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Case Report

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# Severe aortic coarctation in a patient with refractory hypertension

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Acronyms
**HT:** hypertension
**CoA:** coarctation of the aorta

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**ABSTRACT**
Idiopathic hypertension is common in adults, and some patients are considered resistant or refractory to treatment. In such cases it is often associated with a pathological process which hinders its control, in spite of changes in lifestyle and the proper use of drugs. This article is about an adult female patient with refractory hypertension due to aortic coarctation. CT scan and angiographic images are shown.
***Key words:*** Hypertension, Treatment, Aortic coarctation

## *Coartación aórtica crítica en paciente con hipertensión arterial refractaria*

**RESUMEN**
La hipertensión arterial idiopática en el adulto es frecuente y en algunos pacientes suele considerarse resistente o refractaria al tratamiento. En esos casos casi siempre se asocia a algún proceso patológico que dificulta su control, a pesar de los cambios en el estilo de vida y el uso adecuado de fármacos. En este artículo se presenta una paciente adulta con hipertensión refractaria debido a una coartación aórtica. Se muestran las imágenes tomográficas y angiográficas.
***Palabras clave:*** Hipertensión arterial, Tratamiento, Coartación aórtica

## INTRODUCTION

Hypertension (HT) in adults is defined taking into account the average of two or more accurate blood pressure measurements, at two or more visits, and according to the internationally accepted values that are stated in the seventh report on HT (JNC 7 Report)[1].

HT is defined as resistant or refractory to treatment when a therapeutic plan that has included lifestyle modification and the prescription of at least 3

drugs (including a diuretic), in sufficient doses, has not reduced systolic and diastolic blood pressure to the recommended values, below 140/90 mmHg. According to this definition, the prevalence of resistant hypertension is high. Some studies estimate that up to 8% of patients are prescribed four drugs or more without achieving the control of HT[2].

This article reports the case of an adult female patient with refractory hypertension due to coarctation of the aorta (CoA).

## CASE REPORT

A 25-year-old female patient, with a history of systemic hypertension for 17 years, was treated with atenolol (100 mg/day), hydrochlorothiazide (25 mg/day) and enalapril (40mg/day), to which it was added amlodipine, due to resistance to treatment in recent months.

From a clinical point of view, the patient complained of episodes of occipital headache, ringing in the ears, occasional precordial pain, plus intermittent claudication while walking. Therefore, the case was consulted with Angiology specialists, on suspicion of peripheral arterial disease.

On physical examination, slightly delayed femoral and posterior tibial pulses were found, as well as aortic systolic ejection murmur grade III/VI, without being interscapular, with marked hypertension in the upper limbs (180/100 mmHg, despite the 4 antihypertensive drugs) in sharp contrast to a normal to low blood pressure in the lower limbs, with 90/60 mmHg, and a gradient which largely exceeded 20 mmHg between upper and lower limbs.

Blood count, blood chemistry testing, electrolytes and blood gases were normal. The electrocardiogram showed no signs of hypertrophy or left ventricular systolic overload (**Figure 1**). The transthoracic echocardiography showed the presence of a continuous flow in the abdominal aorta and a moderate septal hypertrophy, which together with the clinical data, early onset and refractory nature of hypertension led to the suspicion of CoA. Therefore, a CT angiography was performed (**Figure 2**). It confirmed the diagnosis and identified the coarcted area, which was circumscribed and very narrow, with a large post-coarctation aneurysm.

Percutaneous treatment was attempted, but it was impossible to implant the stent. A virtually closed vessel was visualized in the angiography, with the above mentioned post-coarctation aneurysm (**Figure 3**). Surgical treatment was then performed, with a satisfactory outcome. Currently, the nulliparous patient, in fully fertile stage, awaits the right moment to plan her first pregnancy, with one antihypertensive drug as maintenance therapy.

