Yonsei Medical Journal
Vol. 38, No. 3, pp. 190~192, 1997

# Bilateral Coronary Arteriovenous Fistula Coexistent with Atrial Septal Defect and Pulmonary Stenosis

**Jong-Won Ha, Hyung-Joon Lee, Joo-Yong Lee, Ho-Young Kim, Junghan Yoon, and Kyung-Hoon Choe**

*A coronary arteriovenous fistula consists of a communication between a coronary artery and a cardiac chamber, a great artery or the vena cava. It is the most common anomaly that can affect coronary perfusion. Bilateral involvement of coronary fistula, however, constitutes an uncommon subgroup of coronary arteriovenous fistulas. We report a case which shows a rare occurrence of bilateral coronary arteriovenous fistula coexistent with atrial septal defect and pulmonic stenosis.*

**Key Words:** Bilateral coronary arteriovenous fistula

A coronary arteriovenous fistula consists of a communication between a coronary artery and a cardiac chamber, a great artery or the vena cava. It is the most common anomaly that can affect coronary perfusion. Usually, both coronary arteries arise normally from the aorta at their normal sites, in contrast to the anomalous origin of a coronary artery from the pulmonary trunk. Approximately half of all patients with coronary fistulas develop symptoms of congestive heart failure, myocardial ischemia or myocardial infarction resulting from a coronary steal. The other half may remain totally asymptomatic. This anomaly is usually congenital in origin but it can be acquired in some clinical settings (Rose *et al.* 1978; Sandhu *et al.* 1988). Bilateral involvement of coronary fistula, however, constitutes an uncommon subgroup of coronary arteriovenous fistulas. This report describes the bilateral involvement of coronary arteriovenous fistula coexistent with atrial septal defect and pulmonary stenosis.

Received April 30, 1997
Accepted June 5, 1997
Division of Cardiology, Department of Internal Medicine, Wonju College of Medicine, Yonsei University, Wonju, Korea
Address reprint request to Dr. J.W. Ha, Cardiology Division, Yonsei Cardiovascular Center, Yonsei University College of Medicine, C.P.O. Box 8044, Seoul, Korea

## CASE REPORT

A 47-year-old male was referred for evaluation of electrocardiographic abnormalities discovered during a routine checkup. He had no particular prior medicosurgical history. Physical examination revealed wide, fixed-splitting of S2 with grade 3/6 ejection systolic murmur at the left upper sternal border. Chest X-ray revealed an enlarged main and left pulmonary artery without increased pulmonary vascular markings. Transthoracic echocardiography demonstrated mild systolic doming of the pulmonic valve and an approximately 2.2 cm-sized secundum atrial septal defect with left-to-right shunt. On cardiac catheterization, there was 8% oxygen step-up between mixed venous blood and the right atrium.

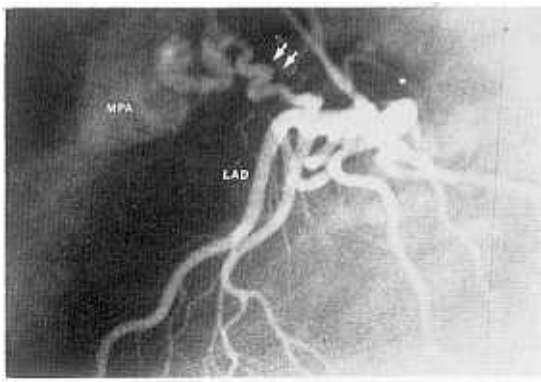


*Fig. 1. Left coronary arteriography at lateral projection revealed an abnormal tortuous vessel that coursed superiorly and drained into the main pulmonary artery.*

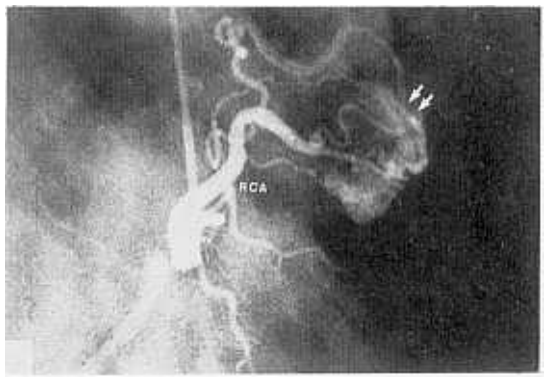


*Fig. 2. Right coronary arteriography at right anterior oblique projection revealed the coronary arteriovenous fistula through conus branch.*

