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Diagnostic Work-Up of Pulmonary Hypertension – Think About Shunt Flow! A Case Report

Authors' Contribution:
Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
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Patient: Male, 65
Final Diagnosis: Shunt vitium
Symptoms: Dyspnea
Medication: —
Clinical Procedure: —
Specialty: Cardiology

Objective: Unusual clinical course

Background: The diagnostic work-up and treatment of pulmonary hypertension can be complex. Pulmonary arterial hypertension (PAH), pulmonary hypertension second to lung or heart diseases and thromboembolic pulmonary hypertension, and other rare causes of pulmonary hypertension such as congenital heart diseases must be considered in the differential diagnostic work-up.

Case Report: We report on a patient who has been treated for PAH over many years. At the age of 65, progressive symptoms required a complete re-evaluation. Here, a complex shunt vitium with a partial anomalous pulmonary venous return (PAPVR) and a sinus venosus defect (SVD) was diagnosed.

Conclusions: PAPVR is a rare congenital heart disease that is often associated with an SVD. It is usually diagnosed during childhood but may also be detected in adult patients who develop pulmonary hypertension and dyspnea as primary symptoms. The initial predominant left-to-right shunting associated with this disease may be undetected for years, with a slow development of right heart failure with right heart volume overload and pulmonary hypertension. Early detection is important, with a subsequent surgical intervention.

MeSH Keywords: Heart Defects, Congenital • Hypertension, Pulmonary • Scimitar Syndrome

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Background

Pulmonary hypertension is diagnosed by an increase in mean pulmonary pressure ≥25 mmHg using right heart catheterization at rest [1]. According to mean pulmonary pressure and in relation to pulmonary wedge pressure, cardiac output, diastolic pressure gradient, and pulmonary vascular resistance, pulmonary hypertension can be classified into pre- and post-capillary sub-forms [1]. In addition to PAH, pulmonary hypertension secondary to lung or left heart diseases and thromboembolic pulmonary hypertension, as well as other rare causes of pulmonary hypertension such as congenital heart diseases, must be considered in the differential diagnostic work-up of this disease.

Case Report

A 65-year-old man was admitted to our cardiology department with progressively worsening shortness of breath (New York Heart Association-Classification [NYHA] III) and palpitations. In our outpatient heart rhythm care center, atrial fibrillation had previously been diagnosed and he had been scheduled for transesophageal echocardiography and electrical cardioversion. The patient's records listed coronary artery disease with multiple coronary interventions and a history of PAH, which was first diagnosed in 2007 and treated with several drugs, including macitentan 10 mg per day and bosentan 125 mg twice a day. However, his exertional dyspnea had been increasing steadily over the years.

Upon admission, physical examination revealed a right ventricular cardiac decompensation. Mild bibasilar crackles were audible and bilateral lower-extremity edemas were present. Blood pressure was 106/71 mmHg, respiration rate 15 per min, and peripheral oxygen saturation 93%. We documented an atrial fibrillation with a rate of 133 beats per min. The n-terminal prohormone of brain natriuretic peptide (NT-proBNP) level was increased to 8483 pg/ml. Cardiac enzyme levels, including troponin and creatine kinase, were within normal limits.

Transthoracic echocardiography windows were poor, particularly for the parasternal views, due to obesity. A global systolic left ventricular function within normal range, a dilated right ventricle with normal systolic function (tricuspid annular plane systolic excursion [TAPSE] 28 mm), severe tricuspid regurgitation, and dilated inferior vena cava with reduced respiratory