## COMMENTS

The etiologic classification of refractory HT identifies

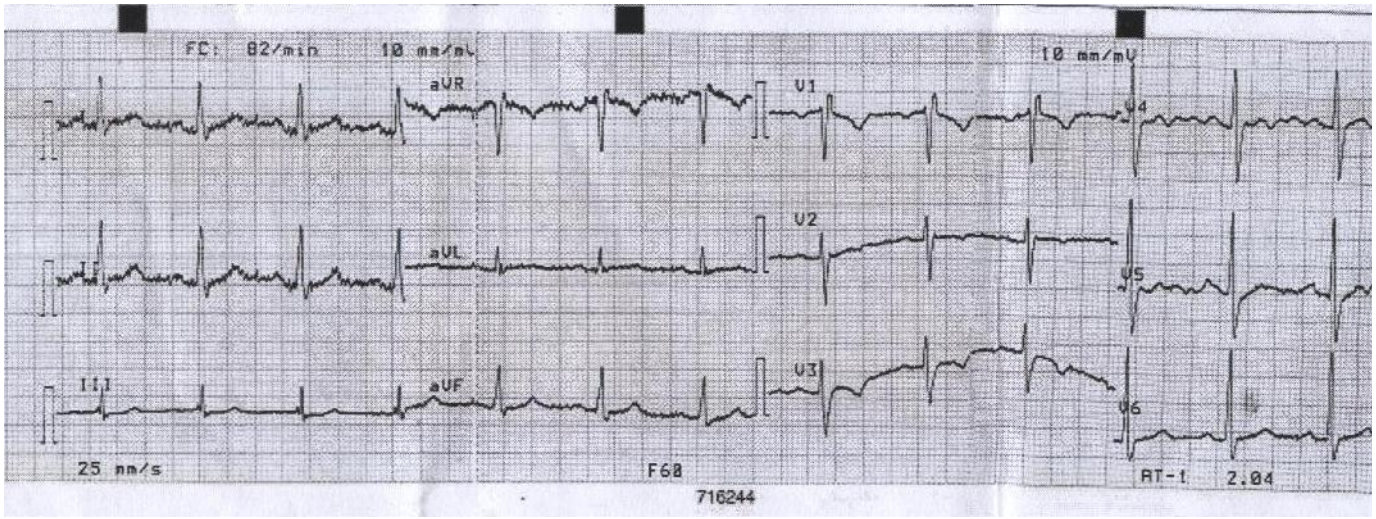


**Figure 1.** 12-lead electrocardiogram. Sinus rhythm, electrical axis and heart rate are normal. Hypertrophy and systolic overload of the left ventricle, typical signs accompanying the CoA, are not evident.

