Radiology Case Reports 20 (2025) 1243–1246

Available online at www.sciencedirect.com

ScienceDirect

journal homepage: www.elsevier.com/locate/radcr

Case Report

Supracardiac type of total anomalous pulmonary venous connection: Diagnosis and demonstration by multidetector CT angiography ☆,☆☆

Neha Singh, MD[a,*], *Guriya Kumari, MD*[a], *Deepak Kumar Singh, MCh*[b]

[a] *Department of Radiodiagnosis and Imaging, Dr. Ram Manohar Lohia Institute of Medical Sciences, Lucknow, India*
[b] *Department of Neurosurgery, Dr. Ram Manohar Lohia Institute of Medical Sciences, Lucknow, India*

ARTICLE INFO

Article history:
Received 4 October 2024
Revised 5 November 2024
Accepted 7 November 2024

Keywords:
Supracardiac TAPVC
TAPVC
TAPVR
MDCT

ABSTRACT

Pulmonary venous developmental anomalies have historically been evaluated using echocardiography and catheter-based angiography. In recent years, however, multidetector CT angiography (MDCTA) and MR angiography have become increasingly important tools for detailed characterization of these anomalies. This case report provides an in-depth review of the radiologic findings in a 15-year-old patient diagnosed with the supracardiac type of Total Anomalous Pulmonary Venous Connection (TAPVC). The report emphasizes the imaging features that were instrumental in the diagnosis and underscores the crucial role of advanced imaging techniques in the management of this serious congenital heart defect.

Background

Total anomalous pulmonary venous connection (TAPVC) is a rare congenital anomaly where the pulmonary veins fail to connect to the left atrium. Instead, they drain into the systemic venous circulation, causing oxygenated and deoxygenated blood to mix and flow to the body through an atrial septal defect (ASD), patent foramen ovale (PFO), or patent ductus arteriosus (PDA). This leads to significant hemodynamic disturbances and hypoxemia. TAPVC represents about 1.5%-3% of all congenital heart defects and is classified into four types based on the location of the abnormal venous connection: supracardiac, cardiac, infracardiac, and mixed. The supracardiac type is the most common, accounting for 50%-55% of cases, where the pulmonary veins drain into the superior vena cava or innominate vein via a vertical vein, bypassing the left atrium entirely. Advances in imaging techniques, such as multidetector CT (MDCT), now allow for highly accurate, noninvasive diagnosis of this condition [1,2].

Abbreviations: TAPVC, Total anomalous pulmonary venous connection; SVC, Superior vena cava; LA, Left atrium; ASD, Atrial septal defect; PFO, Patent foramen ovale; PDA, Patent ductus arteriosus; ECG, Electrocardiography; MDCTA, Multidetector Computed tomography angiography; MPR, Multiplanar reformation.

☆ Competing Interests: The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

☆☆ Acknowledgments: This research received no specific grant from any funding agency in public, commercial, or not-for-profit sectors.

* Corresponding author.
E-mail address: neha.singh.dr@gmail.com (N. Singh).

https://doi.org/10.1016/j.radcr.2024.11.018

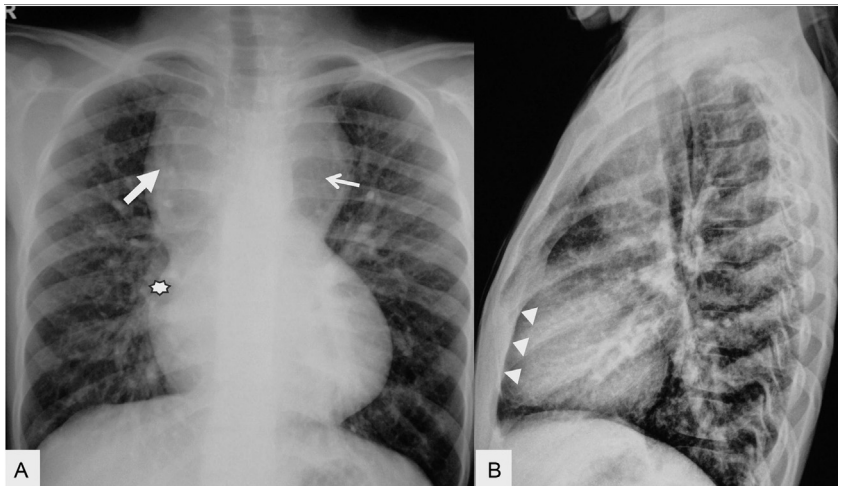

Fig. 1 – Chest Radiography PA (A) and lateral (B) view show cardiomegaly and a "snowman" or "figure-of-eight" sign. Head of the snowman is formed by the abnormal vertical vein (thin arrow) and the dilated superior vena cava (thick arrow) while the enlarged right atrium (*) forms the "body". Arrowhead in image (B) shows obliterated retrosternal space.

