

## Case Report

## Echocardiographic Demonstration of Coronary Artery to Left Ventricle Fistulas: Case Report and Review of the Literature

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We describe the case of a 45-year-old female patient with coronary fistulas arising from both the left and right coronary artery system and emptying in the left ventricle. Only sporadically do coronary artery fistulas drain into the left ventricle. In our patient, the most likely explanation of the fistulous communications was a congenital cause. We review the literature on coronary cameral fistulas and discuss the etiology of the diagnostic findings. Small coronary artery fistulas are generally well-tolerated and should impose no significant restriction on daily routine and activities. Nevertheless, small fistulas may under certain conditions produce a "steal" phenomenon and shunt blood flow away from the myocardial capillary network, causing ischemia.

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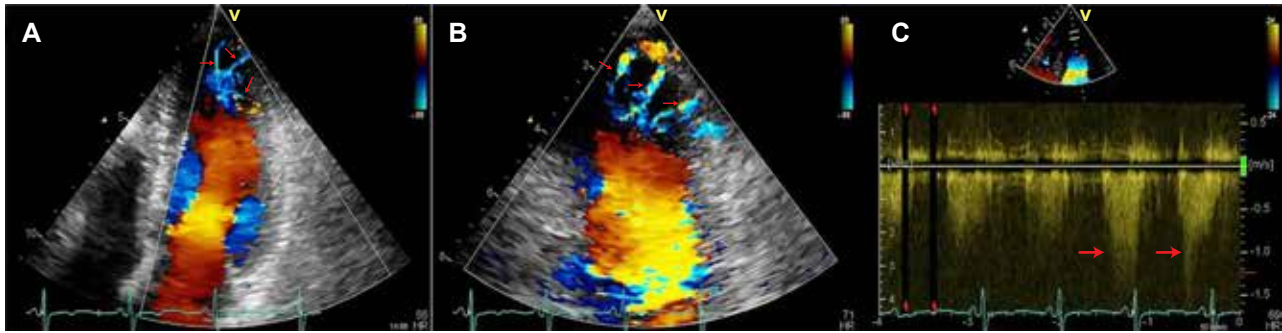
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**C**oronary artery fistulas are rare coronary anomalies. In early fetal development, the persistence of myocardial sinusoids and the subsequent connection with the endothelial buds that originated from the base of the *truncus arteriosus* form the basis for abnormal coronary artery fistulas. However, in certain instances they might be acquired, usually secondary to either trauma or invasive cardiac procedures.<sup>1-3</sup> We describe the case of a female patient with coronary fistulas arising from both the left and right coronary artery system and emptying in the left ventricle, as demonstrated by her echocardiography study.

### Case presentation

A 45-year-old female was hospitalized complaining of chest pain and palpitations. For at least 2 months, she had experienced exertional chest pain that resolved upon rest. She had none of the classical risk factors for coronary artery disease

and was not taking any prescribed medications. The ECG showed normal sinus rhythm with negative T waves in leads II, III, aVF, and all precordial leads. Cardiac enzymes and routine laboratory index findings were normal. A complete standard two-dimensional echocardiography and Doppler interrogation was performed. Transthoracic echocardiography revealed normal left and right ventricular size and function, without significant valvular disease. Color Doppler showed multiple turbulent jet areas, probably originating from the distal left anterior descending artery and draining into the left ventricle (Figure 1). Contrast-enhanced echocardiography more accurately depicted the presence of a fistulous network tract. Our patient underwent coronary angiography, which showed multiple coronary fistulas arising from both the left and right coronary artery system and emptying in the left ventricle. Thus, shunting of blood between the epicardial coronary arteries and the left ventricular chamber was confirmed.



**Figure 1.** Four-chamber (A) and 2-chamber (B) view with color Doppler, showing multiple fistulas between the left anterior descending artery and the left ventricle (arrows). Figure C illustrates the CW-Doppler recording of a coronary fistula flow that is predominantly diastolic (arrows).

Atherosclerotic lesions were not visible. The contrast medium streamed into the left ventricle via a maze of fine vessels from both left and right coronary arteries. Left ventriculography showed normal global and regional left ventricular function. The left-to-left shunting of blood was not hemodynamically significant. In order to exclude the coronary artery steal phenomenon a treadmill exercise test was performed, as well as SPECT perfusion scanning. The exercise test was interrupted during the 5th minute of the Bruce protocol owing to chest pain and pseudo-normalization of the resting T-wave inversion, seen both at peak exercise and in recovery. Thallium-201 myocardial perfusion imaging revealed an apical region with reversible ischemia. During her hospitalization no arrhythmia was detected by routine ECG examination, while 24-hour Holter monitoring showed brief episodes of sinus tachycardia. The patient's symptoms improved after titration of metoprolol.

