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An exceptional case of coronary artery collateralization in unilateral pulmonary artery agenesis: implications for right heart failure in the elderly

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Unilateral pulmonary artery agenesis (UPAA) is a rare congenital cardiovascular anomaly defined by the complete absence of one pulmonary artery, most often affecting the right side.[1] Although typically identified in childhood due to symptomatic presentation, certain cases may remain undiagnosed until adulthood, particularly in asymptomatic patients or those with subtle clinical signs. The absence of a pulmonary artery significantly alters pulmonary hemodynamics, prompting the development of systemic collateral vessels to maintain perfusion to the affected lung. However, this compensatory mechanism often contributes to the progression of pulmonary hypertension (PH), imposing an increased hemodynamic burden on the right ventricle. This case underscores the critical role of early diagnosis and intervention in mitigating the development of *cor pulmonale*, a severe complication resulting from prolonged right heart strain in patients with UPAA.

A 68-year-old female patient presented to the cardiology clinic with complaints of worsening shortness of breath. Her medical history included diabetes mellitus, hypertension, and chronic obstructive pulmonary disease. She also reported intermittent emergency department visits for respiratory issues. During prior evaluation for chronic obstructive pulmonary disease, right pulmonary artery agenesis had been identified. Approximately one year earlier, she had been admitted to the intensive care unit for pneumonia, after which progressive dyspnea began to develop. Echocardiography revealed a significantly elevated systolic pulmonary artery pressure of 100 mmHg. Physical examination identified 2^+ pretibial edema, diminished breath sounds, and coarse crackles in the upper zones of the right lung. Vital signs included a respiratory rate of 18 breaths/min, a pulse rate of 89 beats/min, and oxygen saturation of 90%. A previous pulmonary computed tomography angiography confirmed the absence of the right pulmonary artery. Coronary angiography and right heart catheterization were performed for further assessment. Angiography revealed extensive collateral vessels originating from both the circumflex and right coronary arteries, as well as additional collaterals from the right subclavian artery (Figure 1). Right heart catheterization demonstrated combined pre- and post-capillary PH (Table 1). Initial treatment was directed at managing heart failure with preserved ejection fraction. The patient was prescribed dapagliflozin 10 mg, torasemide 20 mg, diltiazem 60 mg (2 × 1), and sacubitril/valsartan 49/51 mg (2 × 1). While edema resolved, pulmonary artery pressure showed no significant improvement. Tadalafil 20 mg (1 × 2) was subsequently initiated, yielding partial symptomatic relief. However, systolic pulmonary artery pressure remained elevated around 100 mmHg. Given the persistent PH, dual therapy was implemented by adding bosentan 62.5 mg (2 × 1). This adjustment led to clinical improvement, including reduced oxygen requirement. The patient was discharged with scheduled follow-ups in the outpatient clinic for continued monitoring and management.

UPAA is a rare congenital vascular anomaly, typically presenting as the complete absence of one pulmonary artery, most commonly the right, due to the failure of embryonic development of the proximal segment of the sixth aortic arch.[1] The estimated incidence is approximately 1 in 200,000 individuals.[2] The condition results from the regression of the proximal portion of the sixth aortic arch during embryogenesis, leading to the absence of the proximal pulmonary artery. Blood supply to the affected lung is typ-

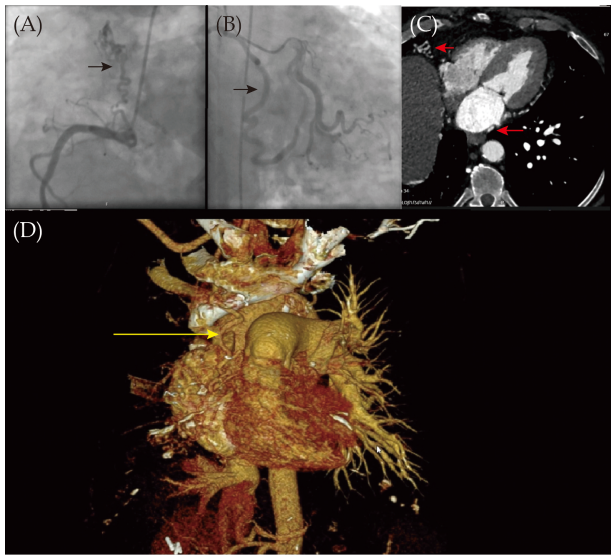

Figure 1 Coronary angiography and computed tomography. (A): Collaterals from right coronary artery; (B): collaterals from circumflex artery; (C): image of collaterals in computed tomography (red arrow); and (D): image of dense collateral networks in coronary computed tomography angiography (yellow arrow).