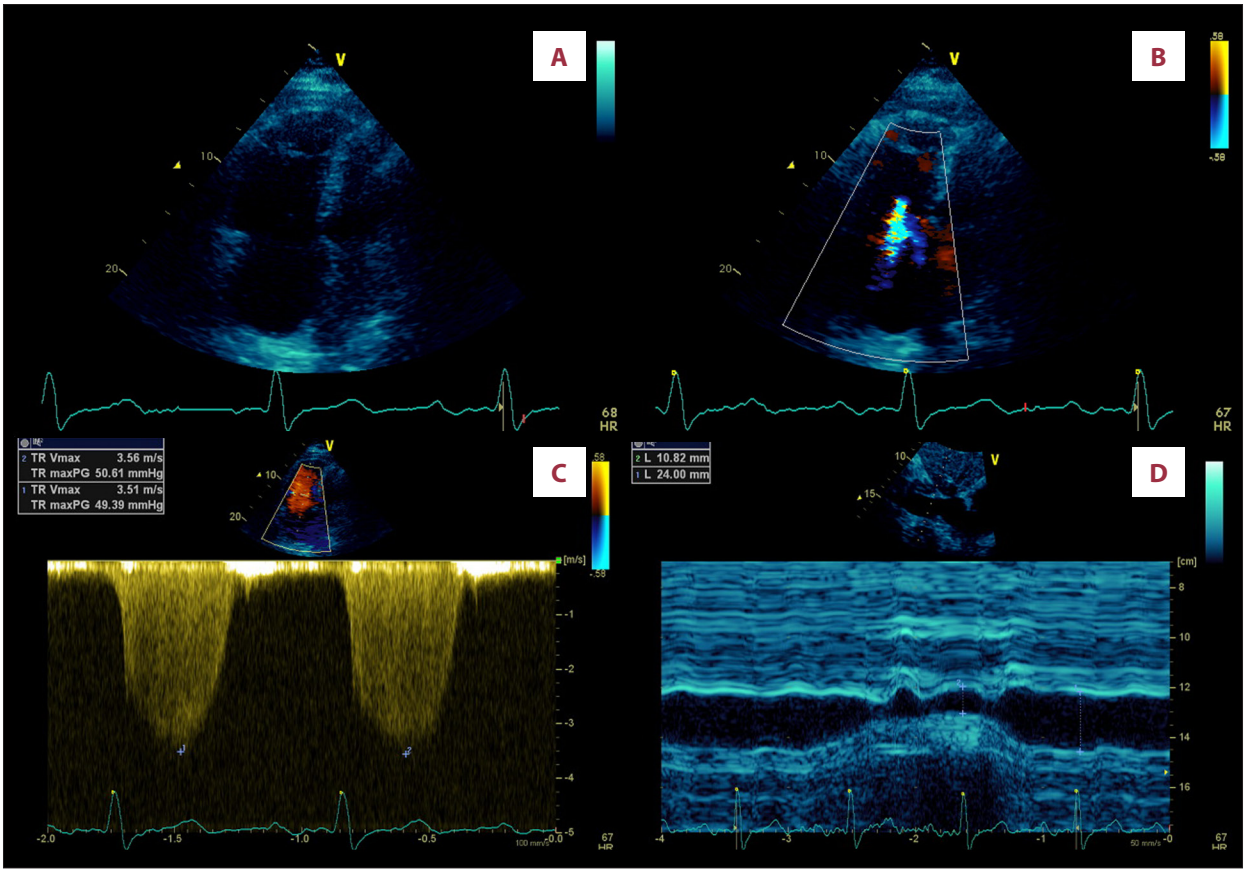

Figure 1. Transthoracic echocardiography showed a dilated right ventricle (**A**), a severe tricuspid regurgitation (**B**) with a peak tricuspid regurgitation velocity of 3.56 m/s (**C**), and a dilated inferior vena cava with reduced respiratory collapse (**D**).

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collapse were detected. In addition, we documented signs of pulmonary hypertension with a peak tricuspid regurgitation velocity of 3.56 m/s and an estimated systolic pulmonary pressure of 49 mmHg plus central venous pressure (Figure 1).

After exclusion of intracavitary thrombi via transesophageal echocardiography, an electrical cardioversion into sinus rhythm was successfully performed. Due to the reported increased dyspnea and the previously known coronary heart disease, a heart catheter investigation was performed. Significant coronary lesions were excluded. The previous cardiac catheter examinations had diagnosed PAH. We now measured a mean pulmonary pressure of 32 mmHg with a pulmonary wedge pressure of 16 mmHg. Thus, a PAH, as previously diagnosed, was now excluded and a combined post-capillary and pre-capillary pulmonary hypertension was determined. Blood samples for oximetry were taken from the high superior vena cava, inferior vena cava, and pulmonary arteries. Surprisingly, the right ventricular catheter examination revealed high oxygen saturation in the pulmonary arteries, with a hitherto undetected jump in the oxygen saturation measurement between the right atrium (70%) and the pulmonary artery (85%), indicating a left-to-right shunt. Figure 2 outlines the oximetric results of the patient, with high oxygen saturations in the lower right atrium and pulmonary artery. The examination revealed a shunt volume with a Qp/Qs (pulmonary blood flow/systemic blood flow) of 3.8: 1 and a pulmonary vascular resistance of 2 WU. The low aortic oxygen saturation (90%) indicated a beginning right-to-left shunt. An angiogram of the superior vena cava revealed a wash-out of the contrast agent due to a PAPVR (Figure 3).

For further characterization of this suspected shunt, magnetic resonance imaging (MRI) was performed, which showed a relevant vitium with a PAPVR with an anomalous connection of the right superior and middle lobe vein with the superior vena cava, as well as an atrial septum defect type sinus venosus (Figure 4).

Due to the severe discomfort of the patient and the recurrent cardiac decompensation, we saw an indication for an operative correction of the shunt vitium. A minimally invasive PAPVR-correction using allogenic pericardium and an atrial septum defect occlusion were successfully performed. The anomalous pulmonary vein was redirected into the left atrium through a tunnel via the atrial septal defect. To avoid constriction of the superior vena cava through this tunnel, the upper vena cava was additionally widened with a tissue patch.

