



# A Case of Coronary Cameral Fistula: When and How to Intervene?

Mehdi Peighambari, MD, Marziyeh Pakbaz, MD, Azin Alizadehasl, MD\*,  
Saeid Hosseini, MD, Hamidreza Pouraliakbar, MD

*Rajaie Cardiovascular, Medical and Research Center, Iran University of Medical Sciences, Tehran, Iran.*

Received 09 October 2019; Accepted 13 July 2020

## Abstract

Coronary artery fistulas constitute a rare anomaly defined as an abnormal communication between a coronary artery and a great vessel or any cardiac chamber. The majority of these fistulas arise from the right coronary artery and the left anterior descending coronary artery; the circumflex coronary artery is rarely involved. We present an unusual case of a coronary artery fistula in a middle-aged woman who presented with symptoms of heart failure and abnormal auscultation. Echocardiography and conventional and computed tomography angiography showed that the coronary fistula originated from the left circumflex coronary artery and drained majorly into the right ventricle. Given the complex anatomy of the fistula, we managed it surgically rather than percutaneously. There were no complications early after surgery and at 1 year's follow-up.

*J Teh Univ Heart Ctr 2020;15(4):189-194*

**This paper should be cited as:** Peighambari M, Pakbaz M, Alizadehasl A, Hosseini S, Pouraliakbar H. A Case of Coronary Cameral Fistula: When and How to Intervene? *J Teh Univ Heart Ctr 2020;15(4):189-194.*

**Keywords:** Heart Defects, Congenital; Fistula; Computed tomography angiography

## Introduction

Coronary artery fistulas (CAFs) constitute a rare congenital or acquired anomaly of coronary arteries defined as an abnormal communication between a coronary artery and a great vessel (the vena cava, the pulmonary vein, or the pulmonary artery) or any cardiac chamber.<sup>1</sup> The majority of these fistulas arise from the right coronary artery and the left anterior descending coronary artery; the circumflex coronary artery is rarely involved.<sup>2</sup> The termination site of a fistula in order of frequency may be the right ventricle, the right atrium, the pulmonary artery, the coronary sinus, the left atrium, the left ventricle, and the superior vena cava.<sup>3</sup> CAFs are found in 0.05% to 0.25% of patients undergoing coronary angiography.<sup>4</sup> Generally, the shunt produced by the fistula

is small; and in most cases, the patient is asymptomatic. In such cases, the fistula is detected incidentally by coronary angiography or echocardiography for other reasons. In uncommon cases of larger fistulas, symptoms and complications such as dyspnea, congestive heart failure, angina, endocarditis, or arrhythmias may be encountered.<sup>2</sup> We present an unusual case of a CAF in a middle-aged woman originating from the left circumflex coronary artery and draining majorly into the right ventricle and also faintly into the left atrium.

## Case Report

A 52-year-old woman with diabetes was referred to

\*Corresponding Author: Azin Alizadehasl, Associate Professor of Echocardiography, Department of Echocardiography, Rajaie Cardiovascular, Medical and Research Center, Vali-e-Asr Street, Adjacent to Mellat Park, Tehran, Iran. 1996911151. Tel: +98 21 23922190. Fax: +98 21 22042026. E-mail: alizadeasl@gmail.com.



