Case 1/2013 – 20-Year-Old Man with Native and Evolutionary Coarctation in Aortic Arch

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Clinical data: After correction of severe aortic coarctation (AoC) in the isthmus through the end-to-end technique and closing of large Ventricular Septal Defect (VSD) with 18 days of life, in addition to correction of moderate subaortic stenosis with intraventricular pressure gradient of 40 mmHg with 3 years of age, a good clinical outcome was later observed. The patient currently tolerates routine exercising and does not refer any symptoms. Over time, he maintained upper and lower limb gradient between 10 and 15 mmHg and discreet systolic murmur in the aortic area by left ventricular outflow tract gradient around 15 mmHg. In the last routine evaluation, blood pressure in the right upper limb was 140 x 70 mmHg, and in the left upper and lower limbs, the systolic pressure was 90 mmHg, which characterized diagnosis of aortic arch obstruction between the upper limbs.

Physical examination: Good general condition, eupneic, acyanotic, contrasting pulses between the right upper limb and the other ends. Weight: 65 Kg. Height: 165 cm, average systolic and diastolic blood pressure: 140/70 mmHg, PAMSE = PAMID = 90 mmHg, HR: 78 bpm. The aorta was clearly palpable palpable at the suprasternal notch with systolic fremitus and murmur of ++ intensity.

In the precordium, apical impulse was not palpable and there were no systolic impulses. The heart sounds were normal and moderate systolic murmur of ++ intensity was harsh across the left sternal border, accompanied by fremitus. The liver was not palpable and the lungs were clear.

Complementary tests

Electrocardiogram showed sinus rhythm, signs of complete right bundle branch block and anterior superior division block of the left branch (unchanged since the neonatal correction of aortic coarctation and VSD). AP: +50º, AQRS: +245º, AT: +12º.

Chest radiograph showed normal and globular heart size on myocardial hypertrophy and normal pulmonary vasculature (Figure 1).

Echocardiogram (Figure 1) showed cardiac cavities with normal dimensions, discrete myocardial hypertrophy (septum = posterior wall = 10 mm), left intraventricular pressure gradient of 15 mmHg in the left outflow tract and 58 mmHg in the aortic arch.

CT of thoracic aorta showed three-level aortic occlusion. In the ascending aorta (15.9 x 18.5 mm), the arch after the emergence of the left carotid artery (11 x 14.6 mm) and in the isthmus after the left subclavian artery at the level of the previous correction of aortic coarctation (15.1 x 15.1 mm) compared to normal diameters of 24.9 x 23.9 at the end of the ascending aorta and 20.5 x 19.8 mm in the descending aorta (fig. 2).

Clinical diagnosis: Coarctation in the aortic arch in natural evolution coupled with discrete recoarctation of the aorta in the isthmus, stenosis of the ascending aorta by prior surgical incision and discrete subaortic restenosis.

Clinical reasoning: Evolutionary clinical findings were compatible with the diagnosis of aortic coarctation in the arch between the upper limbs through the demonstration of the clear pressure gradient between them. The absence of symptoms, with good physical tolerance and no signs of heart failure were also compatible with a favorable dynamic situation. Cardiac auscultation was conducive to the diagnosis of associated subaortic restenosis. The absence of a gradient between the left upper limb and the lower limbs predicted slight narrowing of the aortic isthmus. These elements were confirmed by the echocardiography images, especially by chest CT.

Differential diagnosis: Other diseases that are accompanied by aortic obstruction at different levels should be recalled, especially vasculopathies such as Kawasaki's disease and Takayasu's disease. However, they are accompanied by inflammations early in life.

Conduct: Given the greater impact of aortic arch obstruction, presence of systolic hypertension and myocardial hypertrophy, hence the need for surgical correction, it was thought that the most appropriate procedure was redirecting blood by placing a plastic tube between the ascending aorta and the descending aorta in order to better adjust the systemic circulation.

Comments: The evolutionary aortic arch coarctation, even in the long term follow-up post-correction of the aortic coarctation in the usual area after the emergence of the left subclavian artery, has been observed at a rate of 10% to 20% of these cases. Considering that, the aortic coarctation correction at an early age has been extended to the aortic arch on a technique known as "extended end-to-end anastomosis" of the aorta. The diagnosis of aortic arch coarctation is easily determined later during evolution, provided that there is medical concern of the blood pressure measurement performed systematically in all four limbs. It is worth noting that the narrowing of the aortic arch becomes sharper during the late phase, and there are no previous parameters to establish its earlier diagnosis with precision. Hence, in suspected cases, the correction extended to the arch becomes the method of choice.

Keywords

Coarctation of the aorta, aortic arch coarctation.

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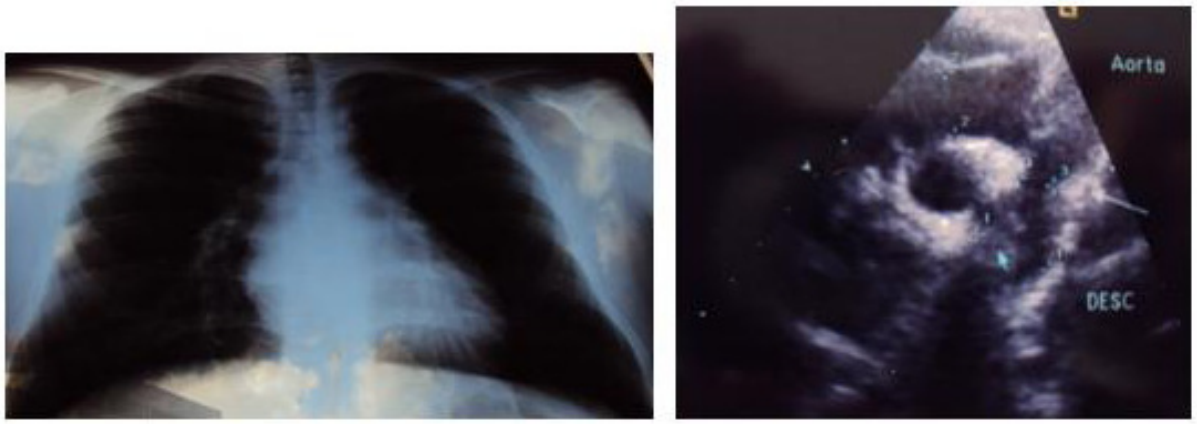

Figure 1 – *Chest radiograph shows normal cardiac area and echocardiography stresses the sign of coarctation of the aorta in the aortic arch (longer arrow) after the emergence of the left carotid artery and discrete narrowing in the isthmus (shorter arrow).*

Figure 2 – *Chest CT clearly highlights the obstructions in the aorta: in the ascending aorta, due to residual lesion of previous incision for correction of subaortic stenosis, in the aortic arch (between the left carotid and subclavian arteries) and in the isthmus.*