Discussion

PAPVR is a rare congenital heart disease often associated with a SVD [2]. It is usually diagnosed in childhood but may also be detected in adult patients who develop pulmonary hypertension.

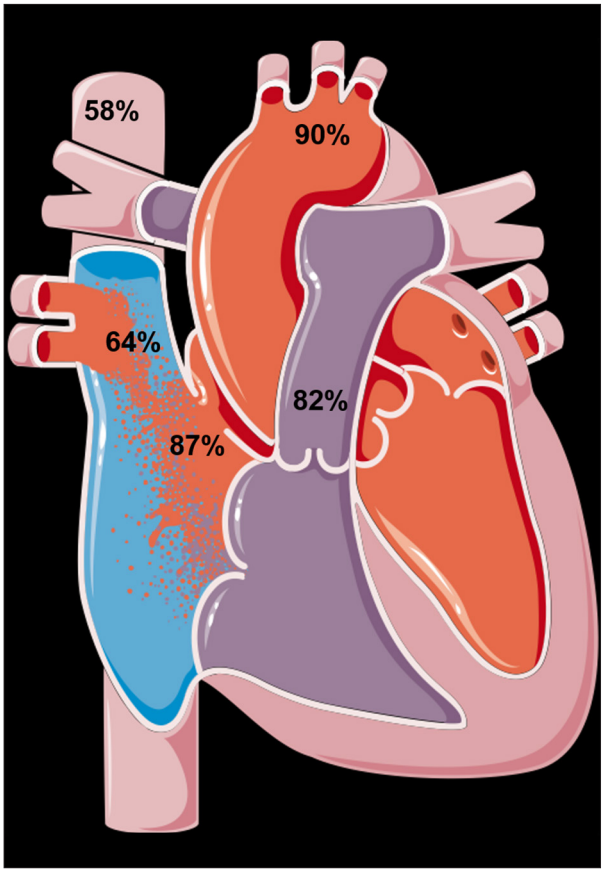

Figure 2. Overview of the oximetric results in the superior and inferior vena cava, the right atrium, the pulmonary artery, and the aorta. The shunt flow into the superior vena cava and the right atrium is pictured. (This figure was produced and modified using Servier Medical Art).

In some cases, PAPVR is concealed and patients are misdiagnosed as suffering from idiopathic PAH. The initial predominant left-to-right shunting associated with this disease may be undetected for years, with a slow development of right heart failure due to right heart volume overload and pulmonary hypertension. The diagnosis and therapy of this anomaly can be challenging. Transthoracic echocardiography cannot always reliably detect PAPVR due to technical limitations [3]. In patients with pulmonary hypertension, MRI may be useful in cases of suspected pulmonary venous anomaly with inconclusive echocardiographic findings [4,5]. MRI is able to define structural abnormalities and is a valuable technique for noninvasive assessment and quantification of shunt volume [5]. Shunt volume is expressed as a ratio of the pulmonary blood flow versus the systemic blood flow (Qp/Qs). The criterion standard for quantifying shunt volume (Qp/Qs) is the analysis of the flow through the pulmonary arteries in relation to the systemic arterial system during left and right heart catheterization (e.g., by Fick equation) [6]. An oxygen saturation assessment at all right heart locations should be performed in cases of suspected

