

CASE REPORT

INTERMEDIATE

CLINICAL CASE

# Percutaneous Coil Embolization of Confluent Bilateral Coronary Artery Fistulas



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ABSTRACT

A 52-year-old woman presented with a continuous heart murmur at an annual health examination. Coronary computed tomography angiography showed confluent coronary artery fistulas meeting to form a large aneurysm fistulizing to the pulmonary artery. Percutaneous coil embolization was performed, resulting in the disappearance of the coronary artery fistulas and aneurysms. (**Level of Difficulty: Intermediate.**) (J Am Coll Cardiol Case Rep 2020;2:991-5)  
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HISTORY OF PRESENTATION

A 52-year-old woman presented with a continuous heart murmur from childhood observed at her annual health examination. Chest pain and shortness of

breath were absent. Her vital signs were within normal limits.

She had dyslipidemia and migraine, both stable by medication, and no family history of cardiac disease.

DIFFERENTIAL DIAGNOSIS

Continuous murmurs are hallmarks of left-to-right shunting, as seen in patent ductus arteriosus and ruptured aneurysm of sinus of Valsalva.

INVESTIGATIONS

The only significant blood abnormality was a slightly elevated brain natriuretic peptide concentration (45.6 pg/ml). Twelve-lead electrocardiography (ECG) showed normal sinus rhythm, whereas a double Master stress test with ECG showed no ischemic changes. Transthoracic echocardiography showed normal left ventricular function

LEARNING OBJECTIVES

- Coronary fistulas are relatively rare coronary artery abnormalities, but myocardial ischemia and congestive heart failure may occur if there is a large amount of shunt.
- When an aneurysm is also present, there is a risk of rupture. Treatment of coronary artery fistulas should be considered.
- Surgical ligation and transcatheter embolization can be used to treat coronary fistulas. Because the latter may be safer and less invasive, it should be used when the risk of adverse events is high.

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The authors attest they are in compliance with human studies committees and animal welfare regulations of the authors' institutions and Food and Drug Administration guidelines, including patient consent where appropriate. For more information, visit the *JACC: Case Reports* [author instructions page](#).

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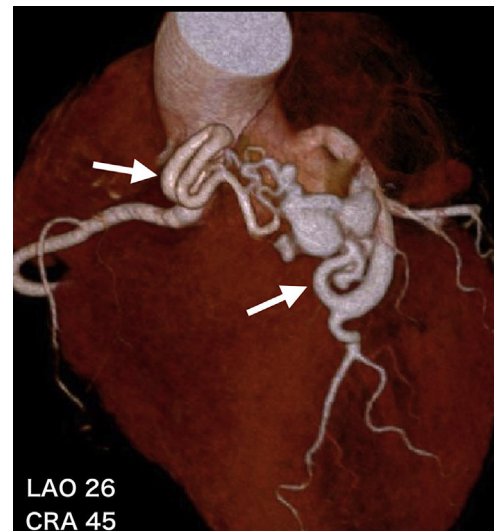
**ABBREVIATIONS  
AND ACRONYMS****CAF** = coronary artery fistula**CAG** = coronary angiography**CTA** = computed tomography angiography**ECG** = electrocardiography**PA** = pulmonary artery **$Q_p/Q_s$**  = ratio of pulmonary blood flow to systemic blood flow**RCA** = right coronary artery**RHC** = right-sided heart catheterization**RV** = right ventricle

with normal wall motion, valvular function, and dimension of the right ventricle (RV).

Coronary computed tomography angiography (CTA) revealed confluent coronary artery fistulas (CAFs) from the right coronary artery (RCA) and the left anterior descending artery meeting to form a 12.5-mm aneurysm fistulizing to the pulmonary artery (PA) (**Figure 1**). These findings were confirmed with invasive coronary angiography (CAG) (**Figures 2A to 2C**). Right-sided heart catheterization (RHC) showed significant oxygen step-up between the RV and the PA. Oxygen saturation was 70.3% and 80.2%, whereas the mean PA pressure (16 mm Hg) and mean pulmonary capillary wedge pressure (11 mm Hg) were within normal limits. The ratio of pulmonary blood flow to systemic blood flow ( $Q_p/Q_s$ ) was 1.47 on the basis of RHC.

