Mixed Total Anomalous Pulmonary Venous Connection to Superior Cavoatrial Junction

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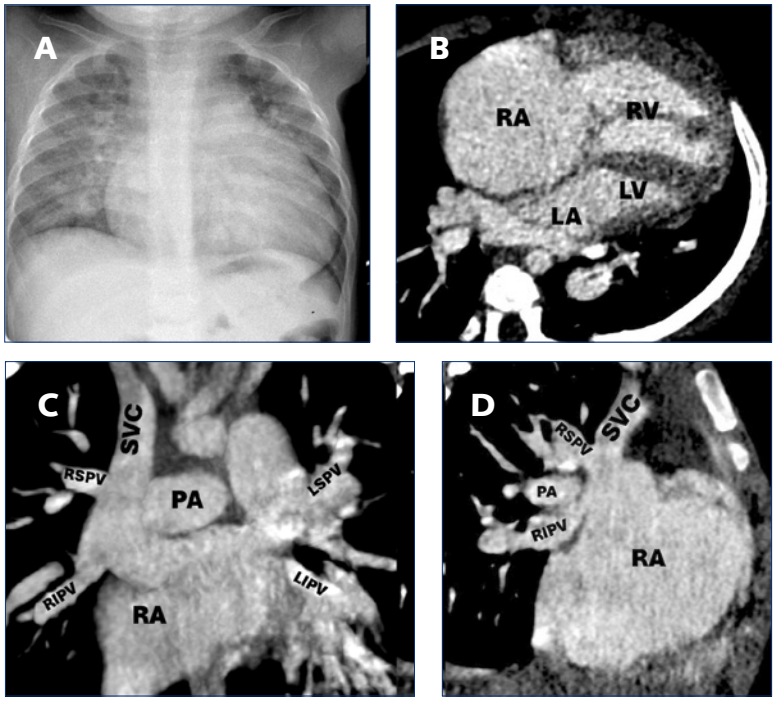
CASE PRESENTATION

An 11-month-old male with a seven-month history of diaphoresis and malaise during breastfeeding was referred for evaluation after a pulmonary systolic murmur was noted on physical examination. Chest radiography revealed cardiomegaly mainly affecting the right heart chambers [**Figure 1A**]. Transthoracic echocardiography (TTE) demonstrated right heart chamber dilation and a secundum atrial septal defect. Cardiac computed tomography angiogra- (CCTA) demonstrated a mixed-type total anomalous pulmonary venous connection (TAPVC), with the left pulmonary veins (PVs) and the right inferior PV draining via a common confluence into an enlarged right atrium; additionally, the right superior PV drained directly into the origin of the superior vena cava [**Figures 1B-D, Figure 2**]. The cardiovascular surgery department was consulted to perform surgical correction and establish pulmonary venous return to the left atrium.

Figure 1. Mixed Total Anomalous Pulmonary Venous Connection to the Superior Cavoatrial Junction

[A] Chest X-Ray showed right chamber cardiomegaly along with bilateral enlarged pulmonary hilum. CCTA showing an enlarged right atrium and hypertrophied right ventricle, along with right inferior pulmonary vein and left pulmonary veins draining to a common collector which flows to the right atrium as well as the right superior pulmonary vein draining independently to the origin of the superior vena cava; **[B]** Axial MPR view, **[C]** Coronal MIP view, **[D]** Sagittal MIP view.



CCTA: Cardiac Computed Tomography Angiography, LA: Left Atrium, LIPV: Left Inferior Pulmonary Vein, LSPV: Left Superior Pulmonary Vein, LV: Left Ventricle, MPR: Multiplanar Reconstruction, MIP: Maximum Intensity Projection, PA: Pulmonary Artery, RA: Right Atrium, RIPV: Right Inferior Pulmonary Vein, RSPV: Right Superior Pulmonary Vein, RV: Right Ventricle, SVC: Superior Vena Cava.

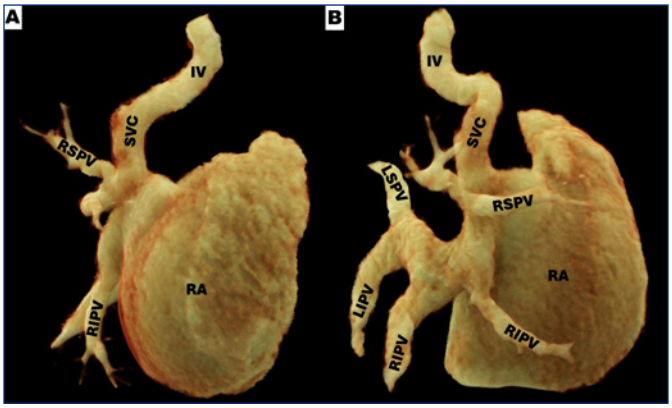
DISCUSSION

Mixed-type TAPVC, characterized by more than one level of pulmonary venous drainage, is the least frequent subtype, representing approximately 5% to 10% of all TAPVC cases.[1] Among these, variants in which all PVs drain into the superior cavoatrial junction are considered rare.[2] Although transthoracic echocardiography (TTE) is typically the first-line imaging modality and is sufficient in many cases to detect anomalous PV connections and obstruction, its diagnostic yield may be limited. Frequently, only two or three PVs are visualized, which can complicate accurate anatomical assessment. Consequently, up to 44.4% of mixed-type TAPVC cases are diagnosed only after surgery.[1] Given these challenges, a multimodality imaging approach is indicated in all patients with unrepaired TAPVC, especially before surgical intervention or in the presence of clinical deterioration or new symptoms.[3] CCTA provides comprehensive evaluation, allowing for precise identification of all four PVs, definition of their drainage pathways, and detailed delineation of PV anatomy through high-resolution, three-dimensional reconstructions.[1] Accurate characterization of PV anatomy is critical in mixed-type TAPVC, as surgical correction is technically complex due to the heterogeneous and variable arrangement of pulmonary venous confluences. The primary goal of surgical repair is to establish a non-obstructive connection between the pulmonary venous confluence and the left atrium, minimizing the risk of postoperative complications and ensuring anatomical compatibility with patient growth.[2] Among patients with TAPVC who have undergone

Figure 2. Mixed Total Anomalous Pulmonary Venous Connection to the Superior Cavoatrial Junction.

CCTA 3D cinematic volumetric rendering reconstruction:

[**A**] Anterior view, [**B**] Lateral view.



CCTA: Cardiac Computed Tomography Angiography, IV: Innominate vein, LIPV: Left Inferior Pulmonary Vein, LSPV: Left Superior Pulmonary Vein, RA: Right Atrium, RIPV: Right Inferior Pulmonary Vein, RSPV: Right Superior Pulmonary Vein, SVC: Superior Vena Cava

surgery, the overall survival rate is 87.2%, with higher survival observed in children (94.1%) and in those with mixed-type TAPVC (96%).[4] Postoperative follow-up is essential and should include TTE within the first 30 days after surgical repair, followed by surveillance imaging at three to six months in asymptomatic patients with no or mild sequelae, and at one to two-year intervals thereafter in patients who remain clinically stable.[3]

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Disclosure

We confirm that there are no conflicts of interest or disclosures to report for all authors of this study. No financial support was received for this publication.

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