

Correction of anomalous origin of left coronary artery with mitral insufficiency and mechanical hemolysis

Correção da origem anômala de artéria coronária esquerda com insuficiência mitral e hemólise mecânica

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Abstract

A 2-month-old girl presenting with heart failure and murmur was investigated using Doppler echocardiography and cardiac catheterization revealing an anomalous left coronary artery origin from the pulmonary trunk and massive mitral regurgitation. The patient was submitted to surgery under cardiopulmonary bypass: the anomalous coronary artery was implanted in the aorta and the mitral valve repaired by annuloplasty. In the postoperative period the patient had important mechanical hemolysis caused by the mitral annuloplasty. The patient underwent a second intervention to remove a piece of bovine pericardium. A postoperative echocardiogram revealed mild mitral insufficiency and the patient is free of symptoms.

Descriptors: Coronary vessel anomalies, surgery. Pulmonary artery, abnormalities, surgery. Anemia hemolytic. Hemolysis. Mitral valve insufficiency.

Resumo

Criança do sexo feminino, 2 meses de idade, apresentando quadro clínico de insuficiência cardíaca e sopro. Durante a investigação, foi realizado ecocardiograma e estudo cineangiográfico que evidenciaram origem anômala da artéria coronária esquerda do tronco pulmonar e insuficiência mitral importante demonstrada ao doppler. A paciente foi submetida a tratamento cirúrgico com auxílio de circulação extracorpórea, que consistiu no reimplante da artéria coronária esquerda na aorta, associado à anuloplastia da valva mitral. No pós-operatório tardio, houve importante hemólise mecânica ocasionada pela plastia mitral. A paciente foi reoperada para remoção de retalho de pericárdio bovino. O ecocardiograma pós-operatório mostra insuficiência mitral leve e o paciente apresenta-se em classe funcional I (NYHA).

Descritores: Anomalias dos vasos coronários, cirurgia. Artéria pulmonar, anormalidades, cirurgia. Anemia hemolítica. Hemólise. Insuficiência da valva mitral.

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INTRODUCTION

The anomalous origin of the left coronary artery from the pulmonary trunk is more frequent in the left posterior pulmonary sinus. Rarely the abnormal coronary ostium is located near to the other pulmonary sinus and when this occurs, it is more frequently in the right posterior sinus than the right anterior. After an initial section of variable length, the coronary trunk divides into the anterior descending and circumflex arteries. The right coronary artery is generally dilated, whilst the left is normal or has a smaller caliber.

Because the pressure and oxygen saturation is similar in the aorta and in the pulmonary artery during intra-uterine development, this anomaly does not have repercussions on the fetus. After birth, however, the arterial duct closes and the pulmonary blood resistance progressively drops causing chronic hypoperfusion of the corresponding myocardial region. The severity of the myocardial consequences depends of the relative dominance of the right and left coronary arteries and of the significance of collateral arteries that develop between them. When the left coronary artery is dominant, dilatation with hypokinesia of the left ventricle occurs, which is frequently associated with mitral valve insufficiency due to ischemia of the papillary muscles. The clinical state is characterized by severe dilated heart disease so the prognosis depends of immediate surgical intervention.

The recommended surgical treatment of this anomaly is the re-implantation of the coronary artery in the aorta and annuloplasty of the mitral valve [1,2].

In this report we describe a case of an anomalous origin of the left coronary artery from the pulmonary trunk associated with a massive mitral regurgitation in a 2-month-old infant who underwent a successful surgery.

CASE REPORT

A 2-month-old girl, weighing 5430g, was hospitalized with congestive heart insufficiency. The blood pressure was 95 x 54 mmHg, heart rate was 140 bpm and her respiration was 72 breaths per minute. She was pale, hydrated and acyanotic, presenting with medial-sternal thoracic arching, hyperdynamic precordium, an audible systolic murmur +++/6 at the left sternal border with irradiation to the axilla and dorsum. Her pulses were of normal amplitude, lung auscultation was normal, but she had hepatomegaly with the liver 4 cm from the right costal edge. A chest radiogram showed an increased cardiac area, a bulged pulmonary artery trunk and increases in the left atrium and ventricle. The ECG was sinus, axis at + 40°, the PR and QTc were normal, with a overload on the left atrium and ventricle and a necrotic Q wave at DI and AVL. The Doppler echocardiograph showed

significant overload of the left atrium and ventricle, diffuse contractile deficiency of the left ventricle and severe mitral insufficiency. Additionally, the left coronary artery measured 2 mm at its root, the right coronary artery was 3 mm at its root and 2 mm approximately 1 cm from the right coronary ostium.

Cardiac catheterization identified an anomalous origin of the left coronary artery from the pulmonary artery trunk, dilated right coronary artery with normal origin and course connected to the left coronary artery by collateral arteries. A surgery was performed with transversal opening of the pulmonary trunk and identification of the left coronary artery ostium near to the right posterior sigmoid artery. The ostium was removed with an extensive flap of the pulmonary wall and the lateral orifice in the ascending aorta was opened. Re-implantation of the ostium in the posterior edge of the aortic orifice followed. The anterior wall was repaired with autologous pericardium treated in 2% glutaraldehyde. The pulmonary trunk was also repaired with an autologous pericardium patch shaped similar to that which had previously been removed (Figure 1).

