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Abnormal Drainage of Inferior Vena Cava to Left Atrium Together with a Partial Anomalous Pulmonary Venous Drainage to Right Atrium: A Case Report

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Abstract

Partial anomalous pulmonary venous connection (PAPVC) is a rare congenital anomaly often associated with sinus venosus atrial septal defect (SVASD). We report a unique case of a 4-year-old male presenting with dyspnea on exertion, diagnosed with right-sided PAPVC involving all three right pulmonary veins draining into the right atrium (RA), an inferior SVASD, and anomalous inferior vena cava (IVC) drainage into the left atrium (LA). Diagnosis was confirmed with echocardiography and CT imaging. Surgical correction involved rerouting the anomalous venous connections using a Dacron patch and augmenting the IVC-RA junction with a pericardial patch. The patient had an uneventful recovery. This case highlights the importance of precise imaging, thorough anatomical understanding, and tailored surgical techniques in managing rare and complex congenital heart defects.

Keywords: Partial Anomalous Pulmonary Venous Connection; Sinus Venosus Atrial Septal Defect; Inferior Vena Cava; Left Atrium

Abbreviations

PAPVC: Partial Anomalous Pulmonary Venous Connection; SVASD: Sinus Venosus Atrial Septal Defect; RA: Right Atrium; IVC: Inferior Vena Cava; LA: Left Atrium; ASD: Atrial Septal Defects; RIPV: Right Inferior Pulmonary Vein.

Introduction

Partial anomalous pulmonary venous connection (PAPVC) is a rare congenital heart defect where one or more pulmonary veins fail to follow their usual pathway to the left atrium (LA). Instead, they drain abnormally into the right atrium (RA) either directly or through systemic venous connections [1]. This misdirected blood flow can disrupt the heart's normal function and lead to long-term complications if left untreated. Another uncommon congenital heart condition, sinus venosus atrial septal defect (SVASD), occurs at the junction of the right atrium and vena cava. It represents about 5%–10% of all atrial septal defects (ASD) [2]. SVASD is frequently associated with PAPVC, as the abnormal anatomy of the septum and pulmonary veins often go hand-in-hand [3]. Together, these defects create unique challenges in both diagnosis and management. In this case report, we discuss

an especially rare and fascinating combination: PAPVC associated with SVASD, along with an anomalous drainage of the inferior vena cava (IVC) into the left atrium (LA). This unique arrangement defies the usual patterns of venous return and has only been documented in a handful of cases worldwide. Its rarity highlights the complexity of congenital heart defects and the critical importance of careful imaging and surgical planning to achieve the best outcomes for these patients.

Case Description

A 4-year-old boy presented with symptoms of breathlessness on exertion. His oxygen saturation on room air was 94%, and he exhibited no signs of cyanosis or clubbing. He had been diagnosed with congenital heart disease and was referred to our center for further evaluation and management. Preoperative echocardiography revealed a rare and complex anatomy. The diagnosis was consistent with right-sided partial anomalous pulmonary venous connection (PAPVC) and an inferior sinus venosus atrial septal defect (SVASD) measuring 2 × 1 cm. All three right lobar pulmonary veins were seen draining abnormally into the right atrium (RA) as separate openings (Figure 1A). During the echocardiographic evaluation, an additional unusual finding was identified: the inferior vena cava (IVC) was seen draining directly into the left atrium (LA) (Figure 1B). To confirm these findings and better delineate the anatomy, a CT scan was performed, which corroborated the echocardiographic findings (Figures 2A and 2B).

