Left anterior descending artery fistula to pulmonary artery

Fístula da artéria descendente anterior para artéria pulmonar

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ABSTRACT - Congenital anomalies and within them anatomical variations of coronary arteries, although uncommon, are being increasingly studied and diagnosed due to the close relationship of symptoms such as angina, dyspnea, syncope or arrhythmias in young people without comorbidities. This study aimed to present the case report of a patient with ischemic symptoms secondary to malformation of a coronary artery. We describe the moment of patient admission, diagnosis by complementary tests and therapeutic resolution of the case. Coronary fistulas, despite having a low incidence among congenital heart diseases, have become increasingly frequent, being better characteristic due to new diagnostic methods.

Keywords: Coronary vessel anomalies; Coronary angiography; Arterio-arterial fistula

INTRODUCTION

Coronary artery anomalies are congenital alterations including an extensive group of malformations with a wide variety in origin, path and distribution.¹ ²

Coronary artery fistula (CAF) is defined as a very rare anomalous connection between a coronary artery and a cardiac chamber or a main vessel, it is present in 0.002% of population, accounts for 0.4%³ of all cardiac malformations, and may be asymptomatic and benign or present with symptoms, such as chest pain and cardiac dysfunction in young adults, depending on the location. Fistulas are usually diagnosed by computed tomography angiography, coronary angiography, transesophageal echocardiography, or cardiac magnetic resonance imaging.⁴ ⁶

In the present study, we report the case of a young male patient diagnosed with fistula from the left anterior descending (LAD) artery to the pulmonary artery (PA) trunk with successful percutaneous treatment.
graphy was ordered, which showed the presence of an im-
portant fistula from the LAD artery to the PA trunk with a
QP/QS of 1.2 (Figures 1 to 3), and a later intervention was
scheduled.

The patient underwent percutaneous closure of the
LAD-AP fistula by means of coil interlock embolization
(Figures 4 to 6). The procedure was successful, and a con-
trol echocardiography showed no segmental changes, and a
preserved ejection fraction.

**DISCUSSION**

Coronary fistulas were first described in 1865, with
the first successful surgical repair in 1947. 5,7 Coronary ar-
tery fistulas are congenital anomalies with an estimated
incidence of 0.2 to 0.4%. 7,8 and result from persistence of
primitive myocardial sinusoidal circulation and primordial
epicardial vessels and may be associated with other con-
genital heart diseases, such as patent ductus arteriosus,
tetralogy of Fallot and ventricular septal defect. 8 However,
CAFAs may also be acquired, secondary to complications
from procedures like coronary angioplasty, coronary ar-
tery bypass graft surgery or after heart transplantation and
myocardial biopsy. 9

![Figure 1. Coronary angiography showing a coronary fistula (red arrow) arising from the left anterior descending artery (white arrow).](image)

![Figure 2. Coronary angiography, left oblique projection, showing in greater detail the presence of a coronary fistula (arrows).](image)

![Figure 3. Coronary fistula (red arrows) with drainage to the pulmonary artery trunk (white arrows).](image)

![Figure 4. Coronary fistula catheterization.](image)
Coronary artery fistulas are classified according to their origin, drainage site or complexity, which includes the number of fistulas and the presence of other accompanying anomalies (Table 1). The most frequent origin is the right coronary artery (52%), followed by the LAD artery (30%) and circumflex artery (18%).

The most frequent drainage sites are right ventricle (41%), right atrium (26%), pulmonary trunk (17%), coronary sinus (7%), left atrium (5%), left ventricle (3%) and the superior vena cava (1%).

