Navigating complexity: A pictorial representation of anomalous pulmonary venous connection classification in the pediatric population with volume rendering and multiplanar imaging technique

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Abstract

Anomalous pulmonary venous connections represent a heterogeneous group of congenital heart diseases in which a part or all of the pulmonary veins drains into the right atrium instead of draining into the left atrium. Pulmonary venous anomalies can manifest as partial or total anomalous drainage due to the abnormal embryological development. Multidetector CT angiography, with its multiplanar reformatting and volume rendering techniques, precisely offers the information about the three-dimensional anatomy and spatial relationships of the cardiovascular structures. Clinical features of anomalous pulmonary venous connections may be silent or have variable features like neonatal cyanosis, volume overload, and pulmonary arterial hypertension due to the left-to-right shunt, which are often associated with other congenital cardiac disease, so that accurate diagnosis is essential for treatment planning.

Keywords: *TAPVC, volume rendering images, anomalous pulmonary drainage, scimitar syndrome*

Abbreviations: RSPV - right superior pulmonary vein, **RIPV** - right inferior pulmonary vein, **LSPV** – left superior pulmonary vein, **LIPV** - Left inferior pulmonary vein, **TAPVC** - Total anomalous pulmonary venous connection, **PAPVC** – Partial anomalous pulmonary venous connection

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Methodology

At our institution, the Department of Radiology in Madras Medical College, we performed the scan for patients (45 days–10 years) who were referred from the pediatrics department for CT pulmonary angiogram having symptoms of cyanosis, lower respiratory infections, breathlessness, and failure to thrive.

Multidetector computed tomography angiography (MDCT) examination done using GE 128 SLICE by giving contrast material (iohexol) 1 ml/kg to 1.5 ml/kg with the region of interest and field of view from the inferior margin of the aortic arch up to below the diaphragm. Image acquisition (0.5 mm, section thickness: ≤1 mm) is done with 0.75 mm collimation and reconstructed in axial, sagittal, coronal projections. Retrospective 3D reconstruction of data was performed to obtain a volume-rendered image.

Introduction

Normal pulmonary venous drainage (Fig.1)

The normal arrangement has four separate pulmonary veins: the right and left superior and inferior veins, which drain individually into the left atrium.

The right superior pulmonary vein (RSPV) drains the right upper and middle lobes of the lung while the right lower lobe drained by the right inferior pulmonary vein (RIPV). The left superior pulmonary vein (LSPV) drains the lingula and left upper lobe, while the left lower lobe drains the left inferior pulmonary vein (LIPV) (1).

