

## An Unusual Case of Dual Coronary Artery Fistulas to Main Pulmonary Artery

Mei-Cheng Lai, MD; Wei-Jan Chen, MD, PhD; Cheng-Wen Chiang<sup>1</sup>, MD;  
Yu-Lin Ko, MD, PhD

Coronary artery fistula is an anomaly in which a coronary artery directly connects to a cardiac chamber or great vessel. Its incidence is around 0.1 to 1 % in the adult population. Dual coronary artery fistulas are far less common and their incidence is estimated to be around 5% in patients with this anomaly. Closure of the fistulas is indicated in patients with myocardial ischemia, large left to right shunt, congestive heart failure or other complications. Herein, we report a 64 year-old man with dual coronary artery fistulas presenting with exertional chest pain. The fistulas were initially suspected on transesophageal echocardiogram because of abnormal flow with a mosaic pattern between the left anterior descending and main pulmonary arteries. Selective coronary angiogram confirmed the diagnosis and revealed fistulous connections from the proximal left anterior descending and ostial right coronary arteries to the main pulmonary artery. The patient became symptom-free after surgical closure of the fistulas. (*Chang Gung Med J* 2002;25:51-5)

**Key words:** coronary artery fistulas, echocardiography, coronary angiography.

Coronary artery fistula, first reported by Krause in 1865,<sup>(1)</sup> was thought to be a rare anomaly of the coronary arteries. It has been reported more frequently in recent years because of the increase in coronary artery procedures performed. The incidence in the adult population is around 0.1 to 1%<sup>(2-4)</sup> and it may be either congenital or acquired. Only 5% of the fistulas are dual. However, patients with ischemic chest pain owing to dual or multiple coronary fistulas without significant coronary artery stenosis are rather rare.<sup>(5-7)</sup>

### CASE REPORT

A 64 year-old man was admitted to our hospital because of progressive chest tightness and breath-

lessness for 2 months. He had been well in the past and there was no previous history of chest trauma. The only risk factor for coronary artery disease was advanced age. His chest discomfort was effort-related, and relieved by rest, and was accompanied by diaphoresis and radiation to the bilateral jaws and shoulders. The duration was less than 10 minutes. On examination, the only positive finding was a grade II/VI continuous murmur loudest at the left 2nd intercostal space. His baseline electrocardiogram revealed non-specific ST-T changes. He was then referred for a Treadmill exercise test (Cornell's protocol) for the possibility of ischemic heart disease. However, it was non-diagnostic because of inadequate exercise load. Transesophageal echocardiography in the short axis view showed abnormal

---

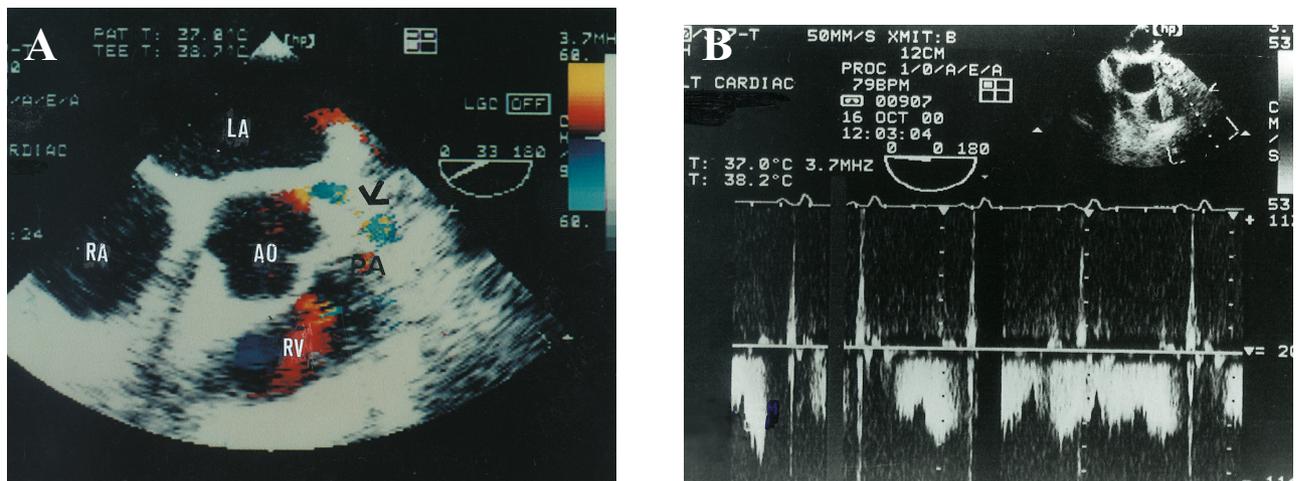
From the First Cardiovascular Division, Department of Internal Medicine, Chang Gung Memorial Hospital, Taipei; <sup>1</sup>Division of Cardiovascular, Department of Internal Medicine, Cathay General Hospital, Taipei.