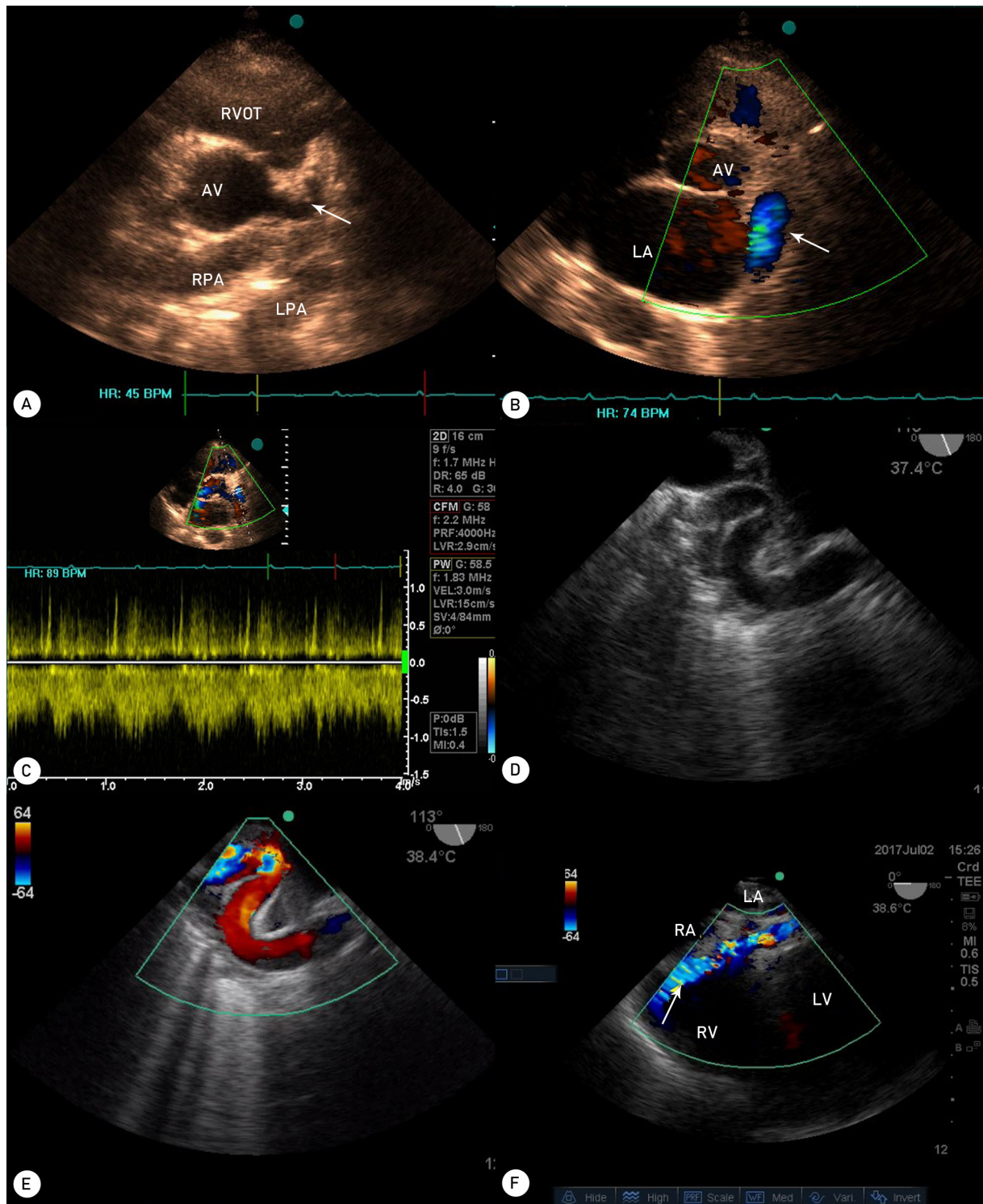


Figure 1. A) Transthoracic echocardiography (short-axis view at the AV level) shows an aneurysmal dilation in the left main coronary artery (arrow). B) Color Doppler interrogation (short-axis view at the AV level) shows an abnormal continuous flow (arrow) that starts from the dilated left main coronary artery and runs between the aorta and the pulmonary artery toward the posterior aspect of the LA. C) Pulsed Doppler study shows a continuous low-velocity flow, predominantly in diastole, which is consistent with a coronary artery fistula. D and E) Two-dimensional and color Doppler transesophageal echocardiography shows a severely dilated and tortuous left circumflex artery passing through the left atrioventricular groove toward the posterior aspect of the heart. F) An angulated view of transesophageal echocardiography (0° at the mid-esophageal level) shows the fistulous communication between the left circumflex artery and the RV inflow (arrow).

