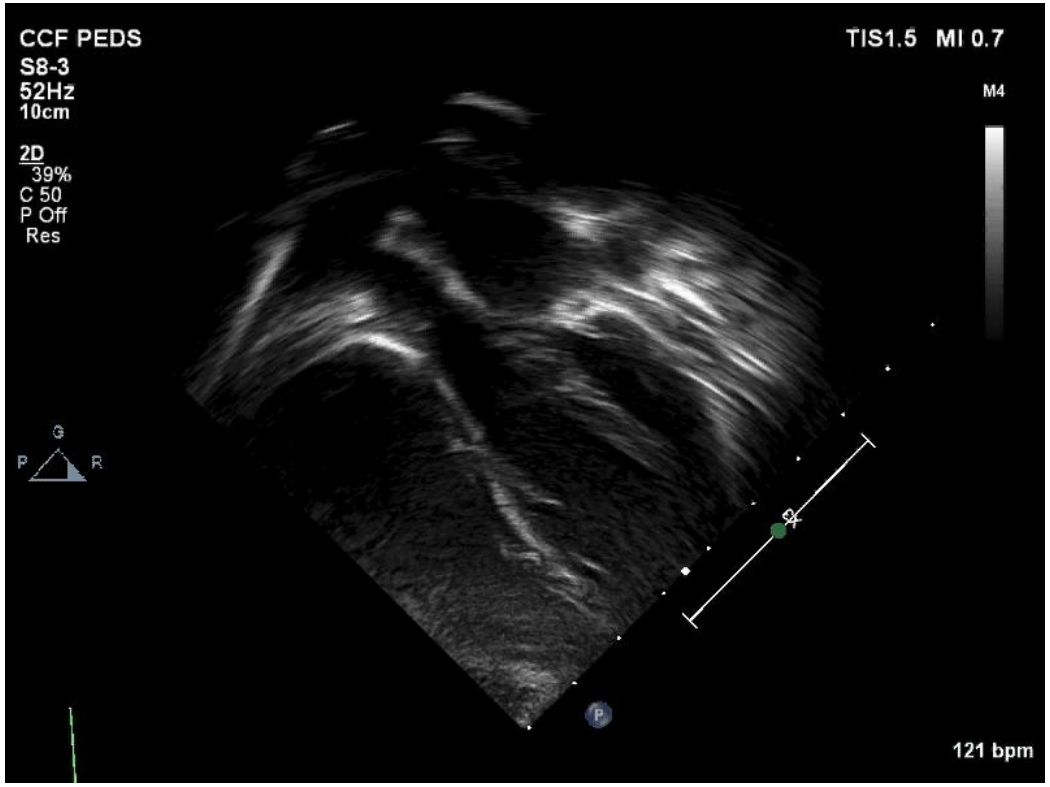


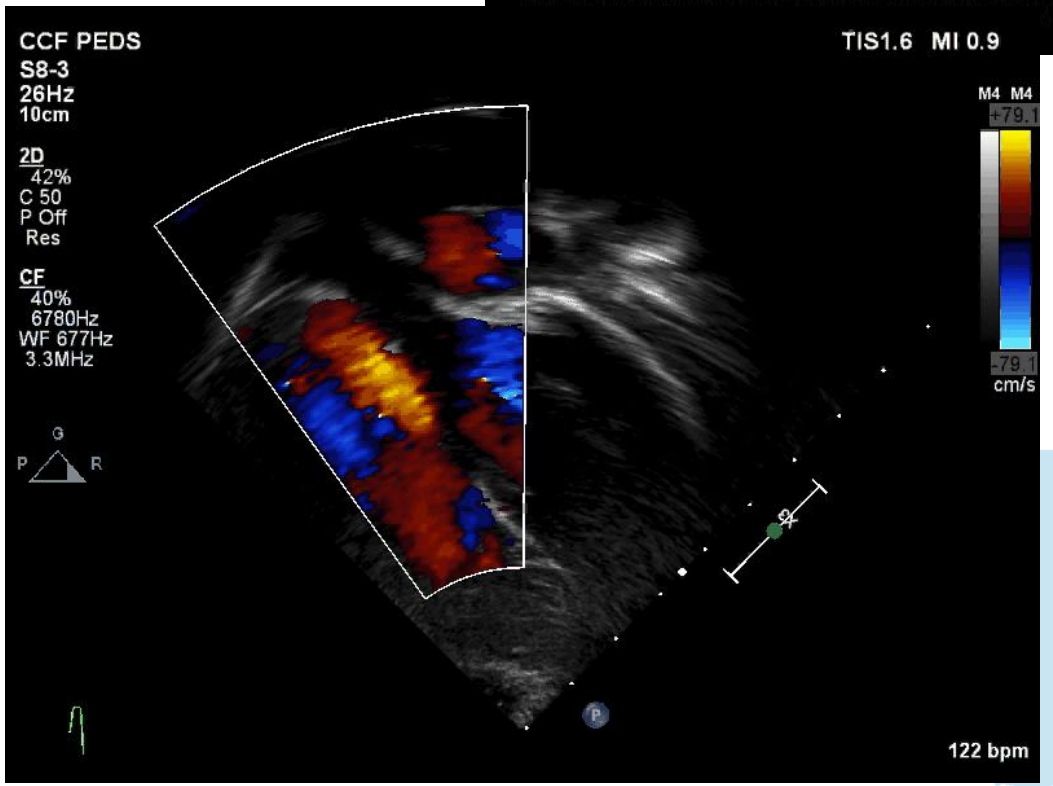


Assess For LVOT obstruction

- Use Apical 5 chamber to rule out LVOT obstruction
