

# Deciphering a case of pulmonary hypertension in a young female: Partial anomalous pulmonary venous drainage the culprit

Hiten R. Patel, Suchit Bhutani<sup>1</sup>, Fayez Shamoan, Hartaj Virk

Department of Cardiology,  
Saint Joseph's Regional  
Medical Center,  
New York Medical  
College, New Jersey,  
<sup>1</sup>Department of Internal  
Medicine, Abington  
Hospital-Jefferson Health,  
Philadelphia, USA

**Address for  
correspondence:**

Dr. Hiten R. Patel,  
Saint Joseph's Regional  
Medical Center, New York  
Medical College,  
703 Main Street, Paterson,  
New Jersey - 07503, USA.  
E-mail: hiten0409@yahoo.  
com

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**Abstract:**

Partial anomalous pulmonary venous drainage (PAPVD) is a rare congenital cardiac defect and is associated with sinus venosus atrial septal defect. While most cases are asymptomatic, a patient can present with pulmonary hypertension (PHTN) and it can be difficult to diagnose. Here, we discuss the case of a young female with PHTN who was found to have two right-sided PAPVD. Through this case, authors try to emphasize the importance of meticulous and thorough investigation when evaluating PHTN, which allows for correct diagnosis and a timely intervention before PHTN becomes irreversible.

**Keywords:**

Partial anomalous pulmonary venous drainage, pulmonary hypertension, sinus venosus atrial septal defect

Partial anomalous pulmonary venous drainage (PAPVD) is a rare congenital cardiac defect with the incidence of 0.4%–0.7% and is associated with sinus venosus atrial septal defect (ASD). While most cases are asymptomatic, a patient can present with pulmonary hypertension (PHTN) and it can be difficult to diagnose. Here, we discuss the case of a young female with PHTN where thorough investigations lead to a correct diagnosis of PAPVD and ultimately a timely intervention.

## Case Report

Our case is a 33-year-old female with morbid obesity who needed preoperative cardiac risk stratification for gastric bypass surgery. She complained of mild dyspnea on exertion for 2–3 months but denied any history of chest pain, leg swelling, palpitations, syncope, or cardiac problems in the past. Her cardiac examination

revealed normal S1, loud P2, and 3/6 systolic murmur in the left 3<sup>rd</sup> intercostal space. Her electrocardiogram revealed the right bundle branch block [Figure 1] and her chest X-ray revealed right ventricular (RV) enlargement [Figure 2]; hence, a transthoracic echocardiogram was ordered for further evaluation. Later showed normal left ventricular systolic and diastolic function, moderately dilated RV and right atrium (RA) [Figure 3], and pulmonary artery (PA) systolic pressure was estimated at 50 mmHg. Computed tomographic angiography for pulmonary embolism was negative. Right heart catheterization was performed which revealed elevated mean RA pressure of 11 mmHg, borderline elevated mean PA pressure of 25 mmHg, mean capillary wedge pressure of 13 mmHg, and a high oxygen saturation difference between PA and superior vena cava (SVC) of 23% ( $\geq 8\%$ ); hence, a detailed oximetry run was done which showed a step-up in the oxygen saturation from 69% to 92% between

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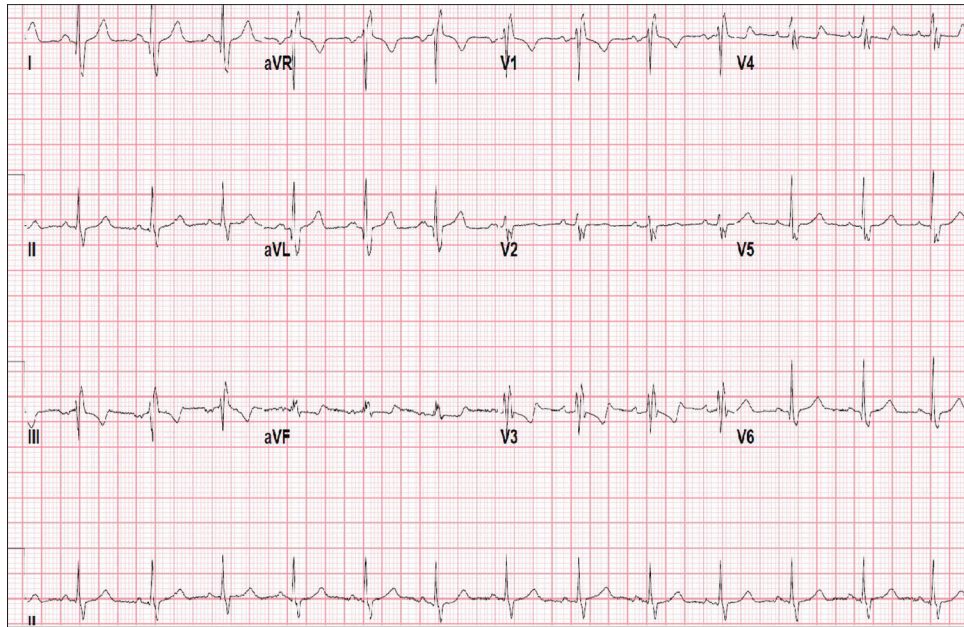


