Case Reports

Surgical Repair of a Giant Aorto–Left Ventricular Fistula

Charles Mve Mvondo, MD
Marta Pugliese, MD
Ellen M. Dailor, MD

Aortoventricular fistula, a rare congenital or acquired defect of the aortic wall, is characterized by an abnormal connection between the aorta and one of the ventricles. Symptom severity correlates with the diameter of the fistula and with the acute or chronic timing of presentation. The diagnosis is usually made by using echocardiography, and surgical treatment is necessary to avoid progression to heart failure. We describe the case of a 27-year-old woman who underwent successful surgical repair of an aortoventricular fistula that originated from the right coronary sinus and extended into the left ventricle through the interventricular septum. In addition to the patient's case, we briefly discuss this unusual condition ***(Tex Heart Inst J 2019;46(2):133-5)***

Key words: *Aortic diseases/diagnosis/etiology/surgery; aortic valve insufficiency/complications; heart defects, congenital/complications/diagnosis/surgery; heart ventricles/surgery; treatment outcome; vascular fistula/diagnosis/etiology*

From: *Division of Cardiac Surgery (Dr. Mve Mvondo), Cardiac Center of Shisong, Kumbo 8, Cameroon; Division of Cardiac Surgery (Dr. Pugliese), Tor Vergata University of Rome, 00133 Rome, Italy; and Department of Anesthesiology (Dr. Dailor), School of Medicine & Dentistry, University of Rochester, Rochester, New York 14642*

Address for reprints: *Marta Pugliese, MD, Division of Cardiac Surgery, Tor Vergata University of Rome, Viale Oxford 81, 00133 Rome, Italy*

E-mail: *marta_pugliese@hotmail.com*

Aortoventricular fistula (AVF), a defect of the aortic wall, is characterized by an abnormal connection between the aorta and the right ventricle or left ventricle (LV). Its estimated incidence is 0.14% to 0.96%.[1] The most prevalent cause of AVF is the rupture of a congenital or acquired sinus of Valsalva aneurysm. The aneurysm may be associated with congenital lesions such as a subarterial ventricular septal defect or coronary artery abnormalities. Rarer causes are aortic dissection, penetrating chest trauma, infective endocarditis, aortic valve (AV) surgery, and percutaneous endovascular procedures.[2-6] We report the case of a patient whose aorto–LV fistula we surgically treated, and we discuss the nature of AVFs and their treatment.

Case Report

In July 2014, a 27-year-old woman with atypical chest pain and exertional dyspnea (New York Heart Association class III) was referred to our institution. Her clinical history included recurrent episodes of fever 4 months earlier. A grade 3/4 diastolic murmur was audible at the left parasternal border. Transthoracic echocardiograms showed an AVF arising from the right coronary sinus and extending through the interventricular septum into the LV (Fig. 1). Mild-to-moderate AV regurgitation was also found. Selective coronary angiography and aortography revealed no evidence of coronary abnormalities. The patient tested positive for human immunodeficiency virus (HIV) with a normal CD4 count. Test results for infection, such as leukocyte count, sedimentation rate, and C-reactive protein level, were normal on admission. The patient underwent closure of the AVF with the use of an autologous pericardial patch and transaortic resection of the ventricular tissue of the fistula (Fig. 2). For technical reasons, complete evaluation with histologic analysis of the fistula was not possible.

A postoperative transesophageal echocardiogram showed no residual shunt and minimal AV regurgitation (Fig. 3). The patient was transferred to the intensive care unit, recovered uneventfully, and was discharged from the hospital on the 6th postoperative day. As of October 2018, she was asymptomatic.

