Successful percutaneous occlusion of a large left circumflex coronary artery fistula draining into the coronary sinus using a ventricular septal defect occluder: a case report

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Background
Coronary artery fistula (CAF) is a congenital anomaly of the coronaries that can lead to significant intracardiac shunting and myocardial ischaemia.

Case summary
We describe the case of a 15-year-old male with an incidentally documented precordial cardiac murmur. An evidently dilated coronary sinus (CS) on transthoracic echocardiography prompted further investigation. A computed tomography (CT) revealed the presence of a large CAF from the left circumflex coronary artery to the CS. No other structural heart defects were detected. A haemodynamically significant intracardiac shunt was confirmed during cardiac catheterization, and it was decided to close the fistula. This was successfully performed using a ventricular septal defect (VSD) occluder (Konar 10-8, Lifetech Scientific) that was deployed through a 6 Fr right coronary guiding catheter. A partial thrombotic occlusion of the CS behind the closure device was noted during follow-up which led to anticoagulation in a higher target INR range and concomitant start of low dose carbasalate calcium to reduce further retrograde thrombus extension. Patient is doing well at over 1 year of follow-up, and no further thrombotic extension into the CS was seen on a recent CT.

Discussion
This report illustrates the diagnostic workup and a percutaneous treatment strategy of a CAF using a VSD occluder. We also describe a not previously reported complication, thrombotic CS occlusion. Improving transcatheter techniques and marketing of novel devices with a broad spectrum of applications can offer new opportunities for treating CAF and avoiding surgical correction often involving cardiopulmonary bypass, reserving this option for patients with complex anatomy or failed transcatheter closure.

Keywords
Congenital heart disease • Coronary anomaly • Coronary fistula • Structural heart disease intervention • Case report

Learning points
• Coronary artery fistula (CAF) is a rare form of an arterio-venous communication between one of the coronary arteries and a cardiac chamber or a great vessel.
• The majority of CAFs will remain silent. Large fistulae can be a haemodynamic burden and result in significant shunting and volume overload or myocardial ischaemia due to a steal phenomenon.
• Transcatheter or surgical closure of a CAF should be considered in all haemodynamically relevant lesions.
Introduction

Coronary artery fistula (CAF) is a rare form of arterio-venous malformation entailing a direct communication between one of the coronary arteries and a cardiac chamber or a great vessel. As most CAFs are congenital, haemodynamically insignificant and will remain silent, the true incidence of the defect in the general population is not known. This anomaly is diagnosed in 0.1–0.3% of patients undergoing a diagnostic heart catheterization. Most fistulae arise from the right coronary artery (RCA), left anterior descending, and left circumflex artery (LCx, named in order of decreasing incidence). The vast majority of CAFs (> 85%) drain into the right heart chambers or the pulmonary artery. Only several cases of fistulae between LCx and the coronary sinus (CS) have been reported in the literature, and this anatomical variant is estimated to represent only 7% of CAFs.

The size of the communicating orifice, pressure difference between the artery and the compartment it is draining into and the relative position of the fistula are all important determinants in the magnitude of the shunt and the clinical consequences. Large CAFs can be a haemodynamic burden and result in significant shunting and volume overload or myocardial ischaemia due to a steal phenomenon. Experts recommend elective closure of large CAFs, even in asymptomatic patients, to reduce associated long-term mortality and morbidity, irrespective of ischaemia stress testing. The treatment options entail surgical ligation or catheter-based alternatives. Surgically treated patients show a reduced long-term survival compared to age matched controls and about 11% of the patients experience post-operative myocardial infarction due to low flow state in the dilated coronary artery proximal to the fistula or progressive retrograde thrombosis involving the coronary arteries. Over the recent years, various catheter-based techniques have proven to be an effective and safe alternative with non-inferior long-term outcomes in comparison to surgery. In this report, we describe a giant CAF between the LCx coronary artery and the CS in an adolescent male, which was successfully managed percutaneously using a newly marketed Lifetech Konar-MF ventricular septal defect (VSD) occluder device.

