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CASE REPORT
Adult congenital heart disease

Minimally invasive coil embolization for significant left-to-right shunts due to giant coronary-to-pulmonary artery fistulas: a case report

Sato Namura [1], Hiroki Konishi [1]*, Ryo Nishio [1], and Hiroshi Yamamoto [2]

[1]Department of Cardiology, Yodogawa Christian Hospital, 1-7-50, Kunijima, Higashiyodogawa-ku, Osaka 533-0024, Japan; and [2]Department of Diagnostic Radiology, Sumitomo Hospital Interventional Radiology Center, 5-3-20, Nakanoshima, Kita-ku, Osaka 530-0005, Japan

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Background Coronary-to-pulmonary artery fistula (CPF) is a rare disease, and its optimal treatment strategy remains controversial. Herein, we report a rare case of minimally invasive coil embolization of giant CPFs.

Case summary A 78-year-old man with a history of persistent atrial fibrillation and lumbar canal stenosis presented to our hospital with breathlessness. Cardiac computed tomography revealed giant CPFs inducing a significant left-to-right shunt (Qp/Qs 1/2.1) with a coronary artery aneurysm smaller than the size indicated for surgical treatment. To reduce the left-to-right shunt flow, coil embolization procedures for the fistulas were performed twice. Initially, the fistula arising from the right coronary artery was embolized using three Target® XXL (6 × 40 mm, 5 × 20 mm) and two Target® XL SOFT (4 × 12 mm) coils (Stryker Inc., Tokyo, Japan). One month later, the fistulas arising separately from the left coronary artery were embolized. After the procedures, the major shunt flow disappeared angiographically, and Qp/Qs significantly decreased to 1/1.2. Additionally, the fractional flow reserve of the left coronary artery increased from 0.79 to 0.93, and cardiopulmonary exercise testing showed an improvement in his exercise tolerance.

Discussion In similar cases, a surgical procedure with ligation of the CPFs combined with resection of a small aneurysm and coronary artery bypass grafting would normally have been considered the best approach. However, endovascular treatment targeting only the fistulas was a superior strategy considering the patient's age. The coil embolization technique effectively controlled the shunt flow of the CPFs. This technique is considerably less invasive than surgical therapy.

Keywords Aneurysm • Case report • Coil embolization • Coronary-to-pulmonary artery fistula

ESC curriculum 9.7 Adult congenital heart disease • 6.1 Symptoms and signs of heart failure • 6.3 Heart failure with preserved ejection fraction • 9.6 Pulmonary hypertension

Learning points

- The coil embolization technique for coronary-to-pulmonary artery fistulas (CPFs) is considerably less invasive and may be a valid alternative to surgery in selected cases.
- For CPFs, the coil embolization can significantly reduce arteriovenous shunt flow and concomitant volume overload.

* Corresponding author. Tel: +81 6 6322 2250, Fax: +81 6 6320 6308, Email: hiroky_k@yahoo.co.jp
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Introduction

Coronary artery fistula (CAF) is a rare heart disease characterized by malformations forming abnormal communications on the coronary arteries connecting to the cardiac chambers, coronary sinus, pulmonary arteries, and vena cava. Coronary-to-pulmonary artery fistulas (CPFs) account for 17% of all CAFs. Various treatments for symptomatic patients have been reported.[1–4] However, the optimal treatment strategy remains controversial. We report a rare case of minimally invasive coil embolization of giant CPFs.

Summary figure

Date	Events
Summer 2021	New York Heart Association (NHYA) class II dyspnoea.
May 2022	Admitted to our hospital because of his dyspnoea with NHYA class III. Transthoracic echocardiography and cardiac computed tomography revealed giant coronary-to-pulmonary artery fistulas extending from the left anterior descending artery (LAD) and right coronary artery (RCA) to the pulmonary artery.
July 2022	Revealed a significant left-to-right shunt (Qp/Qs 1/2.1) by coronary angiography and right heart catheterization. The first coil embolization for the fistula arising from the RCA.
August 2022	The second coil embolization for the fistulas arising from the LAD. After the procedures, Qp/Qs decreased to 1/1.2 and dyspnoea was improved to NYHA class I.
January 2023	Follow-up coronary angiography. No change in symptoms.