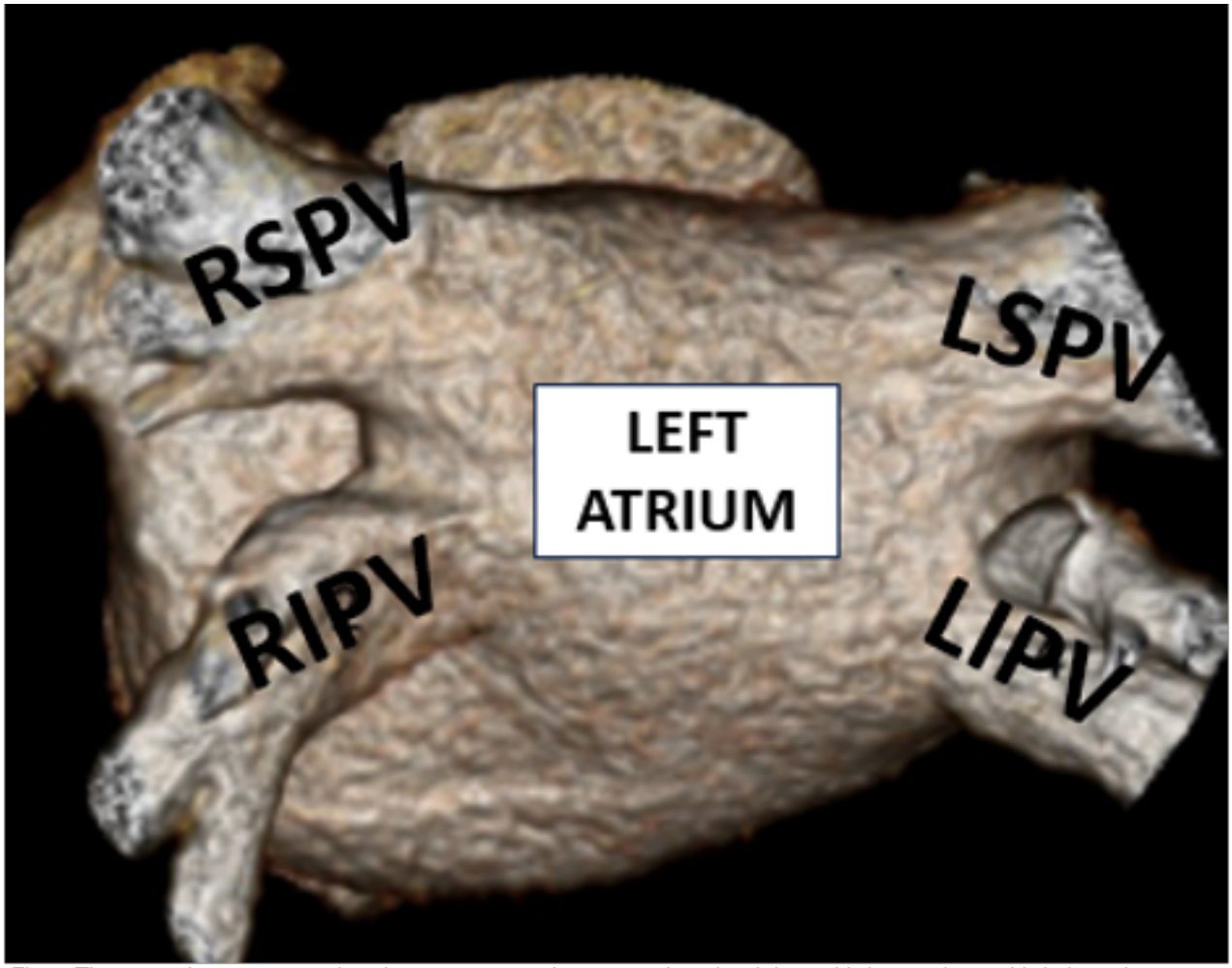


Fig. 1 The normal arrangement has four separate pulmonary veins: the right and left superior and inferior veins, which drain individually into the left atrium.

CASE SERIES

Case 1: A case of supracardiac TAPVC with heterotaxy syndrome - (Fig. 2 a-g)

A 10-year-old female presented with complaints of recurrent respiratory symptoms and a gradual exertional dyspnea. Echo findings show dilated pulmonary arteries and a double SVC. patient was subjected to a CT angiogram (128 slices).

MDCT shows all four pulmonary veins joining together to form a common vertical vein draining into the right SVC, as shown in " figure 2 here". A posterior view of the VR image of the 3D image shows double SVC.

The coronal reformatted image shows the midline liver with asplenia and features of heterotaxy syndrome which is diagnosed as supracardiac TAPVC with heterotaxy syndrome—right-sided isomerism.

