Percutaneous closure of coronary artery fistulae: is it time for a UK registry?

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Abstract

Most coronary artery fistulae are asymptomatic but there may be complications such as rupture and myocardial infarction. Percutaneous intervention is an attractive alternative to open surgical repair that offers lower procedural risk.

Increasing numbers of fistulae are being discovered incidentally during angiography. They present challenges in assessment and management. For example, there is poor correlation between symptoms and the size and flow rate of fistulae.

Key words: coronary artery fistulae, percutaneous intervention, surgical repair, scintigraphy, registry.

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Background

Coronary artery fistulae are rare, occurring in 0.1% of the general population,[1,2] but are the largest subgroup of haemodynamically significant congenital coronary artery anomalies.[3] Most are asymptomatic, and are thus diagnosed incidentally at angiography. Presentation can occur either through the development of complications or by detection of a continuous murmur. Coronary artery fistulae are associated with complications such as rupture,[4] congestive cardiac failure,[5] myocardial infarction,[6] arrhythmias,[7] endocarditis[4,6] and death.[4,8] They may also lead to symptomatic myocardial ischaemia.[5,9] Open surgical repair is generally effective and safe.[10-12] However, there can be considerable difficulty gaining surgical access to a fistula and ensuring that the whole anatomical communication has been treated by this method.[13]

Percutaneous intervention is an attractive option that offers lower procedural risk. Percutaneous closure has been performed since the 1980s both in adults and children, using diverse techniques such as detachable coils and balloons,[14-16] covered stents, umbrella devices, cyanoacrylate, polyvinyl alcohol foam and even the floppy tips of guidewires.[17-19] There is no central database or registry of the management of coronary artery fistulae and it is therefore not possible to determine the comparative frequency of medical, percutaneous and surgical treatment.

Here we report on four consecutive cases of percutaneous closure of coronary artery fistulae that were performed in a single centre. In all cases the patients were symptomatic with angi-

Figure 1. Images showing successful closure of left anterior descending coronary artery (LAD) to pulmonary artery fistula with a covered stent

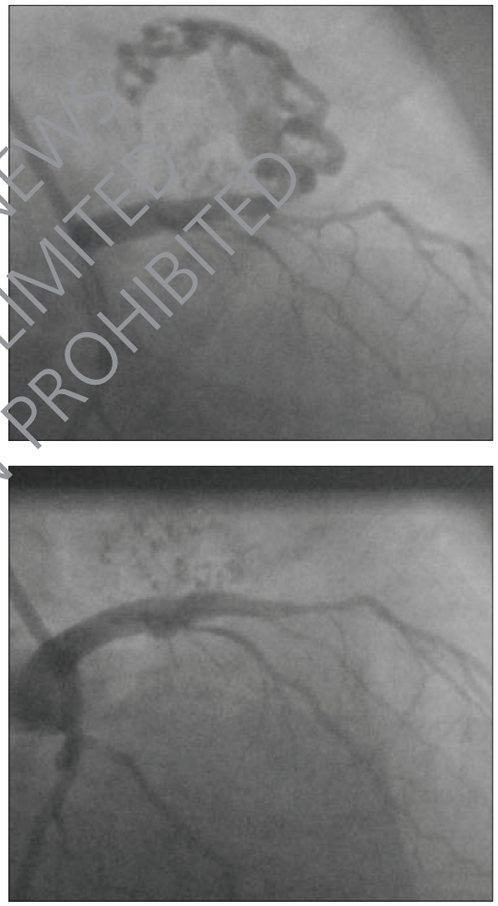

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Figure 2. Successful closure of a left anterior descending coronary artery (LAD) to pulmonary artery fistula with four coils