Case presentation

A 78-year-old man with a history of persistent atrial fibrillation, on rivaroxaban 15 mg, presented to our hospital with breathlessness. His New York Heart Association (NYHA) class was III. His $CHADS_2$ score was 2, and his HAS-BLED score was 1. His blood pressure was 134/83 mmHg, heart rate 90 b.p.m., oxygen saturation 98% (room air), with no fever. Physical examination revealed mild pitting oedema and no chest murmurs or rales. Transthoracic echocardiography revealed a left ventricular ejection fraction (51%) and left atrial and ventricular enlargement (left atrial dimension, 53 mm; left ventricular end-diastolic dimension, 55 mm; and left ventricular end-systolic dimension, 40 mm). Doppler ultrasonography revealed left atrial pressure elevation (E/e': 15.8), trivial tricuspid regurgitation, and a large shunt flow into the pulmonary artery (*Figure 1*). Cardiac computed tomography revealed giant CPFs from the left anterior descending artery (LAD) and right coronary artery (RCA) to the pulmonary artery. Furthermore, a small coronary artery aneurysm (10 × 12 mm) was observed in the ostium of the LAD (*Figure 2*). Coronary angiography revealed that the coronaries had no significant obstructive lesions. Right heart catheterization revealed a significant left-to-right shunt (Qp/Qs 2.1), and oximetry analysis revealed a significant oxygen step-up of 15% (72–87%) in the pulmonary artery. Right atrial pressure was 10/9/8 mmHg; right ventricular pressure, 35/6/8 mmHg; mean pulmonary artery pressure, 37/19/29 mmHg; pulmonary capillary wedge pressure, 21/21/18 mmHg; cardiac output, 5.92 L/min; cardiac index, 3.48 L/min/m^2. Thus, the CPFs were considered the main cause of the symptoms.

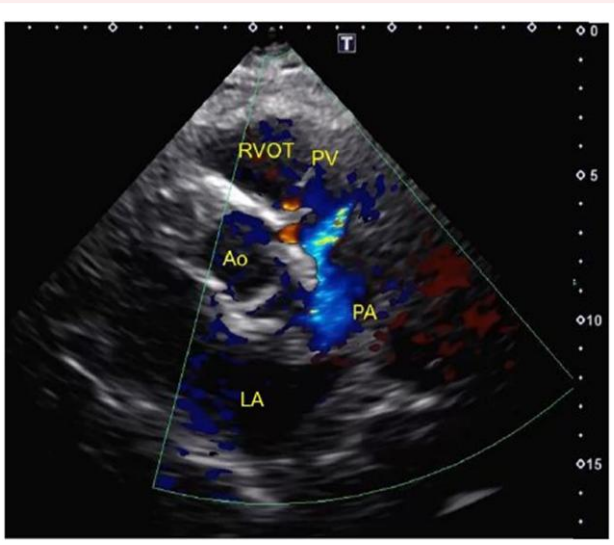


Figure 1 Echocardiography results reveal shunt flow into the pulmonary artery. The shunt flow into the pulmonary artery showed a Qp/Qs of 1/2.1. Ao, aorta; LA, left atrium; PA, pulmonary artery; PV, pulmonary valve; RVOT, right ventricular outflow tract.

