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# Better Late Than Never – A Case of a Congenital Left Partial Anomalous Pulmonary Venous Drainage Diagnosed and Treated in The Sixth Decade of Life

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## ABSTRACT

Partial anomalous pulmonary venous drainage (PAPVD) is a relatively uncommon cardiac anomaly. The diagnosis might be challenging as are the presenting symptoms. Its clinical course mimics more familiar diseases, e.g., pulmonary artery embolism. We present a case of PAPVD, which had been misdiagnosed for more than two decades. After establishing the correct diagnosis, the patient got his congenital anomaly surgically corrected and showed excellent cardiac recovery in the six months follow up.

## INTRODUCTION

**Background**: Pulmonary vein anomalies are rare, with an incidence of 0.4-0.7% [Adler 1973]. This anomaly is more commonly right-sided (90%) [Haramati 2003]. The incidence of left upper lobe partial anomalous pulmonary venous drainage (PAPVD) is occasionally accompanied by cardiac anomalies, such as mitral stenosis, pulmonic stenosis, and a patent ductus arteriosus [Pennes 1986]. No conclusive statistics exist to determine if PAPVD is more prevalent in men or women. There are no identified risk factors connected with its development. It is plausible that PAPVD is either overlooked or misdiagnosed, given the scarcity of research on the subject.

In a complete left PAPVD (left upper and lower pulmonary veins), the deviant vein returns oxygenated blood from the left lung to the left innominate vein. (Figure 1) This then empties into the normally located superior vena cava. Consequently, this circulation pattern may significantly alter the pulmonary to systemic blood flow; this alteration eventually will cause a right ventricle volume overload and a rise in pulmonary pressure. Ultimately, the patient may develop right-sided heart failure. Due to the absence of a systemic supply of oxygenated blood from the left lung, the right lung primarily is responsible for peripheral gas exchange.

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## CASE PRESENTATION

A 61-year-old Caucasian male (1.75m, 98kg, BSA 2.13m², BMI 31.9) presented to the emergency department (ED) in a tertiary healthcare facility complaining of worsening dyspnea (NYHA III). He markedly gained weight just two weeks prior, despite loss of appetite. He sought professional medical advice in a specialized cardiac center after unsatisfactory results of his prior treatment and deterioration of his condition, even though he strictly adhered to his medical therapy.

He was a chronic active smoker (15 pack years) and reported previous treatment for dyslipidemia.

On presentation, blood pressure was 100/96 mmHg, heart rate 109 beats per minute, respiratory rate 21 per minute, temperature 37 degrees Celsius using an ear thermometer, and oxygen saturation of 88% on ambient air. The central venous blood gas analysis was unremarkable except for increased oxygen saturation (Spo2 86%). Physical examination was remarkable for a systolic murmur. Lung auscultation revealed bibasilar crackles. Neck veins were distended with canon systolic wave, and massive lower extremity edema was detected in both lower limbs.

The patient signed informed consent related to the clinical course; therefore, the Institutional Review Board was waived due to the retrospective nature of the educational case report.

**Investigations**: Initial laboratory test results were remarkable for an elevated red blood cell count of 6.9 cells/mcL

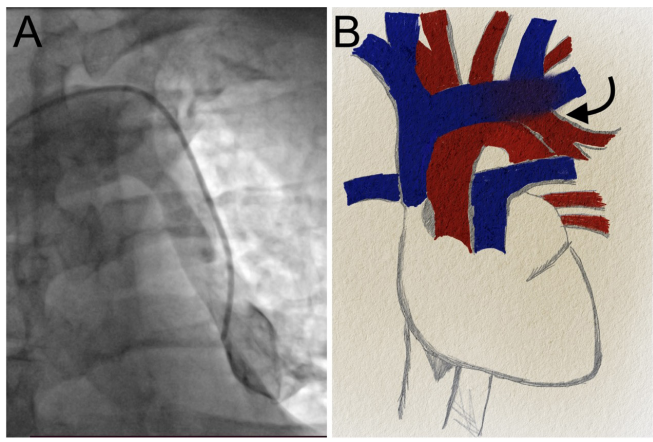


Figure 1. (A) catheterization of the left pulmonary vein through the left brachiocephalic vein with an injection of contrast dye, (B) schematic representation of the anomaly. "Black arrow" is pointed toward the deviant courses of the left pulmonary vein.