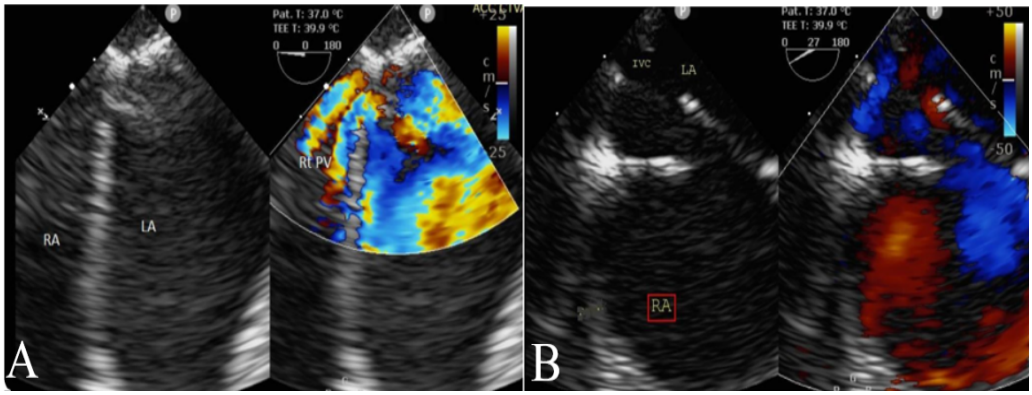

Figure 1: Trans-Esophageal Echocardiography. **A** Right Pulmonary Veins opening into RA. **B** dilated RA, with IVC opening to LA.

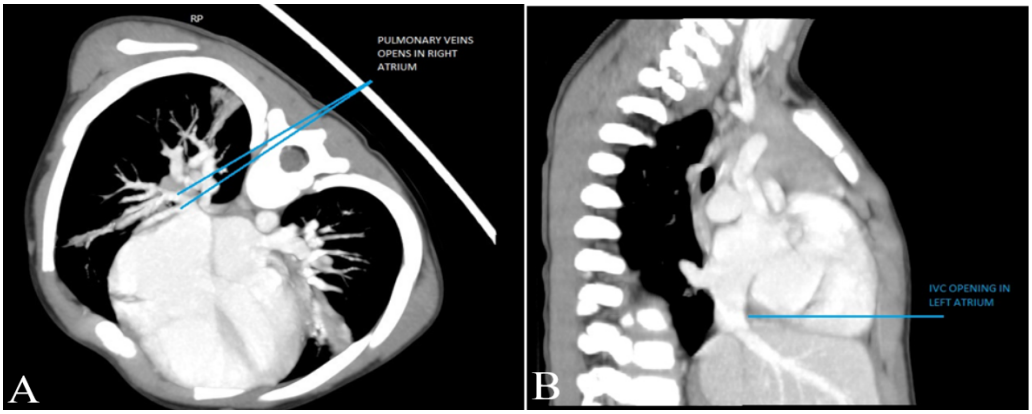

Figure 2: CT scan. **A** transverse section of CT scan showing right pulmonary veins opening into RA. **B** Sagittal section of CT scan showing IVC draining into LA.

The patient underwent surgical correction via a standard median sternotomy approach. After instituting cardiopulmonary bypass with aortobicaval cannulation, we snugged the IVC, which presented some technical difficulty due to its abnormal drainage pattern. Upon opening the significantly enlarged right atrium, a large atrial septectomy was performed, extending from the patent foramen ovale to the inferior SVASD, to provide better exposure to the anomalous pulmonary veins and the IVC. The intraoperative findings were consistent with the preoperative diagnosis. All three right pulmonary veins were clearly visualized draining into the right atrium (Figure 3A). The right inferior pulmonary vein (RIPV) was particularly notable, as it entered the RA near the IVC, just above the diaphragm. The IVC was

also confirmed to drain anomalously into the LA (Figure 3B).

