Clinical-Surgical Correlation

Case 1/2004 – Pediatric cardiology department, Hospital de Base, Medical School of São José do Rio Preto

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CLINICAL DATA
The patient is a pre-term male Caucasian of 36 weeks of gestation weighing 2.61 kg. Since the second day of life he presented with episodes of cyanosis, tachydyspnea and hypoglycemia, with suspicions of pneumonia. Antibiotic therapy was initiated. The patient had a good general state but was discolored (+/4). He had rhythmic noise, a systolic murmur of +++/6 at the left sternal margin and lungs with diffuse stertor crackles. The liver was 2 cm from the right costal edge. His peripheral pulses were palpable in the four limbs, however the pulse was stronger in the right arm. An infusion of prostaglandin E1 was initiated giving an improvement of the congestive heart insufficiency, diuresis and peripheral saturation.

ELECTROCARDIOGRAM
Sinusal rhythm was recorded with 155 beats per minute. Electrical axis of the QRS + 160º was seen with a broad R wave at V1 and good P wave at D2 suggesting right atrial and ventricle overloads.

RADIOGRAM
Bilateral diffused pulmonary a hypotransparent area was evidenced suggestive of pulmonary venous congestion due to hypertension. The cardiothoracic index was 0.67.

ECHOCARDIOGRAM
Situs solitus at levocardia was seen and the veno-atrial, atrioventricular and ventriculoarterial connections were in accordance. An interruption of the aortic arch after the left common carotid artery was identified, as was a foramen ovale type interatrial communication. The interventricular communication was dually related. There was a patent but
restricted arterial canal supplying the left subclavian artery and descending aorta.

DIFFERENTIAL DIAGNOSIS
Truncus arteriosus, total anomalous connection of the pulmonary veins, severe coarctation of the aorta with interventricular communication, aorta-pulmonary window and complex heart diseases with single ventricles should be the main considerations.

DIAGNOSIS
A cineangiocardio graphic study was not necessary and could even be a disadvantage due to the use of contrast. The echocardiogram clearly demonstrated the anatomy of the aortic arch and the intracardiac characteristics, confirming the diagnosis of a type-B aortic arch interruption with an interventricular communication.

SURGERY
On the 21st day of life the patient underwent an operation to totally correct the anomaly in a single surgery. Median transsternal thoracotomy with the establishment of a cardiopulmonary bypass with two arterial and two venous cannulae was performed. After deep hypothermia at 16 ºC and intermittent anterograde sanguineous cardioplegia at 4 ºC were employed. Hypoflow at 40 mL/kg/h during 25 minutes was chosen with a total perfusion time of 164 minutes and anoxia of 47 minutes was recorded. Initially the arterial canal was ligated and sectioned with ample sectioning of the great vessels, aortic arch and the descending aorta. End-to-side anastomosis of the descending aorta on the posterior wall of the aortic arch was performed. A ventriculoseptoplasty was easily achieved through the pulmonary branch. The interatrial communication was closed by direct suturing. In the postoperative period the patient evolved with renal insufficiency and required peritoneal dialysis during four days. An unspecific infectious process was treated with broad-ranging antibiotic therapy. Episodes of hypoglycemia in spite of an endocrinological investigation not demonstrating alterations of the metabolism called our attention. The patient was released from hospital on the 22nd postoperative with a control echocardiogram demonstrating the absence of a gradient in the region of the aortic anastomosis and the interventricular communication closed.