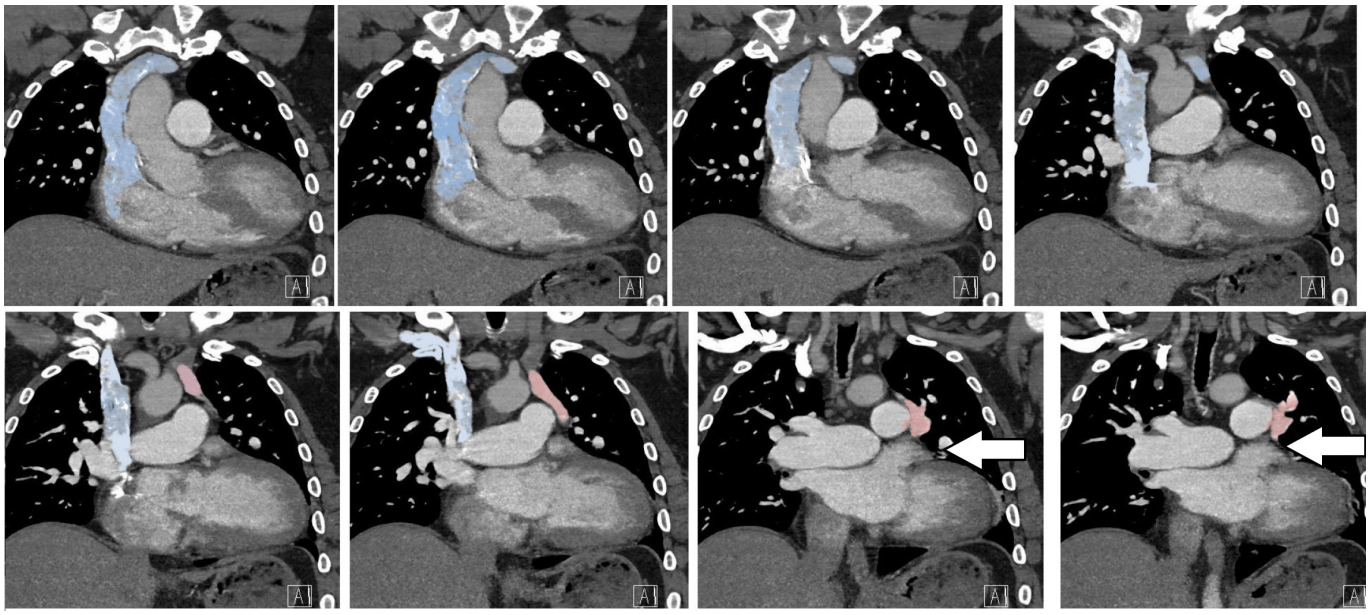

Figure 2. Coronal CT angiography sequential series showing the course of the brachiocephalic vein joining with the superior vena cava (blue) and the anomalous drainage of the left pulmonary vein into the brachiocephalic vein (red). Note: The arrow shows the lack of pulmonary veins entering the left atrium.

(from 4.7 to 6.1 million cells per microliter) concomitant with high serum hemoglobin of 18.5 g (from 14 to18 grams), revels a polycythemia vera secondary to the chronic hypoxia, and renal function and liver function tests were within normal limits. The electrocardiogram showed atrial fibrillation with a right bundle branch block. Transthoracic echocardiography revealed a left ventricle ejection fraction (LVEF) of 65% and a dilated right ventricle with a fractional area change (FAC) of 30% with a right atrial enlargement and severe tricuspid valve insufficiency.

A contrast enhancement chest CT demonstrated in the venous phase that the left pulmonary veins presented an anomalous route: It first ran under the pulmonary artery, then went up medially to it, running via the aortopulmonary window, and finally joined the left brachiocephalic vein. The remaining pulmonary veins and cardiovascular structures were normal in appearance and connection. (Figure 2)

A right heart catheterization was done to quantify the shunt and objectively measure the pulmonary pressure. The patient was noted to have a left to right shunt with a QP: Qs ratio of 1.5:1, as well as a mildly elevated pulmonary pressure with systolic ($P_s$), mean ($P_{mean}$), and diastolic ($P_d$) pulmonary pressures of 35, 22, and18 mmHg, respectively (Figure 1).

