Coarctation of the aorta – a case report

Vanda Areias, Ália Givá, José Arduán, Catarina Mendonça, Pastor Santos e Silva

Abstract

The authors report the case of a 15-years-old male football player, referred to the outpatient hypertension clinic for investigation. The clinical manifestations included headache, epistaxis and dizziness evolving for 5 years.

The physical examination showed values of blood pressure in the upper limbs higher than percentile 99; the difference in blood pressure between the upper and lower limbs was over 40 mmHg, femoral pulses were narrow, symmetrical and with femoral delay; precordial systolic murmur II / VI radiating to the back.

According to the clinical data, the most probably diagnosis was aorta coarctation. A thorax angio-CT scan was performed, confirming our clinical suspicion. The patient was referred to cardio-thoracic surgery and dilatation with stent by percutaneous catheter was carried out.

Key words: Hypertension, teenager, aorta coarctation.

INTRODUCTION

High blood pressure (HBP) in the pediatric population is defined as a systolic and/or diastolic blood pressure (BP) higher than or equal to the 95th percentile, according to charts adjusted for age, gender and height, on three occasions. Pre-HBP is defined as systolic and/or diastolic blood pressure higher than or equal to the 90th percentile and lower than the 95th percentile.[1]

In children over three years, BP should be regularly taken in the routine consultations. High BP in childhood is a risk factor of early HBP in adulthood. Primary HBP in children is often associated with excessive weight and a family history of HBP or cardiovascular disease. Secondary HBP is more common in children than in adults, and the younger the patient and the more serious the HBP, the higher the likelihood that it will to occur. In adolescents, the main causes of secondary HBP are renal parenchymal disease and coarctation of the aorta. A thorough clinical history and objective examination are essential in the initial evaluation of a patient with persistently high BP.[1-4]

Medicine Service I, Hospital de Faro, EPE

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Coarctation of the aorta is a narrowing of the aorta, most frequently occurring in the thoracic aorta distal to the origin of the left subclavian artery. Nevertheless, it can occur proximal to the left subclavian artery, and in rare cases, occurs in the abdominal aorta.

This entity accounts for around 6% to 8% of congenital cardiopathies, and is a rare cause of HBP. It is two to five times more common in males and seven times more common in Caucasians than in Asians. Most cases occur sporadically, and familial occurrence is rare. In approximately 50% of patients, heart changes are observed, particularly bicuspid aortic valve (25-85%), ventricular septal defect, left sided heart obstructive lesions or hypoplastic left heart syndrome, and aortic arch hypoplasia. In 25% of patients, extra-cardiac changes are observed, in the musculoskeletal, genitourinary, gastrointestinal or urinary systems. The prevalence of coarctation of the aorta is higher in some diseases, like Turner Syndrome (35% of patients).[5,6]

CLINICAL CASE

A male patient, fifteen years of age, Caucasian, born in Lisbon, a student and registered footballer, was referred to us for investigation of HBP. The patient reported frontoparietal headaches, dizziness, frequent epistaxis and roncopathy with five years of evolution. The patient did not report any other symptoms.

Personal history included allergy to dust mites and adenoidectomy at the age of eight; the patient reported that he did not smoke or drink, and did not use drugs. His diet was balanced and varied. Family history included a grandmother on the mother's side with HBP since the age of fifty.

On objective examination, the patient had good

general condition, normal color and hydrated mucosa; weight was 58kg, height was 165 cm (25th percentile), body mass index was 21.5 Kg/m^2; fundoscopy presented no alterations. The following BP were recorded: right upper limb - 144/75 mmHg (higher than the 99th percentile); left upper limb - 145/62 mmHg (higher than the 99th percentile); right lower limb - 106/42 mmHg (50th percentile); left lower limb - 102/59 mmHg (50th percentile); the difference between BP in the upper limbs and lower limbs was higher than 40 mmHg. Heart auscultation showed a heart rate of 62 beats per minute, as well as grade II/VI pan-systolic murmur radiating to the back; weak, symmetrical and delayed femoral pulses; and abdomen presenting no masses, enlarged organs, or murmurs.

When the patient arrived at the clinic for investigation of HBP/ CVRF, he had already undergone additional diagnostic tests and analyses (Table 1) that did not show any changes. Chest X-ray showed no changes; electrocardiogram revealed sinus rhythm, 55 bpm and isolated supraventricular extrasystoles; ambulatory blood pressure measurement (ABPM) measured mean, systolic and diastolic, daytime and night-time blood pressure were 143/78 mmHg and 122/61 mmHg, respectively, with BP values above normal during the practice of sports; echocardiogram did not show any changes; renal and suprarenal echography and Echo Doppler of the renal arteries, and abdominal, renal and suprarenal CT scan did not show any changes.

A hypothesis of coarctation of the aorta was proposed. CT Angiography of the chest was requested, which revealed stenosis in the aortic arch at its transition into the descending aorta, with a diameter of only 7.68 mm, confirming the presence of this change. The aortic outflow tract measured around 19.59 mm and at the aortic arch, below the coarctation, the aorta measured 16 mm *(Fig. 1)*.

