Case 6/2010 - 14 Months Female Infant with Double Aortic Isthmus Coarctation

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Clinical data
The patient remained asymptomatic with good height and weight development since the discovery of congenital heart disease, with 12 days of life, in the presence of neonatal infection.

On physical examination, the patient presented good general condition, eupneic and blushed, with large pulses in upper limbs and missing pulses in lower limbs. The patient weighed 9,150 g, right upper limb average blood pressure: 120/80 mmHg, right lower limb average blood pressure: 80 mm Hg, Heart rate: 120 bpm. The aorta was not palpable at suprasternal notch.

In the precordium, there were no impulses in the left sternal edge and ictus was not palpable. The heart sounds were normal and there was a mild systolic murmur of greater intensity in the aortic area without irradiation. Liver could not be felt.

Electrocardiogram revealed sinus rhythm and there were no atrial and ventricular overloads. The QRS complex showed an rS morphology in V1, qRs in V6 and normal ventricular repolarization. AQRS was at +90 degrees, AP at +60° and AT at +50°.

Radiographic image
The image shows a slightly increased cardiac area due to rounded ventricular arch with normal pulmonary vasculature and a slightly increased vascular pedicle (Figure 1).

Diagnosis impression
The image is consistent with the diagnosis of heart disease that is accompanied by obstruction of blood flow on the left side of the heart. The greater prominence of the vascular pedicle may influence the presence of a dilated aorta, suggesting the diagnosis of aortic coarctation.

Differential diagnosis
All other acyanogenic congenital heart diseases with obstruction of blood flow should be considered, such as aortic stenosis and even those on the right side, such as pulmonary stenosis, even in the absence of middle arch dilation.

Diagnosis confirmation
Clinical findings were decisive for the diagnosis of aortic coarctation. Echocardiogram confirmed the existence of vascular defect in the isthmus, 3 mm in diameter. Magnetic resonance imaging revealed the presence of two strictures in this region. The first one, right after the emergence of the left subclavian artery and the second one right after mild dilation of the descending aorta (Figure 2).

Conduct
Upon the surgery, through posterior lateral-left incision, two constrictions at the aortic isthmus were identified, the first at the ductus arteriosus, and the other after the emergence of left subclavian artery. Resection of this region through termino-terminal anastomosis with the arch, enabled a proper diameter of the aorta. As a result, there was resolution of the aortic obstruction condition after control of paradoxical hypertension using sodium nitroprusside, amlodipine, propranolol, and captopril.

Comments
In this case of aortic isthmus coarctation, presence of two sharp and subsequent strictures with tissue dilatation between them draw attention. These constrictions may have been formed by prominent ductal tissue with greater preservation among them. Such anatomic diversity is unusual in aortic isthmus coarctation. There was a false impression of hypoplastic aortic arch evidenced by magnetic resonance imaging.
Figure 1 - Chest radiography reveals normal cardiac area and pulmonary vasculature close to normal. The rounded ventricular arch directs myocardial hypertrophy. Slightly broader vascular pedicle at right.

Figure 2 - Magnetic resonance imaging shows aortic isthmus coarctation, after the emergence of the left subclavian artery with two adjacent strictures (arrows). The impression of aortic arch hypoplasia was false.