

Giant Coronary Fistula Between Circumflex Artery and Coronary Sinus: Initial Diagnosis by Echocardiography

Fístula Coronária Gigante da Artéria Circunflexa para o Seio Coronariano: Diagnóstico Inicial por Ecocardiografia

Israel Nilton de Almeida Feitosa¹, Maria Rafaela Viana de Sá², Jonas Silva Andrade², Ana Raquel Avelino Diniz Gonzaga²

¹Department of Echocardiography, Federal University of Campina Grande, Campina Grande, PB; ²School of Medical Sciences of Campina Grande, Campina Grande, PB, Brazil.

Introduction

Coronary fistula is defined as communication between the termination of a coronary artery (or its branches) and a cardiac chamber, a large vessel, or other vascular structure.¹ The term coronary-cameral fistula refers to communication between a coronary artery and a cardiac chamber.

This entity is responsible for 0.2–0.4% of cardiac anomalies, with a prevalence of approximately 0.7% based on coronary computed tomography angiography. About 20% of patients with coronary fistula have associated congenital heart disease (septal defects, bicuspid aortic valve, aortic coarctation, tetralogy of Fallot, transposition of the great arteries and, more commonly, pulmonary atresia with an intact ventricular septum). It was postulated that the embryological basis for congenital coronary fistulas is the persistence of portions of the embryonic coronary sinusoids that connect the primitive coronary arteries to the cardiac chambers.²

Case report

A 34-year-old woman undergoing a perioperative evaluation of the surgical correction of an umbilical hernia reported dyspnea on moderate effort, orthopnea, and tachycardic palpitations occurring at least weekly. She also affirmed that, during her first pregnancy at 16 years of age, she experienced severe dyspnea and that episodes of convulsive crises began in this period.

The patient's blood pressure was normal, and cardiac auscultation revealed a regular heart rhythm in two stages, with evidence of a systolic murmur (+3/+6) that was more clearly audible in the lower left sternal border. General and cardiovascular physical examinations revealed no other findings. Electrocardiography showed an unchanged sinus rhythm.

Two-dimensional transthoracic echocardiography showed dilation of the left chambers, right atrium, and coronary sinus. The trunk of the left coronary artery was dilated (Figure 1) and originated a dilated circumflex artery (Figure 2), with

continuous and turbulent flow at the color Doppler, and a fistulous path, which drained into the coronary sinus and, sequentially, into the right atrium (Figures 2 and 3). Her biventricular systolic function was normal.

The patient was sent to a referral cardiology center and underwent coronary computed tomography angiography, which showed a long and dilated left coronary artery trunk and a circumflex artery with a fistulous and dilated path (Figure 4A) draining into the coronary sinus.

Coronary cineangiography showed a long coronary-cameral fistula with large caliber and tortuosity (Figure 4B) and the impression of a path that followed to the coronary sinus and, sequentially, the right atrium.

After a period of clinical follow-up, the patient underwent surgical correction of the fistula. The intraoperative description was of an aneurysmatic circumflex artery (approximately 2 cm in diameter) from its origin with a markedly tortuous path draining into the coronary sinus (referred to as a “great lake”). The intervention was successful and consisted of dissection and triple ligation of the circumflex coronary artery close to its origin. The patient's postoperative clinical progression was satisfactory, and she was discharged 6 days after the procedure.

Discussion

Coronary fistulas are mainly congenital. Acquired causes can be infectious, traumatic, or iatrogenic (percutaneous balloon coronary angioplasty, myocardial revascularization surgery, heart transplantation, permanent pacemaker placement, accessory anomalous bundle ablation, and endomyocardial biopsy). There are also reports of coronary fistulas being associated with myocardial infarction, hypertrophic cardiomyopathy, dilated cardiomyopathy, and tumors.³

Coronary fistulas can remain silent for years and be discovered incidentally during noninvasive or invasive procedures. However, patients can become symptomatic with age and/or increasing shunts. Symptoms include dyspnea on effort, fatigue, and angina pectoris; occasional complications can arise such as congestive heart failure, myocardial infarction, pericardial or pleural effusion, cardiac arrhythmias, and rupture of dilated aneurysmatic coronary arteries.⁴

Most statistics show greater involvement of the right coronary artery (60% of cases).² Fistulas rarely appear in the circumflex artery. The most common drainage area is the right ventricle, followed by the right atrium, pulmonary artery, and coronary sinus.⁵

Keywords

Diagnosis; Echocardiography; Fistula.

