CARDIOLOGY 2025

HOPE. HEAL. LEARN.

Imaging Evaluation of a Fontan Pathway

February 21, 2025

Children's Hospital of Philadelphia®
Cardiac Center

Overview

- Imaging highlights
- Limitations
- Exemplary cases

Intracardiac anatomy

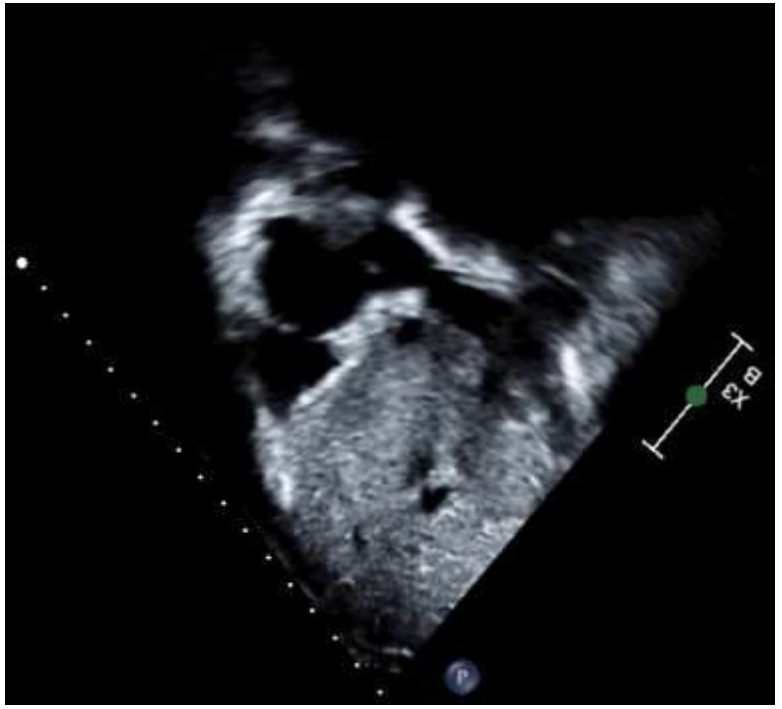
- Many anatomic variations culminate with Fontan circulation
- Knowing intracardiac anatomy is key
  - Atrioventricular valve type/number
  - Atrial septum
  - Semilunar valve and connections
  - VSDs (is restriction important)
  - Pulmonary veins
  - Native aorta, DKS/Norwood
  - Antegrade pulmonary blood flow
  - Ventricular Anatomy

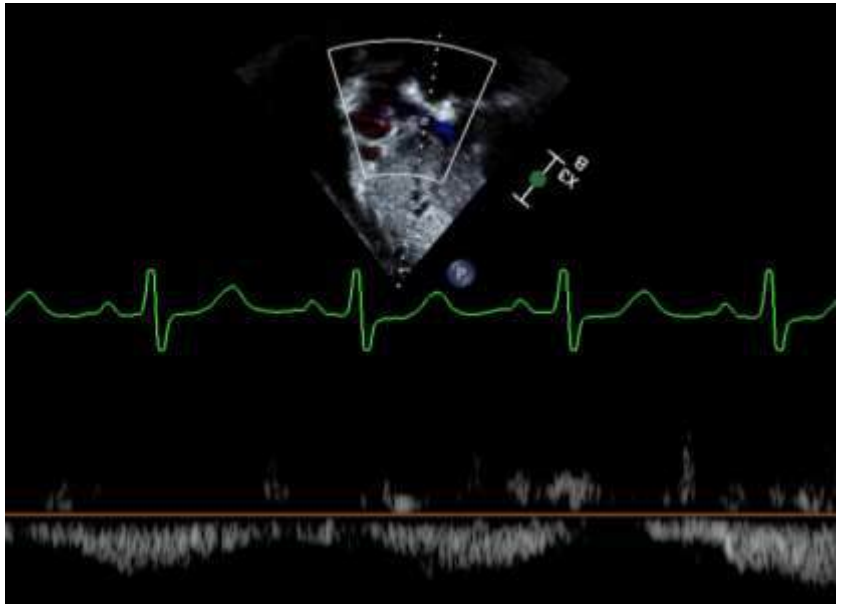
Key Elements to Echo

- Assessment of the Fontan pathway:
  - Presence of fenestration and transfenestration mean gradient
  - Presence of thrombus in Fontan pathway
  - Obstruction of Fontan connections
- Assessment of the pulmonary venous chamber:
  - Document unrestricted atrial communication
  - Assess for pulmonary venous obstruction
- Assessment of AV valve function:
  - AV valve regurgitation: severity grade and mechanism
  - AV valve stenosis: mean inflow gradient
- Assessment of single ventricular function:
  - Ventricular size measurements
  - Systolic performance
- Assessment of outflow tracts, semilunar valve(s), ascending aorta, and aortic arch:
  - Determine semilunar valve competency
  - Assess for outflow obstruction to aorta
  - Assess the DKS connection
  - Assess for residual or recurrent arch obstruction
- Assessment of branch pulmonary arteries:
  - Determine narrowing, stenosis, and competitive flow

Initial Sweep

- IVC-Fontan connection
  - Low velocity, phasic flow (<20-30cm/s)
  - Flow increases with inspiration
  - Absence of variation abnormal
    - Obstruction, AVVR, competitive flow (collaterals)





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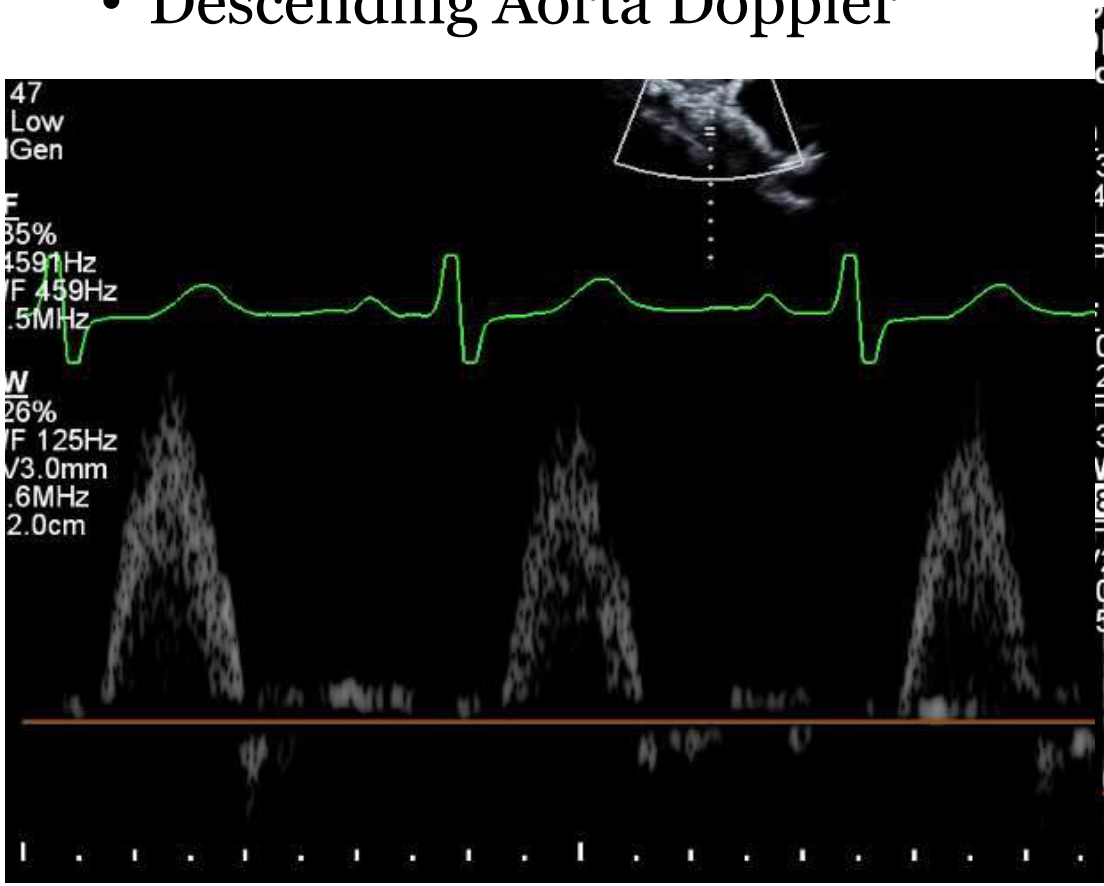
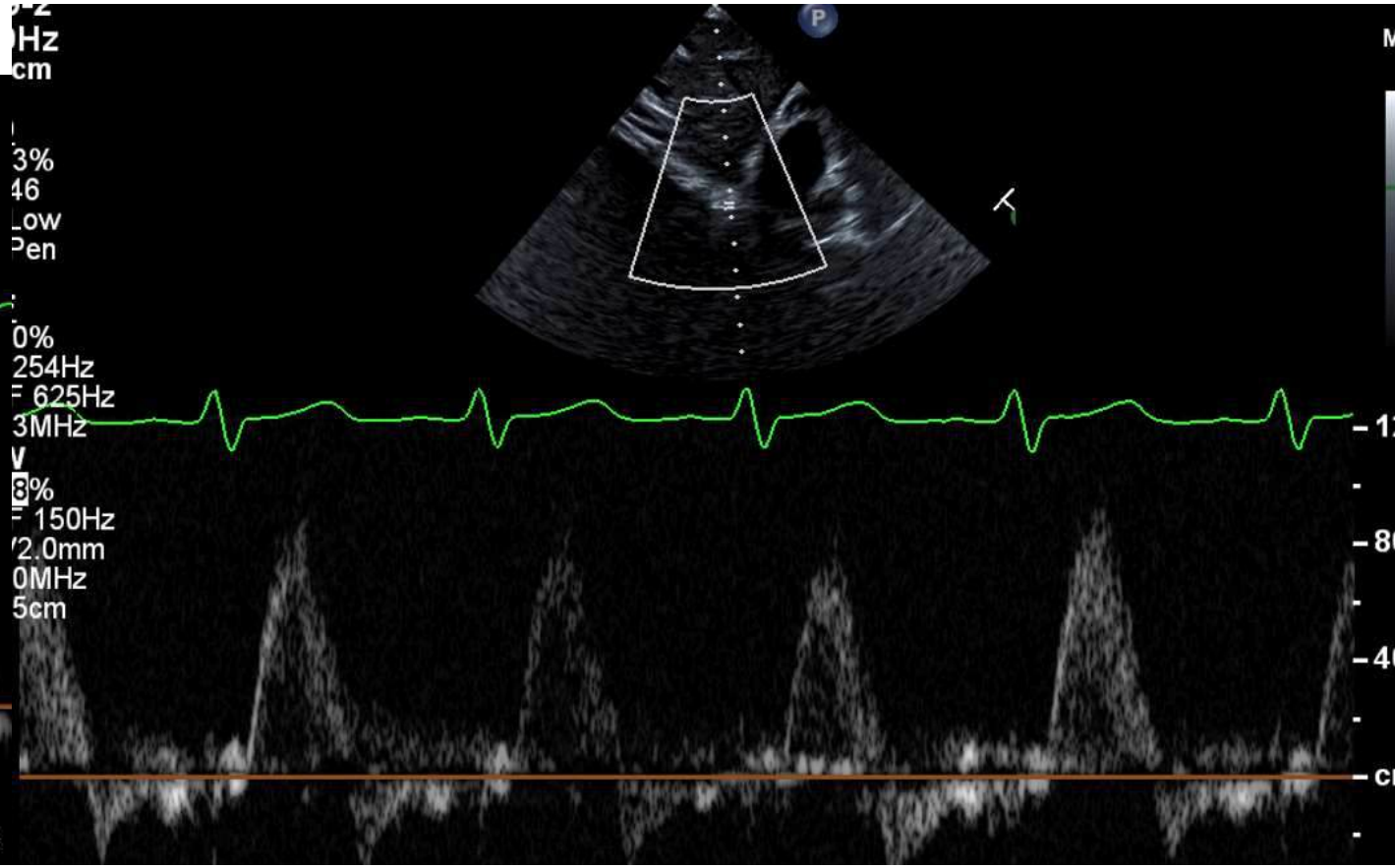
Subcostal

