

# Partial anomalous pulmonary venous connection in a 72-year-old woman: A case report

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Lucas Molinari Veloso da Silveira<sup>1,2</sup> , Miguel Gus<sup>2,3</sup>,  
Felipe Soares Torres<sup>2</sup>, Flávio Danni Fuchs<sup>2,3</sup> and  
Sandra Costa Fuchs<sup>2</sup>

## Abstract

Partial Anomalous Pulmonary Venous Connection is a congenital abnormality characterized by drainage of one or more, but not all, pulmonary veins to the right atrium or to one of the systemic veins. This pathology has low prevalence, although it probably is underestimated and is rarely diagnosed in adults. This report describes a case of a 72-year-old woman with long-term worsening shortness of breath and elevated pulmonary artery systolic pressure in which Partial Anomalous Pulmonary Venous Connection was occasionally diagnosed through imaging methods.

## Keywords

Cardiology, partial anomalous pulmonary venous connection, hypertension, pulmonary, aged

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## Introduction

Partial anomalous pulmonary venous connection (PAPVC) is a congenital abnormality characterized by the failure of fusion of embryologic pulmonary venous system with the left atrium.<sup>1</sup> Its prevalence ranges between 0.1% and 0.2%<sup>2,3</sup> among adults; however, it might be higher since most cases can progress unnoticed. Patients with PAPVC usually have a clinical course that consists in worsening of dyspnea. It is considered a benign lesion with no propensity to cause significant pulmonary vascular changes. Nonetheless, development of pulmonary vascular obstructive disease with markedly increase in pulmonary vascular resistance (PVR) has been described, with shortened life expectancy.<sup>4</sup> The presentation of this case report aims to forewarn that the diagnosis of PAPVC should be considered in elderly patients with occasional unexplained breathing complains. In addition, patients with unexplained raise in the pulmonary pressure should have this condition in the differential diagnosis of primary pulmonary hypertension (PH).<sup>5</sup>

## Case report

A female patient, aged 72-year-old, obese, hypertensive and with a previous ischemic stroke, reported progressive breathlessness during the previous 2 years, with no other clinical

complaints. Physical examination revealed controlled blood pressure and heart rate of 55 beats per minute. There was no jugular venous distention or left parasternal lift. A midsystolic 2+/6 murmur was heard over the precordium in the mitral area. The pulmonary and abdominal examinations showed no abnormalities. The transthoracic echocardiography disclosed mild dilatation of the right chambers and an intra atrial septum aneurysm with no visible communication. None segmental abnormality compatible with ischemic disease was disclosed in this exam.

A transesophageal echocardiography revealed a dilatation of the right chambers and pulmonary artery systolic pressure of 58 mmHg. Besides, it was observed an interatrial septal

<sup>1</sup>Centro de Pesquisa Clínica, Hospital de Clínicas de Porto Alegre, School of Medicine, Universidade Federal do Rio Grande do Sul, Porto Alegre, Brazil

<sup>2</sup>Postgraduate Studies Program in Cardiology, School of Medicine, Universidade Federal do Rio Grande do Sul, Porto Alegre, Brazil

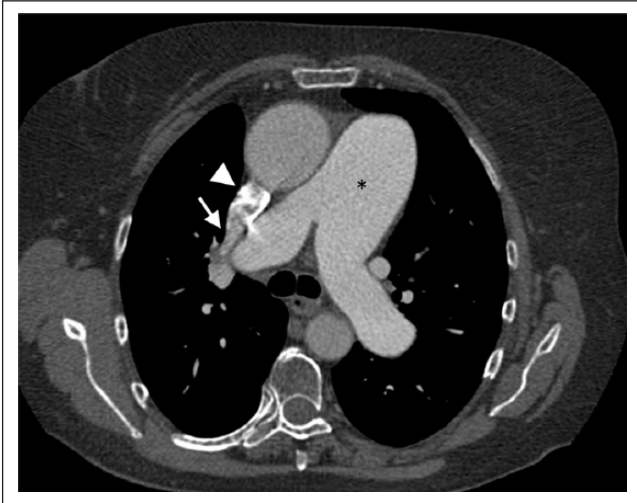
<sup>3</sup>Division of Cardiology, Hospital de Clínicas de Porto Alegre, Porto Alegre, Brazil

## Corresponding Author:

Lucas Molinari Veloso da Silveira, Centro de Pesquisa Clínica, Hospital de Clínicas de Porto Alegre, School of Medicine, Universidade Federal do Rio Grande do Sul, 5º andar, Estudo PREVER, Rua Ramiro Barcelos, 2350, CEP 90.035-903, Porto Alegre, RS, Brazil.

Email: lmvsilveira@gmail.com





**Figure 1.** Contrast-enhanced computed tomography transaxial image demonstrating the right upper pulmonary vein (arrow) draining to the superior vena cava (arrowhead). Note the enlarged main pulmonary artery (asterisk).