Coronary-to-pulmonary artery fistula coil embolization was performed twice to reduce the left-to-right shunt flow. Initially, the fistula arising from the RCA was embolized using a 6 Fr Judkins Right guiding catheter (Launcher®, Medtronic, Minneapolis, MN, USA), a guide-extension catheter (Guideplus™ Ⅱ, Nipro, Osaka, Japan), and a guidewire (ASAHI SUOH 03®, ASAHI Inc., Aichi, Japan) running through a strongly bent malformed vessel. After inserting a microcatheter (Excelsior®1018®, Stryker, Kalamazoo, MI, USA) into the vessel, the malformed blood vessel was embolized using detachable coils (three Target XXL®s: 6 × 400 mm, 5 × 200 mm, and two Target XL® Softs: 4 × 120 mm) (Stryker Inc., Tokyo, Japan) (*Figure 3*, Supplementary material online, *Video S1*). The procedure improved Qp/Qs from 1/2.1 to 1/1.5. One month later, a second embolization procedure was performed for fistulas arising from the LAD. Intravascular ultrasonography (Opticross™; Boston, Natick, MA, USA) identified two fistulas arising separately from the LAD. A 6 Fr EBU 3.5 guiding catheter (Launcher®, Medtronic, Minneapolis, MN, USA), a guide-extension catheter (Guideplus™ II, Nipro, Osaka, Japan), and a guide wire (SION blue, Asahi Inc., Aichi, Japan) were inserted into the LAD. A steering microcatheter (Excelsior®1018®, Stryker, Kalamazoo, MI, USA) was inserted into the malformed vessel. Rapid pacing was delivered via a temporary pacing catheter placed in the right ventricle to reduce the coronary blood flow and prevent the coils from being washed away. Two fistulas were embolized using detachable coils (two Target XXL®s: 8 × 400 mm and four Target XL® Softs: 8 × 300 mm and 4 × 120 mm) (*Figure 4*, Supplementary material online, *Video S2*). After coil embolization, the major shunt flow disappeared angiographically, and Qp/Qs significantly decreased to 1/1.2. Further, the LAD's resting full-cycle ratio, tested using Pressure Wire™ X (Abbott, Illinois, USA), increased from 0.79 to 0.93. Bepridil 100 mg was prescribed the day after the reduced cardiac load due to the reduction or amelioration of shunt flow. His atrial fibrillation became sinus rhythm.

One month post-procedure, no shunt flow due to the CPF was detected using transthoracic echocardiography. His breathlessness