- Descending Aorta Doppler





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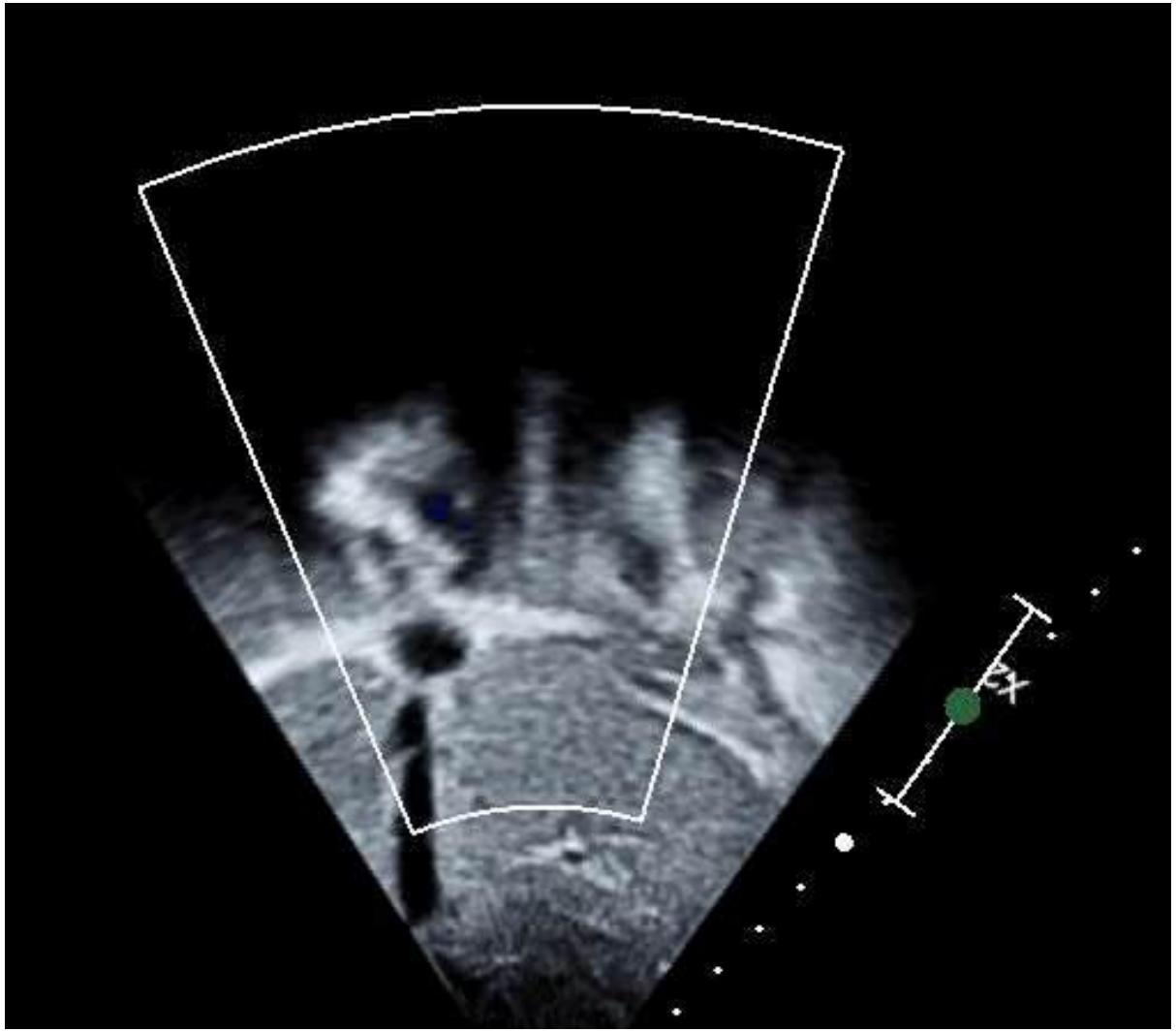
Atrial Septum



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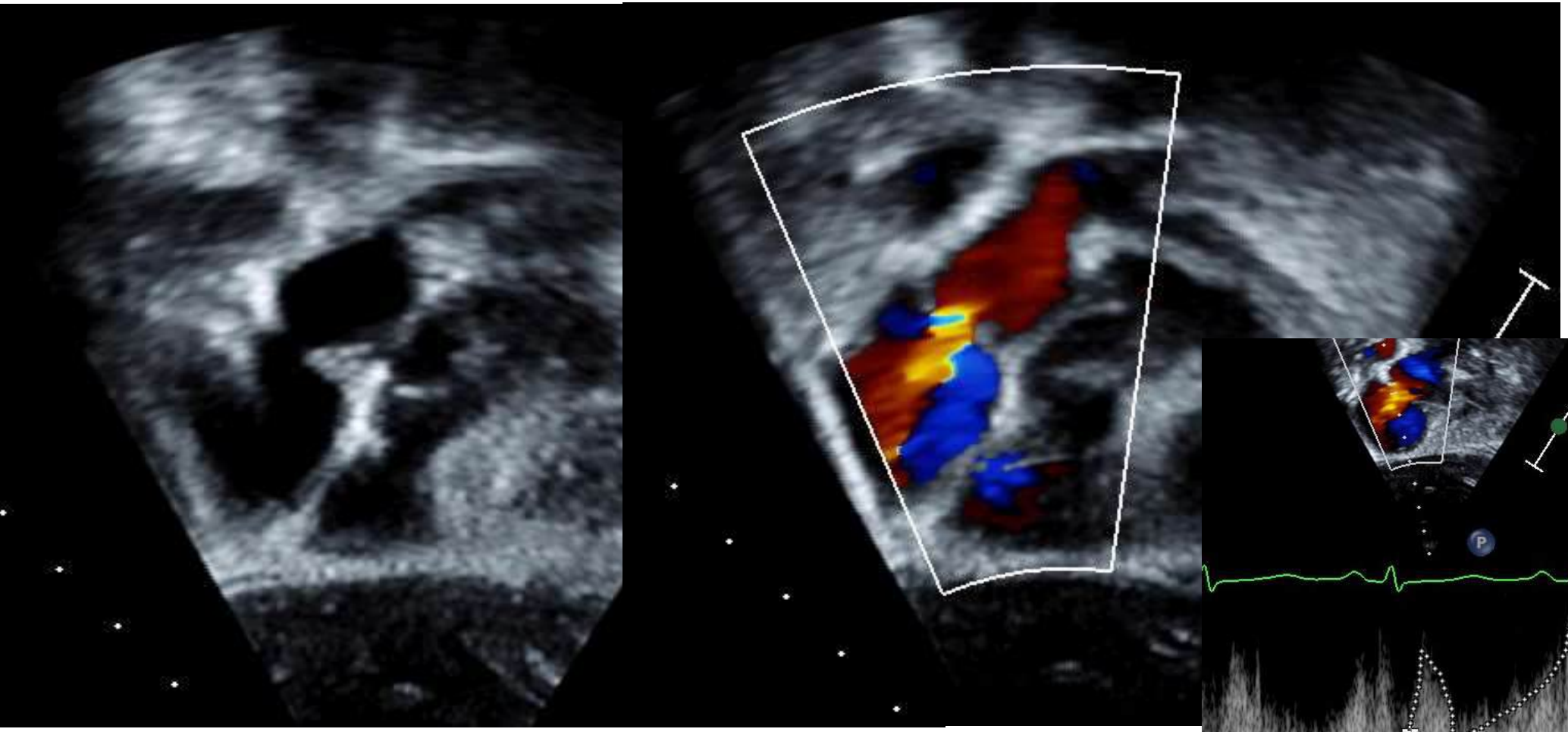
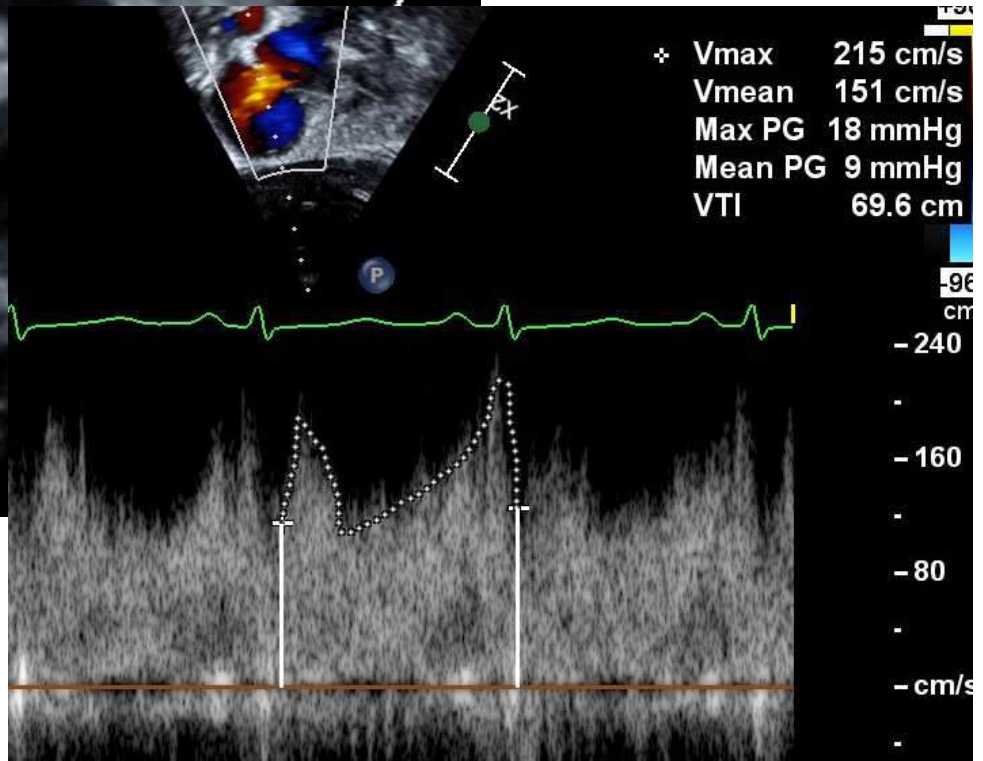
Atrial Septum