Case description, diagnosis and management

A 15-year-old boy presented with worsening cyanosis and shortness of breath. His medical history revealed childhood-onset symptoms, including breathlessness, recurrent chest infections, cyanosis and easy fatigability. On examination, he exhibited tachypnea, tachycardia, digital clubbing and cyanosis of the lips. A prominent right ventricular impulse was palpated, along with a loud, fixed split second heart sound and a systolic murmur, best heard at the left upper sternal border. The ECG showed a prolonged PR interval, peaked P waves in the right atrium, tall R waves in the right precordial leads, inverted T waves, a deep S wave in the left precordial leads, and right axis deviation, all consistent with right ventricular enlargement.

Chest radiography revealed cardiomegaly with prominent pulmonary arteries, along with a "snowman" or "figure-of-eight" sign, suggesting a potential diagnosis of supracardiac TAPVC (Fig. 1). Echocardiography confirmed that all four pulmonary veins converged into a common venous chamber, which drained via a vertical vein into the left innominate vein, and subsequently into the superior vena cava (SVC). The left atrium was underfilled and appeared smaller than normal, while the right atrium and right ventricle were significantly enlarged. Evidence of right ventricular hypertrophy indicated increased workload due to the abnormal pulmonary venous return. Based on these findings, a provisional diagnosis of supracardiac TAPVC was made, and the patient was referred for 64-slice CT angiography to confirm the diagnosis.

A retrospective ECG-gated cardiac CTA was performed with the administration of 70 ml of nonionic contrast agent (iohexol 350 mg I/ml), followed by a 30 ml saline flush. To minimize patient radiation exposure, ECG-gated tube current modulation was applied. The images were reconstructed during the diastolic phase. Three-dimensional (3D) reconstructions in various projections along with multiplanar reformations (MPR) and volume rendering (VR), demonstrated all four pulmonary veins converging into a common channel posterior to the left atrium (LA) before draining into the left innominate vein, and subsequently entering the superior vena cava (SVC). The CT images revealed markedly dilated right-sided cardiac chambers and an enlarged pulmonary artery (Figs. 2 and 3). Notably, there was no evidence of pulmonary vein obstruction, a condition sometimes associated with TAPVC. The three-dimensional reconstructions from the CT scan were particularly valuable for surgical planning, offering a clear depiction of the vertical vein's course and its connections.

Discussion

Total anomalous pulmonary venous connection (TAPVC) is a cyanotic congenital heart defect in which all pulmonary veins, carrying oxygenated blood from the lungs, incorrectly drain directly or indirectly into the right atrium instead of the left. This results in oxygenated blood bypassing the left atrium, returning to the right side of the heart. TAPVC is categorized anatomically into four subtypes based on the location of the abnormal connection: supracardiac, cardiac, infracardiac, and mixed [1,2] In the supracardiac type, the pulmonary veins connect abnormally to the left innominate vein, superior vena cava (SVC), or azygos vein via an ascending vertical vein. In the cardiac type, the pulmonary venous confluence drains into the coronary sinus, or, in rare cases, individual pulmonary veins drain directly into the right atrium [3]. In the infracardiac type, pulmonary veins form a descending vertical vein that drains below the diaphragm into the portal vein, hepatic vein, or inferior vena cava (IVC). The mixed type involves combinations of connections at different anatomical levels [4].

Supracardiac TAPVC is typically diagnosed in infancy due to severe symptoms like cyanosis, respiratory distress, recurrent chest infections, and failure to thrive [5]. However, in

