

## Echocardiographic Reverse Flow Pattern Seen in the Distal Segment of Left Anterior Descending Coronary Artery: An Important Finding in Anomalous Left Coronary Artery from Pulmonary Artery Syndrome

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### Introduction

The anomalous left coronary artery from the pulmonary artery is a rare disease. Therefore, it is a major congenital coronary anomaly. The clinical course depends on the formation and development of collateral arteries. The pathophysiology is explained by the low pressure in the pulmonary arterial system, with the right coronary artery (RCA) filling the left coronary artery in a retrograde manner through collateral arteries, working as a phenomenon of coronary flow theft. This study reports a case in which another finding was observed. The flow pattern in the left coronary artery in its distal segment evaluated by pulsed Doppler confirms the diagnosis confirmed by angiography.

### Case Report

Male 32-year-old patient complaining of fatigue on exertion since childhood when exercising, especially when playing soccer, and currently with palpitations not related to exertion, was referred to Doppler echocardiography, which revealed increased left ventricular diameters with mild ventricular dysfunction and hypokinesia of apical segments. Ejection fraction calculated by the Simpson method was 48% (Figure 1A). Presence of thickened mitral valve with mild regurgitation (Figure 1B). Presence of right coronary artery dilatation (1.0 cm) in the transverse section of the aorta, as shown in Figure 1 C. Systolic-diastolic pulmonary artery flow in the transverse section of the aorta (Figure 1D), in addition to the finding of systolic-diastolic reverse flow with diastolic predominance, velocity around 1m/s in the anterior descending artery in the distal segment acquired in apical modified two-chamber view (Figures 2A and B) showing open vessel without obstruction. The combination of the findings described above suggests the diagnosis of anomalous origin of the left coronary artery from the pulmonary artery.

### Keywords

Coronary Vessel Anomalies; Congenital Abnormalities; Echocardiography; Pulmonary Artery/abnormalities; Syndrome.

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Patient referred to coronary angiography, in which the diagnosis was confirmed (Figures 3 and 4). After the tests, the patient underwent cardiac surgery for a successful correction of the anomaly.

### Discussion

Anomalous origin of the left from the main pulmonary artery coronary artery is a major congenital anomaly, considered the most common defect of coronary anomalies in childhood associated with ischemia and myocardial infarction. It affects one in every 300,000 live births<sup>1</sup>. It was described in 1886 by Brooks and then in 1908 by Abbott. However, it was officially confirmed in 1933 with a case of a child who presented acute myocardial infarction confirmed in autopsy by Bland, Garland and White<sup>2</sup>. This anomaly may occur alone or associated with other lesions such as patent ductus arteriosus (PDA), atrial septal defect or ventricular septal defect<sup>3,4</sup>. Associated lesions are important because they increase the pulmonary artery pressure, maintaining coronary perfusion. The literature presents cases of patients who had the PDA closed and died because they had no diagnosis of anomalous origin of the left coronary artery from the pulmonary artery associated<sup>5</sup>. Diagnosis is not simple and requires a high clinical suspicion.

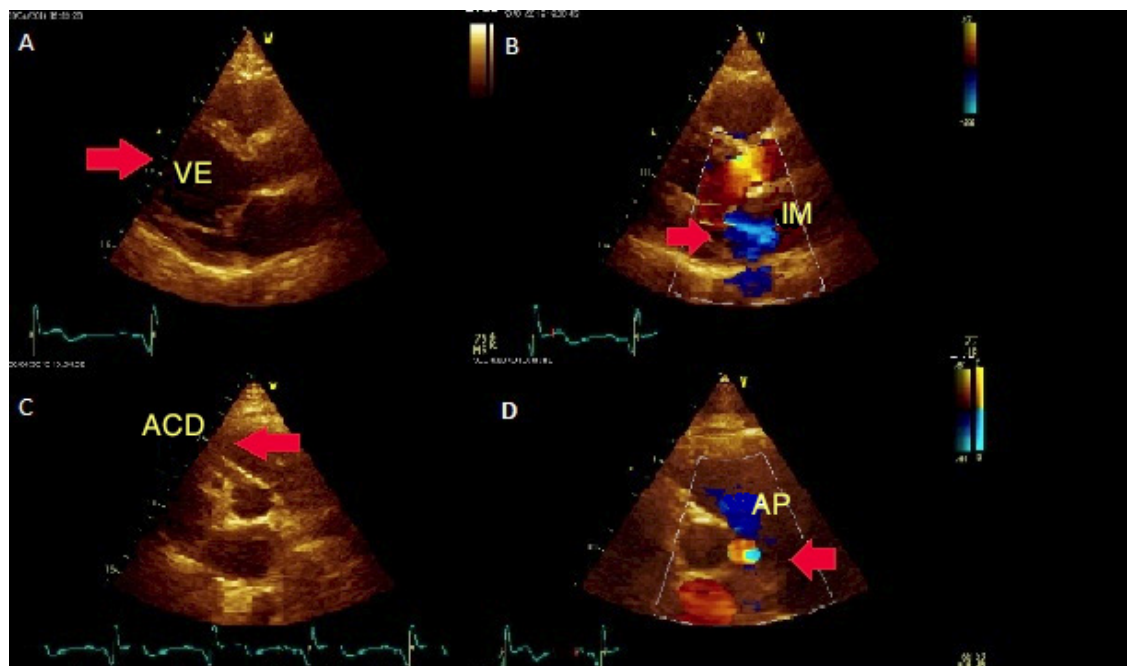
The pathophysiology of this disease is explained by the decrease in pulmonary vascular resistance and the left anomalous coronary artery with origin in the pulmonary artery works as a vessel draining the myocardial blood instead of supplying it. The clinical evolution of these patients will depend on the formation of collateral circulation. If the collateral arteries are not enough, the patient will present segmental abnormalities, myocardial infarction, mitral valve abnormalities and ventricular aneurysms that may be related to cases of sudden death. As a result, most patients die in early childhood<sup>6</sup>. They need to have an extensive set of collateral arteries to allow them to survive.