The patient was referred to cardio-thoracic surgery and dilation was performed with percutaneous catheter implantation of a stent.

TABLE I

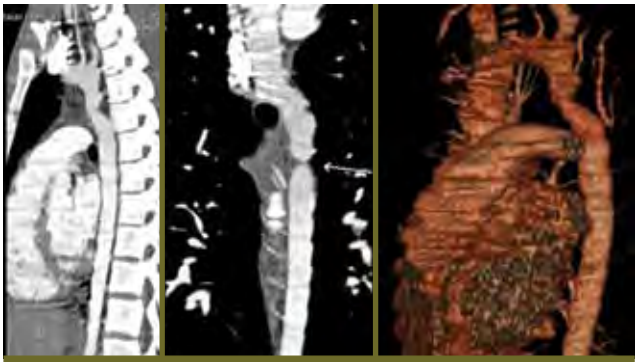
Analyses

Hb (g/L)	151
Leukocytes (cells/µL)	4.800
Glucose (mg/dL)	89
BUN (mg/dL)	18
Creatinine (mg/dL)	1,0
Ionogram	Normal
Potassium (mmol/l)	4,5
Chlorine (mmol/l)	92,6
Total cholesterol (mg/dl)	162
Triglycerides (mg/dl)	65
ALT (IU/L)	21
ALT (IU/L)	14
g-GT (IU/L)	19
TSH (µIU/mL)	2,566
Cortisol (µg/dL)	15,9
Aldosterone (µg/dL)	26
24-hour Urine (1,200 mL): • Creatinine clearance 90 mL/min • Normal urinary ionogram • Microalbuminuria 0.2 mg/L • Normal aldosterone, active renin, cortisol and 17-hydroxycorticosteroids. Normal metanephrine, normetanephrine and vanillylmandelic acid.	

DISCUSSION

The clinical manifestations of coarctation of the aorta vary according to age, location and severity of the coarctation. In the first years of life, depending on the severity of the obstruction and the associated heart lesions, patients can have congestive heart failure, severe acidosis or poor perfusion of the lower limbs. During childhood, patients are usually asymptomatic, and can have HBP, headaches, epistaxis, claudication of the lower limbs, muscular weakness, coldness of feet and hands, or neurological changes. Bearing in mind that the most common location is distal to the origin of the left subclavian artery, the traditional clinical changes are: HBP in the upper limbs, weakened or delayed femoral pulses and reduced BP in the lower limbs.[5]

Even though coarctation of the aorta has a clinical diagnosis (the difference in BP and delayed pulses are pathognomonic), echocardiogram tests enable us to confirm the diagnosis and detect associated heart defects; NMR and CT Angiography of the chest are more sensitive tests to detect the location and extent of the coarctation.[5]



CT Angiography of the thorax.

FIG. 1

The severity of HBP does not depend only on the BP values, but also on the presence of other cardiovascular risk factors, lesions of the target organ, and cardiovascular and renal complications. Taking these factors into account, the risk for these patients is classified as low, moderate, high and extremely high, in comparison with normotensive, healthy individuals with no risk factors.[6,7]

Patients with congenital heart disease who engage in competitive sports may be exposed to high levels of physical and mental stress, presenting a higher risk of sudden cardiac death or clinical aggravation, compared with healthy patients. Little is mentioned in the literature about physical exercises and congenital heart disease; therefore, a restrictive approach seems to be reasonable. The recommendations for athletes with heart pathology engaging in competitive sports are based on the results of the risk assessment and classification. Dynamic exercises are preferable to static exercises, but only in low to moderate- intensity training sessions.[6,7]

This patient was a registered footballer, engaged in dynamic, high-intensity training, and who had been indicated to give up this type of training and maintain only physical exercises of lower intensity (low or moderate) and avoid contact sports. The practice of isometric high-intensity, prolonged training poses a higher risk of dissection, even in cases of correction of the coarctation.[8]

Surgery was initially the only available treatment, the first having been performed in 1944. With this therapy, surgical mortality has increased with age, and can reach 4.5% in individuals aged over 30 years. Later, balloon angioplasty was developed (in 1982), which was preferable to surgery in very localized coarctation. Endovascular treatment with insertion of a stent has showed good results in the short and medium terms, compared with balloon angioplasty; it reduces the re-incidence of stenosis and aneurysm formation, and maintains the haemodynamic benefits. Endovascular treatment is currently the therapy of choice in patients with recurrent coarctation, if the anatomical location of the coarctation permits.[8]

Consensus has not yet been reached in the literature regarding the best therapeutic option - surgery or angioplasty - for the correction of coarctation of the aorta. The patient's age, whether it is a first-time or recurrent coarctation, and the presence of associated lesions should be considered, in addition to the experience of each department.