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Fontan Conduits

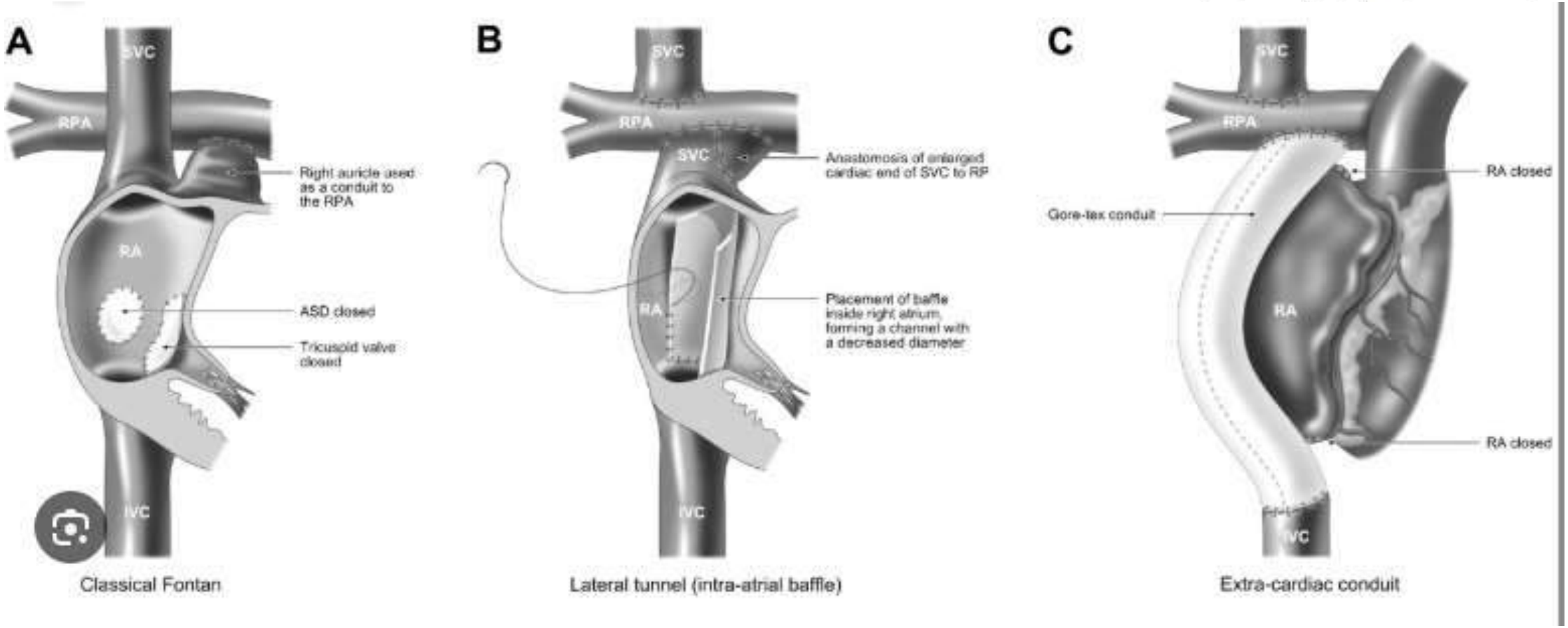
Circulation
Volume 110, Issue 11_supplement, 14 September 2004: Pages I-157-I-164
https://doi.org/10.1161/CIRCULATIONAHA.104.482445

SURGERY FOR CONGENITAL HEART DISEASE

The Fontan Procedure

Contemporary Techniques Have Improved Long-Term Outcomes

Yves d'Udekem, MD, PhD, Ajay J. Iyengar, BmedSci, Andrew D. Cochrane, MD, FRACS, Leeanne E. Grigg, MBBS, FRACP, James M. Ramsay, MD, FRACP, Gavin R. Wheaton, MD, FRACP, Dan J. Penny, MD, PhD, FRACP, and Christian P. Brizard, MD



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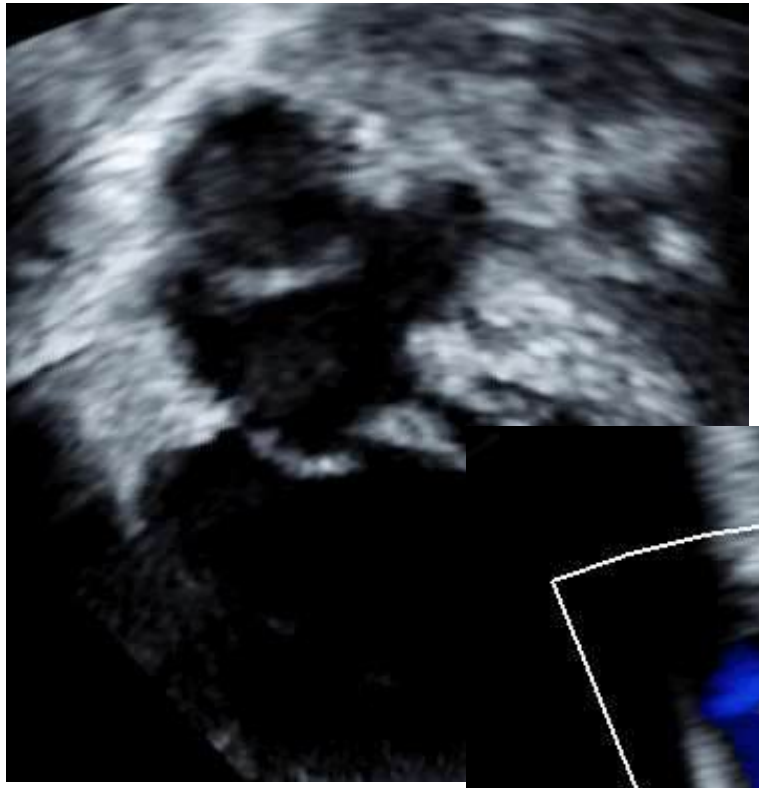
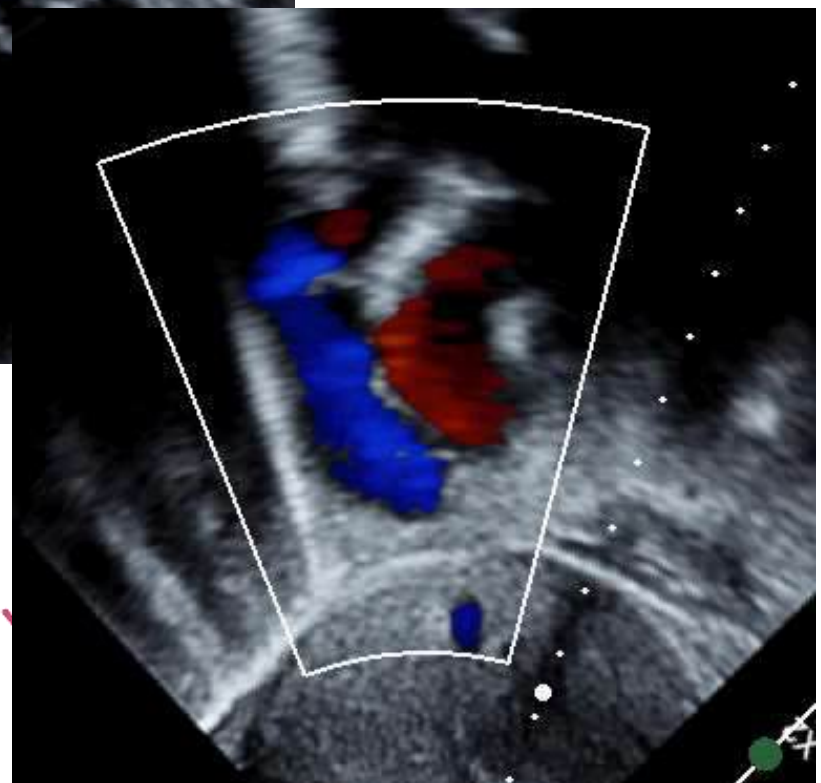
Lateral Tunnel vs. Extracardiac





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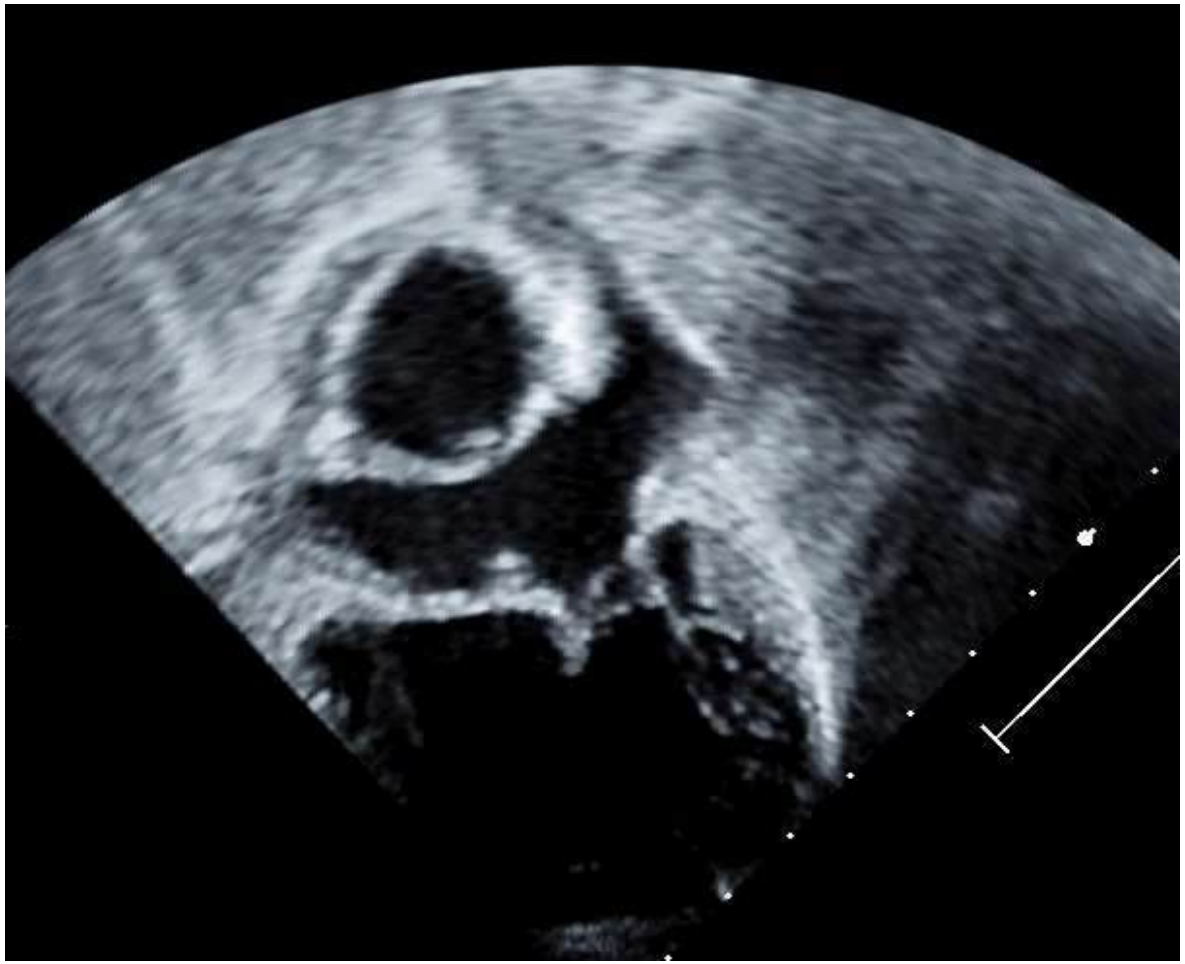
Apical View: Fontan Pathway