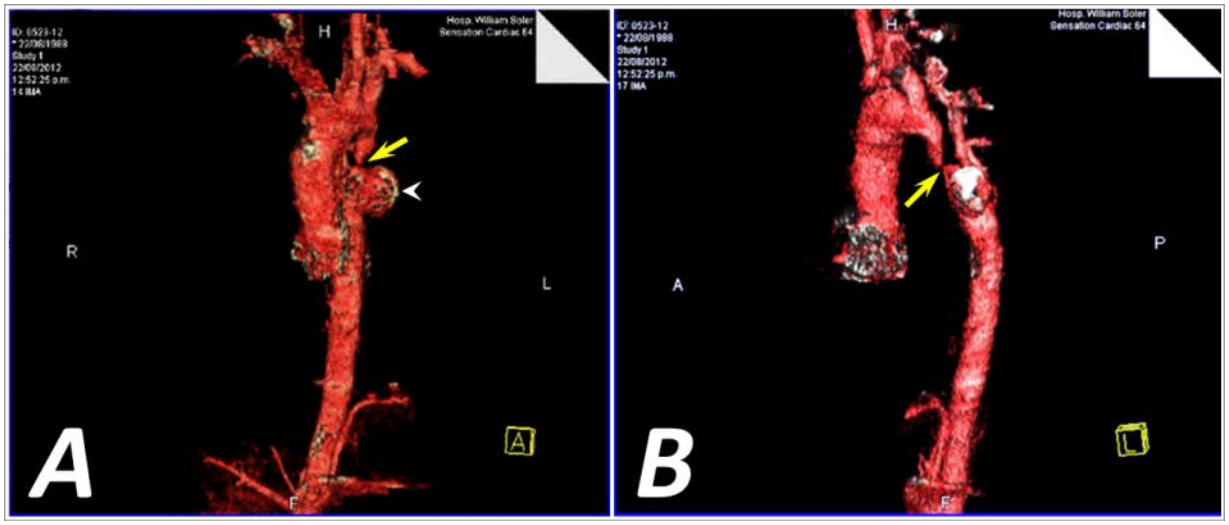


**Figura 2.** CT angiography views of the aorta (VRT reconstruction) showing CoA with small hypoplastic segment before coarcted area, ending in an image with "pencil point" (arrows), followed by a calcified saccular aneurysm, post-coarctation (arrow). Anteroposterior view (**A**) and left anterior oblique view (**B**).

the causes of resistant hypertension and of false resistant hypertension (**Table 1**)[2].

The presence of refractory HT in a young adult requires the ruling out of secondary causes. The patient was studied with this approach. We began by screening for vascular-renal conditions, which happen to be the most frequent ones, especially fibromuscular dysplasia usually seen in young women between the third and fifth decades of life.

The term CoA refers to a narrowing of the aorta causing an obstruction to blood flow[3-5]. It is typically located on the descending thoracic aorta, distal to the origin of the left subclavian artery. Most are located in the region of the posterior wall of the aorta opposite the insertion of the arterial duct (*ductus*), and are classified according to their relationship as preductal, juxtaductal and postductal coarctation.

The CoA accounts for between 3 and 10% of all congenital malformations. Its prevalence is estimated at 2.09 per 10 000 live births and ranks eighth in order of frequency of congenital heart disease. It may be isolated or associated with several conditions of which the most common are bicuspid aortic valve, ventricular septal defect and Turner's syndrome[3-5]. Most often, it appears as an isolated case that respond to the multifactorial inheritance pattern described for

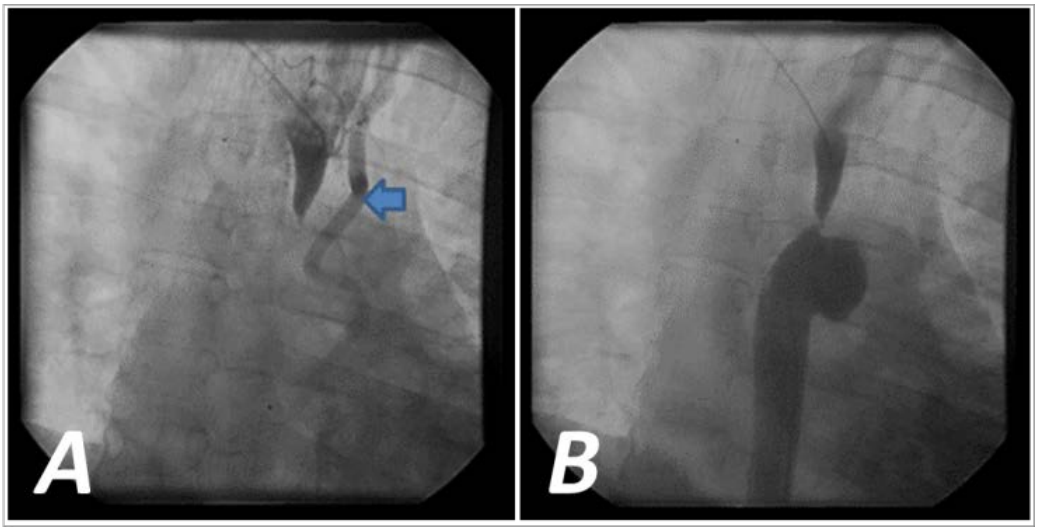


**Figura 3. A.** Angiogram showing total obstruction of aortic flow at the level of the aortic isthmus. The arrow points to the large collateral branch corresponding to the left internal thoracic artery. **B**. Simultaneous injection of contrast using two catheters, across the aortic isthmus and the descending portion. Critical CoA and post-coarctation aneurysm are seen.

