

A rare variant of congenital heart disease: Infradiaphragmatic Total Anomalous Pulmonary Venous Return

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Section: Cardiovascular

Area of Interest: Cardiac Cardiovascular system

Paediatric Vascular

Procedure: Diagnostic procedure

Procedure: Comparative studies

Imaging Technique: MR

Imaging Technique: MR-Angiography

Special Focus: Congenital Case Type: Anatomy and Functional Imaging

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Patient: 2 days, male

Clinical History:

A 2-day-old full term African American male patient was referred for further evaluation of a prominent grade IV systolic murmur observed immediately following delivery. The infant arrived in respiratory distress on high flow oxygen. Respiratory support was escalated from CPAP to intubation with prostaglandin administration for congenital cardiac abnormality concerns.

Imaging Findings:

Pre-operative MRA was performed using steady-state free precession localisers in axial, coronal, and sagittal projections under general anaesthesia. Images demonstrated a Type III infradiaphragmatic TAPVR with all four pulmonary veins draining into the vertical collecting vein draining into the hepatic vein through ductus venosus. Kinking of the ductus venosus, causing turbulent flow with pre-stenotic dilation of the vertical vein was also observed. Severe tricuspid regurgitation was present as well as an enlarged central pulmonary artery due to right to left shunt with volume overload.

Discussion:

Background: Infradiaphragmatic total anomalous pulmonary venous return (TAPVR) is one of multiple types of congenital heart defects that results in sub-optimal oxygen rich blood return to the left atrium. From an embryological perspective, TAPVR results from the failure of the left atrium to link with the primitive pulmonary venous plexus, maintaining the early umbilicovitelline and cardinal venous systems. This lack of connection can result in multiple variants which are classified based on site of the abnormal pulmonary venous drainage. In our patient, all four pulmonary veins formed an abnormal connection below the heart as a consequence of continued pulmonary vein linkage to the umbilicovitelline venous system. These veins joined to form a common vertical collecting vein draining into the hepatic vein via the ductus venosus [1].

Clinical Perspective: Acute management of congenital cardiac defects is vital for newborn survival. TAPVR is one of

the cyanotic newborn syndromes in which the patient will present with characteristic cyanosis. Additionally, patients will likely present in respiratory distress which should suggest underlying cardiopulmonary pathology. Prostaglandins should be delivered immediately in situations suggestive of congenital defects.

Imaging Perspective: A complete trans-thoracic echocardiogram and cardiac MRI with gadolinium were obtained, which showed structural malformations consistent with infradiaphragmatic TAPVR with all four pulmonary veins leading into a vertical vein collector draining into the hepatic vein via the ductus venosus. Findings are demonstrated in Figures 1-4.

Outcome: Surgical intervention was recommended and pursued within 2 days of identification. Repair was uncomplicated and the patient remains under routine surveillance to monitor appearance of anastomotic gradient which could complicate the prognosis.

Teaching Points: Infradiaphragmatic TAPVR is the second least common variant resulting in sub-optimal oxygen-rich blood return to the left atrium [2]. It is necessary to maintain a patent ductus arteriosus via prostaglandin administration in these patients to ensure adequate venous return prior to further evaluation. Intraoperative findings confirmed the above detailed imaging findings. Surgical repair of the defect was uncomplicated and recent literature reports a 5-year post-operative survival rate of 92.6% without a gradient across the anastomosis [3].

Differential Diagnosis List: Total Anomalous Pulmonary Venous Return, infradiaphragmatic type, Congenital heart disease, Cardiac valvular pathology, Infant respiratory distress syndrome

