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Case Report

Total Anomalous Pulmonary Venous Connection with Rare Direct Right Atrial Drainage and Unprecedented Array of Coexistent Multisystem Variations

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Abstract

Total anomalous pulmonary venous connection (TAPVC) is anomalous drainage of all pulmonary veins into systemic circulation. The intracardiac type typically entails the drainage of all the pulmonary veins into the right atrium, via the coronary sinus. The connection of the pulmonary veins directly into the right atrium is exceptionally rare and has been primarily reported with right atrial isomerism. Herein, we presented a remarkable case of TAPVC in a 10-year-old male child, distinguished by an unconventional drainage of all the pulmonary veins directly into the right atrium, with normal coronary sinus and absent right atrial isomerism. Intriguingly, computed tomography imaging revealed a combination of incredibly rare coexistent pulmonary, vascular, and skeletal anomalies. These anomalies included absence of pulmonary fissures in the right lung, presence of left circumflex aortic arch with bovine branching pattern, bilateral cervical ribs, and C7 vertebral fusion anomalies. To our knowledge, this unique combination of coexistent anomalies has not been previously reported in scientific literature in the background of rare drainage pattern of TAPVC.

Keywords
- TAPVC
- cardiac TAPVC
- MDCT
- pulmonary venous drainage
- circumflex aortic arch

Introduction

Total anomalous pulmonary venous connection (TAPVC) is a common cause of cyanotic cardiac disorders. An obligatory shunt is required to direct the blood to the left side of the heart. The supracardiac type is the most common type, followed by the infracardiac and cardiac types.[1] The most common intracardiac pattern is the drainage of all veins into the right atrium (RA) via the coronary sinus (CS). The direct drainage of the pulmonary veins into the RA is an extremely rare occurrence, usually associated with right atrial isomerism and requires distinct treatment strategies. distinct treatment strategies. In this article, we presented a rare drainage pattern of TAPVC in a 10-year-old male child with abnormal drainage of all four pulmonary veins directly into the RA, with normal CS. Computed tomography (CT) also revealed a rare combination of cardiovascular, pulmonary, and skeletal anomalies.

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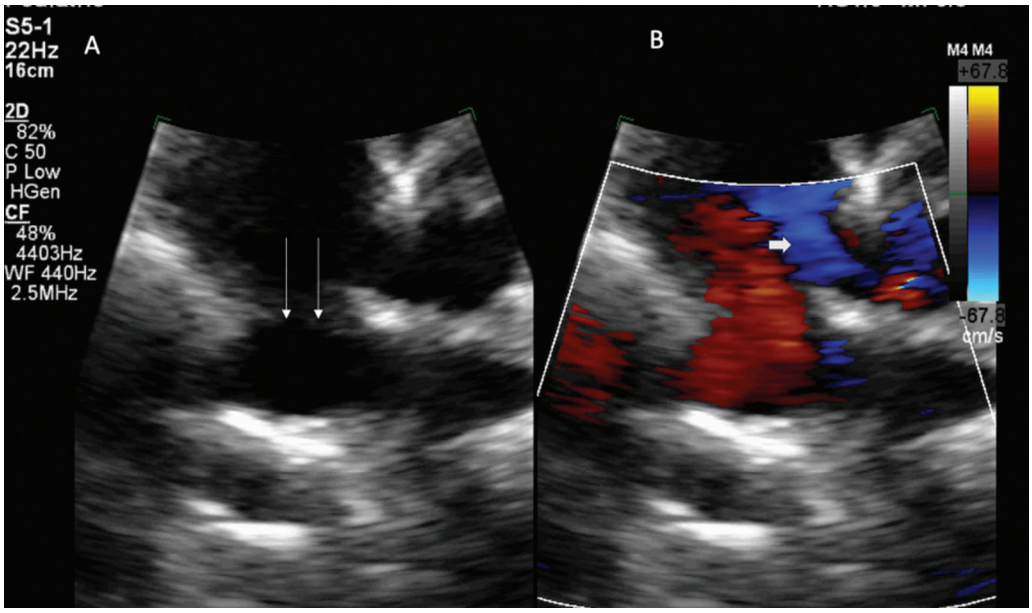

Fig. 1 (A, B) Transthoracic echo image depicting pulmonary confluence (*double arrow*) entering into the RA, ASD (*arrow*) shunting; right→left shunt. ASD, atrial septal defect; RA, right atrium.