**Table 1.** Etiologic classification of refractory hypertension.

| Causes of resistant hypertension | Causes of false resistant hypertension |
|---|---|
| Poor adherence to treatment | |
| Lack of lifestyle modification[a] | |
| Continued consumption of substances that increase blood pressure[b] | White coat hypertension |
| Obstructive sleep apnea | Inadequate diameter of the cuff |
| Non suspected secondary cause[c] | Pseudohypertension |
| Irreversible or poorly reversible organ injuries | |
| Volume overload[d] | |

[a] Weight gain and heavy drinking (intermittent binge drinking)
[b] Liquorice, cocaine, glucocorticoids, nonsteroidal anti-inflammatory drugs
[c] Aortic coarctation, vascular-renal causes, hyperaldosteronism
[d] Insufficient diuretic treatment, progressive renal failure, high sodium intake, hyperaldosteronism

most congenital heart disease, but also familial cases have been described with Mendelian inheritance[6].

The clinical presentation of CoA has a broad spectrum. It is a disease with high mortality in neonates, with refractory heart failure, as it depends on the ductus arteriosus and can lead to right ventricular failure or biventricular failure. In infants and schoolchildren, it shows very little symptoms, is virtually asymptomatic. The first clinical signs, typical of the adults, appear in adolescents[7].

At that stage of life, the patient present with symptoms such as occipital headache, occasional epistaxis and weakness in the lower limbs, which may rarely reach claudication while walking. Other symptoms appear due to the refractory nature of HT, such as tinnitus, dizziness or facial redness. If there is a well-established left ventricular hypertrophy, dyspnea and effort angina may occur.

On physical examination, there is absence or decrease in the amplitude of the pulses in the lower limbs; HT in the upper limbs with pressure gradient of 20 mm Hg or more, compared with the lower limbs; interescapular aortic ejection systolic murmur; and if there were important aorto-bronchial or aorto-pulmonary collaterals, continuous back murmurs may be heard[8]. The electrocardiogram shows signs of left ventricular hypertrophy, with varying degrees of sensitivity and specificity[8].

The assessment of the symptoms and clinical signs is reinforced with a telecardiogram, in which the 3 sign is displayed by the configuration adopted by the thoracic aorta with pre- and post-coarctation dilation, and the appearance of rib notching between the second and ninth ribs (Roesler sign), which show the development of collateral circulation through the intercostal arteries and the internal thoracic artery[8].

The echocardiography is a diagnostic tool of extraordinary value to locate the site and size of the CoA, calculate the trans-coarctation peak and mean pressure gradients, assess the impact of the obstruction in the systo-diastolic left ventricular function, the size of the hypoplastic segment that accompanies it, and clarify the association with other heart diseases.

The CT angiography is considered the main technique for assessing extracardiac vascular structures, as it allows the assessment of the coarctation site, the degree of aortic hypoplasia, the magnitude of pre- and post-stenotic dilation, and the number of aorto-pulmonary collaterals; however, it has the disadvantage of exposure to iodinated contrast and ionizing radiation. Cardiac magnetic resonance imaging is a diagnostic technique that is experiencing a boom in contemporary cardiology. It is free of radiation, provides detailed anatomical information and is routinely used in some centers, although its high cost is an inconvenient[9].

A definitive treatment of CoA may be accomplished by surgical repair of the defect or by percutaneous intervention, whereby the lesion is only dilated or, in addition, a stent is implanted. The choice of one of these techniques depends on the weight of the patient, the anatomy of the site, the degree of hypoplasia of the aortic arch and of the pre-coarctation area[7,8].

The most common complications are the recoarctation of the site; residual systemic hypertension, especially in those treated in older ages; true aortic aneurysm after surgery; paraplegia due to damage to the spinal arteries, and late aortic dissection, after surgical repair[8].

Recoarctation of the aorta has been reported between 7 and 60% of cases, paraplegia due to spinal cord ischemia is rare (0.4%). The incidence of true aneurysm is estimated between 2 and 27%, while late dissection is an extremely rare complication.

Mortality is estimated at 1% after surgical repair[8].

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