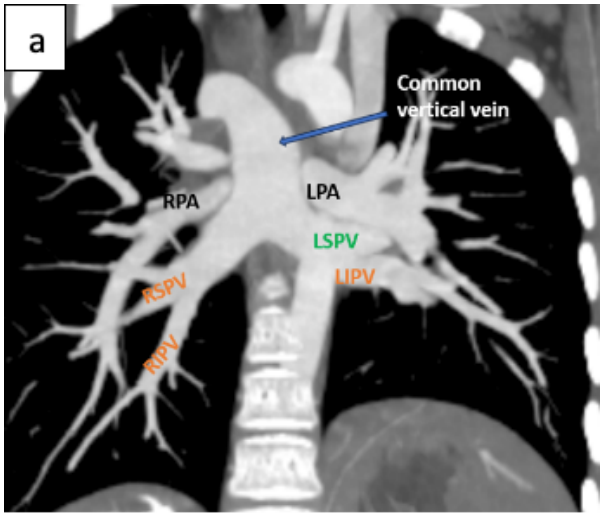


Fig. 2 a

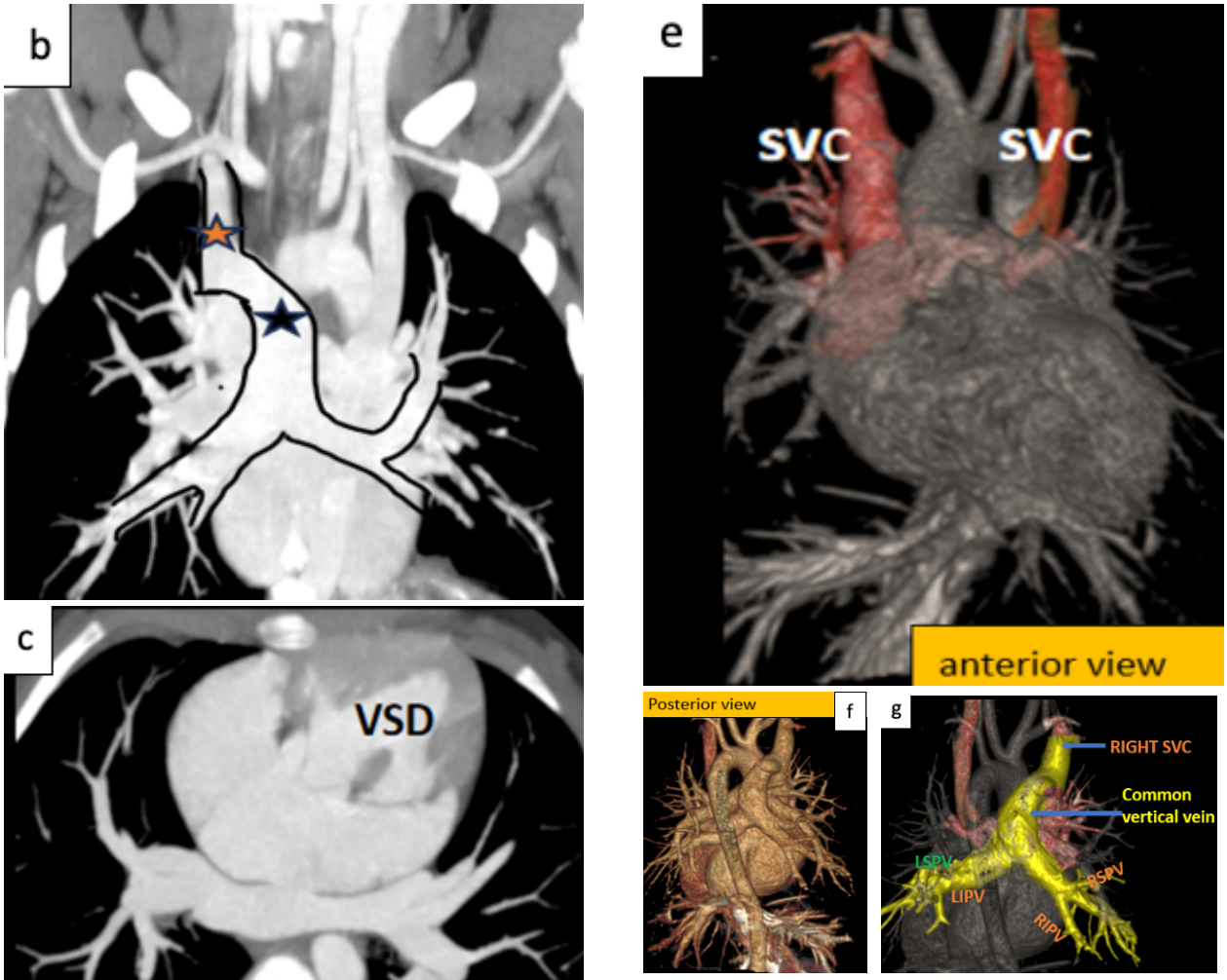


*Fig. 2 (a-g) case of supracardiac TAPVC with heterotaxy syndrome – right sided isomerism a) and b) Coronal reformatted image all 4 pulmonary veins joins together to form a common vertical vein (arrow& *) draining into SVC as shown in fig1b, fig1c) posterior view of the VR image, d) anterior aspect of the 3D image shows double SVC (red), e) coronal reformatted image shows midline liver with asplenia. f) and g) volume rendering image shows common vertical vein draining into right SVC.*

Case 2: A case of cardiac total anomalous pulmonary venous return - (Fig. 3 a-e)

A 45-day-old female child presented for evaluation of cyanosis during feeding and crying. The echo finding shows a dilated coronary sinus. patient was subjected to a CT angiogram 128 slice in which a reformatted coronal and axial image shows all 4 pulmonary veins instead of draining into the left atrium, joining together and draining into the coronary sinus, which then drains into the right atrium as in reflecting the cardiac type of total pulmonary venous anomaly (Fig. 3)

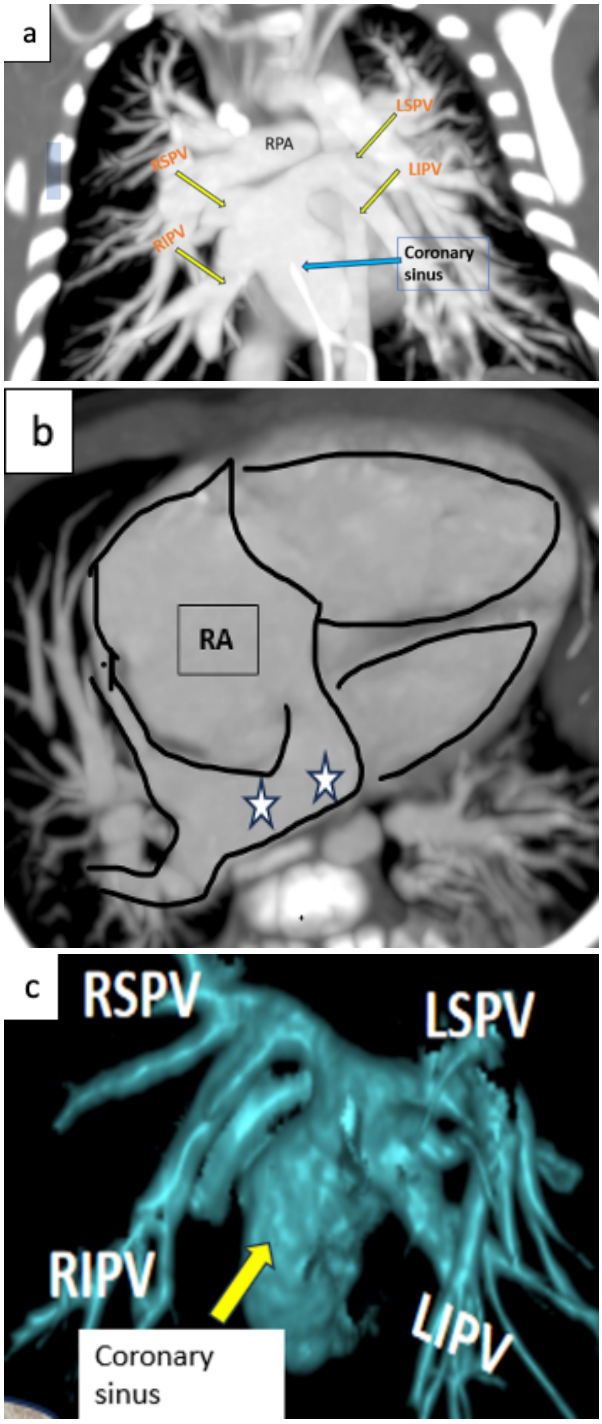


Fig. 3 (a-e)– A case of cardiac TAPVC a) and b) reformatted coronal and axial image shows all 4 pulmonary veins joining together draining into the coronary sinus which then drains into right atrium (annotated) c) shows the 3D reformatted image showing the venous mapping of all 4 pulmonary veins draining into coronary sinus d) 3D image posterior aspect of heart shows TAPVC e) shows the 3D reformatted image of cardiac type of total pulmonary venous anomaly.