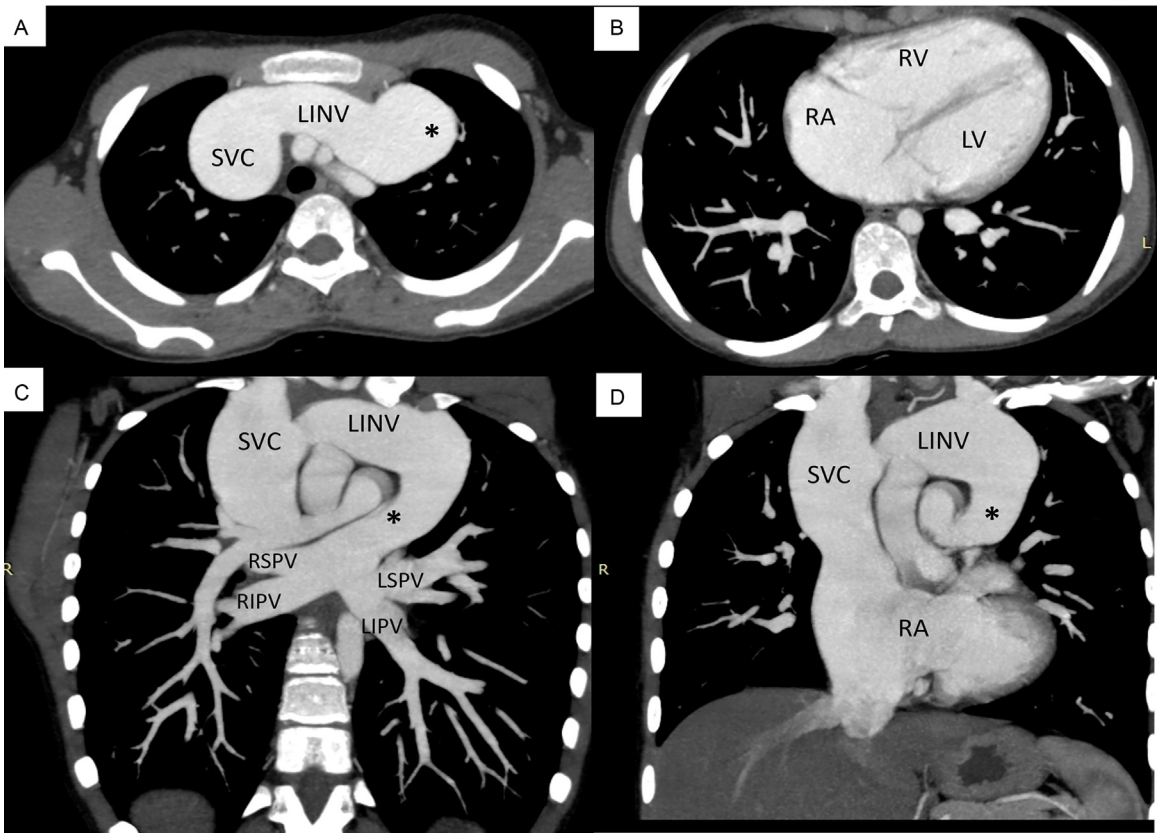

Fig. 2 – Axial (A and B) cardiac CT angiography and coronal reformatted images (C and D) show all the four pulmonary veins (RSPV- Right superior pulmonary vein, RIPV- Right inferior pulmonary vein, LSPV- Left superior pulmonary vein, LIPV- Left inferior pulmonary vein) converging to form ascending vein (*) which is ascending superiorly and draining into left innominate vein (LINV). Image (B) shows right heart enlargement.

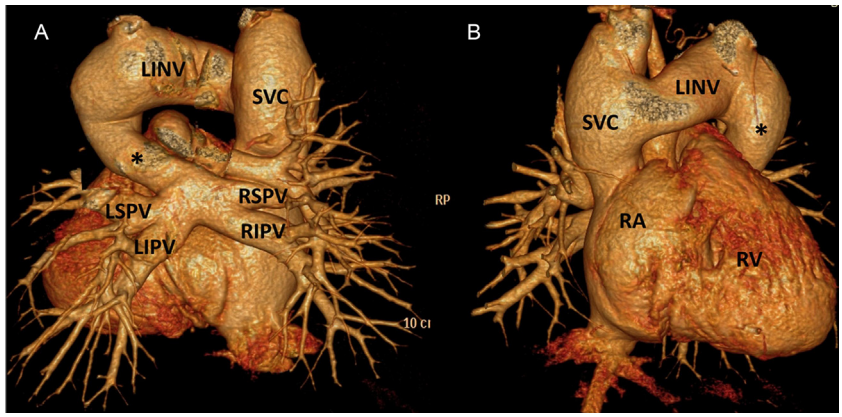

Fig. 3 – Volume rendered cardiac CT angiography images (A- from posterior aspect and B- from anterior aspect) show all the four pulmonary veins converging to form ascending vein (*) which is ascending superiorly and draining into left innominate vein (LINV). Left innominate vein and superior vena cava (SVC) appear dilated.

this case, the diagnosis was made in an adolescent, which is unusual. The late presentation in this 15-year-old patient highlights the variability in clinical course of TAPVC and the importance of maintaining a high index of suspicion for the condition in adolescents and older children with unexplained cyanosis and right heart failure.

In this case, radiological findings played a key role in diagnosing supracardiac TAPVC. The chest X-ray revealed the characteristic 'snowman' sign, a well-known radiographic feature of this condition. The 'head' of the snowman represents the abnormal vertical vein and dilated superior vena cava (SVC), while the "body" is formed by the enlarged right atrium

and pulmonary artery. Although highly suggestive of the diagnosis, this sign is not pathognomonic, and further imaging with echocardiography, CT, or MRI is required to confirm the diagnosis and guide surgical planning [6].

