Percutaneous Coronary Arteriovenous Fistula Occlusion in Infants with Left Ventricular Dysfunction

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Introduction

Congenital coronary fistulas are rare anomalies in which there is connection from the coronary arteries to large vessels or cardiac chambers, without the need for anomalies of number, origin or course of coronary arteries. They usually present in an isolated way, and can have severe hemodynamic consequences. Diagnosed in one of every 50,000 live births, it has an incidence of approximately 0.4% among congenital heart diseases.1-3 Most of them involve the right coronary artery (50%), followed by the left coronary artery (30%) and both left and coronary arteries (5%). They most commonly drain into a right sided cardiac chamber (more than 90%).1,4 The clinical presentation depends upon the size and location of the fistula, and may vary from just auscultation of heart murmurs, in general continuous, to severe heart failure.5,6

The diagnosis after the clinical suspicion can be confirmed by color Doppler two-dimensional echocardiography, but the angiographic study of the coronary arteries is essential for better anatomical definition and treatment decision-making.2,7 Nowadays the therapeutic option of choice is the percutaneous occlusion, with excellent short- and long-term results.1,5,8

Case report

Eight-month-old infant, male, born by c-section, weighing 2.9 kg and with a body length of 48 cm, forwarded to our service for assessment due to moaning, fatigue and irritability during breastfeeding. On physical examination, he presented good height and weight development (weight 8.1 kg, length 70 cm), in regular general condition, normal colored, hydrated, acyanotic, tachypneic, with mild intercostal retraction. In respiratory auscultation, he presented mild basal crackles. Cardiac auscultation showed regular rhythm, normal heart sound and presence of a high-frequency continuous murmur in high right sternal border, in addition to a third heart sound in the mitral area. He presented mild hepatomegaly on the palpation of the abdomen.

We initiated a treatment of heart failure with diuretics (Furosemide and Spironolactone) and an inhibitor of angiotensin-converting enzyme (Captopril) and requested an electrocardiogram, echocardiogram and chest X-ray. The electrocardiogram did not show any alterations. The echocardiography showed the presence of a left CAF draining into the right atrium – right atrium; dominant right coronary artery of good caliber, and with normal course. The test revealed dilation of left coronary artery trunk extending to the circumflex artery, which was enormously dilated in its entire course (Figure 1A), ending in a blood pool that drained through a double-outlet right atrium (Figure 1B). The inferior cardiac vein was filled from the aneurysmal sac. The anterior descending artery showed adequate caliber in its initial portion and tapered in the distal portion, due to major steal of coronary flow caused by...
by the left circumflex fistula. The angiography revealed mild-to-moderate global LV systolic dysfunction, absence of mitral regurgitation and increased cavity dimensions, without interventricular communication. There was mild pulmonary artery hypertension with elevated RVEDP and RVIDP, elevated right atrium mean pressure and LVEDP.

The patient was re-evaluated after 30 days of treatment, with good clinical improvement but, since the fistula was large and had hemodynamic consequences, we decided to perform a percutaneous occlusion.

Through puncture of the right femoral artery, a 5F introducer was inserted, through which a guide catheter (Judkins 5F) was introduced, and the left coronary trunk was selectivated. A 0.014” guidewire was introduced through the guide catheter and positioned distally in the left circumflex artery near the right atrium outflow. The guide catheter was removed and an Envoy 5F (Cordis) catheter was introduced over the guidewire and positioned distally in the left circumflex artery. The guidewire was removed and an Amplatzer Vascular Plug I (Figure 2A) was introduced through the Envoy guide catheter, being released in the mean left circumflex artery. After release of the Amplatzer Vascular Plug, which was well adapted, without any mobility, there was a progressive decrease in flow through the left circumflex artery, until complete disappearance of flow through the fistula. Left coronary trunk angiography, performed 30 minutes after plug release, showed complete occlusion of the AVF (Figure 2B), with clear flow improvement in the LAD and LMA due to the elimination of coronary steal.