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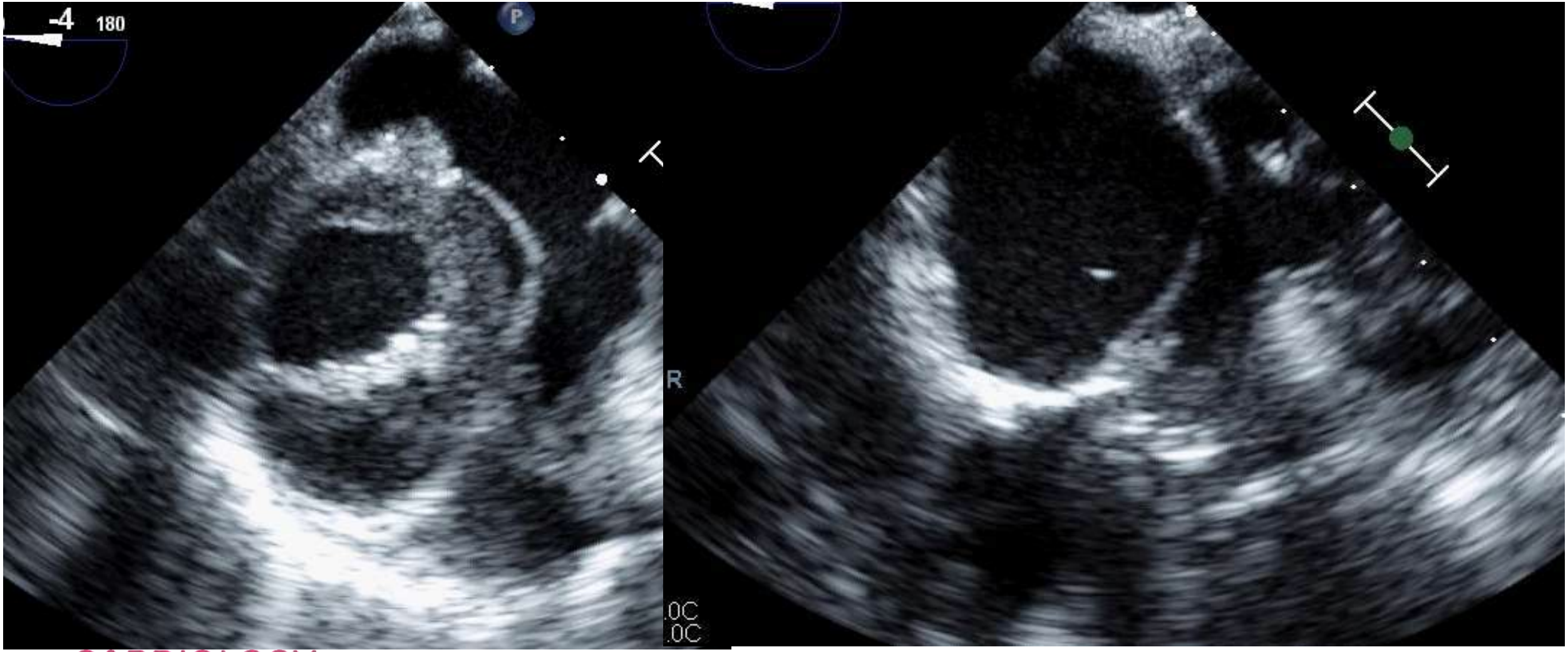
TEE: Fontan Thrombus



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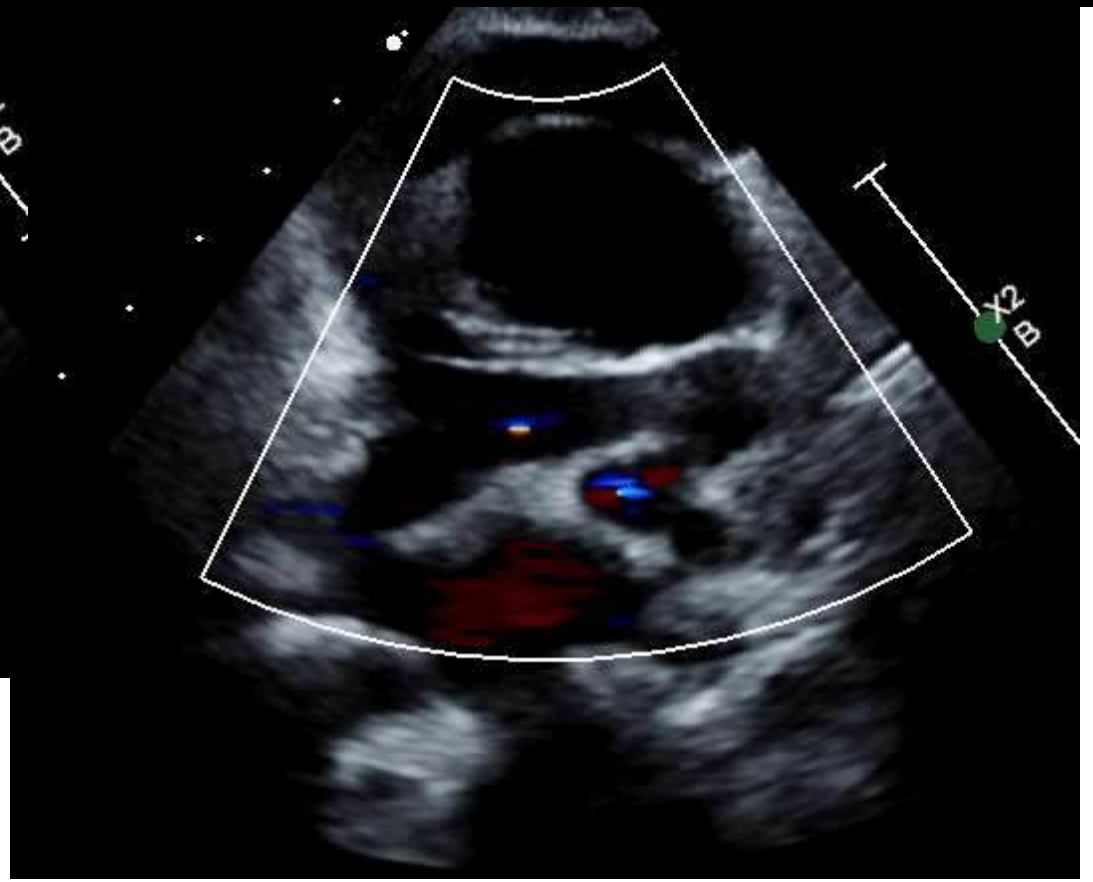
Suprasternal: SVC/PA Connect



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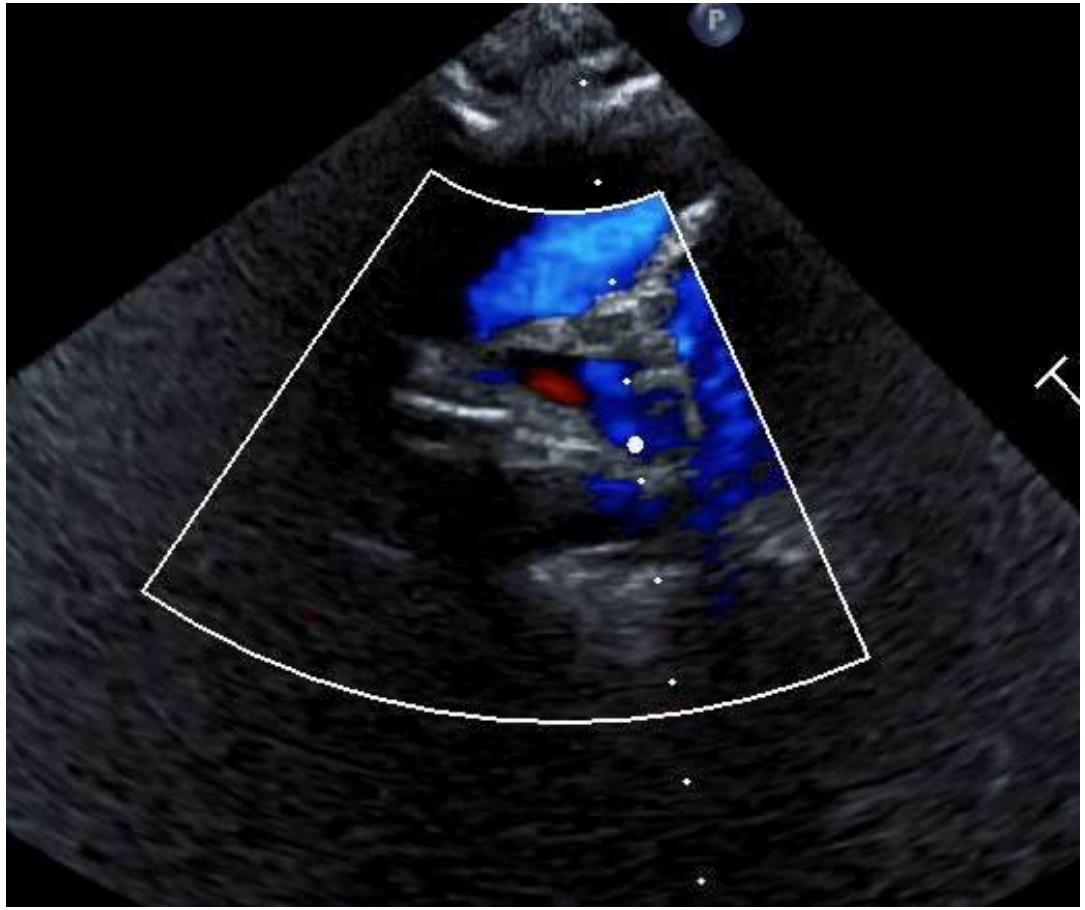
Pulmonary Arteries: Bilateral SVCs