- Record peak LVOT velocity

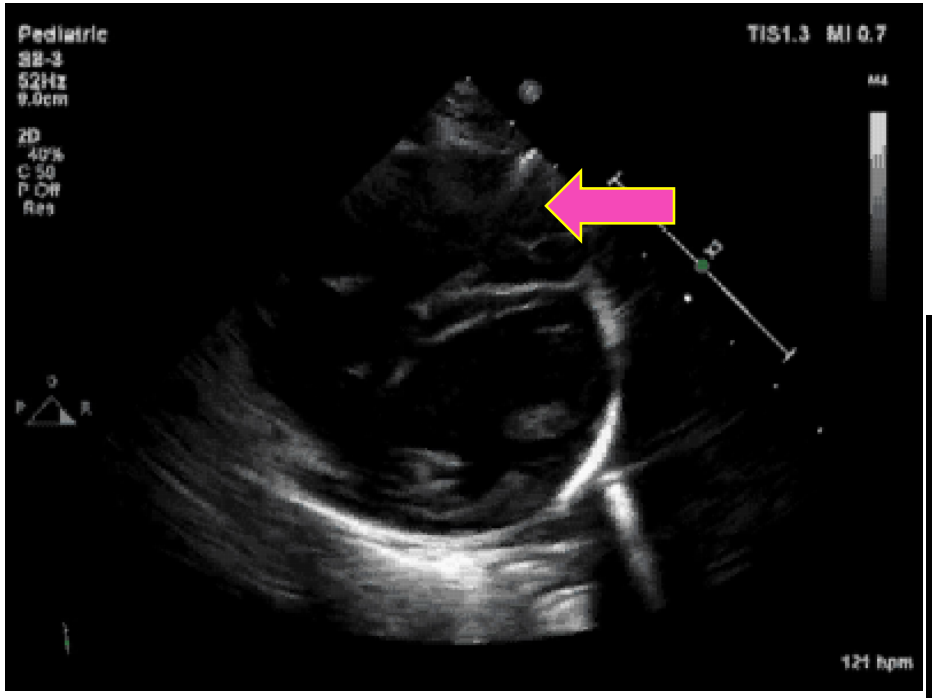


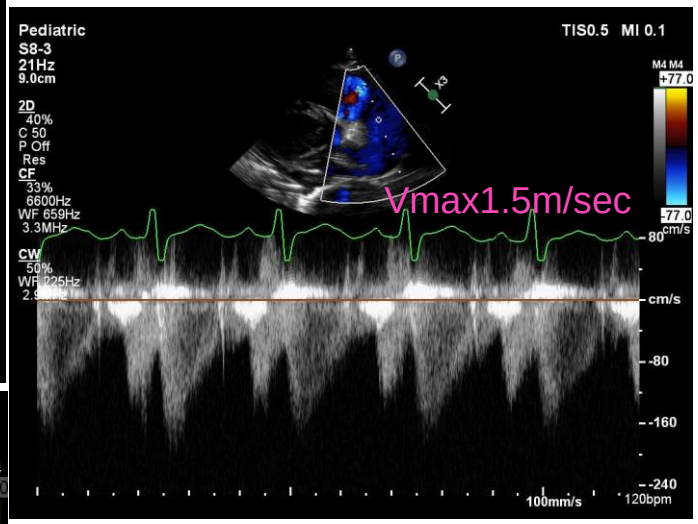


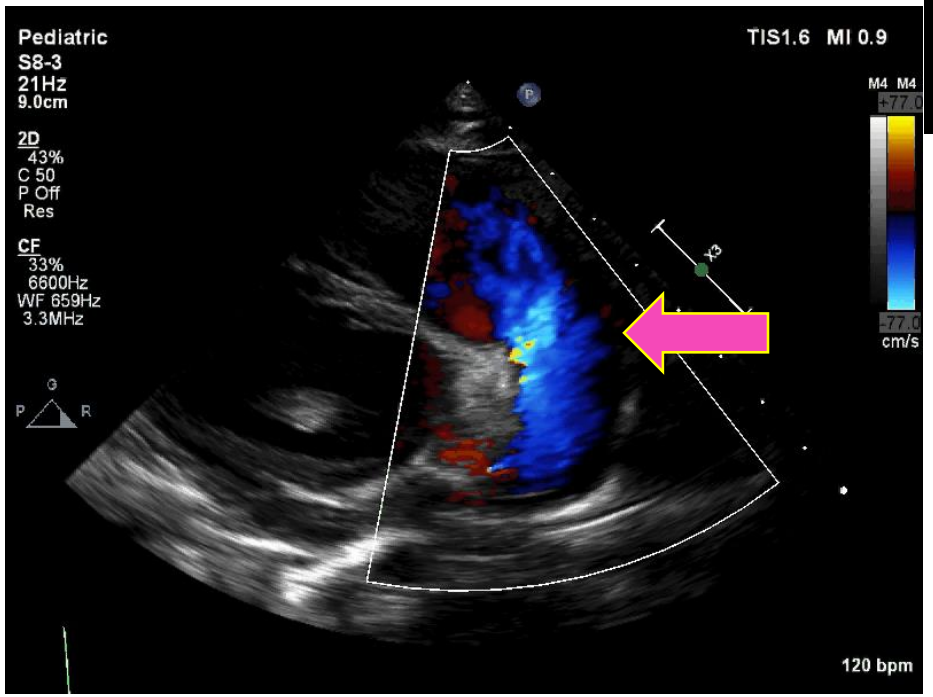


Evaluate Hemodynamic Load On Ventricles

- Dilation of the RV
- Increased flow across the pulmonary valve
- Increased AV valve inflow