Figure 1: Electrocardiogram showing normal sinus rhythm with right bundle branch block

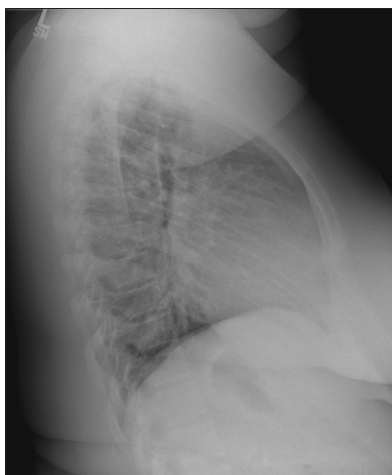


Figure 2: Chest radiography in lateral view showing prominent right ventricle

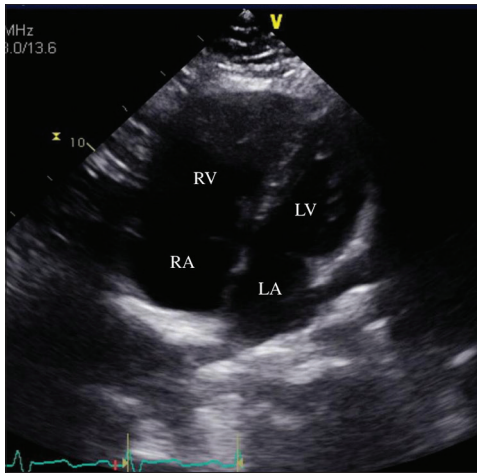
SVC and high RA, which suggests that the intracardiac shunt is at the high RA level. Differential diagnosis includes ASD, PAPVD, ruptured sinus of Valsalva into the RA, and coronary artery fistula to RA.<sup>[1]</sup> Finally, cardiac magnetic resonance angiography (MRA) was done that revealed enlarged right side of the heart with preserved RV ejection fraction of 54%, large anomalous right upper pulmonary vein (RUPV) draining into the SVC-RA junction [Figure 4], and small anomalous right middle pulmonary vein draining into the left atrium (LA) with sinus venosus ASD [Figure 5]. Ratio of pulmonic to systemic blood flow ( $Q_p:Q_s$ ) was 2.78:1. Therefore, cardiothoracic surgery was consulted and correction of the defect was recommended before bariatric surgery. A pericardial patch was used to create a baffle to redirect pulmonary venous blood from the anomalous RUPV beneath the baffle through the ASD into the LA. The

patient tolerated the procedure well and was discharged in a stable condition.

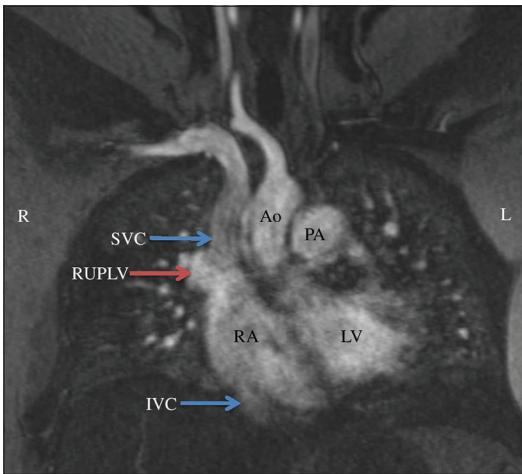
## Discussion

First described by Winslow in 1739,<sup>[2]</sup> PAPVD is a rare congenital cardiac defect which is more common in females with an incidence of 0.4%–0.7% in autopsy series;<sup>[3]</sup> this may overestimate the clinical significance because most cases are asymptomatic. It is different from total anomalous pulmonary venous drainage in which all or most pulmonary veins drain into the right side of the heart. In PAPVD, usually a single pulmonary vein is anomalous, but there can be some exceptions like in our patient wherein two right-sided pulmonary veins were draining into RA. The etiology is unknown, but it represents the persistence of embryonic anastomosis between the systemic and pulmonary vein plexus, resulting in one or more anomalously connecting pulmonary veins. Patients with Turner syndrome, in particular, are at increased risk for PAPVD.<sup>[4]</sup>

