Truncus Arteriosus

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06/08/21

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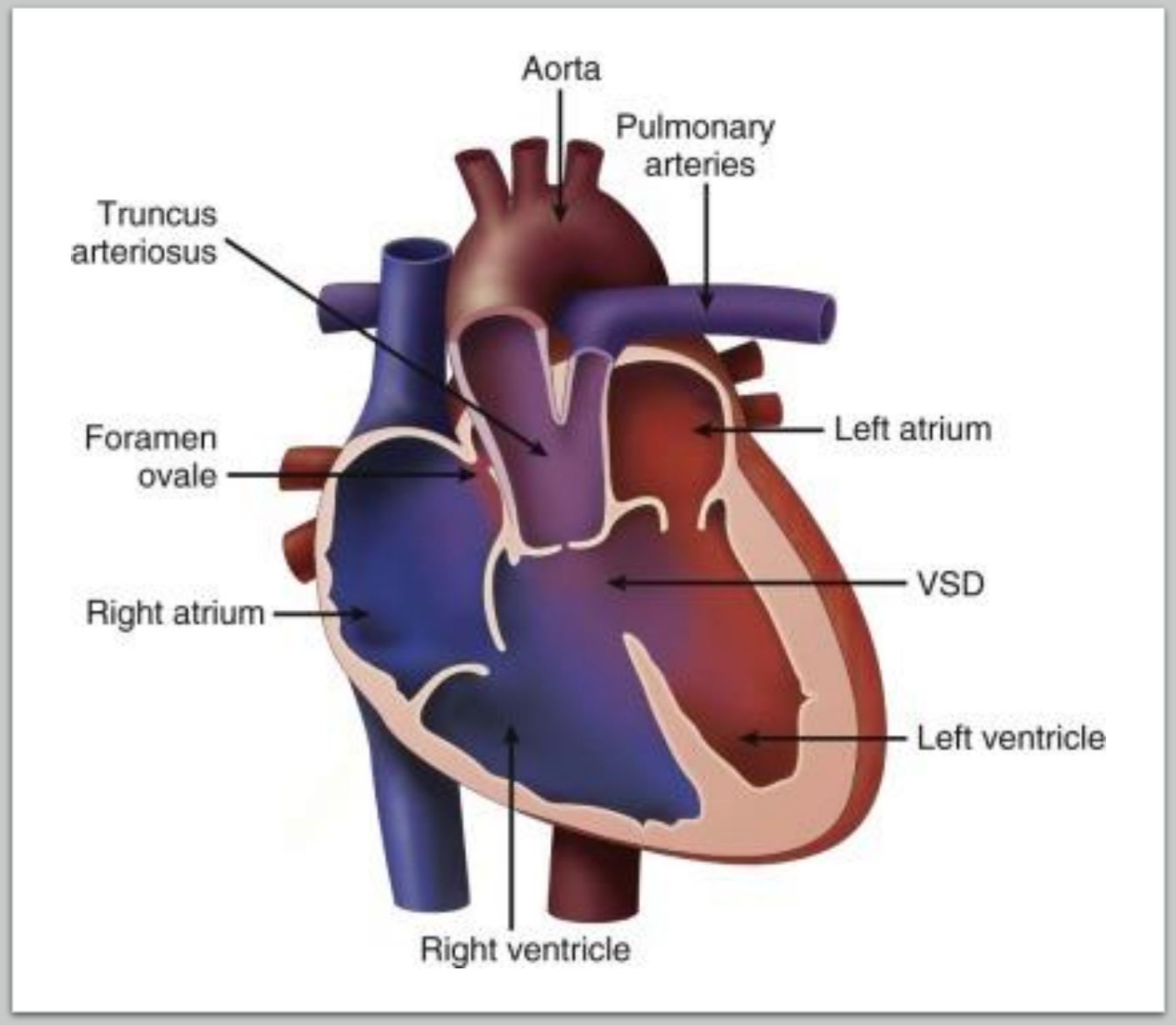
Outline

- Review Definition, Anatomy and classification
- Fetal echo diagnosis and differentials
- Prenatal counselling and Repair

Definition of TA

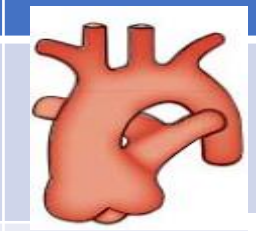
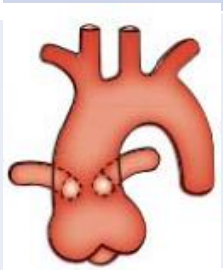
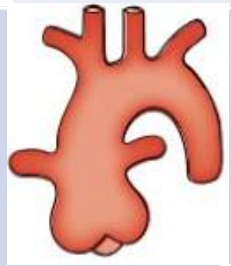
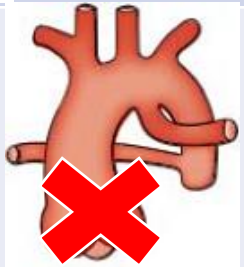
- Conotruncal defect with a single great arterial trunk and gives rise to
  - Aorta
  - At least one pulmonary artery
  - And at least one coronary artery
- Differentiating TA from PA(TOF,PA,MAPCAS),AA



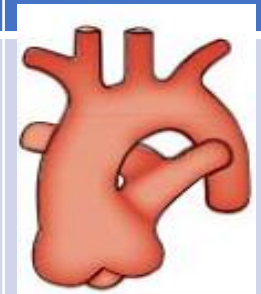
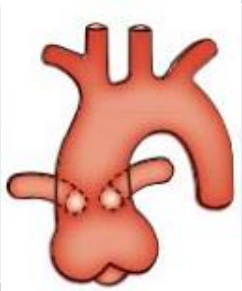
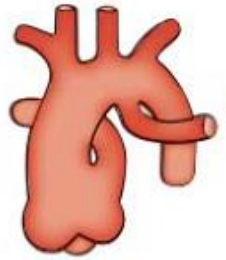
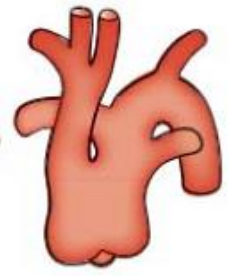
Classification and types

- Collett and Edwards
- Van Praagh
- Robert Anderson

Collet and Edwards

Type		
1	Branch PA s arise from a separate MPA segment	
2	Branch PA s arise directly from the trunk(with 1 single or 2 very close orifices)	
3	Branch PA s arise directly from the trunk (with 2 separate orifices on either side)	
4	Branch PA s arise from DA → Not Truncus! TOF/PA/MAPCAS	

Van Praagh(modified Weinberg)

Type		
A1	Branch PA s arise from l[illegible] MPA segment) Most common present in 60% of cases.	 PDA absent
A2	Branch PA s arise from trunk posteriorly(No MPA segment)	 PDA absent
A3	One branch PA is "isolated“ off a ductus arteriosus or comes from a collateral artery	PDA present 
A4	Discrete coarctation, tubular hypoplasia or interrupted aortic arch	PDA present 

Robert Anderson

- Common arterial trunk with **aortic dominance** (RPA and LPA arising from a confluent pulmonary segment)
- Common arterial trunk with **aortic dominance** (RPA and LPA arising separately from posterior aspect of intrapericardial trunk)
- Common arterial trunk with **aortic dominance** (RPA arising from trunk and LPA from ductus arteriosus or collateral)
- Common arterial trunk with **pulmonary dominance** (Branch PA s arising from a confluent pulmonary segment and aortic arch is interrupted)

Other anatomic features

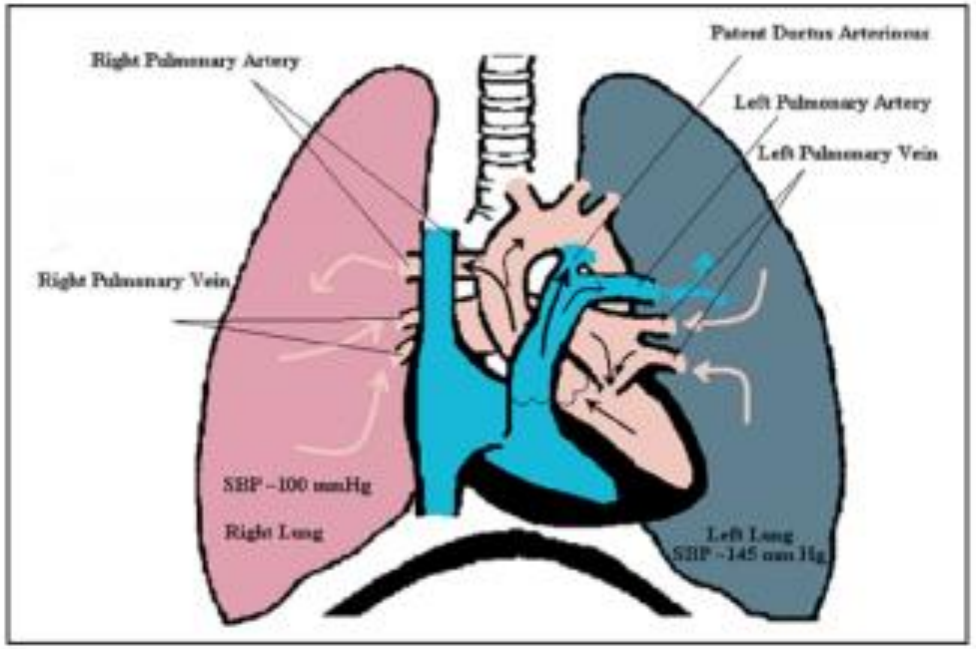
- Outlet VSD
- No sub truncal conus typically
  - Mitral valve to truncal valve continuity
  - Truncal valve sits over both ventricles and VSD
- Truncal valve morphology: dysplastic valve, variable number of leaflets and asymmetry. Stenosis or insufficiency.
  - 3>4>2>5>1
- Coronary artery origins: abnormal or eccentric, narrow or slit like or single coronary artery
- Right aortic arch is common

Associations

Associated findings	% of patients
Coronary artery anomalies	30-50%
Right aortic arch	30-40%
ASD	10-20%
Interrupted Aortic arch	10-20%
LSVC to CS	5-15%
Di George syndrome	Up to 35%
Extracardiac anomalies	Up to 30%

Hemitruncus

- Anomalous origin of one of the branch pulmonary arteries from the aorta and the other arises normally from the right ventricle in the presence of two semilunar valves.
- R Hemi truncus common with RPA from Aorta and LPA coming from pulmonary valve



