AMSER Case of the Month March 2026

43d old male presenting with hypoxia

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Patient Presentation

- 43d old male born via C-Section on 8/25/2025 at 38w4d
- APGAR score of 8 and 9 at 1 and 5 minutes
- Weight 4lb12.9oz
- Concern for congenital heart abnormality
- Admitted to NICU after birth for further management
- On 10/6/25 he developed severe hypoxia with saturations in the 40-50%s for 2-3 hours despite 100% FiO2

What Imaging Should We Order?

Select the applicable ACR Appropriateness Criteria

Variant 1: **Child or adult. Repaired tetralogy of Fallot or pulmonary valve stenosis with concern for pulmonary valve dysfunction or branch pulmonary artery stenosis. Incomplete or inadequate assessment of cardiovascular morphology and function after transthoracic echocardiography. Next imaging study.**

Procedure	Appropriateness Category	Relative Radiation Level
Radiography chest	Usually Appropriate	☢
MRA chest without and with IV contrast	Usually Appropriate	O
MRI heart function and morphology without and with IV contrast	Usually Appropriate	O
MRI heart function and morphology without IV contrast	Usually Appropriate	O
CTA chest with IV contrast	Usually Appropriate	☢☢☢☢
CT heart function and morphology with IV contrast	Usually Appropriate	☢☢☢☢
US echocardiography transesophageal	May Be Appropriate	O
Arteriography coronary with ventriculography	May Be Appropriate	☢☢☢☢
Arteriography pulmonary	May Be Appropriate	☢☢☢☢
MRA chest without IV contrast	May Be Appropriate	O
CTA coronary arteries with IV contrast	May Be Appropriate	☢☢☢☢
Perfusion scan lung	May Be Appropriate	☢☢☢☢
MRA abdomen without and with IV contrast	Usually Not Appropriate	O
MRA abdomen without IV contrast	Usually Not Appropriate	O
MRA neck without and with IV contrast	Usually Not Appropriate	O
MRA neck without IV contrast	Usually Not Appropriate	O
MRI heart function with stress without and with IV contrast	Usually Not Appropriate	O
MRI heart function with stress without IV contrast	Usually Not Appropriate	O
FDG-PET/CT heart	Usually Not Appropriate	☢☢☢☢
SPECT or SPECT/CT MPI rest and stress	Usually Not Appropriate	☢☢☢☢☢