RVOT, Right ventricular outflow tract; AV, Aortic valve; RPA, Right pulmonary artery; LPA, Left pulmonary artery; LA, Left atrium; RA, Right atrium; LV, Left ventricle; RV, Right ventricle



our center for the repair of a large CAF diagnosed by echocardiography and coronary angiography in another center. The patient had complaints of exertional chest pains and dyspnea with a functional class of II-III of 8 months' duration. Physical examination showed stable vital signs and a continuous murmur with IV/VI intensity. Electrocardiography showed normal sinus rhythm and nonspecific ST-T changes.

Transthoracic echocardiography illustrated severe left ventricular enlargement with moderate systolic dysfunction and global hypokinesia (ejection fraction=35%), mild right ventricular enlargement with mild systolic dysfunction, up-to-moderate mitral regurgitation, mild-to-moderate tricuspid regurgitation, and a normal pulmonary artery pressure. In the short-axis view at the aortic valve level, an aneurysmal dilation in the left main coronary artery and a dilation in the pulmonary artery and its branches were observed (Figure 1A & Video 1). Color Doppler interrogation showed an abnormal continuous flow that started from the dilated left main coronary artery and ran between the aorta and the pulmonary artery in the left atrioventricular groove toward the posterior aspect of the left atrium. The finding was compatible with the pathway of the left circumflex coronary artery (Figure 1B, Video 2). Pulsed-wave Doppler study showed a low-velocity continuous flow, which was present predominantly in diastole but extended to systole, consistent with a CAF (Figure 1C). Transesophageal echocardiography, performed for a more accurate evaluation of the structure and the course of the fistula, revealed a dilated left circumflex coronary artery (size=1.5 cm) exhibiting a very tortuous course in the atrioventricular groove (Figure 1D & Figure 1E; Video 3 and Video 4) with its fistulous tract ultimately terminating in the right ventricular inflow tract (Figure 1F & Video 5). An additional finding was a moderate-sized patent foramen ovale with a left-to-right shunt.

The findings of cardiac catheterization, previously performed on the patient in another center, were reviewed. The left anterior descending coronary artery and the right coronary artery had a normal appearance with no significant stenosis. The aneurysmal left main coronary artery and the left circumflex coronary artery with its tortuous course were readily observed, as was the fistulous communication between the left circumflex coronary artery and the right ventricle (Figure 2 & Video 6).

In order to better appreciate the surface anatomy of the CAF and accurately delineate its destination site, we performed a multislice (384-slice) computed tomography (CT) angiography of the coronary arteries. Via maximum intensity projection and the volume-rendering technique, a giant left circumflex coronary artery fistula was depicted and its drainage into the base of the right ventricle was confirmed (Figure 3).