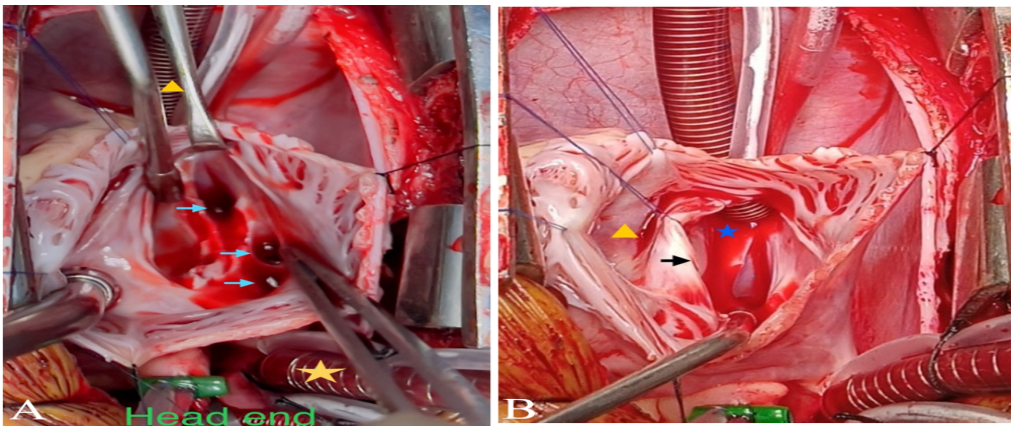

Figure 3: Intraoperative picture. **A** Three Right Pulmonary Veins opening into RA (light blue arrows), SVC (yellow star), IVC (yellow triangle). **B** IVC opening into LA (blue star), RA (yellow triangle), atrial septectomy edge (black arrow).

The surgical repair involved the use of a Dacron patch to close the defect, effectively rerouting the anomalous pulmonary veins to the LA and redirecting the IVC to the RA. To prevent any obstruction or gradient at the IVC-RA junction, we augmented the anterior surface of the IVC with a pericardial patch near the Eustachian valve. The procedure was completed smoothly, and the patient was successfully weaned off cardiopulmonary bypass without complications. This case highlights the complexity of congenital heart anomalies and the necessity of meticulous preoperative imaging and surgical planning to achieve optimal outcomes.

Discussion

Partial anomalous pulmonary venous connection (PAPVC) encompasses a spectrum of congenital heart defects in which one or more, but not all, pulmonary veins drain abnormally into the right atrium (RA). This can occur either directly into the RA or indirectly through systemic venous connections [4]. Among the various forms of PAPVC, the most common is where the left upper pulmonary vein drains into the left innominate vein, which eventually channels blood to the RA [4]. Other variations include pulmonary veins draining into the superior vena cava (SVC), coronary sinus, inferior vena cava (IVC), or azygos veins [4]. The physiologic consequence of PAPVC is similar to that of an atrial septal defect (ASD), as both involve a left-to-right shunt at the atrial level. This results in the recirculation of oxygen-rich blood through the pulmonary vasculature, leading to an increased volume load on the right heart [4]. The extent of the left-to-right shunting is influenced by several factors, including the number and size of the anomalous veins, their site of origin, the size of any associated ASD, and the pulmonary vascular resistance [4]. Although PAPVC is rare, the coexistence of PAPVC with anomalous IVC drainage into the left atrium (LA) represents an even more exceptional congenital anomaly. This rare combination has been documented in only a few reports [5-7]. During embryologic development, the right and left venous valves play a critical role in separating the systemic sinus venosus from the primary RA. The right venous valve contributes to the formation of the inferior border of the atrial septum, while the left venous valve typically fuses with the developing atrial septum [8].

In cases where the IVC abnormally drains into the LA, it is thought that an excessively large right venous valve, or eustachian valve, may coexist with an ASD. This developmental anomaly prevents the normal integration of the IVC into the RA, instead directing its flow to the LA. This, in turn, creates a complex anatomic and hemodynamic scenario that can be challenging to diagnose and treat [8]. In this case, the diagnosis and surgical management required a comprehensive understanding of these anomalies. The patient presented with PAPVC, where the right inferior pulmonary vein drained into the IVC-RA junction, coupled with the anomalous drainage of the IVC into the LA. The presence of an associated ASD further complicated the clinical picture. The surgical correction addressed these abnormalities by rerouting the anomalous pulmonary and systemic venous drainage to their appropriate chambers while ensuring that no obstruction or gradient occurred at the IVC-RA junction. This case underscores the importance of detailed imaging, a thorough understanding of embryologic development, and meticulous surgical planning in managing such rare and intricate congenital heart defects.

Conclusion

This case illustrates a rare combination of PAPVC, SVASD, and anomalous IVC drainage into the LA, highlighting

the complexity of such congenital anomalies. Accurate diagnosis through detailed imaging and precise surgical planning were key to achieving a successful outcome. Surgical correction involved rerouting the anomalous venous connections and optimizing the IVC-RA junction, ensuring normal hemodynamic. This case emphasizes the importance of a thorough understanding of cardiac anatomy and individualized approaches in managing rare congenital heart defects.

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