

## Case Report

## Bilateral Fistulas: A Rare Cause of Chest Pain. Case Report with Literature Review

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## Key words:

**Bilateral fistulas,  
angina, chest pain.**

A coronary artery fistula is a direct communication between a coronary artery and one of the cardiac chambers or vessels around the heart. These fistulas are usually diagnosed by coronary arteriography. Clinical presentations are variable depending on the type of fistula, shunt volume, site of the shunt, and presence of other cardiac conditions. Bilateral coronary fistulas between coronary arteries and the pulmonary artery are very rare. This report describes a 51-year-old man without any previous medical history, who presented to our hospital one hour after the acute onset of severe substernal chest pain associated with shortness of breath and nausea. Coronary angiography revealed two fistulas arising from the left anterior descending and right coronary arteries and draining at exactly the same site in the pulmonary artery. There was no evidence of atherosclerotic coronary artery disease in either the left or right coronary arterial tree.

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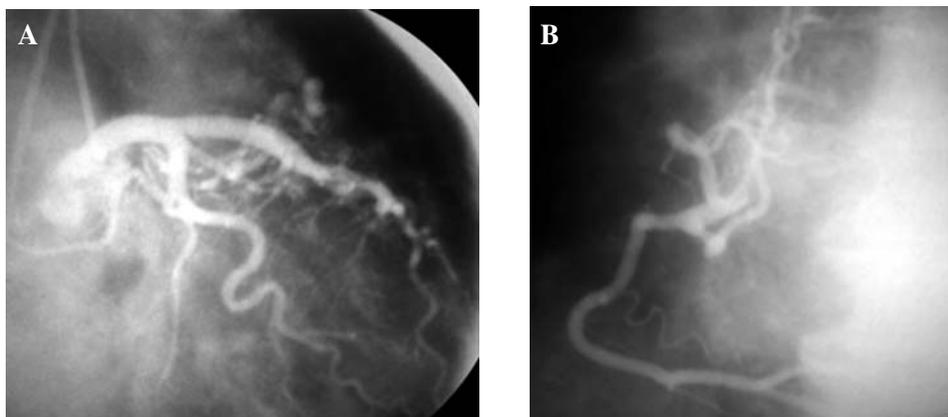
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**C**oronary artery fistulas are rare congenital or acquired coronary artery anomalies that can originate from any of the three major coronary arteries and drain into any of the cardiac chambers and great vessels. The incidence is 0.13% of congenital cardiac lesions, while its incidence in the overall population is estimated to be about 0.002%.<sup>1,2</sup> Over 90% of these fistulas drain into the systemic venous side of the circulation. Drainage of the fistula into the pulmonary trunk has been reported in 17% of cases.<sup>3</sup> To the best of our knowledge, a connection of both the left anterior descending artery (LAD) and the right coronary artery (RCA) to the main pulmonary artery has very rarely been reported as a case in the literature.

### Case presentation

A 51-year-old man without any previous medical history presented to our hospital one hour after the acute onset of severe substernal chest pain, which was associat-

ed with shortness of breath and nausea. The patient denied chest trauma, fever or chills. The ECG showed 2 mm ST elevation in leads D<sub>2</sub>, D<sub>3</sub>, aVF. Maximal creatine kinase elevation was 1500 IU at 24 hours after the onset of clinical symptoms. He was treated with aspirin, nitroglycerin, and low molecular weight heparin with resolution of his chest pain. In view of the resolution of chest pain and the decrease of ST-segment elevation fibrinolytic therapy was not administered and the patient was transferred to the catheterisation laboratory for further management. Selective coronary angiography revealed two fistulas arising from the LAD and RCA and draining into the pulmonary artery, without evidence of atherosclerotic coronary artery disease in either the left or right coronary arterial tree (Figure 1). Right heart catheterisation was performed and multiple blood samples were obtained to calculate the magnitude of the left-to-right shunt. There was no significant step-up of oxygen saturation in the right heart and pulmonary arteries, and



**Figure 1.** Bilateral fistulas from the left anterior descending artery (A) and right coronary artery (B) draining into the pulmonary artery.

pulmonary artery pressure was normal. Exercise stress showed no myocardial perfusion abnormality that might be due to a steal phenomenon. Because of the insignificant left-to-right shunt and normal myocardial perfusion, interventional therapies such as transcatheter embolisation or surgical closure were not recommended, and the patient was treated with the usual medical therapy (nitrates, aspirin, and beta-blockers). He is still under follow up without any cardiac event.

## Discussion

Coronary arteriovenous fistula (CAF) is a very uncommon entity. In a review<sup>4</sup> of 363 cases with CAF, 50% of the fistulas were found to arise from the RCA, 42% from the left coronary artery, and 5% from both coronary arteries. Approximately 5% of coronary arteriovenous fistulas are bilateral. The most common site of drainage is the right ventricle (41%), followed by the right atrium (26%) and the pulmonary artery (17%).<sup>4</sup> Approximately 56% of the bilateral fistulas and only 17% of the unilateral fistulas have been found to drain into the pulmonary artery.<sup>5</sup> Multiple coronary-pulmonary artery fistulas have also been reported. Abhyankar reported a case of a patient with non-exertional chest pain and a normal nuclear stress test who had 3 coronary artery fistulas arising from the LAD, 2 from the left circumflex artery and 2 from the RCA, all of which drained into the main pulmonary artery, without concomitant coronary artery disease.<sup>6</sup>

Patients with CAF may have symptoms of pulmonary congestion due to left-to-right shunt, or they

may be asymptomatic. Complications such as congestive heart failure, infective endocarditis, and myocardial infarction have also been reported.<sup>4,7</sup>

The management of CAF is controversial, particularly in patients with small shunts. Liberthson<sup>7</sup> reviewed 187 cases and found that operative mortality (1%) and complication rates (7%) were low if patients underwent operation below the age of 20 years. However, in those older than 20 years at operation, significant postoperative mortality (7%), post-operative myocardial infarction (7%) and other complications (23%) occurred. The authors recommended surgery in all cases of CAF in childhood irrespective of symptoms or size of the shunt.<sup>8-12</sup>

A new therapeutic option for the treatment of CAF is the percutaneous transcatheter embolic occlusion or transcatheter closure technique, which has been advocated as a minimally invasive alternative to surgery. A variety of materials have been used, including Gianturco coils, covered stainless steel coils, detachable balloons, coaxial embolisation with platinum microcoils, double umbrella device, and the Gianturco Grifka vascular occlusion device (as cited by Armsby et al,<sup>13</sup> Dorros et al,<sup>14</sup> Reidy et al,<sup>15</sup> and Mavroudis et al<sup>8</sup>).

The feasibility of closure by a device was determined by the number and location of drainage sites, the ability to cannulate the distal fistula, and the proximity of coronary branches to the optimal occlusion site. Special care should be taken to avoid allowing the device to interfere with flow into any visible coronary artery. The selection of occlusion device was based on the anatomical features of the fistulas.<sup>9</sup> Coils have been used in smaller CAF and the double umbrella

device in larger CAF. Device deployment has been either antegrade (via the femoral vein) or retrograde (via the femoral artery).<sup>16</sup>

In conclusion, with increased experience and improved devices and techniques transcatheter closure of CAF is emerging as a successful therapeutic strategy. The preferred method of approach for any individual patient will depend on the anatomy of the fistula, the presence or absence of associated defects, and the experience of the interventional cardiologists and surgeons.

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