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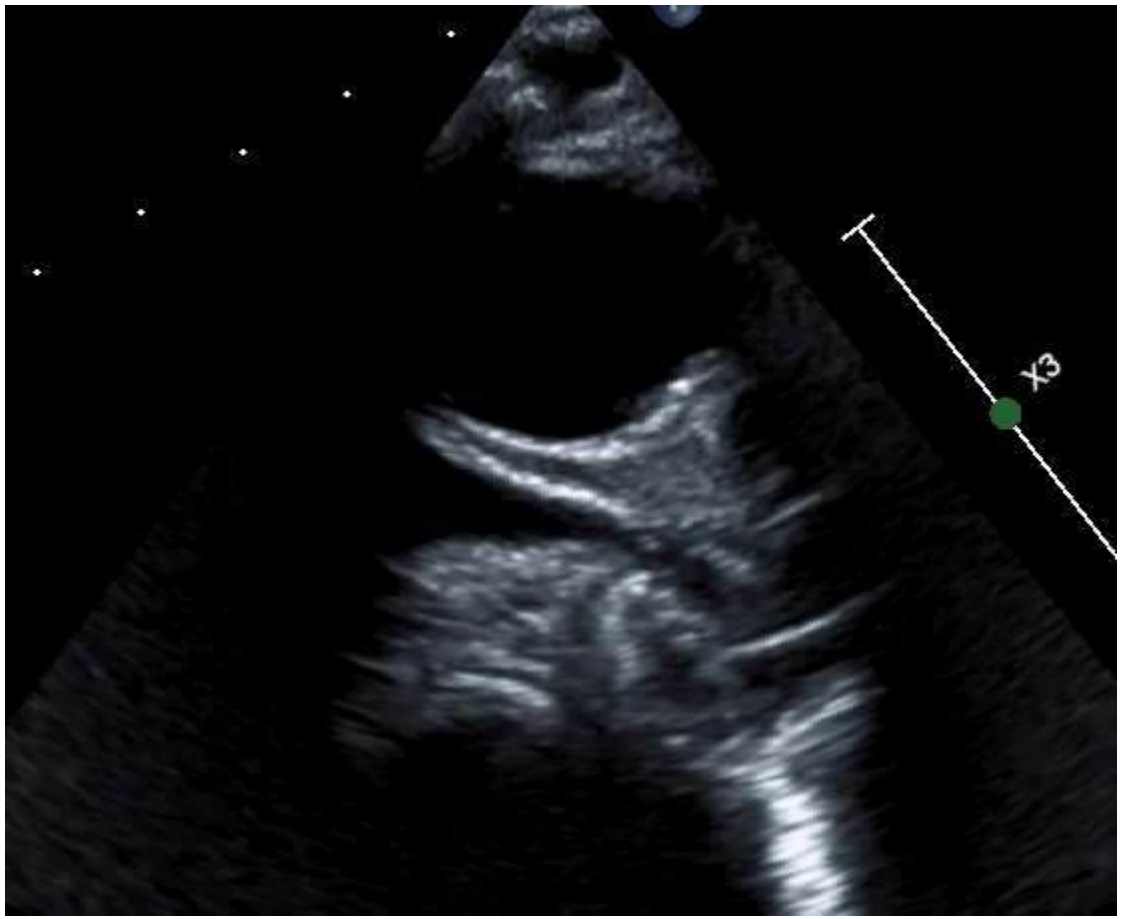
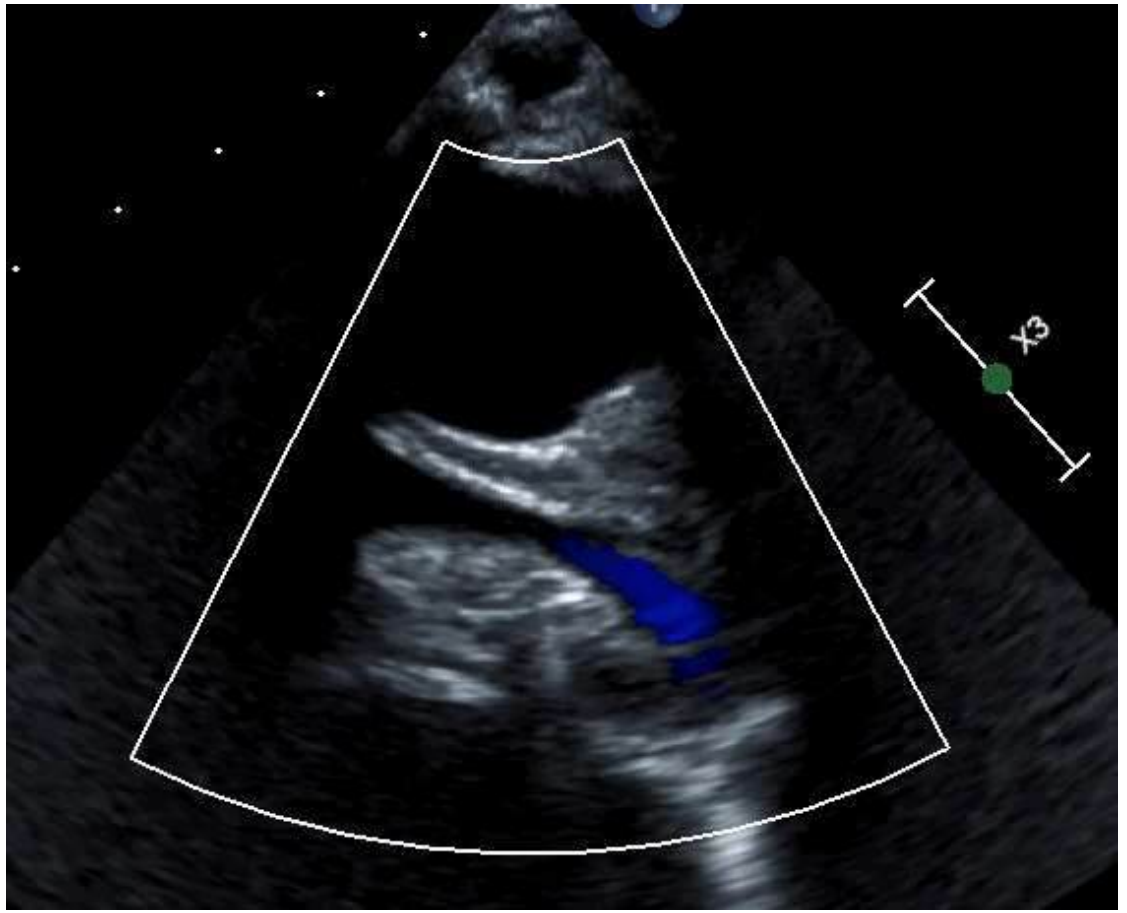
Central PA Narrowing





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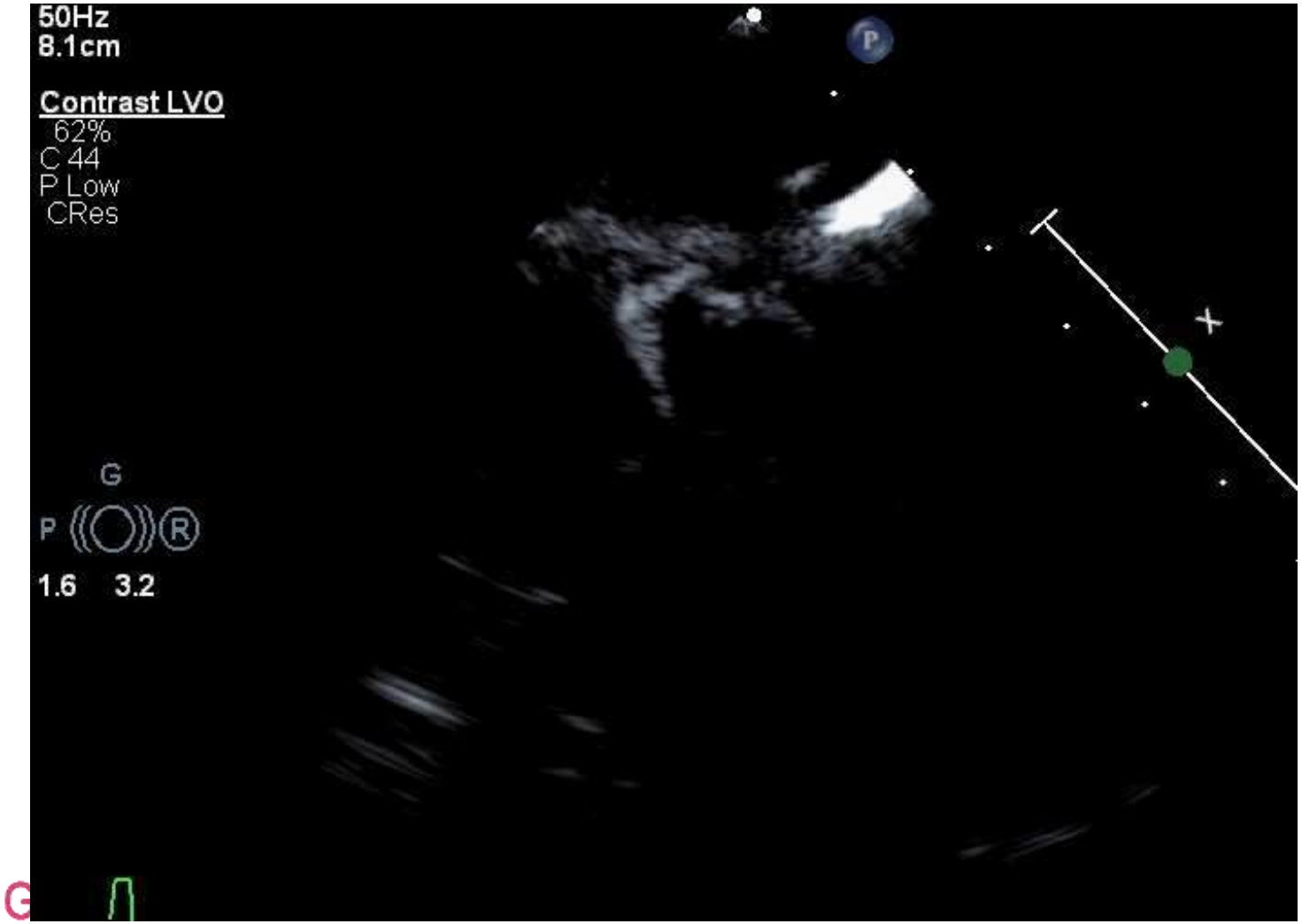
Difficult Windows: Contrast imaging of PAs with Lumason