Discussion

Approximately 94% of AVFs caused by sinus of Valsalva aneurysm rupture originate from the right coronary sinus and involve primarily the right ventricle.[7] However, AVF location may be less predictable when the defect is a complication of aortic dissection, infective endocarditis, or AV surgery. The main differential diagnosis is a congenital aortoventricular tunnel (AVT) connecting the ascending aorta and the ventricular cav-

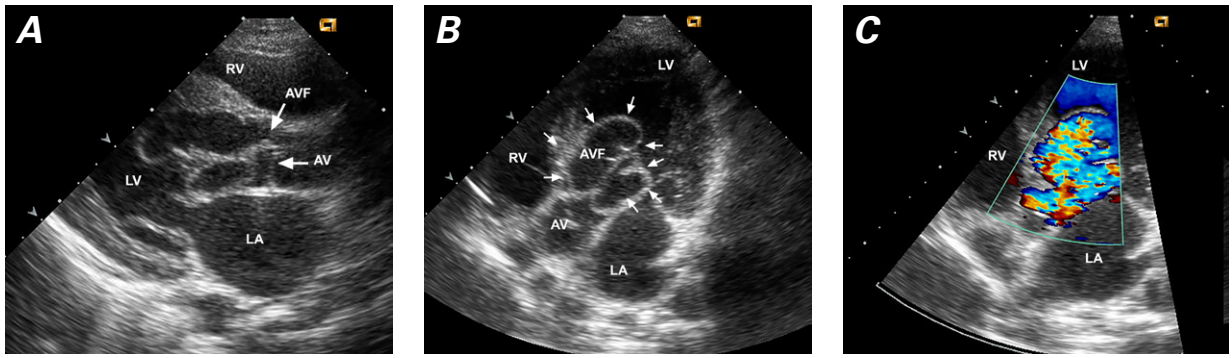

Fig. 1 *Preoperative transthoracic echocardiograms show the aortoventricular fistula (AVF) that extends from the right coronary sinus into the left ventricle (LV) through the basal septum in the* ***A****) left parasternal view (diastole),* ***B****) apical 5-chamber view (arrows delineate the AVF), and* ***C****) apical 5-chamber view in color-flow Doppler mode.*

AV = aortic valve; LA = left atrium; RV = right ventricle

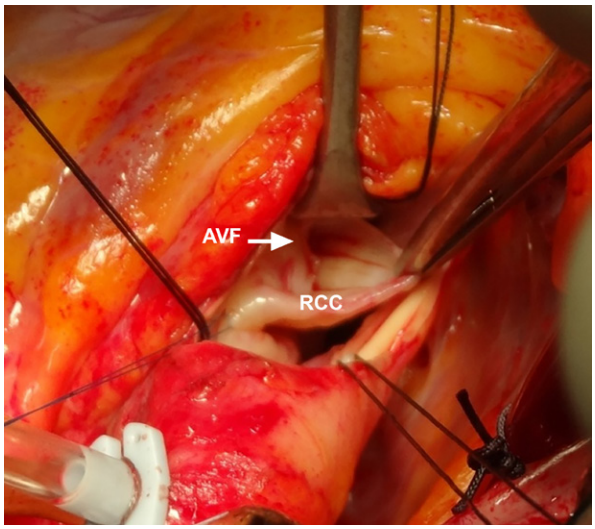

Fig. 2 *Intraoperative photograph shows the aortoventricular fistula (AVF).*

RCC = right coronary cusp

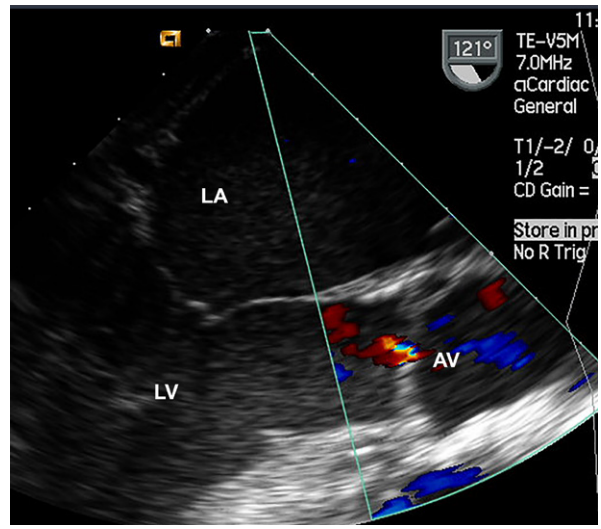

Fig. 3 *Postoperative transesophageal echocardiogram (color-flow Doppler mode) shows a minimal residual central aortic jet.*

