Successful coil embolization of a large right coronary artery-coronary sinus fistula causing a significant left-to-right shunt: a case report

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Background
This case reviews a challenging but successful transcatheter coil embolization of a large congenital coronary artery fistula (CAF) causing a significant left-to-right shunt.

Case summary
A 51-year-old female with no significant prior history presented with symptoms of dyspnoea and chest discomfort. Extensive evaluation revealed a large CAF between a tortuous right coronary artery (RCA) and the coronary sinus (CS) composed of three aneurysmal pseudochambers. Closure of the RCA-CS fistula was attempted through coil deployment into the fistula neck. However, due to the brisk flow through the fistula, both coils embolized into the fistula sac. An alternative location was subsequently identified on three-dimensional rendering of a computed tomography angiography scan, which revealed a sharp bend in the RCA prior to the fistula neck and distal to the posterior descending artery (PDA) takeoff. Repeat attempt at embolization was accomplished using a telescoping system to reach and occlude the targeted bend. The coil mass remained stable and angiography demonstrated reduced flow through the fistula and preserved patency of the PDA. The decreased residual flow through the fistula secondary to the initial embolization attempt likely aided the successful deployment of coils in the second and final attempt. At 1 year, the patient was doing well with resolution of her symptoms and no clinical symptoms of coronary ischaemia.

Discussion
We suggest that an initial unsuccessful attempt at transcatheter embolization of a CAF should not preclude subsequent attempts for closure when there exists an appropriate indication.

Keywords
Embolization • Therapeutic • Coronary vessels • Heart defects • Congenital • Heart aneurysm • Heart catheterization • Case report

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Coronary artery fistulas (CAFs) are rare congenital anomalies for which the ideal management is not well studied. Large CAFs may present with multiple high-flow chambers that complicate attempts at transcatheter embolization. However, reduction in residual flow through the fistula secondary to initial coil embolization may aid subsequent attempts. An initial unsuccessful attempt at transcatheter embolization should not preclude subsequent attempts for fistula closure.

**Timeline**

| Presentation | Dyspnoea, chest discomfort, and atrial flutter |
|-------------|------------------------------------------------|
| One month later | Transthoracic echocardiogram: right atrial and right ventricular enlargement |
| Catheterization: $Q_p:Q_s$, 1.4 |
| Computed tomography angiography (CTA): large, tortuous fistula between the right coronary artery (RCA) and the coronary sinus (CS) composed of three aneurysmal pseudochambers |
| Three months | Initial attempt to embolize RCA-CS fistula at the fistula neck |
| Four months | Atrial flutter ablation with no subsequent episodes of palpitations |
| Six months | Continued dyspnoea |
| Eight months | Transthoracic echocardiogram: coils from initial embolization procedure lodged within fistula sac |
| Second and final coil embolization of RCA-CS fistula through an alternate location identified on three-dimensional CTA with preservation of the posterior descending artery |
| Twenty months | Clinically well and asymptomatic with no symptoms of coronary ischaemia |
| Cardiac magnetic resonance imaging: $Q_p:Q_s$, 1.1 |

**Introduction**

Coronary artery fistulas (CAFs) are rare congenital anomalies; incidence varies from 0.002% to 0.3%. The right coronary artery (RCA) is reportedly the most common site of origin of CAFs, accounting for 50–55% of cases; fistulas draining to the coronary sinus (CS) account for 7% of cases. Though most CAFs are asymptomatic, symptoms of CAFs include dyspnoea on exertion and palpitations. The ideal management of CAFs is not well studied due to the rarity of the condition. Previously, the presence of a large CAF was a class I indication given the extensive reports of coronary thrombosis following surgery in this particular scenario. The patient was taken to the catheterization lab and coiling of the RCA-CS fistula neck was attempted by deploying a 12 mm × 60 cm packing coil and a 14 mm × 60 cm packing coil (Penumbra, Inc., CA, USA). However, due to the brisk flow through the fistula, both coils embolized into the fistula sac (Figure 4A).

