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A Novel Presentation of Hemodynamic Decompensation With Cardiogenic Shock in a Patient With Partial Anomalous Pulmonary Venous Return

Authors' Contribution:
Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
Funds Collection G

AEF 1 Wafaa Mansour
EF 1 Jamil Qiqieh
E 2 Jonathan Stephan
EF 3 Sara Gezahegn
E 1 Fadi Ayoub
E 3 Dawit K. Worku
EF 3 Alehegn Gelaye

1 Department of Internal Medicine, Henry Ford Providence Hospital, Southfield, MI, USA
2 Department of Cardiology, Henry Ford Providence Hospital Southfield, MI, USA
3 Department of Pulmonology and Critical Care, Henry Ford Providence Hospital, Southfield, MI, USA

Corresponding Author: Wafaa Mansour, Department of Internal Medicine, Henry Ford Providence Hospital, 16001 W Nine Mile Rd, Southfield, MI 48075, USA, Phone: (248) 849-3281, e-mail: wmansou2@hfhs.org

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Patient: Male, 51-year-old
Final Diagnosis: Partial anomalous pulmonary venous return (PAPVR)
Symptoms: Shortness of breath
Clinical Procedure: —
Specialty: Critical Care Medicine
Objective: Congenital defects/diseases

Background: Partial anomalous pulmonary venous return (PAPVR) is a rare congenital cardiovascular anomaly that can remain clinically silent until significant shunting results in pulmonary hypertension and right ventricular dysfunction. We report a rare presentation of PAPVR complicated by acute right-sided cardiogenic shock in a previously stable adult, highlighting the potential for abrupt decompensation in advanced disease.

Case Report: A 51-year-old African American man with known PAPVR and pulmonary hypertension presented with 1 week of progressive dyspnea, rapidly developing acute respiratory failure requiring BiPAP. His condition deteriorated within 24 hours, requiring intubation and intensive care unit (ICU) admission. Transthoracic echocardiography demonstrated severe right ventricular dilation, reduced systolic function, and septal flattening consistent with pressure overload. He developed right-sided cardiogenic shock requiring inotropes, vasopressors, and aggressive diuresis. After stabilization, he was extubated and discharged with plans for surgical evaluation. This case report highlights a rare presentation of PAPVR in which cardiogenic shock was caused by initial critical decompensation. Despite initial recovery, he did not undergo corrective intervention, as he later fell at home and had a fatal intracranial hemorrhage, illustrating the importance of early recognition, close follow-up, and timely surgical consideration.

Conclusions: Because PAPVR is often asymptomatic and considered benign, it is frequently managed conservatively; however, risk stratification based on shunt magnitude, right ventricular remodeling, and pulmonary pressures is essential. Patients with large shunts and pulmonary hypertension, as in our case, may be at risk of rapid decompensation, requiring prompt diagnosis, close monitoring, and early surgical evaluation.

Keywords: Cardiology • Cardiomyopathies • Critical Care • Pulmonary Adenomatosis, Ovine • Pulmonary Circulation • Ventricular Dysfunction, Right

Abbreviations: **PAPVR**, partial anomalous pulmonary venous return; **RV**, right ventricle; **LV**, left ventricle; **ICU**, Intensive Care Unit; **BiPAP**, bilevel positive airway pressure

Full-text PDF: https://www.amjcaserep.com/abstract/index/idArt/953243

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Introduction

Partial anomalous pulmonary venous return (PAPVR) is a rare congenital cardiovascular anomaly in which 1 or more pulmonary veins drain into the right atrium or systemic venous circulation rather than into the left atrium. It is estimated to occur in approximately 0.4% to 1% of the population [1]. Because only a portion of pulmonary venous return is anomalous while the remaining veins drain normally into the left atrium, many individuals remain asymptomatic, and the condition is often identified incidentally [2]. However, patients with larger left-to-right shunts may develop progressive pulmonary hypertension and right ventricular (RV) volume overload. This occurs as oxygenated blood is redirected into the systemic venous circulation—most commonly the superior vena cava, inferior vena cava, or right atrium—resulting in recirculation through the right heart and pulmonary vasculature. The magnitude of shunting is quantified by the Qp: Qs ratio, defined as the ratio of pulmonary to systemic blood flow, with values ≥1.5: 1 considered hemodynamically significant [3,4]. Chronic volume overload leads to progressive RV dilation and dysfunction. Although compensatory mechanisms can persist for years, abrupt decompensation can occur, resulting in acute right-sided heart failure [5]. We present a rare case of PAPVR complicated by right-sided cardiogenic shock in a previously stable adult, highlighting the importance of early recognition, risk stratification, and timely surgical intervention.