Differential diagnoses include Kawasaki disease, dilated cardiomyopathy, unexplained mitral regurgitation, endomyocardial fibrosis, coronary fistula, vasculitis (polyarteritis nodosa, Takayasu arteritis)<sup>7</sup>.

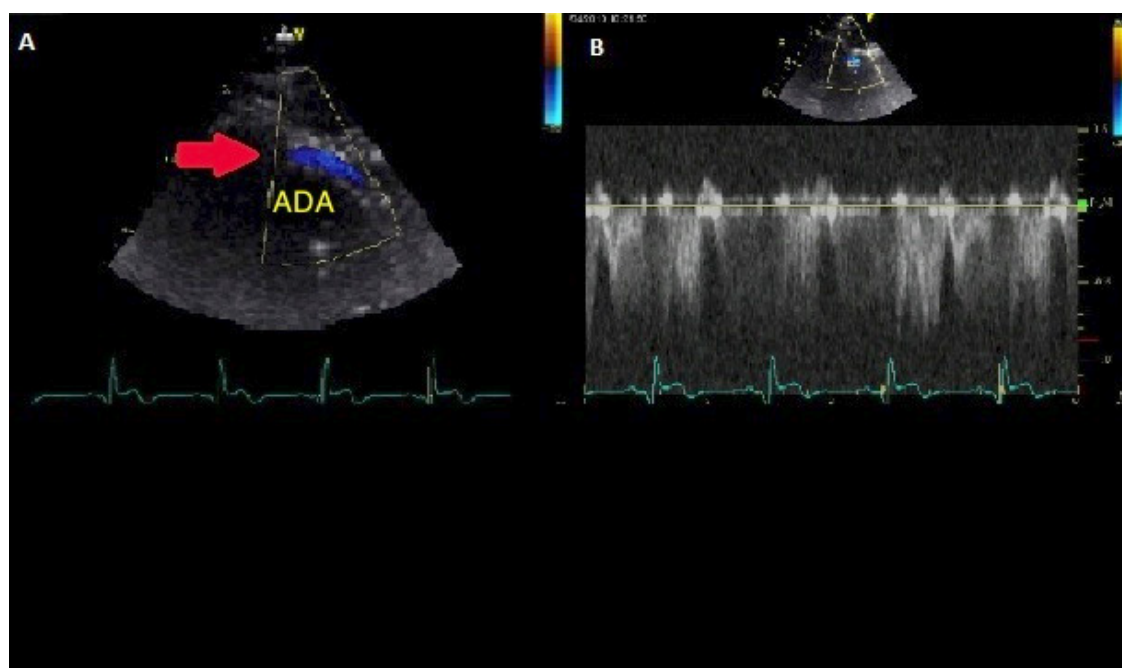
Coronary angiography usually establishes diagnosis as it directly shows the left coronary artery emerging from the pulmonary trunk, but it is not a risk-free invasive method<sup>8</sup>.

Doppler echocardiography plays a key role in the suspicion of this syndrome, since it is able to demonstrate myocardial

