Diagnostic Approach to Coronary Artery Fistulae with Magnetic Resonance, Echocardiography and Coronary Angiography

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Learning objectives

We reviewed more than 2800 cardiac MRI studies performed in our center until October 2011.

We describe the imaging findings of 4 out of 5 patients with coronary fistulas evaluated by MRI, echocardiography and invasive coronary angiography.

In this poster we detail the information provided by each technique for the diagnosis and its main advantages and limitations.

Background

The coronary artery anomalies include abnormalities in the number, origin, course, termination and structure. The coronary artery fistulas (CAF) are classified as termination abnormalities and can be considered as a major anomaly.

A coronary artery aneurysm, is a focal dilatation that exceeds 1.5 times the size of the largest diameter measured in the coronary tree of the patient.¹

Coronary fistulas are direct connections between precapillary coronary artery and a cardiac chamber or vessel, as the superior vena cava, the coronary sinus or pulmonary circulation.²

Imaging findings OR Procedure details

The fistulas can be congenital or acquired. The acquired can be traumatic, more frequent between the right coronary artery and the right margin of the heart, or iatrogenic, usually secondary to endomyocardial biopsy, pacemaker implantation, percutaneous transluminal coronary angioplasty or cardiac surgery.

This anomaly represents the 0.4% of congenital heart defects and approximately 50% of pediatric coronary vascular anomalies. 20% of patients with CAF have other concomitant cardiac abnormalities, the most common are aortic and pulmonary atresia, patent ductus arteriosus, Fallot’s tetralogy, coarctation of the aorta and hypoplastic left heart syndrome. 3% of congenital fistulas are associated with absence of contralateral coronary artery.
Generally there is only one CAF in each patient. Multiple CAF are rare, with an uncertain etiology, but have been reported associated with polimiositis.\textsuperscript{3}

There is no predilection for sex or race, and age at diagnosis is variable, but usually they are suspected in children, when a continuous heart murmur is heard. If the shunt is large, may present with signs of congestive heart failure or coronary insufficiency.

The CAF originate from the right coronary artery (RCA) in approximately 50\% of patients, in the left coronary artery (LCA) in approximately 42\% and 5\% in both. Most symptomatic fistulas originate from the RCA, whereas asymptomatic usually arise from LCA. Over 90\% of fistulas drain to venous circulation, resulting in a left-to-right shunt. In order of frequency, the drainage points are: right ventricle (41\%), right atrium (26\%), pulmonary artery (17\%), coronary sinus (7\%), left ventricle (3\%) and superior vena cava (1\%).\textsuperscript{4}

Most arterial fistulas are small and asymptomatic, being clinically undetectable until echocardiography or coronary arteriography are performed for another reason. Usually they do not cause any complication and may even resolve spontaneously. However, large fistulas are typically three times the size of a normal coronary artery and may or may not be symptomatic. When symptomatic, a coronary artery steal phenomenon usually exists, that leads to myocardial ischemia of the territory perfused by the vessel.\textsuperscript{5}

The mechanism is related to a diastolic pressure gradient and to blood steal from the coronary system to a low pressure cavity. If the fistula is large, intracoronary diastolic perfusion pressure decreases pathologically.\textsuperscript{6} This results in a progressive increase of the ostium and coronary artery diameter, leading to the formation of a true aneurysm\textsuperscript{7}, intimal ulceration or rupture, media degeneration, atherosclerosis, calcification, mural thrombosis and rarely artery rupture. The factors determining the hemodynamic significance of the fistula are the size of the communication, the receiving chamber pressure and the risk of developing myocardial ischemia.

**Diagnostic techniques:**

- Chest x-ray is usually normal, except in cases of significant shunt, in which cardiomegaly, signs of pulmonary venous congestion and interstitial edema, can be detected.

- Echocardiography is usually able to diagnose fistulae, although not always identify the full path of the fistula or the precise drainage point. It can also provide information about:
- Left cardiac chamber enlargement, secondary to significant shunt (volume overload) or systolic dysfunction secondary to myocardial ischemia.

- Dilatation, tortuosity and ectasia of the coronary artery that nourishes the malformation.

- Turbulent flow detected by color Doppler at the source (origin) or along the vessel lumen.

- Drainage point of the fistula: often detectable as a distortion of the flow signal. Using ultrasonographic contrast media, improves the drainage point detection.