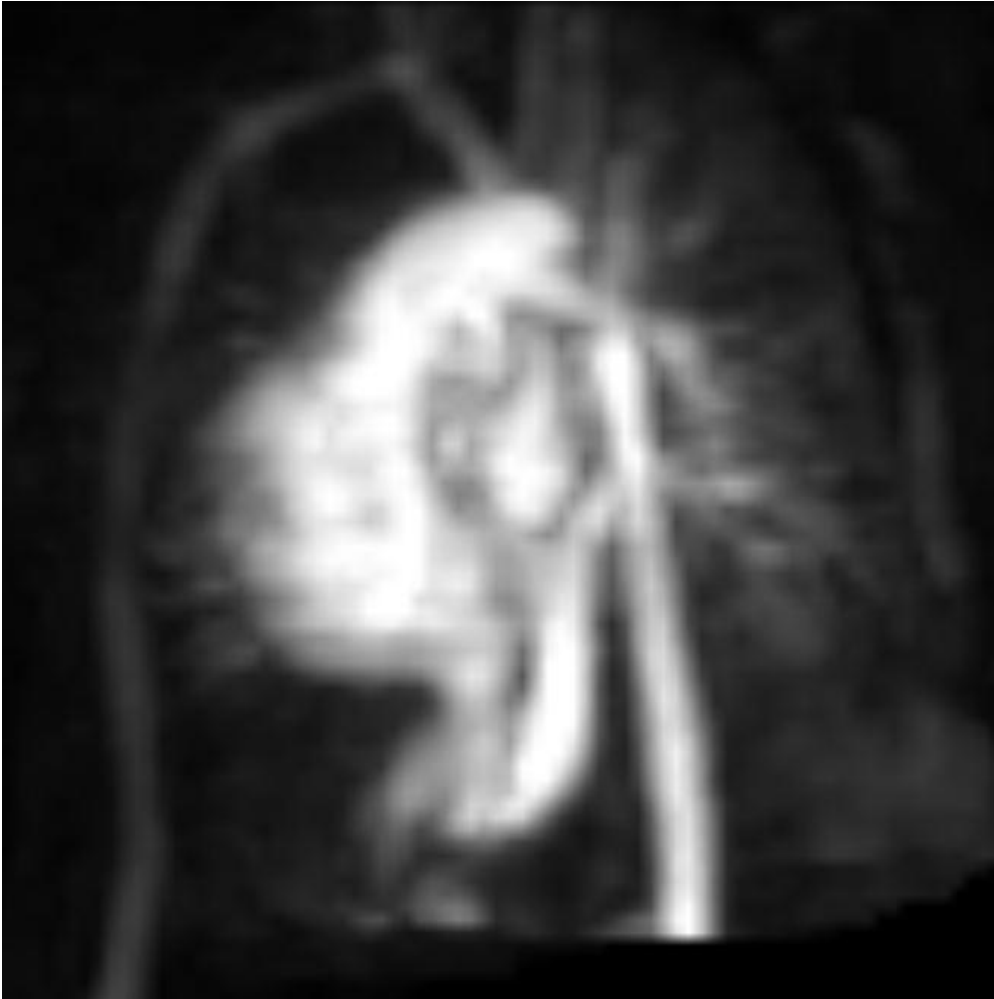
Final Diagnosis: Total Anomalous Pulmonary Venous Return, infradiaphragmatic type

References:

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- Douglas YL, Jongbloed MR, den Hartog WC, Bartelings MM, Bogers AJ, Ebels T (2009) Pulmonary vein and arterial wall pathology in human total anomalous pulmonary venous connection. *Int J Cardiol* 134(2):302-12 (PMID: [19117620](#))
- Zhao K, Wang H, Wang Z, Zhu H, Fang M, Zhu X, Song H (2015) Early-and intermediate-term results of surgical correction in 122 patients with total anomalous pulmonary venous connection and biventricular physiology. *J Cardiothoracic Surg* (PMID: [26602895](#))

