Right Coronary Artery Fistula with Congestive Heart Failure in the Neonate.
Doppler Echocardiographic Diagnosis and Closure with Detachable Balloon

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We report here a case of coronary artery fistula in a neonate with clinical signs of heart failure. The electrocardiogram showed signs of left ventricular hypertrophy and diffuse alterations in ventricular repolarization. Chest X-ray showed an enlargement of the cardiac silhouette with an increase in pulmonary flow. After echocardiographic diagnosis and angiographic confirmation, closure of the fistulous trajectory was performed with a detachable balloon with an early and late successful outcome.

Coronary artery fistula is a rare disease accounting for approximately 0.2% to 0.4% of all congenital cardiac defects. It was first described by Krause in 1865 and, since then, approximately 400 cases have been reported with a large variation in regard to the site of origin and drainage in the cardiac chambers. Congestive heart failure in the neonatal period resulting from coronary artery fistula is extremely infrequent, and most of the time it is diagnosed in a routine examination because of the presence of a continuous murmur.

With the appearance of Doppler echocardiography, the diagnosis could be established in a noninvasive way, leaving the invasive procedures for confirmation and treatment of the disease.

We report a case of right coronary artery fistula with congestive heart failure in a neonate diagnosed through Doppler echocardiography and confirmed with angiography. The fistula was closed in the catheterization room.

The patient is a full-term male neonate weighing 3,900g. He was born from a normal delivery, the first child, and from the São Paulo area. No abnormalities in regard to the gestational antecedents were reported. The main complaint was dyspnea on feeding since the first day of life. On the third day of life, the patient was admitted to the hospital because of dyspnea and jaundice.

The patient was transferred to our service at the age of 40 days in regular condition, pale (+/+), hydrated, anicteric, acyanotic, afebrile, tachycardic, tachypneic, active, and reactive.

On physical examination, the liver was palpated 3cm from the right costal margin, the spleen was unpalpable, and lung auscultation revealed symmetric respiratory sounds, without other noises. Diastolic thrill could be heard on the lower left sternal margin and cardiac auscultation showed rhythmic beats of normal intensity with a continuous murmur (3+/4+), more intense in the lower left parasternal region irradiating to the base of the back. Hemoglobin and hematocrit values were 10.5g/dL and 31%, respectively.

The electrocardiogram showed a sinus rhythm, heart rate of 160 bpm, biventricular hypertrophy, with predominance of the left ventricle, and diffuse alterations in the ventricular repolarization (primary inversion of the T wave in some leads, without necrotic areas). Chest X-ray showed global cardiomegaly (2+/4+) with bilateral increase in pulmonary flow.

The patient received furosemide, spironolactone, and Cedilanid-D. Doppler echocardiography was then performed showing situs solidus, atrioventricular and ventriculoarterial concordance, intact interatrial and interventricular septa, moderate dilation of the right ventricle, and significant reduction in its systolic function (hypokinesia of
the right ventricular free wall and of the posterior region of the interventricular septum). The right coronary artery showed a significant dilation in its origin (9mm), which represented approximately 75% of the diameter of the aortic root, and after a short trajectory in the atrioventricular sulcus the coronary artery drained right below the sulcus in the lateral region of the right ventricle. No aneurysm in the site of drainage was observed (figure 1 and 3A).

Coronary angiography confirmed the echocardiographic findings (figure 2) showing a narrowing (neck) at the point of drainage in the myocardium, allowing a possible closure with a balloon. In addition, a retrograde filling of the distal region of the right coronary artery by collaterals from the anterior descending artery was observed. We chose to occlude the fistulous trajectory with a detachable balloon GVB 16, whose diameter was larger than that of the fistulous neck (figure 3). No complications occurred during introduction, transportation, and inflation of the balloon. Then, manual injection of the contrast medium was performed showing a complete closure of the fistula (figure 3B) with no change in the distal filling of the right coronary artery and with neither hemodynamic nor electrocardiographic repercussions. A new injection of the contrast medium was performed in the right coronary ostium after balloon deployment, and the fistulous trajectory and the contrast medium inside the ventricle could no longer be observed (figure 2C). Injection of the contrast medium in the ostium of the left coronary artery showed good perfusion of the right coronary artery through the collaterals (figure 2D).

The patient had a good clinical and echocardiographic evolution with reduction in the right ventricular diameter, improvement in ventricular function, and disappearance of the previous flow through the fistulous trajectory. Chest X-ray performed after the procedure allowed the visualization of the topographic location of the balloon in regard to the cardiac chambers (figure 4).

**Discussion**

Coronary artery fistula is an infrequent disease,
whose diagnosis may be easily established echocardiographically\(^3\).

Approximately 55% to 60% of the fistulas involve the right coronary artery and 90% of them drain into the small circulation (right ventricle, 45%; right atrium, 25%; pulmonary artery, 15-20%; coronary sinus, 7%). Small fistulas draining into the pulmonary artery are more frequent in the left coronary artery, and in 4-5% of the cases both coronary arteries may be involved. In this last case, survival of 9 hours to 5 months has been reported\(^4\).

Usually symptoms begin late (75% of the patients older than 40 years of age have dyspnea or congestive heart failure), which makes our case even rarer. Our patient had signs of heart failure in the first days of life, probably due to the significant dilation of the coronary artery with right-left shunt. He also had signs of ischemia on the electrocardiogram (alterations in the ventricular repolarization) and on the echocardiogram, with significant areas of hypokinesia, which is also more frequent (80%) in individuals older than 50 years of age\(^5\).

Left ventricular hypertrophy on the electrocardiogram is a common finding in large fistulas because of the increase in pulmonary flow (shown in the angiography in our case).

Even though surgical mortality is low in some centers with experience in this therapeutical option (annual mortality of 0-4% in reports of children older than 2 years of age, morbidity of 7%, postoperative myocardial infarction of 3.6%, and recurrence of the fistula of approximately 9%)\(^5\), occlusion through catheterization is usually a procedure with low morbidity and mortality, in addition to reducing the hospitalization costs. Little experience exists with this procedure, especially in neonates, because of the rarity of the disease and the difficulty in occluding fistulas with increased flow. In
addition, potential risks of manipulation of the coronary arteries occur. Use of the detachable balloon has been described as a precise method that produces rapid occlusion of the fistula. Among the disadvantages, we can cite the need for a longer guide catheter and the risk of precocious emptying of the balloon with unexpected systemic or pulmonary embolism. This procedure, however, has not been reported in children less than 2 months of age, as in our case, probably because of the rarity of symptomatic children in this age group.

Our case was selected for closure of the fistula after demonstration of an adequate “neck” and verification of collaterals to the right coronary artery, which allowed its filling through retrograde via without impairment of the circulation depending on this coronary artery.

Data on long-term evolution of patients undergoing this type of treatment are still lacking, and we do not know the chances of recurrence of the fistula in this case. Early and late (2 years) follow-up, however, was satisfactory with improvement in ventricular function and absence of the continuous murmur on cardiac auscultation.
References