Case Report

A 10-year-old male child presented to the pediatric cardiology department with complaints of easy fatiguability, poor weight gain, and bluish discoloration of skin. On examination, his pulse rate and respiratory rate were elevated, with oxygen saturation of 91% on room air and left parasternal (second intercostal space) systolic murmur. Echocardiography (►**Fig. 1**) showed TAPVC with associated atrial septal defect (ASD). CT angiography (►**Figs. 2**–**3**) nicely demonstrated the anomalous drainage of all the four pulmonary veins along the posterosuperior aspect of the RA. The CS was normal in size (►**Fig. 4D**). A large ASD was seen (►**Fig. 4A**). The main pulmonary artery and its branches were dilated with dilated right-sided cardiac chambers. Interestingly, multiple other associated anomalies were also identified. Anterior protrusion of the sternum (pectus carinatum) was observed (►**Fig. 4B**). Major and minor fissures were both absent in the right lung (►**Fig. 4B**). The left lung was normal. In addition, the left circumflex aortic arch with a common origin of the left common carotid artery (CCA) and brachiocephalic trunk was also noted (►**Fig. 4F-G**). No patent ductus arteriosus (PDA) was seen. Skeletal variations like the presence of bilateral cervical ribs and fusion anomaly of the C7 vertebra were also seen (►**Fig. 4**). The child is presently awaiting surgery.

Discussion

TAPVC refers to the anomalous drainage of all the four pulmonary veins into systemic circulation. The estimated incidence of TAPVC is 7 to 9 per 100,000 births, constituting 0.7 to 1.5% of all congenital cardiac disorders.[1–3] Patients can present with cyanosis, respiratory distress, feeding intolerance, and failure to thrive. TAPVC can be defined

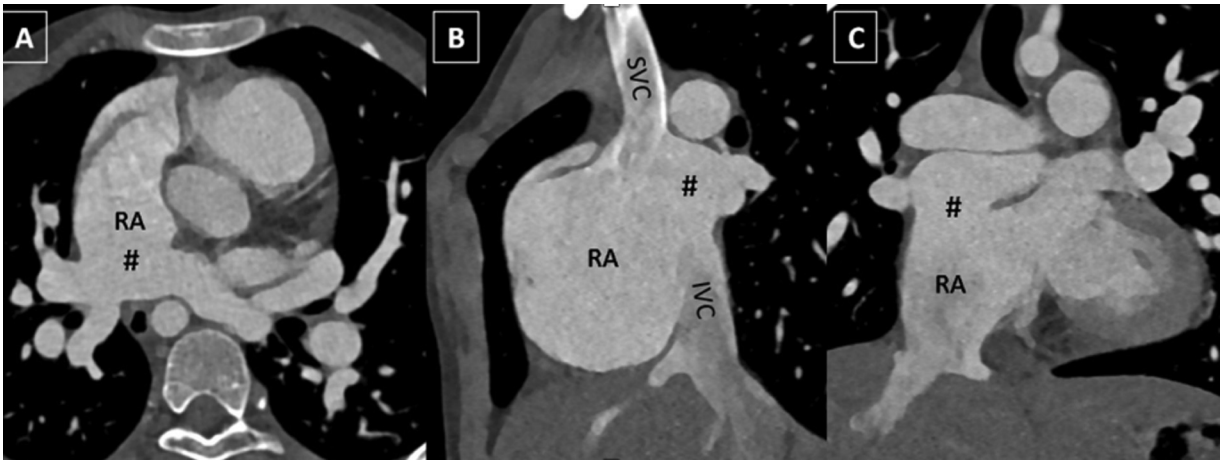

Fig. 2 (A) Axial, (B) oblique sagittal, and (C) oblique coronal reformatted images depicting the anomalous drainage of the pulmonary veins (#) into the right atrium (RA). IVC, inferior vena cava; SVC, superior vena cava.