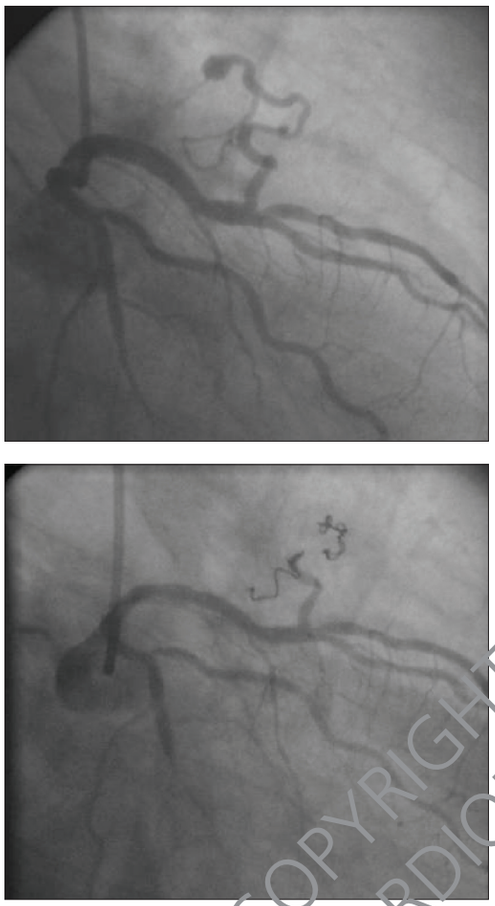
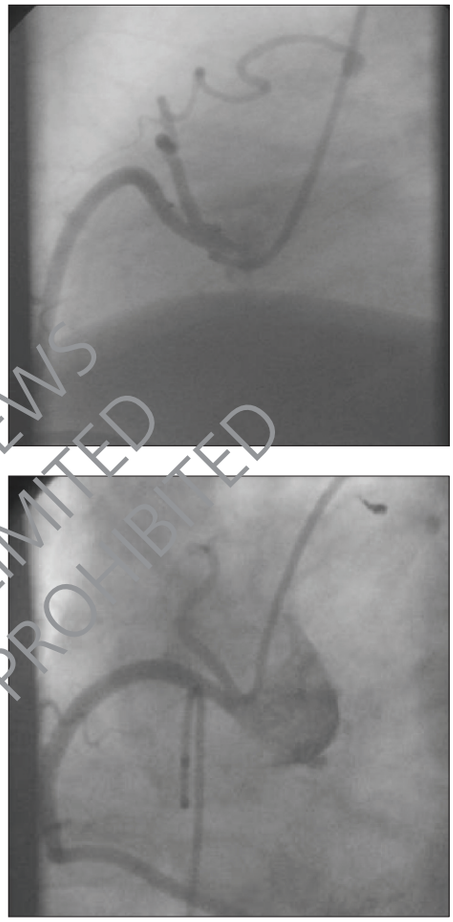


Figure 3. Proximal right coronary artery (RCA) to pulmonary artery fistula, showing cessation of flow following insertion of two coils. Note pacing wire inserted during procedure for transient bradycardia



na pectoris and had objective evidence of myocardial ischaemia on myocardial scintigraphy. We describe their percutaneous fistula closure and subsequent clinical outcome. In all cases written informed consent for the procedure was obtained.

