# Pulmonary artery from the left main coronary artery

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#### **ABSTRACT**

The usual sources of pulmonary blood flow in pulmonary atresia (PA) with(VSD) are patent ductus arteriosus and aortopulmonary collaterals. However, rarely fistulous collaterals may also arise from the coronary arteries which usually open into the main pulmonary trunk or branch pulmonary arteries. In such cases, selective coronary angiogram may be required for the demonstration of pulmonary arterial anatomy. A case of PA with VSD with failure to demonstrate pulmonary arteries on routine catheterization study (ventricular, aortic root, and descending aortic angiograms) is being presented here. A coronary artery-to-pulmonary artery fistula was suspected in view of dilated left main coronary artery, and pulmonary arteries were well demonstrated with selective coronary angiogram.

**Keywords:** Collateral circulation, coronary-pulmonary artery fistula, ventricular septal defect with pulmonary atresia

### **CASE REPORT**

A 15-year-old girl presented with deep cyanosis and easy fatigability without preceding a history of recurrent respiratory tract infections or cyanotic spells. Physical examination revealed a right ventricular apex and absence of cardiac murmur. A diagnosis of pulmonary atresia (PA) with ventricular septal defect (VSD) was confirmed with transthoracic echocardiography. Corrective surgery was planned after a preoperative catheterization study. In the catheterization laboratory, pulmonary arteries could not be profiled after right ventricular, left ventricular, aortic root, and descending thoracic aortic angiograms. However, a closer inspection of the aortic root angiogram revealed a dilated left main coronary artery. The left main coronary artery was hooked selectively with a Judkins left 4-curve catheter. Selective left injection revealed confluent good-sized pulmonary arteries arising from the left main coronary artery through a tortuous coronary-to-pulmonary artery fistula (CPAF) [Figure 1 and Video 1]. Later,

the girl underwent successful intracardiac repair with transannular patch and clipping of the fistula with no complications.

#### **DISCUSSION**

The usual sources of pulmonary blood flow in VSD with PA are patent ductus arteriosus and major aortopulmonary collaterals (MAPCAs). Rarely, a fistulous connection between coronary artery and main pulmonary artery is the main supply to the pulmonary blood flow. In contrast to MAPCAs which usually connect to branch pulmonary artery at the hilum or inside lung parenchyma, CPAF is intrapericardial and usually connected to the main pulmonary trunk through a tortuous channel. [1] To the best of our knowledge, only 52 cases of CPAF have been reported in the literature. However, in a case series of 87 patients with VSD with PA, Amin *et al.* reported an incidence of 10% (9 patients) and suggested commoner occurrence and emphasized the importance of routine ascending aortogram to avoid missing the

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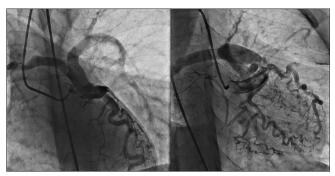


Figure 1: Selective angiogram of the left coronary artery with Judkins left catheter demonstrates filling of confluent good-sized pulmonary arteries through a tortuous coronary to pulmonary artery fistula in the anteroposterior view with cranial tilt (left side of the image) and right anterior oblique view with caudal tilt (right side of the image)

diagnosis.<sup>[2]</sup> CPAF most commonly arise from the left coronary artery (65%), followed by the right coronary artery (23%) and single coronary artery (5%). Rarely, they may arise from both the coronary arteries (2%).<sup>[3]</sup> Additional MAPCAs may also be present along with the CPAF. Intuitively, CPAF may result in coronary steal phenomenon. However, coronary steal is extremely rare due to compensatory dilatation and increase in flow in the proximal coronary artery which maintains both the pulmonary and the coronary blood flow.<sup>[3]</sup> Typically, the fistulous connection is restrictive, and there is no significant elevation of the pulmonary pressures.<sup>[1]</sup>

Surgical management entails clipping the fistula along with the usual intracardiac repair with or without a conduit.

To conclude, the possibility of a CPAF should always be borne in mind with failure to demonstrate pulmonary blood flow during a routine catheterization study in VSD with PA and may require selective coronary angiogram to demonstrate the CPAF.

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#### Conflicts of interest

There are no conflicts of interest.

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