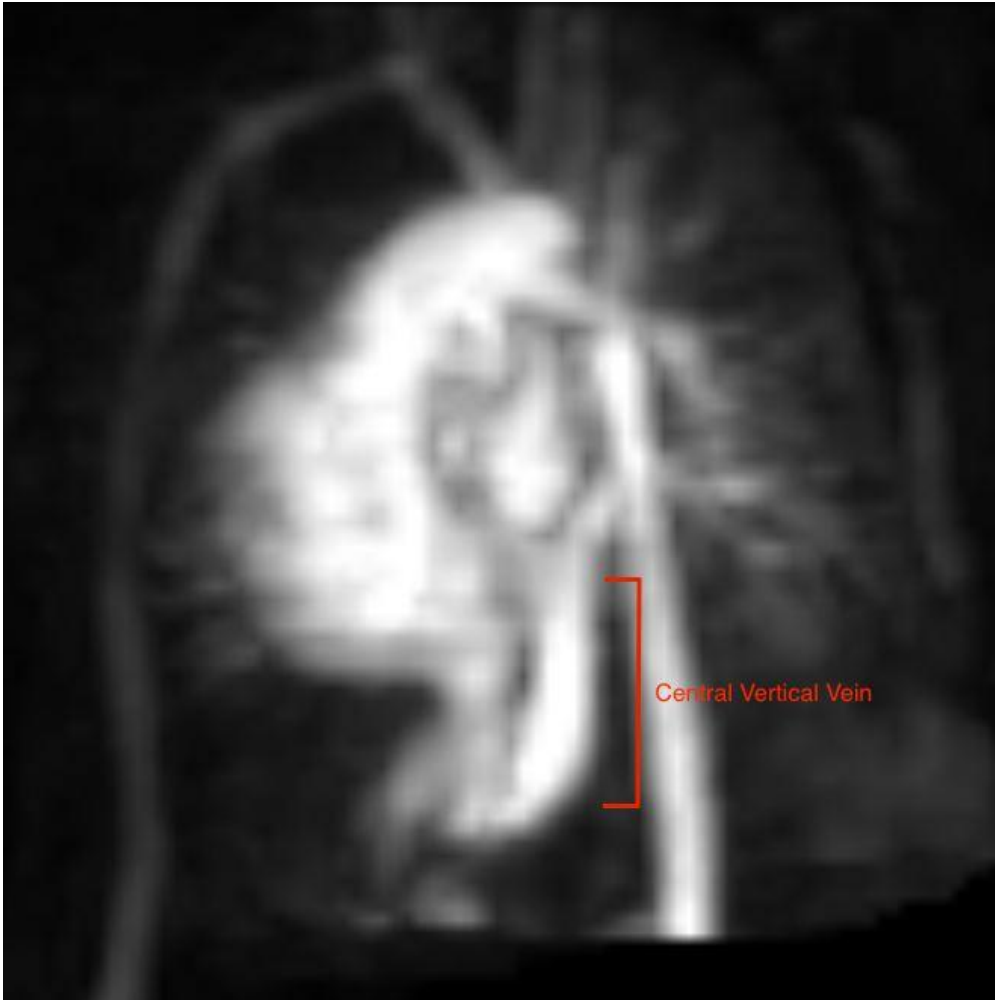
Figure 1

a



Description: Contrast-enhanced MR angiogram demonstrates a vertical collecting vein draining into a hepatic vein below the diaphragm. **Origin:** Keshavamurthy J, Department of Radiology, Medical College of Georgia, Augusta, Ga

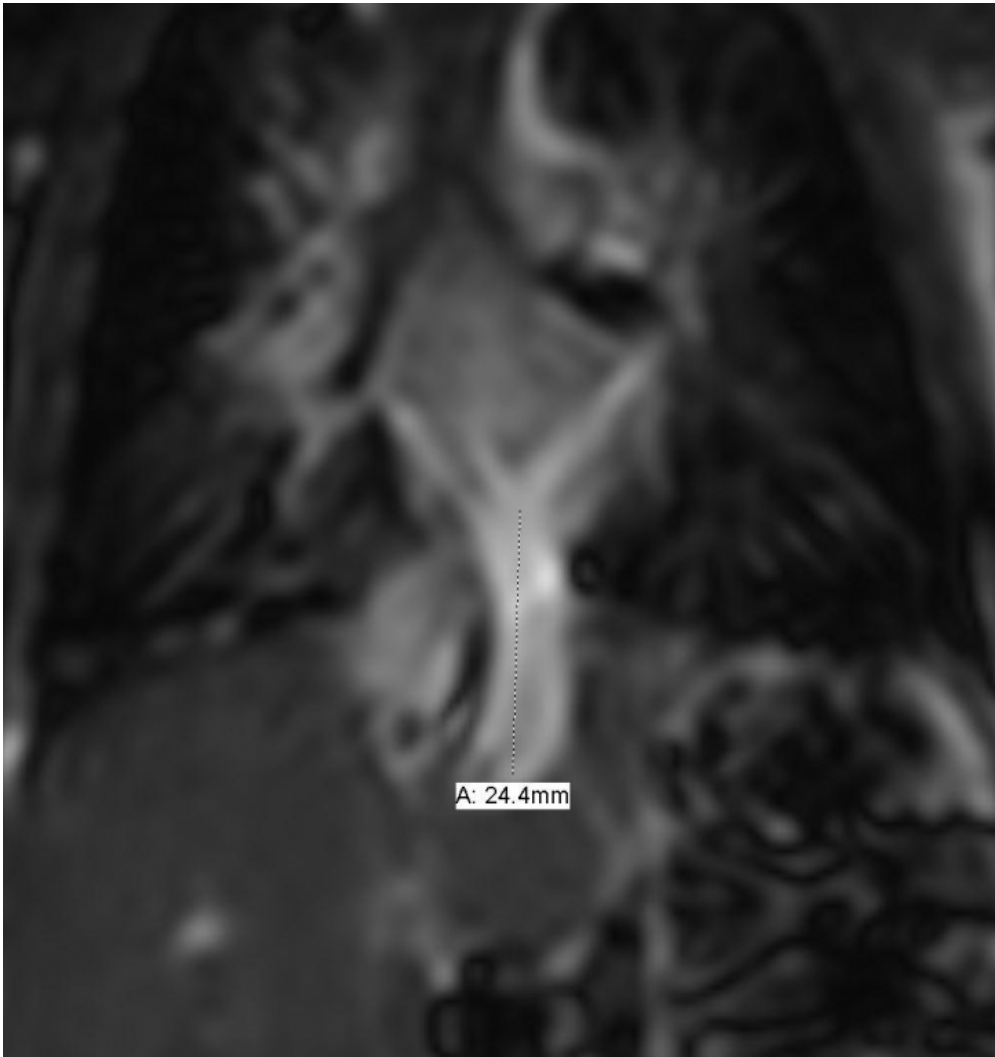
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Description: Contrast-enhanced MR angiogram demonstrates a vertical collecting vein draining into a hepatic vein below the diaphragm. **Origin:** Keshavamurthy J, Department of Radiology, Medical College of Georgia, Augusta, Ga, USA