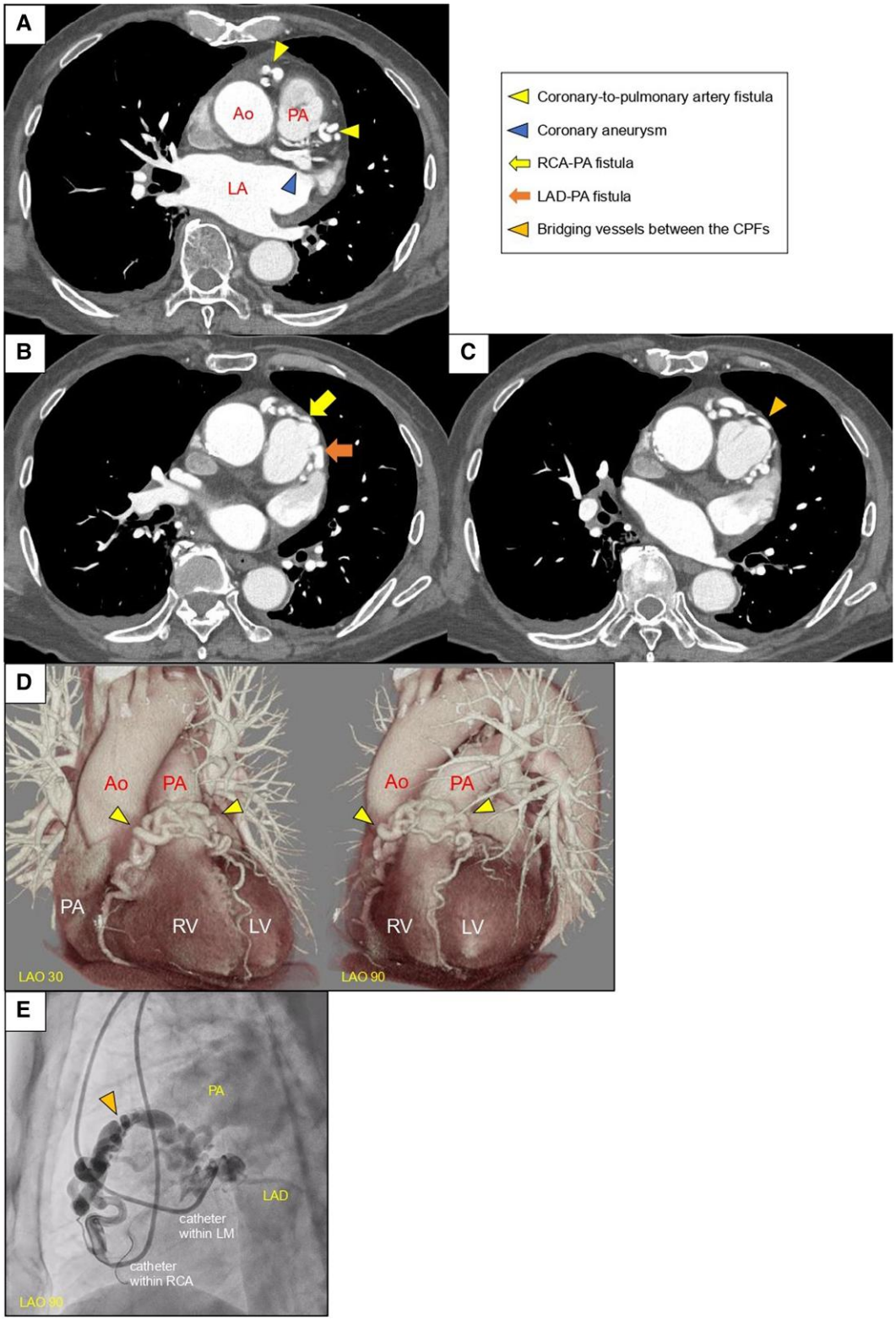


Figure 2 Coronary computed tomography images and angiography of coronary-to-pulmonary artery fistulas. (*A*) Axial coronary computed tomography detected CPFs and a coronary aneurysm. (*B*) Axial coronary computed tomography detected an RCA-PA fistula and LAD-PA fistula. (*C*) Axial coronary computed tomography detected bridging vessels between the CPFs. (*D*) Coronary computed tomography detected CPFs from LAO 30° CRA 0° and LAO 90° CRA 0°. (*E*) Contrast was simultaneously injected from the coronary ostia of the RCA and LAD. The CPFs from the RCA and LAD flowed into the same point of the PA. The bridging vessel between the CPFs from the RCA and LAD was revealed by coronary angiography. Ao, aorta; CPF, coronary-to-pulmonary artery fistula; LAD, left anterior descending artery; RCA, right coronary artery; LM, left main coronary artery; LV, left ventricle; PA, pulmonary artery; RV, right ventricle.

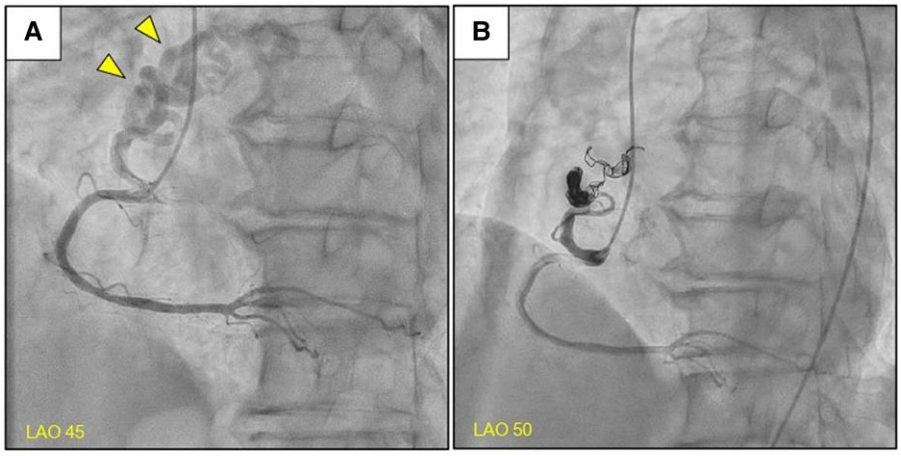


Figure 3 The coronary angiography results after the first procedure. (*A*) A strongly winding CPF flowed into the PA from the RCA. (*B*) The CPF arising from the RCA disappeared after the coil embolization. The bridging vessels between two CPFs from the RCA and LAD also disappeared. CPF, coronary-to-pulmonary artery fistula; LAD, left anterior descending artery; PA, pulmonary artery; RCA, right coronary artery.

