

# A CASE OF CORONARY ARTERIOVENOUS FISTULA ASSOCIATED WITH GIANT CORONARY ARTERY ANEURYSM

SOO-HYUN KIM, MD, JUM-SUK KO, MD, HYUN-JU YOON, MD, SANG-CHUN LIM, NR,  
SOOK-HEE CHO, NR, HEY-SOOK KIM, NR, JI-SUN LEE, NR AND JONG CHUN PARK, MD

DEPARTMENT OF CARDIOLOGY, THE HEART CENTER, CHONNAM NATIONAL UNIVERSITY HOSPITAL, CARDIOVASCULAR  
RESEARCH INSTITUTE, CHONNAM NATIONAL UNIVERSITY MEDICAL SCHOOL, GWANGJU, KOREA

Coronary arteriovenous fistula is a more prevalent, hemodynamically significant congenital malformation. Both coronary arteries arise normally from their aortic sinuses, but the branches of fistula communicate directly with cardiac chamber, pulmonary trunk, coronary sinus, superior vena cava, or pulmonary vein. Fistula associated with coronary aneurysm is an uncommon finding. We report a rare case of 76-year-old female patient who had a coronary arteriovenous fistula with giant coronary artery aneurysm. This case is clearly diagnosed by echocardiography, three-dimensional computed tomography (3D-CT), and coronary angiography (CAG).

**KEY WORDS:** Coronary arteriovenous fistula · Coronary artery aneurysm.

## INTRODUCTION

Coronary arteriovenous fistula (CAVF) is an infrequent vascular anomaly that establishes a direct link between an epicardial coronary artery and a cardiac chamber, major vessels, or other vascular structures. We describe a case of 76-year-old woman who develop huge coronary artery aneurysm arising from Left main coronary artery and tortuous fistula form proximal right coronary artery drained to aneurysm.