This imaging modality was ordered by the physician

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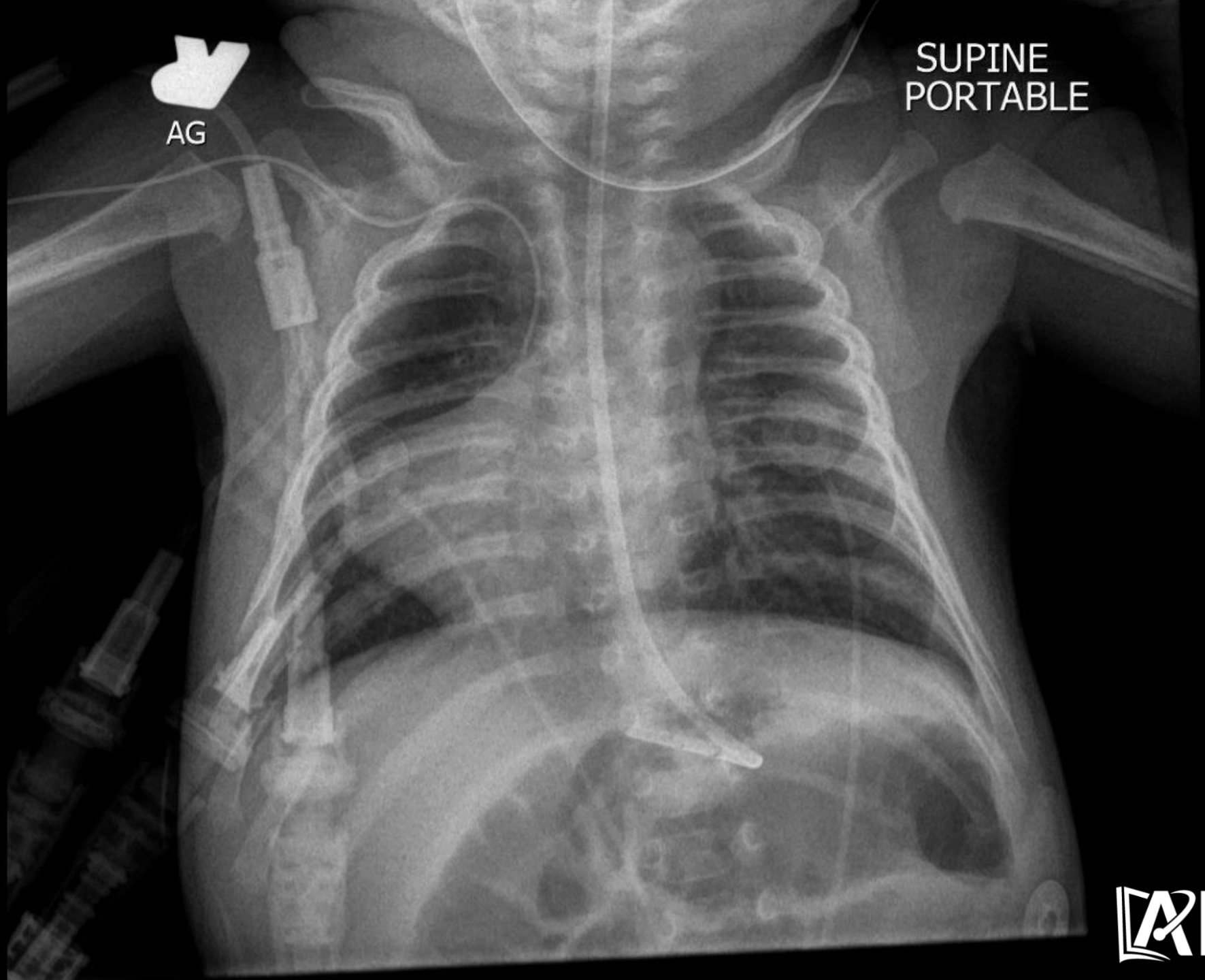