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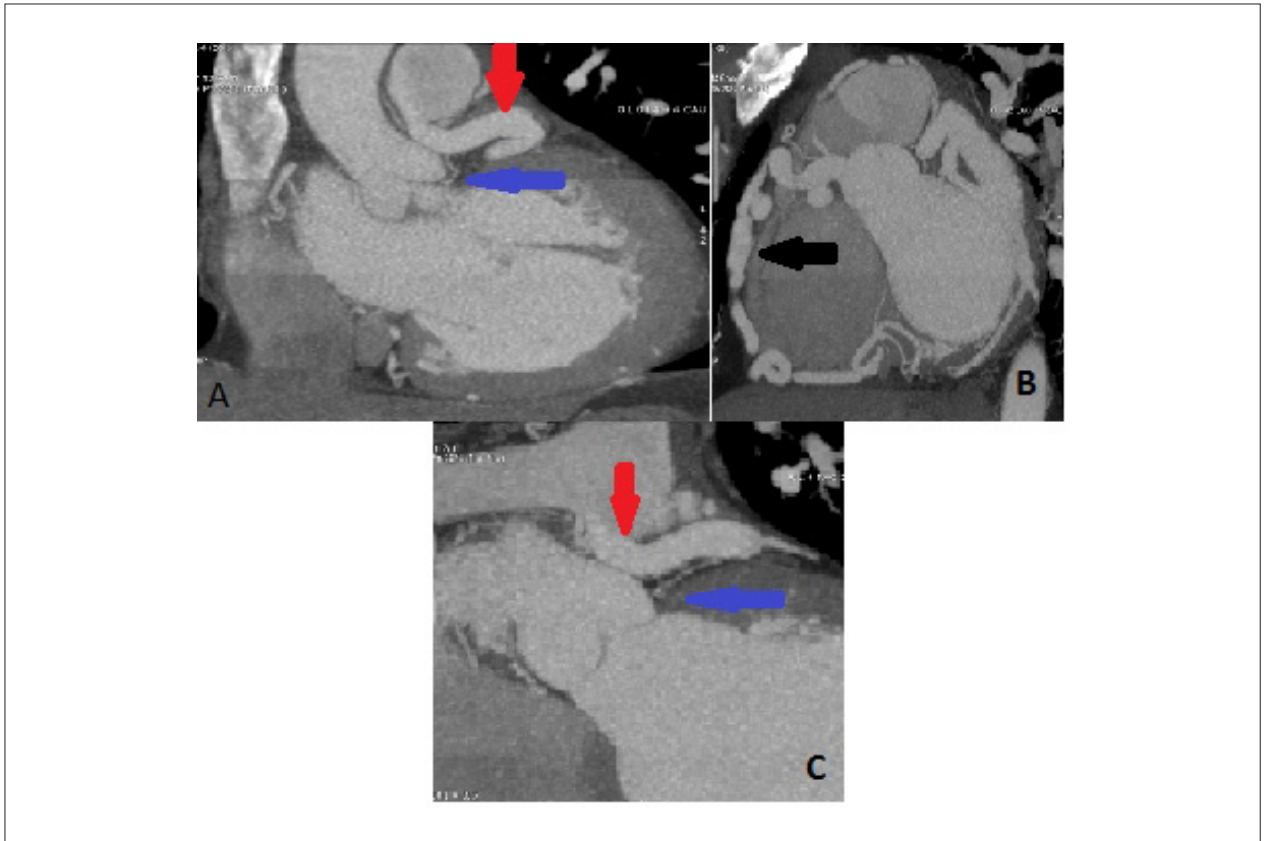
**Figure 1** - Echocardiographic findings in the anomalous origin of the left coronary artery from the pulmonary artery showing in figure A: Doppler echocardiography demonstrating increased LV cavity diameters in the longitudinal parasternal section (arrow); B: Mild mitral regurgitation viewed in longitudinal parasternal section (arrow); C: Dilated right coronary artery (arrow); D: Presence of systolic-diastolic flow in the pulmonary artery at the aorta level in the transverse section of the aorta (arrow). LV: Left ventricle; MR: Mitral regurgitation; RCA: Right coronary artery; PA: Pulmonary artery.



**Figure 2** - Retrograde flow in the distal segment of the Anterior Descending Artery (ADA) in apical modified two-chamber view by color Doppler (red arrow) in A and pulsed Doppler in B.



**Figure 3** - Coronary CT angiography with volume rendering. Tortuous and hypertrophic right coronary artery (blue arrow) and major collateral arteries to the left coronary artery (black arrow).



**Figure 4** - A, B and C - MIP (maximum intensity projection) and oblique reformations of coronary CT angiography. The left coronary artery comes directly from the pulmonary artery (red arrow) and does not communicate with the sinus of Valsalva (blue arrow). Hypertrophic and tortuous right coronary artery (black arrow).

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segmental abnormalities, contractile dysfunction with increased left ventricular diameters, right coronary artery dilatation, mitral valve abnormalities due to papillary muscle ischemia with increased echogenicity, and may progress to rupture. Through the pulsed Doppler, it is possible to see low-velocity systolic-diastolic flow in the pulmonary artery trunk in the transverse section of the aorta and reverse flow in the anterior descending artery in its distal segment acquired in apical modified two-chamber view with diastolic predominance, where the latter is one more finding to support the diagnosis of anomalous origin of the left coronary artery from the pulmonary artery.

The evolution of multislice computed tomography, due to the ability to acquire and reconstruct three-dimensional images provided an alternative tool to confirm the syndrome. Since it is non-invasive, it is capable of demonstrating the anatomy of the coronary arteries with high diagnostic accuracy<sup>9,10</sup>.

Therefore, the study of coronary blood flows through Doppler echocardiography should be recommended for all cardiomyopathies without an explainable cause in children and young adults, since it is a simple noninvasive test that does not require the use of contrast for the diagnosis of coronary anomalies.

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