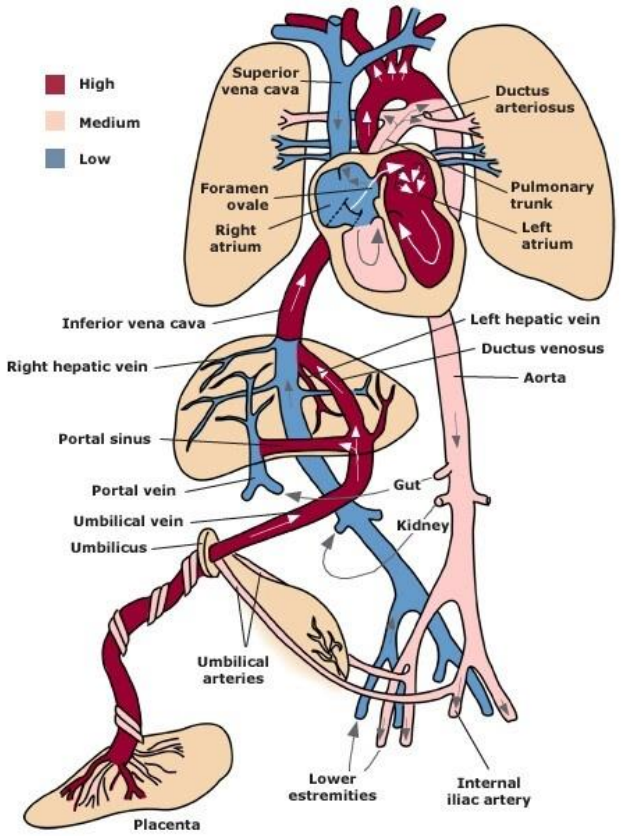
Gaint AP window

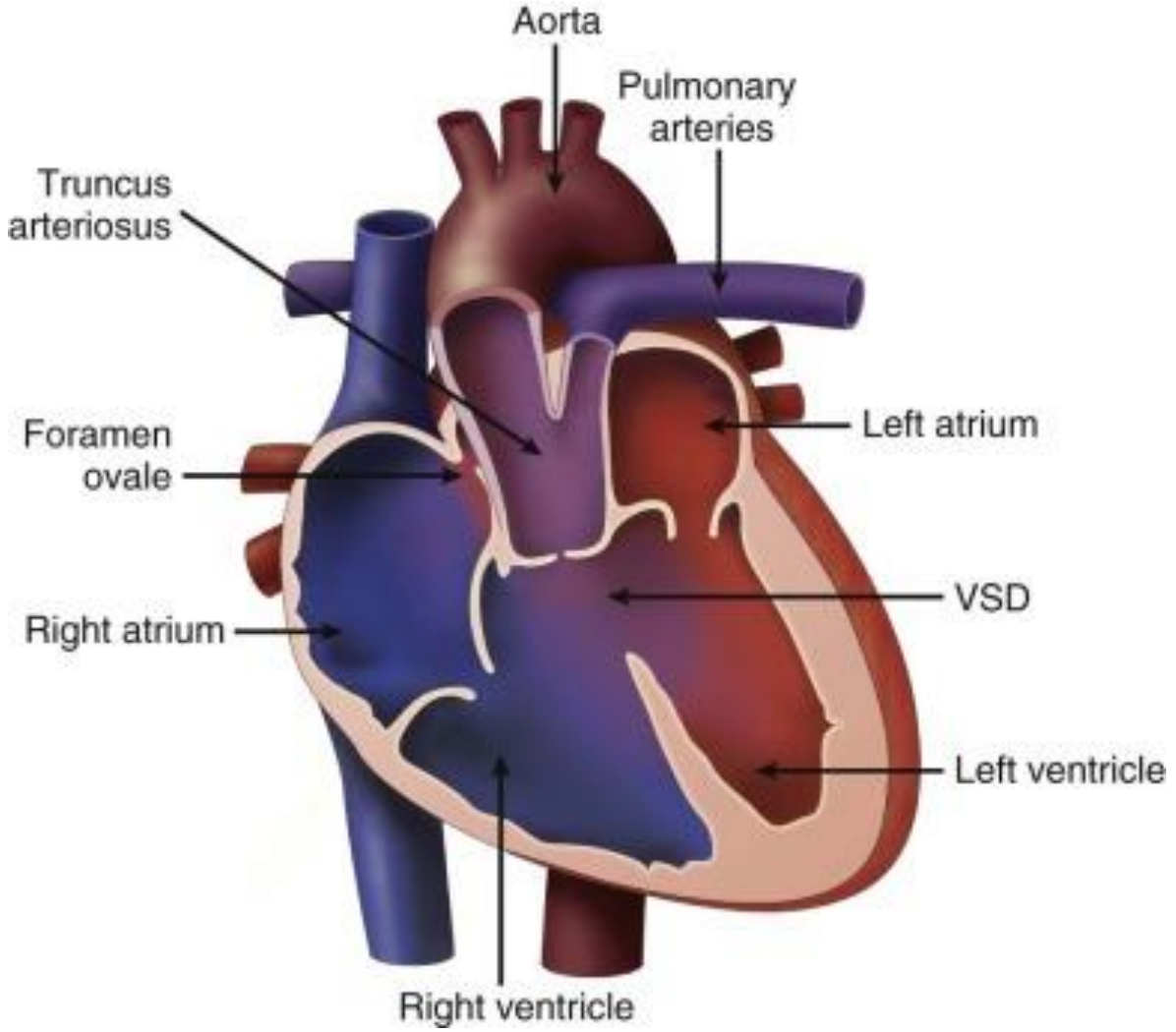
- 2 semilunar valves, no VSD
- Absence of aorticopulmonary septum and no clear demarcation between aorta and pulmonary artery

Epidemiology, Genetics

- TA - 1% to 2% of all forms of CHD.
- Extracardiac anatomic anomalies and chromosomal abnormalities exist in nearly 1/2 of patients with TA
- Most common genetic abnormality is microdeletion of chromosome 22q11 (22q11 del)
  - Over 1/3rd of patients with TA have been identified to have 22q11 del
  - 50% of cases with an associated interrupted aortic arch (Van Praagh type IV) have 22q11 del.

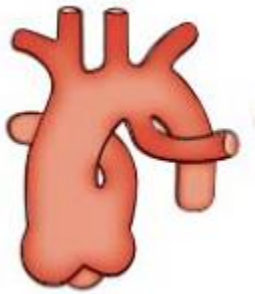
Prenatal physiology





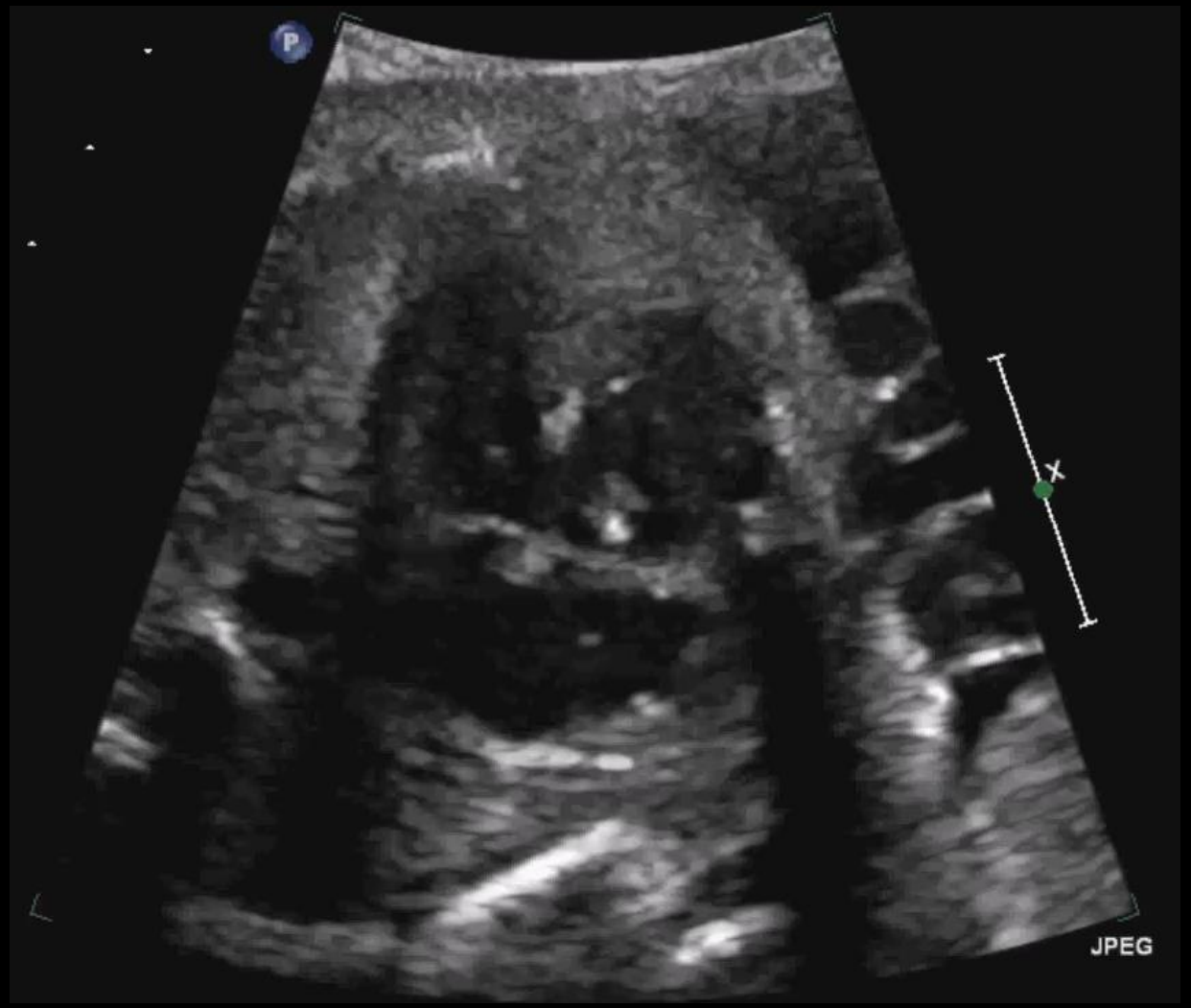
Prenatal physiology

- Significant abnormalities of truncal valve result in fetal distress in TA
- Stenosis and regurgitation tend to occur together in dysplastic Truncal valves leading to ventricular dilation, heart failure, hydrops.
- If the pulmonary arteries are discontinuous like in TA Van Praagh type III, disparity in flow between the lungs may cause differences in pulmonary vascular development, which may be problematic after birth.
- IUGR is common in fetus with DiGeorge



