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Total Anomalous Pulmonary Venous Return: Challenges in fetal diagnosis

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Suma Goudar, M.D.
Cardiology

Disclosures

- None

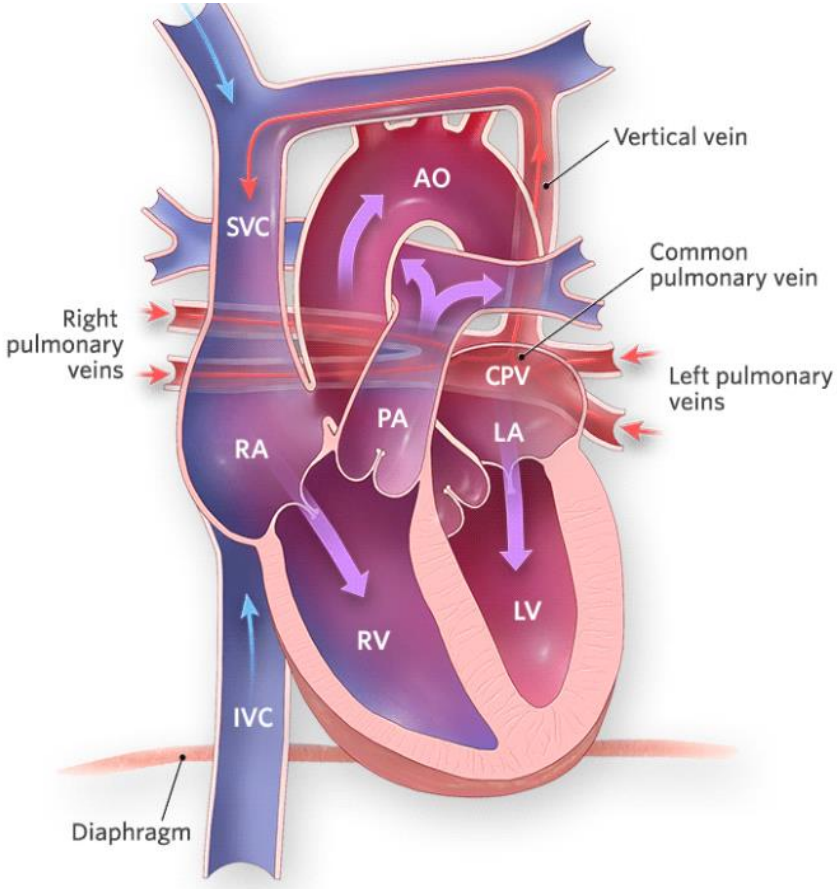
Learning Objectives

- Describe total anomalous pulmonary venous return, treatment, and outcomes
- How to identify TAPVR on fetal echo
- Risk stratification and delivery planning for TAPVR

Total anomalous Pulmonary venous return

- All four pulmonary veins do not connect to the left atrium
- Typically drain into the right atrium via an abnormal venous channel
- Requires adequate ASD for intracardiac mixing
- 3 types:
    - Supracardiac
    - Infracardiac
    - Intracardiac

Supracardiac total anomalous pulmonary venous return



- Pulm veins drain to superior caval/innominate vein via a vertical vein
- Vertical vein can be obstructed (narrowed or compressed by airway)
- Dilated caval veins and right atrium
- Most common (43%)