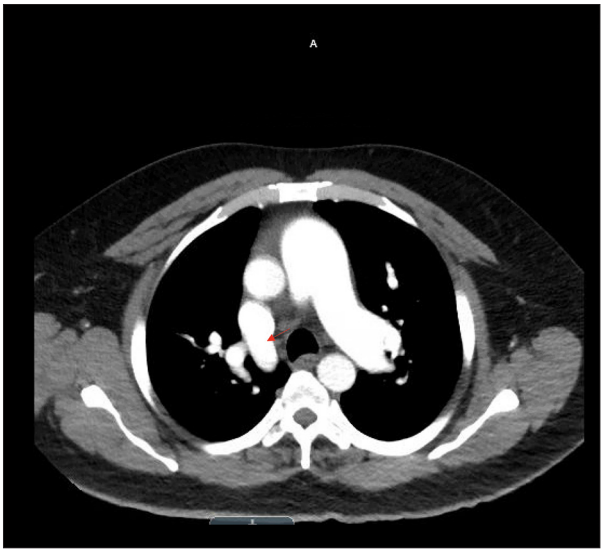

Figure 1. Contrast-enhanced computed tomography scan of the chest demonstrating anomalous drainage of a right pulmonary vein into the azygos vein (red arrow), findings consistent with partial anomalous pulmonary venous return. Image quality is limited due to the use of externally obtained CTA images without access to the original dataset for additional reconstruction.

Case Report

A 51-year-old African American man with a history of PAPVR and pulmonary hypertension presented with 1 week of progressive dyspnea. His PAPVR had been diagnosed 2 years earlier at an outside facility and involved a large right lower lobe pulmonary vein draining into the azygos vein (**Figure 1**), with an estimated 2: 1 left-to-right shunt and a severely elevated right ventricular systolic pressure of 101 mmHg. Pulmonary blood flow was markedly elevated at 8.79 L/min, and pulmonary vascular resistance was 5.2 Wood units, consistent with significant pulmonary hypertension (**Table 1**). Over the preceding 2 years, he had multiple admissions for heart failure exacerbations but had never required intubation or intensive care unit (ICU)-level care. On presentation, examination revealed volume overload. Chest radiography demonstrated cardiomegaly and moderate pulmonary congestion. He was admitted for acute decompensated heart failure and treated with diuresis. On hospital day 1, his respiratory status worsened, prompting a rapid response. Arterial blood gas revealed severe hypercapnia with $PaCO_2$ 116 mmHg and pH 7.13, consistent with acute hypercapnic respiratory acidosis and failure. Despite bilevel positive airway pressure (BiPAP), his condition worsened during the same day, necessitating intubation and transfer to the ICU. He was managed on ventilator settings with PEEP of 5 cmH_2O, FiO_2 of 40%, and a respiratory rate of 12 breaths per minute, with tidal volumes initially set at 450 mL and subsequently reduced to 400 mL following improvement in arterial blood gas parameters on day 2 of intubation (**Table 2**). Transthoracic echocardiography demonstrated a hyperdynamic

Table 1. Hemodynamic parameters at diagnosis showing a significant left-to-right shunt with elevated right-sided pressures and pulmonary vascular resistance consistent with pulmonary hypertension.

Parameter	Value	Reference range/normal value
Left-to-right shunt ratio (Qp: Qs)	2: 1	~1: 1
Right ventricular systolic pressure	101 mmHg	15-30 mmHg
Pulmonary blood flow (Qp)	8.79 L/min	~4-6 L/min
Pulmonary vascular resistance (PVR)	5.2 Wood units	0.25-1.6 Wood units

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Table 2. Arterial blood gad trends show severe hypercapnic respiratory acidosis pre-intubation with improvement after ventilation; persistent hypercapnia with normal pH suggests chronic CO_2 retention with metabolic compensation.

Parameter	Initial (pre-intubation)	Post-intubation	Reference range
pH	7.13	7.45	7.35-7.45
$PaCO_2$	116 mmHg	70 mmHg	35-45 mmHg
PaO_2	96 mmHg	97 mmHg	80-100 mmHg