Fetal echo findings

- Identify the origin and course of the branch pulmonary arteries.
- PA origins should be identified to appropriately classify the type of TA and determine whether postnatal PgE therapy is needed or not.
- The base of the trunk should be identified in either long- or short-axis view and a slow, careful sweep cephalad should reveal the origin of the pulmonary arteries. Alternatively, if not found in this manner, a reverse search back from “lung-to-trunk” may be helpful.
- By identifying the branch pulmonary artery in the lung parenchyma using color Doppler imaging, one can track the branch pulmonary artery vessel back to the heart and trace its origin from the trunk.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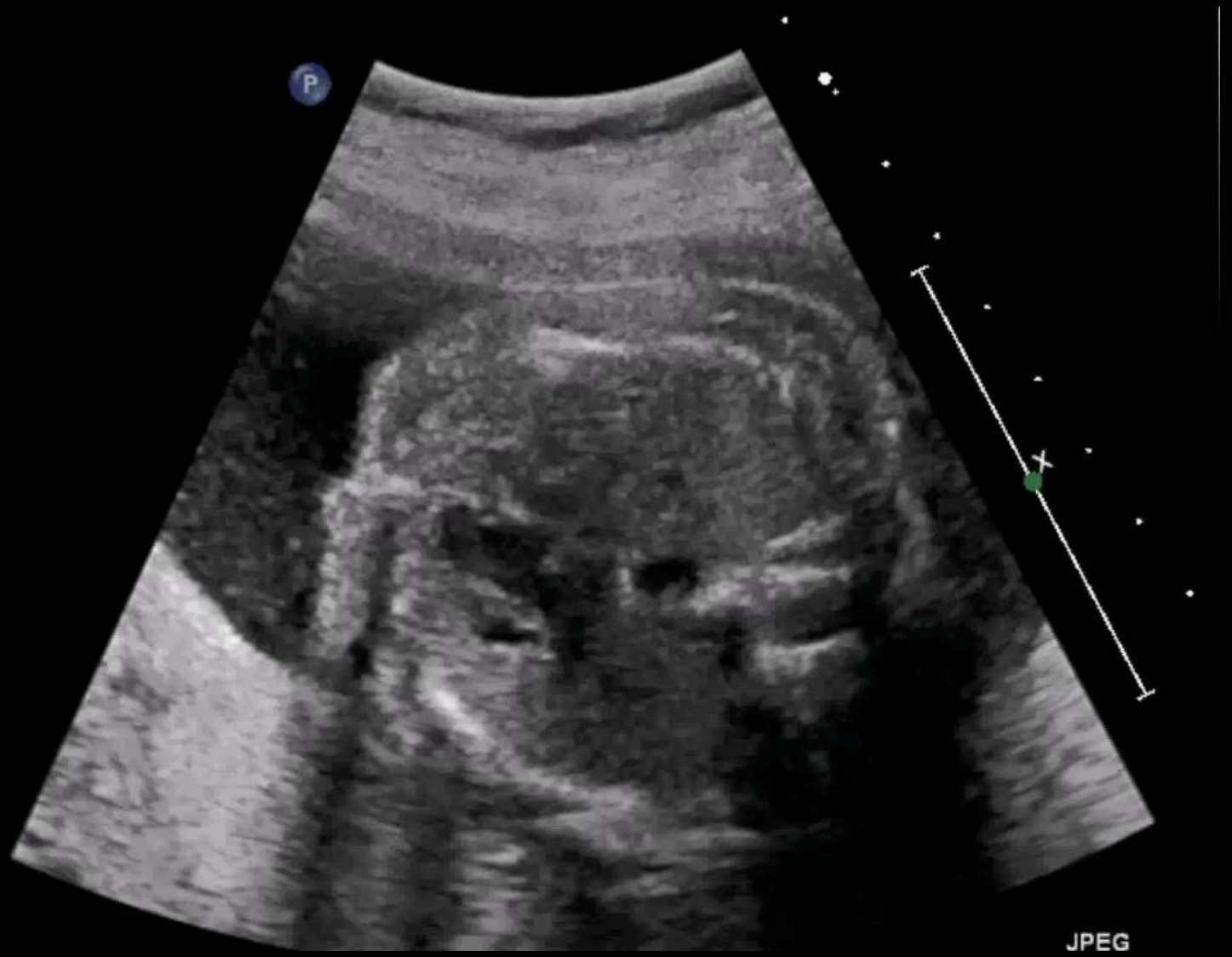
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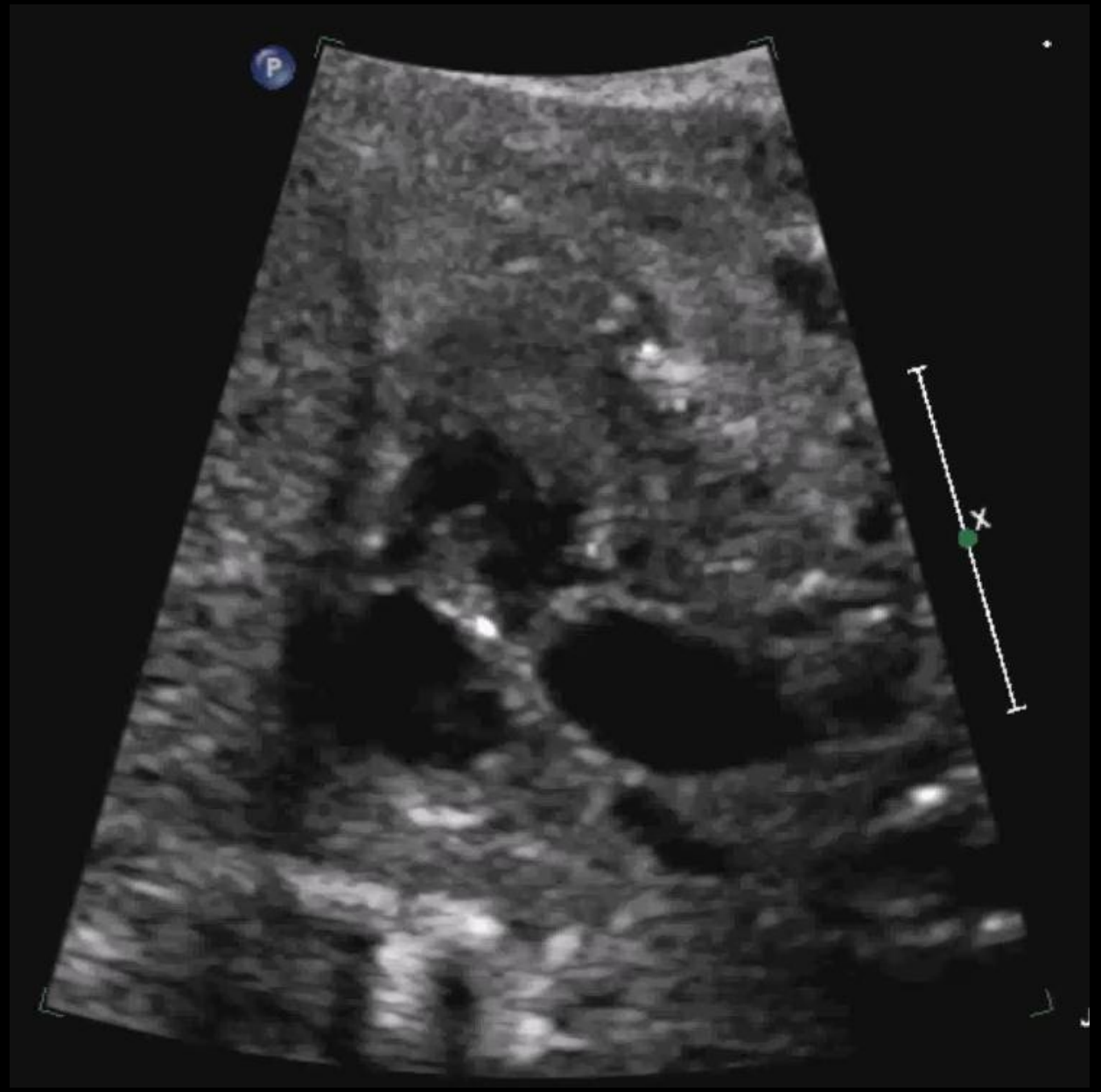
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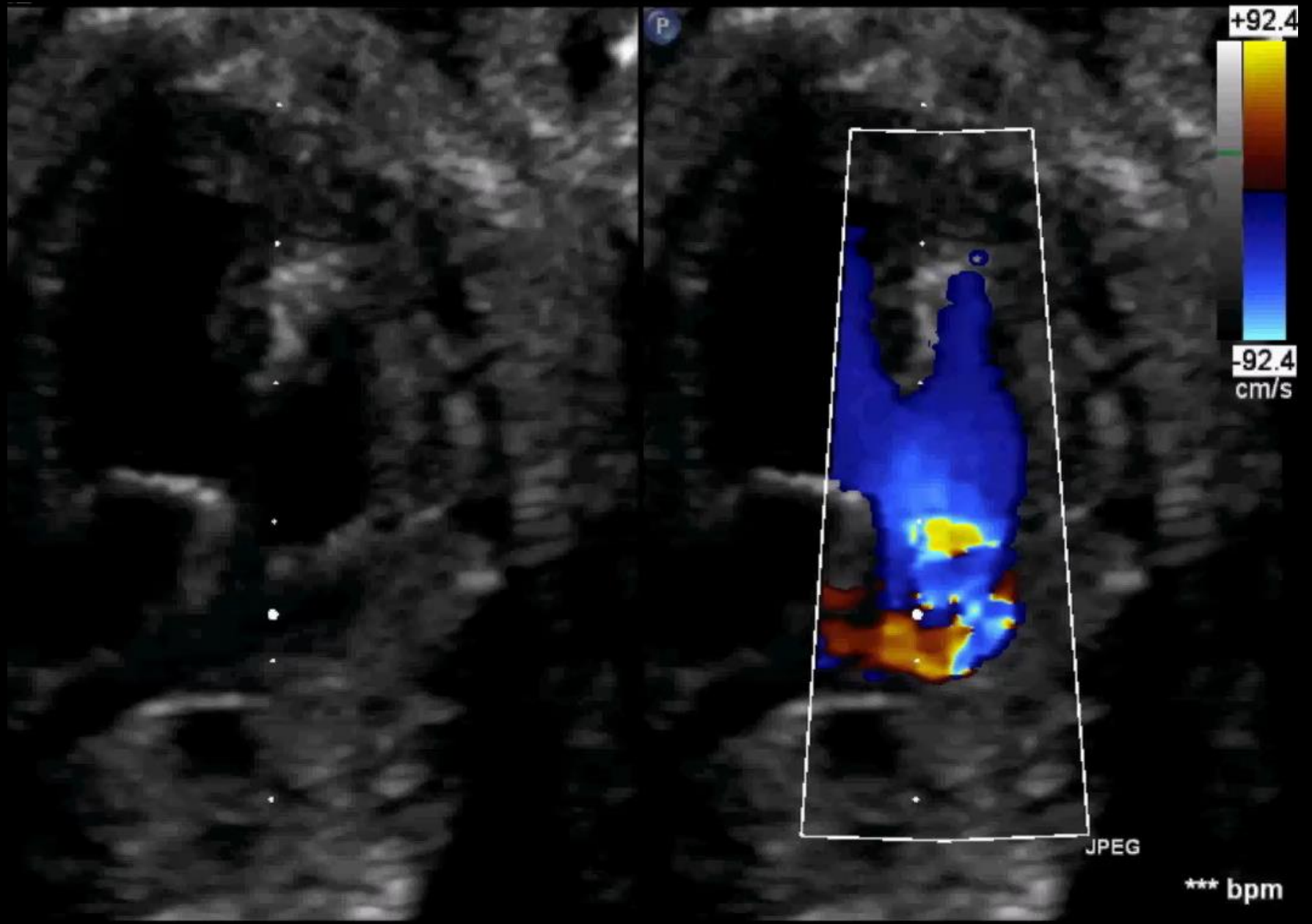
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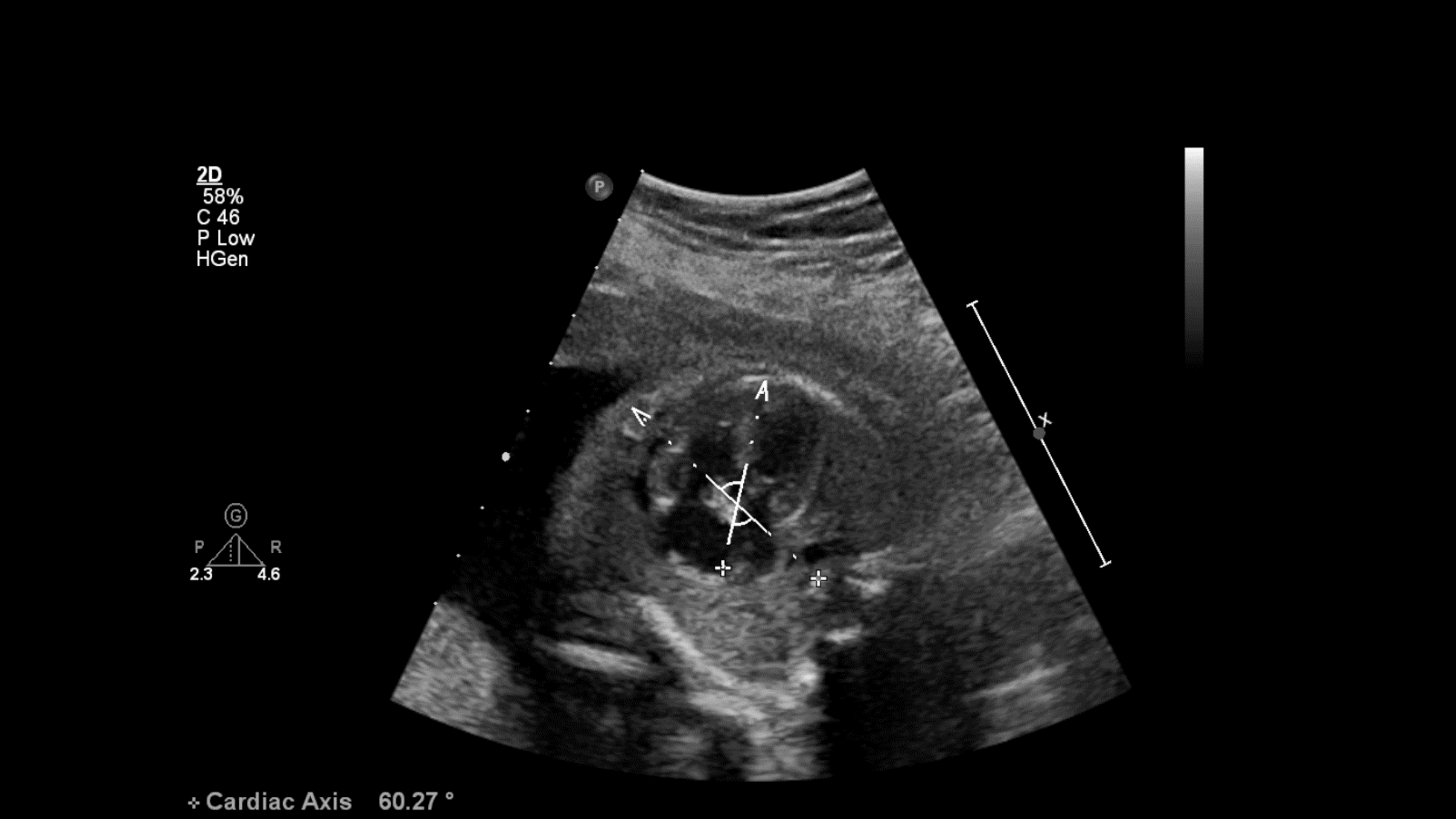
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C 46
P Low
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2.3
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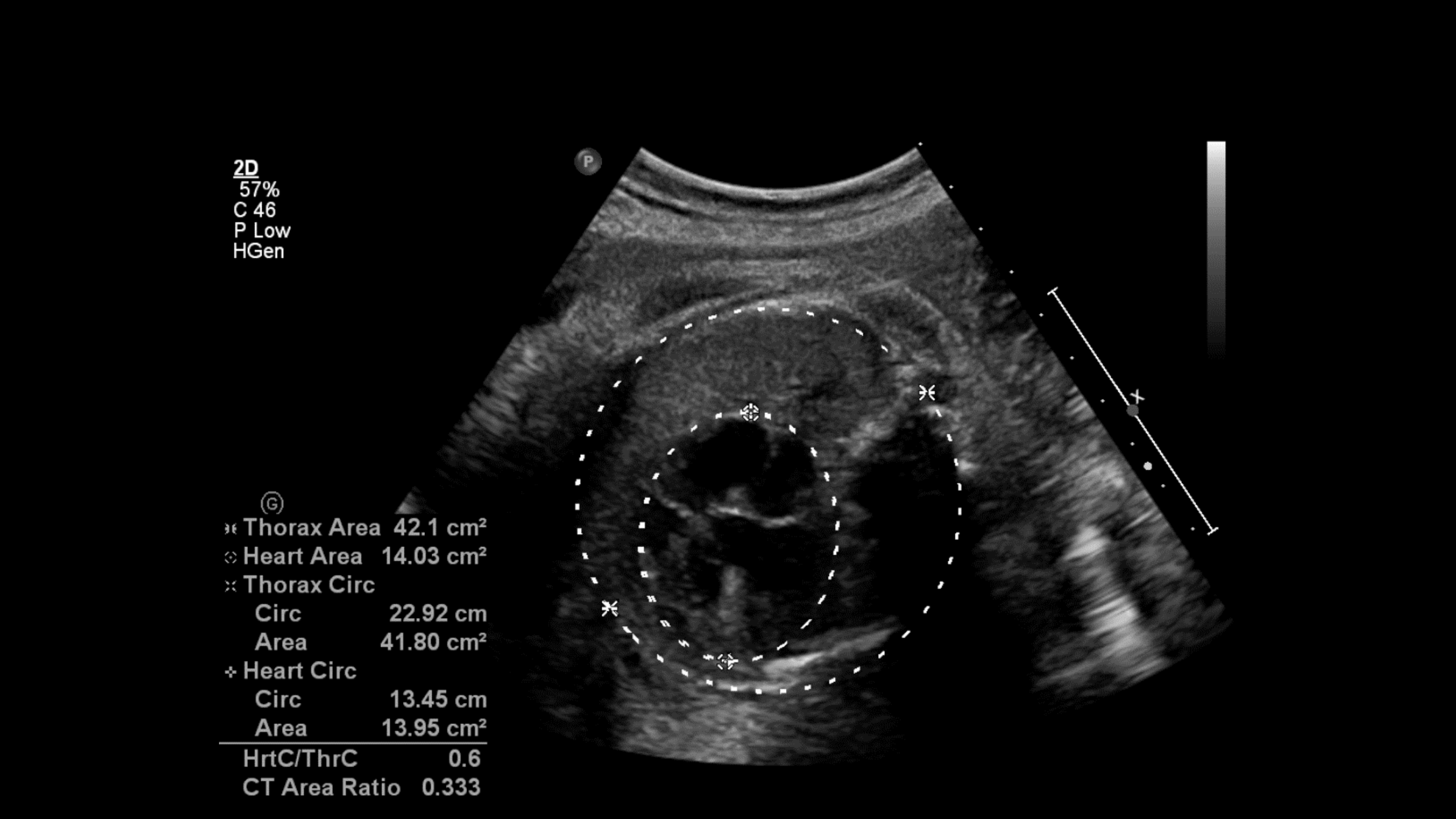
Cardiac Axis 60.27 °