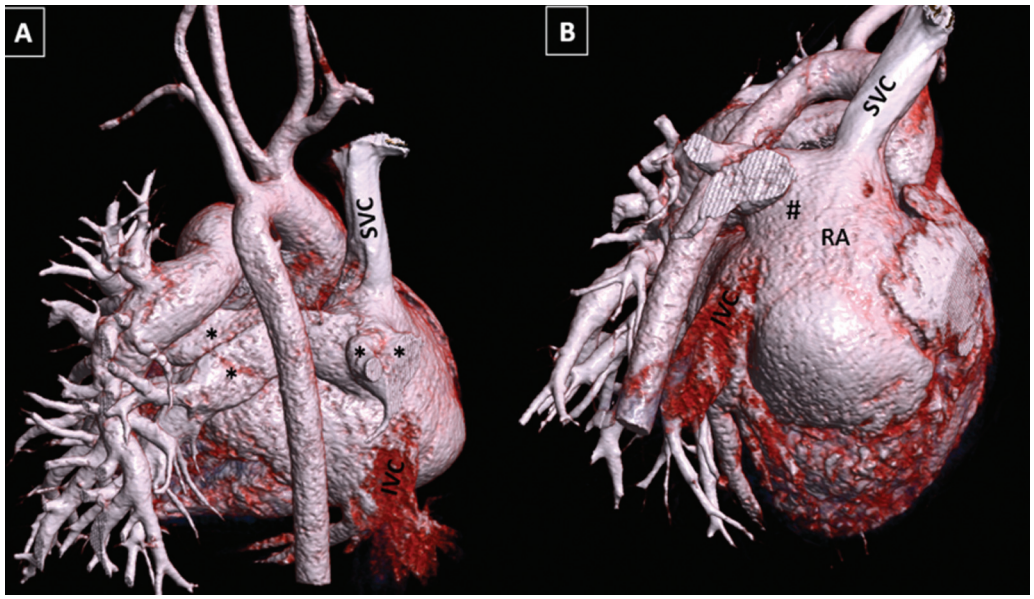

Fig. 3 (**A**, **B**) Volume-rendered images depicting the anomalous drainage of the pulmonary veins (*) into the right atrium (#). RA, right atrium; SVC, superior vena cava.

by the drainage into the systemic circulation as the supracardiac (45%), infracardiac (25%), cardiac (25%), and mixed (5%) subtypes.[4–6] The cardiac variant of TAPVC is a relatively infrequent subtype in which the common pulmonary venous chamber drain into the RA usually through the CS, after formation of a common confluence (CS type).[7] The direct drainage into the RA is extremely rare and is mostly reported in association with atrial isomerism.[8] It is extremely rare in the presence of normal CS and situs solitus.[8] Echocardiography is the initial screening tool. Important clues to the identification of this rare pattern include normal-sized CS, absence of any ascending or descending vein with tracing of the individual pulmonary veins to their site of entry to the RA.[7] CT angiography is a