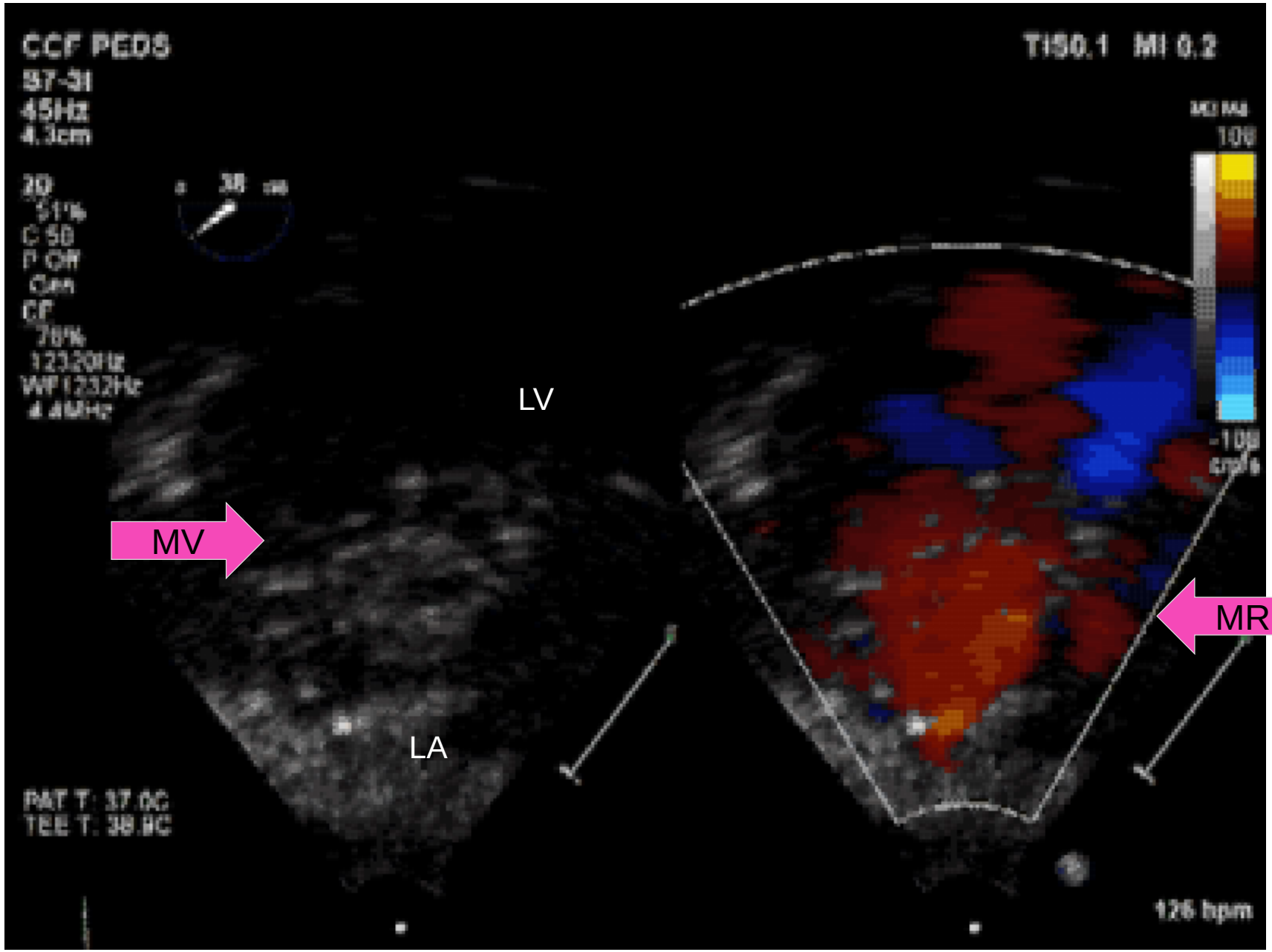


Approximately 1 month later, the child underwent complete