2D
57%
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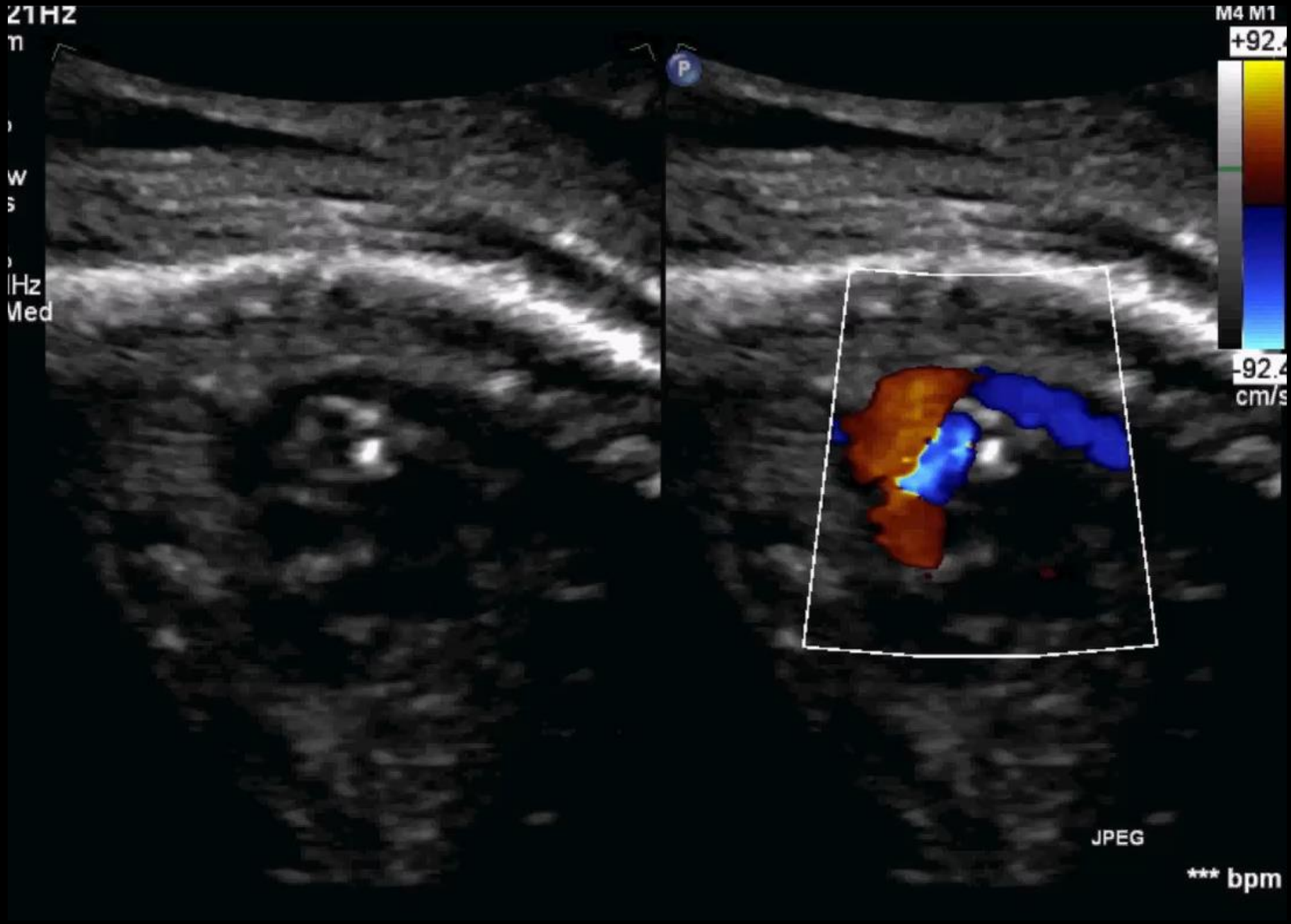
Thorax Area 42.1 cm²
Heart Area 14.03 cm²
Thorax Circ
Circ 22.92 cm
Area 41.80 cm²
Heart Circ
Circ 13.45 cm
Area 13.95 cm²
HrtC/ThrC 0.6
CT Area Ratio 0.333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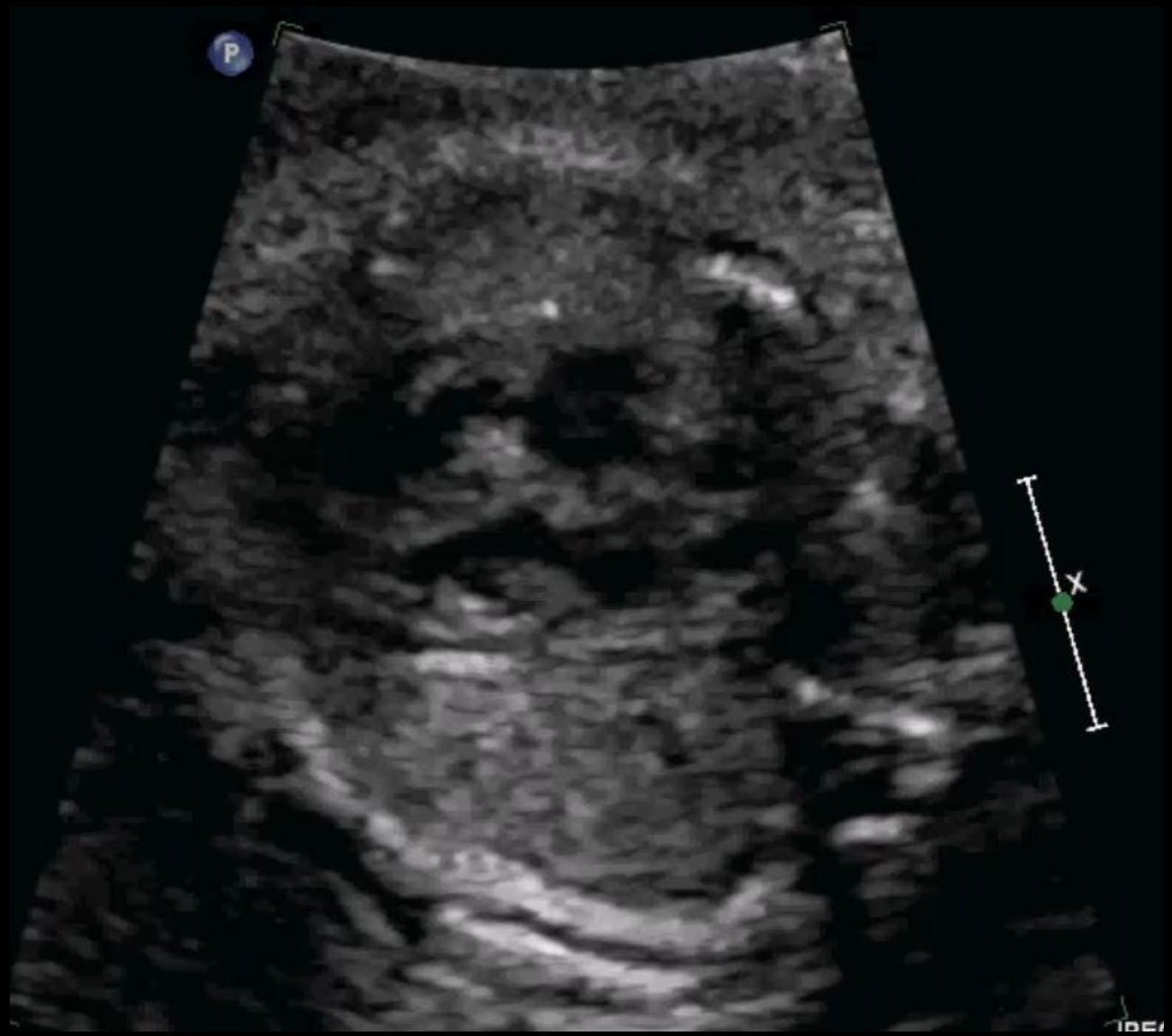