Case presentation

The patient, a 15-year-old male, had no relevant cardiovascular history or complaints and was being analysed due to an incidentally documented precordial gross systolic-diastolic murmur without radiation. His electrocardiogram showed regular sinus rhythm and normal de- and repolarization. Transthoracic echocardiogram (TTE) showed a non-dilated left ventricle with normal systolic function and revealed an evidently dilated CS (Figure 1A) and a dilated tortuous vascular structure running per continuum from the anterior to the inferior basal epicardial cardiac borders (Figure 1B). A systolic and diastolic jet could be demonstrated in the region of inflow of the CS in the right atrium (Figure 1C) as well as a clearly dilated left main coronary artery (Figure 1D). A contrast-enhanced electrocardiogram (ECG)-gated computer tomography angiography (CTA) confirmed the presence of a large CAF from the LCx to the CS (Figure 2).

A cardiac catheterization was performed during which a pulmonary artery pressure of 24/13 mmHg and a left to right intracardiac shunting with the magnitude of Qp:Qs 1.6:1 was identified. A vastly dilated left main and LCx coronary artery was visualized, which fed a distal fistula with the CS (Figure 3A), while the rest of the coronary anatomy was normal with a dominant RCA. The diameter of the fistula at the entrance to the CS was about 4–5 mm, the LCX was dilated to 17 mm. Due to the large size of the CAF, the significant left to right shunt and the evident dilation of the left main and LCX arteries prone to lead to myocardial steal phenomenon in a young active patient it was decided to close the fistula percutaneously. This was successfully performed using a Konar VSD-occluder (10-8, Lifetech Scientific; Figure 3B, C). The occluder was advanced and deployed through a 6 Fr right coronary guiding catheter. There was no residual flow at the end of the procedure, as demonstrated by the selective angiography (Figure 3D). The patient was started on vitamin K antagonist anticoagulant (phenprocoumon) with target International Normalized Ratio (INR) 2.0–3.0 and was discharged home in good condition after a 48-hour observation period on the ward. Although no recommendations currently exist on the optimal antplatelet or anticoagulation treatment of patients after CAF closure, it is common practice in many institutions to give therapeutic

Timeline

| Presentation       | Incidentally documented cardiac murmur |
|--------------------|----------------------------------------|
| 1 month later      | Transthoracic echocardiogram: dilated coronary sinus (CS), suspicion of a coronary fistula |
| 3 months           | Computed tomography angiography (CTA): large coronary artery fistula from the left circumflex artery to CS |
| 6 months           | Catheterization: Qp:Qs 1.6:1 shunting |
|                    | Percutaneous closure using a ventricular septal defect occluder |
|                    | Uneventful 48-hour clinical observation |
|                    | Discharge home with a vitamin K antagonist [target International Normalized Ratio (INR) 2.0–3.0] |
| 6 months and 1 week| Readmission with non-specific chest complaints |
|                    | CTA: a partial thrombotic occlusion of the CS |
|                    | Increase in target INR to 3.0–4.5 and start carbasalatecadium |
|                    | Uneventful 10-day clinical observation |
| 12 months          | Clinically well and asymptomatic |
|                    | CTA: no residual shunting, no further thrombotic extension into the CS |
anticoagulation for patients with large CAFs, given the higher risk of thrombotic complications in this group.

The patient was readmitted a week after the procedure with transient non-specific chest complaints without any ischaemic ECG changes and no significant increase in high sensitive cardiac troponin T levels. Additional TTE and CTA imaging showed the VSD occluder in situ without signs of migration or residual flow over the fistula, however, a partial thrombotic occlusion of the CS proximal to the closure device was noted (Figure 4). The patient reported adequate self-monitored anticoagulation. No thrombosis of the macroscopic coronary artery system branches could be identified and the patient could be discharged home after a 10 day ward observation period without having had any complaints or signs of ischaemia. We anticoagulated him with an increased target INR 3.0–4.5 and started on low-dose carbasalate calcium to reduce further retrograde thrombus extension. Currently over 12 months after the procedure, he is clinically well and the follow-up CTA showed no further thrombotic extension into the CS.

The Lifetec Konar-MF™ VSD Occluder (Lifetech Scientific, Shenzhen, China) received the CE marking in 2018. The occluder is a self-expandable, double-disc device made from a nitinol wire mesh and the two discs are connected by a cone-shaped waist (Figure 3B)11. Each disc has a screw that can be connected the delivery cable. Polytetrafluoroethylene (PTFE) membranes are sewn inside the waist of the four large models to increase the ability of closure. The occluder has excellent closure properties and can be delivered through a relatively small delivery catheter (in this case a 6 Fr guiding catheter).