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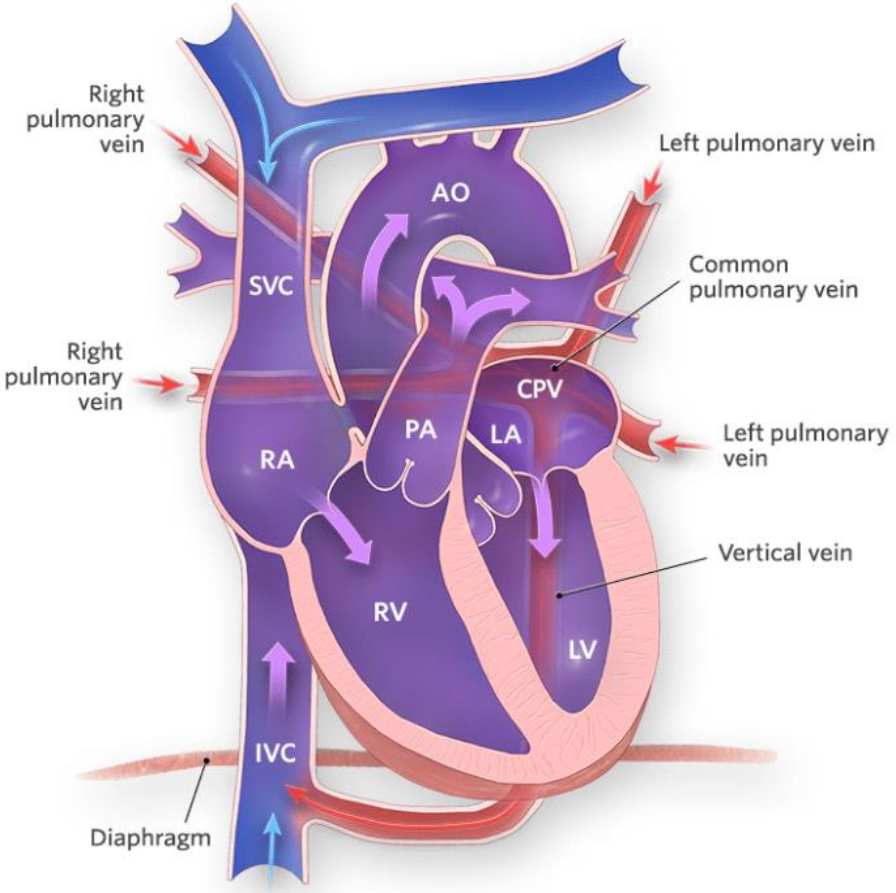
Infradiaphragmatic total anomalous pulmonary venous return



- **Pulm veins drain below the diaphragm to join the IVC, portal vein, ductus venosus**
- **Vertical vein courses anterior to aorta**
- **commonly obstructed**