Mailing Address: Israel Nilton de Almeida Feitosa •

Rua José Bonifácio, 67 – Apartamento 1202 - Centro – Campina Grande/PB. CEP 58400-250

E-mail: inafeitosa@hotmail.com

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Case Report

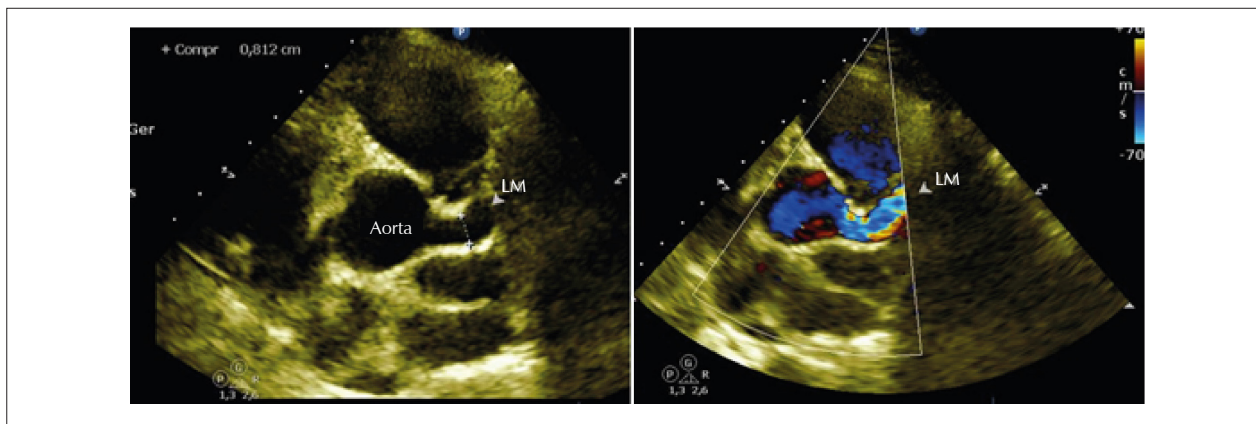


Figure 1 – Transthoracic echocardiography (short-axis imaging plane at the level of the base vessels). Left coronary artery trunk dilated to two-dimensional and demonstration of turbulent flow on color Doppler.