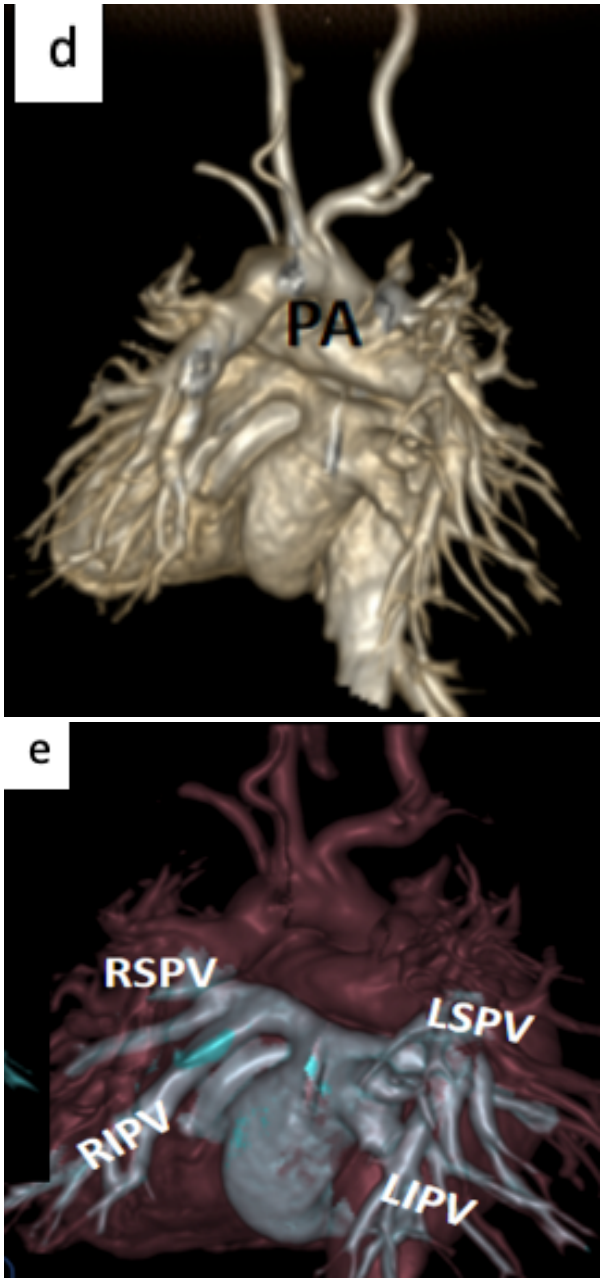
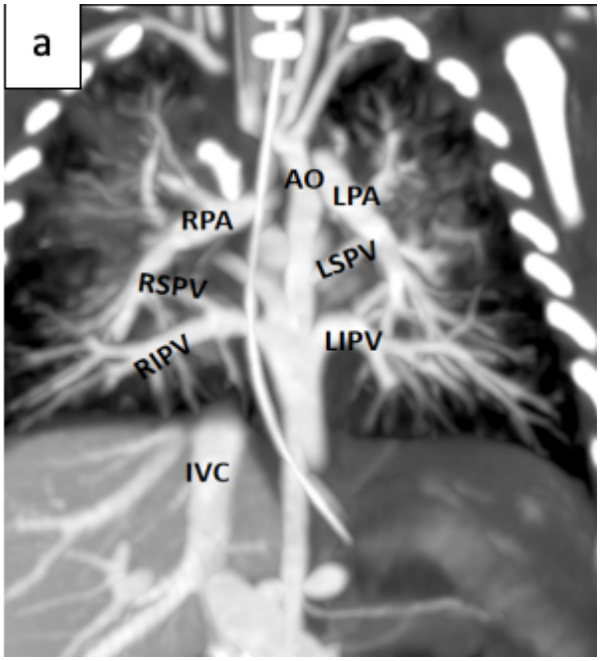


Fig. 3 (d-e)

Case 3: Infra-Cardiac Total Anomalous Pulmonary Venous Connection - (Fig. 4 a-e)

A 3-year-old male child presented as failure to thrive . child was subjected to a CT angiogram. The reconstructed coronal image shows all four pulmonary veins joining together to form the common vertical vein and draining into the portal vein, as shown in Fig 4. 3D reconstructed image showing venous mapping shows partial short segment stenosis in the vertical vein before draining into the portal vein which is an infra-cardiac total pulmonary venous anomaly - obstructed type (Fig.4).



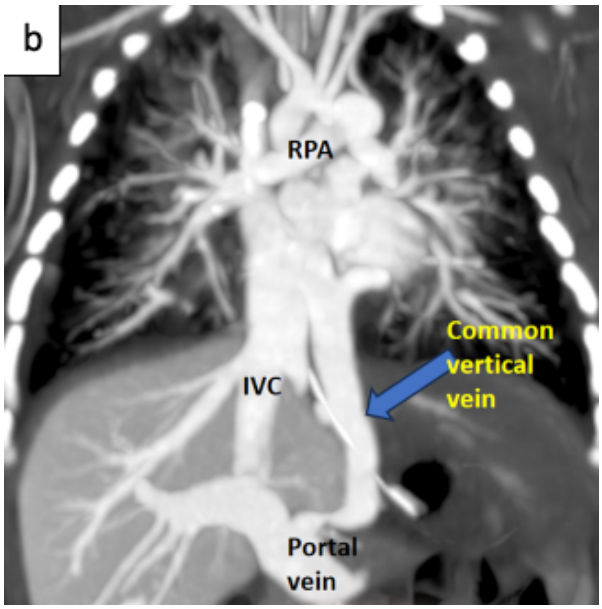
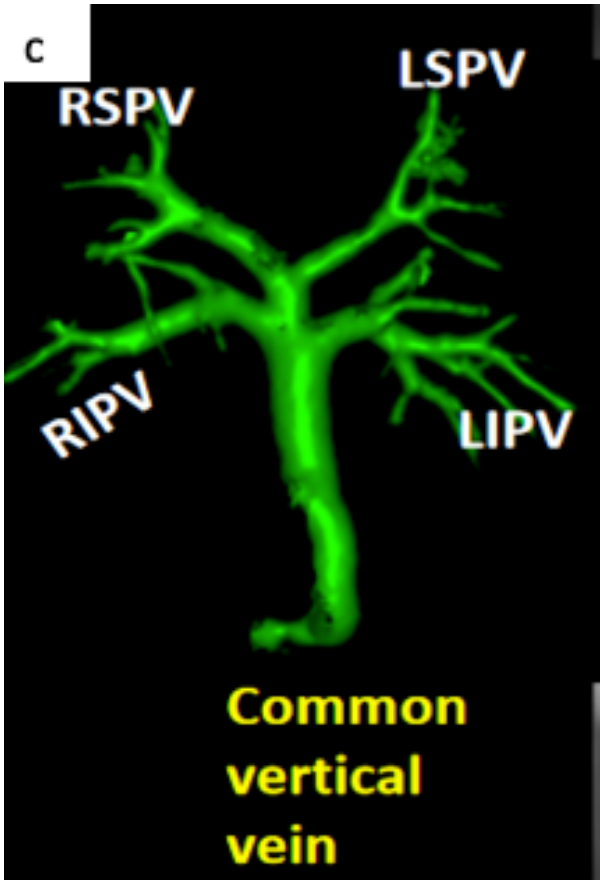