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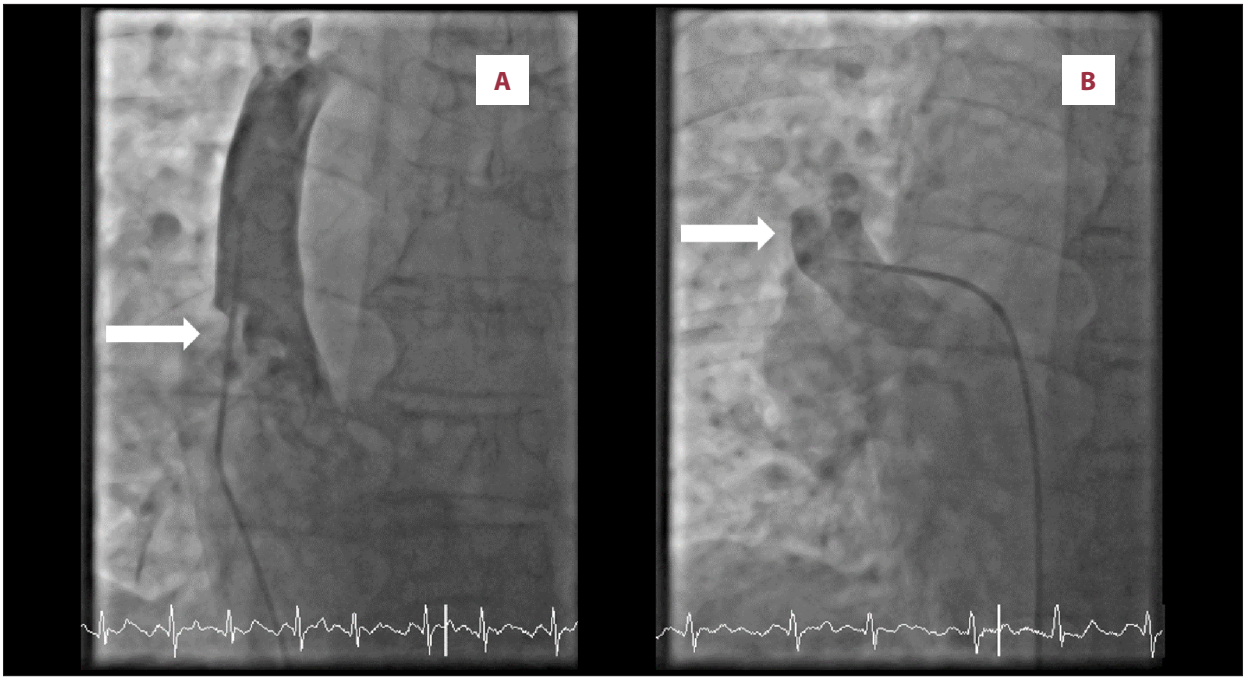

Figure 3. Angiogram of the superior vena cava with turbulent flow into the superior vena cava (arrow) (**A**) and the anomalous pulmonary venous return into the superior vena cava (arrow) (**B**).

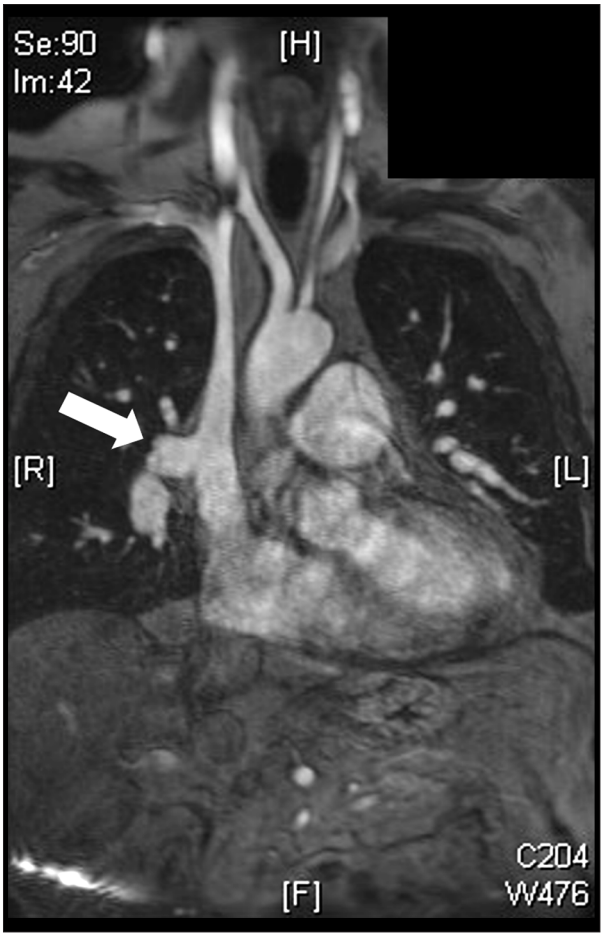

Figure 4. Contrast-enhanced MRI scan of the chest showing the anomalous pulmonary venous return of the right upper and middle pulmonary veins into the superior vena cava (arrow).

left-to-right shunt and in cases with a pulmonary arterial oxygen saturation >75% [6]. Surgery may be considered in patients with a predominant left-to-right shunt. The criteria for shunt closure depend on baseline pulmonary vascular resistance. If the pulmonary vascular resistance is <2.3 WU, the congenital heart disease is considered correctable [7,8]. Further criteria include the type of the defect, the age of the patient, the pulmonary vasculare resistance/systemic vascular resitance ratio (PVR/SVR ratio), and the Qp/Qs ratio [9]. Surgical repair of PAPVR is generally the best treatment option for correcting such complex shunt vitium [10]. Surgical techniques for treatment of this shunt vitium are continuously being revised and improved [11–13]. In patients with severe elevated pulmonary vascular resistance, cardiopulmonary transplantation is the only curative therapy [6].

Conclusions

PAPVR is often concealed and patients are misclassified as suffering from idiopathic PAH. Awareness of the disease is important, as early detection potentially leads to surgical therapeutic options.

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Figure 2 was produced using Servier Medical Art.

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