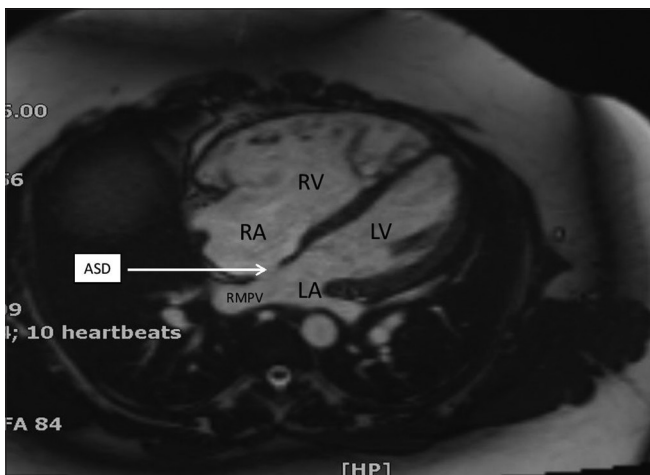
The most common ASD associated with PAPVD is sinus venosus type of ASD (80%–90% of cases). In about 10% of cases, the ASD is of secundum type. Normally, each PV contributes an average of 25% of the total pulmonary blood flow; however, when a PV connects anomalously to the RA or SVC, blood is preferentially shunted to this anomalous vein because of the lower RA pressure, compared with LA pressure, producing significant volume overload. This is especially true in the presence of systemic hypertension, mitral valve disease, or left ventricular dysfunction, which increases LA pressures. The clinical evidence may not be apparent



**Figure 3:** Transthoracic echocardiogram (in apical 4-chamber view) showing moderately dilated right ventricle and right atrium



**Figure 4:** Cardiac magnetic resonance angiography showing large anomalous RUPLV draining into the SVC-RA junction. RUPLV=Right upper pulmonary vein, SVC=Superior vena cava, RA=Right atrium, IVC=Inferior vena cava, Ao=Aorta, PA=Pulmonary artery, LV=Left ventricle



**Figure 5:** Cardiac magnetic resonance imaging showing small anomalous RMPV draining into the LA with sinus venosus ASD. RMPV=Right middle pulmonary vein, ASD=Atrial septal defect, RA=Right atrium, RV=Right ventricle, LA=Left atrium, LV=Left ventricle

until the patient reaches middle age. Some authors have suggested that this defect becomes clinically significant when 50% or more of the pulmonary veins anomalously return.

Adult patient may present with dyspnea and occasionally palpitations. Physical examination findings can reveal signs of the right-sided heart failure and PHTN. The diagnosis can be confirmed with transthoracic echocardiogram,<sup>[5]</sup> but all the pulmonary veins may not be identified, especially in adults. Cardiovascular magnetic resonance imaging (CMR) is rapidly becoming the procedure of choice to diagnose and characterize congenital heart disease, including PAPVD. CMR also provides additional information including quantitation of heart chamber volumes, ventricular mass, and blood flow through the great vessels, especially when other modalities such as echocardiography yield equivocal findings.<sup>[6-8]</sup> Several techniques used in CMR are particularly useful in the diagnosis of PAPVD; these include cardiac MRA which provides enhanced visualization of the pulmonary vasculature including the anomalous pulmonary veins and phase velocity mapping which directly measure the shunt volume (Qp:Qs) noninvasively. Contrast-enhanced computed tomography scanning is an alternative imaging modality that can help in preoperative planning.<sup>[9]</sup>

Medical therapy is indicated for patients with heart failure or arrhythmias, but the definite treatment is surgical repair, especially when the Qp:Qs is >2.1:1. For the PAPVD to the SVC, the repair techniques may include internal patch technique with or without SVC enlargement, or the caval division technique with atriocaval anastomosis known as Warden technique.<sup>[10,11]</sup> Patients with internal patch technique must be observed for obstruction of the SVC with SVC syndrome, obstruction of the pulmonary veins, sick sinus syndrome, and supraventricular tachyarrhythmias.<sup>[12]</sup> The perioperative mortality rate is comparable to that for ASD repair (<0.1%). Prognosis is excellent if surgical repair is done early,<sup>[13]</sup> but it becomes more guarded if the lesion is undetected for a long period.

This case emphasizes the importance of meticulous and thorough investigation when evaluating PHTN. This allows for correct diagnosis and correction of cardiac abnormalities before any noncardiac surgery and before PHTN becomes irreversible.

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**Conflicts of interest**

There are no conflicts of interest.

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