

# Coronary Artery Fistulae: 4 cases repaired surgically

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## Abstract

Coronary artery fistulae involve a communication between a coronary artery and a heart chamber or part of the pulmonary circulation. Most are asymptomatic and discovered incidentally, whilst larger ones may cause coronary steal syndrome. Fistulae may produce continuous murmurs and are diagnosed at echocardiography or angiography. Treatment is by percutaneous coil embolisation or open surgery. We review four cases treated with surgical closure. All patients were asymptomatic and diagnosed incidentally at angiography. One case involved a failed attempt at percutaneous coil embolization requiring immediate open surgery. The other three cases required other operative procedures and the fistulae were oversutured during the same procedure.

## Introduction

Coronary artery fistulae, although rare, are amongst the commonest congenital cardiac anomalies. They involve a communication between a coronary artery and a heart chamber or part of the pulmonary circulation. Most are asymptomatic and discovered incidentally. Larger ones may cause coronary steal syndrome, as blood is shunted from the coronary artery to the ventricle or pulmonary circulation, bypassing the myocardium, and causing symptoms of angina, arrhythmias or high-output heart failure.<sup>1</sup>

First described in 1908 by Maude Abbott, fistulae can be classified as a congenital abnormality of termination<sup>2</sup>. Embryologically coronary artery fistulae are thought to arise as a persistence of sinusoidal connections between the lumens of the primitive tubular heart that supply myocardial blood flow in the early embryologic period. Most coronary artery fistulae arise from the right coronary artery (40-60%) and those arising from the left coronary artery are predominantly from the left anterior descending artery. The right atrium or ventricle and pulmonary artery are the site of termination in 90% of cases.

Small fistulae may remain clinically silent and are recognized at routine angiography, echocardiography or autopsy. In small fistulae, the myocardial blood supply is not sufficiently compromised to cause symptoms. Although spontaneous closure usually occurs, some can dilate over time. The increased flow in the feeding artery may result in dilatation resulting in the commonly associated finding of coronary aneurysms. Fistulae may give rise to various complications, the commonest being myocardial ischaemia, including ischaemic cardiomyopathy, papillary muscle rupture from chronic ischaemia and congestive cardiac failure from volume overload. Older patients may present with signs of congestive heart failure, arrhythmias, syncope or chest pain. Bacterial endocarditis and sudden cardiac death have also been described<sup>3</sup>. Rare cases are described of multiple microfistulae causing angina. The management of such fistulae is with antianginal medication and risk factor control. However these reports are rare and no evidence based management is offered in the literature<sup>4</sup>.

## Case reports

The four cases described were discovered incidentally on angiography and were treated surgically.

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## Case Report

One followed a failed attempt at coil embolisation and open surgery was performed urgently to retrieve the coil. The other three were repaired electively as the patients required coronary bypass grafting or valve replacement. All cases were performed on cardiopulmonary bypass. Two of the cases presented with multiple fistulae.

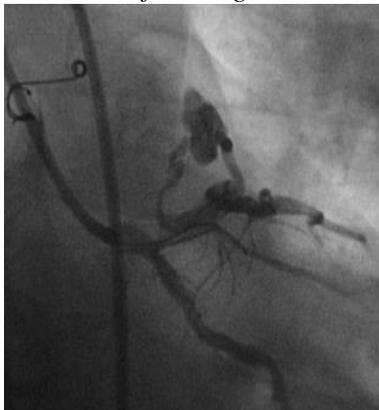
### Case 1

A 49 year-old male was investigated for syncope. Exercise stress testing was positive and the patient underwent coronary angiography. He was found to have two fistulae, one arising from the ostium of the right coronary artery (RCA), draining into the right pulmonary artery (PA) (fig 1) and the other arising from the left anterior descending artery (LAD), bifurcating into two vessels both draining into the pulmonary trunk. Successful coil embolisation of the right fistula was performed angiographically (fig 2). The fistula arising from the LAD was entered and during deployment the coil migrated distally into the LAD (fig 3). Immediate surgery was performed, the coil was retrieved and the fistulae were oversutured with 5-0 non-absorbable suture throughout their length.