Received: Mar. 8, 2001; Accepted: Jun. 11, 2001

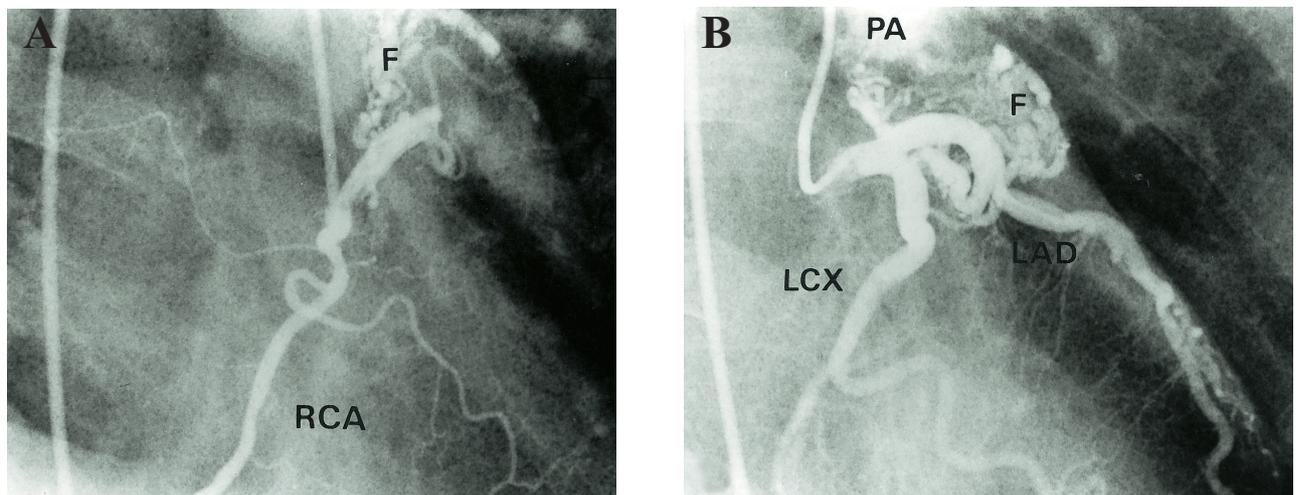
Address for reprints: Dr. Mei-Cheng Lai, The First Cardiovascular Division, Chang Gung Memorial Hospital, 5, Fu-Shin Street, Kweishan, Taoyuan, 333, Taiwan, ROC. TEL: 886-3-3281200 ext. 8162; E-mail: ufjl0378@ms5.hinet.net

flow with a mosaic pattern between the left anterior descending artery and the main pulmonary artery (Fig.1A), which was continuous on continuous-wave Doppler (Fig.1B). Under the suspicion of coronary artery fistula, he underwent cardiac catheterization. Coronary angiogram revealed bilateral coronary artery fistulas from the ostial right coronary artery and proximal left anterior descending artery draining

into the main pulmonary artery (Fig. 2). Left ventricular function was normal (ejection fraction 78%). No O<sub>2</sub> step-up was found during a pull-back tracing of the right heart channel. Due to persistent symptoms even with anti-angina therapy, he was referred to cardiovascular surgery for fistula closure. The orifices of the fistulas were closed with 4-0 prolene via pulmonary arteriotomy under partial cardiopul-



**Fig. 1** (A) Transesophageal echocardiogram in the short axis view showing abnormal flow with mosaic patterns on color Doppler between the left anterior descending artery and the main pulmonary artery ( arrowhead ). Ao = aorta; PA = pulmonary artery, LA = left atrium; RV = right ventricle; RA = right atrium. (B) Continuous-wave Doppler echocardiogram revealing a continuous jet at the drainage site.



**Fig. 2** Selective coronary angiograms showing 2 fistulas connecting from the ostial right coronary artery (A) and proximal left anterior descending artery (B) to the main pulmonary artery in a 30-degree right anterior-oblique projection. F = fistula; RCA = right coronary artery ; LAD = left anterior descending artery; LCX = left circumflex artery; PA = pulmonary artery.

monary bypass. He has been free of chest pain during the follow-up period of one year.

## DISCUSSION

Coronary artery fistula, a direct connection of the coronary artery with a cardiac chamber or great vessel, may develop due to enlargement of the capillary network during cardiac embryogenesis or main coronary arteries being attached to the pulmonary trunk at the time of separation.<sup>(8)</sup> The fistulas originate slightly more often from the right than the left coronary artery with only 5% originating from both. The right side of the heart is the drainage site in more than 90% of cases of single coronary artery fistula. Furthermore, the main pulmonary artery is the site of drainage in more than 50% of dual coronary fistulas as was the case in our patient.<sup>(9)</sup>

It is note-worthy that one fistula originates from ostium of right coronary artery in our patient. Coronary steal phenomenon may be prominent in this situation resulting from proximal location of the fistulas and its bilaterality.