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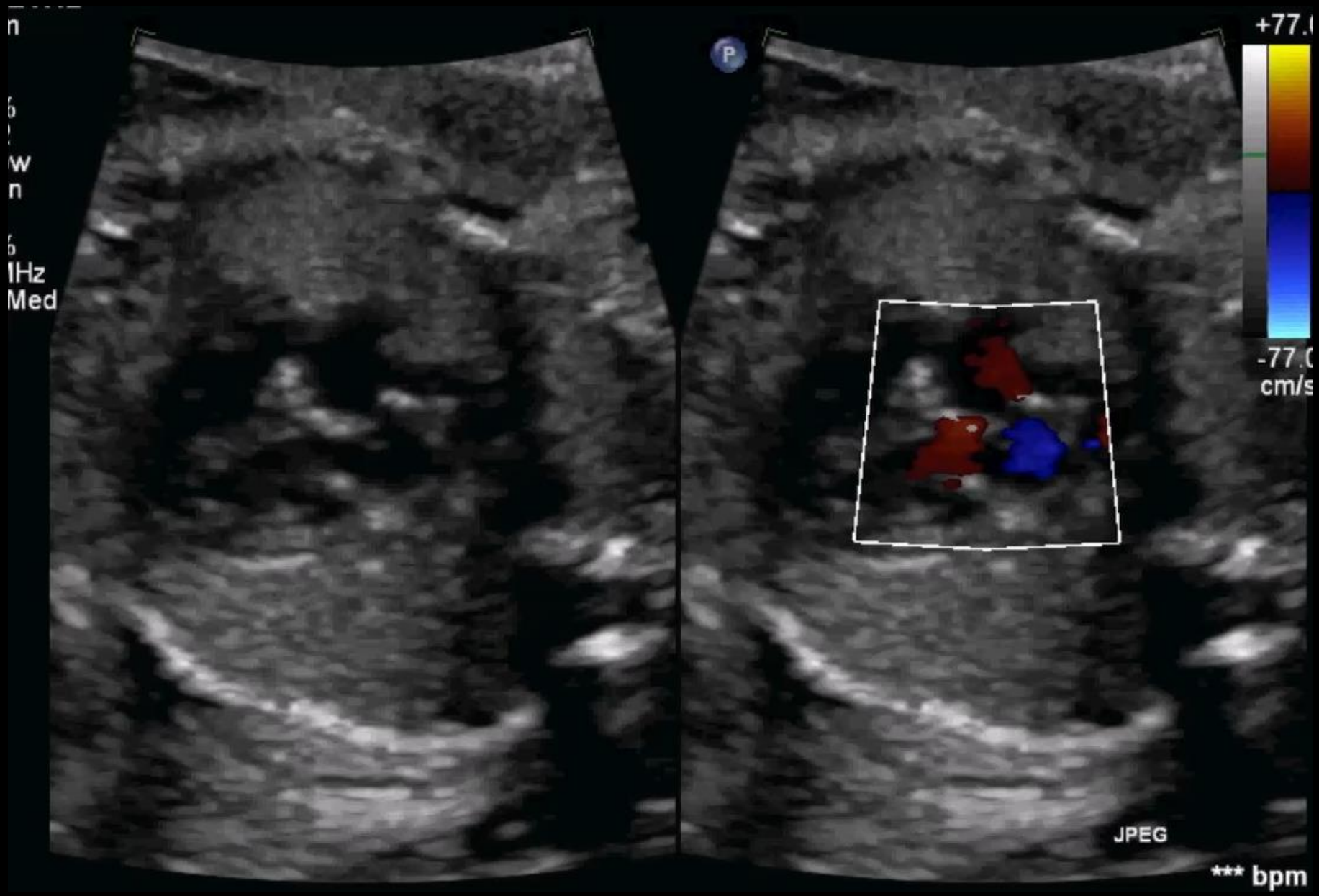
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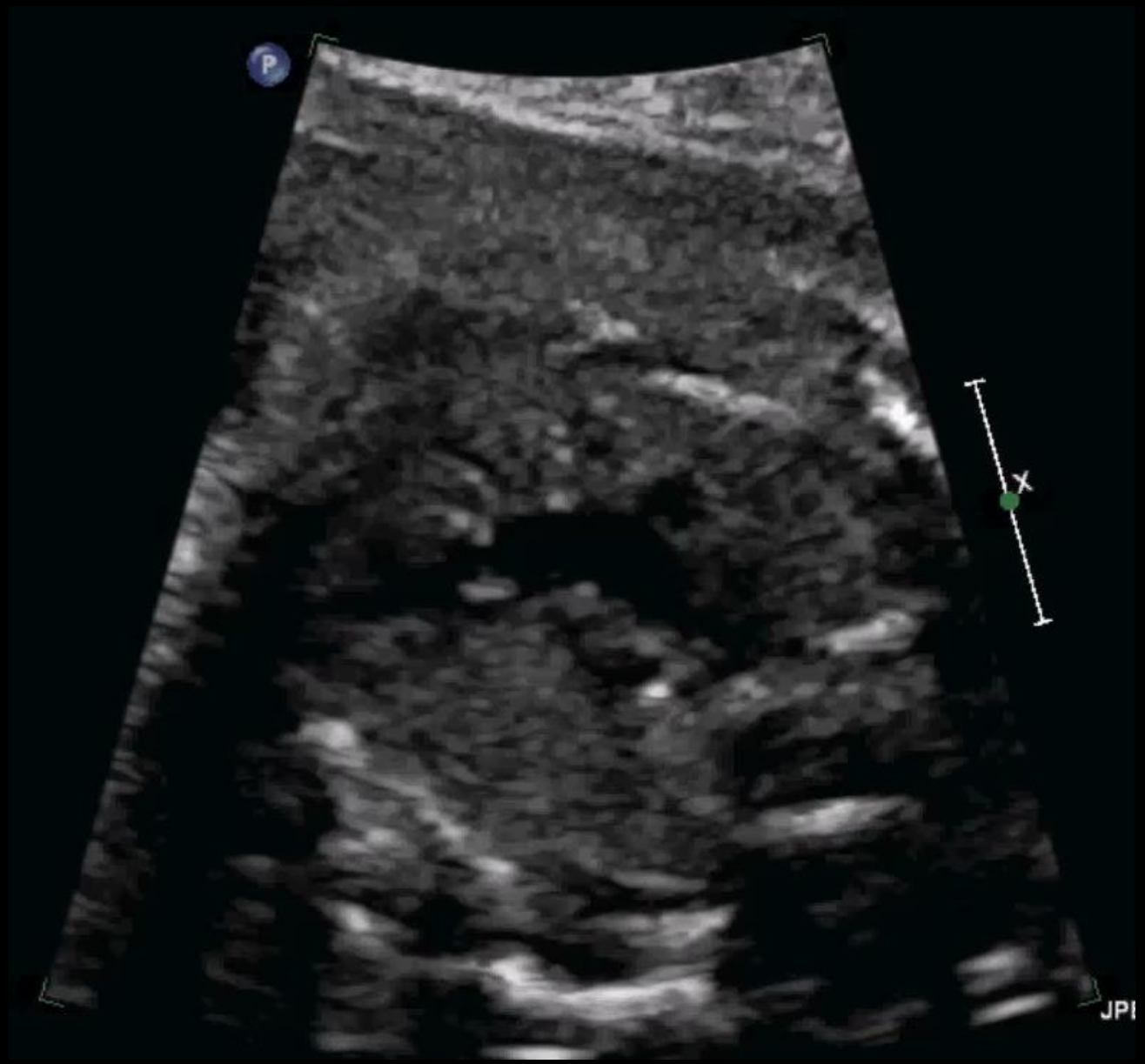
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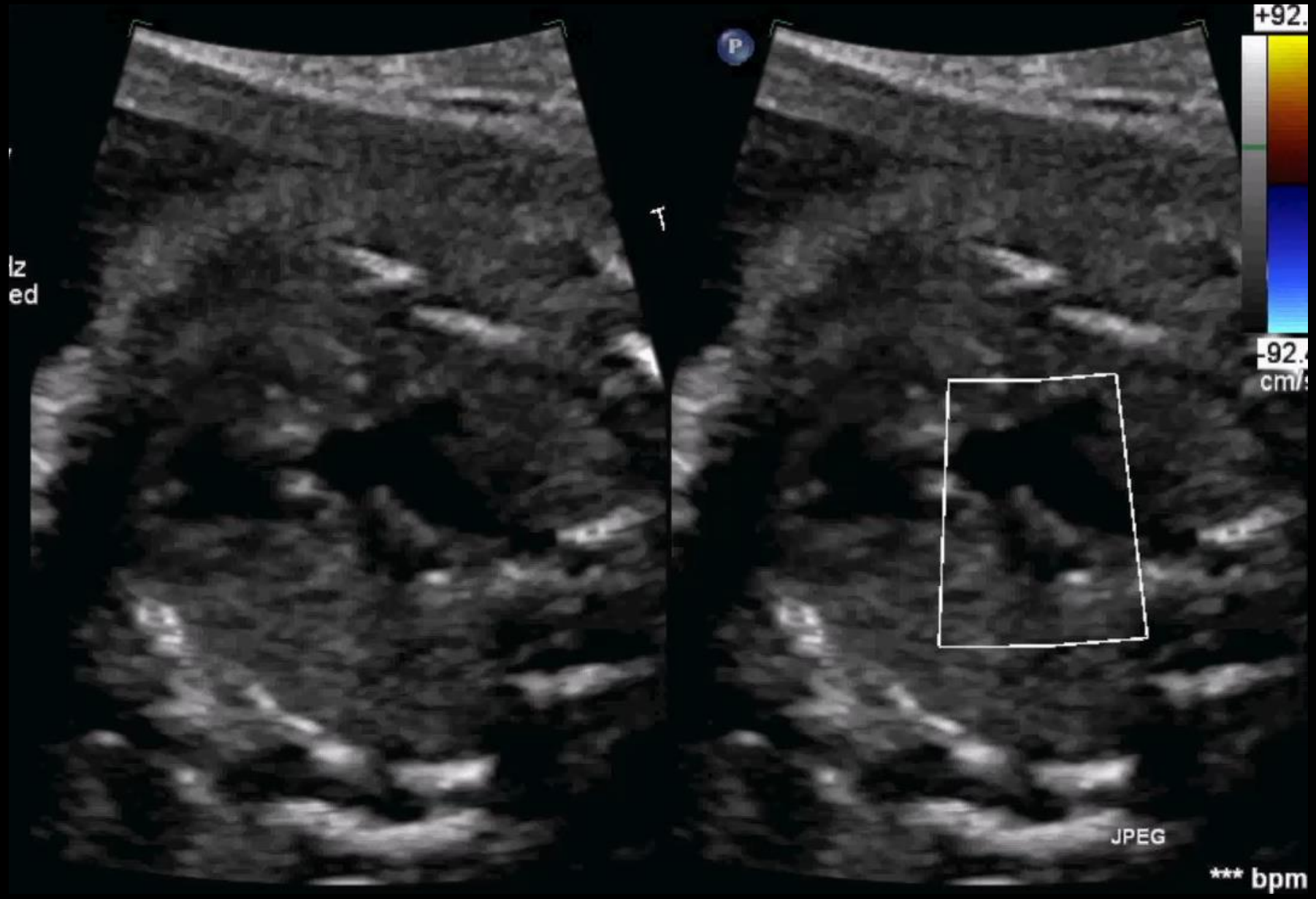
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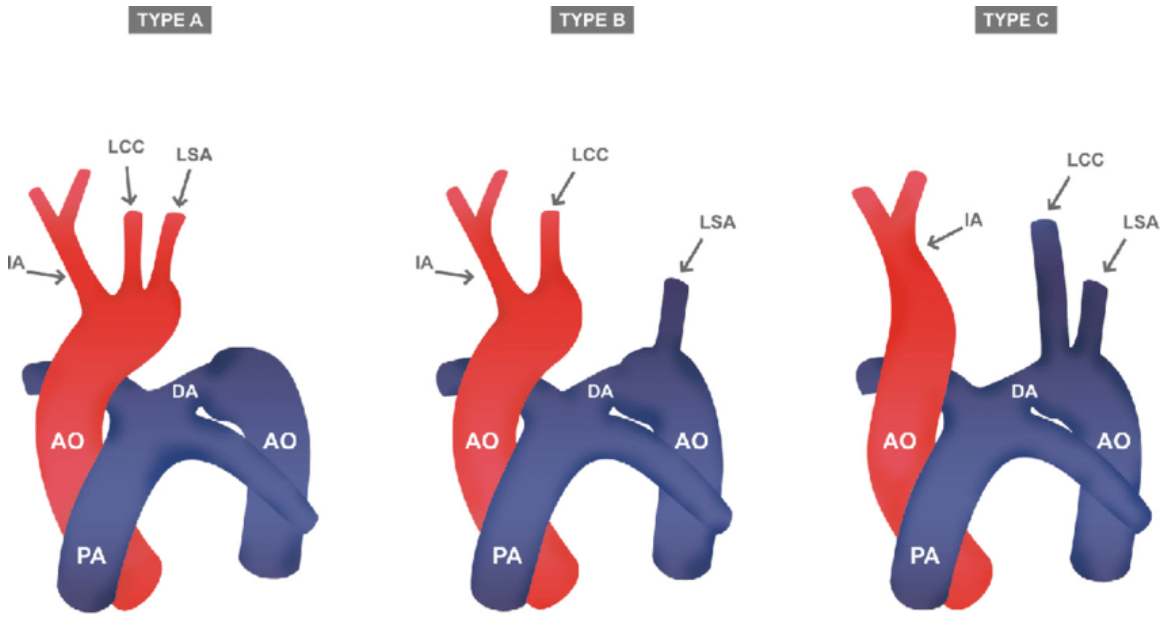
Type 4 Truncus with Interrupted aortic arch