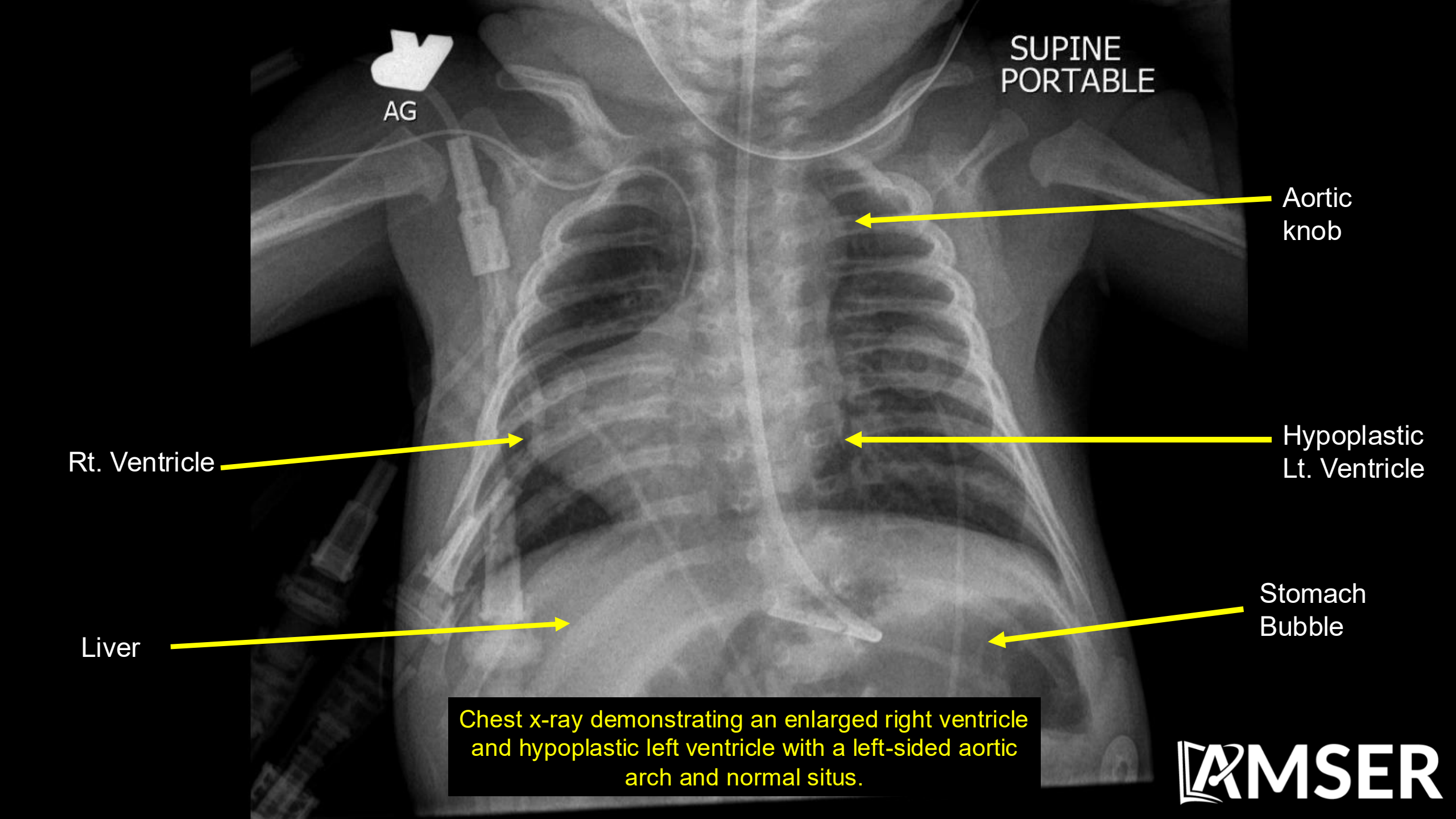
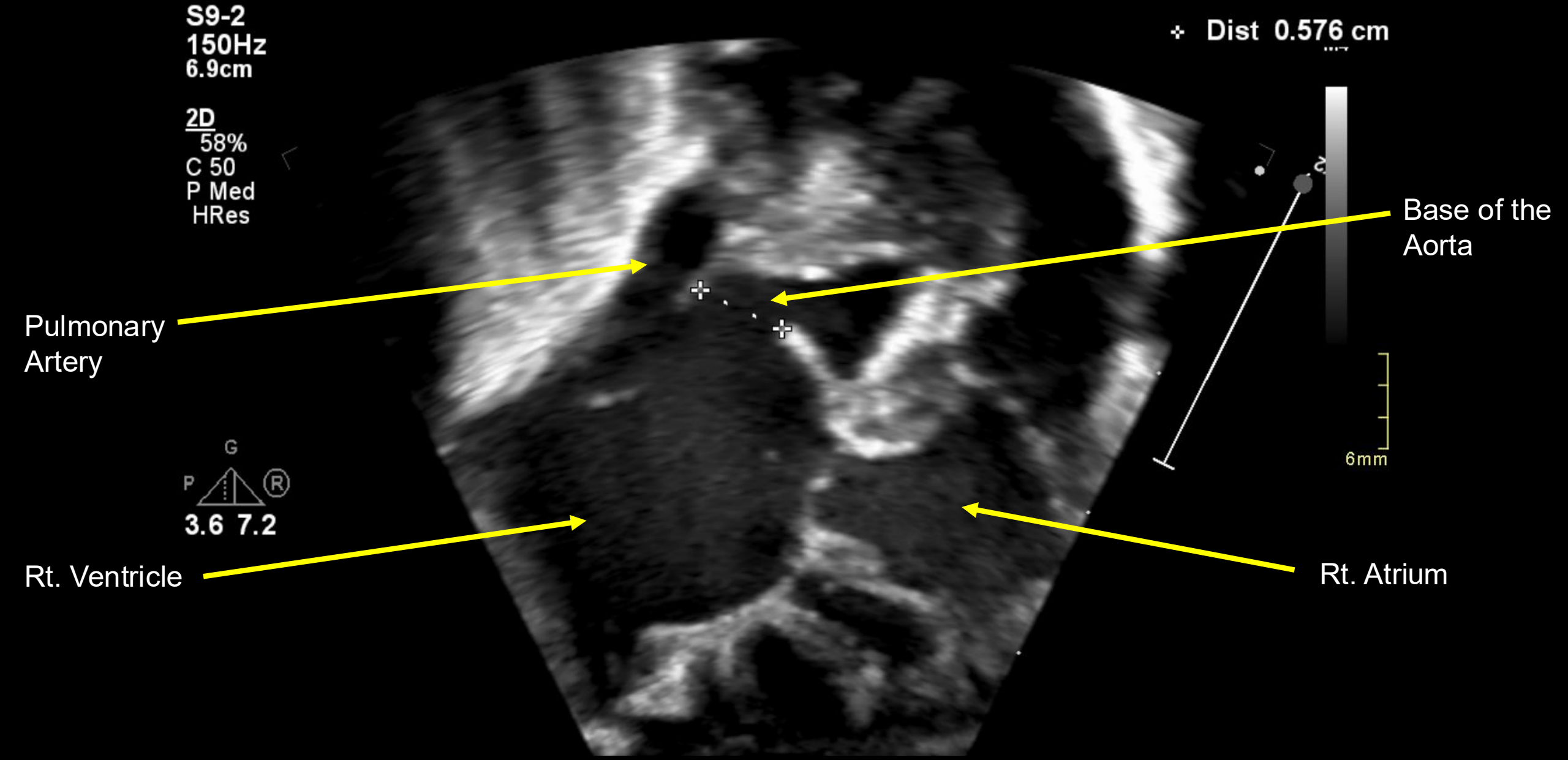

