

# Transcatheter Coil Embolization for Coronary Artery Fistula with Giant Aneurysm Originating from Left Circumflex Artery and Drainage to Right Ventricle

Chiung-Zuan Chiu<sup>1,2</sup> and Kou-Gi Shyu<sup>1,3</sup>

Coronary artery fistulae (CAF) represent the most frequent congenital anomalies influencing hemodynamic parameters of coronary arteries. However, CAF complicated with a huge aneurysm does not frequently occur. Determining the optimal management of symptomatic patients with CAF and aneurysm still remains a challenging problem, and has been traditionally considered as an indication for cardiac surgery. The use of transcatheter coil embolization for CAF has emerged as a less invasive strategy, and has recently been considered as an alternative to surgical correction. We reported a 71-year-old male patient with a congenital CAF, originating from the left circumflex artery and draining into the right ventricle, complicated with a giant aneurysm presenting with typical angina pectoris and exertional dyspnea. This patient underwent successful transcatheter closure of both the fistula and aneurysm using a coil embolization technique, and had an uneventful recovery after treatment.

**Key Words:** Aneurysm • Angina pectoris • Coil embolization • Coronary artery fistulae • Transcatheter closure

## INTRODUCTION

Coronary artery fistulae (CAF) represent a relatively rare clinical manifestation, with an estimated prevalence of 0.2% to 0.6% in patients undergoing coronary angiography.<sup>1-4</sup> CAF are either congenital or an acquired coronary artery anomaly, characterized by a fistula communicating between the coronary artery and cardiac chambers or great vessels, bypassing the myocardial capillary network to result in coronary artery steal phenomenon. The

clinical presentations or symptoms/signs are dependent upon the severity of the shunt and may include angina pectoris, myocardial ischemia, myocardial infarction, congestive heart failure, arrhythmias, or syncope.<sup>4</sup> Myocardial ischemia, without evidence of coronary artery stenosis, is related to reduced coronary blood flow distal to the fistula. Other rarer severe complications, including infective endocarditis, aneurysm formation with or without rupture, pericardial effusion, stroke, and sudden cardiac death have been described previously.<sup>4,5</sup> Current indications for CAF closure are limited to patients with clinical symptoms/signs, or to asymptomatic patients with evident left to right shunts to prevent complication development, especially in the pediatric or younger patients.<sup>6,7</sup> The optimal strategy to manage symptomatic patients still remains controversial due to the lack of long-term clinical follow-up. Consequently, conservative medical treatment remains the major treatment for most CAF patients with limited symptoms/signs.<sup>7</sup> Al-

Received: July 20, 2011

Accepted: January 19, 2012

<sup>1</sup>Division of Cardiology, Shin Kong Wu Ho-Su Memorial Hospital, Taipei; <sup>2</sup>School of Medicine, Fu-Jen Catholic University, New Taipei City; <sup>3</sup>Graduate Institute of Clinical Medicine, College of Medicine, Taipei Medical University, Taipei, Taiwan.

Address correspondence and reprint requests to: Dr. Kou-Gi Shyu, Division of Cardiology, Shin Kong Wu Ho-Su Memorial Hospital, No. 95, Wen Chang Road, Taipei 111, Taiwan. Tel: 886-2-2833-2211; Fax: 886-2-2812-9733; E-mail: juice243@hotmail.com

though spontaneous closure or regression of CAF was reported in previous articles,<sup>8,9</sup> surgical correction has been traditionally considered as the treatment of choice for CAF closure, with excellent long-term results.<sup>6,7</sup> Transcatheter closure approaches, including covered stent, coil embolization or occluder, have emerged as less-invasive strategies since 1983 and are recently considered an alternative treatment with similar outcomes.<sup>7,10,11</sup> However, transcatheter management of patients with fistulae complicated with aneurysms remains a challenging problem and has been traditionally considered an indication for surgical correction.<sup>6,7</sup> We reported an extremely rare case of a patient with a symptomatic congenital CAF originated from the left circumflex artery (LCX) draining into the right ventricle (RV), complicated with a huge aneurysm who underwent successful transcatheter closure of both the fistula and the aneurysm using a coil embolization technique.