The left-sided cardiac catheterization showed no evidence of obstructive coronary artery disease. Consequently, the patient was transferred to our department for a surgical correction of his anomaly.

**Management**: After induction of general anesthesia, the patient was prepped and draped in usual sterile fashion. After the median sternotomy, the patient was heparinized by checking activated clotting time. Aortic and right atrial cannulation was installed, and a cardiopulmonary bypass was initiated for 58 minutes. The aortic cross-clamp was then applied for

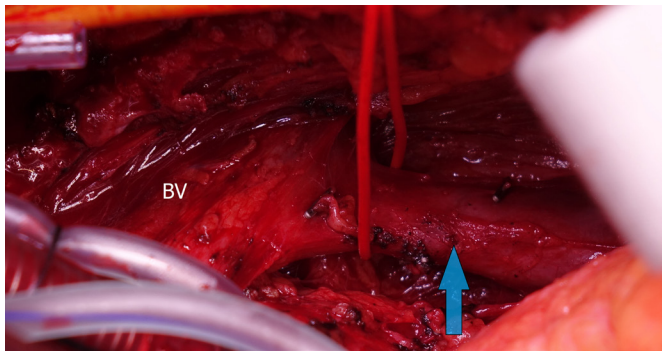


Figure 3. Intraoperative anatomy showing the anomalous drainage of the left pulmonary vein (arrow) into the brachiocephalic vein (BV).

39 minutes. A normothermic blood cardioplegia was administrated through the aortic root, achieving a satisfactory diastolic arrest. The left pulmonary vein was clamped and resected; a pericardial patch was used to anastomose the left pulmonary vein into the left atrium wall. (Figure 3) (Figure 4) (Figure 5) The aortic root was vented. The heart was deaired, and the aortic cross-clamp was removed. The weaning off the cardiopulmonary bypass was routinely performed after the spontaneous return of the heartbeats. Protaminized decannulation was done, and hemostasis was secured. The chest was closed in layers. The patient tolerated all the following procedures well and was monitored in an intensive care unit for 24 hours and then shifted to the surgical ward later for other postoperative care, which was uneventful.

**Follow up**: In the postoperative follow-up transthoracic echocardiography, the patient had mild tricuspid valve regurgitation with preserved ejection fraction. The patient

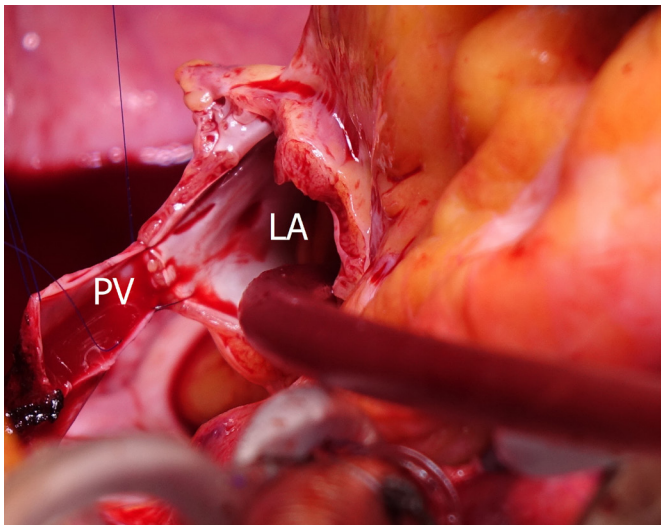


Figure 4. Anastomosis of the left pulmonary vein (PV) to the left atrium (LA).

was discharged free from symptoms to an inpatient rehabilitation facility.

In 6 months follow up after surgery, the patient showed freedom from hospitalization for heart failure. And his dyspnea symptoms revert to NYHA I. His peripheral edema regressed dramatically, and he lost about 9 kilograms from his weight.

## DISCUSSION

Partial anomalous pulmonary venous drainage (PAPVD) is a congenital anomaly in which one or more (but not all) pulmonary veins drain directly or indirectly into the right atrium.

Embryogenic development of the pulmonary veins occurs in the first two months of fetal development. The prevailing theory is that initial drainage is via the splanchnic plexus into the cardinal and umbilicovitelline veins, a craniocaudal outpouching forms in the sinoatrial region of the heart with extension to the lung buds. With caudal regression, the cranial portion develops into the common pulmonary vein, which then incorporates into the left atrial wall. The partial anomalous pulmonary venous return occurs, due to the failure of connection between the common pulmonary vein and the splanchnic plexus [Kao 2017]. When occurring in the left lung, a vertical vein is seen collecting oxygenated blood from the upper and lower left lobe and then rains into the brachiocephalic vein.