Most patients with coronary artery fistula are asymptomatic at diagnosis and it is often an incidental finding on coronary angiogram. Some patients are referred for cardiac evaluation for a continuous murmur at the site of drainage. But complications do occur and patients may present with heart failure, chest pain, infective endocarditis, arrhythmia, aneurysmal changes or fistula rupture. As found in our patient, coronary steal phenomenon is the main manifestation of fistula. It should be noted that patients with dual coronary artery fistulas presenting with angina pectoris usually have other associated lesions, such as significant stenosis due to coronary atherosclerosis, aortic stenosis, hypertrophic obstructive cardiomyopathy or others,<sup>(7)</sup> all of which may also cause coronary ischemia. This is the first reported case of dual coronary artery fistulas presenting with exertional chest pain without definite coronary artery stenosis or other associated lesions.

Surgical management of coronary artery fistula is clearly indicated in patients with evidence of myocardial ischemia, a large left to right shunt or congestive heart failure.<sup>(7)</sup> It has been reported that the fistulas can be repaired without cardiopulmonary bypass with low morbidity and mortality.<sup>(10)</sup>

Recently, successful transcatheter coil embolization of the fistula has also been reported.<sup>(11)</sup> Some authors advocate fistula closure even in asymptomatic patients for prevention of complications because of the high success rate and low risk of complications.<sup>(12,13)</sup> We believe that fistula closure was clearly indicated in this patient because of symptomatic coronary steal. On the other hand, non-surgical modalities such as transcatheter coil embolization of the fistula may be promising. They may become an attractive alternative for surgical intervention after the procedures and devices are standardized.

## REFERENCES

1. Krause W. Ueber den Ursprung einer akzessorischen A. coronaria aus der A. pulmonalis. *Z Ratl Med* 1865; 24:225-9. [German]
2. Fernandes ED, Kadivar H, Hallamnn GL, Reul GJ, Ott DA, Cooley DA. Congenital malformations of the coronary arteries: the Texas Heart Institute experience. *Ann Thorac Surg* 1992;54:732-40.
3. Osamu Yamanaka, Robert E. Hobbs. Coronary artery anomalies in 126,595 patients undergoing coronary arteriography. *Cathet Cardiovasc Diagn* 1990;21:28-40.
4. Vavuranakis M, Bush CA, Boudoulas H. Coronary artery fistulas in adults: incidence, angiographic characteristics and natural history. *Cathet Cardiovasc Diagn* 1995;35:116-20.
5. Shu-Meng Cheng, Shih-Ping Yang, Ween-Yuang Lai, Fang-Yian Lee, Philips Yu-An Ding. Congenital coronary artery anomalies - a retrospective analysis of 28 patients and literature review. *Acta Cardiol Sin* 1997;13:85-92.
6. Davendra Mehta, David Redwood, David E. Ward. Multiple bilateral coronary arterial to pulmonary artery fistulae in an asymptomatic patient. *Int J Cardiol* 1987; 16:96-8.
7. Baim D, Kline H, Silverman J. Bilateral coronary artery to pulmonary artery fistulas : Report of 5 cases and review of literature. *Circulation* 1982;65:810-815.
8. Dervan J, Vlay S. Coronary artery to pulmonary artery fistula: A potential new mechanism. *Am Heart J* 1989; 117:921-3.
9. Levin DC, Fellows KE, Abrams HL. Hemodynamically significant primary anomalies of the coronary arteries. *Circulation* 1978;58:224-35.
10. Goto Y, Abe T, Sekine S, Iijima K, Kondoh K, Sakurada T. Surgical treatment of the coronary artery to pulmonary artery fistulas in adults. *Cardiology* 1998;89:252-6.
11. Wax DF, MaGee AG, Nykanen D, Benson LN. Coil embolization of a coronary artery to pulmonary artery fistula from an antegrade approach. *Cathet Cardiovasc*

- Diagn 1997;42:68-9.
12. Liberthson RR, Sagar K, Berkoben JP, Weintraub RM, Levene FH. Congenital coronary arteriovenous fistulae. Report of 13 patients, review of literature and delineation of management. *Circulation* 1979;59:849-54.
  13. Hitoshi Hirose, Masatake Takagi, Naotaka Miyagawa, Hiroshi Hashiyada, Takafumi Yamada, Seiichi Tada and Toshiyasu Kugimiya. Coronary atherosclerosis with dual coronary artery fistulas. *Scand Cardiovasc J* 1998;32:312-4.