However, there was no significant oxygen step-up between the right ventricle and pulmonary artery. There was an approximately 28 mmHg peak-to-peak pressure gradient between the right ventricle and main pulmonary artery. Right pulmonary venous angiography at the left anterior oblique projection revealed left-to-right shunt through the atrial septal defect. Coronary arteriography revealed left anterior descending artery, giving rise to a tortuous vessel that coursed cephalad in direction and drained into the main pulmonary artery (Fig. 1). The right coronary artery had two separated conus branch ostium, which gave rise to abnormal tortuous vessels that drained into the main pulmonary artery (Fig. 2). A treadmill exercise test according to the Bruce protocol was performed and revealed no significant ST segment change during exercise.

## DISCUSSION

Coronary arteriovenous fistulas have been well characterized (Gobel *et al.* 1970; Levine *et al.* 1978). Bilateral involvement in a coronary fistula, however, constitutes an uncommon subgroup of coronary artery fistulas. It has been found with a frequency of 0.002~0.013% in cardiac catheterization (Vanselow *et al.* 1996). In 36 reported cases of bilateral coronary fistula, it has been noted that drainage into the pulmonary artery is more common than is the case with single coronary artery fistula (Vanselow *et al.* 1996). This report describes the rare occurrence of bilateral coronary arteriovenous fistula coexistent with atrial septal defect and pulmonic stenosis. Approximately half of all patients with coronary fistulas develop symptoms of congestive heart failure, myocardial ischemia or myocardial infarction resulting from a coronary steal. However, the other half, similar to our patient, is totally asymptomatic. Tai *et al.* reported a patient with bilateral coronary arteriovenous fistula coexistent with apical hypertrophic cardiomyopathy(Tai *et al.* 1992). In their case, similar to our patient, no evidence suggesting significant myocardial ischemia was found. In contrast, Castelo *et al.* reported a case of bilateral coronary artery fistula into the main pulmonary artery which resulted in a severe angina pectoris attack (Castelo *et al.* 1994). In their case, ligation of the fistula was performed successfully and uneventfully. The treatment of a coronary arteriovenous fistula depends on its presentation and the magnitude of pulmonary-to-systemic flow. It is suggested, however, that surgical ligation, with or without the use of extracorporeal support, could be a form of treatment due to the natural history of the disease, which is not always benign, and low incidence of spontaneous fistula closure in patients with large coronary arteriovenous fistula. In conclusion, we report a case which shows a rare occurrence of bilateral coronary arteriovenous fistula coexistent with atrial septal defect and pulmonic stenosis.

## ACKNOWLEDGMENT

We wish to thank Seung-Nyun Kim, Myung-Ok Lee, Won-Taek Cho, Sung-Soo Shin, Myung-Ae Kye at the catheterization laboratory for the technical assistance.

## REFERENCES

Castelo V, Gonzalez-Juanatey JR, Amaro A, Iglesias C, Rubio J, Gil M: Myocardial ischemia secondary to a bilateral coronary fistula with drainage into the pulmonary artery trunk. *Rev Esp Cardiol 47: 497-499, 1994*

Gobel FL, Anderson CF, Baltaxe HA: Shunts between the coronary and pulmonary arteries with normal origin of the coronary arteries. *Am J Cardiol 25: 655-661, 1970*

Levine DC, Fellows KE, Abrams HL: Hemodynamically significant primary anomalies of the coronary arteries. Angiographic aspects. *Circulation 58: 25-34, 1978*

Rose AG: Multiple coronary arterioventricular fistulae. *Circulation 58: 178, 1978*

Sandhu JS, Uretsky BF, Zerbe TR: Coronary artery fistula in heart transplant patients: a potential complication of endomyocardial biopsy. *Circulation 78: II-253, 1988*

Tai YT, Fong PC, Chow WH: Bilateral coronary artery to pulmonary artery fistula coexistent with apical hypertrophic cardiomyopathy - a case report. *Angiology 43: 72-75, 1992*

Vanselow B, Fallen H, Zimmermann A, Henkel B, Seybold-Epting W: Bilateral coronary fistula: a case report and review. *Z Kardiol 85: 214-220, 1996*