AG
SUPINE
PORTABLE



Chest x-ray demonstrating an enlarged right ventricle and hypoplastic left ventricle with a left-sided aortic arch and normal situs.


S9-2
150Hz
6.9cm
2D
58%
C 50
P Med
HRes
G
P
R
3.6 7.2
Dist 0.576 cm
6mm
163bpm



Echocardiogram showing both the pulmonary artery and aorta arising off the right ventricle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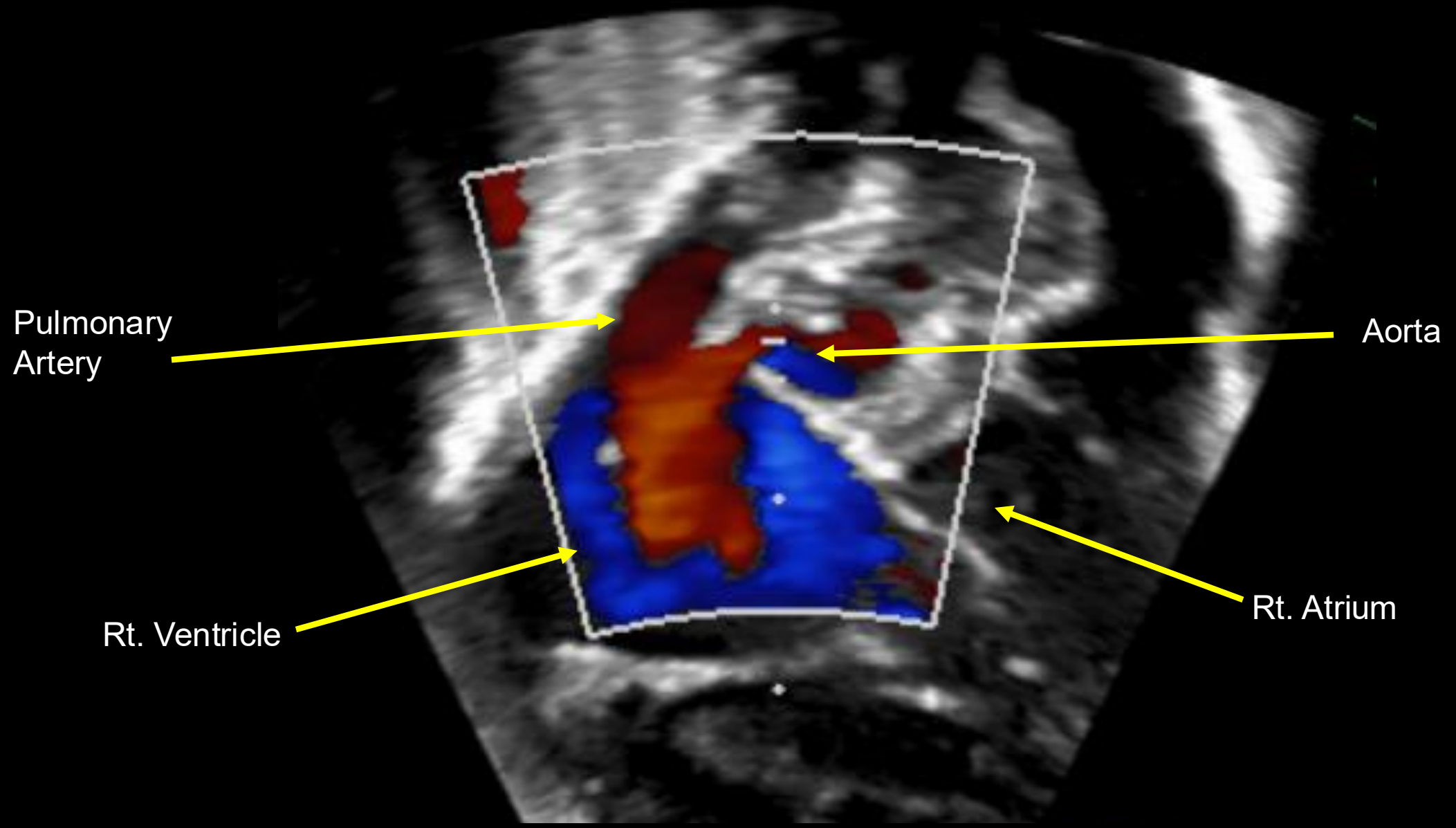
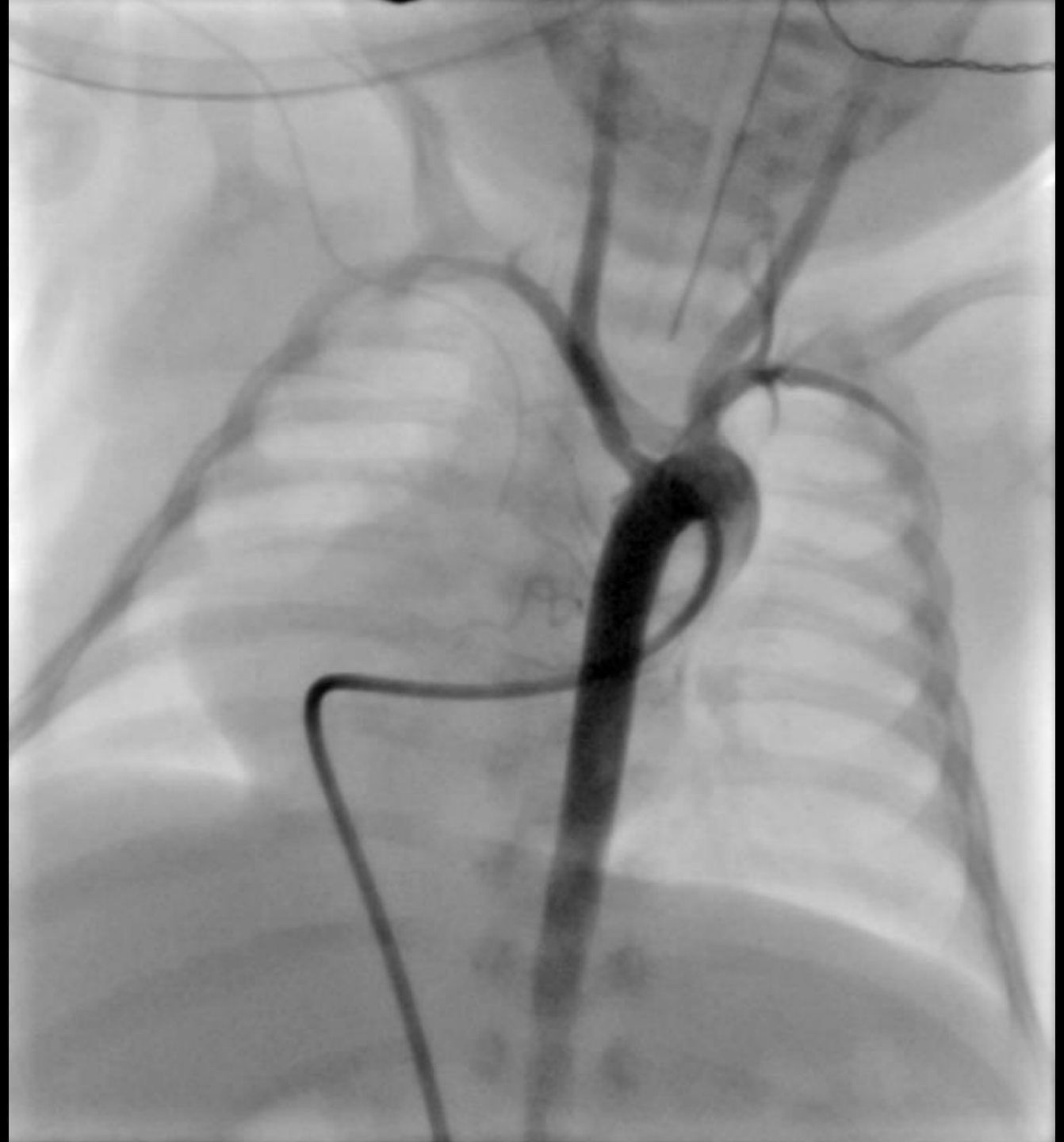
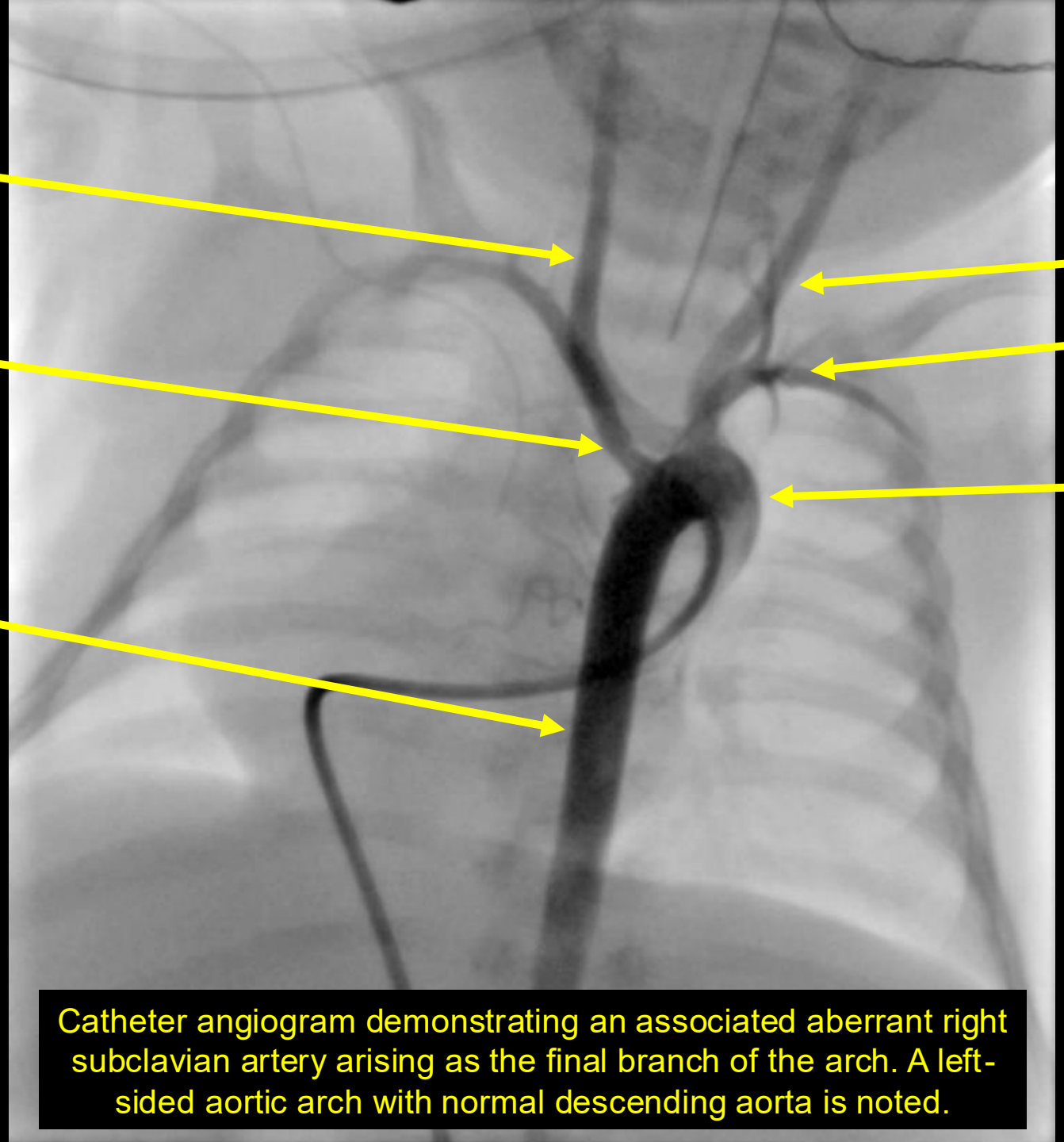