The patient later underwent a 2nd embolization attempt. To prevent its inadvertent embolization, the PDA was identified on computed tomography angiography (CTA) pre-procedure and again on fluoroscopy intra-procedurally. Importantly, the flow through the RCA seemed to have slowed with better contrast opacification during the 2nd catheterization, likely related to partial occlusion of the distal segments of the fistula. In order to overcome the tortuosity of the RCA, a telescoping delivery system was created using a 7-Fr 90 cm Flexor® guiding sheath (Cook Medical LLC, IN, USA) (Figure 4A) within which was advanced a Benchmark™ mother catheter (Penumbra, Inc., CA, USA) to the mid-RCA (Figure 4B), and a daughter catheter to the distal RCA (Figure 4C). A 150 cm LANTERN microwire (Penumbra, Inc., CA, USA) was then advanced into the fistula neck (Figure 4D). A stable coil mass was then attempted with deployment of a 14 mm × 34 cm AZUR framing coil (Terumo, NJ, USA) into the fistula neck (Figure 4E); however, this also embolized into the fistula sac (Figure 4F).

**Learning points**

- Coronary artery fistulas (CAFs) are rare congenital anomalies for which the ideal management is not well studied.
- Large CAFs may present with multiple high-flow chambers that complicate attempts at transcatheter embolization. However, reduction in residual flow through the fistula secondary to initial coil embolization may aid subsequent attempts.
- An initial unsuccessful attempt at transcatheter embolization should not preclude subsequent attempts for fistula closure.

**Case presentation**

A 51-year-old female with no significant prior history presented as an outpatient with dyspnoea on exertion and palpitations secondary to atrial flutter. Physical exam revealed a systolic murmur with a grade of 2/6. Transthoracic echocardiogram revealed moderate right atrial (RA) and right ventricular enlargement with normal systolic function. Right heart catheterization revealed a left-to-right shunt with step-up in oxygen saturation from the superior vena cava (SVC) (73%) to the pulmonary artery (80%). $Q_p:Q_s$ was 1.4 and pulmonary vascular resistance was 1.4 WU. Selective coronary angiogram revealed a tortuous, ectatic RCA with an aneurysmal connection through three pseudochambers to the CS and subsequent drainage into the RA (Figure 1). The posterior descending artery (PDA) takeoff appeared normal (Figure 1). Venography revealed a severely dilated SVC. Computed tomography confirmed the presence of three pseudochambers constituting the fistula sac: a 5 cm chamber immediately distal to the RCA, followed by a 2nd and 3rd chamber connected by a narrow neck (Figures 2 and 3).

Given the significant left-to-right shunt causing right heart enlargement, which was the likely aetiology for her atrial flutter, a decision was made to treat the patient’s fistula. Multidisciplinary discussion with cardiac surgeons led to the decision of a transcatheter approach given the extensive reports of coronary thrombosis following surgery in this particular scenario. The patient was taken to the catheterization lab and coiling of the RCA-CS fistula neck was attempted by deploying a 12 mm × 60 cm packing coil and a 14 mm × 60 cm packing coil (Penumbra, Inc., CA, USA). However, due to the brisk flow through the fistula, both coils embolized into the fistula sac (Figure 4A).

The patient later underwent a 2nd embolization attempt. To prevent its inadvertent embolization, the PDA was identified on computed tomography angiography (CTA) pre-procedure and again on fluoroscopy intra-procedurally. Importantly, the flow through the RCA seemed to have slowed with better contrast opacification during the 2nd catheterization, likely related to partial occlusion of the distal segments of the fistula. In order to overcome the tortuosity of the RCA, a telescoping delivery system was created using a 7-Fr 90 cm Flexor® guiding sheath (Cook Medical LLC, IN, USA) (Figure 4A) within which was advanced a Benchmark™ mother catheter (Penumbra, Inc., CA, USA) to the mid-RCA (Figure 4B), and a daughter catheter to the distal RCA (Figure 4C). A 150 cm LANTERN microwire (Penumbra, Inc., CA, USA) was then advanced into the fistula neck (Figure 4D). A stable coil mass was then attempted with deployment of a 14 mm × 34 cm AZUR framing coil (Terumo, NJ, USA) into the fistula neck (Figure 4E); however, this also embolized into the fistula sac (Figure 4F).
An alternative target location for the coils was subsequently identified in a sharp bend in the RCA distal to the PDA takeoff and coursing towards the fistula sac (Figure 4G), into which a series of coils were rapidly deployed (Figure 4H). The coil mass remained stable, and angiography demonstrated reduced flow through the fistula and preserved patency of the PDA. One year later, the patient was doing well with resolution of her symptoms, no clinical symptoms of coronary ischaemia and a reduced Qp:Qs of 1.1.