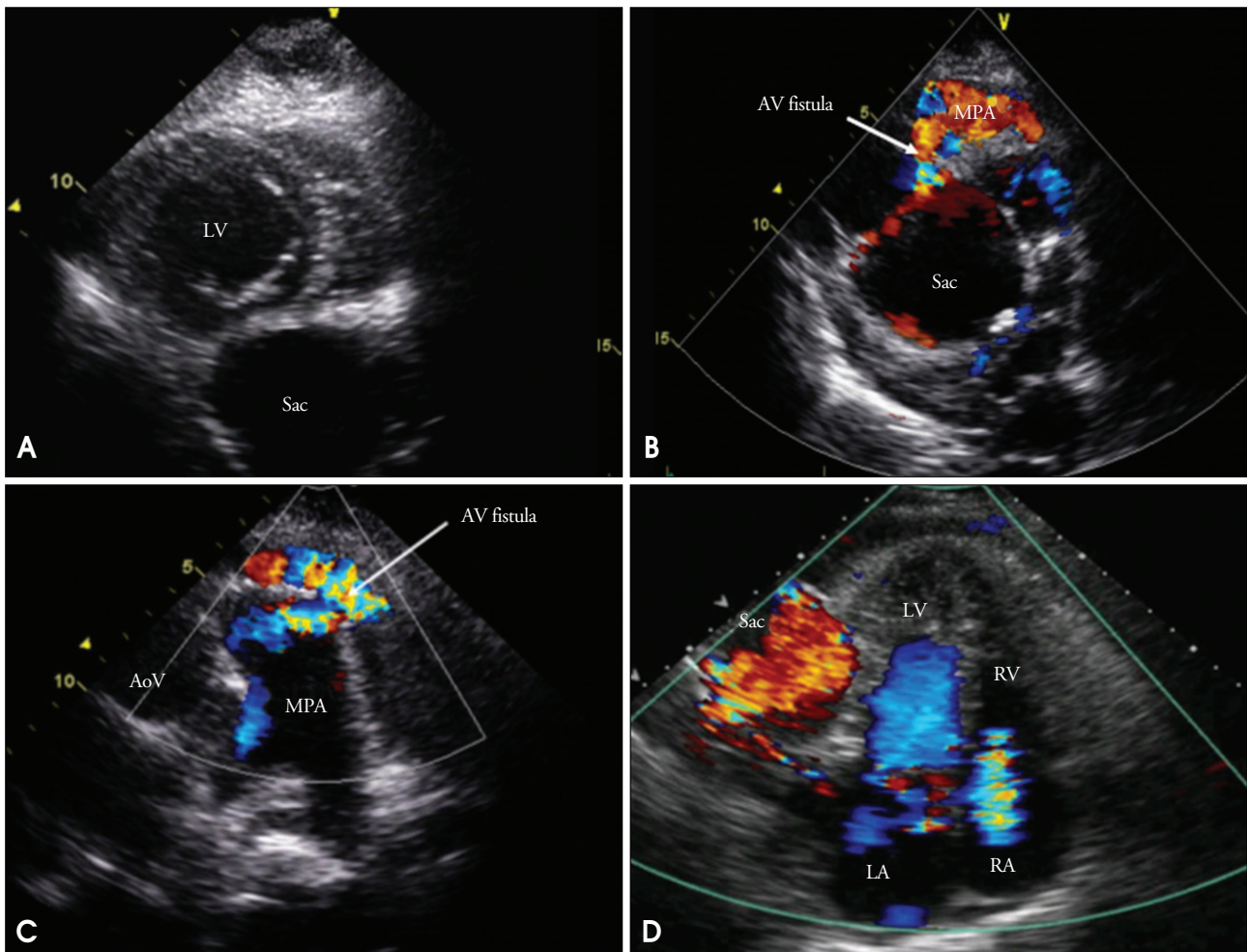
## CASE

A 76 year-old female patient presented with exertional dyspnea and effort angina for about 1month. She had a history of hypertension 2 years ago and medication of calcium channel blocker once daily. On physical examination, her blood pressure was 140/95 mmHg and chest auscultation revealed a continuous murmur graded IV/VI on her right second intercostal space along the right upper sternal border and crackles on both lower lung field. Electrocardiography revealed atrial fibrillation with rapid ventricular rhythm and left ventricular hypertrophy by voltage. Chest X-ray showed severe cardiomegaly and edematous change in both lower lung fields.

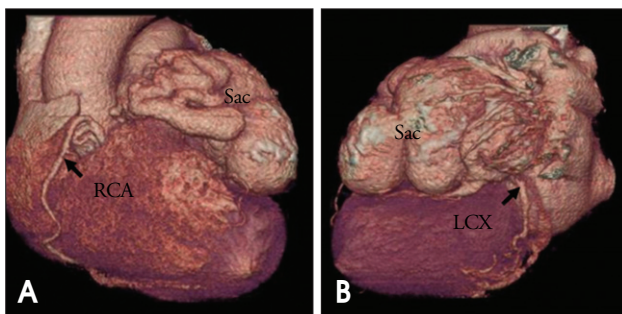
Echocardiography revealed huge saccular aneurysm in posterolateral side of left ventricle (Fig. 1A). Flow communication between aneurysmal sac and main pulmonary trunk was observed (Fig. 1B). Color Doppler with continuous wave revealed arteriovenous fistula with huge aneurysm which drained to main pulmonary artery (Fig. 1C). Left ventricular ejection fraction was 45% and global hypokinesia was noted. Left ventricle was compressed by multiple fusiform aneurysms (maximal diameter of largest one: about 6.7 cm) in apical 4 chamber view (Fig. 1D). Moderate pulmonary hypertension (systolic pulmonary arterial pressure=46.1 mmHg) and small amount pericardial effusion was noted. Cardiac computed tomography angiography (CTA) showed markedly enlarged tortuous anomalous vessel from left main coronary artery along course of left circumflex artery and several small tortuous anomalous vessels from proximal right coronary artery (Fig. 2). We performed coronary angiography for further evaluation of coronary artery anomaly. In coronary angiography, left main coronary artery was dilated and communicated to aneurysmal sac. Left anterior descending artery (LAD) was not visible. Instead, distal branch from left circumflex artery supplied left anterior