**MANAGEMENT**

Although there were no signs of myocardial ischemia or congestive heart failure, transcatheter embolization for CAFs was planned because of the high  $Q_p/Q_s$  and the large aneurysm. The initial intervention was for the CAF of the left anterior descending artery. A 7-F guide catheter (JL-4.0, Heartrail II, Terumo Corp., Tokyo, Japan) was engaged in the left coronary artery through the right femoral artery. Unfractionated heparin maintained an activated whole blood clotting time of >250 s. A guidewire (Runthrough NS floppy, Terumo Corp.) and a 2.9-F microcatheter (Excelsior angle, Stryker, Kalamazoo, Michigan) were inserted into the tortuous vessel. The abnormal vessel was embolized with 2 detachable coils (Target XL soft, Stryker) up to 9 mm in diameter and 30 cm in length. Next, embolization of the vessel proximal to the RCA was performed. Using an AL-2.0 guide catheter (Heartrail II) and the same microcatheter, the fistula was embolized with 6 detachable coils (Target XL soft 360) up to 7 mm in diameter and up to 80 cm in length (**Figures 3A and 3B**). Post-embolization CAG confirmed the occlusion of all abnormal vessels (**Figures 4A and 4B**). Post-embolization RHC revealed  $Q_p/Q_s$  improvement to 0.7 and disappearance of the oxygen saturation step-up (RV, 70.1%; PA, 71.7%).

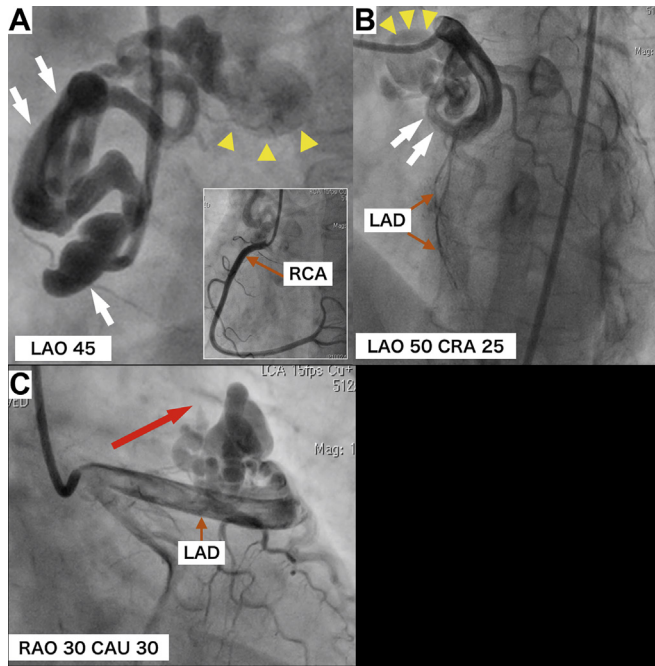
**FIGURE 1** Coronary Computed Tomography Angiography

The abnormal tortuous vessels originated proximal to the right coronary artery and the middle left anterior descending artery (**arrows**) with a common giant aneurysm. CRA = cranial; LAO = left anterior oblique.

**DISCUSSION**

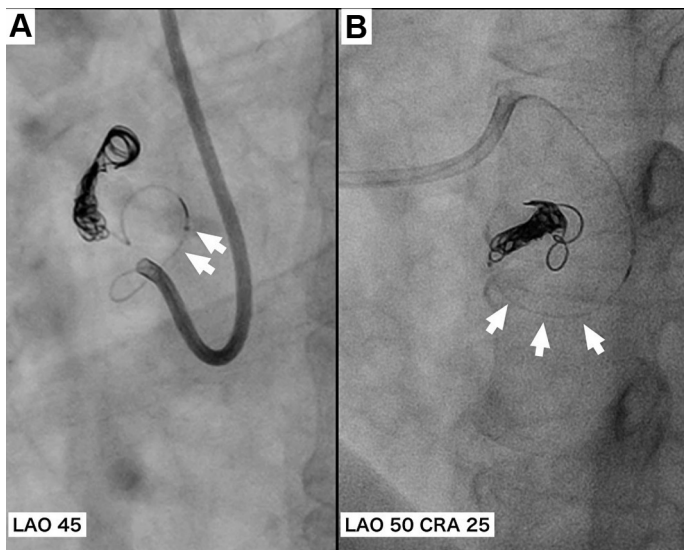
Recently, more CAFs have been incidentally diagnosed because of developments in CAG and coronary CTA quality (1). Most CAFs are congenital, developing in the early embryonic period (2). CAFs are reportedly present in 0.002% of the general population and represent 0.4% of all cardiac malformations (2,3). CAFs are incidentally found in 0.13% to 0.22% of adults undergoing CAG (4). A single fistula is the most common presentation (70% to 90% of cases); multiple fistulas involving both coronary arteries account for 10% to 20% of cases (1-3). Although some studies have reported that RCA-originating CAFs are the most common presentation (2,3), Said (1) reported that CAFs originating from the left coronary artery were the most common in the decade of the 2000s. CAFs draining into right-sided heart structures (e.g., PA, right atrium, and RV) are common, whereas cases involving left-sided heart structures (e.g., left ventricle, coronary sinus, and superior vena cava)