## CASE REPORT

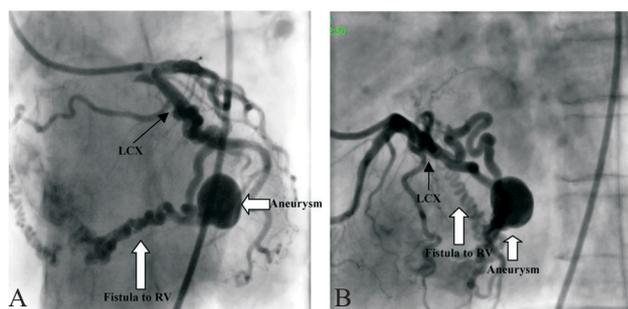
This patient was a 71-year-old heavy smoker with a past history of chronic obstructive pulmonary disease (COPD). He had chronic exertional dyspnea and was under regular medical treatment for his pulmonary problem at our chest outpatient clinic. He denied any other systemic disease, such as hypertension, diabetes, or dyslipidemia. The patient was admitted to our medical center due to aggravated chest tightness/chest pain and dyspnea caused by exertion during the week prior to admission. His symptoms persisted and became exacerbated even under previous medications for his COPD. He came to our cardiovascular clinic for help, whereafter aspirin and calcium channel blockers (diltelan) were prescribed after a preliminary diagnosis of coronary artery disease. The patient's situation did not improve under medical management, and the patient was referred to coronary angiography.

On examination, the patient was alert and communicative. His temperature was 37.0 °C, with blood pressure of 132/78 mmHg, a little bit higher than his usual measurements. Cardiac examination demonstrated a regular pulse with 88 beats per minute, and no heart murmur could be detected. Neither gallop nor rub was found. The abdominal examination was normal and no abdominal

aortic bruit was heard. Radial, brachial, femoral, and dorsal pedal pulses were 2+, and both feet were warm to the touch. Laboratory evaluation demonstrated normal findings. The patient characterized his chest pain as dull and compressive, lasting for more than three days. Subsequent electrocardiography showed a bifascicular block with non-specific ST-T changes. Coronary angiography revealed a macro-fistula which originated from the LCX, complicated with a huge aneurysm (14.8 mm × 16.2 mm in diameter) and drained into the RV without evidence of obstructive coronary artery disease (Figure 1). We thought that the patient's typical ischemic symptoms were caused by coronary steal phenomenon secondary to the fistula. Consequently, transcatheter closure of both CAF and aneurysm was performed to relieve the patient's clinical symptoms.

### Transcatheter closure technique

Transcatheter closure was attempted by a retrograde approach via femoral artery access with a 7 French (Fr) sheath, and using a standard microcoil embolization technique as previously described.<sup>10,11</sup> Angiographic views allowed adequate visualization of the fistula (Figure 1) in order to determine the appropriate site for device delivery, and the coil size required. A 7 Fr XB 3.50 guide catheter (Cordis Corp., Miami, Florida, USA) was positioned at the left coronary ostium. The delivery catheter, a 2.6 Fr × 150 cm Excelsior 1018 (144181) microcatheter guide (Boston Scientific, Natick, Massachusetts, USA), was advanced under fluoroscopic guidance over a ASAHI NEO's Fielder FC guide wire (ASAHI INTEC., Nagoya, Japan) directly to the distal coronary artery (LCX) in order to occlude the drainage orifice and minimize the risk of distal embolization. Hemodynamic parameters were assessed by use of microcatheter at the level of the fistula. The microcatheter was then positioned in the proximal segment of the fistula (diameter of the vessel at this point: 2.25 mm) and a single stainless-steel MWCE<sup>®</sup> 18S-4/2-TORNADO microcoil (4-2 mm; Cook Medical, Inc., Bloomington, Indiana, USA) was deployed and advanced by using a straight 0.038-inch guidewire (Figure 2, panels A and B). After delivery of the device, coronary angiography confirmed partially successful occlusion of the fistula with less visible residual flow from the fistula. An additional two microcoils (stainless steel