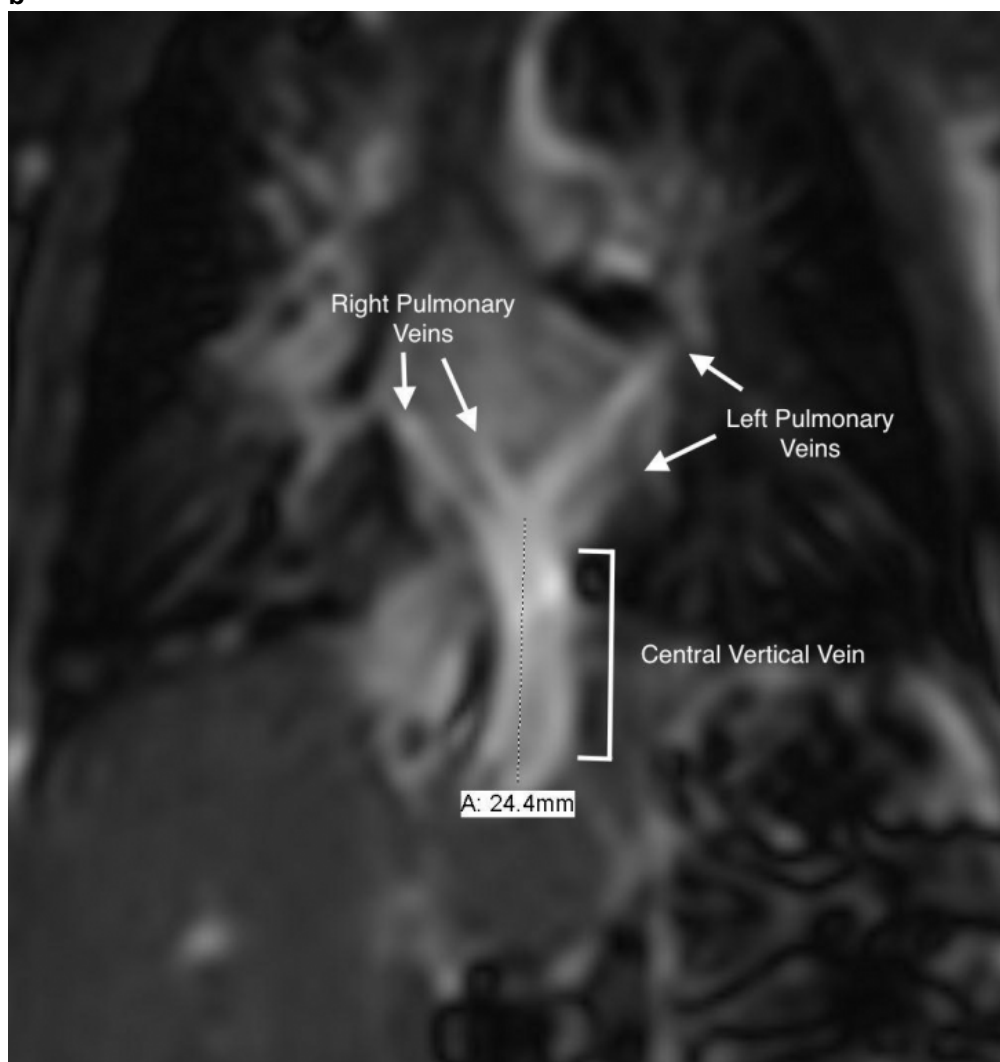
Figure 2

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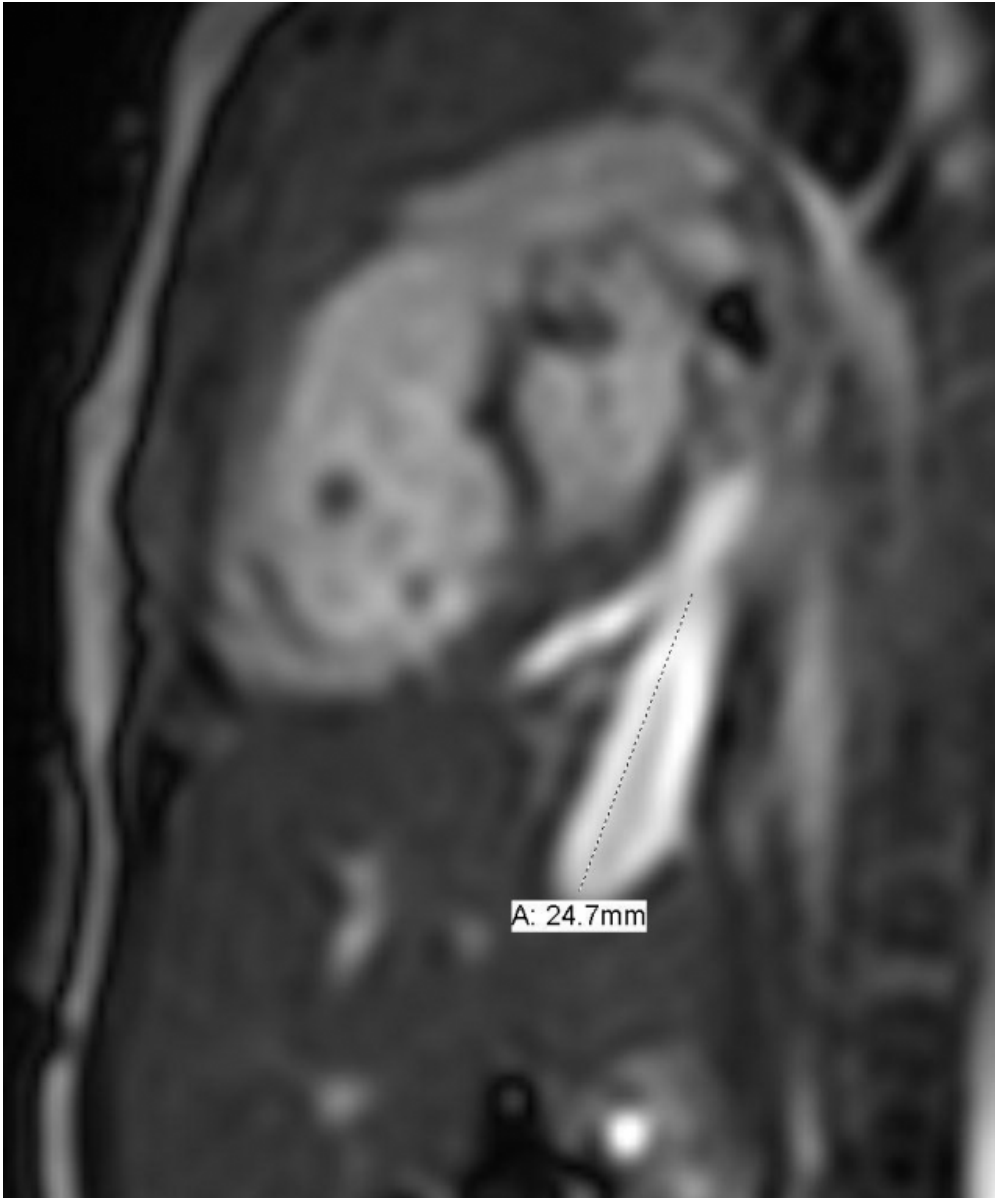
Description: Coronal steady-state free precession MRI demonstrates all four pulmonary veins draining into a vertical collecting vein extending below the diaphragm. **Origin:** Keshavamurthy J, Department of Radiology, Medical College of Georgia, Augusta, Ga

b



Description: Coronal steady-state free precession MRI demonstrates all four pulmonary veins draining into a vertical collecting vein extending below the diaphragm. **Origin:** Keshavamurthy, J, Department of Radiology, Medical College of Georgia, Augusta, Ga, USA

c



Description: Coronal steady-state free precession MRI demonstrates all four pulmonary veins draining into a vertical collecting vein extending below the diaphragm. **Origin:** Keshavamurthy J, Department of Radiology, Medical College of Georgia, Augusta, Ga, USA