Echocardiogram showing both the pulmonary artery and aorta arising off the right ventricle





Catheter angiogram demonstrating an associated aberrant right subclavian artery arising as the final branch of the arch. A left-sided aortic arch with normal descending aorta is noted.

Final Dx:

Double outlet right ventricle (DORV)

Case Discussion

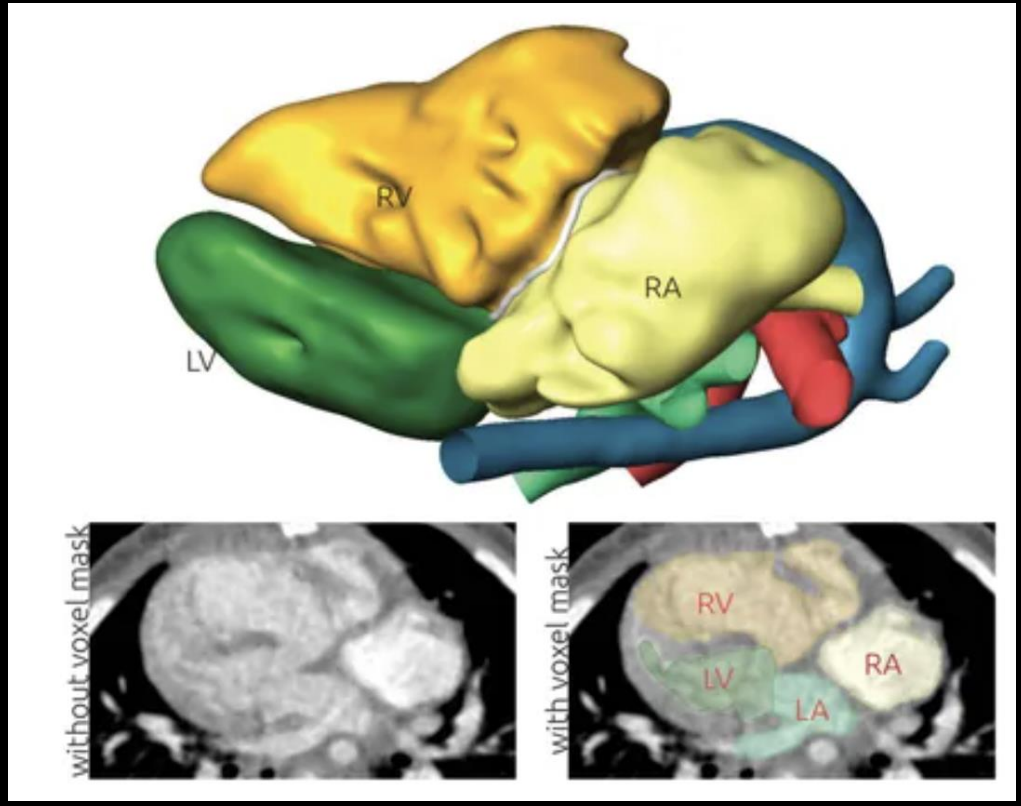
- DORV is a rare congenital heart defect with an abnormal ventriculoarterial connection in which both great arteries (pulmonary artery and aorta) connect to the right ventricle[4]
- The global incidence of DORV is about 1-1.5/10,000 live births, representing 1-3% of all congenital heart defects[6]
- Classification and surgical strategies differ considerably across centers due to wide anatomical variability[6]
- DORV is frequently associated with other cardiac malformations such as an obligate ventricular septal defect, atrioventricular septal defect, and outflow tract obstruction[6]
- The type of concurrent cardiac malformation significantly alters treatment, which includes biventricular vs. univentricular repair[6]
- Multimodal imaging is essential for preoperative planning and optimizing surgical outcomes[8]

Case Discussion

- Image Findings:
  - Chest X-Ray:
    - Mild to moderate cardiomegaly
    - Prominent right ventricle
    - Small left ventricular contour
    - Left sided arch most common, but right sided arch can occur
    - Normal situs (stomach bubble on left, liver shadow on right)
  - Echocardiography:
    - Initial diagnostic tool which shows both the aorta and pulmonary artery arising from the right ventricle[5,12]
    - VSD is always present as the left ventricle has no other outlet. [5,12]
    - Other cardiac malformations often present as well (atrioventricular septal defect, outflow tract obstruction).[6]
  - Angiography:
    - Provides detailed anatomical information and associations[2,9,10]
    - Useful for surgical planning[2,9,10]

Case Discussion

- Three-dimensional printing is increasingly used in the diagnosis and surgical treatment of congenital heart anomalies such as DORV[2]
- 3D printed models are derived from patient-specific CT or MRI imaging[2]
- The Radiological Society of North America recommends 3D printing for complex congenital heart anomalies where precise pre-op understanding can reduce operative and cardiopulmonary bypass time[2]



3D model used for surgical planning in complex double outlet right ventricle. Image adapted from Brüning et al., Frontiers in Cardiovascular Medicine (2022)

References:

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