

UNMASKING AN ANOMALOUS VEIN: SURGICAL REPAIR OF LEFT UPPER PULMONARY VEIN DRAINAGE INTO THE INNOMINATE VEIN

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Abstract

Partial anomalous pulmonary venous return (PAPVR) is a rare congenital anomaly characterized by one or more pulmonary veins draining into the systemic venous system. We present the case of a 51-year-old woman with PAPVR involving the left upper pulmonary vein (LUPV) draining into the left innominate vein via a vertical vein, diagnosed after investigation of progressive dyspnea and lower limb edema. The diagnosis was confirmed through contrast-enhanced CT and echocardiography. The patient underwent surgical correction of the anomalous return along with tricuspid valve repair. Postoperative recovery was uneventful. This case highlights the importance of recognizing PAPVR in adult patients presenting with unexplained right heart dilation or symptoms of right heart overload.

Keywords: PAPVR, anomalous pulmonary venous drainage, vertical vein, tricuspid repair, adult congenital heart disease

INTRODUCTION

Partial anomalous pulmonary venous return (PAPVR) is a rare congenital cardiovascular defect found in approximately 0.4–0.7% of the population.¹ It results from an abnormal drainage of one or more pulmonary veins into the systemic venous circulation instead of the left atrium. While right-sided PAPVR involving the superior vena cava is more common, left-sided forms are less frequently reported, particularly those involving the left upper pulmonary vein (LUPV) draining into the left innominate vein via a vertical vein.² These anomalies are often asymptomatic but may lead to right heart dilation, pulmonary hypertension, or symptoms of heart failure depending on the magnitude of the left-to-right shunt.^{1,2}

CLINICAL CASE

A 51-year-old woman was referred for evaluation due to a one-year history of progressive fatigue, bilateral lower limb edema, palpitations, and episodes of chest tightness both at rest and during exertion. She also reported frequent dizziness but denied syncope or claudication. Her past medical history

included hypertension, dyslipidemia, irritable bowel syndrome, ankylosing spondylitis, recurrent urinary tract infections, renal lithiasis, and migraine. She had undergone previous cervical spine, nasal septum, and maxillofacial surgeries.

On physical examination, her vital signs were stable. Electrocardiogram showed sinus rhythm. Transthoracic echocardiography revealed right ventricular (RV) dilation with preserved systolic function and moderate functional tricuspid regurgitation associated with annular dilation. Estimated pulmonary artery systolic pressure was 30 mmHg. Left ventricular (LV) size and function were normal.

Contrast-enhanced cardiac CT confirmed partial anomalous pulmonary venous return: the LUPV drained into the left innominate vein via a vertical vein (Figure 1). No other congenital anomalies or coronary artery disease were identified.

The patient underwent surgical correction of the PAPVR. Median sternotomy under general anesthesia and conventional cardiopulmonary bypass was carried. The vertical vein draining the left upper pulmonary vein into the left innominate vein was identified, mobilized along its course and divided from the innominate vein, preserving adequate length to allow a tension-free anastomosis. (Figure 2). Attention was

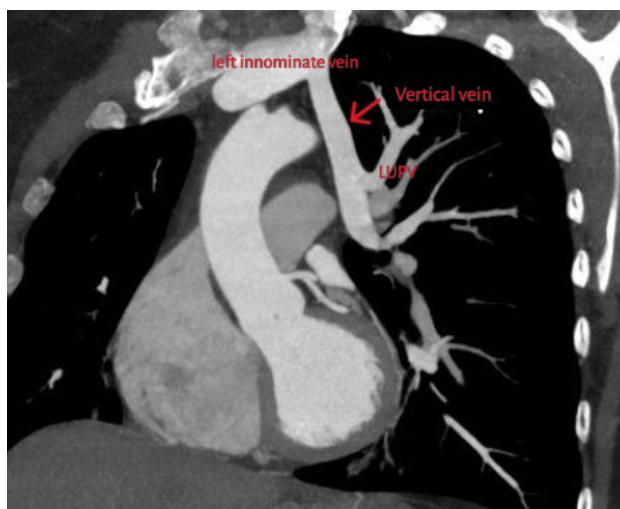


Figure 1

CT angiogram showing partial anomalous pulmonary venous return: LUPV draining into the left innominate vein via a vertical vein.