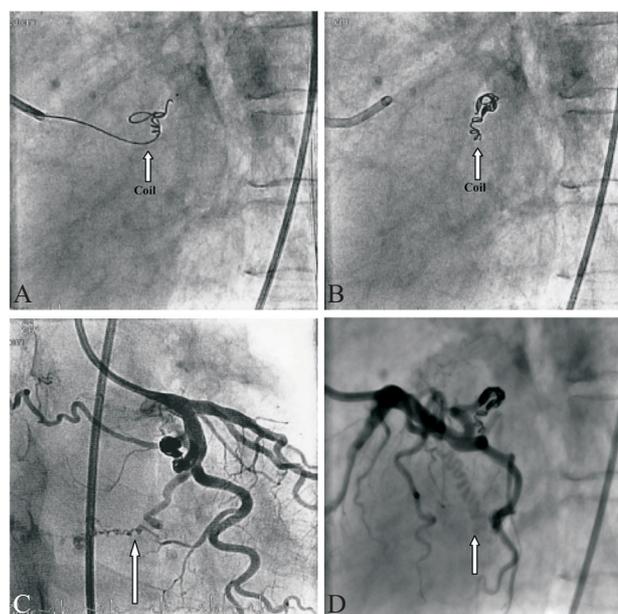


**Figure 1.** Coronary angiography showed macro-fistula from left circumflex artery (LCX) to right ventricle (RV) with a huge aneurysm. (A) Left anterior oblique (LAO) 15 degree and caudal 38 degree. (B) True posterior anterior (PA) 0 degree and lateral 90 degree.

MWCE<sup>®</sup> 18S-6/2-TORNADO; 6-2 mm) were deployed again with no more aneurysm noted and only minimal residual fistula flow could be seen after embolization (Figure 2, panels C and D). Following the above procedures, coils were implanted so as to merge together forming a conglomeration. Coronary angiography was repeated to confirm the quality of the occlusion, and heparin (100 U per kg) was given during the procedure. There were no ECG changes noted which might suggest myocardial ischemia following the procedure; no procedural complications occurred and the patient was discharged on the following day. The patient subsequently reported that his earlier clinical symptoms had completely disappeared, and that his recovery had continued uneventfully.

## DISCUSSION

CAF are the most frequent congenital anomalies to influence hemodynamic parameters of the coronary arteries. We described here a unique case with a symptomatic congenital CAF complicated with a huge aneurysm originating from the LCX and draining into the RV. The patient underwent successful transcatheter closure of both the fistula and aneurysm using a microcoil embolization technique. Congenital CAF are usually solitary anomalies, and approximately 20% of them may be complicated with aneurysm formation.<sup>7</sup> The incidence of CAF from LCX occurs with a lesser frequency as compared with those from the left anterior descending artery or right coronary artery in previous reports.<sup>1-4</sup> However, congenital CAF which originated from the LCX and drained into the RV are even more atypical, and rarely



**Figure 2.** After coil embolization (A and B), aneurysm disappeared and the flow of fistula to RV also decreased significantly (C and D). (C) LAO 15 degree and caudal 38 degree. (D) True PA 0 degree and lateral 90 degree.

seen. Interestingly, and for the first time, we demonstrated the transcatheter closure of a macrofistula complicated with a huge aneurysm. In our case, an evident shunt from the coronary fistula resulted in clinical ischemic symptoms in the patient. In addition, the huge aneurysm may become complicated with thrombus formation or a ruptured aneurysm if left untreated.<sup>5,12,13</sup> In 1983, Reiday et al. first successfully performed transcatheter closure of CAF.<sup>7</sup> Transcatheter closure approaches have emerged as a less invasive strategy for surgical correction, since the safety and efficacy of percutaneous closure techniques have been demonstrated in previous reports.<sup>7,10,11,14</sup> Currently, microcoil embolization is considered the predominant method for transcatheter closure, and recent developments in catheter and coil technologies have made it a safe procedure.<sup>10,11</sup> Coils have been used most commonly in small to medium sized CAFs. Mavroudis et al.<sup>7</sup> recommended elective coil occlusion in patients who satisfy the following criteria: 1) absence of multiple fistulae; 2) a single narrow drainage site; 3) absence of large branch vessels; 4) safe accessibility to the coronary artery supplying the fistula. Less common complications of coil embolization include transient ischemic electrocardio-

graphic changes, arrhythmias, device embolization, and myocardial infarction.<sup>7,10,11</sup> Close long-term follow-up with coronary angiography and myocardial scintigraphy after transcatheter closure of the CAF is very important. Recently, transcatheter closure techniques have also been suggested as an effective alternative to cardiac surgery for certain select patients, particularly when a single drainage site is documented.<sup>10,11</sup> However, limited data are available regarding the long-term results of percutaneous management of patient CAF as a course of treatment. A detailed evaluation of the origin and drainage sites of the fistulae is essential before selecting the appropriate closure technique. We demonstrated here that a transcatheter retrograde closure approach using a microcoil embolization technique for a fistula with huge aneurysm is relatively safe and effective. However, the need for appropriate patient selection before considering percutaneous embolization closure of symptomatic fistulae should be emphasized.