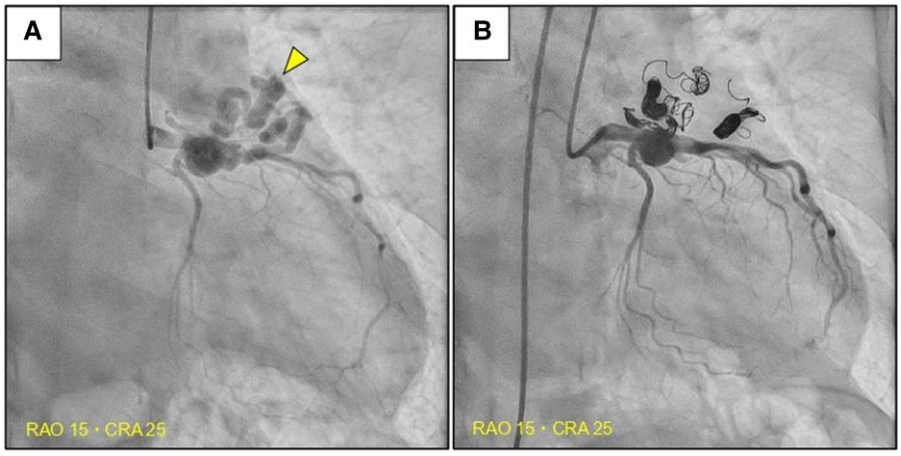


Figure 4 The coronary angiography results after the second procedure. (*A*) The CPFs flowed into the PA from the LAD, and the aneurysm was on the proximal region of the LAD. (*B*) The two fistulas were embolized with coils. The CPFs arising from the LAD disappeared, and blood flow into the LAD increased. CPF, coronary-to-pulmonary artery fistula; LAD, left anterior descending artery; PA, pulmonary artery.

improved (NYHA class I). Six months later, follow-up coronary angiography revealed a small shunt flow from both CPFs; however, the aneurysm size never changed angiographically (*Figure 5*). We planned to monitor the coronary artery aneurysm using echocardiography and coronary angiography; if it enlarges to meet the surgical criteria, surgery will be considered.

Discussion

Coronary artery fistulas are rare heart malformations, accounting for 0.13% of all congenital coronary anomalies and affecting 0.002% of the general population.[1] In adults, most CAFs are incidentally detected using coronary computed tomography angiography and coronary angiography. The prevalence of CAF on coronary computed tomography angiography is 0.9%, and that on coronary angiography is 0.05–0.25%.[5] Their most common origin is the LAD (84%), followed by the RCA (38%). The most common drainage site is the main pulmonary artery (89%). Approximately 45% of CPFs have multiple fistulas, and 19% have coronary-to-pulmonary fistulas.[5]

Although most CPFs are asymptomatic, angina (39.0–62.5%) and dyspnoea (3.6–25.0%) have been reported. Coronary-to-pulmonary artery fistulas can induce palpitations, syncope, arrhythmias, pulmonary hypertension, congestive heart failure, ischaemic heart disease, rupture, or sudden death.[1,2,5] If the shunt ratio (Qp/Qs) is above 1.5, it can help diagnose functional severity in patients with possible coronary steal using fractional or coronary flow reserve.[6]

The optimal treatment strategy remains controversial due to the low prevalence of CPFs. There is no clear definition of fistula size; however, fistulas are classified into small, moderate, or large according to the

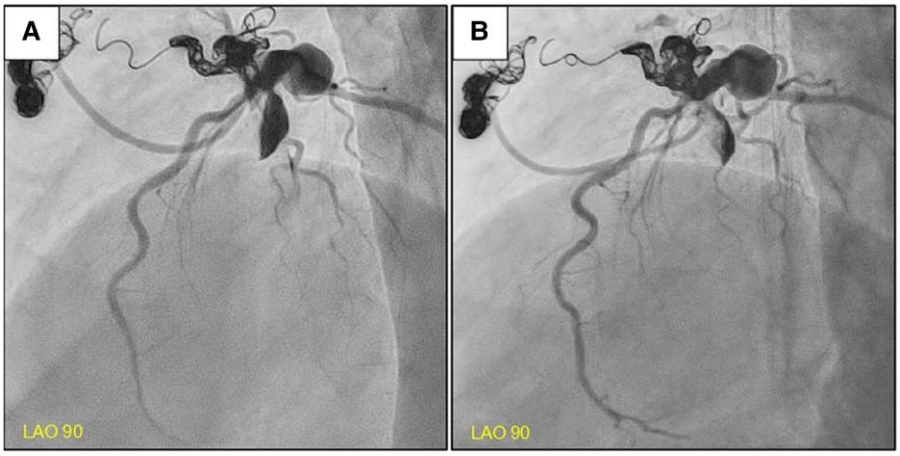


Figure 5 The follow-up coronary angiography results, 6 months after the procedure. The coronary angiography results showed that the size of the aneurysm 6 months after the procedure (*B*) was almost the same as that before the procedure (*A*).

