

Surgical correction of partial anomalous pulmonary venous return in a patient with persistent left superior vena cava: cannulation techniques and challenges

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SÚHRN

Parciálny anomálny návrat plúcnych žíl asociovaný so sínusovým venóznym defektom predsieňového septa a perzistujúcou ľavou hornou dutou žilou predstavuje významnú chirurgickú výzvu. Tieto vrodené anomálie si vyžadujú dôkladné plánovanie mimotelového obehu a stratégií venózne kanulácie.

Tridsaťosemročná pacientka sa prezentovala s anamnézou supraventrikulárnej tachykardie a hemodynamicky významným ľavo-pravým skratom ($Q_p : Q_s = 1,87$) v dôsledku sínusového venózneho predsieňového septa a parciálny anomálny návrat plúcnych žíl. Predoperačné zobrazovacie vyšetrenia preukázali drenáž pravej hornej plúcnej žily do perzistujúcej ľavej hornej dutej žily. Pacientke bola úspešne vykonaná chirurgická korekcia metódou dvojitej náplasti s podporou mimotelového obehu, pričom sa využila komplexná stratégia kanulácie oboch horných dutých žíl. V pooperačnom období sa pacientka zotavila bez známok reziduálneho skratu alebo významnej chlopňovej dysfunkcie.

Případ poukazuje na chirurgický prístup pri zriedkavej kombinácii vrodených anomálií a diskutuje alternatívne možnosti kanulácie pri mimotelovom obehu u pacientov s perzistujúcou ľavou hornou dutou žilou.

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ABSTRACT

Partial anomalous pulmonary venous return (PAPVR) associated with a sinus venosus atrial septal defect (SVASD) and a persistent left superior vena cava (PLSVC) presents significant surgical challenges. These anomalies require careful planning of cardiopulmonary bypass (CPB) and venous cannulation strategies.

A 38-year-old woman presented with a history of supraventricular tachycardia (SVT) and a hemodynamically significant left-to-right shunt ($Q_p : Q_s = 1.87$) due to SVASD and PAPVR. Preoperative imaging revealed drainage of the right upper pulmonary vein into the PLSVC. The patient underwent successful surgical correction using the double-patch technique with cardiopulmonary bypass support via a complex cannulation strategy involving both superior venae cavae. Postoperatively, the patient recovered well, with no evidence of residual shunt or significant valvular dysfunction.

The case highlights the surgical approach to a rare combination of anomalies and discusses alternative cannulation strategies for CPB in patients with PLSVC.

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Introduction

Sinus venosus atrial septal defect (SVASD) with partial anomalous pulmonary venous return (PAPVR) represents a rare congenital anomaly with significant implications for cardiac function. Its prevalence is up to 0.5% of the general population and 3% to 10% in patients with congenital heart disease.^{1,2} These defects are often accompanied by additional venous anomalies, such as a persistent left superior vena cava (PLSVC), complicating the surgical approach. The double-patch technique, a well-established method for addressing PAPVR, becomes particularly relevant in such scenarios.³ This case report describes the surgical management of a patient with these anomalies, emphasizing the challenges in cardiopulmonary bypass (CPB) setup and highlighting alternative venous cannulation options.

Case presentation

A 38-year-old woman presented with recurrent episodes of supraventricular tachycardia (SVT), diagnosed as atrioventricular nodal reentrant tachycardia (AVNRT) in 2005 and 2023. She had no significant comorbidities but reported exertional dyspnea and occasional chest pressure. Transthoracic echocardiography (TTE), transesophageal echocardiography (TEE), and cardiac CT confirmed a sinus venosus defect (20 × 15 mm) with associated PAPVR. Imaging revealed anomalous drainage of the right upper pulmonary vein into the right superior vena cava (RSVC) and partial drainage into the left atrium (LA). Additionally, a persistent left superior vena cava (PLSVC) was identified, draining into the coronary sinus (Fig. 1). Hemodynamic studies showed a left-to-right shunt with $Q_p : Q_s = 1.87$, with preserved systolic function and no pulmonary hypertension. On October 31, 2024, the patient underwent surgical correction. Cannulation for CPB was performed using separate cannulas for the RSVC and PLSVC

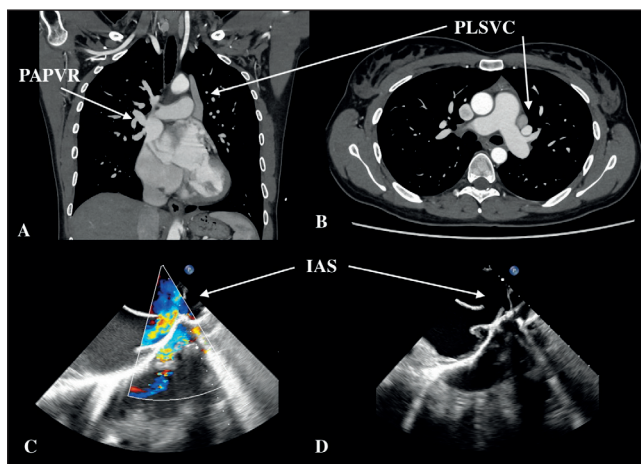


Fig. 1 – (A) Computed tomography coronal view showing the partial anomalous pulmonary venous return. (B) Computed tomography axial view showing the persistent left vena cava superior. (C, D) Transesophageal images showing the interatrial septum. IAS – interatrial septum; PAPVR – partial anomalous pulmonary venous return; PLSVC – persistent left vena cava superior.

to ensure effective drainage, and antegrade cardioplegia was administered. A Foley catheter was temporarily placed into the coronary sinus for PLSVC drainage. The double-patch technique was employed to redirect the anomalous pulmonary venous flow to the left atrium. The sinus venosus defect was closed with a pericardial patch. Intraoperative transesophageal echocardiography confirmed no residual shunt and unobstructed flow through the reconstructed venous pathways.