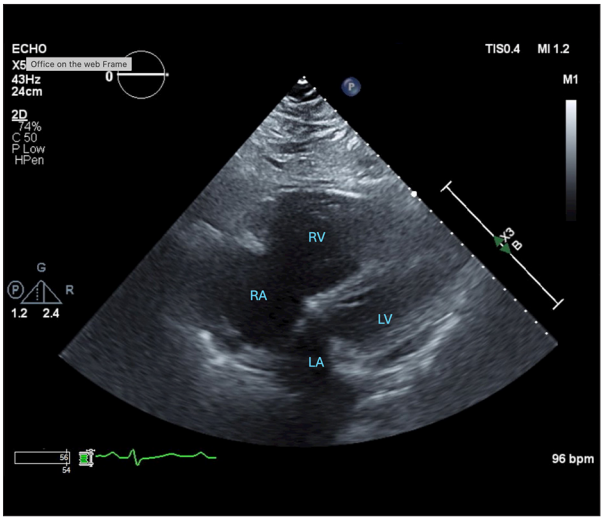

Figure 2. Transthoracic echocardiography (subcostal view) demonstrates right ventricular enlargement with an increased RV/LV ratio, reflecting right ventricular dilation that exceeds the size of the left ventricle. Additionally, the right ventricular wall appears thickened, suggesting chronic pressure overload and compensatory remodeling. These findings are indicative of significant right-sided volume or pressure overload and suggest advanced right ventricular remodeling.

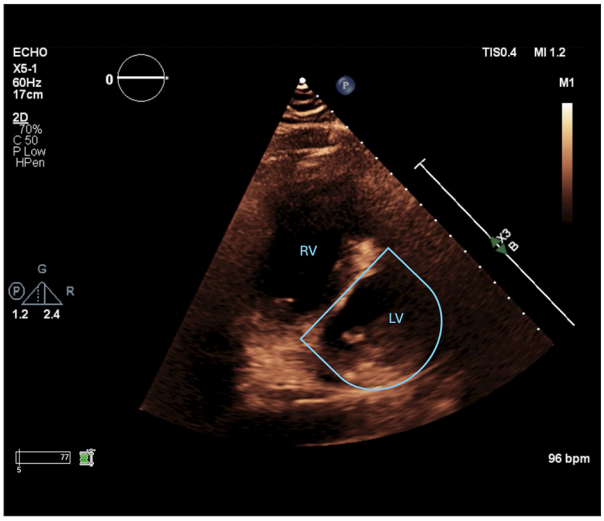

Figure 3. Transthoracic echocardiography (parasternal short axis) demonstrating the classic D-sign, characterized by interventricular septal flattening that results in a D-shaped left ventricle during diastole, consistent with right ventricular volume overload.

left ventricle with preserved ejection fraction of 72%, grade I diastolic dysfunction, and normal left atrial size. In contrast, the right ventricle was severely dilated and thickened (**Figure 2**) with moderately reduced systolic function, and interventricular septal flattening (**Figure 3**), consistent with RV pressure overload. Central venous pressure was elevated at 20 mmHg, and the right atrium was severely enlarged, consistent with long-standing pulmonary hypertension. The patient was treated for right-sided cardiogenic shock with cautious diuresis to reduce RV volume overload and improve ventricular interdependence, along with norepinephrine to maintain systemic perfusion as mean arterial pressure declined below 65 mmHg. Due to persistent hemodynamic instability despite vasopressor support, milrinone was initiated for inotropic support and pulmonary vasodilation. As an inodilator, milrinone enhances right ventricular contractility while reducing pulmonary vascular resistance, thereby decreasing RV afterload and improving forward blood flow. Over the following 3 days, both respiratory and hemodynamic status improved, allowing successful extubation. The patient was subsequently maintained on low-dose bumetanide for volume management and sildenafil for pulmonary hypertension. He was weaned off supplemental oxygen within 1 week and was discharged in stable condition.

Outpatient follow-up with pulmonology, cardiology, and cardiothoracic surgery was arranged for evaluation of surgical repair. However, 1 month following discharge, the patient was rehospitalized with hypercapnic respiratory failure and altered mental status, complicated by a fall resulting in fatal intracranial hemorrhage. This event was considered unrelated to the patient's underlying PAPVR or the index hospitalization described in this report. The sequence of events is outlined in **Table 3**.

Discussion

This case report describes an adult patient with PAPVR and pulmonary hypertension who developed acute decompensation

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Table 3. Clinical timeline of presentation, hospital course, management, and outcomes of the case.