**FIGURE 2** Coronary Angiography

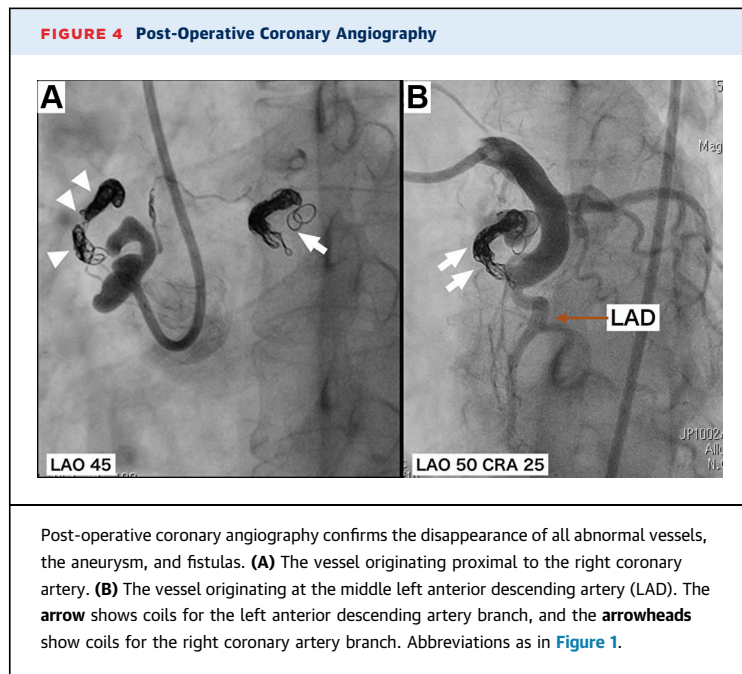


The abnormal tortuous vessels (**short arrows**) and the (**A**) right coronary artery (RCA) and (**B and C**) left anterior descending artery (LAD) fistula (**long arrow**). A small window shows the relationship between the right coronary artery and the abnormal vessels. Feeding vessels are indicated with **arrows**. **Arrowheads** indicate the common aneurysm. CAU = caudal; RAO = right anterior oblique; other abbreviations as in [Figure 1](#).

**FIGURE 3** Coil Embolization



(**A**) The abnormal vessel originating proximal to the right coronary artery was embolized with 2 detachable coils (**arrows**), and (**B**) the vessel originating at the middle left anterior descending artery was embolized with 6 detachable coils. Abbreviations as in [Figure 1](#).