• Received: December 31, 2008 • Accepted: March 5, 2009

• Address for Correspondence: Jong Chun Park, Department of Cardiology, The Heart Center, Chonnam National University Hospital, Cardiovascular Research Institute, Chonnam National University Medical School, 671 Jebong-ro, Dong-gu, Gwangju 501-757, Korea Tel: +82-62-220-6244, Fax: +82-62-225-6260, E-mail: jcpark54@hanmail.net



**Fig. 1.** A: Echocardiogram in parasternal short axis view show extrinsic cardiac compression in left ventricle side due to multiple fusiform aneurysms (maximal diameter of largest one: about 6.7 cm). B: Modified apical 2 chamber view showed a fistula from aneurysmal sac to main pulmonary artery. C: Parasternal short axis window in aortic valve level revealed a arteriovenous fistula from coronary artery to main pulmonary trunk. D: Apical 4 chamber view showed mild regurgitation of tricuspid valve. Sac: aneurysmal sac, LV: left ventricle, AoV: aortic valve, MPA: main pulmonary artery, AV: arteriovenous, LA: left atrium, RV: right ventricle, RA: right atrium.



**Fig. 2.** A 64-detector row cardiac computed tomography with 3D reconstruction showed several small tortuous anomalous vessels from proximal right coronary artery (A) and enlarged tortuous anomalous vessel from left main coronary artery along course of left circumflex artery (B). Arrow indicates RCA and LCX. Sac: aneurysmal sac, RCA: right coronary artery, LCX: left circumflex artery.

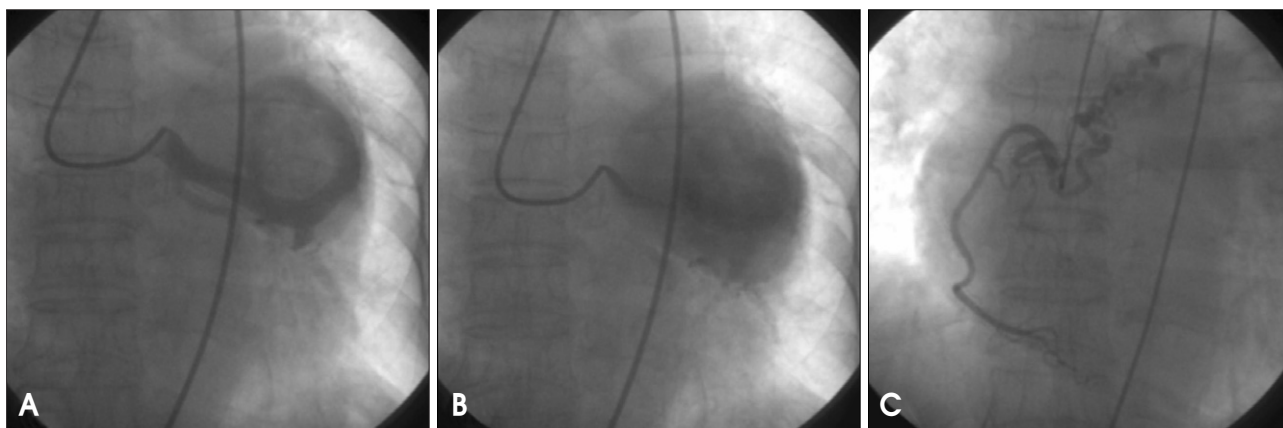
descending artery territory (Fig. 3). The pulmonary to systemic flow ratio was 1.7 by echocardiography. Reversible perfusion defect in left anterior descending artery and right

coronary artery territory was identified in myocardial single-photon emission computed tomography (M-SPECT) which represented myocardial ischemia result from coronary steal.

We recommended surgical treatment to patient. But patient refused surgery. After intensive medical treatment, patient showed improved clinical status and uneventful follow up.