**Discussion**

Coronary artery fistula is a rare, yet important to recognize congenital anomaly of the coronaries that can lead to significant volume overload/shunting and myocardial ischaemia due to the coronary steal phenomenon, especially in case of large or complex fistulae.12 Surgical or transcatheter repair should at least be considered in all haemodynamically non-trivial CAF patients, irrespective of their complaints.6,7,12 Although the best approach is still a matter of ongoing debate, transcatheter closure has been shown to be an effective and safe primary intervention of choice, avoiding cardiopulmonary bypass in this often young patient group.8–10,13–15 Here we describe the case of a large LCx-CS fistula that was...
successfully closed percutaneously using the Konar 10-8 VSD occluder, which has only been available on the European market since spring 2018 and is to our current knowledge, the first time this occluder was used to close a CAF in Europe. The device can be delivered through a relatively small catheter (a particularly important feature in congenital cardiology patients), we used a 6 Fr right coronary guiding catheter. The haemodynamic and angiographic result of the procedure was satisfying and the post-procedural admission course uneventful.

During follow-up, we were confronted with a rather extensive thrombus formation proximal to the closure device, to our knowledge not a previously reported complication of percutaneous CAF closure. The therapeutic anticoagulation proved to be insufficient to prevent this thrombotic complication. Given the large calibre of the CAF, the complete closure leading to flow stagnation and the fine pro-thrombotic nitinol cage structure of the percutaneous closure device, the risk of thrombosis was not entirely unforeseen in this patient. This highlights the importance of post-procedural monitoring and structural imaging during follow-up and calls for further research on optimal post-procedural antiplatelet/vitamin K antagonist or novel oral anticoagulation treatment post-CAF closure. Long-term anticoagulation is probably warranted in such cases to prevent progressive thrombosis and coronary ischaemia. Recent case series reports up to 11% of treated patients with CAFs developing thrombosis mediated myocardial infarction. Incidence and risk factors for coronary artery fistula (CAF), coronary sinus (CS), left circumflex artery (LCx), left atrium (LA).

Figure 2. A volume rendering reconstruction of a computed tomography angiography confirming the presence of a large coronary artery fistula from the left circumflex artery to the coronary sinus. Evident dilatation of the left main and the left circumflex arteries is seen.

Figure 3. Angiographic aortic root injection showing the course of the dilated left circumflex artery to the coronary sinus (A). A schematic representation of the ventricular septal defect occluder (Lifetech Scientific) (B). Post-procedural angiography showing the ventricular septal defect occluder in situ (C) resulting in good angiographic result without any rest shunt through the coronary artery fistula (D). Ao, aorta; CAF, coronary artery fistula; CS, coronary sinus; LCx, left circumflex artery. Courtesy of Lifetech Scientific.
thrombosis after percutaneous CAF closure are not yet clearly defined and a literature search suggests that the details of these patients are not well documented and possibly underreported. If there is no extension of the thrombus into the coronary artery system, occlusion of the CS could prone maturation of the cardiac collateral venous circulation (amongst others the anterior cardiac and Thebesian veins), avoiding long-term functional problems.

Improving transcatheter techniques and marketing of novel devices with a broad spectrum of applications such as the Konar VSD occluder can offer new opportunities for treating CAF and avoiding surgical correction often involving cardiopulmonary bypass, reserving this option for patients with complex anatomy or failed transcatheter closure.

Lead author biography

Anastasia Dmitrievna Egorova (1987) is an adult congenital and implantable rhythm device cardiologist at the Leiden University Medical Centre in the Netherlands. She defended her PhD thesis on ‘Ciliary regulation of endothelial response to shear stress’ in 2012 and completed her cardiology training in Leiden in 2018. During her congenital cardiology fellowship, she spent several months in training at the Deutsche Herzzentrum in München, Germany, where the work described in this case report was performed.

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 author/s confirm that written consent for submission and publication of this case report including image(s) and associated text has been obtained from the patient in line with COPE guidance.

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Figure 4 Post-procedural transthoracic echocardiography (modified four-chamber view, A) and computed tomography angiography (B) shows the ventricular septal defect occluded effectively closing off the left circumflex artery fistula with the coronary sinus, resulting in extensive thrombus formation proximal to the closure device, however, not involving any coronary arterial branches. CS, coronary sinus; LV, left ventricle; RA, right atrium; RV, right ventricle.
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