surgical repair

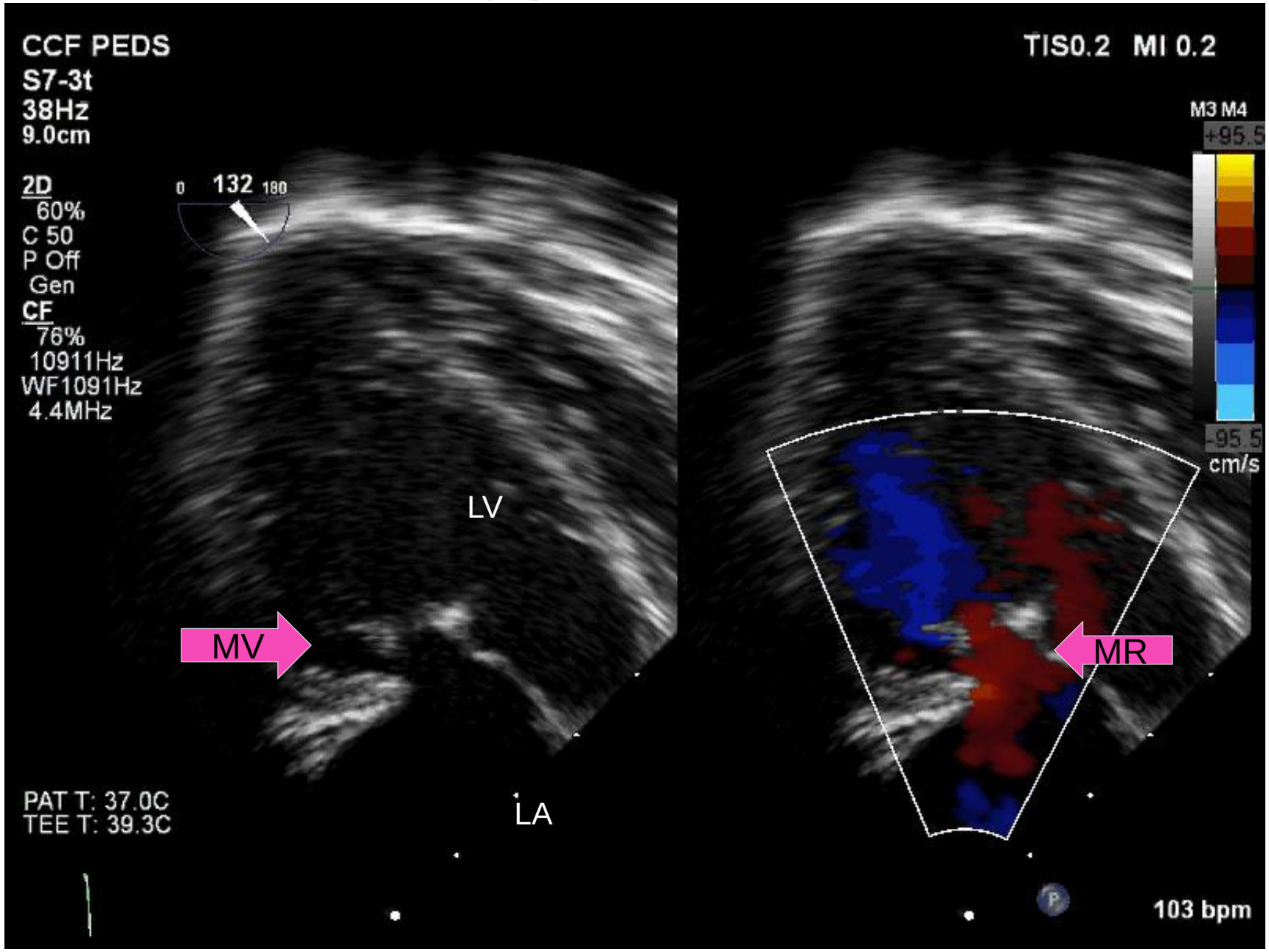
- Primary (suture) closure of primum ASD