diameter of the coronary artery proximal to the fistula. If patients experience symptoms, the shunt flow of the fistula should be managed. Treatment for asymptomatic patients should be considered in cases with moderate or large fistulas; the resting or stress electrocardiogram shows ischaemic changes; the Qp/Qs is above 1/1.5; or there is a possibility of progression of pulmonary hypertension or congestive heart failure, a history of infective endocarditis, or risk of coronary aneurysm rapture.[3] Some treatment options include surgery and percutaneous transcatheter closure (various occlusion types).[4]

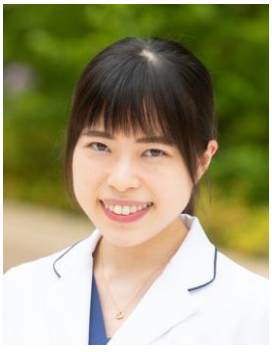
Surgery is suitable for extreme vessel tortuosity, multiple drainage sites, and coronary branches at sites of optimal device positioning or for cases involving intracardiac haemangiomas, side branch obstruction, coronary rupture, or aneurysmal degeneration.[7] The major advantages of transcatheter closure over surgery are avoiding cardiopulmonary bypass or median sternotomy and related iatrogenic complications, shorter recovery time and morbidity, improved cosmetic results, and lower cost. Transcatheter closure is possible in most patients; however, it may not be suitable depending on the running of the vessels and the position of the fistula. In our case, there was a high probability of selecting ligation of the fistulas combined with resection of the small aneurysm and coronary bypass grafting as the operative procedures. Thus, endovascular treatment targeting only fistulas was a better strategy, considering the patient's age. Detachable coils are considered safer than pushable coils because they can be retrieved and indwelled before detachment. The coil embolization technique effectively controlled the shunt flow of CPFs.

Clinical follow-up with echocardiography 1 month after percutaneous or surgical CAF closure is usually recommended.[4] If the patient remains asymptomatic and echocardiography shows improved results, close follow-up every 6–12 months should be conducted initially and then every 2 or 3 years. Patients who undergo fistula ligation require post-operative angiography or other imaging modalities to ensure that the fistula does not recanalize.[7] Moreover, patients who undergo percutaneous transcatheter closure also need regular angiographic follow-up to avoid short- or long-term complications because the coils may produce an artefact on cardiac computed tomography imaging.[6]

In this case, although slight recanalization of the left and right CPFs occurred, coil embolization continuously contributed to increased blood flow in the LAD and reduced symptomatic arteriovenous shunt flow. Moreover, although we were concerned that the aneurysm would enlarge due to increased blood flow in the LAD following CPF embolization, an increase in blood flow never affected the size of the coronary aneurysm during the subacute phase. The indications for surgical treatment of coronary artery aneurysms generally include severe coronary stenosis, complications (e.g. fistula formation), cardiac chamber compression, a high likelihood of rupture (e.g. rapidly increasing size of the aneurysm or pseudoaneurysm), and any type of aneurysm developing after coronary intervention. Coronary artery aneurysms should be monitored to determine if they meet the surgical criteria.[8] The coronary aneurysm in this case was not indicated for surgery, and follow-up coronary angiography showed no significant enlargement; therefore, it was followed up. In the future, if the fistulas get recanalized, coil embolization could be necessary. If the coronary artery aneurysm enlarges, surgery could be necessary.

There are few reports of transcatheter closure of CPF with detachable coils. Furthermore, to our knowledge, this is the first case report of transcatheter closure with detachable coils for multiple CPFs with a coronary artery aneurysm. Treatment with detachable coils has fewer complications and patients can be discharged early post-operatively. Therefore, it was very safe and effective.

Lead author biography



Sato Namura, MD, graduated in 2020 from the University of Okayama, Japan. She was a junior resident physician at the Yodogawa Christian Hospital until 2021. After completing junior residency, she worked as a clinical fellow in cardiovascular medicine at Yodogawa Christian Hospital. She is currently a cardiology specialist registrar and trained in general cardiovascular practice.

Supplementary material

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

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 patients in line with COPE guidance.

Conflict of interest: None declared.

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Data availability

The data underlying this article are available in the article and in its online Supplementary material.

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