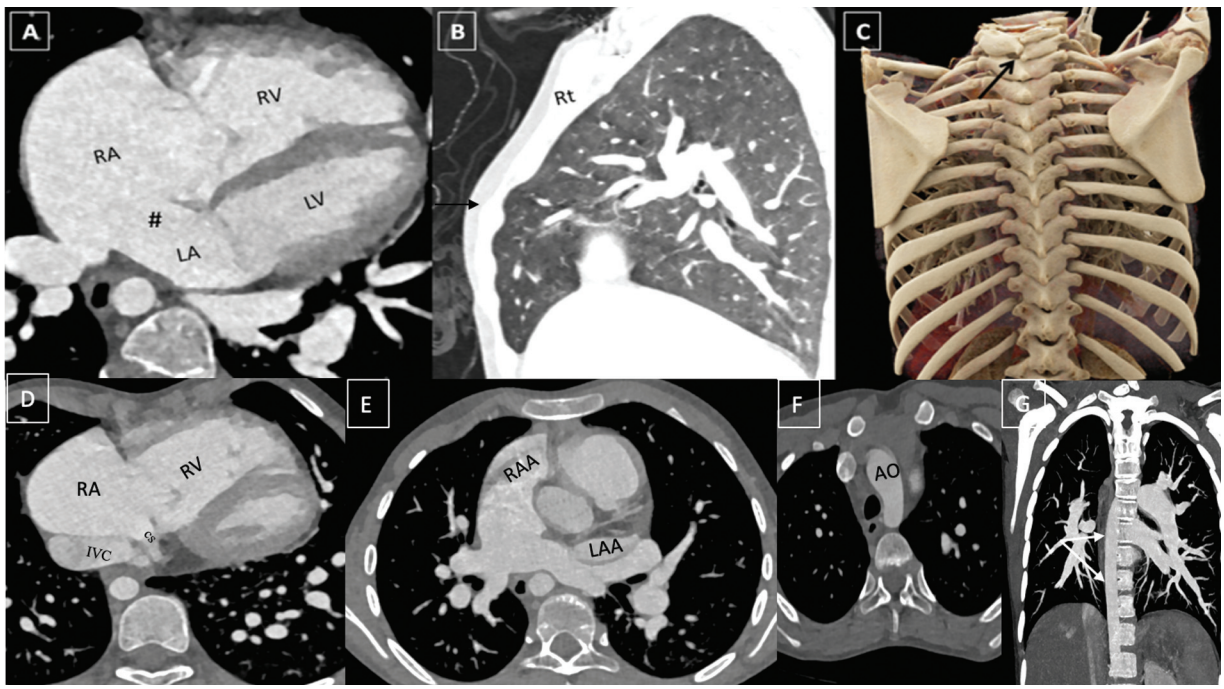

Fig. 4 (**A**) Axial image showing large atrial septal defect (#). (**B**) Sagittal reformatted image in the lung window showing the right lung with absent pulmonary fissures and anterior protrusion of the sternum—pectus carinatum (*black arrow*). (**C**) Volume-rendered image depicting fusion anomaly of the spinous process of C7 (*arrow*). (**D**) Axial image showing normal coronary sinus (CS). (**E**) Normal right atrial appendage (RAA) and left atrial appendage (LAA) morphology. (**F**) Axial and (**G**) coronal images depicting left circumflex aortic arch (AO) crossing the midline with the descending thoracic aorta on the right side (*grouped arrows*). IVC, inferior vena cava; LV, left ventricle; RA, right atrium; RV, right ventricle.

noninvasive investigation with good temporal and spatial resolution that allows confirmation of the echocardiography findings and excellent delineation of not only the detailed cardiac anatomy but also associated coexistent congenital anomalies like the left circumflex aortic arch imperative for presurgical planning.[9] The objective of the surgical management is to redirect the pulmonary venous flow into the left atrium, and the approach depends upon the anatomical course. In the CS type, incision is given at the CS along its anterior wall to establish the direct communication with the left atrium, along with the patch repair of ASD and coronary ostium. However, the RA type is usually repaired by creating a baffle with pericardial patch to redirect blood flow through the ASD to the left atrium.[10] In the case of anomalous pulmonary venous drainage into the superior vena cava (SVC)/inferior vena cava (IVC), the presurgical planning also depends upon the level of opening into the systemic veins.[11] The CT images can be reformatted into different planes and volume-rendered images, which provide comprehensive understanding of anatomical details that are imperative for appropriate tailored surgical approach.

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Conflict of Interest
None declared.

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