The patient experienced an uneventful recovery. Echocardiography prior to discharge showed preserved left and right ventricular function, absence of residual atrial septal defects, and no significant valvular dysfunction. She was discharged on the ninth postoperative day in a stable condition.

Discussion

SVASD with PAPVR and PLSVC represents a unique surgical challenge due to the anatomical complexity of venous drainage and potential difficulties in establishing adequate venous return during CPB. The presence of a PLSVC often necessitates modifications to the standard cannulation strategy, as the anomalous vessel may impede venous drainage if not properly addressed.

PLSVC is the most common venous anomaly of the thoracic circulatory system, with an incidence of 0.5% of the general population, and in 90% of the cases it is drained into the coronary sinus, like our patient. These patients remain asymptomatic and the diagnosis is mostly an incidental finding. Only in 10% of the patients it is drained into the left atrium, resulting in a right-to-left shunt, with systemic hypoxemia and an increased risk of paradox embolization.⁴ Its presence requires some consideration in the management of myocardial protection and intraoperative conduct. It is a contraindication to retrograde cardioplegia, since the flow may result in cardioplegia wash-out. In order to overcome the aforementioned problems, PLSVC can be momentarily occluded in case the anomalous vein is present. However, even antegrade cardioplegia may fail, due to the steal effect by the hemiazygos venous system linked to PLSVC.⁵ In our case, antegrade cardioplegia was used without any complications.

It is more common in patients undergoing surgical procedures congenital heart disease, and if it is suspected, computed tomography (CT), magnetic resonance imaging (MRI), venous angiography or a digital subtraction venogram can be performed for the correct diagnosis.⁶ Moreover, TTE and TEE can be performed for the diagnosis, but sometimes these studies do not necessarily reveal a PLSVC that drains to the left atrium unless a bubble study with agitated saline is performed. In some cases where the PLSVC is identified intraoperatively, the cannulation technique should be reconsidered and managed accordingly.⁴

In our case report, the PLSVC was identified preoperatively by CT so the cannulation strategy and planning were decided before the operation.

Different cannulation techniques for the PLSVC are proposed. Direct cannulation of the PLSVC is an effective method and maybe feasible when properly visualized in-

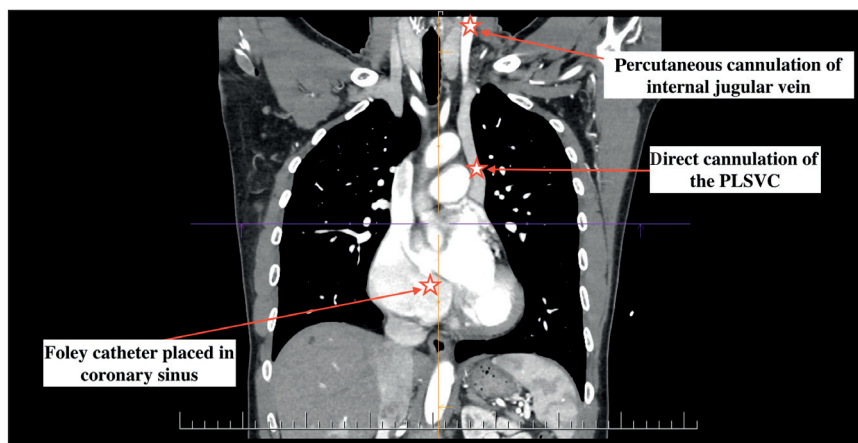


Fig. 2 – Proposed cannulation techniques.

side the pericardium. Based in our experience, we propose using Foley catheter placed in coronary sinus to be used for venous drainage during CPB. This method is simple, can be performed easily without any complications, and can provide adequate drainage of the PLSVC. Another method as proposed by G. Bianchi et al⁵ is a hybrid cannulation by direct percutaneous cannulation of internal jugular vein. The proposed cannulation techniques are shown in **Figure 2**.

The surgical correction using the double-patch technique is safe approach for addressing PAPVR with associated SVASD. This technique allows effective redirection of anomalous pulmonary venous flow to the left atrium while preserving normal venous pathways. The main advantage of the two-patch technique is the lower incidence of SVC stenosis on long-term follow-up, however, as many as 55% may have sinus node dysfunction while incising the SVC anteriorly. This may be directly related to the sinus node itself, as well as its supply artery.⁷ The patient's postoperative rhythm disturbances, including junctional rhythm, were transient and resolved with supportive care. This highlights the importance of vigilant postoperative monitoring, particularly in patients with preoperative arrhythmias.

Conclusion

This case underscores the importance of individualized surgical planning for complex congenital cardiac anomalies. The successful use of the double-patch technique, combined with a tailored cannulation strategy, ensured optimal outcomes. Awareness of alternative CPB cannulation approaches is crucial for addressing anatomical variations, such as PLSVC, and minimizing intraoperative complications.

Conflict of interest

The authors declare no conflict of interest.

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Ethical statement

This case report was conducted in accordance with the principles of the Declaration of Helsinki. The authors affirm that all methods were carried out in accordance with relevant guidelines and regulations.

Informed consent

The patient provided an informed consent to present this case report.

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