

## Clinical-Surgical Correlation

### Case 6/2004 – Pediatric Heart Surgery Service – Hospital de Base, Medical School, São José do Rio Preto

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#### CLINICAL DATA

A 5-month-old male child born by normal delivery is reported. The 3325-g child originates from Limeira, São Paulo, Brazil. In the nursery of the hospital he already presented with respiratory difficulties and cyanosis during breastfeeding, which continued in subsequent months. During hospitalization for the treatment of pneumonia, a cardiac murmur was observed and the patient was referred to specialized treatment.

During a physical examination the patient was seen to be in a moderate general state, ruddy, afebrile, dehydrated, cyanotic and agitated. The heart was rhythmic, hyperphonic with a second noise. The boy had a regurgitant systolic murmur at ++/6 focused on the medium left sternal margin and ejective at the right sternal margin. The lungs had a symmetrical vesicular murmur with a bilateral rumble. The abdomen was rounded and the liver was positioned 3 cm from the right edge of the ribcage. The peripheral pulses



Fig. 1 - Intra-operative aspect showing the anterior aorta with the principal left coronary artery deriving from the coronary sinus 1. The pulmonary branch is behind and much dilated.

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were palpable, although reduced in the lower limbs and the left upper limb. The peripheral saturation was 60%.

#### ELECTROCARDIOGRAM

The electrocardiogram evidenced sinus rhythm with the axis of the QRS complex + 90°. The P wave was positive at D1 and the AVF spiked with an overload of the right atrial. The P-R interval was 0.24. The QRS presented V1 with RR', denoting a partial block of the right branch. V5 and V6 presented with wide R and V1 with deep S, indicating great overload of the left ventricle.

#### RADIOGRAM

The cardiothoracic index was 0.64. Levocardia was evidenced with an increase in the cardiac area affecting the left ventricle and pulmonary vascular prominence was seen.

#### ECHOCARDIOGRAM

Situs solitus was present at levocardia. Veno-atrial and atrioventricular connections were concordant and a double-outlet right ventricle type ventriculo-arterial connection was observed as was a subpulmonary perimembranous interventricular shunt of 10 mm. The aortic isthmus hyperplastic with the arch measuring 11 mm and the carotid and left subclavian arteries measured 5 mm. The aorta after the left subclavian artery was 3 mm. The foramen oval was patent. There was a gradient between the right ventricle and the pulmonary branch of 35 mmHg evidenced by Doppler echocardiography.

#### DIFFERENTIAL DIAGNOSIS

Transposition of the great vessels with an interventricular shunt, Fallot's tetralogy, truncus arteriosus, and heart diseases with a univentricular physiology should be considered.

#### DIAGNOSIS

Due to the possible associations, a cineangiographic study was performed that confirmed the diagnosis of Taussig-Bing heart, with double-outlet right ventricle, anterior aorta and posterior pulmonary ("transposition vessels"). Subpulmonary interventricular shunt was also diagnose with significant coarctation of the pre-ductal aorta with hypoplasia of the aortic arch. The pressure

measurements of the right ventricle were 110 and 15 mmHg and the pulmonary branch pressure was 70 mmHg.

#### OPERATION

Median transsternal thoracotomy was performed and cardiopulmonary bypass was established under deep hypothermia at 18 °C. Intermittent antegrade sanguineous cardioplegia at 4 °C was utilized. The arterial canal was ligated and sectioned and the subclavian and the entire hypoplastic region of the aortic arch were sectioned. The descending aorta was anastomised end-to-end to the aortic arch near to the innominate arterial branch. The right atrium was opened and the large interventricular shunt was closed via the atrium using a pericardial bovine patch. The Jatene operation [1] was initiated with the sectioning of the aorta and pulmonary branch, which was dilated and required a reduction in its size to the diameter of the neo-aorta. The left and right coronary ostia were removed and re-implanted in the neo-aorta. The Lecompte maneuver [2] was made with the reconstructed neo-aorta and neo-pulmonary artery. The foramen ovale and right atrium were closed. The thorax was closed and a catheter for peritoneal dialysis was implanted. The perfusion, myocardial ischemia and cardiac arrest times were 207, 171 and 16 minutes, respectively. In the immediate postoperative period, the patient evolved with renal insufficiency requiring peritoneal dialysis for four days. He remained in the intensive care unit for 10 days and was released from hospital on the 23<sup>rd</sup> day. Six months after the operation, an echocardiogram demonstrated absence of residual defects> At this time the patient was functional class 1 (NYHA) and only taking diuretic drugs.

#### BIBLIOGRAPHIC REFERENCES

1. Jatene AD, Fontes VF, Paulista PP, Souza LC, Neger F, Galantier M et al. Anatomic correction of transposition of great vessels. *J Thorac Cardiovasc Surg* 1976;72:364-70.
2. Lecompte Y, Neveux JY, Leca F, Zannini L, Tu TV, Dubois Y et al. Reconstruction of the pulmonary outflow tract without prosthetic conduit. *J Thorac Cardiovasc Surg* 1982;84:727-33.