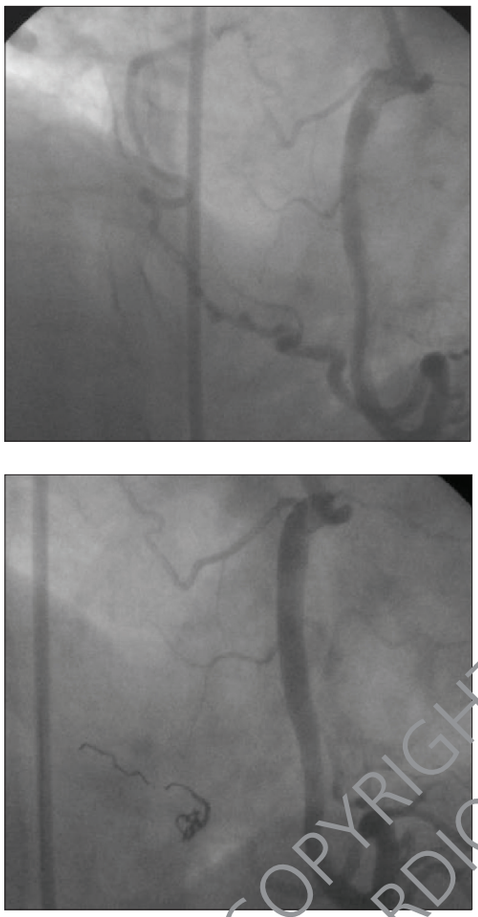
Patient 1

A 53-year-old woman presented with symptoms of chest pain and shortness of breath on exertion. Myocardial scintigraphy revealed reversible ischaemia in the anterior wall. Cardiac catheterisation revealed a tight calcified lesion in the left anterior descending coronary artery (LAD) immediately distal to a fistula, with flow from the LAD into the pulmonary artery. At intervention, following predilatation in the LAD, a 3 mm x 16 mm covered stent was placed across the origin of the fistula and the LAD stenosis (figure 1). There was an excellent angiographic result, with occlusion of the fistula. However, the patient presented again with similar symptoms one month after the procedure. Repeat coronary angiography confirmed that the fistula remained occluded, with no obstruction to flow through the stent. Myocardial scintigraphy, repeated six months after the initial procedure, revealed no evidence of residual ischaemia. In view of her continuing symptoms a further coronary angiogram was repeated three years after the initial procedure. This revealed that the fistula remained occluded and there was mild in-stent restenosis only.

Patient 2

This 58-year-old man presented with angina. Cardiac catheterisation revealed a fistula from the LAD to the pulmonary artery. There was also a 30–40% stenosis at the origin of a large obtuse marginal artery. Myocardial scintigraphy confirmed an area of reversible anterior myocardial ischaemia. Percutaneous fistula closure was performed using four 2 mm Boston Scientific coils, delivered to a total of three collateral channels making up the fistulous connection (figure 2). There was negligible flow through the fistula at the end of the procedure.

Figure 4. Distal right coronary artery (RCA) to pulmonary artery fistula closed with four coils



The patient remained completely symptom-free for one month but then his symptoms recurred. Myocardial scintigraphy was therefore repeated, which confirmed abolition of the perfusion defect identified on the previous scan. Unfortunately, this objective evidence of reversal of ischaemia was not matched by an improvement in his symptoms.

Patient 3

A 61-year-old woman was referred with a long history of exertional chest pain and breathlessness. Previous investigations five years earlier had included a coronary angiogram, which showed unobstructed coronaries and a proximal fistula in a dominant right coronary artery (RCA) draining to the pulmonary artery.

She was treated conservatively. Deteriorating symptoms, however, led to further cardiac catheterisation which showed no atheroma but a significant fistula, as before. Myocardial scintigraphy revealed reversible stress-induced inferior myocardial ischaemia consistent with a steal syndrome from her RCA. A LIMA catheter was used to intubate the fistula selectively. A terumo CT Leggiero guide cath system was used to deploy 2 x 20 mm fibre coils into the distal part of the fistula. This caused a cessation of flow down the fistula (figure 3). During the procedure she has a transient episode of chest pain with ST elevation, associated with profound bradycardia and hypotension, correlating to a period of slow flow in the RCA. She was resuscitated with emergency temporary pacing, intravenous fluids and atropine. At the end of the procedure, flow in the RCA was normal and she was haemodynamically stable.

Whilst there was an initial resolution of her symptoms she was re-investigated for recurrence of exertional chest tightness three years later. Repeat myocardial scintigraphy revealed normal perfusion, with resolution of the previous area of reversible ischaemia.

Patient 4

A 49-year-old man with a history of angina pectoris and a strongly positive exercise test underwent cardiac catheterisation. This showed unobstructed coronary arteries with a large fistula arising from the distal RCA and draining into the pulmonary artery. There was also a small fistula arising from the distal circumflex artery, which also drained into the pulmonary artery. Myocardial scintigraphy revealed reversible ischaemia in the inferior wall of the left ventricle. Percutaneous closure was therefore undertaken using four 2.0 x 20 mm fibre coils that were deployed into the two channels of the fistula (figure 4). At the end of the procedure there was virtual total occlusion of flow into the fistula (TIMI 0). The fistula from the circumflex artery was small and distal and closure was not attempted. At nine month follow-up he remained well.