Fig. 4 (a-e) – A case of Infracardiac TAPVC a) and b) reformatted coronal image shows all 4 pulmonary veins joining together to form the common vertical vein and draining into the portal vein (annotated) c) shows the 3D reformatted image showing the venous mapping of all 4 pulmonary veins combines to form the vertical vein, d) and e) shows the 3D reformatted image of showing venous mapping shows partial short segment stenosis(yellow arrow) in vertical vein before draining into portal vein of infracardiac total pulmonary venous anomaly.



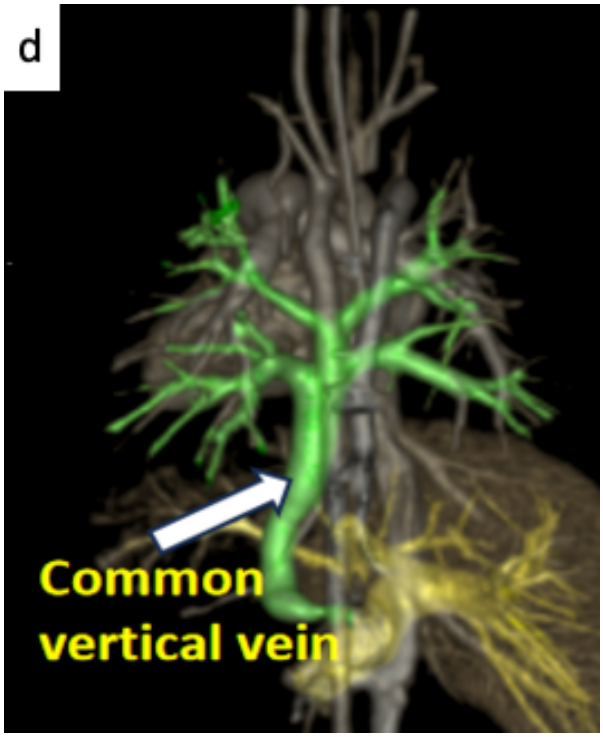


Fig. 4 (c-d)

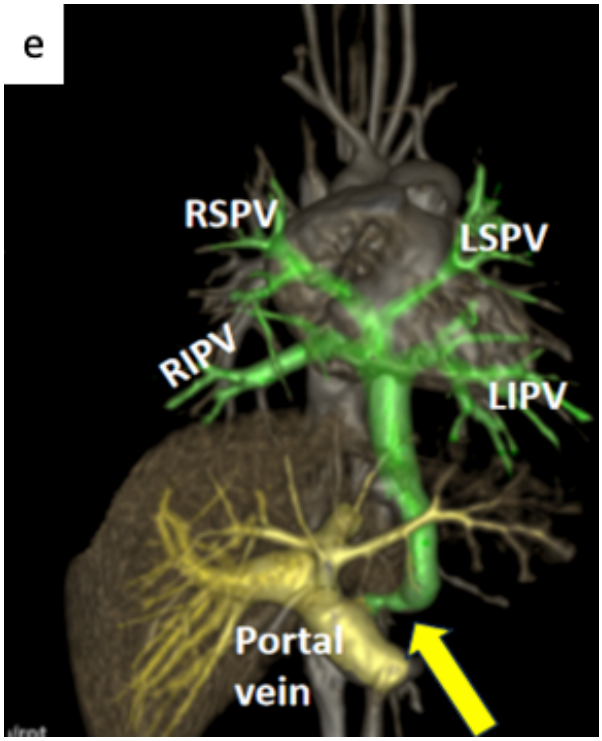


Fig. 4 (e)

And another case:
of a 2 Year Old Child (Fig. 5 a-d)
of ***non obstructed variant*** *is presented in Fig. 5.*