Timepoint	Clinical events
2 years prior	Diagnosed with PAPVR involving right lower lobe pulmonary vein draining into azygos vein; Qp: Qs ~2: 1; RVSP 101 mmHg; PBF 8.79 L/min; PVR 5.2 Wood units
Prior 2 years	Multiple admissions for heart failure exacerbations; no prior intubation or ICU admission
1 week prior to admission	Progressive dyspnea
Day 0 (presentation)	Volume overload on exam; CXR: cardiomegaly and pulmonary congestion; admitted for acute decompensated heart failure; initiated on diuresis
Hospital Day 1	Respiratory deterioration → rapid response; ABG: severe hypercapnia ($PaCO_2$ 116 mmHg, pH 7.13); BiPAP initiated but failed; required intubation and ICU transfer
ICU course (Days 1-3)	Ventilated (PEEP 5 cmH_2O, FiO_2 40%, RR 12); improvement in ABG by day 2; tidal volumes reduced from 450 mL to 400 mL
Cardiac evaluation	TTE: LVEF 72%, severe RV dilation, reduced RV function, septal flattening; CVP 20 mmHg; severe RA enlargement
Shock management	Cardiogenic shock treated with cautious diuresis and norepinephrine; milrinone added for inotropic support and pulmonary vasodilation
Hospital Days 2-5	Gradual improvement in respiratory and hemodynamic status
Post-ICU course	Successfully extubated; continued diuresis (bumetanide) and sildenafil
Within 1 week after extubation	Weaned off supplemental oxygen; discharged in stable condition
Post-discharge plan	Follow-up arranged with pulmonology, cardiology, and cardiothoracic surgery for surgical evaluation
1 month after discharge	Fatal intracranial hemorrhage

with right ventricular (RV)–predominant cardiogenic shock following a period of progressive dyspnea, an outcome infrequently reported in adults with this condition. PAPVR is an uncommon congenital anomaly that is often clinically silent and discovered incidentally [1,6]. In a cohort of 53 patients with isolated PAPVR and an intact atrial septum, more than half were asymptomatic at presentation, with diagnoses frequently made incidentally [7]. Similarly, incidental detection has been reported in adults with small shunts and minimal hemodynamic impact [8].

In contrast, our patient demonstrated objective evidence of advanced pulmonary vascular disease, including markedly elevated pulmonary blood flow (8.79 L/min), pulmonary vascular resistance (5.2 Wood units), and RV systolic pressure (101 mmHg), consistent with a high cumulative hemodynamic burden. These findings support pathophysiological progression from chronic left-to-right shunting to pulmonary vascular remodeling, increased RV afterload, and eventual RV dysfunction [9]. Echocardiography further demonstrated severe RV dilation, reduced systolic function, interventricular septal flattening, and elevated central venous pressure, all consistent with RV pressure overload and failure [7,9]. The development of severe hypercapnia despite preserved oxygenation can reflect impaired pulmonary perfusion and ventilation–perfusion mismatch in the setting of RV dysfunction and elevated pulmonary pressures [10]. Alternative contributors to shock, including left ventricular failure, were not supported by the preserved ejection fraction and normal left atrial size.

Although cardiogenic shock is not commonly described in isolated PAPVR, progressive pulmonary hypertension and RV failure have been reported, particularly in patients with larger shunts or delayed diagnosis [7]. Disease progression appears to correlate with cumulative hemodynamic burden and age. In a 10-year single-center experience, age was significantly associated with RV enlargement (r=0.398, *P*=0.03) and increased pulmonary artery pressures (r=0.423, *P*=0.02), supporting the role of chronic remodeling in adverse outcomes [11]. In our patient, diagnosis delayed until he was in his 50s likely allowed for prolonged exposure to increased pulmonary blood flow and progressive vascular changes.

While many adults with PAPVR and intact atrial septum are managed conservatively when clinically stable [4], surgical repair has been shown to provide excellent outcomes when

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appropriate criteria are met. In a cohort of 45 patients with PAPVR and intact atrial septum undergoing surgical correction, there was no postoperative mortality, with survival rates of 100%, 95%, and 95% at 1, 5, and 10 years, respectively. Multiple surgical approaches—including direct reimplantation, intracardiac baffling, and the Warden procedure—were performed, with low rates of complications and minimal incidence of postoperative pulmonary or systemic venous obstruction [12]. These findings support the safety and durability of surgical intervention and emphasize the importance of timely referral before the development of irreversible pulmonary vascular disease.

Conclusions

Despite often being considered benign, patients with PAPVR and significant left-to-right shunting can develop progressive pulmonary vascular disease leading to right ventricular failure over time. Acute decompensation can occur when pulmonary pressures exceed the compensatory capacity of the right ventricle, highlighting the importance of close surveillance, accurate risk stratification, and serial monitoring with timely escalation of supportive therapies.

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Department and Institution Where Work Was Done

Henry Ford Providence Hospital, Southfield, MI, USA.

Informed Consent

Written informed consent was obtained from the patient for publication of this case report.

Declaration of Figures' Authenticity

All figures submitted have been created by the authors who confirm that the images are original with no duplication and have not been previously published in whole or in part.

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