Discussion

Coronary artery fistulae are most commonly congenital but can be iatrogenic[20] or caused by trauma.[21] The majority of fistulae (55%) arise from the RCA, with 35% arising from the LAD. The distal communication is variable with 41% draining into the right ventricle, 26% into the right atrium and 17% into the pulmonary artery. Whilst the majority are asymptomatic, and found incidentally, there is a risk of complications which increases with age, including myocardial ischaemia and infarction, congestive cardiac failure, arrhythmias, infective endocarditis and rupture.[1-9]

As the frequency of diagnostic angiography increases so do the numbers of coronary artery fistulae that are found incidentally in the adult population. This presents the attending cardiologist with several important management considerations. First, is the fistula an 'innocent bystander' or is it contributing to the patient's clinical syndrome? Secondly, does the risk of a larger fistula render the patient at high enough risk of complications for the fistula to be better treated prophylactically? Third, the experience of any individual cardiologist with these lesions is likely to be limited. Certainly, closure by percutaneous means requires a diverse set of interventional skills relating to access and the method of closure, which can be either by coil or by covering the origin of the fistula. These skills are partially available to most angioplasty operators, but in our series the closures were per-

formed in association with an expert interventional radiologist with no coronary experience but considerable experience of coil embolisation in other areas of the circulation. This multidisciplinary approach provides a wide-ranging skill base with which to tackle such cases but requires careful coordination. Fourth, the assessment of patients thought to have haemodynamically important fistulae is also challenging. In all the cases in our series some objective evidence of ischaemia was obtained by myocardial perfusion scans with stress. However, even though there was objective evidence from myocardial scintigraphy of abolition of ischaemia by closing these fistulae, the overall symptomatic improvement in our patients has been disappointing.

Previous experience

Previously reported case series have included few adult patients symptomatic with angina. In one case series, Reidy *et al*. reported on three symptomatic adults, two with angina and one with dyspnoea. One of the patients with angina had severe coronary disease, which was treated with angioplasty at the time of fistula closure. He became asymptomatic but, in keeping with our results, the other two patients (who both had evidence of reversible ischaemia on myocardial scintigraphy) remained symptomatic despite successful fistula closure.[15] On the other hand, in the series by Perry *et al.* the one patient (of 12) with angina became asymptomatic following successful closure of two fistulae.[16] Galassi *et al.* reported on an adult with angina and reversible ischaemia on myocardial scintigraphy. This patient became asymptomatic (the area of myocardial ischaemia on scintigraphy was also abolished) following closure of the fistula and treatment of an intermediate coronary stenosis with coronary angioplasty and use of a covered stent. It is, however, impossible to estimate the relative contributions of the fistula and coronary disease to the initial ischaemia in this case.[9]

Additionally, there is poor correlation between the presence of symptoms and the size and flow rate of fistulae, with some patients remaining asymptomatic despite large, high flow fistulae, even under conditions of high physiological stress such as marathon running.[22] These issues raise the question about whether there is a more robust way of ensuring that patients will benefit symptomatically from the procedure.

Although the percutaneous approach has considerable attractions compared with open surgery, it still carries risk. Complications in previously reported cases, for example, have included death related to device recoil into a major epicardial vessel,[23] myocardial infarction,[24] transient ECG changes and arrhythmias[25] (also seen in patient 3 in this series) and coil migration into the pulmonary circulation.[26] The use of cyanoacrylate appears particularly hazardous, with complication rates of 75% (including pulmonary embolus, pericarditis and acute renal failure) in one series.[17] Previous authors have recommended the following for percutaneous closure: ability to cannulate safely the branch coronary artery that supplies the fistula; absence of large branch vessels that may be inadvertently embolised; the presence of a single, narrow, restrictive drainage site into the cardiac chamber or vessel; and the absence of multiple fistulous communications.[25]

Key messages

- Coronary artery fistulae are the largest subgroup of haemodynamically significant congenital coronary artery anomalies
- Percutaneous intervention is an attractive alternative to open surgical repair
- Even with scan evidence of abolition of perfusion defects, symptomatic outcome is uncertain
- A national registry of these cases would be useful

Conclusion

Although percutaneous approaches to coronary artery fistula closure have been utilised for more than 20 years, there is still uncertainty as to how they should be best managed. Percutaneous fistula closure is an attractive and feasible option. As the patients in this series demonstrate, however, there is uncertainty as to the symptomatic outcome of such patients, even when reversible myocardial ischaemia has been proven. There is no available record to tell us how many such cases are undertaken in the UK, who does the cases or indeed what their outcome is. In order to address the important questions raised about this activity, a national registry would prove useful.

Conflict of interest

None declared.

Editors' note

Clinical images of bilateral coronary fistulae, discovered incidentally during cardiac catheterisation, appear in the article by Turley, de Belder and Shyam-Sundar on page AIC 13 of this issue of the journal.

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