**Figure 2.** Contrast-enhanced computed tomography sagittal oblique reconstruction demonstrating the right upper pulmonary vein (arrow) draining to the superior vena cava (asterisk).

aneurysm with a patent foramen ovale. The estimated pulmonary/systemic flow ratio ( $Q_p/Q_s$ ) was 2.2. The chest angiography showed a dilatation in the pulmonary artery trunk, measuring 43 mm (Figure 1). In addition, there was a partial anomalous venous drainage from the right upper lobe to the superior vena cava (SVC) (one vein), with the confluence of the abnormal vein occurring 3 cm above the junction from the SVC with the right atrium (RA; Figures 1 and 2). Normal coronary arteries were disclosed. Thromboembolic events were discarded and primarily lung disease was also discarded. Other possible causes of PH were excluded by pneumology staff after performing specific examinations. However, we highlight that in cases such as this, it is possible that PAPVC coexists with another form of PH, not fully explaining the severity of PH alone. Later, a cardiac magnetic resonance imaging (MRI) was performed. The MRI confirmed the anatomical findings from the computed tomography, however disclosed a left-to-right shunt less important than the one disclosed by the echocardiography ( $Q_p/Q_s$  1.2). Right heart catheterization revealed mean pulmonary artery pressure of 54 mmHg, pulmonary capillary pressures of 12 mmHg and PVR of 4 Wood Units, characterizing a pre-capillary PH. Cardiac output was 10.47 L/min.

Since the patient presented a high surgical risk (Euroscore II 7.97%), both patient and a multi-disciplinary team (cardiology, pneumology and cardiac surgery) opted for conservative treatment in this case. Therefore, the assistant physician decided to prescribe Sildenafil, and the patient had a remarkable improvement in dyspnea.

## Discussion

PAPVC occurs when one or more pulmonary veins (PV), but not all the veins, connects to the RA or one of the venous

tributaries instead of the left atrium, resulting in a left to right shunt. The majority of the patients diagnosed with PAPVC presents with fatigue, dyspnea—both present in about 40% of the patients—and palpitations. It is important to highlight that in some cases, the symptoms caused by PAPV are the same as the ones caused by any other cause of PH. When analyzing right ventricle (RV) function, despite volume overload, these patients usually present with normal or only mild RV dysfunction.<sup>1</sup> Previous series with data from pediatric patients reported increased occurrence in men and anomalous right PV draining to the SVC as the most common abnormality in the pulmonary drainage, situation present in 74% of the patients.<sup>6</sup> These patients usually have a sinus venosus atrial septal defect (ASD).<sup>2</sup> However, in a more recent study focused in adult patients, PAPVC was more diagnosed in women and with anomalous drainage coming in 79% of the cases from the left side. Besides, the association with ASD was not found in this report.<sup>3</sup> These findings disclose differences in the characteristics of PAPVC according to age groups. In this report, the most frequent presentation of PAPVC diagnosed in children was present in an elderly patient, age in which this diagnosis is rarely established.

The definitive management of PAPVC consists in the surgical separation of the pulmonary venous system from the systemic system through redirection of an aberrant pulmonary venous drain into the left atrium.<sup>6</sup> However, it is controversial when to indicate this procedure in adult patients, since the surgical correction is considered complex and has risk of PV stenosis at the anastomotic site, vena caval obstruction and residual shunts.<sup>1</sup> Nevertheless, there were no deaths in the largest series of patients reported to date (306 patients).<sup>6</sup>

Considering the limited data existent about this pathology, current guidelines do not specifically address PAPVC

treatment in adults, and it is considered reasonable to apply the same guidelines established for ASD.<sup>7</sup> In this case, it was decided to initiate Sildenafil, which showed favorable functional and hemodynamic results in patients with pulmonary artery hypertension associated with congenital heart diseases.<sup>8</sup> The decision to use Sildenafil in this case was based on the fact that in patients with severe pulmonary artery hypertension and elevated PVR surgery is unlikely to alter diseases course. These cases may have as only curative option the heart-lung transplant.<sup>9</sup> Besides, our patient had relevant comorbidity, increasing the risks of a major surgical procedure.

The PAPVC diagnoses in adults are mostly made by chest angiotomography, method with extremely high diagnostic accuracy.<sup>2</sup> However, it is important to carefully check the anatomy of PVs, because there is a considerable risk of misinterpretation of exams.<sup>10</sup>

This case reports a classical course of PAPVC—long term of mild dyspnea, a benign clinical course and the occasional finding of PAPVC during investigation of PH. This case highlights the importance to suspect of PAPVC, even in elderly patients, and the fact that patients with an increased pressure in the pulmonary circulation must be presented to the differential diagnosis of primary PH,<sup>5</sup> through careful interpretation of imaging methods, this means that we should always look for coexisting shunt lesions in adults presenting with PH.

## Conclusion

This case reports allows us to observe an important cause of PH that may have been misdiagnosed in many adults presenting with PH just because this diagnosis has not been considered

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## ORCID iD

Lucas Molinari Veloso da Silveira  <https://orcid.org/0000-0002-3543-594X>

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