Echocardiography plays a pivotal role in diagnosing TAPVC, offering detailed visualization of abnormal pulmonary vein connections and their hemodynamic consequences. In this case, it revealed that all pulmonary veins drained anomalously into a vertical vein, completely bypassing the left atrium. The enlargement of the right atrium and ventricle along with right ventricular hypertrophy, suggested chronic volume overload and pulmonary hypertension [7]. CT angiography and MRI provided complementary anatomical and functional data crucial for surgical planning. Three-dimensional reconstructions from CT were particularly useful in mapping the vertical vein's course and its connection to the systemic venous system. MRI can provide valuable insights into ventricular function and the extent of pulmonary overcirculation [8,9].

Although cardiac catheterization is not routinely required for diagnosis in all cases, it was crucial in this instance to evaluate pulmonary pressures and vascular resistance, considering the patient's age and symptoms. The findings of elevated right-sided pressures and an increased pulmonary to systemic flow ratio (Qp:Qs) confirmed the hemodynamic burden caused by the anomalous venous return and emphasized the urgency of surgical correction [10].

The timing of surgical intervention in Total Anomalous Pulmonary Venous Connection (TAPVC) is critical; delays can lead to irreversible pulmonary hypertension and right heart failure. In this patient, the successful rerouting of the pulmonary veins to the left atrium, combined with ligation of the vertical vein, resulted in significant clinical improvement, including normalization of oxygen levels and resolution of right heart strain. This outcome highlights the necessity of early and accurate diagnosis, alongside prompt surgical intervention, to prevent the potentially life-threatening complications associated with this condition [11].

Conclusion

In a young patient presenting with progressive cyanosis, TAPVC should be a primary consideration, warranting a thorough investigation using appropriate imaging techniques. Multidetector CT angiography (MDCTA) serves as a rapid, noninvasive, and precise diagnostic tool, providing a three-dimensional visualization of vascular anatomy in suspected cases of pulmonary venous connection anomalies. A comprehensive imaging approach, incorporating echocardiography, CT, and MRI, is crucial for confirming the diagnosis, elucidating the anatomical details of the defect, and facilitating effective surgical planning for correction.

Patient consent

Written informed consent for patient information and images to be published was provided by the patients.

REFERENCES

[1] Van Praagh R, Van Praagh S. The anatomy of common congenital heart disease. Am J Cardiol 1965;15(3):285–342.
[2] Qu Y, Liu X, Zhuang J, Chen G, Mai J, Guo X, et al. Incidence of congenital heart disease: the 9-year experience of the guangdong registry of congenital heart disease, China. PLoS One 2016;11(7):e0159257. doi:10.1371/journal.pone.0159257.
[3] Singh N, Singh R, Aga P, Singh SK. Cardiac type of total anomalous pulmonary venous connection: diagnosis and demonstration by multidetector CT angiography. BMJ Case Rep 2013;2013:bcr2012007994. doi:10.1136/bcr-2012-007994.
[4] Konduri A, Aggarwal S. Partial and total anomalous pulmonary venous connection. StatPearls. Treasure Island (FL): StatPearls Publishing; 2023.
[5] Mir FA, Mohanty S, Pandey A, Naqashbandi A, Das D. Supracardiac total anomalous pulmonary venous connection in adolescence. Cureus 2023;15(10):e47392. doi:10.7759/cureus.47392.
[6] Ferguson EC, Krishnamurthy R, Oldham SA. Classic imaging signs of congenital cardiovascular abnormalities. Radiographics 2007;27(5):1323–34. doi:10.1148/rg.275065148.
[7] Lai W, M L, Cohen M, Geva T. Echocardiography in paediatric and congenital heart disease from foetus to adult. 2nd ed. Wiley-Blackwell. [Accessed December 2015].
[8] Dillman JR, Hernandez RJ. Role of CT in the evaluation of congenital cardiovascular disease in children. AJR Am J Roentgenol 2009;192(5):1219–31. doi:10.2214/AJR.09.2382.
[9] Files MD, Morray B. Total anomalous pulmonary venous connection: preoperative anatomy, physiology, imaging, and interventional management of postoperative pulmonary venous obstruction. Semin Cardiothorac Vasc Anesth 2017;21(2):123–31. doi:10.1177/1089253216672442.
[10] Kang SL, Benson L. Recent advances in cardiac catheterization for congenital heart disease. F1000Res 2018;7:370. doi:10.12688/f1000research.13021.1.
[11] Yong MS, Zhu MZL, Konstantinov IE. Total anomalous pulmonary venous drainage repair: redefining the long-term expectations. J Thorac Dis 2018;10(Suppl 26):S3207–10. doi:10.21037/jtd.2018.08.05.