Case 2/2012 – Eleven-Day-Old Infant with Marked Coarctation of the Aorta in Cardiogenic Shock

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Clinical data: Slight tiredness had been noticed since birth, with obvious aggravation in the last two days, which prompted the hospital admission. Weight at birth had been 2700 g after an uneventful pregnancy and normal delivery.

At physical examination: poor general status, tachydyspneic, pale, weak pulses, especially in the lower limbs. Weight: 2500 g; Height: 48 cm; BP-RSL: 80/40 mm Hg, BP-RIL: 40 mm Hg, HR: 166 bpm, RR: 100 rpm. The aorta was not palpable at the sternal notch. There were mild precordial systolic impulses in the left sternal border and the apical impulse was not palpable. The heart sounds were very loud and there were no heart murmurs. The liver was palpable 4 cm from the right costal margin and the lungs were clear, with no adventitious sounds.

Complementary Examinations:

Electrocardiogram: showed sinus rhythm, signs of exclusive right ventricular overload (Rs morphology in V1 and rS in V6) and alterations in ventricular repolarization. AQRS:

-170º, AP: +60º; AT: indeterminate (Figure 1).

Chest X-ray: showed markedly enlarged cardiac silhouette at the expense of the left ventricular arch and congested pulmonary vascular network (Figure 2).

Echocardiography: disclosed the critical elements for the diagnosis of coarctation of the aorta in the isthmic region, with a pressure gradient of 40 mm Hg and small *ductus arteriosus* (Figure 1).

Clinical diagnosis: Coarctation of the Aorta in Marked Heart Failure

Clinical Rationale: Clinical findings are consistent with the diagnosis of coarctation of the aorta, considering the difference of peripheral pulses, which albeit slight, showed severe heart failure. Right ventricular overload on electrocardiogram and marked cardiomegaly on chest x-ray supported the diagnosis. In fact, this image was compatible with the diagnosis of acyanotic congenital heart disease with heart failure due to obstruction of the left heart, also considering the lung congestion, along with cardiomegaly.

Differential diagnosis: other obstructive left heart diseases are not accompanied by large cardiomegaly as it occurs in mitral stenosis and *cor triatriatum*. Heart diseases with volume overload of the left heart, such as mitral regurgitation, could also be identified as similar, but with a more pronounced increase of the left atrium. Herat diseases with volume overload and left-to-right shunt are accompanied by pulmonary arterial enlargement.

Clinical conduct and Evolution: After endotracheal intubation and mechanical ventilation with FiO2 at 40% and introduction of intravenous vasoactive (dobutamine = 20 mcg/kg/min) and vasodilator (prostaglandin E1 = 0.08 mcg/kg/min) drugs, plus furosemide, there was improvement in the clinical condition.

Lactic acid decreased from 120 to 20 mg/dL; the metabolic acidosis, which was -23 mmol/L decreased to normal levels, with increased diuresis and reduction of cardiomegaly. Three days later, the patient could be submitted to surgery, considering improvement in general status, despite the greater increase in creatinine levels from 1.3 to 2.6 mg/dL and urea from 48 to 73 mg/dL. During surgery, coarctectomy was performed in the accentuated narrowing in the isthmic region with termino-terminal anastomosis and ligation of a large ductus arteriosus.

Evolution was favorable, with a clear regression of heart failure and endotracheal extubation on the first postoperative day, progressive decrease in dobutamine doses in three days as well as signs of renal failure and paradoxical arterial hypertension (treated with sodium nitroprusside and amlodipine). The patient was discharged after 15 days on digoxin. At the evolution, nine months later, normal weight gain (9 kg) was observed, with no murmurs or signs of heart failure and normalization of arterial pulses and cardiac silhouette. The echocardiogram showed pressure gradient in the aorta of 28 mm Hg, 82% of ventricular function and normal-sized cardiac cavities.

Comments: coarctation of the aorta has an unfavorable evolution when it is already far-reaching in the neonatal period, usually in the first two weeks of life. The initial picture can even be mistaken by cardiomyopathy with severe ventricular dysfunction, without murmurs and without the classic signs of the disease. The contrasting pulses between limbs, characteristic of coarctation of the aorta, are demonstrated in the presence of more preserved ventricular function, obtained after appropriate initial treatment. It is also interesting to note the marked right ventricular overload on

Keywords

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the ECG, due to retrograde pulmonary hypertension in the presence of sudden left ventricular overload, after greater narrowing of the ductus arteriosus. The myocardial failure, however, improves after myocardial dilation with PGE1 and even further after surgical correction. The final result is surprising, as demonstrated in this case.