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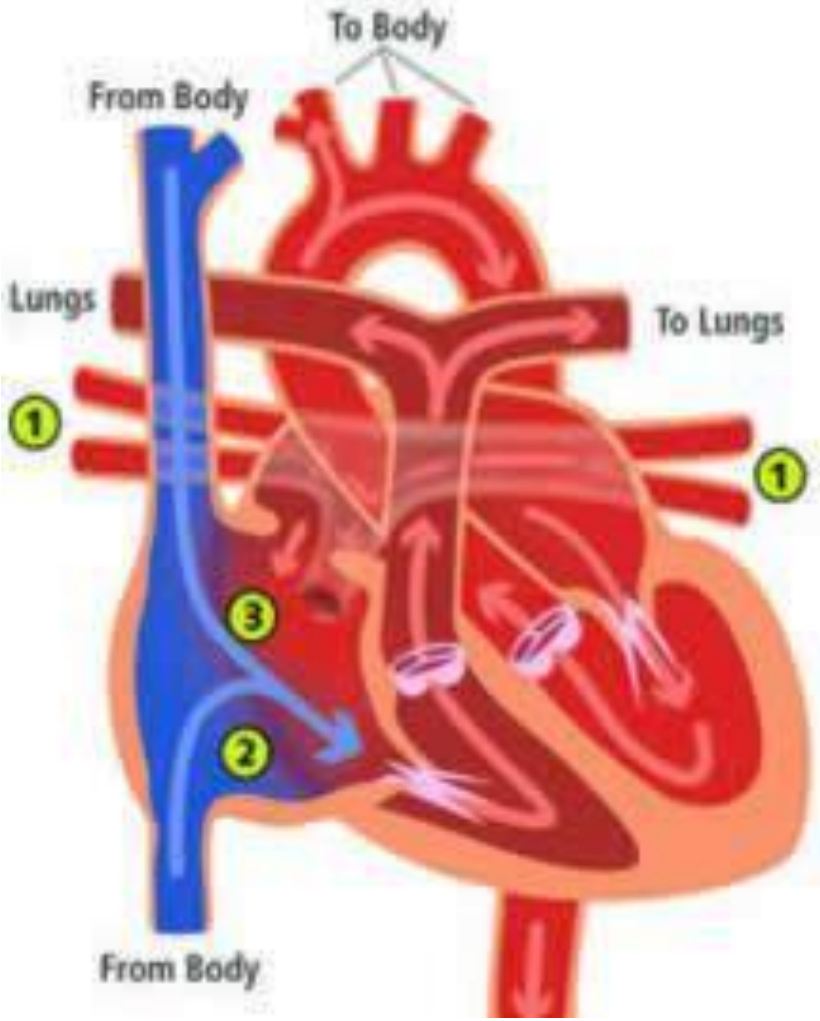
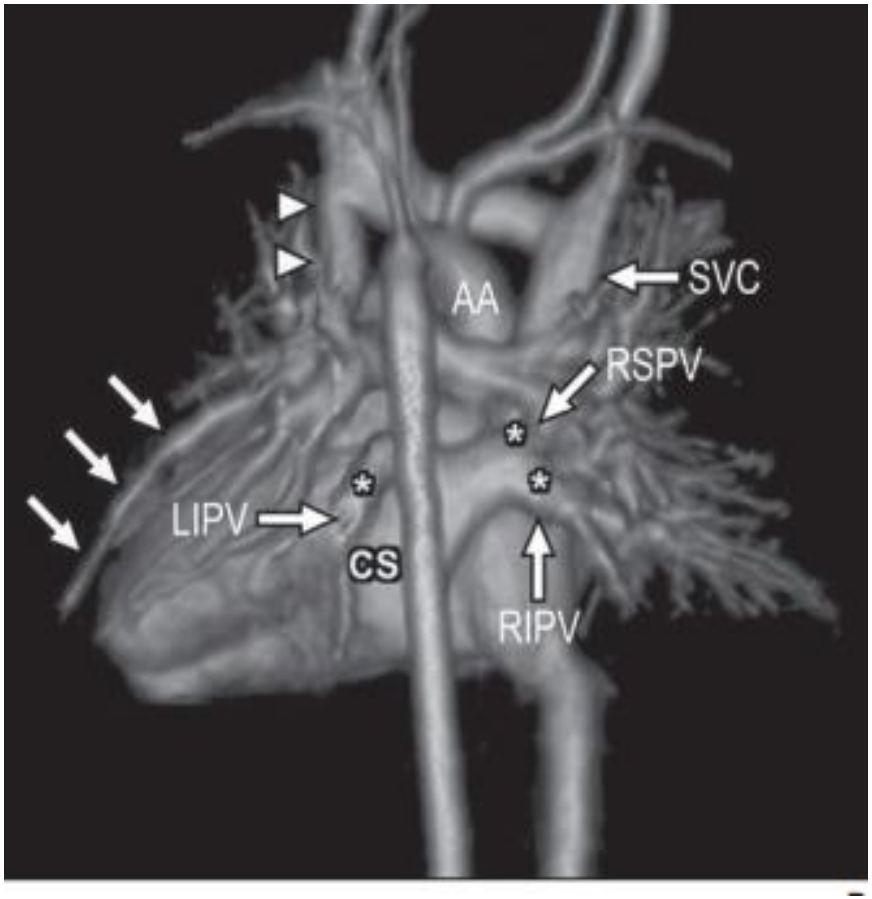
Intracardiac total anomalous pulmonary venous return



- Pulm veins drain to right atrium, usually via coronary sinus
- Mixing of pulmonary venous blood and systemic venous blood in right atrium
- More subacute course of respiratory symptoms
- Difficult to diagnose in the fetus