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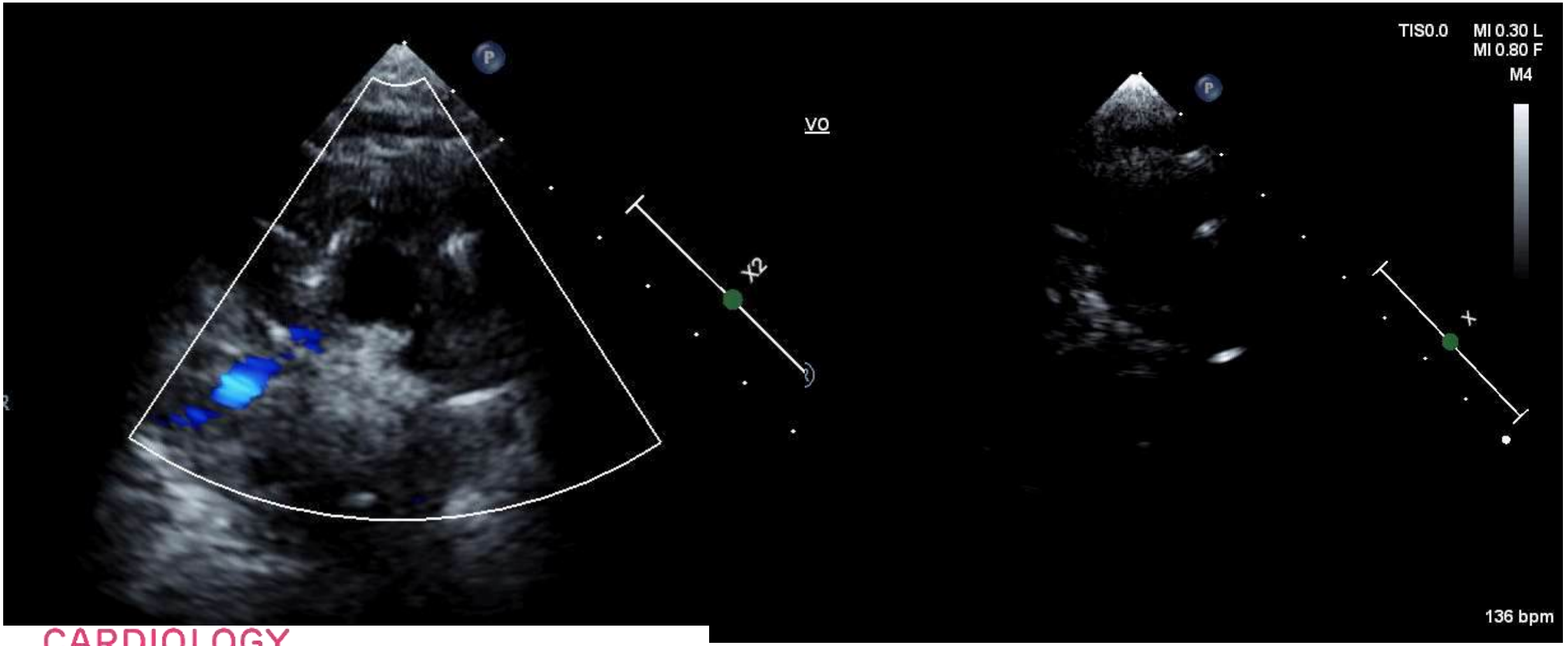
PA obstruction



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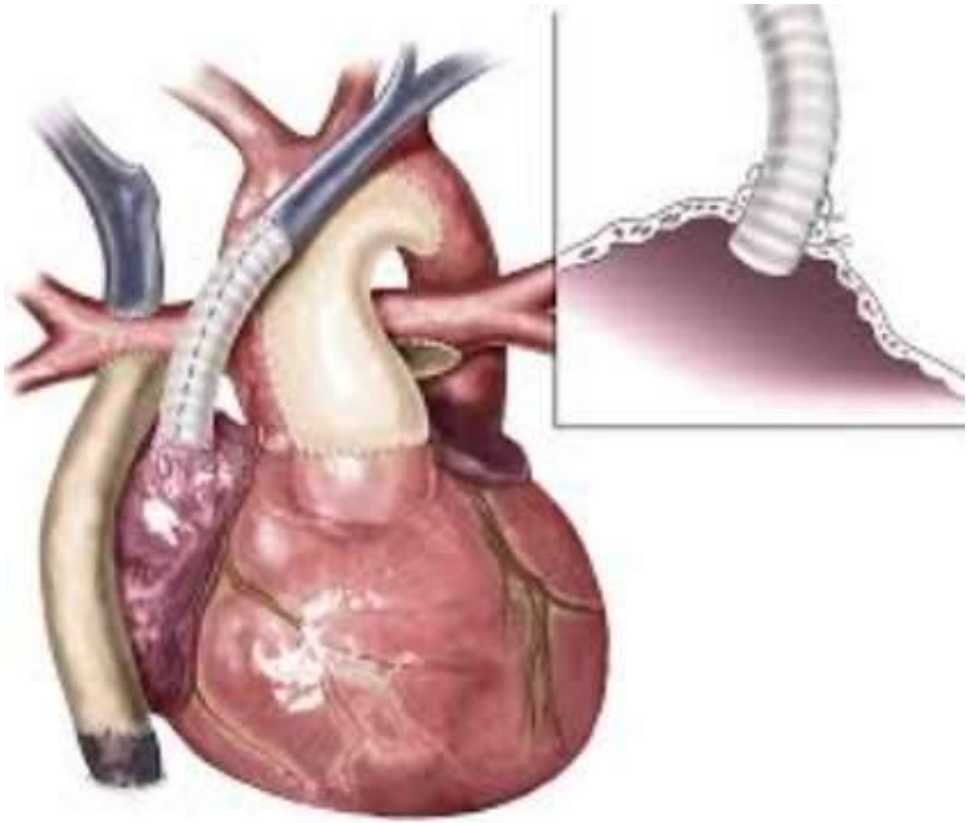
Saline Contrast: Innominate Vein Turn Down (TDD)



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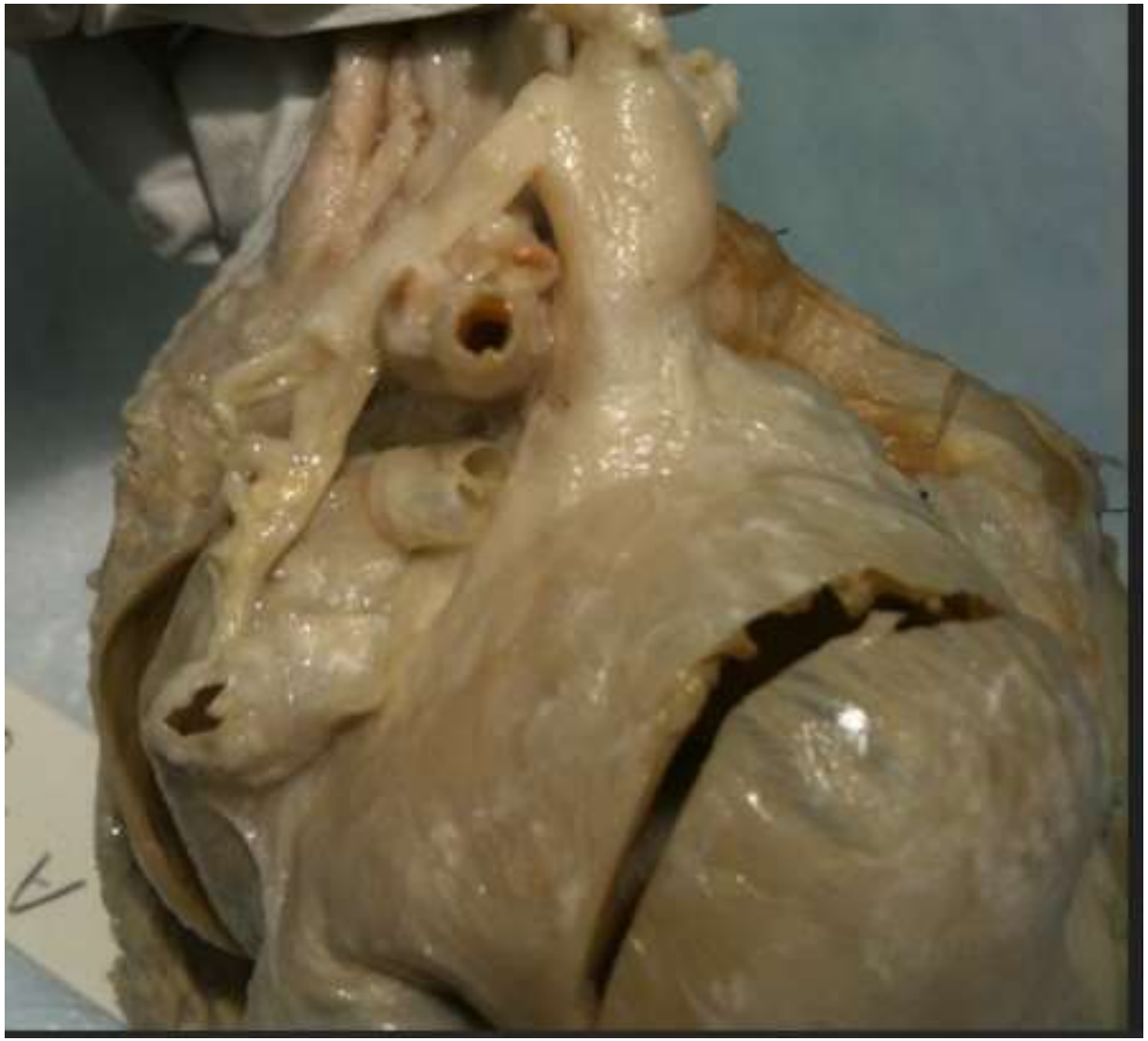
RUPV Obstruction