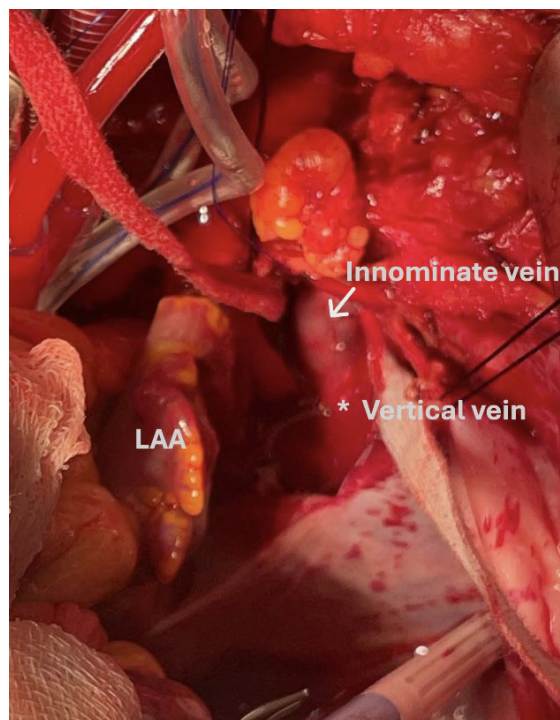


Figure 2

LUPV draining into innominate vein via vertical identified.

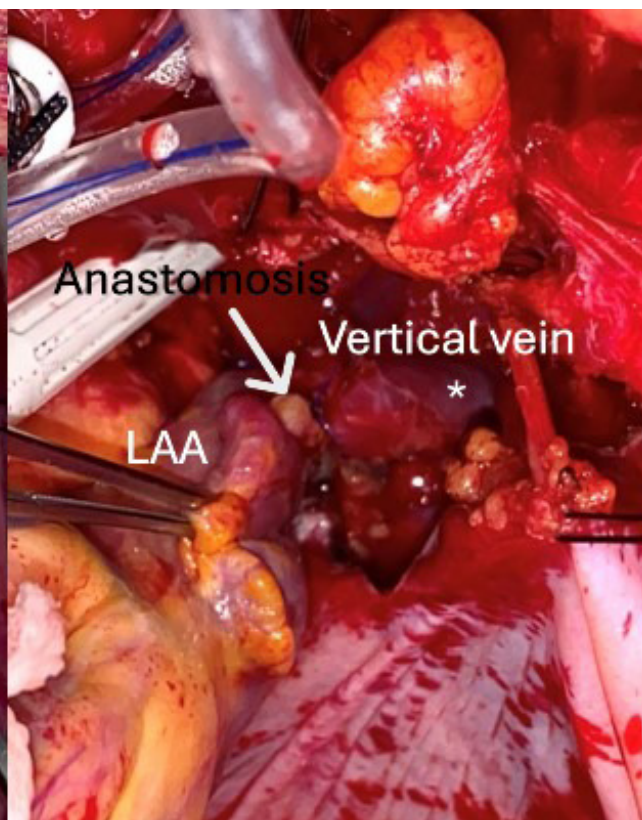
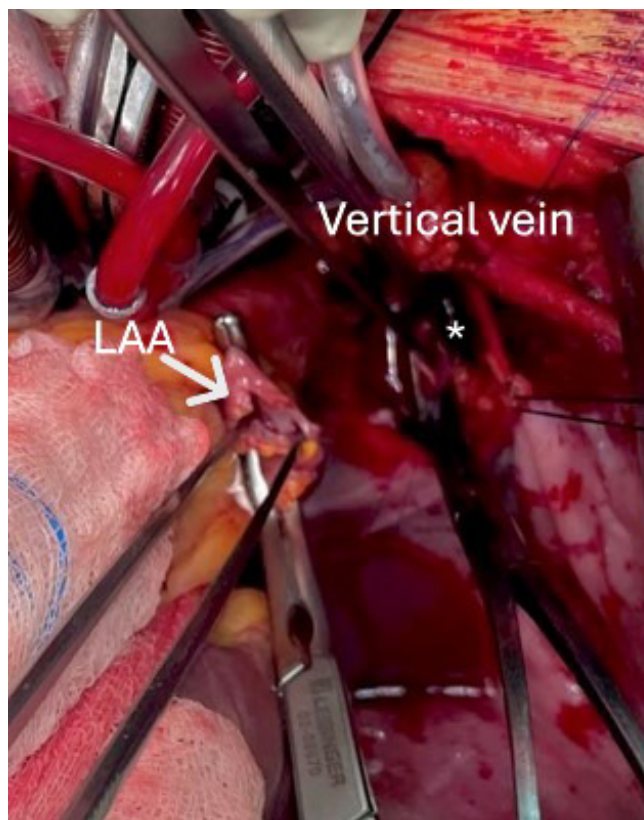


