

# Congenital Circumflex Coronary Arteriovenous Fistula

with Aneurysmal Termination in the Pulmonary Artery

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Coronary arteriovenous fistula is a rare congenital anomaly that is seen in 0.1% to 0.2% of coronary angiograms. Aneurysmal formation in the fistula is even rarer. We report a case of congenital circumflex arteriovenous fistula with aneurysmal formation just near its termination in the pulmonary artery, associated with atherosclerotic left anterior descending coronary artery. The anomaly was successfully repaired. (*Tex Heart Inst J* 2005;32:56-9)

**A** congenital coronary arteriovenous fistula is a direct communication between a coronary artery and the lumen of any of the cardiac chambers, the coronary sinus, the pulmonary artery, or the superior vena cava. The number, origin, and course of the coronary arteries are otherwise normal.<sup>1,2</sup> It is observed in 0.1% to 0.2% of coronary angiographic studies.<sup>3</sup>

## Case Report

In May 2004, a 51-year-old man, a heavy smoker with a history of dyslipidemia and a family history positive for ischemic heart disease, had sudden retrosternal chest pain, with nausea and sweating, for which he was brought to a nearby hospital. Electrocardiographic findings were consistent with anterior myocardial infarction. The patient was admitted, received the necessary treatment for myocardial infarction, improved, and was discharged. Subsequent coronary angiography showed a total occlusion of the mid left anterior descending artery and a circumflex-to-pulmonary artery fistula with aneurysmal dilatation at the distal end of the fistula (Fig. 1). The patient's ejection fraction was 0.35.

The patient presented to our department 5 months after the incident, for management of his condition. His only complaint was easy fatigability, and he reported no chest pain or other symptoms. His examination was unremarkable except for a loud S<sub>2</sub> sound in the pulmonary area upon auscultation. His electrocardiographic and chest radiographic findings were within normal limits. The results of his laboratory tests were all normal, except for elevated low-density lipoprotein and cholesterol levels.

Transthoracic and Doppler echocardiography confirmed the presence of the fistulous tract and its opening into the pulmonary artery, together with aneurysmal dilatation at the end of the fistula.

Surgery was undertaken. Intraoperative transesophageal echocardiography delineated the fistula and showed no other tracts or abnormalities. The aneurysm in the distal end of a tortuous fistula was immediately seen upon opening the pericardium (Fig. 2A). When we followed the fistula tract, we found that it originated in the proximal third of the circumflex artery and extended 2 cm to the aneurysm, which was 1 × 2 cm and fusiform. Due to the difficult location of the fistula and the presence of the aneurysm, the operation was performed with the patient on cardiopulmonary bypass. Myocardial protection was achieved with blood cardioplegic solution administered antegradely, with systemic hypothermia of 32 °C and local cooling. The fistula was approached via a pulmonary arteriotomy. The proximal end was seen to be an opening above the anterior pulmonary cusp (Fig. 2B). After closure of the fistula under direct vision, we opened the aneurysm along its

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length. Revascularization was then performed in the usual manner, by means of a saphenous vein graft to the circumflex artery and a left internal mammary artery graft to the left anterior descending coronary artery.



**Fig. 1** The fistula arises from the side of the circumflex coronary artery, follows a tortuous pathway, and terminates in an aneurysmal dilatation just above its entrance into the pulmonary artery.



**Fig. 2** Intraoperative views show **A)** the fistula's aneurysmal dilatation above the entrance to the pulmonary artery and **B)** the terminus of the fistula after the pulmonary artery has been opened.

Postoperatively, the patient had an uneventful outcome and was discharged from the hospital in good general health. Follow-up echocardiography showed no fistula.

## Discussion

In the general population, coronary artery abnormalities occur in 1% to 2% of people. They are classified into 3 categories: abnormalities of origin, distribution, and termination. Coronary artery fistulae are considered to be termination abnormalities.<sup>4</sup> They are uncommon and are seen in only 0.1% to 0.2% of coronary angiograms. In the majority of reported cases, coronary fistulae were found to originate from the right coronary artery (52% of cases) and to drain into the right ventricle (40% of cases). Fistulae originated from the circumflex artery in 18% of cases and drained into the pulmonary artery in 17% of cases.<sup>3,4</sup>

Clinically, patients with coronary fistulae may present with palpitations or with symptoms of angina, heart failure, or bacterial endocarditis; or they may be totally asymptomatic. There might also be an incidental finding of continuous murmur, characteristically heard over the left sternal border and at the apex.<sup>5</sup> Two-dimensional echocardiography is important in the diagnosis of fistula, and transesophageal echocardiography is superior to transthoracic echocardiography in delineating its characteristics.<sup>6</sup> Definitive diagnosis and planning for treatment can be done with the aid of cardiac catheterization.<sup>1,7</sup>

There has been general agreement that all symptomatic patients with coronary fistulae should undergo closure of the fistulae as soon as diagnosis is made, due to the increased morbidity and mortality associated with delay.

Our patient presented with myocardial infarction, and the discovery of the fistula was purely incidental during 2-dimensional echocardiographic investigation. Confirmation was obtained by cardiac catheterization and transesophageal echocardiography. We believe that most of our patient's symptoms were caused by atherosclerotic occlusion of the left anterior descending artery. This was evident from the presence of good distal flow along the circumflex artery, in spite of the large fistulous tract seen during cardiac catheterization.

Closure of congenital fistulae can be managed by either operative or nonoperative techniques. Nonoperative closure has been accomplished by transcatheter embolization of detachable balloons, platinum microcoils, and steel coils.<sup>1,7</sup> This method could not be applied in our patient due to the presence of an associated lesion of the left anterior descending coronary artery. Surgical ligation of the fistula should be done at its entrance within the recipient cavity, whether that cavity be a cardiac chamber or a vessel. It is generally agreed that bypass is indicated when the fistula is relatively inaccessible (such as in the distribution of the circumflex or distal right coronary artery), when the

fistula is in the course of the coronary artery rather than at its termination, and when an aneurysm requires excision.<sup>1,7,9</sup>

As a result of our findings, we decided to perform surgery using cardiopulmonary bypass.

We believe that congenital coronary fistula can be corrected easily with surgery, especially when it is associated with concomitant disease affecting other vessels. This should be done using cardiopulmonary bypass, because CPB provides excellent results with no apparent mortality or morbidity.

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## Editorial Comment

### Are All Fistulae Worth Closing?

The above report by Darwazah and colleagues mentions the incidental discovery of a small coronary fistula arising from the circumflex and draining into the pulmonary artery, in a patient with fixed coronary artery disease. The fistula was ligated at the time of the surgery, which was indicated by unrelated, apparent coronary obstructive disease. We must emphasize that the indication for surgical or catheter-mediated closure of a coronary fistula should be spe-

cific rather than generic (it is not true that any symptomatic patient who carries a fistula should have it ligated). While there may be borderline cases in which the significance of the fistula is unclear, we can summarize the acceptable indications for intervention to close a coronary fistula in the following terms:

- Ischemic symptoms or positive findings at stress testing in the dependent territory (related to steal of myocardial blood-flow);

- Aneurysmal dilatation, with or without mural thrombus of the feeding coronary artery;
- Cardiac cavity overload, due to substantial blood-shunting.

Such general guidelines are valid even in the context of an intervention (catheter-mediated or surgical) that is required for other reasons.

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