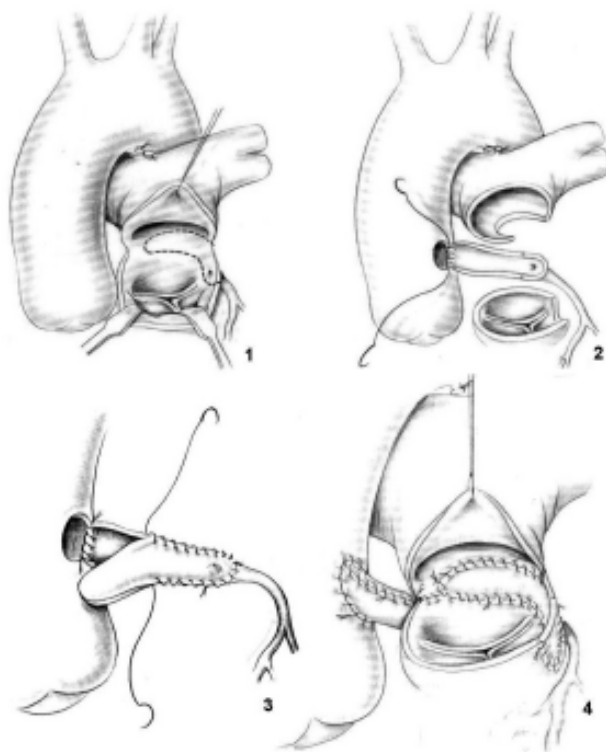


Fig. 1 – Diagram of the re-implantation of the left coronary artery in the ascending aorta

On opening the left atrium, significant mitral annular dilation was evidenced with lengthening of the chords and slight prolapse. Plicature of the two commissures was performed according to the Reed technique [6] using four fragments of bovine pericardium. The sternum was only closed five days after the procedure. The patient evolved with hemolytic anemia in the postoperative period (progressive icterus and hematuria). The patient presented with a positive direct Coombs test on the ninth postoperative day suggestive of immune hemolysis due to multiple transfusions. From then on only irradiated erythrocytes were transfused.

Although, the hemolytic anemia continued, the direct Coombs appeared negative in subsequent examinations and a urinary iron test was positive and so microangiopathic hemolytic anemia due to mechanical trauma was diagnosed – probably related to the mitral valve annuloplasty. A Doppler echocardiograph was performed which verified normal left ventricular function, the mitral valve with good diastolic movement and a regurgitation streaming directly over the ring and one of the commissures. The patient underwent a valvar plastic surgery to remove the fragments of bovine pericardium and to make annuloplasty without using pericardium. The patient left the hospital on the 10th postoperative day.

In the follow-up, the infant is free of symptoms and in functional class I (NYHA).

COMMENTS

Anomalous origin of the left coronary artery from the pulmonary trunk, when diagnosed in a neonate or infant, is generally severe and the left ventricular function is greatly affected [2].

The ideal surgical treatment of this anomaly is anatomic correction that consists of the removal of the left coronary ostium of the pulmonary trunk and its re-implantation in the ascending aorta [3]. In most cases the ostium is found in the left posterior pulmonary sinus, that is, next to the ascending aorta. In these circumstances, transfer is made without technical difficulty.

In rare cases, as in this one, in which the coronary ostium from the pulmonary trunk is at a greater distance from the aorta, anatomic correction is still possible [4,5]. For this, it is necessary to remove a large section of the posterior wall of the pulmonary artery along with the coronary ostium (Figure 1) and anastomose it on the posterior border of the newly-made aortic orifice thereby forming the posterior wall of the new left coronary trunk. The anterior wall is created from an autologous pericardium patch previously preserved in glutaraldehyde. The pulmonary trunk is reconstituted using a second autologous pericardium patch shaped similar to

what had previously been removed.

In respect to the mitral valve annuloplasty, the ring was placated near the commissures according to the Reed technique [6], using four strips of bovine pericardium to reinforce the sutures. With the progressive improvement of the left ventricular function, there was concomitantly an increase in the mechanic hemolysis caused by a regurgitation flow in the commissural region leading to high serum levels of bilirubin. This type of hemolysis is caused by a direct impact of the regurgitation flow (Figure 2) on fragments of bovine pericardium [7]. When this situation occurs, reoperation to remove the pericardium and possibly even valve replacement must be considered [7]. In the present case, valve replacement was not necessary. Instead the pericardium was removed and annuloplasty using a monofilament in the posterior portion of the ring was performed.

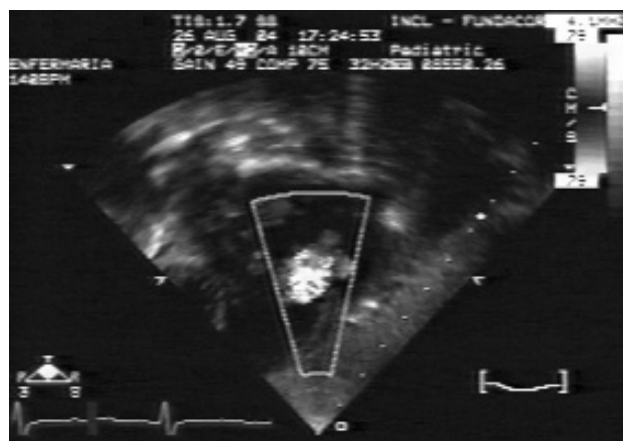


Fig. 2 – Doppler echocardiogram showing mitral regurgitation with a direct flow on the bovine pericardium strips

Based on the good evolution and technical applicability of re-implantation of the left coronary artery, even in exceptions, we believe that the surgical treatment of this congenital anomaly must be employed at the earliest possible moment. We recommend this surgical technique in cases similar to this. Likewise, with persistent hemolysis, reoperation of the mitral valve must be considered when regurgitation flows directly on the material utilized in the annuloplasty.

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