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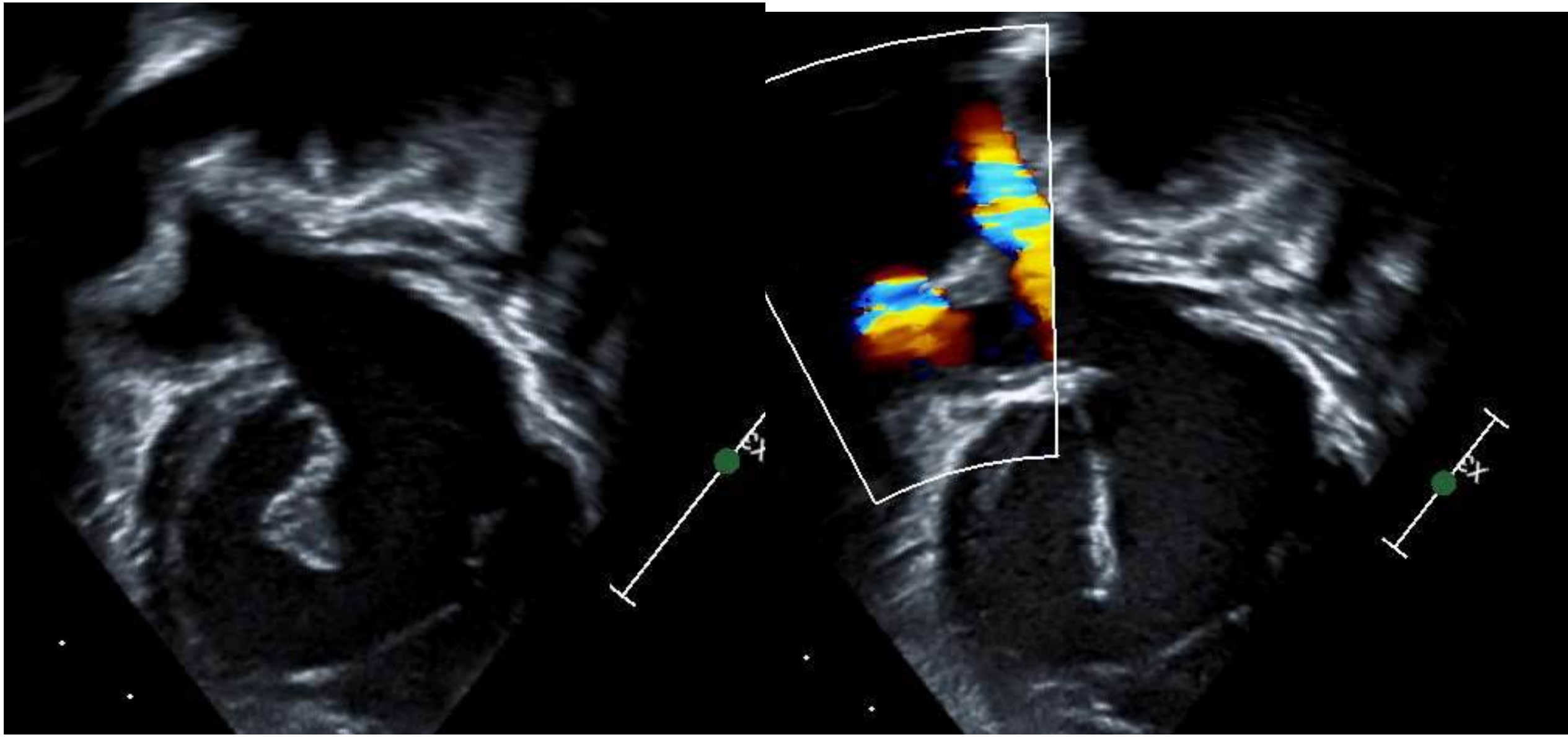
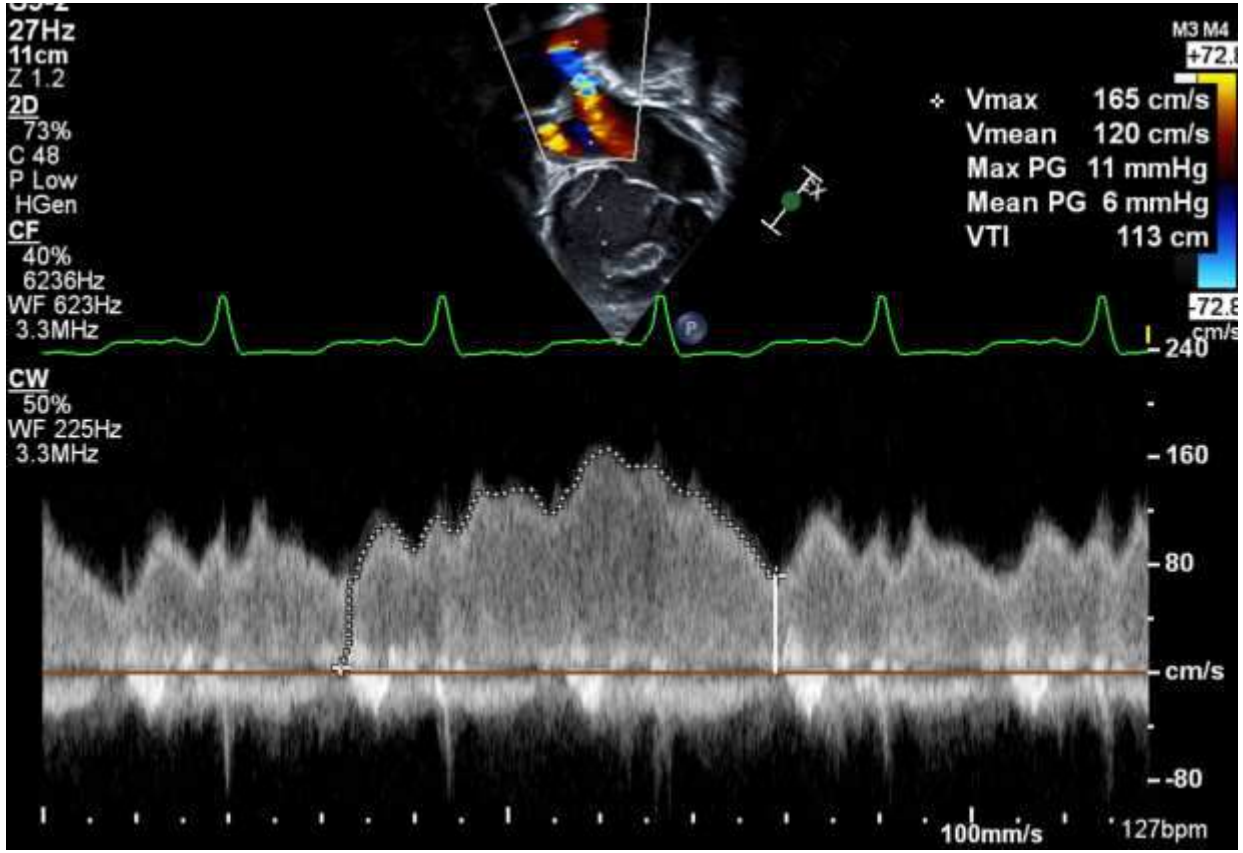
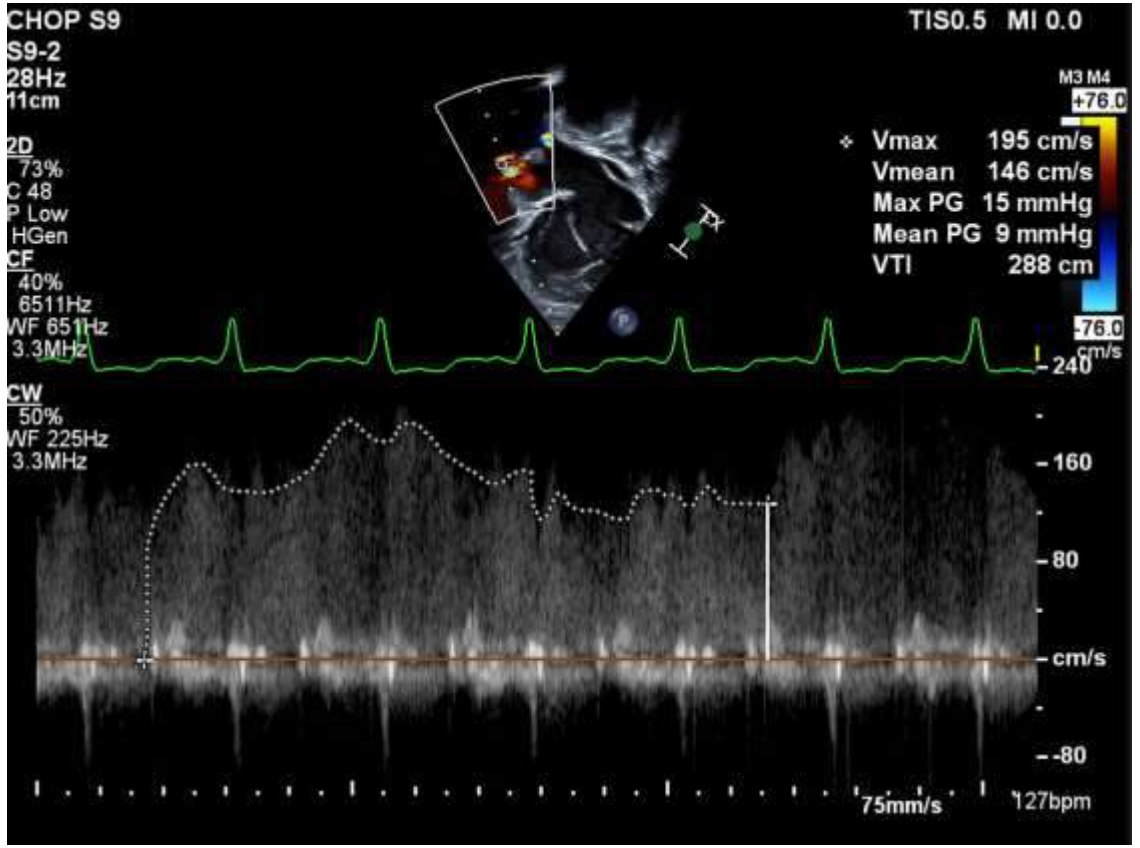
RPV Obstruction