- This combination is very rare
- Associated with a high rate of early mortality.



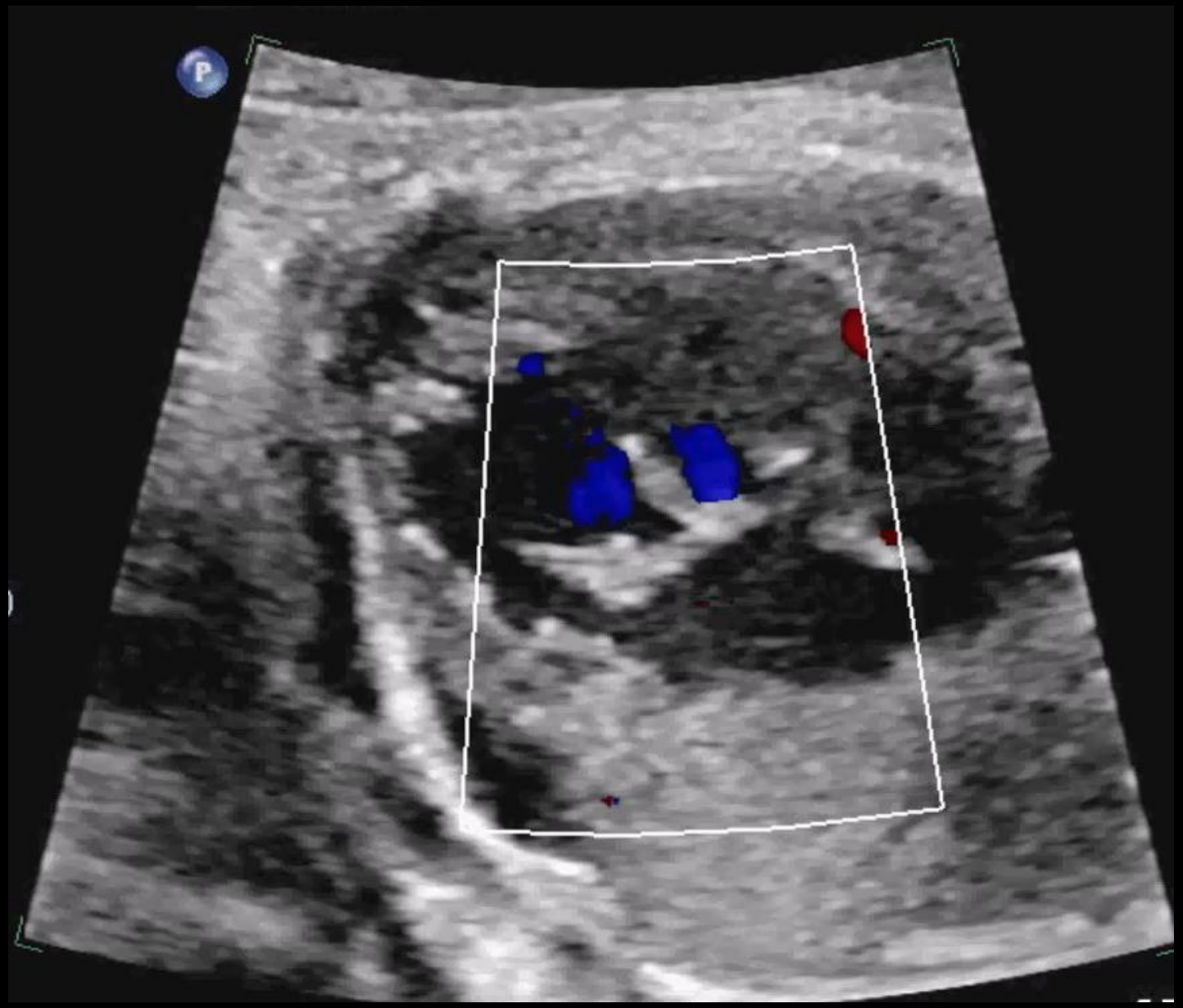
-Type B most common
-**Frequently** associated with 22q 11 del.

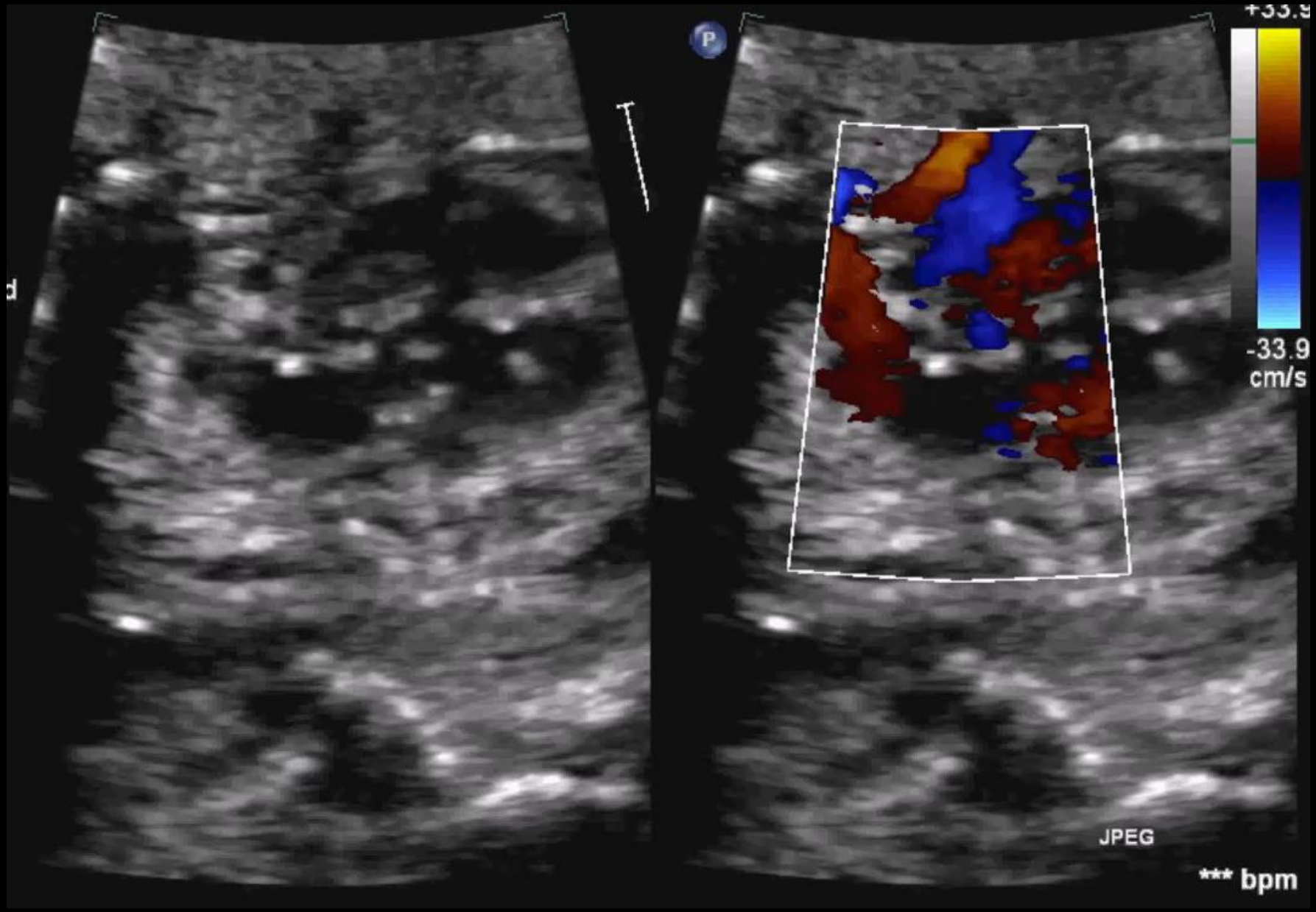
Differential diagnosis

- When a pulmonary valve and right ventricular outflow tract cannot be identified on fetal echo, differentiation between TA and other congenital heart defects with semilunar valve atresia can be difficult.
- TOF-PA, PA/VSD and aortic atresia with ventricular septal defect, involve a single large semilunar valve and a ventricular septal defect.