Legend
1- Pulmonary veins
2-right atrium
3 –mouth of coronary sinus

Mixed type anomomalous pulmonary venous return



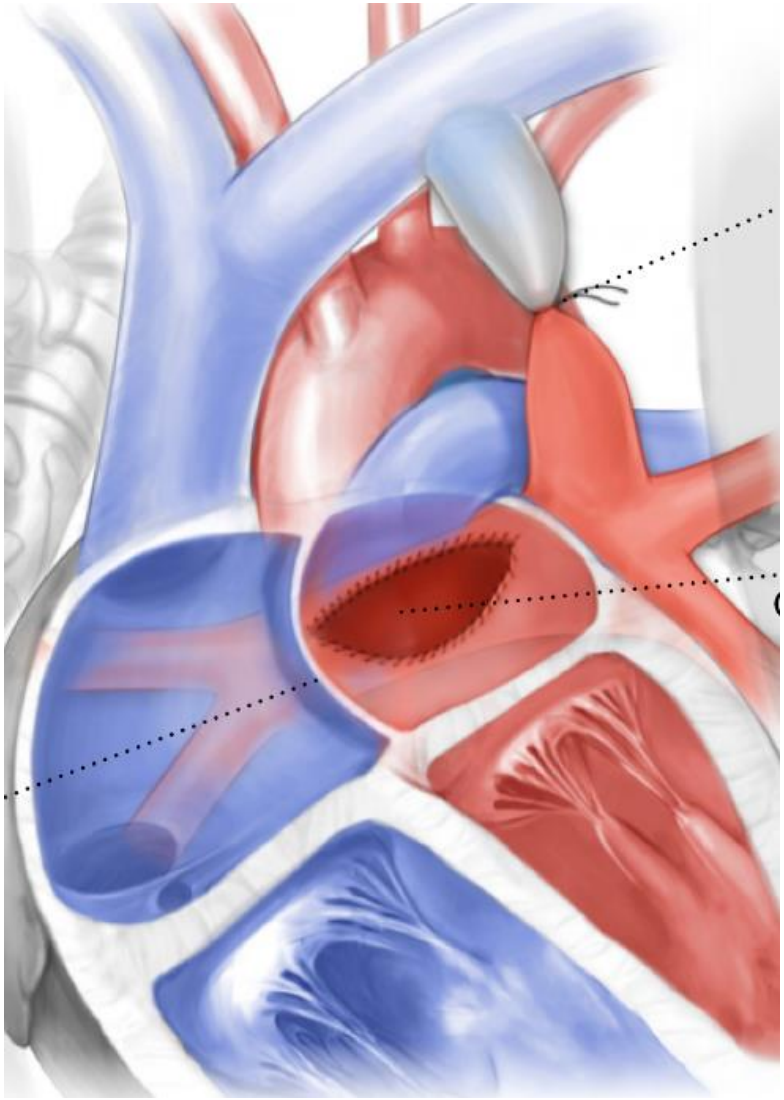
- Pulm veins drain via a combo of above
- This example shows left upper veins to innominate vein, remaining pulm veins to coronary sinus

Clinical presentation

- Cyanosis (mixing cardiac lesion)
    - More severe if obstructed
- Respiratory distress
    - Pulmonary overcirculation
    - Obstructed veins
- CXR: pulm venous congestion, prominent lung interstitum, pleural effusion

Treatment

- Surgery
  - Ligate vertical vein, direct anastomosis of confluence to left atrium
  - ASD closure
  - Obstructed=urgent
- Cath
  - Palliative (premature infants)
  - Stent in vertical vein



Prognosis

- **87% survival at 1 year[1]**
- **Risk factors for mortality:**
    - **Obstructed**
    - **Pulmonary hypertension**
    - **Heterotaxy (30-40% mortality)**
    - **Single ventricle (54% survival)[2]**
        - **Lower if obstructed**
        - **HLHS variant**