Figure 3

LUPV divided and prepared for anastomosis; LAA – Vertical vein anastomosis completed, restoring normal pulmonary venous return.

then directed to the left atrial appendage (LAA). A longitudinal atriotomy was performed along the body of the LAA to create an opening tailored to the diameter of the vertical vein, with care taken to minimise atrial manipulation and avoid unnecessary traction or cautery near atrial conduction tissue, thereby reducing the risk of postoperative arrhythmias. The vertical vein was opened with a corresponding longitudinal venotomy, and an end-to-side "cobra-head" anastomosis was fashioned between the vein and the LAA. The posterior wall was sutured first using a continuous 5-0 Prolene technique, ensuring precise alignment and a smooth, funnel-shaped transition characteristic of the cobra-head configuration. The anterior wall was subsequently completed with the same suture, taking care to avoid tension, twisting, or narrowing of the anastomosis (Figure 3). Concomitantly, tricuspid valve repair was performed using a Carpentier-Edwards Physio® ring (size 30) to address moderate functional tricuspid regurgitation secondary to annular dilation. The procedure was completed without complications, and the patient was weaned from bypass in sinus rhythm with stable hemodynamics. Postoperative recovery was uncomplicated. She was extubated on the first postoperative day, with hemodynamic stability and preserved renal function throughout the ICU stay.

A follow-up echocardiogram showed trivial residual tricuspid regurgitation without stenosis, a pulmonary artery systolic pressure of 23 mmHg, and preserved left ventricular function. Longitudinal systolic RV function was mildly reduced, but radial function was preserved. Patient was discharged on 7th day post-op. At 8 months of clinical follow-up, the patient remains asymptomatic and in stable sinus rhythm, with good functional status and no reported limitations in daily activities. No clinical features suggestive of arrhythmias, heart failure, or recurrent right-sided volume overload were noted during outpatient evaluation.

DISCUSSION

PAPVR involving the LUPV and the left innominate vein is an uncommon variant and may go undetected until adulthood, particularly in the absence of significant shunting. The presence of symptoms such as fatigue, peripheral edema, or signs of right heart strain should prompt further investigation.³ In the case presented, the patient exhibited progressive fatigue, lower limb edema, and palpitations, correlating with right ventricular dilation and moderate tricuspid regurgitation observed on echocardiography.

Advanced imaging modalities, such as contrast-enhanced computed tomography (CT) and magnetic resonance imaging (MRI), play a crucial role in accurately delineating the anomalous pulmonary venous anatomy. These techniques provide detailed visualization, facilitating precise surgical planning.⁴

Surgical intervention is generally recommended for symptomatic patients or those with evidence of right heart dilation and significant shunting. The standard surgical approach involves rerouting the anomalous pulmonary vein to the left atrium and ligating the anomalous connection to prevent ongoing left-to-right shunt.⁵ In this case, the patient underwent successful surgical correction with rerouting of vertical vein to the left atrium and concomitant tricuspid valve repair, resulting in symptomatic improvement and normalization of right heart dimensions.

Long-term outcomes following surgical correction of PAPVR are generally favourable, with most patients experiencing symptom relief and improved cardiac function. However, these results typically rely on extended follow-up, which represents a limitation in the present report, as only 8 months of clinical surveillance are currently available. Although the short-term evolution in this case has been uneventful, regular and lifelong follow-up remains essential to monitor for potential late complications, including pulmonary vein stenosis, thrombosis, arrhythmias, or the need for reintervention.⁵

This case underscores the importance of considering PAPVR in the differential diagnosis of adults presenting with unexplained right heart enlargement or symptoms suggestive of right heart overload. Early recognition and appropriate surgical intervention can lead to excellent outcomes, even in adult patients.

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