TA	TOF/PA
One common trunk	Separate main pulmonary trunk identified even if there is no flow in it
At least one branch PA arises from trunk	No PA artery arises from aorta without an interposed ductus arteriosus or collateral artery
Flow in Pulmonary arteries pulsatile	Continuous
Pulmonary arteries arise before the take-off of the first brachiocephalic vessel	Pulmonary arteries are supplied either by aortopulmonary collateral arteries or a ductus arteriosus, distal to the first brachiocephalic vessel

TA	PA/VSD/MAPCAS
Pulmonary arteries arise before the take-off of the first brachiocephalic vessel	Pulmonary arteries are supplied either by aortopulmonary collateral arteries or a ductus arteriosus, distal to the first brachiocephalic vessel
Flow in Pulmonary arteries pulsatile	Continuous

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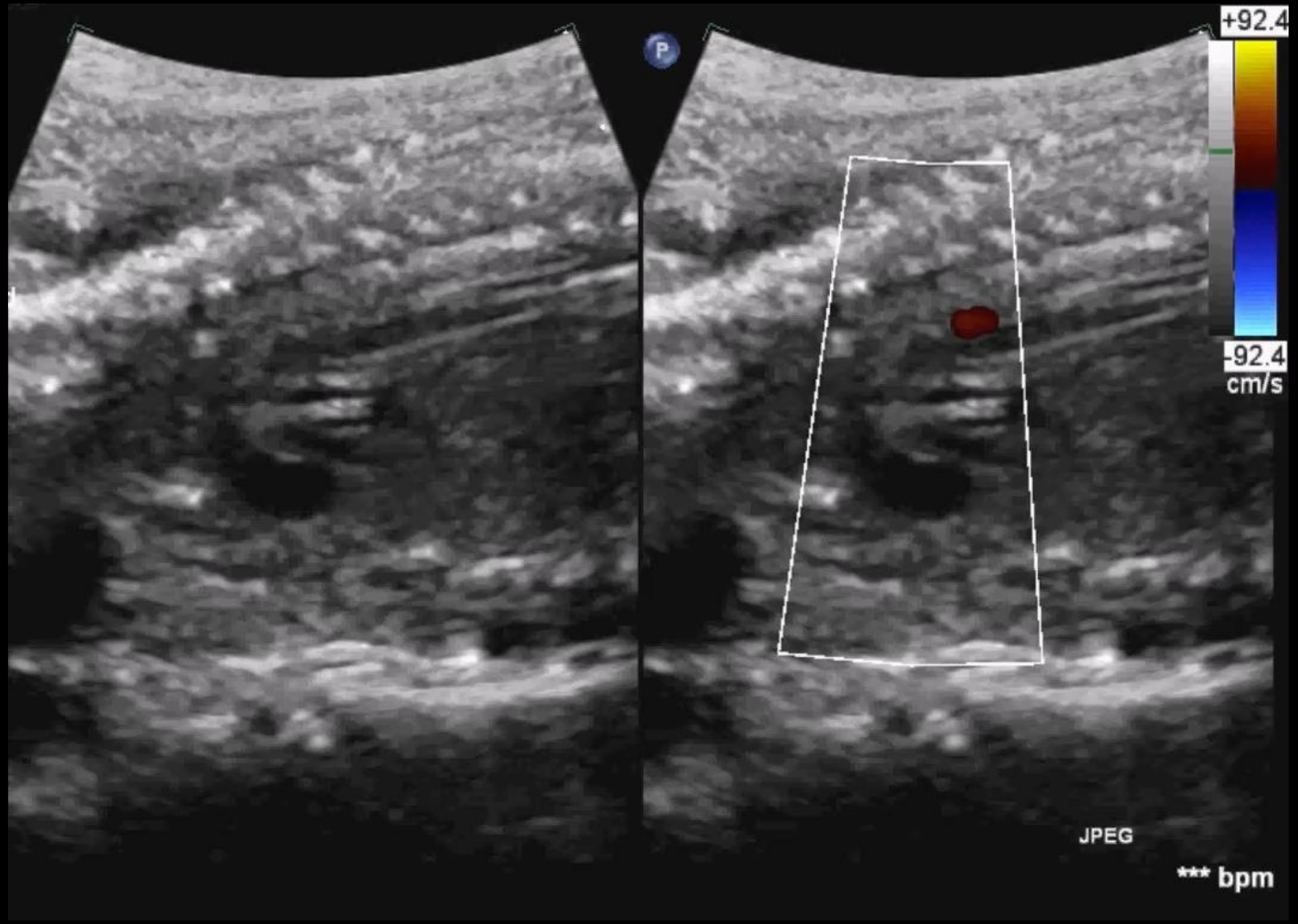
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Truncus Arteriosus: Diagnostic Accuracy, Outcomes, a Impact of Prenatal Diagnosis

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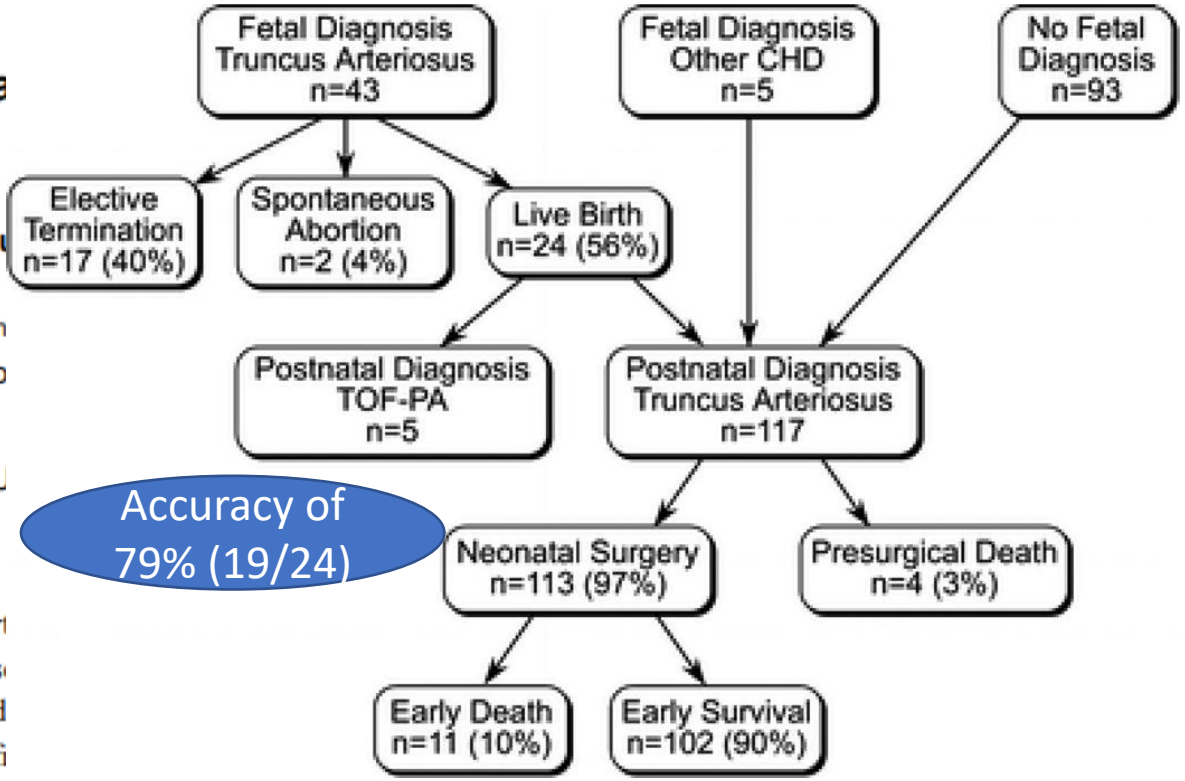
Abstract

Limited data exist on the impact of prenatal diagnosis and outcomes of fetal truncus art (TA). We sought to assess prenatal diagnostic accuracy and prenatal outcomes in fetus and compare postnatal outcomes in neonates with prenatally and postnatally diagnosed Records were reviewed for patients diagnosed with TA in utero or at ≤60 days of life fi 2007. Forty-three (32%) of 136 TA patients had prenatal diagnosis. Five patients with TA were prenatally misdiagnosed, and 5 with other congenital heart diseases were misdiagnosed with TA prenatally. Of 28 fetuses diagnosed at <24 weeks gestation, 19 (68%) did not survive to birth because of spontaneous fetal death ($n = 2$) or because of elective termination ($n = 17$). Pregnancy termination was not more likely for fetuses with extracardiac anomalies. Of 19 live-born patients with correct prenatal diagnosis of TA, 2 (11%) died before surgery, and 4 (24%) died in the early postoperative period. All patients who died presurgically had been diagnosed prenatally. Overall, early postoperative mortality was 10%. Prenatal diagnosis of TA remains challenging and is associated with a high rate of elective termination. Fetal diagnosis was associated with younger age at repair but was not associated with improved neonatal survival.



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Accuracy of Fetal Echocardiography in the Differential Diagnosis between Truncus Arteriosus and Pulmonary Atresia with Ventricular Septal Defect

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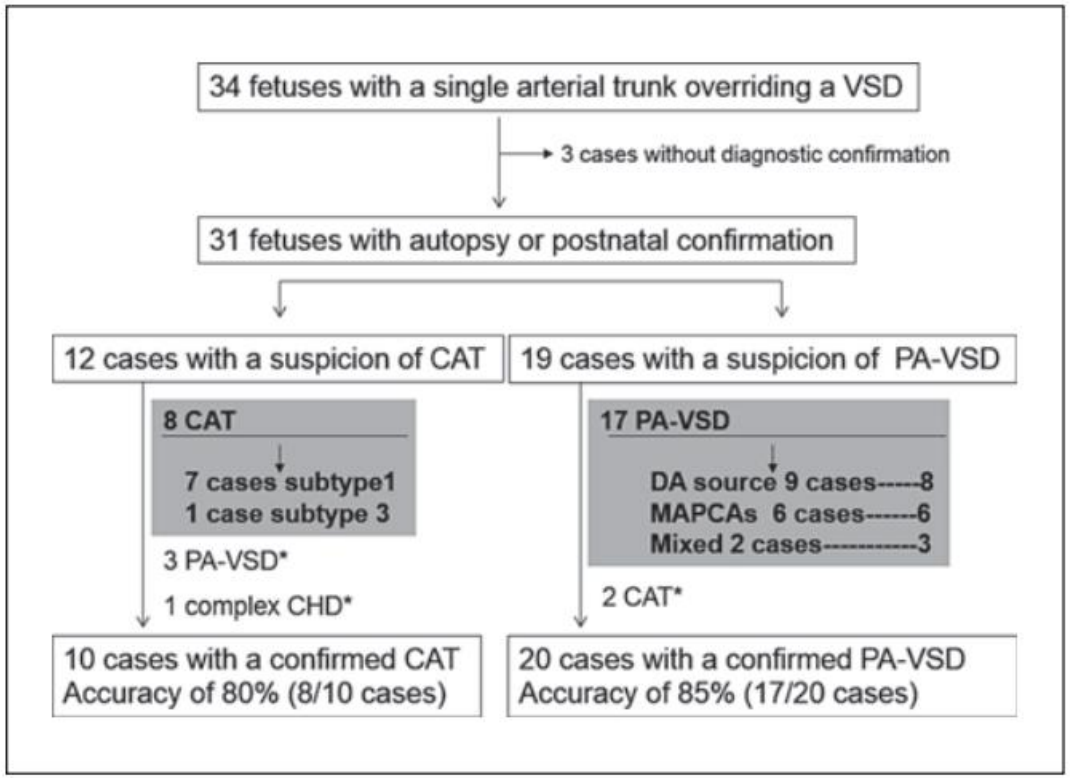


Fig. 1. Flowchart of cases included in the study. * Misdiagnosed cases

Follow up fetally

- In the absence of severe truncal valve abnormalities or extracardiac abnormalities, the fetus with TA is usually stable.
- Serial fetal echocardiography studies are recommended to delineate the morphology and competency of the truncal valve
- If abnormalities of TV stenosis or regurgitation are present, to monitor for signs of heart failure or hydrops.
- Color Doppler flow across the valve as well as pulse Doppler will allow an estimate of the degree of regurgitation or stenosis present.