Schulz A et al. “Outcomes of TAPVD repair in neonates”. JTCVS 2022
Geoffrion T et al. “Contemporary outcomes for functional single ventricle With TAPVC”. JTCVS 2023

Incidence and prenatal detection

- **1 in 7809 or ~ 500 babies per year[1]**
- **5th most common cause of critical heart disease[2]**
- **May require urgent intervention if obstructed**
- **Low detection rates in the fetus (2-12%)[2, 3, 4]**

1 Mai CT et al. National population-based estimates for major birth defects, 2010-2014. Birth Defects Res 2019

2 Allan LD et al. The echo diagnosis of TAPVC in fetus Heart 2001

3 Seale AN et al. Total anomalous pulmonary venous connection. Ultrasound Obstet Gynecol. 2012

4 Domadia S et al. Neonatal outcomes in TAPVR". Pediatr Cardiol .2018

Prenatal evaluation of pulmonary veins

- **Identify at least one pulmonary vein each from the right and left side via color**

- **Pulse Doppler of each pulmonary vein**

Normal: example

Red flags for abnormal PV drainage

- Small left atrium
- Smooth posterior left atrial wall
- Dilated caval veins
- Dilated coronary sinus
- Abnormal pulmonary venous Doppler

Example: Abnormal case #1

Example: Abnormal case # 2

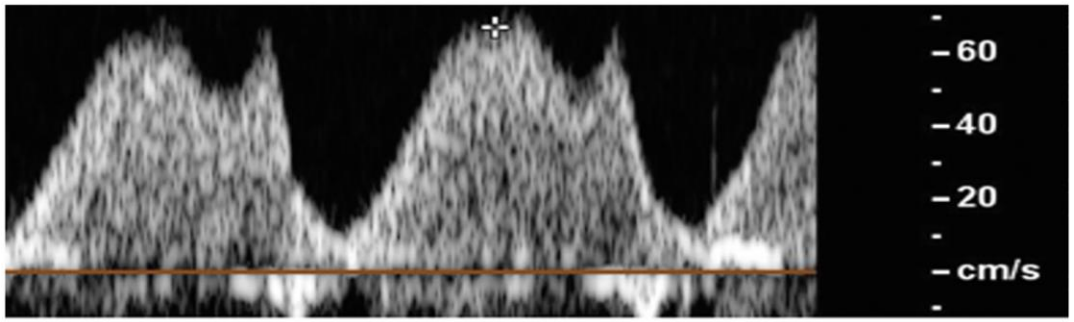
Example: Abnormal case #3

Risk stratification and delivery planning

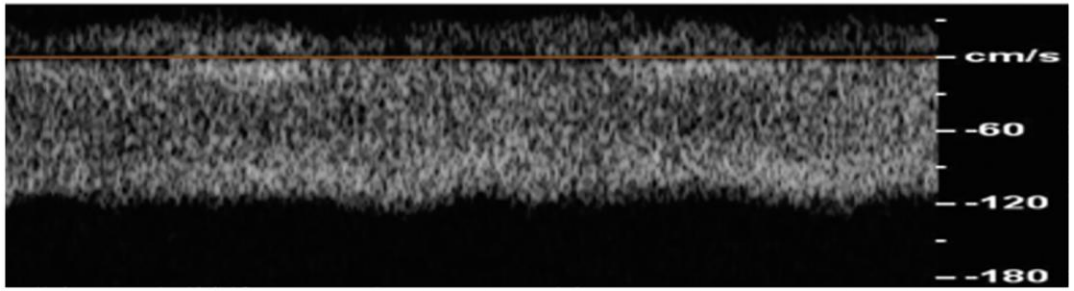
Level of Care	definition	Example CHD	Prenatal planning	Delivery	DR recs
1	CHD without physiologic instability in the 2 weeks	1. Shunt (VSD/ASD) 2. Intracardiac TAPVR	Outpatient in 2 weeks of life	SVD	routine
2	CHD stable at delivery but needs intervention prior to discharge	1. ductal-dependant (HLHS) 2. Truncus arteriosus 3. Unobstructed supracardiac TAPVR	Create plan of care, transport to main hospital	SVD	Neo in DR, umbilical lines, +PGE in some cases
3	CHD with instability at birth and requires specialty care prior to intervention	1. Transposition of great arteries 2. TAPVR (supra/infracardiac	Plan of care in DR Interventional/surg team on standby	Planned induction, bailout c/s for care coordination	Neo and cards in DR, rapid transport
4	CHD with instability requiring urgent intervention/surgery	1. HLHS with intact atrial septum 2. Severe Ebstein's anomaly 3. Obstructed TAPVR	Delivery at Childrens Hospital with specialized team in delivery room	Planned c/s	Specialized teams in DR, rapid transport to OR/cath lab