Regarding aneurysm formation after CAF, the most severe complication is aneurysm rupture. Rupture of congenital aneurysmal fistulae occurred more often in females.<sup>5</sup> Identified risk factors for rupture, cardiac tamponade, and possible death were saccular aneurysm, Asian ethnicity, left coronary artery origin of the aneurysmal fistula, and a history of hypertension.<sup>5,12,15</sup> So it is necessary to manage aneurysm formation after CAF with a high risk of rupture to prevent further complications. Recent studies about management of symptomatic patients with CAF by coil embolization are limited to cases with small patient numbers and short-term follow-up periods. Residual or recurrent shunts after transcatheter closure have been reported in 10-20% of patients, and may require further procedures to achieve complete occlusion. In addition, limited data regarding the optimal follow-up of patients after a successful transcatheter closure of CAF are currently available in the literature. Therefore, the absence of a standardized, detailed follow-up protocol may prevent one from drawing any conclusion concerning long-term results and outcome predictors after percutaneous CAF management.

## CONCLUSION

Coronary artery fistula is a rare coronary artery ab-

normality that is difficult to detect clinically because of nonspecific manifestations. Most CAFs are benign but some may produce life-threatening symptoms. Coronary angiography, which demonstrates the nature of the anatomy of CAF, is crucial to the management of patients with this condition. Transcatheter coil embolization may be indicated in suitable cases with symptomatic CAF, even complicated with an aneurysm, and may replace traditional surgical correction.

## REFERENCES

1. Yamanaka O, Hoobbs RE. Coronary artery anomalies in 126,595 patients undergoing coronary arteriography. *Cathet Cardiovasc Diagn* 1990;21:28-40.
2. Gillebert C, Van Hoof R, Van de Werf F, et al. Coronary artery fistulas in an adult population. *Eur Heart J* 1986;7:437-43.
3. Kardos A, Babai L, Rudas L, et al. Epidemiology of congenital coronary artery anomalies: a coronary arteriography study on a central European population. *Cathet Cardiovasc Diagn* 1997; 42:276-7.
4. Chiu CZ, Shyu KG, Cheng JJ, et al. Angiographic and clinical manifestations of coronary fistulae in Chinese people-15 years experience. *Circ J* 2008;72:1242-8.
5. Said SA, Schroeder-Tanka JM, Mulder BJ. Female gender and the risk of rupture of congenital aneurysmal fistula in adults. *Congenit Heart Dis* 2008;3:63-8.
6. Kamiya H, Yasuda T, Nagamine H, et al. Surgical treatment of congenital coronary artery fistulas: 27 years' experience and a review of the literature. *J Card Surg* 2002;17:173-7.
7. Luo L, Kebede S, Wu S, Stouffer GA. Coronary artery fistulae. *Am J Med Sci* 2006;332:79-84.
8. Schleich JM, Rey C, Gewillig M, et al. Spontaneous closure of congenital coronary artery fistulas. *Heart* 2001;85:E6.
9. Nakatani S, Nanto S, Masuyama T, et al. Spontaneous near disappearance of bilateral coronary artery – pulmonary artery fistulae. *Chest* 1991;99:1288-9.
10. Armsby LR, Keane JF, Sherwood MC, et al. Management of coronary artery fistulae. Patient selection and results of transcatheter closure. *J Am Coll Cardiol* 2002;39:1026-32.
11. Collins N, Mehta R, Benson L, et al. Percutaneous coronary artery fistula closure in adults: technical and procedural aspects. *Catheter Cardiovasc Interv* 2007;69:872-80.
12. Hirose H, Amano A, Yoshida S, et al. Coronary artery aneurysm associated with fistula in adults: collective review and a case report. *Ann Thorac Cardiovasc Surg* 1999;5:258-64.
13. Takahashi M, Sekiguchi H, Fujikawa H, et al. Multiple saccular aneurysm formation in a patient with bilateral coronary artery fistula: a case report and review of the literature. *Cardiology* 1995;86:174-6.

14. Wang W, Wang YJ, Fu SL, Gong FQ. Transcatheter amplatzer vascular plug closure of fistula between right pulmonary artery and left atrium. *Acta Cardiol Sin* 2011;27:128-31.
15. Loo B, Cox ID, Morgan-Hughes GJ, et al. Thrombotic occlusion of giant circumflex artery aneurysm after ligation of arteriovenous fistula. *Circulation* 2010;122:e447-8.