## DISCUSSION

The incidence of the CAVF has been reported to be 0.1-0.2% in patients who undergo coronary artery angiography<sup>1)</sup> and 0.002% for general population.<sup>2)</sup> In about 50-55% of coronary arteriovenous fistulas, the right coronary artery provides the fistulous origin, and about 40% drain to the right ventricle.<sup>2)</sup> Generally, the symptoms develop depending on the amount of the shunt or the presence of coronary steal phenomenon of the fistulas, which tend to present in young adults with angina, exertional dyspnea, syncope, palpitations,



**Fig. 3.** Coronary angiographic finding revealed left main coronary was dilated and communicated to aneurysmal sac (A and B). And left anterior descending artery (LAD) was not visible. Small tortuous anonymous vessel from proximal right coronary artery was drained to aneurysmal sac through a fistula formation (C).

myocardial ischemia and infarction, and to manifest in adults >40 years old with congestive heart failure, atherosclerosis, and cardiac arrhythmias.<sup>1-2)</sup> Though some patients may have symptoms shortly after birth, however most of them are asymptomatic.<sup>1)</sup> Angina pectoris due to blood shunting and perfusion away from the myocardium (coronary steal phenomenon) occur. Steal phenomenon may be a cause of developing coronary artery aneurysmal change.<sup>1-2)</sup> Symptoms of congestive heart failure can result when drainage occurs into the right system resulting in congestion and pulmonary hypertension. The predominant physical finding is a continuous murmur, representing the systolic-diastolic flow in large fistulas. A detailed physical examination combined with an adequate image studies such as echocardiography, computed tomography may provide accurate diagnosis and avoid painful, invasive examinations.<sup>3-4)</sup> The decision to treat a coronary fistula is recommended in the presence of symptoms, a marker of functional significance in the absence of concomitant disease, fistula location, its size, and the resulting shunt volume.<sup>2)</sup> Anti-anginal therapy addressing the demand and supply mismatch induced by the coronary steal phenomenon may be effective for symptom relief.<sup>5-7)</sup> Of further clinical importance is recommended in endocarditis and antibiotic prophylaxis is recommended in CAVF patients. Surgical intervention is generally reserved for single, large, symptomatic fistula (with angina, cardiac

decompression or complications) and circulatory overload warrants surgical treatment even in asymptomatic patients.<sup>7)</sup> The main differences between our case and others are that giant aneurysm is detected in very elderly patient and in atypical location.

#### REFERENCES

1. Shiga Y, Tsuchiya Y, Yahiro E, Kodama S, Kotaki Y, Shimoji E, Fukuda N, Morito N, Urata M, Saito N, Niimura H, Mihara H, Yamanouchi Y, Urata H. *Left main coronary trunk connecting into right atrium with an aneurysmal coronary artery fistula.* *Int J Cardiol* 2008; 123:e28-30.
2. Maleszka A, Kleikamp G, Minami K, Peterschröder A, Körfer R. *Giant coronary arteriovenous fistula. A case report and review of the literature.* *Z Kardiol* 2005;94:38-43.
3. Androulakis A, Chrysohoou C, Barbetseas J, Brili S, Kakavas A, Maragiannis D, Kallikazaros I, Stefanadis C. *Arteriovenous connection between the aorta and the coronary sinus through a giant fistulous right coronary artery.* *Hellenic J Cardiol* 2008;49:48-51.
4. Rangasetty UC, Ahmad M. *Giant coronary artery fistula with aneurysm and multiple openings: A two-dimensional echocardiographic evaluation.* *Echocardiography* 2006;23:611-3.
5. Bobos D, Chatzis AC, Giannopoulos NM, Tsoutsinos A, Antoniadis A, Cokkinos D, Sarris GE. *Successful surgical repair of a giant arteriovenous fistula of the coronary arteries.* *J Card Surg* 2006;21:269-70.
6. Saxena P, Konstantinov IE, Burstow D, Tam R. *Surgical repair of a large coronary artery aneurysm with arteriovenous fistula.* *J Thorac Cardiovasc Surg* 2006;131:1167-8.
7. Olgunturk R, Kula S, Tunaoglu FS. *Transcatheter closure of a rare form of coronary arteriovenous fistula (circumflex artery to coronary sinus).* *Int J Cardiol* 2006;113:261-3.