- Attempted complete closure of mitral valve cleft
  - Significant MR after initial pump run
  - Patient placed back on bypass and additional commissuroplasty and annuloplasty sutures placed
  - Mild to moderate MR still present
  - No additional intervention pursued

Bypass run #1



Bypass run #2

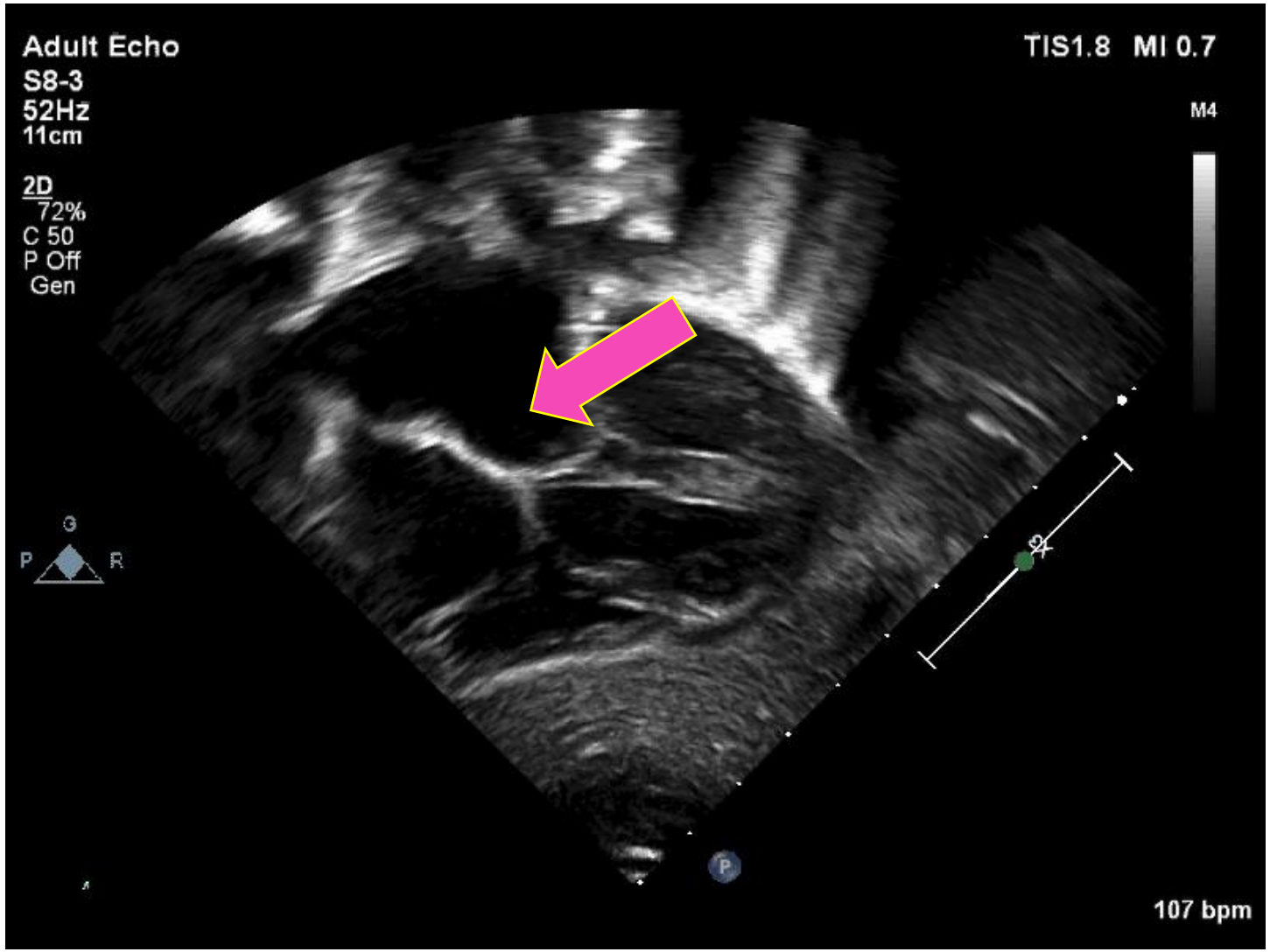

CCF PEDS
S7-3t
38Hz
9.0cm
2D
60%
C 50
P Off
Gen
CF
76%
10911Hz
