

## Incidental finding of a left coronary arteriovenous fistula in a 6-year-old schoolgirl

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### ARTICLE INFORMATION

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The authors declare no competing interests

### Acronyms

**CAVF:** congenital coronary arteriovenous fistulas

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### ABSTRACT

Coronary fistulas are congenital or acquired abnormalities in which there is direct communication between a coronary artery and any of the cardiac chambers or other vessels. An interesting case of a child with a left coronary arteriovenous fistula draining into the right atrium is reported. The diagnosis was made by an echocardiographic finding that was confirmed by cardiac catheterization. Angiographic images are shown, and the literature on this topic is reviewed in order to discuss some aspects of its epidemiology, clinical presentation, diagnostic possibilities, and the final conduct to face this disease.

**Key words:** Coronary fistula, Congenital heart disease, Pediatrics

### *Hallazgo fortuito de fístula arteriovenosa coronaria izquierda en escolar de 6 años*

### RESUMEN

Las fístulas coronarias son anomalías congénitas o adquiridas caracterizadas por la comunicación directa entre una arteria coronaria con cualquiera de las cámaras cardíacas u otros vasos. Se presenta un interesante caso de una niña con una fístula arteriovenosa coronaria izquierda que drena en la aurícula derecha. El diagnóstico fue un hallazgo ecocardiográfico que se confirmó mediante cateterismo cardíaco. Se muestran las imágenes angiográficas y se revisa la literatura al respecto para comentar algunos aspectos de su epidemiología, cuadro clínico, posibilidades diagnósticas, así como la conducta definitiva ante esta cardiopatía.

**Palabras clave:** Fístula coronaria, Cardiopatía congénita, Pediatría

### INTRODUCTION

Coronary fistulas are congenital or acquired abnormalities in which there is di-

rect communication between a coronary artery and any of the four cardiac chambers, the coronary sinus or its tributaries, the pulmonary artery, or a pulmonary vein near the heart. The great majority of congenital coronary arteriovenous fistulas (CAVF) terminate in the right cardiac chambers<sup>1</sup>.

There are two types of CAVF, the primary or isolated ones and the secondary or associated ones, which, as in some cases of pulmonary or aortic atresia, present a very different anatomical and clinical situation.

An interesting case of this disease is reported; in it, the left coronary artery and the right atrium communicate.

### CASE REPORT

A 6-year-old female patient, with a history of apparent good health, was auscultated during an acute respiratory infection and a heart murmur was found. Therefore, she was referred to the William Soler Pediatric Cardiology Center in Havana, Cuba. From a clinical point of view, she did not have any manifestation of cardiac dysfunction and her hemodynamic status was stable, however, hospitalization was decided for additional studies.

Physical examination showed a high pitched continuous murmur at the cardiac base, grade III/VI. Peripheral pulses were present, and were symmetrical and with a normal amplitude. The heart sounds were rhythmic and no third or fourth heart sounds were

auscultated. The nutritional assessment showed that the patient was eutrophic, with a weight-height relationship in the 25 percentile. Blood count, blood chemistry testing, coagulation tests and electrocardiogram were normal. The telecardiogram showed a cardiothoracic index of 0.56, dilatation of the main pulmonary artery and a slightly increased pulmonary flow.

A transthoracic echocardiography was performed to clarify the diagnosis. It showed the existence of a coronary arteriovenous fistula. Therefore, a hemodynamic study was conducted, with a coronary angiography, which was useful to confirm the diagnosis (Figure).

In this patient, percutaneous intervention was attempted but it was not successful, therefore, the final approach to be adopted was deferred, and surveillance was planned for follow-up visits, taking into account that the child remained asymptomatic.