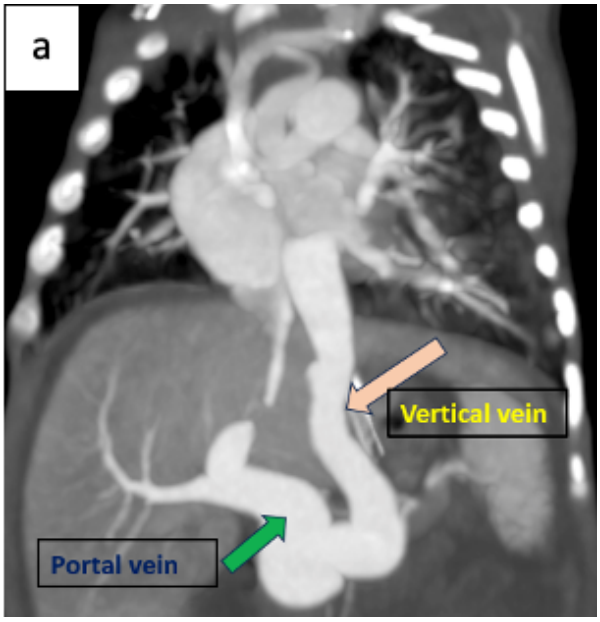
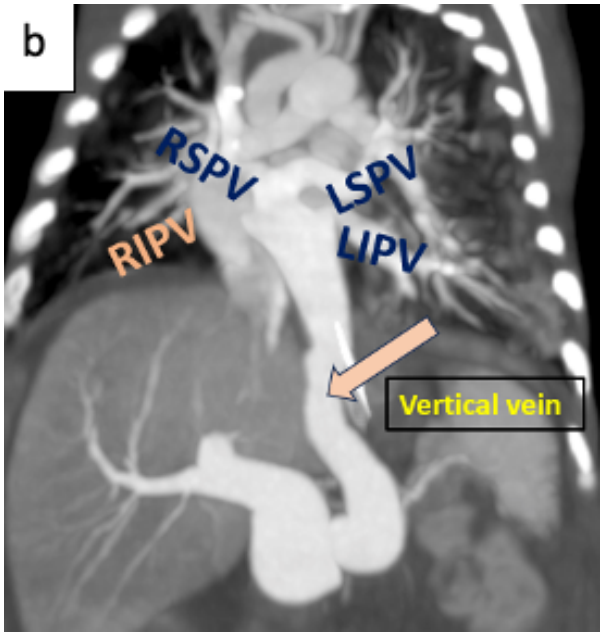


Fig. 5 (a-d)



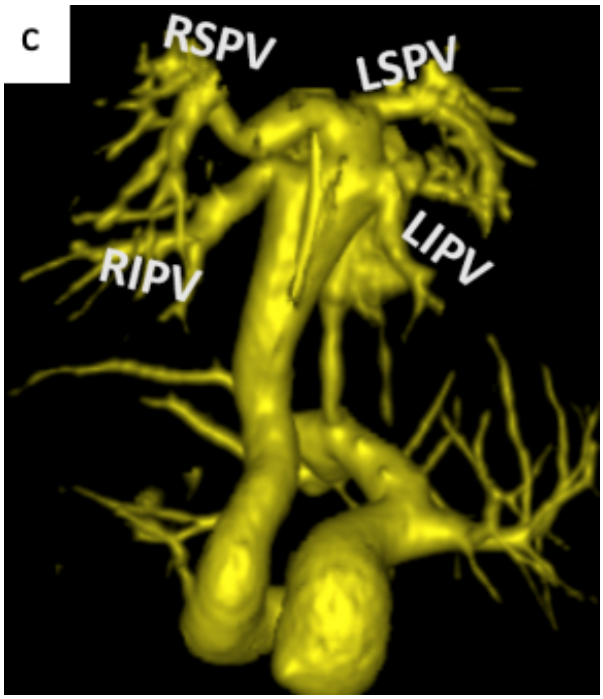


Fig. 5 (a-d) – A case of infra-cardiac TAPVC- Non obstructive type a) and b) reformatted coronal image shows all 4 pulmonary veins joining together to form the common vertical vein and draining into the portal vein (annotated) c) and d) shows the 3D reformatted image of showing venous mapping vertical vein draining into portal vein with no stenosis infracardiac total pulmonary venous anomaly non-obstructive type.

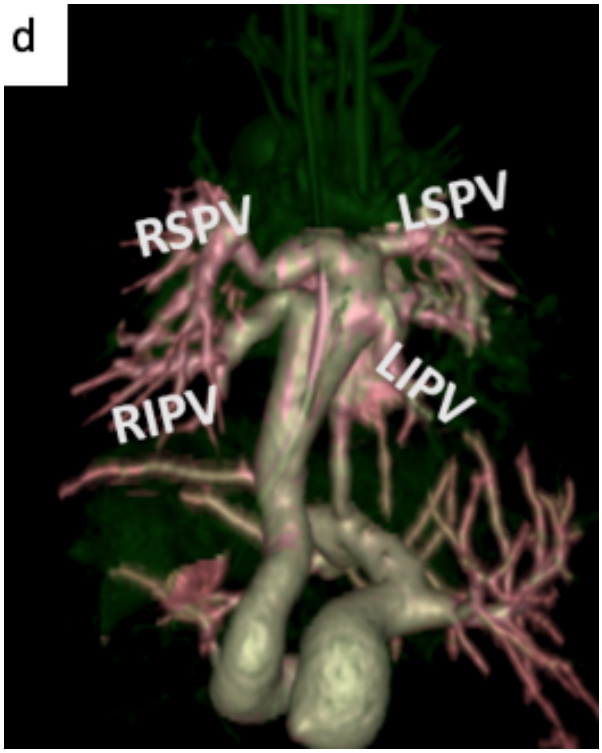
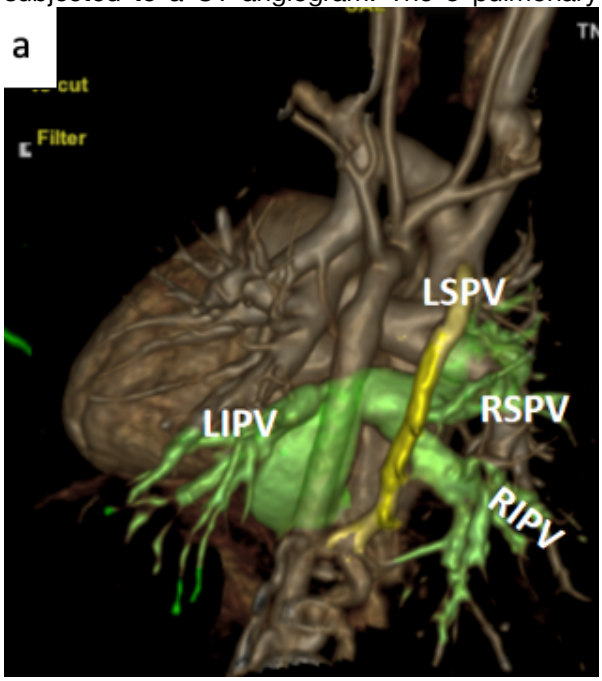