Figure 1 Three-dimensional cinematic rendering of computed tomography angiography (A) and angiography (B) demonstrated a tortuous, ectatic right coronary artery (RCA) with takeoff of the posterior descending artery (PDA) prior to connecting to a large fistula sac.

Figure 2 Craniocaudal computed tomography sequence demonstrating 1st pseudochamber (red), 2nd pseudochamber (green), and 3rd pseudochamber (blue) of the fistula. A narrow neck connected the 2nd and 3rd pseudochambers (arrowhead).
Transcatheter approaches to coronary fistulas offer a safe and effective alternative to high-risk open surgical procedures. Though successfully conducted, Harris et al. noted the challenge posed by large, tortuous, high-flow fistulas in transcatheter coil embolization repair. Indeed, a giant 4 cm × 7 cm RCA aneurysm described by Komoda et al., with a fistula into the CS, was surgically repaired. Due to the lack of an optimal surgical option for our patient, a transcatheter solution was offered, allowing for successful coil embolization of her large RCA-CS fistula.

We encountered multiple challenges during fistula closure. First, the tortuosity of the RCA made it difficult to advance a stable device delivery system to the fistula. Second, the fistula would require closing in a manner that would ensure preservation of the PDA. Third, we needed to achieve a stable coil mass that would withstand the brisk flow through the fistula and adequately occlude it. To address our fistula tortuosity, a telescoping delivery system was used to advance into the fistula neck. An angiographic view illustrating the PDA...
takeoff allowed for avoidance of inadvertent occlusion. Finally, a target location for embolization was identified on three-dimensional CTA imaging. Rather than deploying into the fistula neck, coil embolization was successfully redirected to a sharp bend in the RCA distal to the PDA takeoff and leading towards the fistula sac. A similar case was reported by Tacoy et al. in a transcatheter embolization of a rare congenital fistula draining from the left circumflex artery (LCA) into the CS. Embolization of the severely tortuous, high-flow LCA-CS fistula was achieved through deployment of four 3 mm Guglielmi detachable coils (Boston Scientific/Target, Cork, Ireland) while maintaining patency of the LCA’s distal branches. Report long-term outcomes of CAF repair include morbidity from angina to myocardial infarction. One investigation found that the sole angiographic feature predictive of adverse outcome of CAF closure was termination of the CAF into the CS. Of the 76 patients studied, 11 underwent major complications that included coronary thrombosis; 10 of these patients had a CAF that drained into the CS, and 5 were taking antithrombotic medication at the time of diagnosis. Several potential determinants of thrombosis following CAF repair have been theorized, including fistula length and diameter, coronary tortuosity and abrupt changes in vessel calibre, and abrupt cessation of high-volume flow following repair. Long-term follow-up and anticoagulation should thus be initiated in patients following CAF repair. Our patient was discharged on an anticoagulation regimen of aspirin, clopidogrel, and rivaroxaban.

Despite the challenges and complexity involved in transcatheter approaches to coronary fistulas, we propose that a transcatheter approach can be safely attempted when the procedure is indicated. Successful closure of our patient’s large, high-flow fistula was potentially aided by the patient’s initial embolization attempt, after which the coils had embolized into the fistula sac and caused a reduction in flow rate. Right coronary angiography performed prior to the repeat embolization revealed that the first coil sat in the 1st chamber, while the 2nd coil sat in the narrow neck between the 2nd and 3rd chambers (Figures 2–4A). The reduced residual flow through the fistula allowed for rapid deployment of a series of coils into a sharp bend in the RCA distal to the PDA takeoff, thus reducing flow through the fistula and preserving flow to the PDA. We therefore suggest that an unsuccessful attempt at transcatheter embolization of a CAF should not preclude subsequent attempts for closure when there exists an appropriate indication.

**Supplementary material**

Supplementary material is available at European Heart Journal - Case Reports online.

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**Slide sets:** A fully edited slide set detailing this case and suitable for local presentation is available online as Supplementary data.

**Consent:** The authors confirm that written consent for submission and publication of this case report including images and associated text has been obtained from the patient in line with COPE guidance.

**Conflict of interest:** C.H.L., MD, PhD, is a data monitoring committee member of ACI Clinical, speaker for Abiomed, proctor for Abbott, and course director for Gore Medical. P.C. is a full-time Research Collaborations Manager and Senior Key Expert at Advanced Therapies Division, Siemens Medical Solutions USA, Inc.

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**Lead author biography**

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