

# Isolated Partial Anomalous Pulmonary Venous Connection of the Left Lung

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## ABSTRACT

Isolated partial anomalous pulmonary venous connection (PAPVC) of the entire left lung is a rare congenital anomaly with incidental diagnosis and vague symptoms, if any, until late adulthood. If left untreated, PAPVC may result in severe right ventricular failure and pulmonary vascular disease. We present the case of a 34-year-old woman with isolated PAPVC of the entire left lung. The patient underwent operation with a side-to-side left atrio-vertical vein anastomosis while on cardiopulmonary bypass and under cardioplegic arrest. She was discharged without complications and with a gradient of 2 mm Hg across the anastomosis. Left-sided PAPVC can be repaired with minimal morbidity and mortality. Surgical correction is warranted when patients are symptomatic or show evidence of right-sided overload due to unpredictability of the natural course. Recent data demonstrate that both on-pump and off-pump surgical procedures produce excellent long-term outcomes when performed without persisting gradients.

## INTRODUCTION

Partial anomalous pulmonary venous connection (PAPVC) is a rare congenital anomaly, with left-sided PAPVCs occurring in 10% to 18.2% of all reported PAPVC cases [Ammash 1997; ElBardissi 2008]. Because of the subtlety of the symptoms and the difficulty of diagnosis, particularly in nonreferral centers, the condition may remain undiagnosed until late adulthood if an associated atrial septal defect does not exist. We present the case of a 34-year-old woman with persistent fatigability, palpitations, and dyspnea that rural medical providers had followed without diagnosis for a long time. The diagnosis was isolated PAPVC of the entire left lung without an intracardiac shunt.

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## CASE REPORT

A 34-year-old woman with persistent fatigability, palpitations, and dyspnea was referred to our surgical clinic for isolated PAPVC of the entire left lung. Primary and secondary medical providers followed the patient's symptoms for 4 years despite consecutive echocardiographic examinations. A definitive diagnosis was made from a final echocardiography evaluation that demonstrated the left-sided vertical vein draining all of left pulmonary venous blood to the innominate vein, with consequent enlargement of the right heart chambers. Cardiac catheterization confirmed this diagnosis by demonstrating the anomalous pulmonary connection (Figure 1). The patient was scheduled for surgical correction.

A conventional median sternotomy was performed prior to establishment of cardiopulmonary bypass with mild hypothermia. The surgical exposure of the left-sided vertical vein (Figure 2A) was followed by a short period of cardioplegic arrest. The left pulmonary artery was temporarily occluded with elastic tape. A fine vascular clamp was placed just below the insertion of the vertical vein into the innominate vein, and a long incision (approximately 15 mm) was made along the vertical vein over its fully distended diameter. A corresponding incision was made on the left atrial wall at the base of the left atrial appendage, the tip of which was opened.

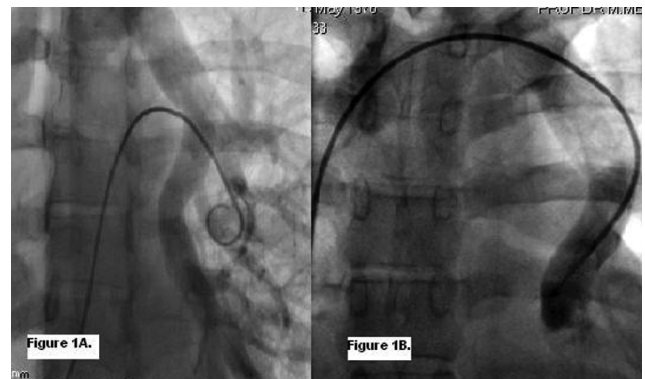


Figure 1. Catheterization demonstrating the left-sided vertical vein draining all of the blood from the left pulmonary vein to the innominate vein.