Patients who are not treated for coarctation of the aorta may live to 35 years of age; less than 20% live to 50 years of age.[8]

If the coarctation is treated when patient is under 14 years of age, the survival rate after 20 years is 91%. If the coarctation is treated when patients are over 14 years of age, the survival rate after 20 years is 79%.[8]

After correction of the coarctation of the aorta, 97%-98% remain in class I of the New York Heart Association (NYHA). The increase in pressure on the site of coarctation during exercise may result in hypertrophy of the myocardium, with changes in the left ventricular diastolic function. Nevertheless, the ventricular systolic function is normal or hyperdynamic.

The recurrence of HBP is common despite successful correction of coarctation of the aorta, being the main risk factor for the occurrence of cerebrovascular disease, aortic rupture, heart insufficiency and progression of coronary disease in the postoperative period. Nevertheless, most patients become normotensive after surgery, and HBP reoccurs only much later. The prevalence of late HBP ranges from 30% to 75% in various studies.[9] The prevalence depends on the age at which the patient underwent surgery. Therefore, for patients who undergo surgery before 1 year of age, the prevalence of late HBP is 7%, while for patients over 14 years, the prevalence is 33%.[9] The cause of late HBP is still unknown, but several theories have been proposed, including decreased aortic compliance, changed baroreceptor functions and neuroendocrine activation. Exercise-induced HBP is common, and an association with increased

exercise-induced pressure in the upper and lower limbs has been found, which may result in a residual, mild coarctation or changed arterial compliance. The relationship of these observations to the onset of late HBP and long-term prognosis remains unclear.[8,9]

Coronary disease is one of the long-term complications described. Lawrie et al claim that two thirds of subsequent deaths are caused by acute myocardial infarction.[9] In a study carried out by Cohen, this rate is 37%. The data obtained by Cohen indicates that deaths due to coronary disease are related to the duration of HBP in the pre-operative period and to the recurrence or persistence of HBP in the postoperative period.

Another complication that has been described are cerebrovascular accidents. Ischemic vascular accidents probably result from early carotid artery disease associated with pre- and postoperative hypertension, while hemorrhagic events can be a consequence of HBP persistence in the postoperative period and existence of berry aneurysm. Their incidences are reportedly different in the different case series, between 0 and 11.5%, as a cause of postoperative death.[9]

Despite anatomical reconstruction, complications like recoarctation, formation of aneurysms and aortic rupture may occur during follow-up, and all patients remain at risk of infective endarteritis. Identifying these complications can be difficult, but echocardiogram, CT scanning and NMR have a crucial role.

In a follow-up study carried out by Koller *et al*, recoarctation occurred in 10.8% of children who underwent surgery before the age of two; in older children this figure was 3.1%, and there were no cases in adults.[9] These results were confirmed by other studies. The occurrence of recoarctation depends not only on the patient's age at the time of the surgery, but also the type of surgery; angioplasty is more common than top end anastomosis (35% *versus* 8%).[9]

The formation of aneurysms is a serious complication that can occur not only at the site of the correction, but also in the interior of the proximal aorta, and is associated with a high risk of aortic rupture. Knyshov *et al* in a case series involving 891 patients followed up over 20 years, observed the formation of aneurysms in 5.4% of patients; of these, 90% had undergone aortoplasty and 37.5% died due to ruptured aneurysm or endarteritis.[9] Its diagnosis is difficult and chest X-ray should be used for screening, followed by aortography or, alternatively, NMR.

Endarteritis, which accounted for around 20% of the pre-operative deaths, is rare in operated patients. The use of prophylactic antibiotherapy before invasive interventions also contributes to this reduced rate.[9]

The bicuspid aortic valve is associated with coarctation of the aorta in 25% to 85% of patients;[8] two thirds of cases develop regurgitation and/or stenosis, and around 10% of patients will need valve replacement.[9] It is associated with risk of aneurysm and aortic dissection, regardless of the occurrence of coarctation, and is responsible for a high number of cases of heart failure, which accounts for more than 20% of subsequent deaths.

The long-term prognosis and high morbidity in these patients after surgical correction of the coarctation make long-term follow-up essential.

Chest X-ray and ECG, although not very specific, provide important information. After surgery, ECG is normal in 25%-48% of patients.[9] The changes observed include right and left bundle branch block, and changes attributable to myocardial ischemia. Chest X-ray shows cardiomegaly in approximately 20% of cases, with most of patients having HBP or aortic valve disease.[9]

Regular evaluation, aggressive treatment of HBP, and investigation of possible recoarctation are essential for high risk patients. In all consultations, the pressure gradient should be measured in the upper and lower limbs; if the gradient is > 20mmHg, an investigation of possible recoarctation should be performed, ideally using NMR. Patients with clinical evidence of aortic valve disease should undergo echocardiogram regularly and control their BP, and valve replacement should be performed according to the guidelines.[9]

It is important to have good control of the cardiovascular risk factors and lifestyle changes on the part of the patient. All patients should be aware of the need to receive prophylactic antibiotherapy, whenever they undergo dental procedures or other invasive interventions.[9] ■

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