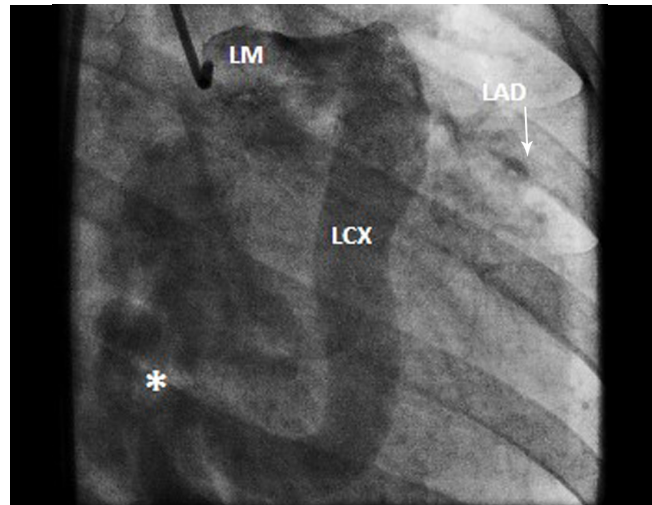
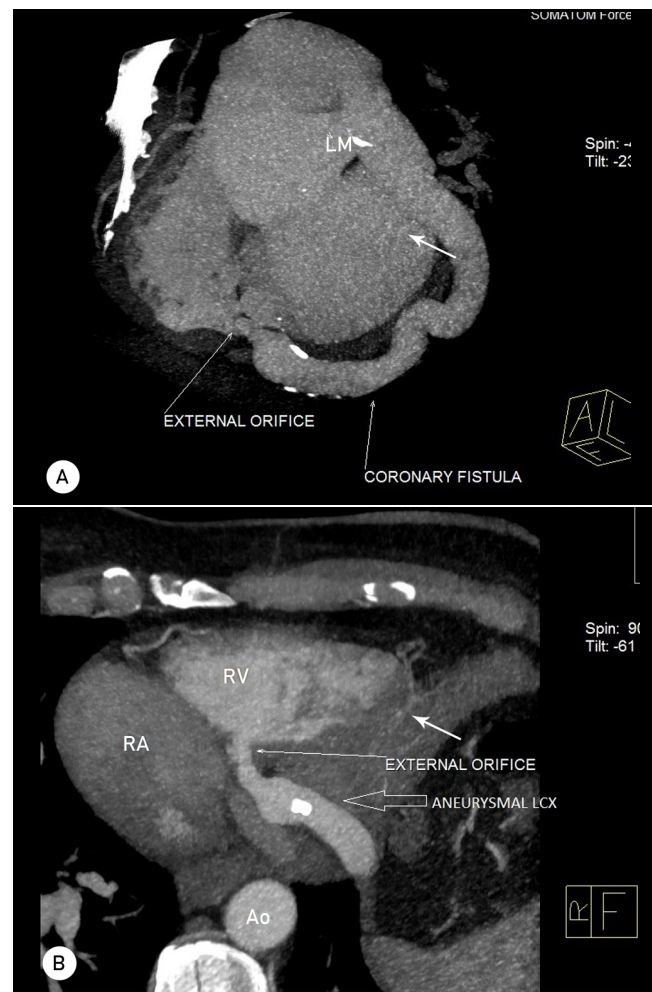


Figure 2. Right anterior oblique view with the caudal angulation view of diagnostic coronary catheterization shows a severely dilated LM, which bifurcates to a small LAD and an aneurysmal LCX. The asterisk shown is the site of the fistulous communication between the LCX and the right ventricle. LM, Left main coronary artery; LAD, Left anterior descending coronary artery; LCX, Left circumflex artery



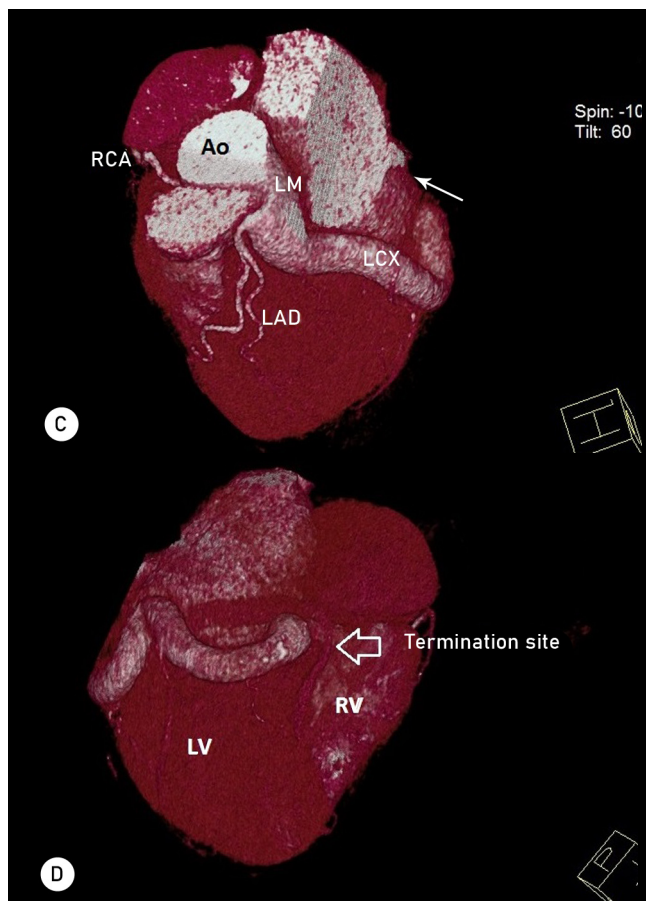


Figure 3. Coronary computed tomography angiography of the coronary arteries using maximum intensity projection (MIP) images (A and B) and volume-rendering technique (VRT) images (C and D). The origin and the termination site of the coronary fistula are clearly depicted in the MIP images. The surface anatomy, course, and shape of the giant tortuous coronary fistula, as well as its anatomic relationships with the RCA and the LAD, are readily appreciated in the VRT images, showing the anterior (C) and posterior (D) aspects of the heart.