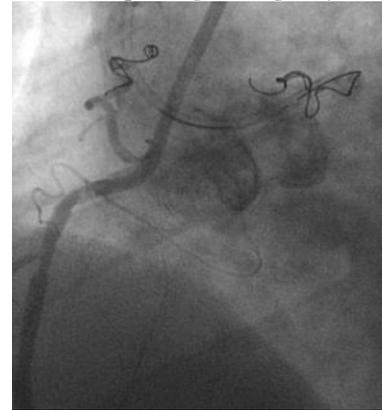
**Figure 1:** *Fistula from RCA to right PA*



**Figure 2:** *Fistula from LAD to pulmonary trunk (coil visible in RCA to PA fistula, guide wire visible in LAD)*



**Figure 3:** *Coil in left fistula, which later migrated into distal LAD requiring emergency surgery*



### Case 2

A 60 year-old female was referred for mitral valve replacement for rheumatic mitral stenosis. On preoperative coronary angiography a fistula was detected originating from the distal right coronary artery, draining into the right ventricle (RV) (fig 4). During the patient's mitral valve replacement the fistula was oversutured using 5-0 nonabsorbable suture along its entire length.

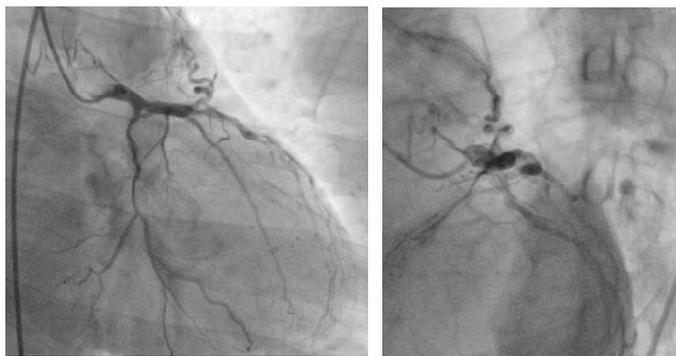
**Figure 4:** *Fistula from RCA to RV*



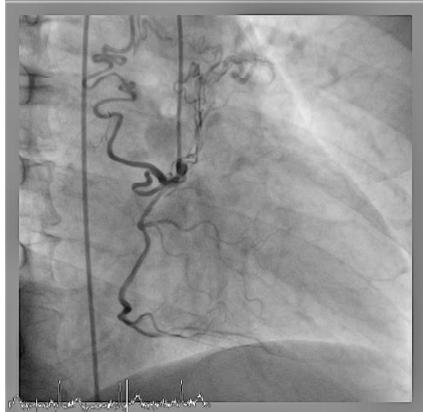
### Case 3

A 70 year-old male with chest pain and a positive exercise stress test underwent coronary angiography. Three fistulae were seen, one from the proximal LAD artery draining into the pulmonary trunk (PT) (fig 5) and the other two originated from the right coronary artery, one drained into the PT and the other into the right atrium (RA) (fig 6). The patient required coronary artery bypass grafting (CABG) and the fistulae were oversutured using 5-0 non-absorbable suture during the same procedure.

**Figure 5: Fistula from LAD to PT**



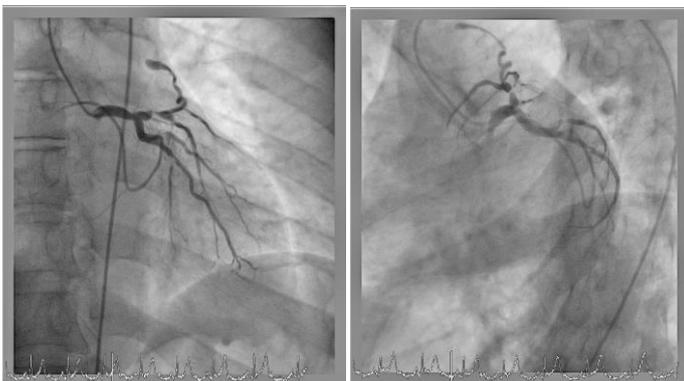
**Figure 6: Fistulae from RCA to PT and RA**



**Case 4**

A 54-year-old male presented with a non-ST elevation myocardial infarction and coronary angiography showed multivessel disease requiring CABG. A fistula was seen arising from the proximal LAD, draining into the pulmonary trunk (fig 7). The fistula was oversutured during the CABG along its entire length using 5-0 non-absorbable suture.

**Figure 7: Fistula from proximal LAD to PT**



**Discussion**

All the cases discussed above had favourable outcomes. The possibility of successful open surgical treatment of these fistulae is clearly identified. Patients being considered for open surgery for other reasons in whom such fistulae are discovered are best treated surgically as definitive cure may be anticipated without increasing the operative risk. The debate on how to treat these fistulae in patients not being planned for other open heart surgery persists.