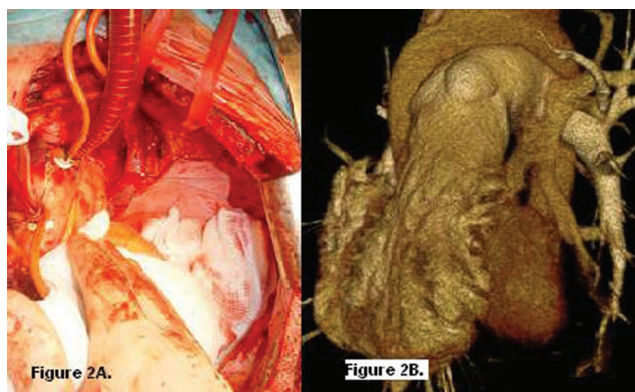


Figure 2. Intraoperative appearance through the median sternotomy. A, Note the left-sided vertical vein lateral to the pulmonary artery. B, 3D reconstruction of the atrio-vertical vein anastomosis. Note that the upper- and the lower-lobe veins connect to form a common vertical vein.

The trabeculations were then trimmed for a broad-based side-to-side anastomosis. The vertical vein was transected and the vertical vein–innominate vein confluence was repaired with 5-0 Prolene suture. Proximal clamping of the vertical vein draining the entire left lung was avoided, and an open anastomosis was possible. Following completion of the anastomosis, air was aspirated with a fine needle, and the clamps to the left atrial appendage and the left pulmonary artery were removed in sequence. The trans-anastomotic pressure gradient as recorded with direct-monitoring lines was 2 mm Hg.

The patient was extubated at the second postoperative hour and discharged to the ward on day 1. The postoperative course was uneventful, and she was discharged on Coumadin on day 6 without complications. After 6 months of follow-up, the patient was well and free of complications. A contrast-enhanced computed tomography scan demonstrated a patent anastomosis with an appropriate configuration.

## DISCUSSION

Isolated PAPVC of the entire left lung is a rare congenital anomaly with an incidental diagnosis and vague symptoms, if any, until late adulthood. Although most patients with PAPVC are asymptomatic, the natural history dictates that patients may develop irreversible pulmonary hypertension, pulmonary vascular obstructive disease, or right ventricular failure if a significant left-to-right shunt exists. Previously, the indication for surgical treatment of PAPVC mimicked that of atrial septal defects, with treatment recommended for patients with a ratio of pulmonary flow to systemic flow ( $Q_p/Q_s$ ) of  $>1.5$ ; however, recent data have suggested early correction irrespective of the  $Q_p/Q_s$  value or when an atrial septal defect is present [ElBardissi 2008]. Our patient had a  $Q_p/Q_s$  value of 1.8 and was evidently symptomatic with enlarged right cardiac chambers and mild to moderate tricuspid valve regurgitation.

Very few data regarding this anomaly have been published.

Until the very recently published series, the single largest reported institutional experience with this anomaly consisted of 13 patients, only 6 of which had PAPVC of the entire left lung [Van Meter 1990]. Both series suggested the use of an anterolateral thoracotomy approach for direct anastomosis without cardiopulmonary bypass. A conversion from an “on-pump and median sternotomy” approach to an “off-pump and thoracotomy” approach was noted in the former series unless an intracardiac procedure was anticipated. Other case reports have described a similar strategy of a median sternotomy and an on-pump approach [Ban 1987; Mukadam 1995].

The importance of a gradient-free anastomosis should be emphasized, because most of the perioperative morbidity as arrhythmias, pulmonary venous obstruction, or recurrent ipsilateral pneumonia is attributed to gradients greater than 4 mm Hg. Postoperative administration of Coumadin has been suggested to avoid any thromboembolic phenomena in and around the anastomotic area. These investigators reported the presence of echocardiographic opacities, although no obstructions were noted [ElBardissi 2008]. In the light of these investigators’ findings, we continue to follow our patient on Coumadin with an international normalized ratio of 1.8 to 2.0 and have encountered no complications. As ElBardissi et al have noted, the presence of foreign material, such as suture, and a low-pressure/low-flow state in the appendix may surrogate for thrombosis in the left atrium.

Left-sided PAPVC can be repaired with minimal morbidity and mortality when the operation is performed correctly. Because the natural history of this disorder includes a wide variety of complications, including arrhythmias and pulmonary obstructive disease, we recommend that surgical correction be performed when patients are symptomatic or show evidence of right-sided overload. Recent data demonstrate that both on-pump and off-pump surgical procedures produce excellent long-term outcomes when surgery is performed without persisting gradients.

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