RCA, Right coronary artery; LAD, Left anterior descending artery; LM, Left main artery; LCX, Left circumflex artery; Ao, Aorta; LV, Left ventricle; RV, Right ventricle; RA, Right atrium

Our patient was symptomatic due to the presence of a large CAF resulting in left ventricular dysfunction and heart failure symptoms and also probably ischemic symptoms; intervention was, therefore, mandated. The aneurysmal dilation and extreme tortuosity of her left circumflex coronary artery and a distally located fistulous lesion rendered the patient an anatomically unsuitable case for percutaneous closure, leaving surgical repair as the viable option. Via median sternotomy, cardiopulmonary bypass utilization, and hyperkalemic cardioplegic arrest, surgery was done. There was a very large left circumflex coronary artery adjacent and parallel to the coronary sinus (Figure 4). Through the right atrium, the opening of the fistula to the right ventricle was just under the posterior leaflet of the tricuspid valve.

It was closed with a 0.6-mm PTFE patch. Thereafter, via a longitudinal incision of the fistulous artery (Video 7) and a partial excision of its wall, arterioplasty was performed to create a near-normal diameter for the left the circumflex coronary artery. There were 2 large obtuse marginal branches of the circumflex artery, which were bypassed with an autologous saphenous vein. After the removal of the aortic cross-clamp, the patient developed ventricular fibrillation, along with bluish discoloration of the left ventricle. The left anterior artery was opened, and it was bypassed as well owing to the presence of scant dark blood. The patient was subsequently weaned from coronary artery bypass.

She experienced an uneventful postoperative period, during which we performed a second multislice CT angiography of her coronary arteries (Figure 5). The patient's symptoms were resolved soon after the surgery, and she was in good condition at 1 year's follow-up.

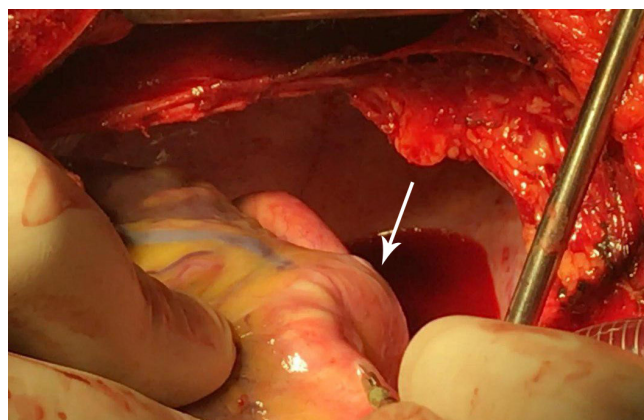


Figure 4. Surgical view of the large aneurysmal circumflex artery (arrow)

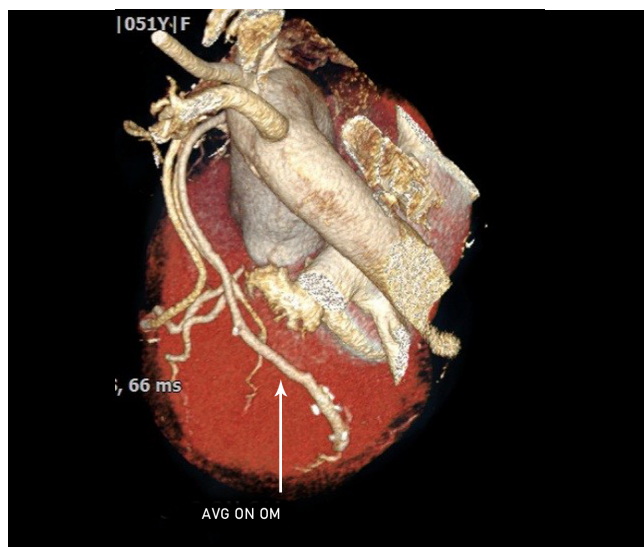


Figure 5. Reconstructed computed tomography angiography (volume-rendering technique images) after the surgical repair of the coronary fistula and coronary artery bypass.