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Description: Coronal steady-state free precession MRI demonstrates all four pulmonary veins draining into a vertical collecting vein extending below the diaphragm. **Origin:** Keshavamurthy J, Department of Radiology, Medical College of Georgia, Augusta, Ga, USA

Figure 3

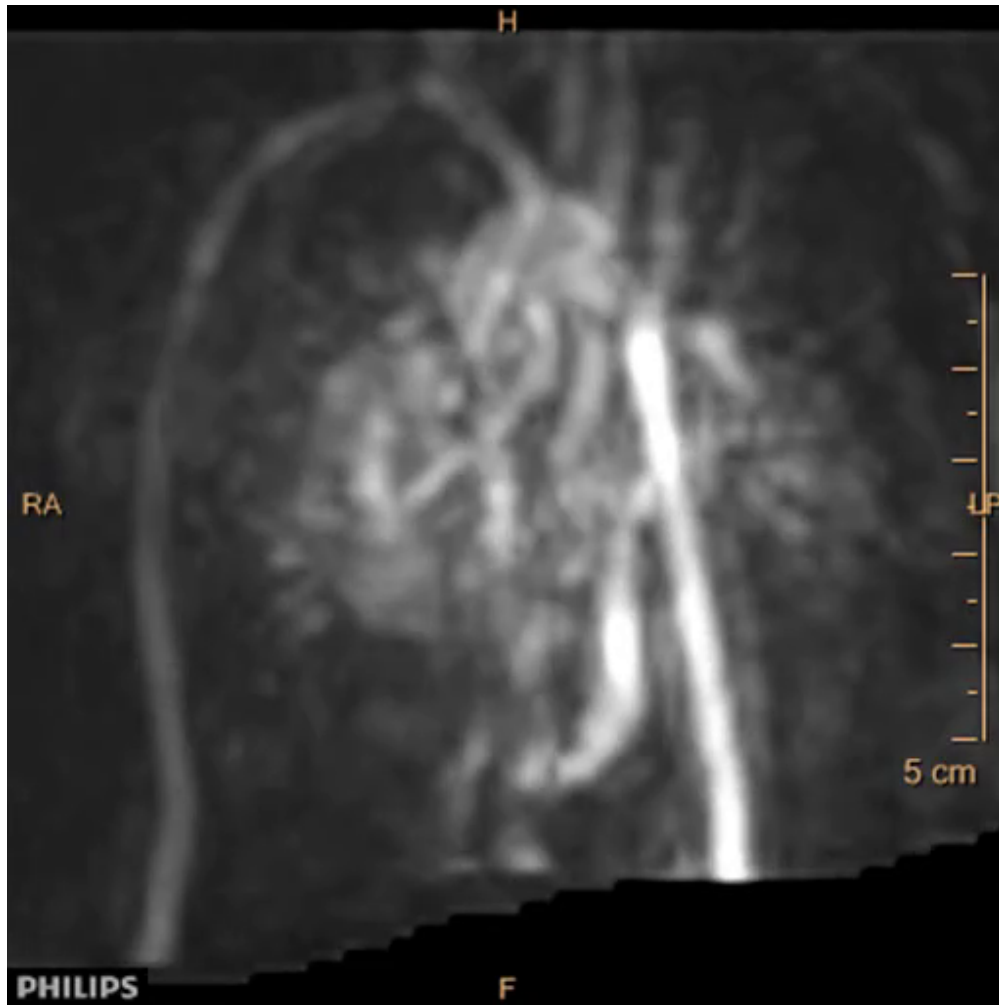
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Description: Chest/abdominal X-ray demonstrates increased pulmonary interstitial thickening and vascular pulmonary plethora without focal consolidation with umbilical venous and arterial catheters in place. Cardiac silhouette enlargement. **Origin:** Keshavamurthy J, Department of Radiology, Medical College of Georgia, Augusta, Georgia, USA

Figure 4

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Description: Contrast-enhanced MR angiogram demonstrates a vertical collecting vein draining into a hepatic vein below the diaphragm. This is a MIP image from the 4D MRA. **Origin:** Keshavamurthy J, Department of Radiology, Medical College of Georgia, Augusta, Ga, USA