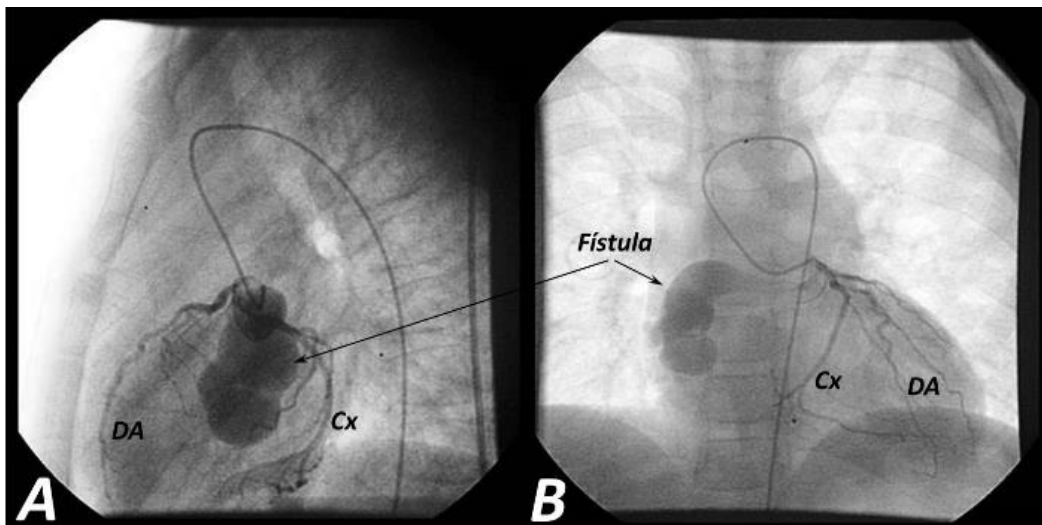
### COMMENTS

#### Epidemiology

In the context of congenital anomalies of coronary arteries, fistulas are classified as an anomaly of termination, and represent between 0.2 and 0.4 % of all congenital heart disease<sup>2</sup>. Despite their low incidence, they are the most common coronary anomaly, and are found in 0.15 % of all coronary angiographies<sup>3</sup>.

It is one of the most common congenital malformations of coronary circulation that allow survival until adulthood<sup>3,4</sup>.

The first description of CAVF was published by Krause<sup>5</sup> in 1865, as Abbot<sup>6</sup> commented in 1906, and the first surgical correction was performed by Björk and Crafoord in 1947<sup>7</sup>. These malformations are rare and their incidence can vary from 0.2 to 1.2 % of all coronary anomalies<sup>2-4</sup>.



**Figure.** Coronary arteriovenous fistula draining into the right atrium. **A.** Side view. **B.** Anteroposterior view with a slight right angle. DA: left anterior descending artery, Cx: circumflex artery.

### Anatomical aspects

The CAVF is a rare disease; Armsby *et al*<sup>8</sup> found two cases among 39 patients treated with coronary fistulas, 50% of these fistulas drain into the pulmonary artery and rarely in the left ventricle. Most often, one coronary artery is affected by the malformation, and it is rare that two or more major coronary arteries show this anomaly in a patient (5-6 % of all cases of coronary arteriovenous fistulas)<sup>4,8</sup>. The development of these alterations has an embryonic basis, because, in the sponge phase of the ventricular walls, the intertrabecular spaces allow communication between the cavities of the ventricles and the primitive coronary arteries of pericardial origin. It is possible the existence of a predetermining hereditary factor of embryonic origin, since there have been reports in the literature of familial diseases associated with these CAVF<sup>1,9</sup>.

In 55% of cases, the site of origin of the fistulous structure is the right coronary artery, in 35% it is of the left coronary artery, and in 5% of cases it occurs in both; 90% of fistulas terminate in the right heart or right circulation. In order of frequency, they may end in the right ventricle, right atrium, coronary sinus and pulmonary circulation. Fistulas rarely end in the left ventricle<sup>4</sup>.

### Clinical presentation

Although its natural history is not entirely clear, coronary fistulas are usually well tolerated for a long time. Those of small dimensions do not show symptoms or progress. Those of large dimensions may be symptomatic in children and young adults; and those of medium size cause progressive ventricular overload, clinical manifestations appear much later.

Therefore, they may be asymptomatic until adulthood, when CAVF may cause angina by coronary steal, dyspnea, pulmonary hypertension, manifestations of infective endocarditis and heart failure<sup>4,10</sup>, and there are anecdotal reports of hemoptysis in the literature<sup>11</sup>. On physical examination, the most important findings may include a continuous murmur, signs of heart failure, pulmonary hypertension and coronary ischemia<sup>4,10,12</sup>.

These manifestations of heart failure, infective endocarditis, and myocardial ischemia increase in frequency with age and are related to the dimensions and hemodynamic characteristics of the shunt<sup>2-4</sup>.

### Diagnosis

Cardiac catheterization is the procedure of choice to determine the anatomy of the coronary anomaly and its hemodynamic consequences, and to identify existing cardiac abnormalities or the existence of coronary obstruction. Transthoracic and transesophageal echocardiography (using Doppler) are also useful, as well as tomography with contrast, and more recently magnetic resonance imaging. Selective angiography may provide additional information<sup>13,14</sup>.

### Management

With regard to the final conduct, opinions are divided. Some authors recommend the closure of all fistulas during childhood, even asymptomatic ones. Others, however, advocate treatment only in symptomatic patients or those at risk of complications, such as cases of coronary steal, aneurysms or significant arteriovenous shunt, which may cause myocardial ischemia<sup>4,12</sup>. Currently, percutaneous treatment is proposed as an elective procedure. It is less radical and requires a shorter hospital stay<sup>15</sup>. Surgery is reserved for cases with multiple fistulas, percutaneous treatment complications (affecting large branches during embolization with coils), or when the fistula tract is narrow, restrictive and drains into a cardiac chamber<sup>16</sup>.

Various products are used to embolize the fistula; they include polyvinyl alcohol foam, balloons<sup>17</sup> and covered stents<sup>18</sup>. However, there is no doubt that the most accepted devices at present are the coils<sup>4,19,20</sup>. Although complications have been reported with the use of coils in some isolated cases, they have been exceptional and with little significance<sup>20,21</sup>.

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