SVG, Saphenous vein graft; OM, Obtuse marginalis artery



## Discussion

Coronary artery anomalies can be classified as the abnormalities of the origin, termination, and course of coronary arteries.<sup>5</sup> CAFs are known as the anomaly of the termination of coronary arteries. In such anomalies, the coronary artery normally originates from the aortic root but has abnormal termination to a great vessel or a cardiac chamber.

CAFs are uncommon anomalies with a reported prevalence rate of 0.05% to 0.25% of all patients undergoing coronary angiography<sup>4</sup> and are even rarer in the general population with an estimated prevalence rate of 0.002%.<sup>3</sup> CAFs are congenital or acquired. Congenital CAFs represent 0.4% of all cardiac malformations and 48.7% of all congenital coronary anomalies.<sup>6</sup> Acquired CAFs are rarer than congenital CAFs and are usually iatrogenic as a complication of cardiac catheterization, percutaneous coronary angioplasty, electrophysiological ablation, pacemaker implantation, endomyocardial biopsy, cardiac transplantation, or coronary artery bypass surgery.<sup>6-8</sup> Acquired CAFs may also be associated with some disease states such as acute myocardial infarction, hypertrophic cardiomyopathy, dilated cardiomyopathy, and cardiac tumors.<sup>6</sup> The majority of CAFs arise from the right coronary artery and the left anterior descending coronary artery; the circumflex coronary artery is rarely involved.<sup>2</sup> Thus, our patient, who had a fistulous left circumflex coronary artery, can be deemed a rare CAF case.

The clinical presentation of CAFs is influenced by many factors such as the age of the patient, the site of the fistulous communication, the advent of ischemia, the size of the shunting, and the resistance of the termination site structure.<sup>9</sup> Most adult patients are asymptomatic given the small size of CAFs and the fact that they are incidentally discovered in physical examinations following the auscultation of a continuous cardiac murmur or during cardiac catheterization or CT angiography for an unrelated reason. In uncommon cases of larger fistulas, symptoms and complications such as dyspnea (60%), congestive heart failure (19%), angina (7%), endocarditis (20%), or arrhythmias may be encountered.<sup>2,6</sup> The pathophysiology of ischemia and related symptoms is a reduction in the blood supply distal to the site of the fistulous communication, which is termed "the coronary steal phenomenon".<sup>8</sup> Blood tends to flow through the lower-resistance trajectory of the fistula rather than the higher-resistance intramyocardial vasculature. The symptoms of heart failure are a consequence of pulmonary congestion and hypertension when a large fistulous communication shunts the blood to the left or right system, producing volume overload.

The first step for the diagnosis of CAFs after a thorough history taking and physical examination is usually echocardiography. Useful findings that may be revealed by this imaging modality include chamber enlargement, a

dilated and tortuous feeding coronary artery, a high-volume flow depicted by color Doppler imaging, and the drainage site of the CAF.<sup>8</sup> Depending on the drainage site in relation to the tricuspid valve, the shunt may be pre-tricuspid or post-tricuspid. A pre-tricuspid shunt (eg, a coronary fistula draining into the right atrium or the coronary sinus) could result in right-chamber enlargement, whereas a post-tricuspid shunt (eg, our patient, whose coronary fistula drained into the right ventricle) could lead to left-chamber enlargement.<sup>10</sup>

The most common imaging modality for diagnosing CAFs still remains the standard coronary angiography.<sup>7</sup> Coronary angiography accurately visualizes the proximal part of the CAF and allows an assessment of the size and also the number of fistulas. However, CAFs drain into the low-pressure chambers of the heart. Due to the significant dilution of the injected contrast agent, these drainage sites may not be well-visualized by conventional angiography.<sup>3</sup> Multidetector CT angiography, by using electrocardiographically gated reconstruction techniques, confers high-resolution imaging of the coronary anatomy and, thus, brings forth an accurate detection of coronary anomalies.<sup>3</sup>