The echocardiographic study performed hours after the percutaneous treatment revealed complete recovery of the LV function (ejection fraction: 71.6%) and absence of flow through the coronary arteriovenous fistula. The patient was discharged on the following day with diuretics (Furosemide and Spironolactone) and an inhibitor of angiotensin-converting enzyme (Captopril). During the 30-day follow-up period he did not present any complications, physical examination showed normalized pulmonary and cardiac auscultation and disappearance of hepatomegaly; echocardiogram showed normal ventricle function (ejection fraction: 68.2%) and absence of flow through the fistula, thus a decision was made to discontinue the medication.

Discussion

Coronary artery fistulas are rare lesions among the pediatric population, with an approximate incidence of 0.3 to 0.8%, and quite variable clinical manifestation. The most frequent symptom in children younger than 2 years is heart failure, whereas in older children dyspnea on exertion is more prevalent. Complications like myocardial ischemia, embolization and arrhythmia are
more frequent after the second decade of life in large-caliber fistulas.\textsuperscript{2,9}

The vast majority of anomalies involve the right coronary artery, with drainage into the venous circulation (right ventricle, right atrium, pulmonary artery and venous sinus); the left coronary is rarely involved.\textsuperscript{5,7,8}

The clinical diagnosis is suspected by the symptomatology and exclusion of other heart diseases that may progress with similar symptoms, but with a cardiac murmur showing different characteristics (patent ductus arteriosus, aortopulmonary window, pulmonary arteriovenous malformations, and pulmonary vein stenosis).\textsuperscript{3} Two-dimensional echocardiography can reveal the consequences of the lesion (dilated chambers and coronary arteries, ventricular dysfunction and elevation of Qp:Qs), and in some cases, it allows for the direct visualization of the fistula and its track.\textsuperscript{3,7} The previous hemodynamic study is essential, because it assesses in detail the anatomy and the site where the fistula drains, allowing for more accurate and less risky therapeutic decision-making.

According to the most recent guidelines, there is consensus that all symptomatic patients should be treated. Occlusion of large fistulas is recommended regardless of symptoms; small- to moderate-sized fistulas, when there is evidence of ischemia, arrhythmia, endarteritis or ventricular dysfunction.\textsuperscript{10} The procedure remains limited when the anatomy of the fistula is not favorable, i.e., when there is a native coronary branch distally to the fistula and high risk for prosthesis embolization.

Although the surgical correction presents a low morbimortality rate and good long-term evolution, nowadays the method of choice for the treatment of coronary fistulas is percutaneous, due to its low cost, length of hospital stay and reduced complications.\textsuperscript{5,6,8} European and Asian cohorts present favorable results in the short and long term, with the elimination of the flow through the fistula and resolution of complications and symptomatology in the vast majority of patients undergoing percutaneous occlusion.\textsuperscript{5,6,8}

Recently, several single and combined devices have been developed and their techniques for implantation have been improved.\textsuperscript{5,8} The most frequently used are the Amplatzer Duct Occluder, Amplatzer Vascular Plug and flipper coils.\textsuperscript{5} A previous and thorough evaluation of the fistula geometry and size is crucial to guide the choice of the best device or combinations of occlusion methods.

We reported the case of left coronary arteriovenous fistula occlusion in an eight-month infant, with symptomatology of heart failure and ventricular dysfunction. The diagnosis was confirmed by echocardiography and the cardiac catheterization was important to assess the fistula size, its hemodynamic consequences and to define the treatment plan. In this report, as well as in the last studies, there were no complications during or after the procedure. The left ventricle function was recovered.
almost immediately and the flow was completely interrupted, as shown by the echocardiographic evaluation performed less than 24 hours after the procedure and during the 30-day follow-up period. As in most of the studies reviewed, in our service this is the approach of choice for congenital coronary fistulas, with optimal results.

Author contributions

Conception and design of the research: Jacob MFFB, Faria DG, Jacob JLB. Acquisition of data: Jacob MFFB, Faria DG, Jacob JLB. Analysis and interpretation of the data: Jacob MFFB, Faria DG, Jacob JLB. Statistical analysis: Jacob MFFB, Faria DG, Jacob JLB. Writing of the manuscript: Jacob MFFB, Faria DG, Jacob JLB. Critical revision of the manuscript for intellectual content: Jacob MFFB, Faria DG, Jacob JLB.

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References