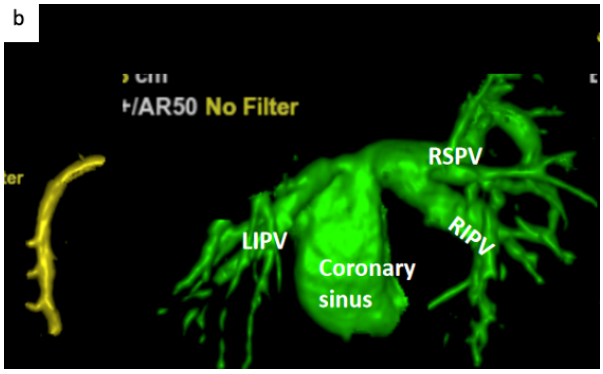
Fig. 5 d

Case 4: Mixed type of total anomalous pulmonary venous connection - (Fig. 6 a-c)

A 55-day-old male child presented for evaluation cyanosis during feeding and irritability which then subjected to a CT angiogram. The 3 pulmonary



veins, including the right superior pulmonary vein, the right inferior pulmonary vein, the left inferior pulmonary vein joining together and draining into the coronary sinus, and the left superior pulmonary vein draining into the left brachiocephalic vein (Fig. 6).



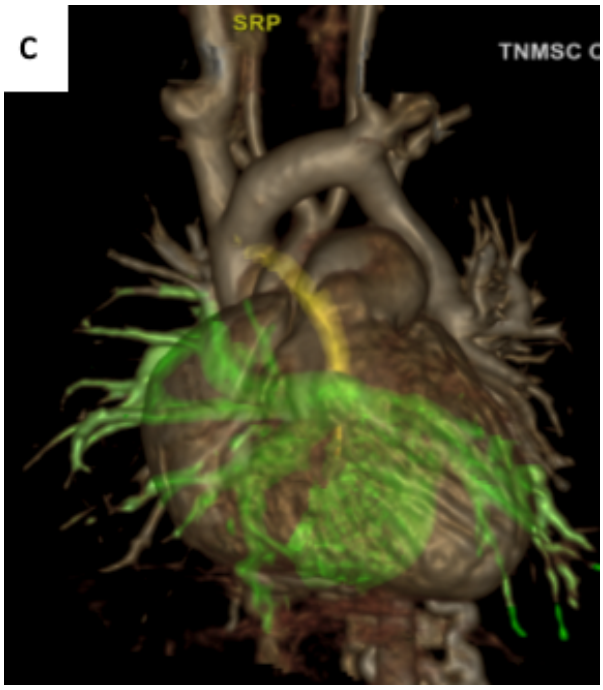


Fig. 6 (a-c) – A case of mixed type of TAPVC a) b) and c) reformatted volume rendered oblique image shows all 3 pulmonary veins (green) joining together and draining into the coronary sinus (annotated) and left superior pulmonary vein draining into left brachiocephalic vein b) and c) shows the 3D reformatted image showing the venous mapping of all 3 pulmonary veins combines draining into coronary sinus, and LSPV into brachiocephalic vein.

Case 5: Partial anomalous venous drainage (Scimitar syndrome)

A 10-year-old female presented with complaints of exertional dyspnea with no other comorbidities. The patient was subjected to a CT angiogram which reveals dextroposition of the heart with hypoplastic right lung, right pulmonary artery

and mediastinal shift towards the right. A reconstructed coronal image shows the right superior and right inferior pulmonary veins draining into the IVC and the left superior and left inferior pulmonary veins draining into the left atrium. Systemic arterial supply from the aorta to the right lobe is also noted (Fig. 7).