The clinical course of CAFs is highly variable. Spontaneous closure has been reported; it usually appears in small-to-medium-sized fistulas.<sup>8,11</sup> According to the latest guideline of the American College of Cardiology/American Heart Association for Management of Adults With Congenital Heart Disease, small-to-medium-sized CAFs that produce no symptom or complication could be managed medically and followed by echocardiography every 3 to 5 years.<sup>12</sup> Percutaneous or surgical closure is a Class I recommendation for large CAFs regardless of symptoms and for small-to-medium-sized CAFs in the presence of myocardial ischemia, arrhythmia, ventricular dysfunction, and enlargement or endarteritis.<sup>12</sup> Deciding which type of intervention should be selected is still controversial. Nonetheless, percutaneous closure may be considered when there is no other reason for surgery, when the CAF has suitable anatomy (eg, a non-tortuous course), when the distal portion of the fistula is accessible to the closure device, and when the CAF has a narrow distal end that prevents embolization to the drainage site.<sup>6,7</sup> As was mentioned previously, our case was not suitable for percutaneous closure and was, thus, surgically managed.

For surgical purposes, CAFs are classified. In type A or proximal type CAFs, the coronary segment proximal to the origin of the fistula is dilated, but the distal part appears normal. This type of CAF can be treated via the epicardial ligation of the CAF. In type B or distal type CAF, the coronary artery is dilated all over its entire length and terminates as a fistula in the right heart. This type of CAF requires ligation with purse-string sutures at the site of termination using cardiopulmonary bypass.<sup>13,14</sup> Patients undergoing the transcatheter or surgical closure of CAFs experience an overall good prognosis with normal life expectancy.<sup>13</sup>

## Conclusion

CAFs constitute a rare cardiac anomaly with variable anatomies and clinical presentations. Large CAFs, regardless of symptoms, and also smaller symptomatic ones should be managed via transcatheter or surgical closure. We herein reported a very rare case of a huge CAF that originated from the left circumflex coronary artery to the right ventricle.

## To watch the following videos, please refer to the relevant URLs:

<https://jthc.tums.ac.ir/index.php/jthc/article/view/970/931>

Video 1. Transthoracic echocardiography (short-axis view at the aortic valve level) shows an aneurysmal dilation in the left main coronary artery.

<https://jthc.tums.ac.ir/index.php/jthc/article/view/970/932>

Video 2. Transthoracic echocardiography with color Doppler interrogation (short-axis view at the aortic valve level) shows an abnormal continuous flow that starts from the dilated left main and runs between the aorta and the pulmonary artery toward the posterior aspect of the left atrium.

<https://jthc.tums.ac.ir/index.php/jthc/article/view/970/933>

Video 3. Two-dimensional transesophageal echocardiography shows a severely dilated and tortuous left circumflex artery, passing through the left atrioventricular groove toward the posterior aspect of the heart.

<https://jthc.tums.ac.ir/index.php/jthc/article/view/970/934>

Video 4. Color Doppler transesophageal echocardiography shows a severely dilated and tortuous left circumflex artery, passing through the left atrioventricular groove toward the posterior aspect of the heart.

<https://jthc.tums.ac.ir/index.php/jthc/article/view/970/935>

Video 5. An angulated view of transesophageal echocardiography (0° at the mid-esophageal level) shows a fistulous communication between the left circumflex artery and the right ventricular inflow.

<https://jthc.tums.ac.ir/index.php/jthc/article/view/970/936>

Video 6. Right anterior oblique view with the caudal angulation view of diagnostic coronary catheterization shows a severely dilated left main coronary artery, which bifurcates to a small left anterior descending artery and an aneurysmal left circumflex artery. The arrow shows the exit site of the fistulous left circumflex artery to the right ventricle.

<https://jthc.tums.ac.ir/index.php/jthc/article/view/970/937>

Video 7. Surgical view of a large aneurysmal circumflex artery shows a longitudinal incision made in the fistulous artery.

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