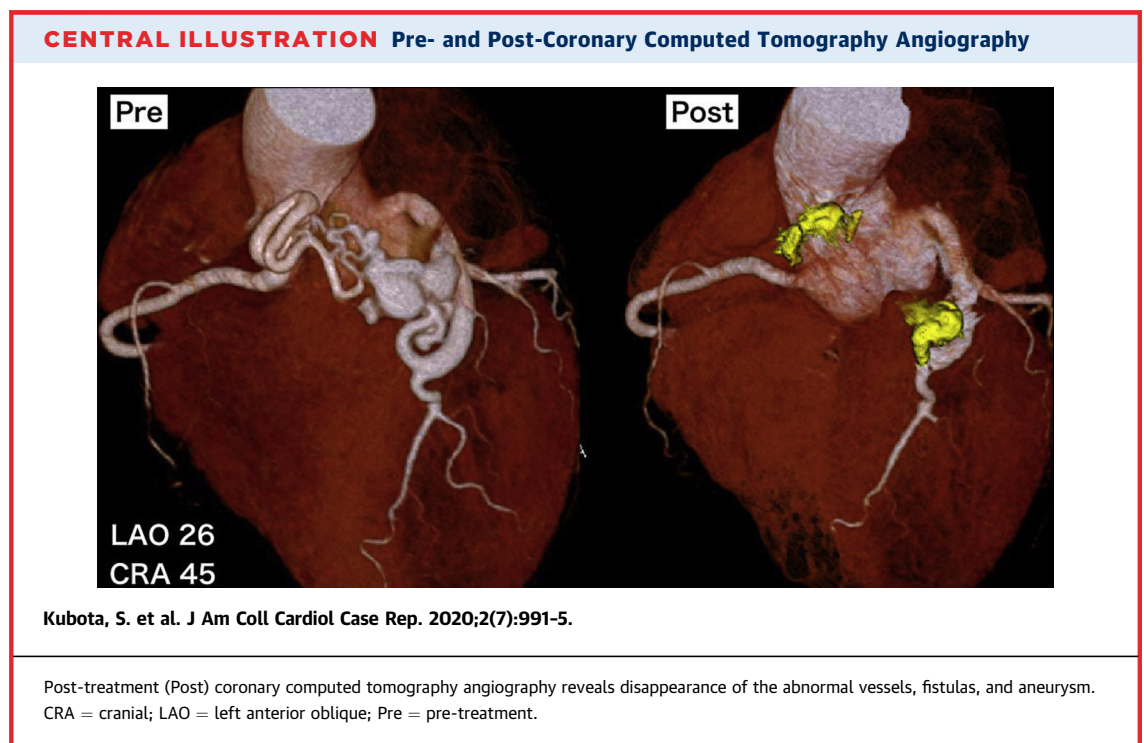


are rare (1,2). Feeding arteries are primarily ectatic and tortuous. For large fistulas, the most common symptoms are dyspnea on exertion caused by congestion and PA hypertension caused by a shunt to right-sided heart structures. Chest pain from the

blood steal phenomenon from the myocardium is another common symptom. Several reports have addressed infectious endocarditis (1,3). Coronary aneurysms involving vessels that are 1.5-fold more dilated than normal vessels are reported in 14% of cases (1).

The appropriate therapeutic indication has remained controversial. According to current guidelines, CAFs causing myocardial ischemia and arrhythmia should be treated if they are small or moderate in size. The size is specified as follows: small,  $<1\times$  distal reference; medium,  $1$  to  $2\times$  distal reference; and large,  $>2\times$  distal reference (5). Several studies reported that indications for closure are high  $Q_p/Q_s$  (1.3 to 1.5), the presence of large fistulas, ECG-indicated myocardial ischemia, PA hypertension progression, congestive heart failure, and/or coronary aneurysm with a high risk of rupture (6).

Transcatheter embolization or closure and surgical obliteration effectively treat CAFs. Transcatheter embolization involving coils, covered stents, and umbrella devices has increased recently because it is less invasive and less risky than surgical treatment (7,8). Post-treatment recurrence is lower following transcatheter embolization than following surgical ligation (9% to 14% vs. 25%) (9).



Although our patient had no symptoms suggestive of myocardial ischemia, we performed transcatheter embolization because of the high  $Q_p/Q_s$  and the giant aneurysm. There are no definite indications for treating coronary artery aneurysms, but some reports describe a risk of rupture for aneurysms 3-fold more dilated than normal vessels (10).

Transcatheter embolization is indicated when the CAF is not severely tortuous and its distal site is accessible and able to be embolized by a closure device, as in this case. The patient avoided all surgery-related complications, including surgical stress, infections, wound healing problems, bleeding, general anesthesia adverse events, and the higher possibility of recurrence.

To our knowledge, there are no available data prognosis for untreated CAFs or aneurysms. However, transcatheter embolization, which is safe and has a low risk, may improve prognosis.

## FOLLOW-UP

A 3-month follow-up coronary CTA confirmed disappearance of the abnormal arteries and fistulas (**Central Illustration**). The patient has had no post-treatment symptoms or complications.

## CONCLUSIONS

Coronary fistulas are relatively rare and may rupture in patients with aneurysm. Transcatheter treatment should be considered because it is safer and less invasive than other methods.

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**KEY WORDS** coronary vessel anomaly, imaging, percutaneous coronary intervention, treatment