Braz J Cardiovasc Surg 2004; 19(3): 323-324

Clinical-Surgical Correlation

Case 5/2004 – Pediatric Heart Surgery Service – Hospital de Base, Medical School, São José do Rio Preto

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RBCCV 44205-702

CLINICAL DATA

A four and a half-year-old white male patient weighing 14 kg was admitted for a physical examination. The patient complained of tiredness after moderate effort over the previous seven months and had been hospitalized with pneumonia on two occasions. He was in a good general state with a rosy complexion and acyanotic. The thorax presented with left anterior bulging, palpable ictus cordis with fremitis at the fifth intercostal space, fixed split S2, systolic murmur of +++/6, ejective at the left sternal border. The lungs had a symmetrical vesicular murmur without adventitious sounds. No alterations were evidenced in the abdomen and the peripheral pulses were palpable and symmetrical.

ELECTROCARDIOGRAM

The electrocardiogram evidenced sinusal rhythm with a heart rate of 145 beats/minute and electrical axis of the QRS complex

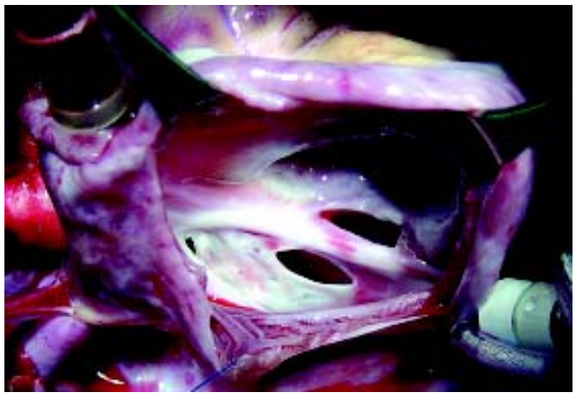

Fig. 1 - Right atriotomy where three orifices can be seen. Two are on the fossa ovalis (ostium secundum-type interatrial shunt). The other is near the right atrioventricular valve corresponding to the unroofed coronary sinus.

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Article received on June, 2004
Article accepted on July, 2004

+ 120º. An ample R-wave at V1 and a great P-wave at D2 were observed suggesting right atrial and ventricular overload.

RADIOGRAM
The cardiothoracic index was 0.47 without evidence of increased heart chambers. A slight pulmonary vascular prominence was seen.

ECHOCARDIOGRAM
Situs solitus was seen at levocardia. The veno-atrial, atrioventricular and ventriculoarterial connections were concordant. There were two interatrial shunts, one, an ostium secundum type was 6 mm in diameter and the other was in the coronary sinus region with 4 mm. There was also a slight reduction in the caliber of the pulmonary branch immediately above the valve. A Doppler echocardiogram demonstrated a turbulent, accelerated blood flow with a maximum gradient of 33 mmHg.

DIFFERENTIAL DIAGNOSIS
The electrocardiogram was compatible with pulmonary stenosis. The radiogram suggested normality or some congenital heart disease with hyper flow however this did not cause any great hemodynamic effect, similar to an interatrial shunt, partial anomalous drainage of the pulmonary veins or partial defect of the atrioventricular septum.

DIAGNOSIS
After identifying the interatrial shunt in the region of the coronary sinus and pulmonary stenosis, catheterization was used to further study the case. Although catheterization did not determine the exact location of the interatrial shunts, it demonstrated a significant shunt between the coronary sinus and the left atrium. This confirmed the suspected diagnosis of unroofed coronary sinus syndrome with an ostium secundum-type interatrial shunt and the presence of supravalvar pulmonary stenosis with a gradient of 30 mmHg.

OPERATION
Median transsternal thoracotomy was performed and a conventional cardiopulmonary bypass was established. The patient was operated on under moderate hypothermia of 28 ºC using intermittent anterograde sanguineous cardioplegia at 4 ºC. The right atrium was opened and the ostium secundum-type interatrial shunt was identified with two fenestrations. In the region of the coronary sinus there was also a substantial shunt between the right atrium and the left atrium. During cardioplegia it was possible to observe drainage into the left ventricle. These two orifices were occluded using bovine pericardial patches with running sutures of 5-0 polypropylene thread. This isolated the left and right atria and leaving the drainage of the coronary sinus to the left atrium. The pulmonary branch was longitudinally opened but supravalvar stenosis was not evidenced, however the pulmonary valve was bivalved and was submitted to a commissurotomy. The patient was released from hospital on the sixth postoperative day using diuretic and corticosteroid drugs owing to pericarditis. An echocardiogram after 90 days demonstrated the absence of the interatrial shunts and a pulmonary transvalvar gradient of 15 mmHg without valvar or supravalvar stenosis.