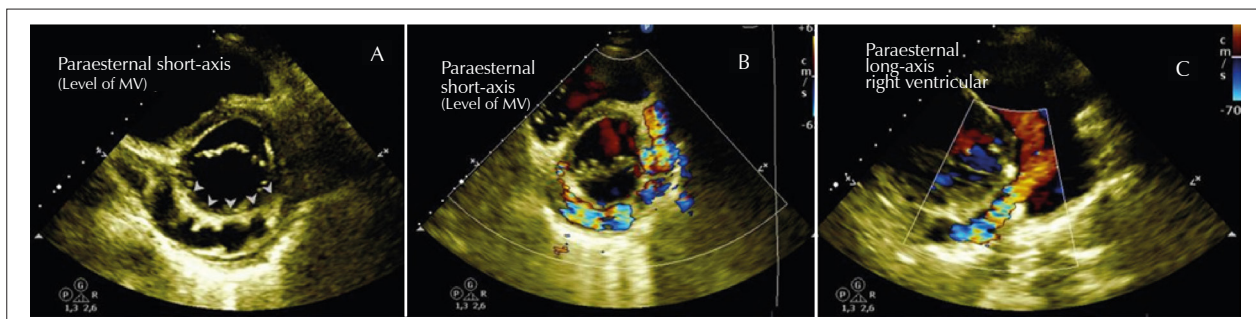
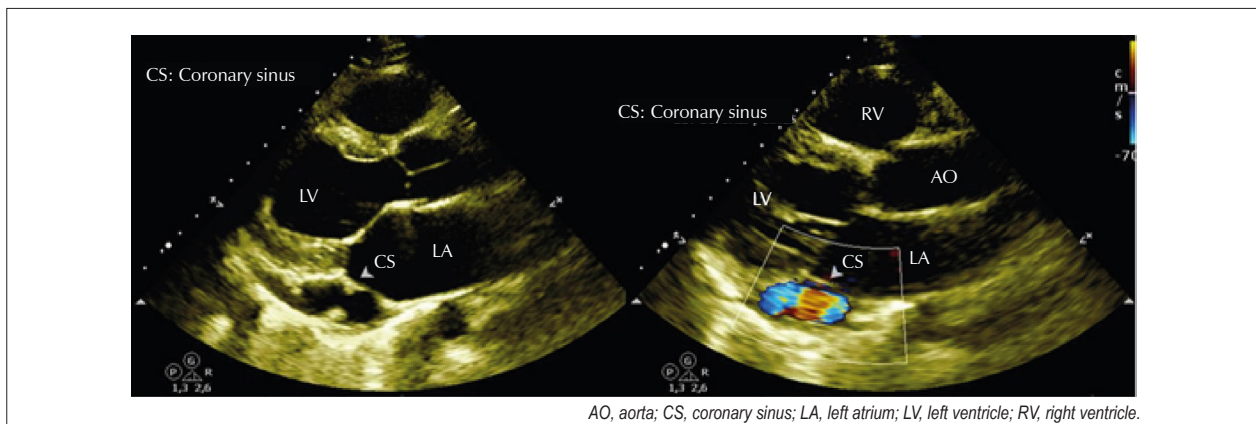


Figure 2 – (A) and (B) Transthoracic echocardiography (short-axis imaging plane at the level of the mitral valve). Circumflex artery dilated and tortuous to two-dimensional with turbulent flow to color mapping. (C) Parasternal view, longitudinal plane of the right ventricular entry path. Color flow mapping shows drainage of the fistula in the coronary sinus and, sequentially, in the right atrium.



AO, aorta; CS, coronary sinus; LA, left atrium; LV, left ventricle; RV, right ventricle.

Figure 3 – Transthoracic echocardiography (longitudinal plane of the left ventricle). Two-dimensional image and color flow mapping revealing an enlarged coronary sinus with turbulent flow inside.

Transthoracic echocardiography, the initial method of investigation for most cardiovascular conditions, is useful in the evaluation and diagnosis of coronary fistula. It is possible to identify a coronary artery or dilated chamber and a variety of congenital or acquired cardiac defects.⁶ Drainage can be verified

through color flow mapping. The use of microbubbles to increase color Doppler signals can help define fistula location and extent.⁷

Conventional angiography is traditionally the reference exam for making the diagnosis of coronary fistulas and can assist the choice of intervention that may be necessary.