Coronary artery fistula are usually asymptomatic during the first two decades of life; afterwards, the frequency of symptoms and complications increases. Symptoms may vary depending on the type of fistula, drainage site and

**Table 1. Classification of coronary fistulas**

<table>
<thead>
<tr>
<th>Classification</th>
<th>Description</th>
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<tbody>
<tr>
<td><strong>Etiology</strong></td>
<td></td>
</tr>
<tr>
<td>Congenital</td>
<td>Embryonic</td>
</tr>
<tr>
<td>Acquired</td>
<td></td>
</tr>
<tr>
<td>Iatrogenic</td>
<td>PCI, CABG, heart transplantation, PM placement</td>
</tr>
<tr>
<td>By diseases</td>
<td>Hypertrophic cardiomyopathy, acute MI, tumor</td>
</tr>
<tr>
<td>By trauma</td>
<td>Penetrating or non-penetrating</td>
</tr>
<tr>
<td>Injury by radiotherapy</td>
<td>Right coronary artery</td>
</tr>
<tr>
<td></td>
<td>Left coronary artery (left anterior descending, left circumflex, ramus intermedium)</td>
</tr>
<tr>
<td></td>
<td>Anomalous coronaries</td>
</tr>
<tr>
<td><strong>Origin segment</strong></td>
<td></td>
</tr>
<tr>
<td>Sakakibara type A</td>
<td>Originates from the proximal segment of the native vessel</td>
</tr>
<tr>
<td>Sakakibara type B</td>
<td>Originates from the distal segment of the native vessel</td>
</tr>
<tr>
<td></td>
<td>Coronary artery is dilated</td>
</tr>
<tr>
<td><strong>Drainage site</strong></td>
<td></td>
</tr>
<tr>
<td>Chamber fistula</td>
<td>Right or left atrium, left or right ventricle</td>
</tr>
<tr>
<td>Arteriovenous fistula</td>
<td>Pulmonary artery, coronary sinus, IVC</td>
</tr>
<tr>
<td><strong>Number</strong></td>
<td>Single or multiple</td>
</tr>
<tr>
<td><strong>Morphology</strong></td>
<td></td>
</tr>
<tr>
<td>Simple</td>
<td>Single vessel with single origin and single termination</td>
</tr>
<tr>
<td>Complex</td>
<td>Fistula with a complex morphology</td>
</tr>
<tr>
<td><strong>Termination mode</strong></td>
<td></td>
</tr>
<tr>
<td>Macrofistula</td>
<td>Vessel visible on angiography</td>
</tr>
<tr>
<td>Microfistula</td>
<td>Vessel not visible but contrast is seen</td>
</tr>
</tbody>
</table>

Source: Reddy et al. PCI: percutaneous coronary intervention; CAGB: coronary artery bypass graft; PM: pacemaker; MI: myocardial infarction; IVC: inferior vena cava.
associated diseases; thus, the most common symptoms in adult patients are dyspnea, fatigue, and angina, with or without complications, such as endocarditis, arrhythmias, and congestive heart failure.9

The diagnosis can be made by Doppler echocardiography, which will show the anomalous flow of the fistula. However, the main diagnostic method is coronary cine-angiography, which will show the increase in pulmonary flow by measuring the ratio between the pulmonary and the systemic gradient (QP/QS) determining the direction of the shunt flow. Therefore, if QP/QS >1, the flow will be systemic-pulmonary, and if QP/QS <1, the flow will be pulmonary-systemic.8,10

The recommendations of the American Heart Association (AHA) and the American College of Cardiology (ACC) regarding treatment are the closure of CAFs associated with symptoms and also in asymptomatic patients with large fistulas, which pose risk of complications, such as endocarditis, pulmonary hypertension or heart failure.8,10,12

Coronary fistulas, despite having a low incidence among congenital heart diseases, have become increasingly frequent due to technological advances in diagnostic methods, especially in hemodynamic studies, which enable a more specific diagnosis and also offer the possibility of minimally invasive treatment and surgical resolution.

SOURCE OF FINANCING

None.

CONFLICTS OF INTEREST

The authors declare there are no conflicts of interest.

CONTRIBUTION OF AUTHORS

Conception and design of the study: DEBG and BDM; data collection: DEBG; data interpretation: BDM; text writing: DEBG; approval of the final version to be published: BDM.

REFERENCES