Clinical-Surgical Correlation

Caso 1/2005 – Pediatric Heart Surgery Service – Hospital de Base, Medical School, São José do Rio Preto

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CLINICAL DATA

A 7-month-old female Caucasian infant weighing 5.3 kg, born at full term was referred to our department. At 2 months of age heart sounds were heard to the right of the chest and there was suspicion of dextrocardia. She was referred to a cardiologist for further investigations. At this time a congenital heart disease was diagnosed which was initially treated with digital and diuretics. The infant evolved with pneumonia and required hospitalization for a period of 30 days. She was in a good general state, ruddy, hydrated, acyanotic and with slight tachypnea. Her thorax was symmetrical; ictus cordis and fremitis were palpable at the 5th intercostal space on the hemiclavicular line. The heart rhythm was regular with two clicks with the second sound hyperphonetic and a systolic murmur of regurgitation of 4+/6 at the lower right sternal border. Pulmonary auscultation was symmetrical without adventitious sounds. The liver was palpable 3 cm from the left costal margin. The extremities had palpable and symmetrical pulses without pressure differences between the limbs.

ELECTROCARDIOGRAM

The electrocardiogram suggested dextrocardia with a negative P wave at the DI deviation on a horizontal plane. The rhythm was sinusal with a heart rate of 136 beats per minute, $SAP + 60^{\circ}$, electrical axis of the QRS complex in the 1^{st} quadrant ($SAQRS + 60^{\circ}$), PR 0.16, QR 0.04, QT 0.24 (QTc 0.36). Biventricular overload (Figure 1).

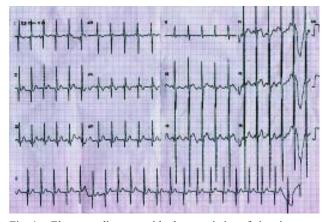


Fig. 1 – Electrocardiogram with characteristics of situs inversus and dextrocardia, obtained with the precordial derivations to the right and without inversion of the electrodes on the limbs

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RADIOGRAPH

The heart area was enlarged with a cardiothoracic index of 0.58 and dextrocardia. Moderate pulmonary vascular prominence was evidenced. Visceral situs inversus seen in an image was compatible with the liver to the left and the gastric "bubble" to the right (Figure 2).

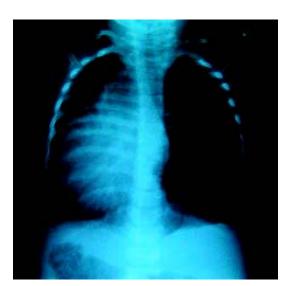


Fig. 2 – Radiogram demonstrating the presence of visceral situs inversus and dextrocardia

ECHOCARDIOGRAM

Situs inversus totalis in dextrocardia was evidenced. Venoatrial, atrioventricular and ventriculo-atrial connections were concordant. A slight increase of the right atrium and moderate increase of the left atrium were identified. The left atrioventricular valve showed slight insufficiency and the right atrioventricular valve moderate insufficiency. Moderate dilation of the pulmonary trunk and branches was evidenced. The echocardiogram evidenced an interventricular connection with extension to the outflow tract of the right ventricle with a diameter of 14 mm². The systolic pressure in the right ventricle was 98 mmHg as seen using Doppler and the mean pulmonary artery pressure was 50 mmHg, suggestive of significant pulmonary hypertension due to hyperflow with Qp/Qs of 2.8.

DIFFERENTIAL DIAGNOSIS

Pulmonary hypoflow diseases should be considered including interventricular or interatrial shunts, atrioventricular septal defect and persistent arterial duct. Occasionally, the presence of situs inversus totalis and

dextrocardia show the existence of a complex heart disease, however, this is not always the case and it is necessary to remember other causes such as atelectasis, diaphragmatic hernia and pneumothorax.

DIAGNOSIS

The echocardiograph was essential and definitive in the diagnosis of this case and a cineangiographic study was not necessary. The diagnosis of situs inversus totalis, dextrocardia, trabecular muscular interventricular shunt extending to the outflow tract of the right ventricle was confirmed during surgery.

OPERATION

Median transsternal thoracotomy was performed with the surgeon to the left of the patient to establish conventional cardiopulmonary bypass and perform the operation. The procedure was performed under hypothermia of 28°C with 20-mL/kg anterograde sanguineous cardioplegia at 4°C every 15 minutes. The perfusion time was 72 minutes and myocardial ischemia time was 57 minutes. The pulmonary trunk was opened above the pulmonary valve and through this incision the interventricular shunt was closed with a bovine pericardial patch fixed with individual 6-0 polypropylene thread sutures which were anchored using small bovine pericardial patches (Figure 3). In the postoperative period the patient evolved with unspecific pulmonary infection and received broad-spectrum antibiotic therapy. The infant developed renal insufficiency requiring peritoneal dialysis during 3 days. She was released from hospital after 21 days of hospitalization in a good general state and after an echocardiogram demonstrated correction of the defect.

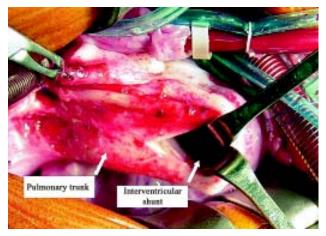


Fig. 3 – View of the interventricular shunt through the lengthwise incision in the pulmonary trunk