Right ventricular dysfunction, pulmonary hypertension, tricuspid regurgitation, and volume overload all are potential consequences of this disorder. PAPVD forms left to right shunt, often silent until adulthood [Edwin 2010]. Our patient started to develop symptoms two decades prior. Unfortunately, his condition was misdiagnosed through his primary health care facility as multiple deep venous thrombi, which

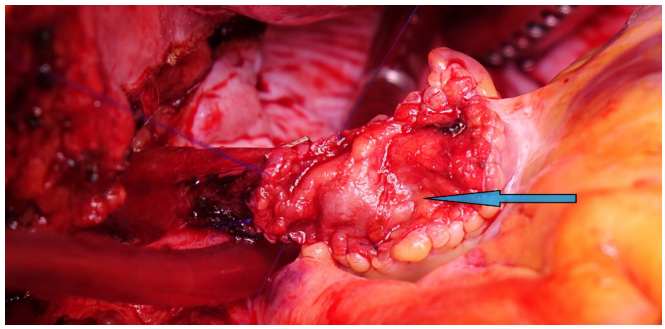

Figure 5. Completed anastomosis of the left pulmonary vein to the left atrium. The arrow shows the use of a glutaraldehyde-fixed autologous pericardial patch on the superior aspect of the anastomosis to prevent stenosis/kinking of the vessel.

led to subsequent pulmonary hypertension and right-sided ventricle failure; due to poor response to treatment and worsening of his symptoms, he sought medical advice in a tertiary health care institute that could detect the anomaly.

In echocardiography, PAPVD may be suspected. However, a definitive diagnosis is challenging, due to a lack of suitable acoustic windows. CT and MRI are the preferred diagnostic modalities for identifying and characterizing congenital pulmonary vein abnormalities [Tricarico 2016].

Regarding the management of the PAPVD, there are two widely accepted opinions. The first is to execute a repair prior to the onset of symptoms. The second is to provide surgical corrections only when symptoms manifest as surgical hazards, including atrial fibrillation, total heart block, cardiac arrest, and pulmonary venous blockage; surgical repair often is advised if there is evidence of mild to severe tricuspid valve regurgitation or right ventricular dilatation, or hypertensive pulmonary vascular disease. Morbidity and mortality are reported low with surgical repair as the operative complications are less likely with improved techniques. The surgical procedure can theoretically be performed with or without cardiopulmonary bypass [ElBardissi 2008].

## CONCLUSION

Isolated right heart strain in middle-aged individuals should be intensively examined to exclude diseases with rare entities.

## ACKNOWLEDGEMENT

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## REFERENCES

Adler SC, Silverman JF. 1973. Anomalous Venous Drainage of the Left Upper Lobe. Radiology. 108(3):563-565.

Edwin F. 2010. Left-sided partial anomalous pulmonary venous connection – should diagnosis lead to surgery? Interact Cardiovasc Thorac Surg. 11(6):847-848.

ElBardissi AW, Dearani JA, Suri RM, Danielson GK. 2008. Left-Sided Partial Anomalous Pulmonary Venous Connections. Ann Thorac Surg. 85(3):1007-1014.

Haramati LB, Moche IE, Rivera VT, et al. 2003. Computed Tomography of Partial Anomalous Pulmonary Venous Connection in Adults. J Comput Assist Tomogr. 27(5):743-749.

Kao CC, Hsieh CC, Cheng PJ, Chiang CH, Huang SY. 2017. Total Anomalous Pulmonary Venous Connection: From Embryology to a Prenatal Ultrasound Diagnostic Update. J Med Ultrasound. 25(3):130-137.

Pennes DR, Ellis JH. 1986. Anomalous pulmonary venous drainage of the left upper lobe shown by CT scans. Radiology. 159(1):23-24.

Tricarico E, Tricarico F, Florio C. 2016. Left-Sided Upper Partial Anomalous Pulmonary Venous Return through a Curved Vein Joining the Left Brachiocephalic Vein. Case Rep Radiol. 1780909.