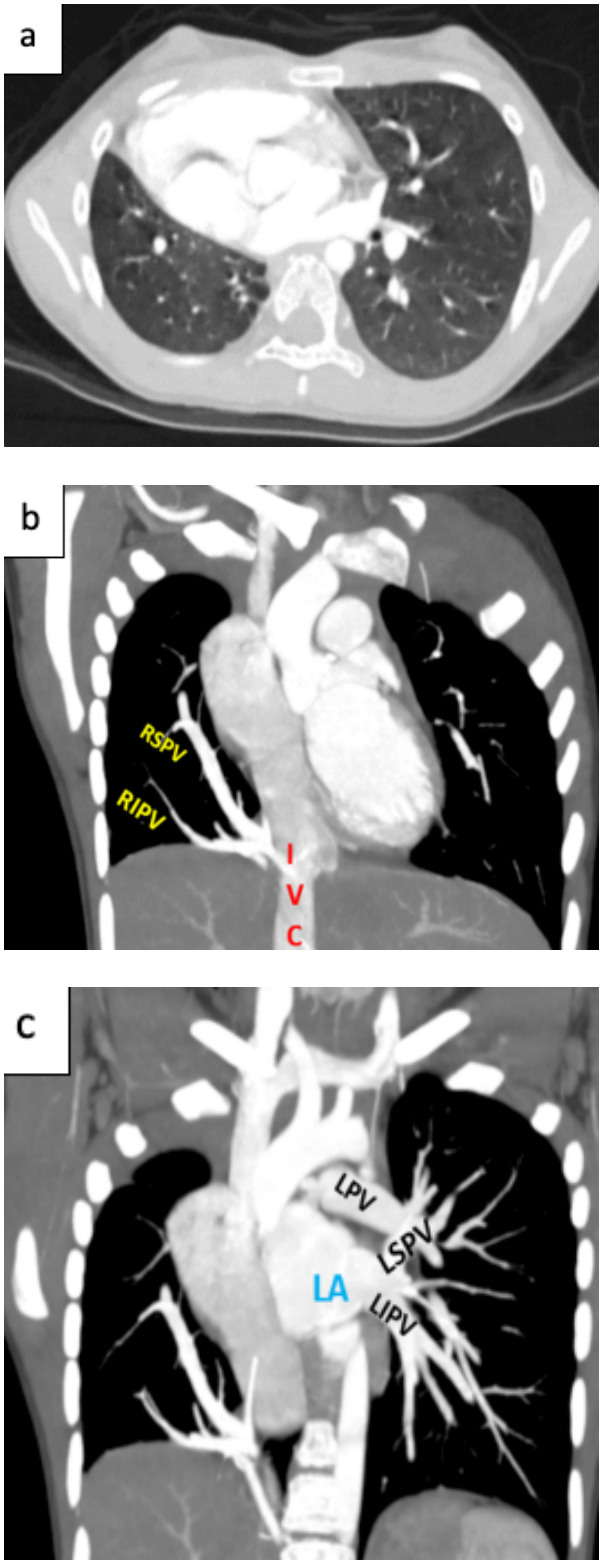


Fig. 7 (a-g) – A case of partial anomalous venous drainage (scimitar syndrome), a) axial image shows dextroposition of heart with hypoplastic right lung and mediastinal shift towards right, b) reformatted coronal image shows right superior and right inferior pulmonary veins draining into the IVC c) reformatted coronal image shows left superior and left inferior pulmonary veins draining into the left atrium d) and e) volume rendered image posterior view shows the RSPV, RIPV (green) drains into IVC, LSPV, LIPV drains into left atrium (yellow) f) volume rendered image shows right pulmonary artery appears hypoplastic g) shows systemic arterial supply from aorta to the right lobe (red arrow).

Discussion

Total anomalous pulmonary venous connection (TAPVC)

TAPVC, or total anomalous pulmonary venous return (TAPVR), accounts for approximately 1–5% of cardiovascular congenital anomalies (2,3). TAPVC implies that all four pulmonary veins connect to right atrium or the systemic veins, either by a common vertical vein or individually. Anatomically, TAPVC can be divided into four subtypes based on the level of the anomalous connection: supracardiac, cardiac, infracardiac, and mixed (4).

- Supracardiac (type 1): Pulmonary veins anomalously connect, most commonly to the left innominate vein. Other locations for connections include the SVC and azygous vein.
- Cardiac (type 2): Pulmonary veins drain into the coronary sinus or right atrium directly.
- Infracardiac (type 3): Pulmonary veins form a vertical vein and drain into the portal vein, hepatic vein, or IVC below the diaphragm.
- Mixed (type 4): In mixed-type, there is a combination of connections at different levels.

The supracardiac variety is the most common of the TAPVC. (5)

Partial anomalous venous drainage (PAPVD)

The prevalence of PAPVD has been reported to be between 0.4 and 0.7% (6,8) and may be incidentally detected on CT or MRI (6). In PAPVC, at least one pulmonary vein drains into another location instead of the left atrium. Clinically, patients with PAPVC are typically acyanotic or mildly symptomatic This partial anomalous drainage results in a left-to-right shunt. Some authors have suggested that PAPVC becomes clinically significant when 50% or more of the pulmonary blood flow returns anomalously (6).

Scimitar syndrome - or hypogenetic lung or pulmonary venolobar syndrome

is a rare form of PAPVC that almost always involves the right lung (7). In this, a portion or all of the right lung pulmonary venous blood flow drains to the IVC (above or below the diaphragm), the azygos system, the right atrium, the portal venous system, or a hepatic vein. Other findings seen in patients with Scimitar syndrome are a small ipsilateral hemithorax, cardiac dextroposition, and pulmonary artery hypoplasia or aplasia, which is seen in our case (Fig. 5). Systemic arterial collateral blood supply to the ipsilateral lung may also be seen arising from the aorta (Fig. 7g) or its branch vessels, such as the celiac axis (3).

Conclusion

MDCT helps in mapping of drainage site of the common pulmonary vein, stenosis of the vertical vein and the course of the atypical vessel into the systemic vein. Accurate characterization of these anomalies is essential for the presurgical planning. Volume rendering is most useful in the evaluation of patients with mixed TAPVC and PAPVC, as well as pulmonary vein stenosis and hypoplasia / atresia, which is difficult to characterize with echocardiography.

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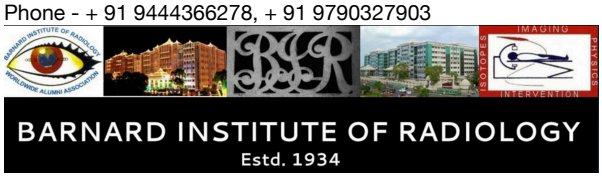

Conflict of interest:

The authors declare that there were no conflicts of interest within the meaning of the recommendations of the International Committee of Medical Journal Editors when the article was written.

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