## Discussion

Coronary artery fistula is an uncommon clinical entity. Its incidence in selected series ranges from 0.26% to 0.40% of congenital cardiac anomalies.<sup>1-3</sup> The etiology may be congenital or traumatic (such as stab or projectile injuries, or following coronary angioplasty), but more typically it arises as a congenital anomaly.<sup>4</sup> It may occur as an isolated abnormality in an otherwise structurally normal heart or in association with congenital outflow obstruction. Multiple fistulous communications to cardiac chambers are a rare anomaly. The pathogenic origin of the malformation is obscure. Morphological studies suggest a partial persistence of embryonic myocardial sinusoids that arise from endothelial protrusions into intertrabecu-

lar spaces.<sup>5</sup> Fetal regression of these structures results in the formation of the Thebesian vessels of the adult heart. Thus, interference with developmental changes might produce an abnormally prominent Thebesian system with the appearance of multiple coronary microfistulae. Coronary artery fistulas may mimic the physiology of various heart lesions.<sup>4-6</sup> Fistulas that drain to the systemic veins or right atrium have a physiology similar to an atrial septal defect; those that drain to the pulmonary arteries have physiology similar to a patent *ductus arteriosus*; those that drain to the left atrium do not cause a left-to-right shunt, but do cause a volume load similar to mitral regurgitation; and those that drain to the left ventricle have a physiology similar to that of aortic insufficiency.

The right coronary artery is the most likely site of origin, in 55% of cases, while the left coronary artery system is involved in 35%. The major receiving chamber is the right ventricle (45%), followed by the right atrium (25%), pulmonary artery (15-20%), and less commonly the coronary sinus (7%).<sup>7</sup> In all reports, coronary cameral fistulas are least often found to be draining into the left atrium or left ventricle.<sup>8</sup>

Many adults are asymptomatic if the fistulas are small, because they do not usually cause any hemodynamic compromise. However, other patients present with symptoms of fatigue, dyspnea, angina, atrial arrhythmia, signs of congestive heart failure, pulmonary hypertension, or infective endocarditis. Larger fistulas are compatible with the above symptoms.<sup>9</sup> They can cause the coronary artery steal phenomenon, which leads to ischemia of the segment of the myocardium perfused by the coronary artery. Over time, the coronary artery leading to the fistulous tract progressively dilates, which, in turn, may progress to frank aneurysm formation, intimal ulceration, medial degeneration, in-

timal rupture, atherosclerotic deposition, calcification, side-branch obstruction, mural thrombosis, and, rarely, rupture.<sup>10</sup> Occasionally, heart failure may develop due to volume overload. Small fistulas remain clinically silent and are recognized at routine echocardiography and autopsy. In the small fistulas, the myocardial blood supply is not compromised enough to cause symptoms. Spontaneous closure usually occurs; however, some can dilate over time.<sup>11</sup> Traditionally, coronary artery fistulas have been diagnosed by invasive investigations such as coronary angiography. Transthoracic echocardiography, supplemented by Doppler and color-flow imaging, is a noninvasive, portable, easily available and fairly accurate technique. Our patient's color Doppler investigation showed multiple turbulent jet areas originating from the left anterior descending artery and draining into the left ventricle. The size and function of the left and right ventricle were normal and there was no valvular disease.

Though larger fistulas may require treatment, with transcatheter embolization or surgical repair,<sup>10</sup> our patient's blood shunting was hemodynamically insignificant. However, considering the clinical presentation, we postulated that the fistulas, although small, might in certain conditions produce a "steal" phenomenon that contributed to the patient's apical wall ischemia and chest pain. Blood follows the path of lower-resistance through a fistula, rather than traversing the smaller arterioles and capillaries of the myocardium; thus, blood is shunted away from the normal coronary pathway. Elective coil occlusion is recommended in patients who fulfill the following criteria: absence of multiple fistulas, a single narrow drainage site, absence of large branch vessels, and safe accessibility to the coronary artery supplying the fistulas.<sup>12-13</sup> Our patient did not meet those criteria, since multiple fine fistulas with multiple drainage sites were revealed by both the echocardiographic and angiographic examinations. Thus, conservative treatment was preferred. She was treated with metoprolol ad-

ministration, acetylsalicylic acid and low dose perindopril. Gradual titration of the metoprolol dose improved our patient's symptoms.

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