WF1091Hz
4.4MHz
0
132
180
TIS0.2 MI 0.2
M3 M4
+95.5
-95.5
cm/s
LV
MV
MR
LA
PAT T: 37.0C
TEE T: 39.3C
103 bpm

1 month Follow Up

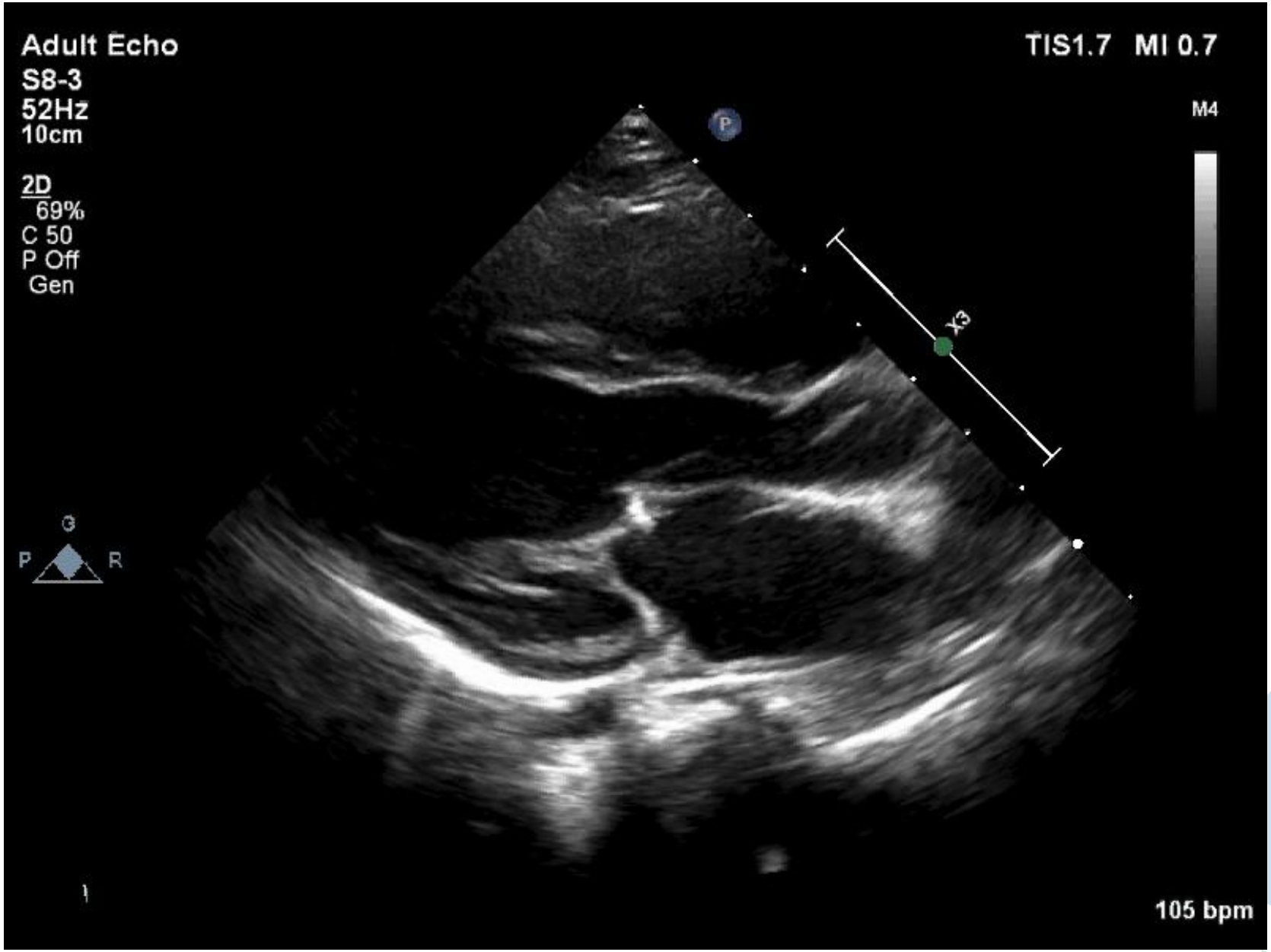
- 90.5 cm (16%ile), 12.2kg (8%ile), BP 88/50, O2 sat 100%
- Normal S1, physiologically split S2, 3/6 medium pitched systolic regurgitant murmur, best heard at left lower sternal border (MR)
- Per mom, he has more energy and a better appetite