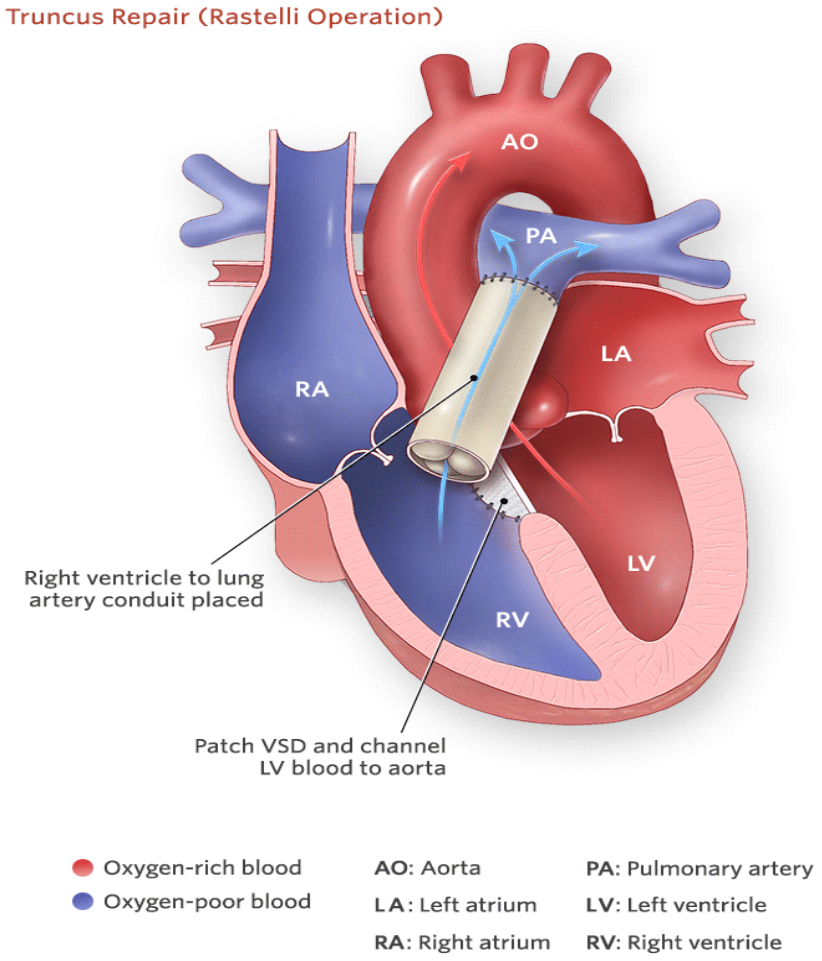
Imaging essentials of TA

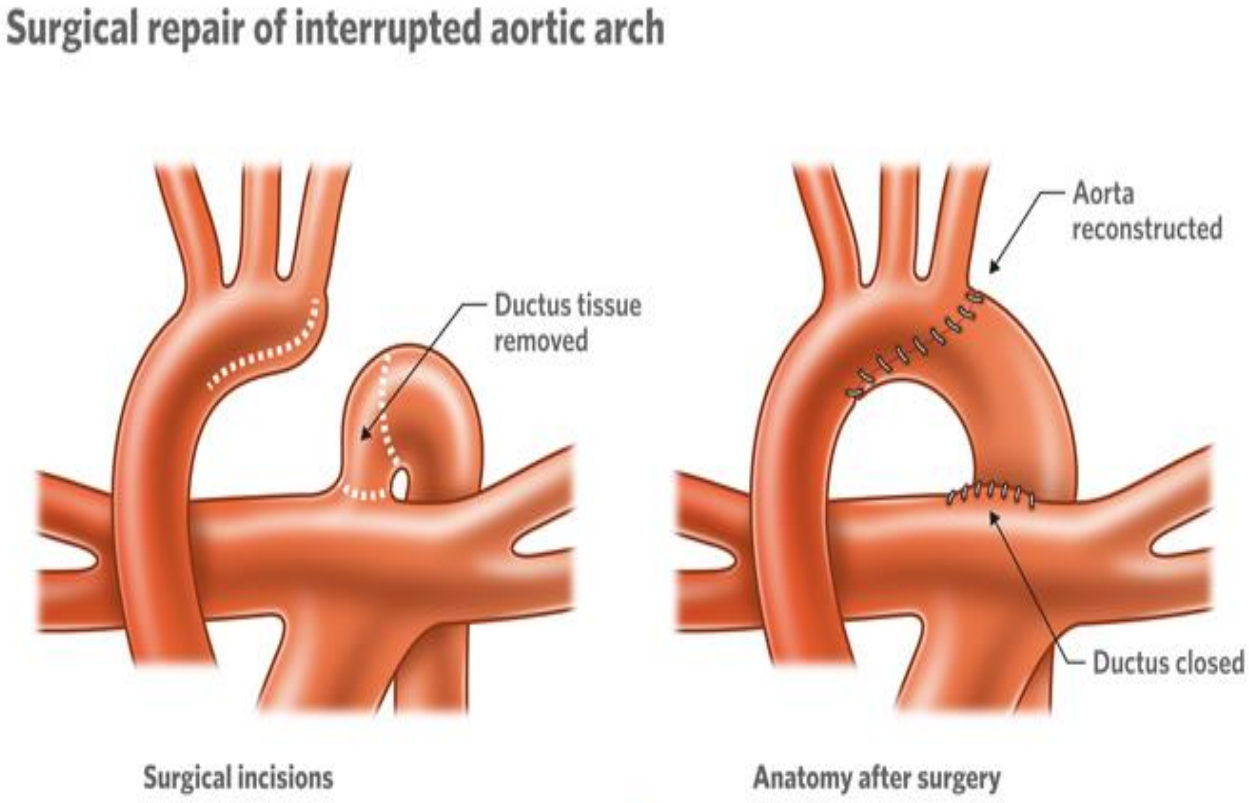
- Identify a large VSD and a single large great artery arising from the heart, positioned over the ventricular septum.
- A true TA gives rise to the aortic arch, at least one pulmonary artery, and at least one coronary artery (the coronary will likely not be visualized in the fetus).
- At least one pulmonary artery arises proximal on the common arterial trunk in TA—if the first pulmonary artery does not originate until one sees the aortic arch or descending aorta, the diagnosis is likely tetralogy of Fallot with pulmonary atresia and not TA.
- A ductus arteriosus will only be seen in a Van Praagh type AIV TA or in a type AIII TA if the ductus arteriosus gives rise to the second pulmonary artery.
- Evaluate the appearance of the truncal valve and assess the truncal valve for insufficiency and stenosis.
- Assess aortic arch sidedness.
- In the setting of significant truncal valve stenosis or insufficiency, evaluate the fetus for signs of ventricular dysfunction and hydrops.
- In type AIV TA, the interruption usually occurs after the left carotid artery.
- Assess for thymic tissue, because its absence may suggest the presence of DiGeorge's syndrome (22q11 deletion).

Counselling

- Needs confirmation post natally
- Requires repair of Truncus as neonate
- If CHF or pulmonary over circulation symptoms develop, may require an intermediate procedure of PA banding before complete repair
- Genetic consultation due to associated extracardiac anomalies and association with Di George
- May need multiple interventions as child or young adult

Repair





Pulmonary artery banding may be needed first if there is pulmonary over circulation before complete repair.

Outcomes

- Overall, surgical outcomes for TA have improved significantly
  - Several centers report early surgical mortality after neonatal repair as 3% to 5% and survival at 6 years as high as 93%.
  - Patients with significant truncal valve disease or an interrupted aortic arch are at higher risk for mortality and complications.
    - A report from the Congenital Heart Surgeons Society 16 pooled results from 33 institutions that performed surgical repair on patients with TA type AIV between 1987 and 1997. **Hospital survival was only 44%** in this group of 50 patients. However, in another single center experience of 16 patients who underwent repair for TA Van Praagh type AIV, **12.5% mortality** was found at a median follow-up of 18 years.

- Patients with TA are at risk for number of morbidities and the need for multiple re-interventions.
  - Almost 50% of neonates will need the right ventricle–to–pulmonary artery conduit replaced by 5 years and 70% by 10 years.
  - In one recent study of TA repair between 1986 and 2003, the need for truncal valve (neo-aortic) re-intervention was nearly 20% at 5 years.
  - There is a correlation between significant truncal valve insufficiency at neonatal presentation and the need for early re-intervention.
  - Finally, the presence of a right ventricle–to–pulmonary conduit places the patient at similar risks as described in the tetralogy of Fallot population, including chronic pulmonary insufficiency, right ventricular dilation and failure, exercise intolerance, and arrhythmias.

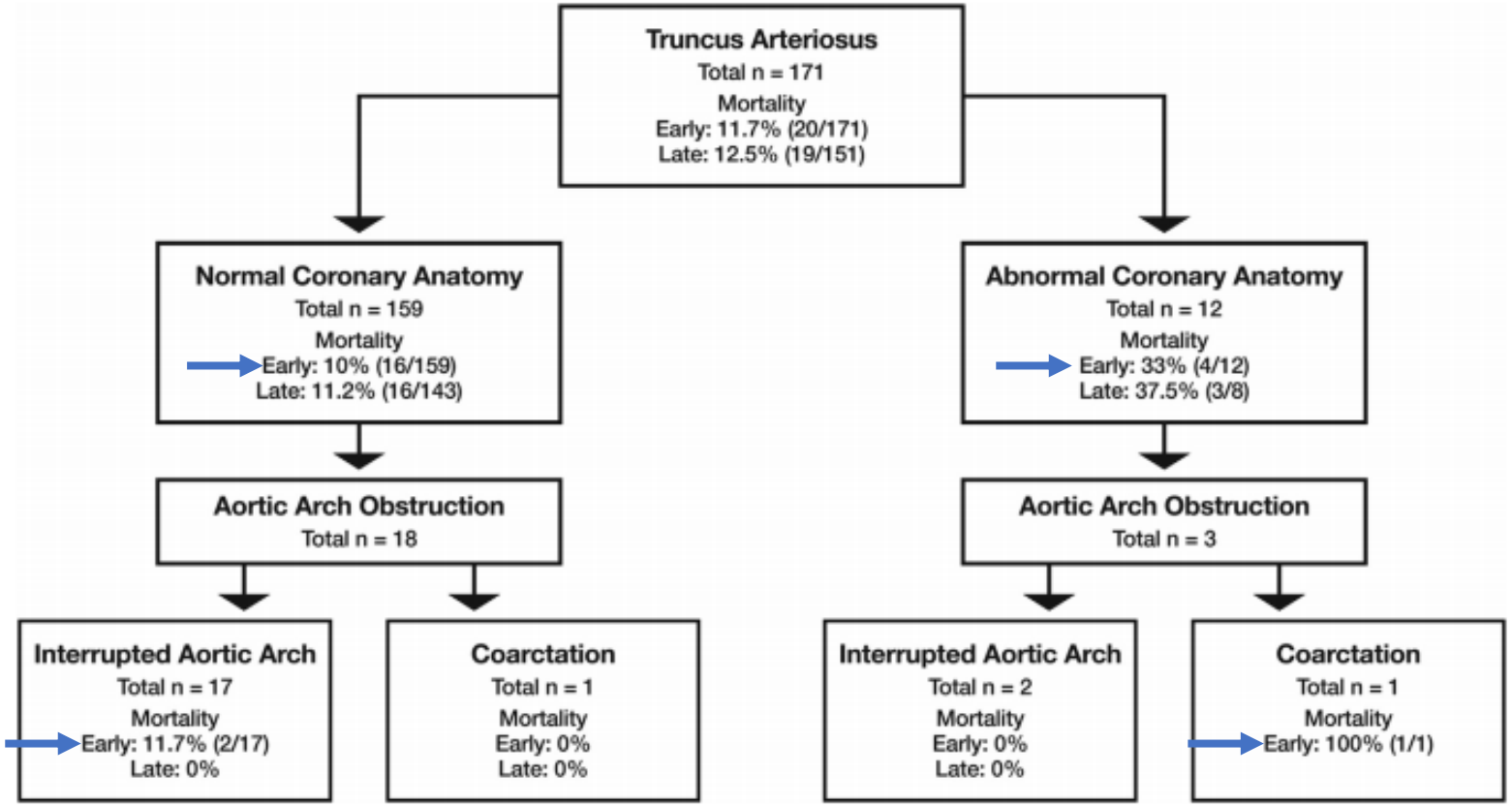


Figure 1. Outcomes of patients with and without coronary artery anomalies and aortic arch obstruction.

Outcomes of Truncus Arteriosus Repair in Children: 35 Years of Experience From a Single Institution

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Seminars in Thoracic and Cardiovascular Surgery, 2016

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Thank you