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Clinical-Surgical Correlation

Case 8/2004 – Pediatric Heart Surgery Service – Hospital de Base, Medical School, São José do Rio Preto

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CLINICAL DATA

We report on a 5-year-old male Caucasian patient weighing 16 kg. In the first year of life a heart murmur was identified during a routine check-up and the patient was clinically followed up until she was referred to our department. She was asymptomatic and did not require medications. She was in a good general state, ruddy, hydrated, acyanotic and with eupnaea. Her thorax was symmetrical, ictus cordis was palpable, normal heart sounds with a systo-diastolic murmur of ++++/6 at the medium left sternal border. The lungs had a symmetrical vesicular murmur without adventitious sounds. The abdomen was unchanged. The peripheral pulses were strong and symmetrical in all four limbs.

ELECTROCARDIOGRAPHY

The electrocardiographic examination showed sinusal rhythm with a frequency of 107 beats per minute and the electrical axis of the QRS complex was + 30º, thus within normal limits.

RADIOGRAPHY

The cardiac area was slightly increased with a cardiothoracic index of 0.53. The greatest increase was seen in the left ventricular chamber and there was a moderate dilation of the aortic arch. The pulmonary territories did not show changes.

Fig. 1 - Transverse aortotomy demonstrating the aortic side of the aorta-to-left ventricle tunnel together with the right coronary artery valve. Note that there is no rupture of the Valsalva sinus characterizing the aorta-to-left ventricle tunnel. The left coronary artery valve is also thick.

Fig. 2 - Aspect of the bovine pericardial patch on the side of the aorta

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ECHOCARDIOGRAPHY

The patient had situs solitus and levocardia. The venoatrial, atrioventricular and ventriculoarterial connections were all concordant. The patient had significant aortic valve insufficiency, dilation of the ascending aorta and of the Valsalva sinus corresponding to the right coronary artery valve with a large fistula between the ascending aorta and the left ventricle compatible with an aorta-to-left ventricle tunnel.

DIFFERENTIAL DIAGNOSIS

With the existing data, basically, a patent arterial duct or interventricular stunt with aortic insufficiency, ruptured aneurysm of the Valsalva sinus and coronary-chamber fistulas should be considered.

DIAGNOSIS

As well as the echocardiogram, a cardio cineangiography was performed which demonstrated significant aortic valve reflux and an image suggestive of an aorta-to-left ventricle tunnel. Magnetic nuclear resonance identified an increase in the calibers of the ascending aorta, pulmonary trunk and a funnel-shaped image that connected the left ventricle with the aorta confirming the initial diagnosis of an aorta-to-left ventricle tunnel.

OPERATION

A transsternal median thoracotomy was performed and cardiopulmonary bypass with antegrade sanguineous cardioplegia with moderate hypothermia at 28ºC was established. After a transverse aortotomy was performed and the aorta-to-left ventricle tunnel was identified (Figure 1), intermittent anterograde sanguineous cardioplegia at 4ºC was used directly in the coronary ostia. An investigation was made through the orifice of the aorta and a bulging of the right ventricle was discovered, which was opened at the outflow tract and a repair was performed in the region of the interventricular septum. Subsequently the orifice was closed through the aorta using a bovine pericardial patch paying special attention to the internal opening (left ventricular wall). A second patch was sutured on to the aorta (Figure 2). The left coronary artery valve was thickened probably due to a jet lesion and this was thinned. In the immediate post-operative period, the patient presented with systemic arterial hypertension which was initially controlled using intravenous nitroprusside and subsequently using an angiotensin converting enzyme inhibitor. The patient was released from hospital on the 8th postoperative day. An echocardiogram after six months demonstrated correction of the aorta-to-left ventricle tunnel, with slight aortic insufficiency and a normal left ventricular function.