1 Month Follow Up Echo

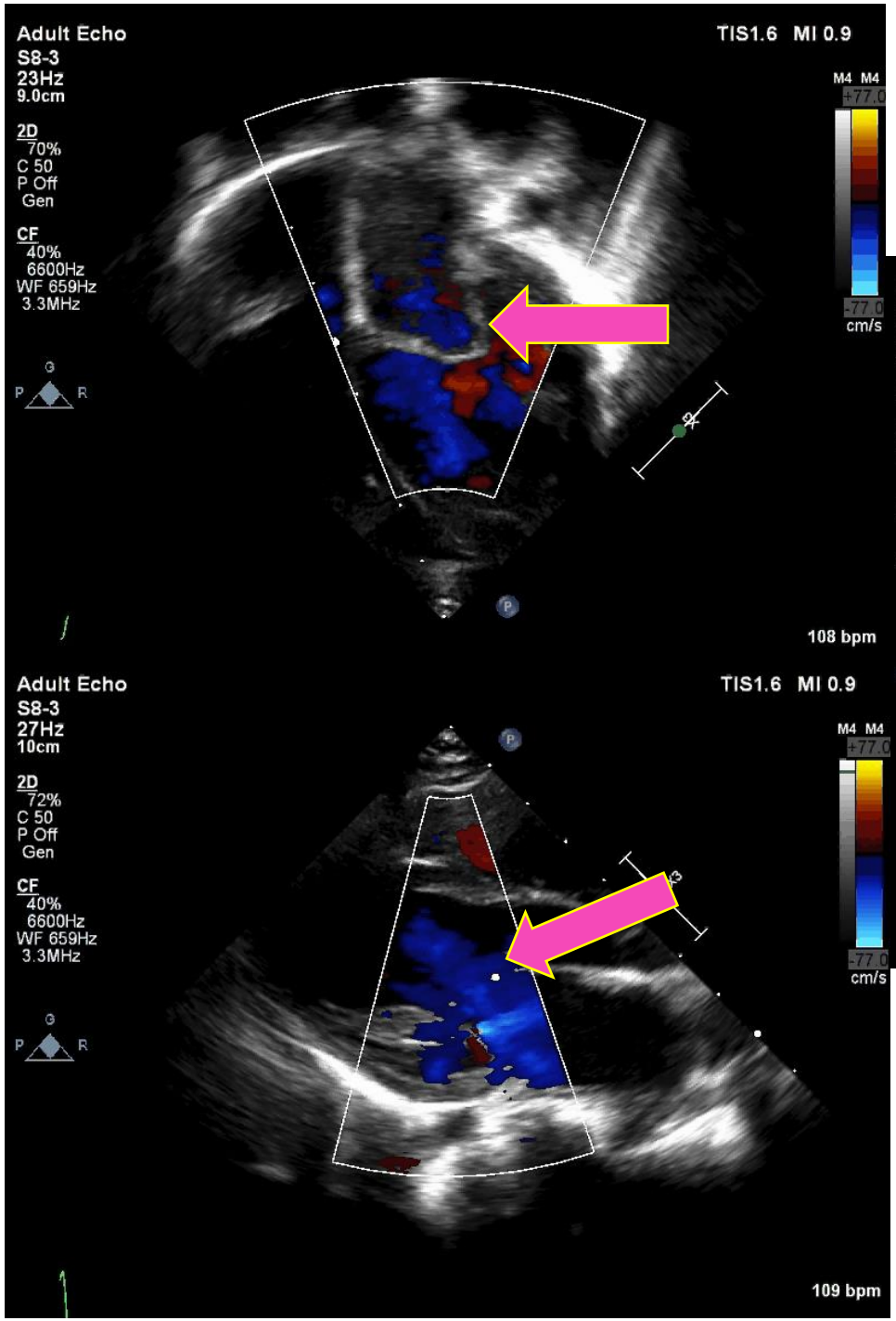
No residual ASD

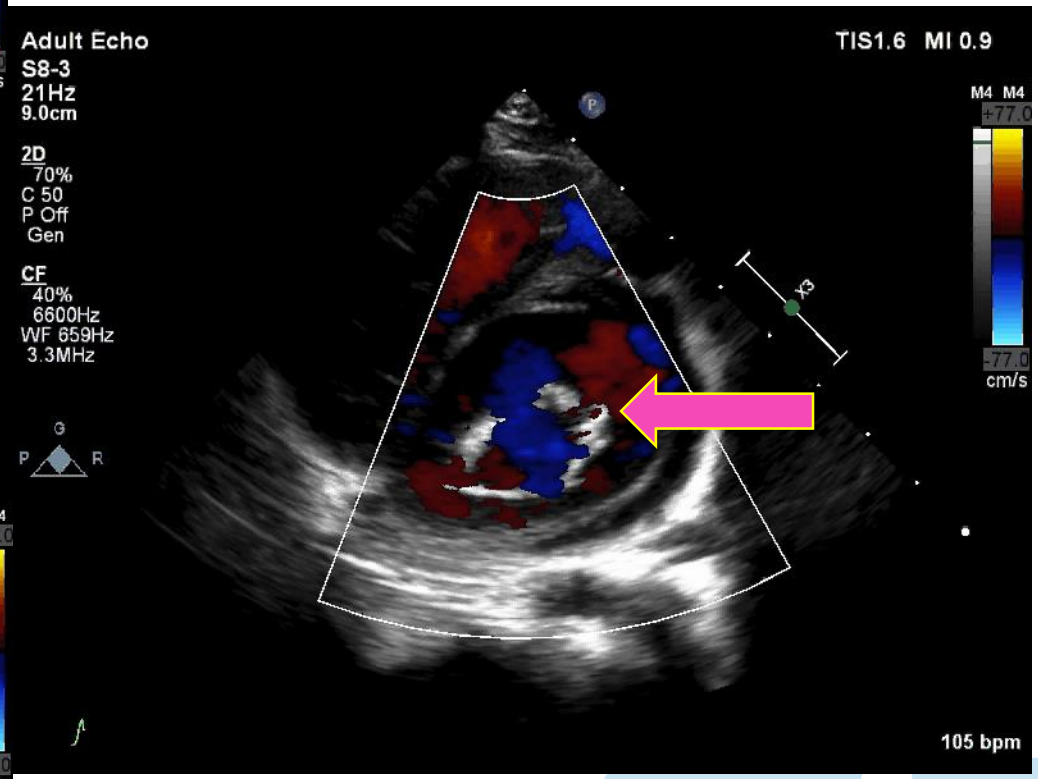


Borderline RV dilation



Moderate residual MR





Recommendations

- No immediate need for repeat intervention
- Close monitoring of LA and LV sizes, PA pressures via serial echocardiograms
- For progression of MR and/or clinical symptoms, surgical intervention with a likely mitral valve replacement would be indicated

Thank you for listening!