



# Coronary Artery Fistula Unmasking the Absence of Left Pulmonary Artery in an Adult

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## Abstract

Coronary artery fistulae are an incidental finding in patients undergoing coronary angiography or computed tomography (CT) coronary angiography. A 60-year-old man with known coronary artery disease presented with dyspnea. Coronary angiography revealed a large fistula arising from the circumflex artery (CX) without a clear intrathoracic target vessel or chamber in the heart. CT angiography revealed the agenesis of the left pulmonary artery. The fistula arising from the CX ensured left lung tissue supply. Unilateral absence of a pulmonary artery is an extremely rare condition. In this case, the identification of a fistula from the heart triggered the correct diagnosis.

## Keywords

- ▶ congenital defect
- ▶ myocardial infarction
- ▶ pulmonary arteries

## Introduction

Coronary artery fistulae are usually abnormal connections between a coronary artery and a cardiac chamber or major intrathoracic vessels with substantially lower pressure. Most coronary fistulae are congenital, but they can also be acquired. In the largest series of cases undergoing coronary angiography, relevant coronary fistulae were found in 62 out of 126,595 cases (0.05%).<sup>1</sup> More recent studies by computed tomography (CT) coronary angiography, screening 15,548, 8,864, and 2,573 patients found higher incidences of 0.19, 0.15, and 0.15%, respectively.<sup>2–4</sup> Most patients are asymptomatic. The most common findings in symptomatic patients are heart failure resulting from left to right shunting, ischemia due to coronary steal, arrhythmia, rupture, thrombosis, and infective endocarditis.<sup>5</sup>

## Case Description

A 60-year-old man suffered a non-ST-segment elevation myocardial infarction at the age of 49 years (year 2009) and underwent interventional treatment of the left anterior

descending artery (LAD, bare metal stent implantation) and mechanical recanalization (balloon only) of the ramus intermedius. At this time, intervention via the femoral route was challenging because of difficulty to selectively intubate the ostium of the left coronary artery and challenging wire passage into the LAD. A large coronary fistula (▶ Fig. 1, panel A) arising from the circumflex artery (CX) had already been described, but at that time, no further action had been taken. Now, more than 10 years later, the patient returned with exertional dyspnea and again underwent coronary angiography. LAD and ramus intermedius showed intermediate lesions without any further progress of his coronary artery disease. Since he was symptomatic, but his lesions did not appear flow limiting, interventional closure of the fistula was considered, but a CT angiography of the chest was performed to further evaluate the fistula and its target vessel or chamber. His chest CT showed a smaller left and larger right hemithorax together with fibrotic and emphysematous changes of the left lung (▶ Fig. 1, panel B). The underlying cause was an agenesis of the left pulmonary artery (▶ Fig. 1, panel B). Collaterals from the heart (fistula coming from the CX), the left and right subclavian arteries, as well as the superior phrenic artery

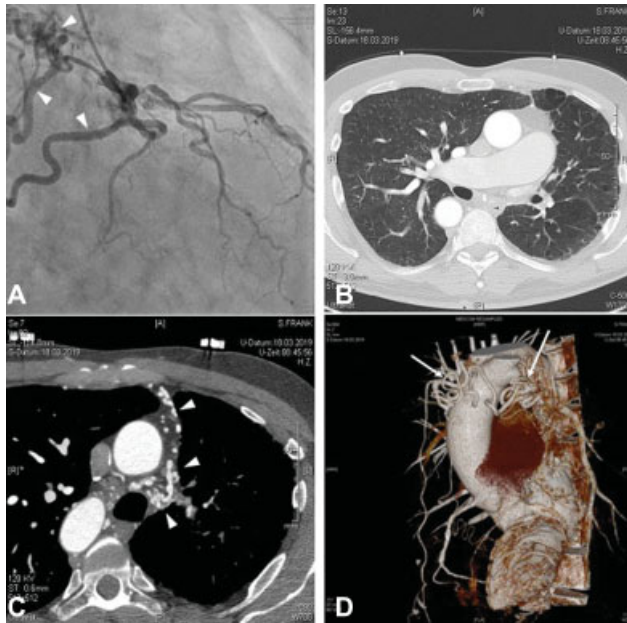
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**Fig. 1** Coronary angiography in right anterior oblique 30-degree projection showing intermediate lesions of the proximal left anterior descending artery and the ramus intermedius as well as a large “fistula” (arrow heads) taking off from the circumflex artery (panel A); CT of the chest indicating a smaller left hemithorax and the absence of the left pulmonary artery (panel B); CT angiography showing the right descending aorta and multiple collaterals (arrow heads) close to the ascending aorta (panel C); and three-dimensional reconstruction of the intrathoracic structures with large collateral vessels arising from both subclavian arteries (arrows, panel D). CT, computed tomography.

(→**Fig. 1**, panels C and D) ensured left lung tissue supply. In addition, it revealed a right descending thoracic aorta. Detailed history revealed dyspnea since childhood which led to discharge from military service later in life due to hemoptysis induced by maximum strength exercise. The decision for conservative treatment was made.

## Discussion

Unilateral absence of a pulmonary artery (UAPA) is an extremely rare condition affecting ~1 in 250,000 persons in the general population and is mostly associated with other congenital defects and is mainly diagnosed in childhood.<sup>6–9</sup> In the studies searching for coronary anomalies in ~175,000 patients, not a single case of UAPA had been described, underlining the uniqueness of this case.<sup>1–4</sup>

Here, a large fistula arising from the CX triggered CT coronary angiography and thereby the diagnosis. It showed that the “fistula” had no clear target vessel or target cardiac chamber but corresponded to collaterals ensuring tissue integrity of the left lung. Since these collaterals supplied the tissue of the left lung, an interventional closure was not indicated. In addition, his coronary lesions were not flow limiting. Thus, the patient’s dyspnea was most likely caused by the congenital defect (UAPA) and associated consequences rather than by a steal phenomenon at the level of the heart. Fistulae arising from the coronary tree which do not have a clear target vessel or target cardiac chamber should therefore be evaluated by CT coronary angiography before considering interventional or surgical closure.

## Conflict of Interest

None.

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