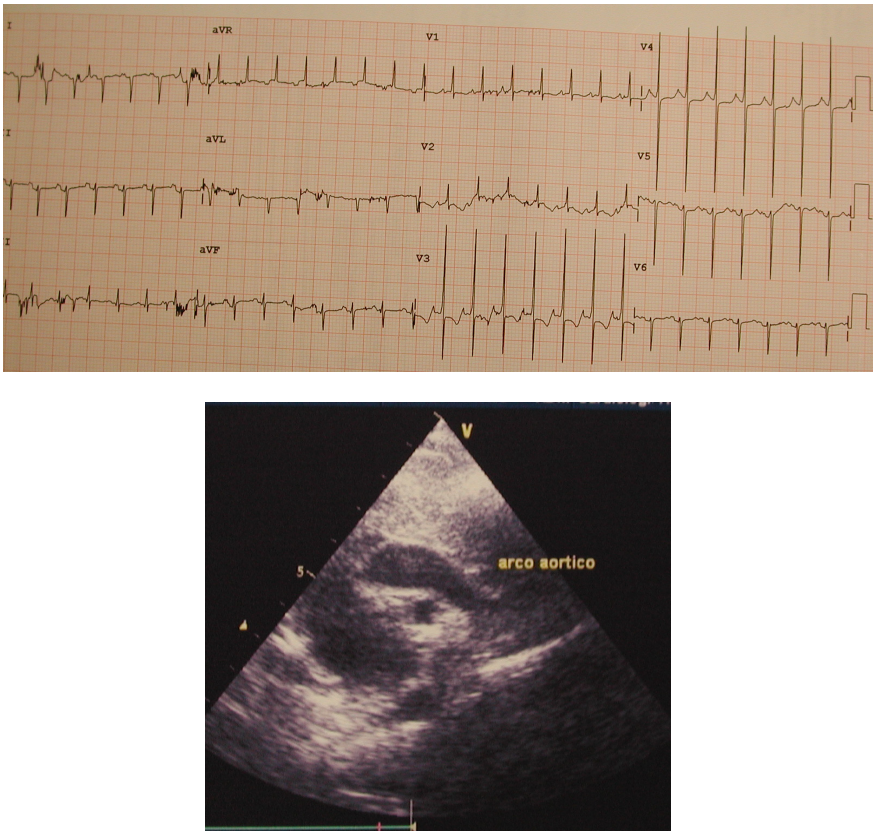

Figure 1 - *Electrocardiogram highlights signs of marked right ventricular overload in the preoperative period and the echocardiogram shows a clear obstruction in the isthmic region.*

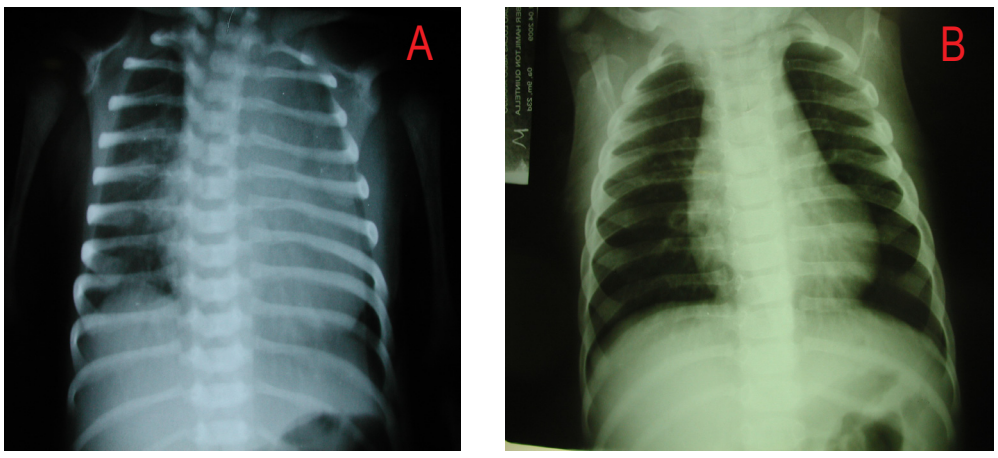

Figure 2 - *Chest X-rays in the preoperative period in A and nine months after surgery in B highlight the clear difference in heart size.*