Risk stratification: fetus with TAPVR

- Based on risk of postnatal obstruction
- Red flags:
  - Vertical vein peak Doppler velocity > 0.74 m/s mmHg[1]
  - Reduced pulsatility of flow in the vertical vein[2]



unobstructed

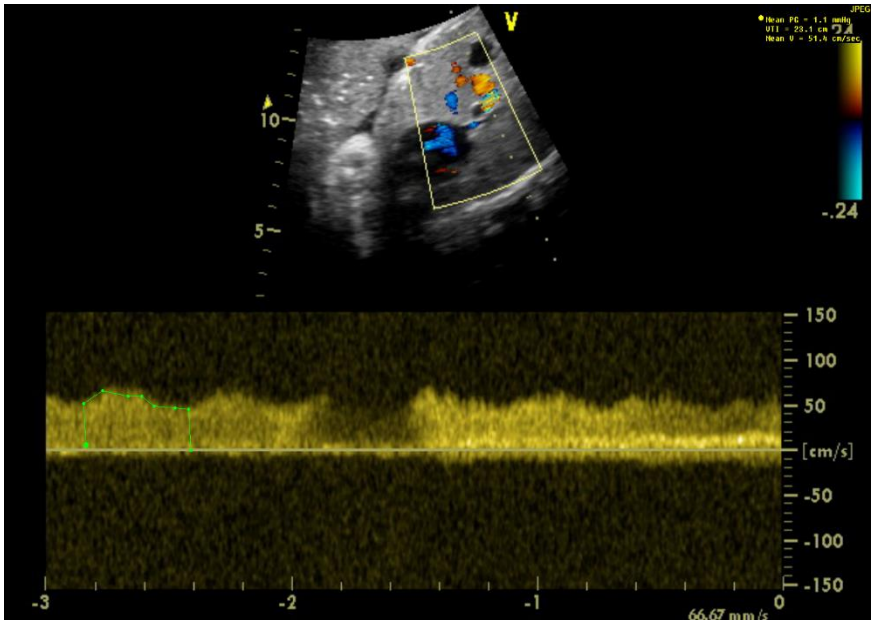


obstructed

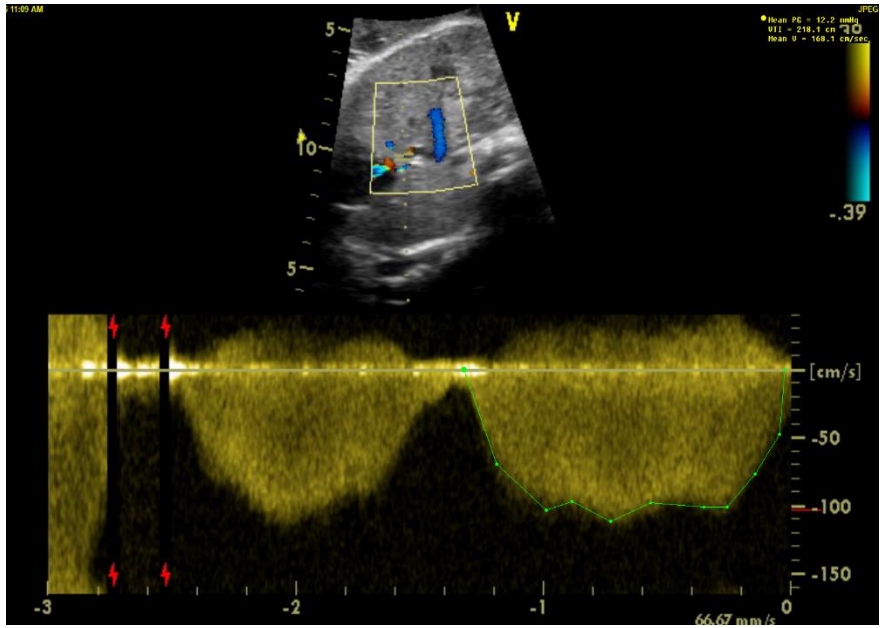
1 Domadia et al. “Neonatal outcomes in TAPVR” Pediatr Cardiol 2018
2Campbell et al” Fetal Doppler Echo assessment predicts severe postnatal obstruction in TAPVR. JASE 2022

Risk stratification: role of hyperoxia

- Obstruction may be underappreciated since pulmonary blood flow is low in the fetus
- Administer oxygen to mother for 10-15 minutes
- Pulmonary blood flow increases with oxygen- unveil gradient
- May be useful in predicting postnatal course in case series[1]



Baseline



Post-hyperoxia

Schidlow D, Donofrio MT. "Prenatal maternal hyperoxygenation testing and implications…" Am J