AV = aortic valve; LA = left atrium; LV = left ventricle

ity through an extracardiac channel. The aortic orifice in an AVT is usually in the ascending aorta above the sinotubular junction, whereas in an AVF, it is usually in the sinus of Valsalva and predominantly communicates with the LV.[8] The involvement of the interventricular septum (Fig. 1A) in our patient was notable because similar echocardiographic findings can be found in aortic root or myocardial abscess. Although we could not exclude an infectious component because of the patient's HIV and recurrent fever 4 months earlier, we found no evidence intraoperatively of previous or active infection. Echocardiograms clearly revealed the origin of the lesion and its saccular extension into the basal septum (Fig. 1A). However, because the patient had reported atypical chest pain, preoperative cardiac angiography was necessary to rule out coronary abnormalities before repair.

When left untreated, AVF leads to heart failure over a variable time; thus, prompt repair is indicated when the diagnosis is made. Despite the availability of percutaneous techniques, especially for symptomatic and high-risk patients,[4] surgical repair is the primary treatment. The timing of surgery is determined by the patient's clinical manifestations, the origin of the lesion, and the presence of associated disease. Surgical techniques for AVF and for AVT are similar, varying from direct suturing of the defect with or without a patch, to more complex procedures such as aortic root or AV replacement.[9]

In our patient, we attained good exposure of the ventricular side of the fistula by retracting the AV cusps, thus facilitating transaortic resection of the ventricular tissue. Subsequent defect closure was exclusively on the aortic side with use of an autologous pericardial patch.

We were concerned that additional patch reinforcement from the ventricular side might compromise the morphology of the corresponding right coronary sinus and cusp and the function of the AV (Fig. 3).

In conclusion, AVF is a rare and insidious lesion with unpredictable anatomic and clinical patterns. Prompt intervention is crucial to avoid progression to congestive heart failure and AV dysfunction.

References

1. Desai S, Flores E, Loli A, Maki P. Sinus of Valsalva fistula to the right ventricle along with coronary artery fistula to the pulmonary artery in a young Native American female. Case Rep Cardiol 2013;2013:674608.
2. Park H, Park TH, Lee DY, Ahn J, Baek HK, Kim MH, et al. A case of aortic dissection with fistula from aorta to right ventricle. Korean Circ J 2012;42(9):629-31.
3. Anguera I, Quaglio G, Miro JM, Pare C, Azqueta M, Marco F, et al. Aortocardiac fistulas complicating infective endocarditis. Am J Cardiol 2001;87(5):652-4, A10.
4. Eng MH, Garcia JA, Hansgen A, Chan KC, Carroll JD. Percutaneous closure of a para-prosthetic aorto-right ventricular fistula. Int J Cardiol 2007;118(2):e31-4.
5. Munoz-Garcia AJ, Rodriguez-Bailon I, Briales JH, Navarro MJ, Garcia JM, de Teresa-Galvan E. Aorto-right ventricular fistula after percutaneous aortic valve implantation of a CoreValve prosthesis. Tex Heart Inst J 2011;38(6):728-9.
6. Samuels LE, Kaufman MS, Rodriguez-Vega J, Morris RJ, Brockman SK. Diagnosis and management of traumatic aorto-right ventricular fistulas. Ann Thorac Surg 1998;65(1):288-92.
7. Ibrahim KS, Waqfi NR, Jarrah MI. Sinus of Valsalva aneurysm with fistula to the right atrium presented as acute heart failure in a young man. Am J Case Rep 2013;14:398-400.
8. McKay R. Aorto-ventricular tunnel. Orphanet J Rare Dis 2007;2:41.
9. Martins JD, Sherwood MC, Mayer JE Jr, Keane JF. Aortico-left ventricular tunnel: 35-year experience. J Am Coll Cardiol 2004;44(2):446-50.