ically provided by systemic collaterals, although in rare cases, anomalous collaterals from coronary arteries may develop.[3] Right-sided pulmonary artery agenesis is more commonly observed and often associated with other cardiovascular anomalies.[4] Patients with UPAA may remain asymptomatic, but the condition can also manifest with recurrent pulmonary infections (37%), exercise intolerance or dyspnea (40%), hemoptysis (20%), or high-altitude pulmonary edema (10%). PH, occurring in up to 44% of cases, is a critical determinant of prognosis, contributing to right heart strain and potential failure.[5] Some patients with UPAA remain asymptomatic, and the diagnosis is often made in adulthood. Although rare, there are documented cases in the literature that present distinct clinical scenarios. The prognosis is primarily influenced by the loss of the pulmonary vascular bed on the side of agenesis and the subsequent hemodynamic burden on the right heart. Without appropriate treatment, complications such as PH, right heart failure, and severe hemoptysis can be life-threatening. The occurrence of coronary collateral circulation in the context of UPAA is exceedingly rare and has been reported in only a limited number of cases. These collaterals may contribute to myocardial ischemia or infarction through the "steal phenomenon", which impairs myocardial perfusion, though in some instances, they may not significantly impact coronary circulation.[6] The management of UPAA is contingent upon the patient's age, clinical presentation, and the severity of the disease. Early surgical revascularization can restore pulmonary circulation, mitigating potential complications.[7] Surgical options such as lobectomy or pneumonectomy may be considered for patients with severe hemoptysis or recurrent infections.[8] Asymptomatic adults should undergo regular monitoring for the development of PH. Medical treatment typically focuses on controlling PH and may involve the use of calcium channel blockers, sildenafil, and endothelin receptor antagonists.[4,5,9] Early diagnosis and prompt, tailored management are critical for improving the patient's quality of life. Considering the patient's age and comorbid conditions, the presence of a phenotype that predisposes to diastolic dysfunction may explain the worsening of symptoms over time. The development of this cl-

Table 1 The patient's findings from echocardiography and right heart catheterization.

Patient's findings		
Echocardiography		
Systolic pulmonary artery pressure	100 mmHg	Right ventricular/Left ventricular > 1 Right atrial area: 24 cm^2
Tricuspid regurgitation velocity	4.7 m^2	Tricuspid annular plane systolic excursion: 13 Tricuspid lateral systolic annular velocity: 11 m/s
Severity of tricuspid regurgitation	Severe	
Severity of mitral regurgitation	Mild	
Ejection fraction	56%	
Right-left heart catheterization and coronary angiography (collateral vessels originating from the osteal portion of the right coronary artery and circumflex to the pulmonary artery)		
Left ventricular end-diastolic pressure	20 mmHg	
Pulmonary vascular resistance	9 wood	
Pulmonary capillary wedge pressure	21 mmHg	
Pulmonary artery pressure	103/56 (72) mmHg	
Combined pre-post capillary pulmonary hypertension		

inical picture may be attributed to a hyperdynamic state induced by collateral circulation, or alternatively, it may reflect the progressive nature of the underlying disease itself.[10] Further research is needed to elucidate the precise mechanisms driving symptom exacerbation in these patients.

The early recognition of this condition is critical for ensuring timely intervention and optimizing patient management. The development of collateral circulation in affected individuals is an important aspect of this disorder. While these collaterals may not always exhibit significant clinical consequences, they can complicate both diagnostic processes and therapeutic approaches. This complexity in management underscores the need for a nuanced understanding of the role of collaterals in UPAA, as they may present additional challenges in the clinical course despite their potential lack of immediate clinical relevance.

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