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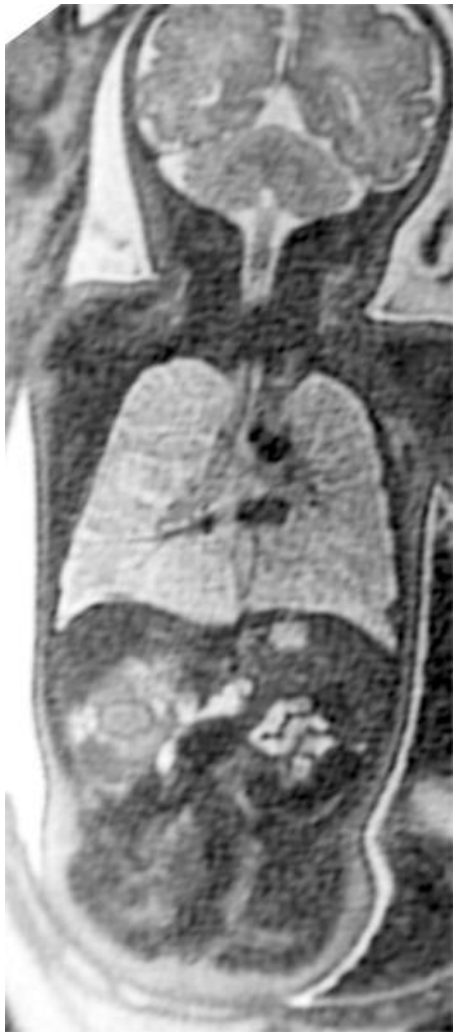
Risk stratification: role of fetal MRI

- Fetal MRI can evaluate lung parenchyma
- Aids risk stratification and prognosis[1,2]
- Typically performed in late gestation (last prenatal visit)
- Obstructed pulmonary veins can lead to lung congestion and lymphangiectasia= worse prognosis and more instability



1 Ryd D et al. Pediatrics 2021
2 Barrera C et al. Pediatr Radiol. 2021

Summary

- TAPVR is critical cyanotic heart defect that frequently requires urgent intervention and good long-term outcomes
- Infrequently detected in-utero but greatly improves morbidity and mortality when it is
- Evaluation of pulmonary veins is critical in performance of fetal echo
- Dilated caval veins/coronary sinus, abnormal venous structures in 3VV and sagittal views and abnormal pulmonary venous Dopplers are red flags and warrant referral

Thank You!