- Continuous flow at some point of the descending aorta, which may lead to suspect a fistula.

- Dilatation of the coronary sinus, when this is the end point of the fistula.

- Diagnosis of pulmonary hypertension.

  • Coronary angiography shows the proximal segment of the CAF and evaluates its size and number. When a fistula drains to a low pressure chamber, the drainage point may not be visible by this technique, due to contrast media dilution.\(^8\)

This is an invasive technique, and due to the complex three-dimensional geometry of fistulae can be difficult to explore it completely, with two-dimensional fluoroscopy. Hemodynamic study may include the determination of the pulmonary artery pressure and quantification of the shunt, when the fistula is significant.

  • Magnetic resonance imaging (MRI): Permits to obtain high quality anatomical images, functional and myocardial viability information, and quantification of the severity of the shunt by calculating the ratio pulmonary flow / systemic flow (Qp/Qs), being more reliable than echocardiography. In addition, MRI is quite accurate in identifying the source and draining point.\(^9\)

  • Multidetector CT: is a very fast technique, acquiring images with higher temporal and spatial resolution than MRI. Detect anomalies in the origin and the course of the coronary arteries, as well as the presence of thrombi and aneurysms.

It is considered an alternative technique to echocardiography and coronary angiography, offering an anatomical cardiac and vascular map, that allows to understand the anatomical complexity of these anomaly, view to its surgical management.
Moreover, it has become a useful exploration for better planning and selecting embolization point and embolization device selection, in cases where percutaneous closure is intended. Another advantage of CT is its capacity to evaluate the venous system of the heart. The main drawback of this technique is exposure to radiation, which can be minimized using dose modulation techniques and bismuth protection covers.

Cases are presented here: Fig. 1 on page 7; Fig. 2 on page 7; Fig. 3 on page 8; Fig. 4 on page 9.

**Treatment:**

The natural history of fistulas is variable and its management is controversial. There have been reports of spontaneous small fistulae closure, secondary to thrombosis, and if the patient is asymptomatic and stable, wait and see policy could be advocated.

In cases where follow-up shows progressive dilatation, with consequent increased risk of thrombosis, endocarditis and tear, the need to treat should be considered by means of percutaneous or surgical closure. Similarly, if the patient is symptomatic with signs of heart failure, myocardial ischemia or high flow shunt, treatment should be advised to prevent further complications.

The treatment option will be defined by careful identification of the number of fistula connections, fistulous vessel characteristics, drainage points and risk estimation of myocardial ischemia and hemodynamic shunt secondary to fistula.

The goal of treatment is to close the fistula, preserving normal coronary flow. Those patients with large fistulas, multiple communications or significant aneurysmal dilatation may not be optimal candidates for transcatheter closure, requiring surgery. Both treatment techniques have similar effectiveness, morbidity and mortality.

There should be a diagnostic catheterization with or without therapeutic intent, which must define the hemodynamic significance of the fistula and provide detailed anatomic assessment of it.

One of the advantages of percutaneous closure is its ability to detect additional fistulas, which only become manifest after percutaneous closure of the primarily diagnosed fistula.
The method of selective embolization is performed with inflatable balloon, embolization with platinum microcoils or vascular plugs. Desirable conditions for transcatheter closure are:

- No significant collateral vessels that may be inadvertently embolized.
- Presence of single, narrow fistula.
- Absence of multiple fistulas.

The outcome of the CAF treatment is good, regardless of the technique used for closure. Approximately 4% of patients will relapse, requiring new treatment.

Images for this section:

![CASE ONE](image)

**Fig. 1:** Case 1
CASE 2

RIGHT CORONARY ARTERY FISTULA TO RIGHT VENTRICLE
An asymptomatic 41 years old female patient

Fig. 2: Case 2
CASE 3

FISTULA FROM ANTERIOR DESCENDANT TO RIGHT VENTRICLE
41 years old patient with the diagnosis of coronary fistula since he was 10. Chronic atrial fibrillation.

Fig. 3: Case 3
Fig. 4: Case 4

CASE 4

CIRCUMFLEX ARTERY FISTULA TO CORONARY SINUS
A 76 years old patient with right ventricular failure
Conclusion

Coronary fistulas are a rare and heterogeneous disease, in which imaging techniques are a cornerstone for diagnosis, treatment planning and subsequent monitoring.

References


**Personal Information**