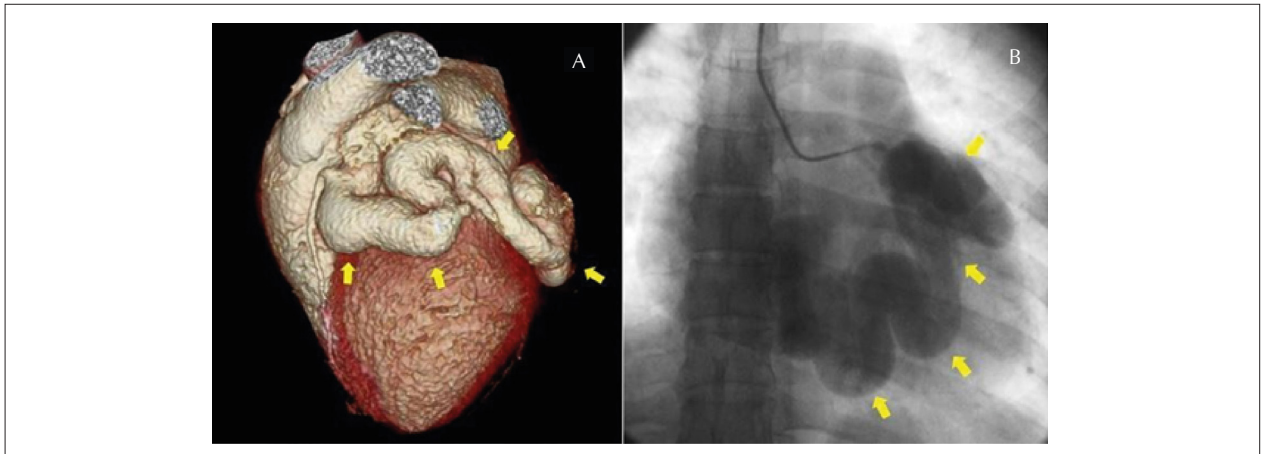


Figure 4 – Dilated circumflex artery with a tortuous path demonstrated by coronary angiotomography (left) and coronary angiography (right).

However, it is not the ideal method for documenting the artery's exact three-dimensional course. Also, drainage sites and aneurysms may not be well visualized using conventional angiography due to the significantly diluted contrast medium. Finally, the invasive nature of conventional angiography has a mortality rate of 0.1%.⁸

Despite the use of ionizing radiation, computed tomography coronary angiography is the reference noninvasive exam for viewing the coronary tree. High spatial resolution facilitates the demonstration of coronary anomalies and the presence of coronary artery disease. The procedure can show the origin of the coronary fistula, the drainage site (chamber or vessel), and the proximal and distal coronary anatomy. Coronary arteries as small as fifth-order branches can be visualized. Three-dimensional imaging establishes fistula size and location in any projection without repeated exposure to radiation or an additional contrast load for the patient.

Magnetic resonance imaging has limited ability to analyze coronary fistulas (trigger and movement artifacts, time consumption) and represents a research field that is not widespread in clinical practice. The coronary display is limited to the proximal course, mainly due to its lower spatial resolution and contrast/noise ratio compared with that of angiotomography.⁹

The management of coronary fistulas is based on the presence or absence of cardiovascular symptoms, degree of cardiac volume overload, and presence or absence of myocardial ischemia or ventricular dysfunction. Current recommendations include closing large fistulas regardless of symptoms.

Subsequent adverse events occur more commonly in patients with fistula draining into the coronary sinus regardless of surgical or percutaneous intervention. Even under open surgical inspection, this coronary anomaly can be particularly difficult to close completely due to multiple connections from

the distal coronary artery to the coronary sinus and the location of the connection at the posterior base of the heart.¹⁰

Surgery can be indicated in cases in which percutaneous treatment cannot be performed (high-risk procedure due to the proximity of an adjacent coronary artery, multiple fistula connections, and difficulty accessing the abnormal coronary artery or its branches). It can also be used when there is a coexisting condition that justifies surgical treatment. Options include external ligation of the coronary fistula, an internal patch, or suture closure of the fistulous communication orifice.

Conclusion

Several diagnostic modalities are currently available to visualize coronary artery fistulas, including noninvasive exams that can provide essential information to guide the most appropriate clinical management and surgical planning.

Considering their clinical variables and anatomical and physiological characteristics, the therapeutic approach to coronary fistulas must be individualized.

Authors' contributions

Research conception and design: Feitosa INA; data collection: Feitosa INA; data analysis and interpretation: Feitosa INA; manuscript writing: Feitosa INA, Gonzaga ARAD, Andrade JS, and Sá MRV; critical review of the manuscript for important intellectual content: Feitosa INA; and collection of echocardiographic images: Feitosa INA.

Conflict of interest

The authors have declared that they have no conflict of interest.

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