Pulmonary Vein Gradient



Fenestration Gradient



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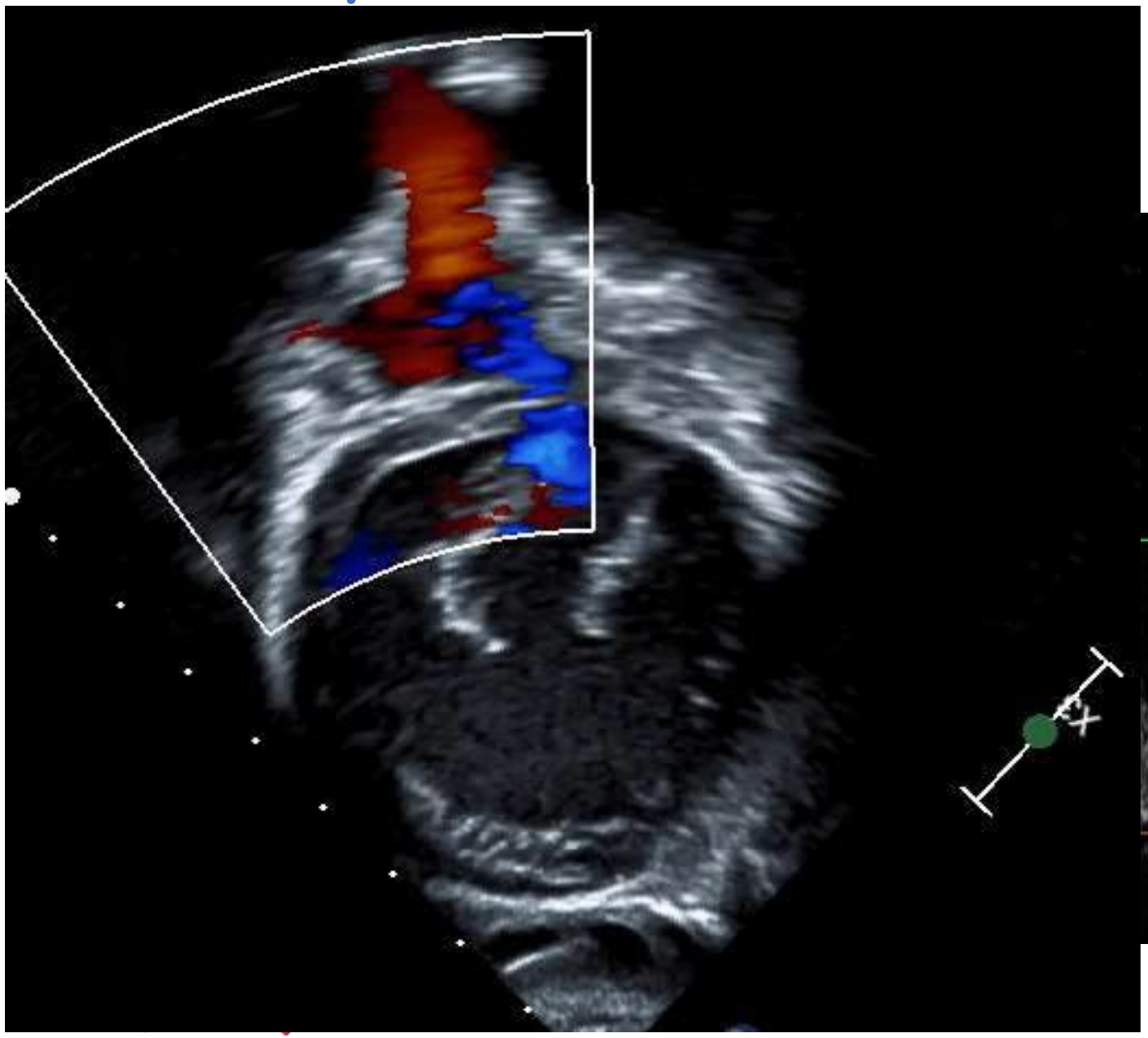
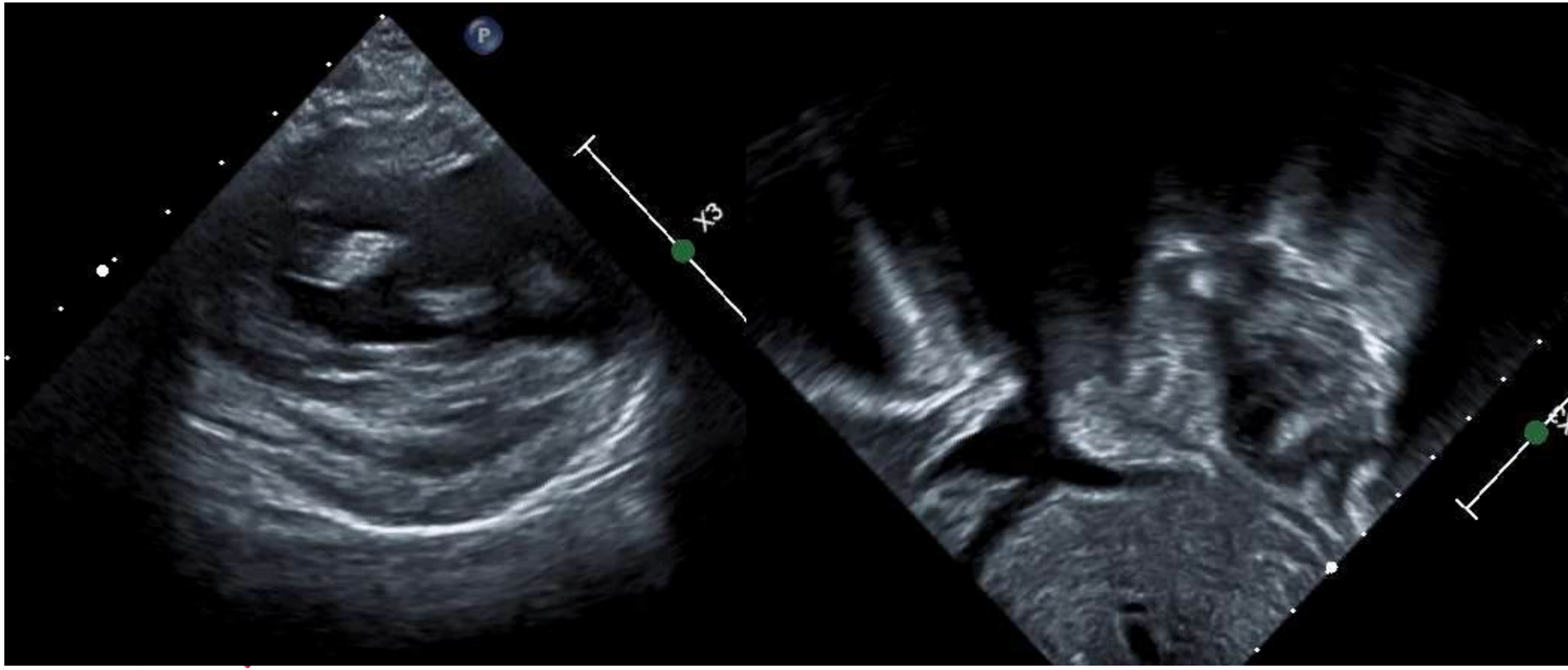
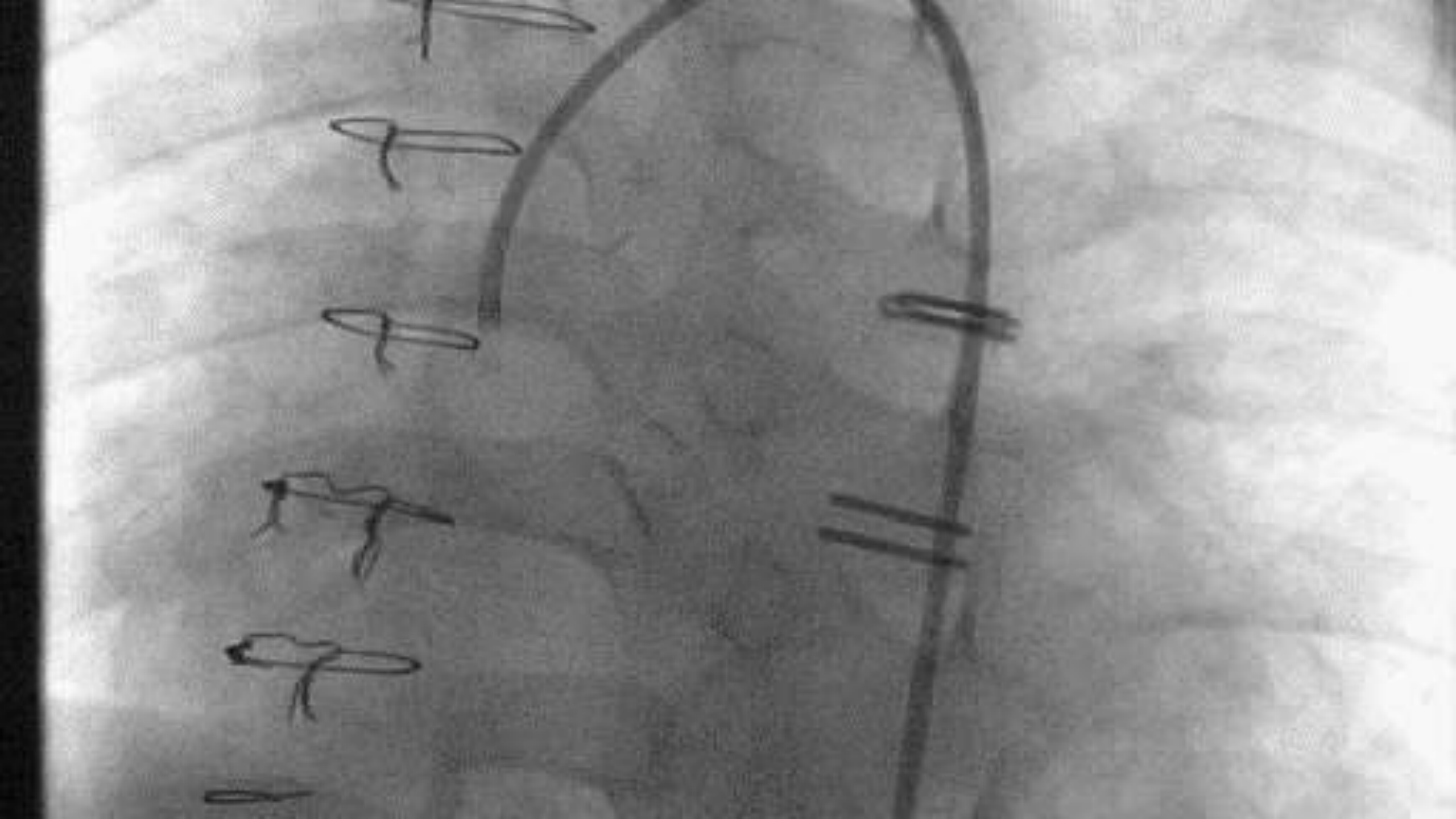
After Repair

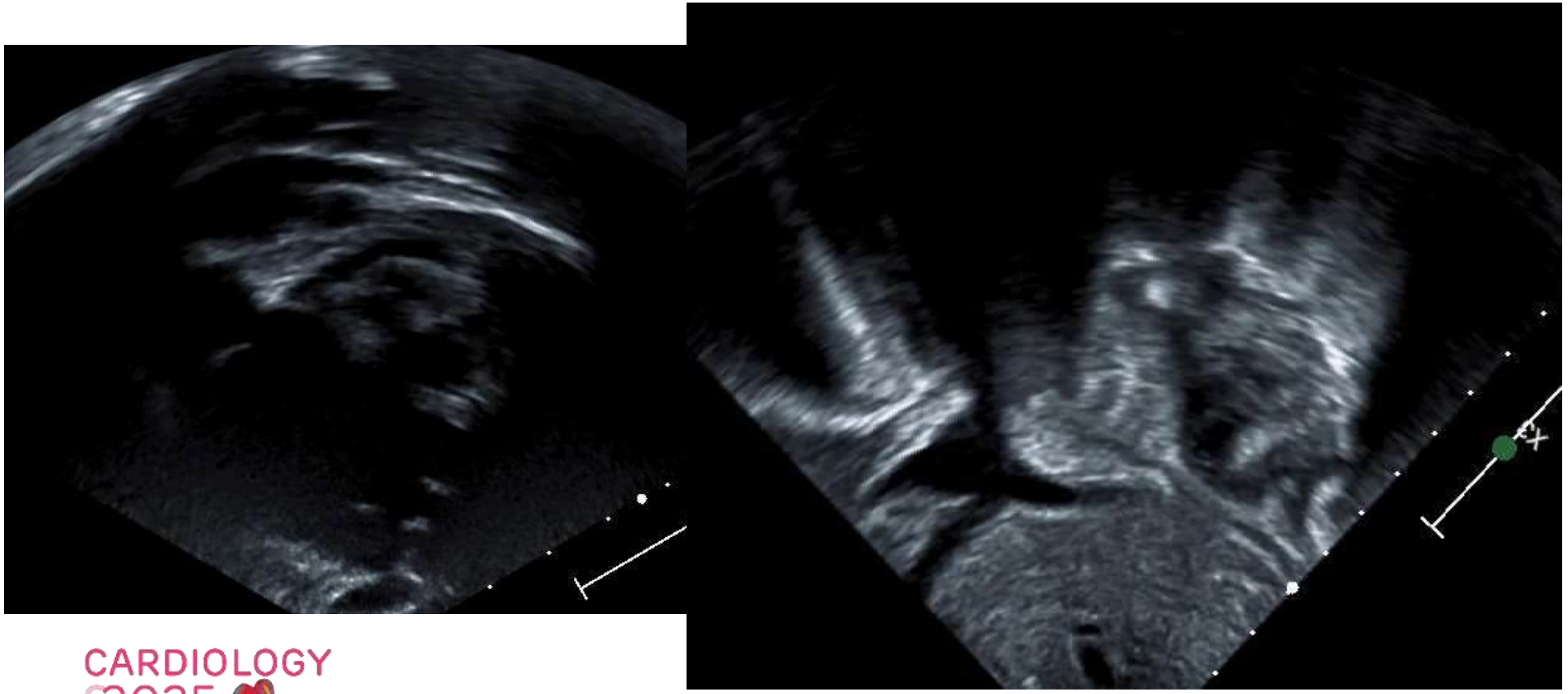


Case: New Right Ventricular Dysfunction in HLHS





Aortic Root



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Summary

- Know underlying anatomy
- Pathway can be seen in multiple views, but windows are often limited
- Doppler patterns can signal abnormal hemodynamics
  - Lack of reparatory variation, high velocity, flow reversal
- Contrast agents can be helpful
- Index of suspicion must be high when changes appear
  - Comparison to previous studies key
  - Clinical picture essential
- Cross sectional imaging when echo not definitive

Thank you

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- Lateral tunnel: https://sketchfab.com/3d-models/46-heterotaxy-with-unbalanced-avc-4c86b147fa9b4f22b7279ed016a15c26

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