Although no randomised control trial exists comparing surgical to percutaneous closure, surgery has been shown to yield lower recurrence rates.<sup>5-9</sup> Surgical techniques include oversuturing the fistula throughout its length, closing the fistula at its distal end, or closing the fistula via its receiving chamber.<sup>11</sup> The last technique requires cardiopulmonary bypass. There is no difference in outcome according to type of surgical repair.<sup>9</sup>

Said (2010) reviewed all coronary artery fistulae cases published between the year 2000 and 2009. 122 patients underwent percutaneous embolisation and 111 underwent surgical treatment. Surgical ligation was performed with cardiopulmonary bypass in 67% of cases and without cardiopulmonary bypass in 33%. Surgical correction yielded a 100% closure rate and 100% survival rate.<sup>5</sup> Percutaneous intervention had less positive results with 87% closure rate.<sup>6</sup> Included in this review was a paper reporting a recurrence rate of 9% with percutaneous intervention. Another study reported residual flow in one of the 25 patients after surgical repair at 9.6 years follow-up.<sup>8</sup>

Although the four cases discussed here were all asymptomatic, the same management can apply to symptomatic cases. Bogers et. al. (1987) presented 23 cases treated surgically. One patient died intraoperatively from other complex congenital cardiac disease. The other 22 achieved full resolution of symptoms and signs and many showed improved cardiac function on echocardiography. The surgical techniques used included oversuturing the fistula from the outside (8 cases), opening the distal fistula and closing the anatomical distal end (6 cases) and opening the receiving cardiac chamber or great vessel and closing the distal end (8 cases). There was no difference in outcome depending on the type of closure. The authors also reviewed the literature and concluded that surgical repair should be offered to all those who are symptomatic.<sup>9</sup>

The possibility of reducing myocardial ischaemia time by performing fistula ligation on the beating heart has been described. Yusuf reported eleven cases of coronary artery fistulae treated surgically. Seven cases were performed on cardiopulmonary bypass while four were performed on a beating heart. One patient underwent surgery for isolated fistula closure whereas all other patients underwent concomitant CABG.

## Case Report

Operative technique included identifying the fistula on the cardiac surface, dissecting it out proximally and clamping it to ensure disappearance of the thrill. The fistula was ligated proximally and distally. One patient required early re-operation as the fistula was still patent in intensive care immediately post-op. At echocardiogram follow-up 3 months later all cases achieved full resolution.<sup>10</sup>

Fistula repair without cardiopulmonary bypass is possible only when the fistulae are accessible without opening any chambers or disrupting the blood flow to the feeding coronary artery.<sup>11</sup> Stroeh et. al. (2012) described the case of a 22 year old patient with a fistula from the right coronary to the right ventricle. The right sinus of Valsalva and proximal right coronary artery were aneurysmal. The aneurysm was incised from the aorta and the fistula was closed with a pericardial patch on cardiopulmonary bypass.<sup>12</sup> Darwazah described the repair of an aneurysmal circumflex fistula draining into the coronary sinus.<sup>13</sup>

Successful percutaneous transarterial coil embolisation has been extensively described.<sup>5-7</sup> Isolated cases of percutaneous closure of large fistulae are also described. Lebreiro et. al. (2010) reported the use of an Amplatzer Vascular Plug (St. Jude Medical, USA) device for percutaneous closure in a 36 year-old patient with a large fistula from the right coronary artery to the superior vena cava.<sup>14</sup> They conclude that although most authors recommend surgical closure, they report a successful outcome despite the significant size of the fistula. Takaaki et. al. (2012) also reported a successful closure of a giant fistula arising from the right coronary artery in a 71-year-old with follow-up limited to 10 months.<sup>15</sup> Panduranga et. al. (2012) described a similar case followed up with coronary angiography 10 years later. The fistula closure remained sound, however there was aneurysmal dilatation of the proximal right coronary artery, around the origin of the fistula.<sup>16</sup>

### Conclusion

The consensus is that all fistulae should be closed. The preferred method of closure, percutaneous or surgical, is open to discussion. Based on results reported in the literature, most authors favour surgical closure, especially in symptomatic patients. Although no randomised control trial exists, several case series show a higher recurrence rate after percutaneous closure. Although size is unlikely to be a contraindication to percutaneous closure, subsequent aneurysmal dilatation of the coronary artery feeding a large fistula at late follow-up would favour initial surgical closure. In cases where the fistulae have multiple connections, circuitous patterns or acute angulations, rendering catheter entry difficult, surgical repair would be the preferred option. In patients requiring sternotomy for other conditions such as valve replacement, congenital defect repair or

bypass grafting then the risks of percutaneous embolisation are best avoided by repairing the fistulae during the same surgical procedure. Percutaneous transarterial embolisation has shown some promising results but as yet open surgical closure of coronary artery fistulae still gives the best patient outcomes.

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