

## Type A Aortic Interruption: Case Report

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### Introduction

Aortic interruption is an anatomical condition that consists in aortic isthmus narrowing between the left subclavian artery and the ductus arteriosus. This condition occurs congenitally, post aortoplasty with balloon catheter or post-surgery.<sup>1</sup> Symptoms may appear in newborns or young adults, depending on its etiology. Clinical manifestations are varied and include findings of pulse asymmetry on physical examination, hypertension in the upper limbs and hypoperfusion of abdominal viscera and lower limbs, which may lead to renal failure, lack of pulse and claudication.<sup>2,3</sup> Aortic interruption may also be asymptomatic and can be identified only because of the presence of systemic arterial hypertension.<sup>4</sup> Diagnosis can be prenatal, through fetal ultrasound or after clinical suspicion, when echocardiography should be performed to confirm the narrowing and detect any associated heart diseases, especially left ventricular dysfunction.<sup>5</sup> Associated with echocardiographic evaluation, tomography angiography or magnetic resonance angiography is performed for confirmation purposes. Treatment is surgical, having several techniques described in the literature, such as extra-anatomical by-pass, stenting, and intra-aortic balloon dilation.<sup>5,6</sup> The choice of technique is fundamentally based on the ductus arteriosus persistence, its relationship with the coarctation site and the presence or absence of collateral circulation. Some complications are likely to occur. The main ones are bleeding, paraplegia, re-stenosis and aneurysms.<sup>6,7</sup> In this report, we describe the case of a female patient with clinical signs of coarctation of the aorta, systemic arterial hypertension and congestive heart failure as comorbidities, treated with the surgical technique of extra-anatomical by-pass.

### Case Report

N.A.O, 60 years old, hypertensive with hypertensive disorder of pregnancy for 22 years and paroxysmal atrial fibrillation, was admitted to a tertiary hospital one month after referring dyspnea on medium exertion, progressing to dyspnea at rest, orthopnea, and nocturnal paroxysmal dyspnea associated with edema of the lower limbs and sharp chest pain diagnosed with

### Keywords

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cardiomegaly and pleural effusion. At hospital admission, the patient was diagnosed with acute pulmonary edema, atrial fibrillation with high ventricular response and pneumonia, and was treated for clinical control. During hospitalization, upon worsening of the condition, she was diagnosed with severe aortic stenosis and coarctation of the aorta. Early surgical intervention was recommended.

At hospital admission, physical examination showed compromised general condition, soft edema with pitting in the lower limbs, paleness, heart rate 150 bpm, normal cardiac auscultation, and pulmonary auscultation with abolished vesicular murmur at the right base.

In the protocol of preoperative admission examinations, electrocardiography revealed atrial fibrillation, left bundle branch block and left ventricular overload. Percutaneous coronary angiography could not be performed as the 0.035<sup>o</sup> cord did not go past the aortic arch at first, but three days later it was performed again via femoral and radial arteries, showing coronary arteries without lesions and aortic occlusion, aortic interruption in the plane of the posterior aortic arch branch immediately after the emergence of the left subclavian artery with an extension of approximately 1 cm and a rich network of collaterals (Figures 1 and 2). Chest X-ray revealed pleural effusion to the right.

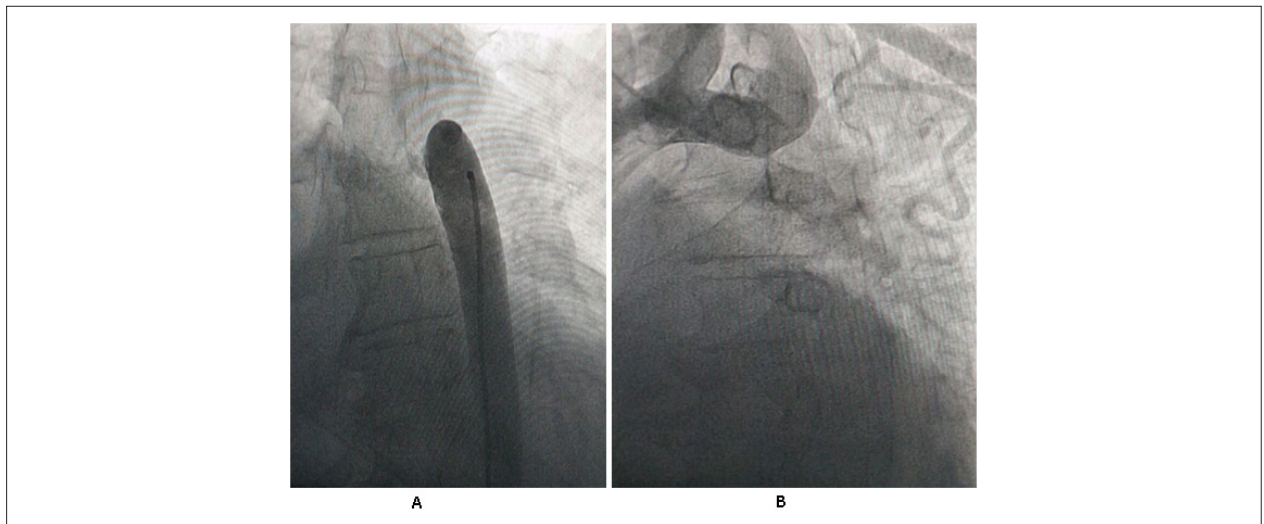
Two-dimensional preoperative Doppler echocardiography showed left ventricular ejection fraction of 63%, left ventricular (LV) concentric hypertrophy, preserved global and segmental LV contractility, signs of diastolic dysfunction, tricommissural aortic valve with thickened calcified cusps with reduced mobility, mild failure and severe stenosis, maximum systolic gradient 91 mmHg and mean systolic gradient 48 mmHg, aortic peak velocity 4.8 m/s, valve opening area of 0.7 cm<sup>2</sup>, VTI LVOT/Ao ratio of 0.22. Transesophageal echocardiography did not show descending thoracic aorta from the distal portion of the arch.

Extra-anatomical aortic bypass surgery was conducted with 18 mm grafting in the supra-diaphragmatic descending aorta to the ascending aorta and implantation of metal aortic prosthesis 21 (Figure 3).

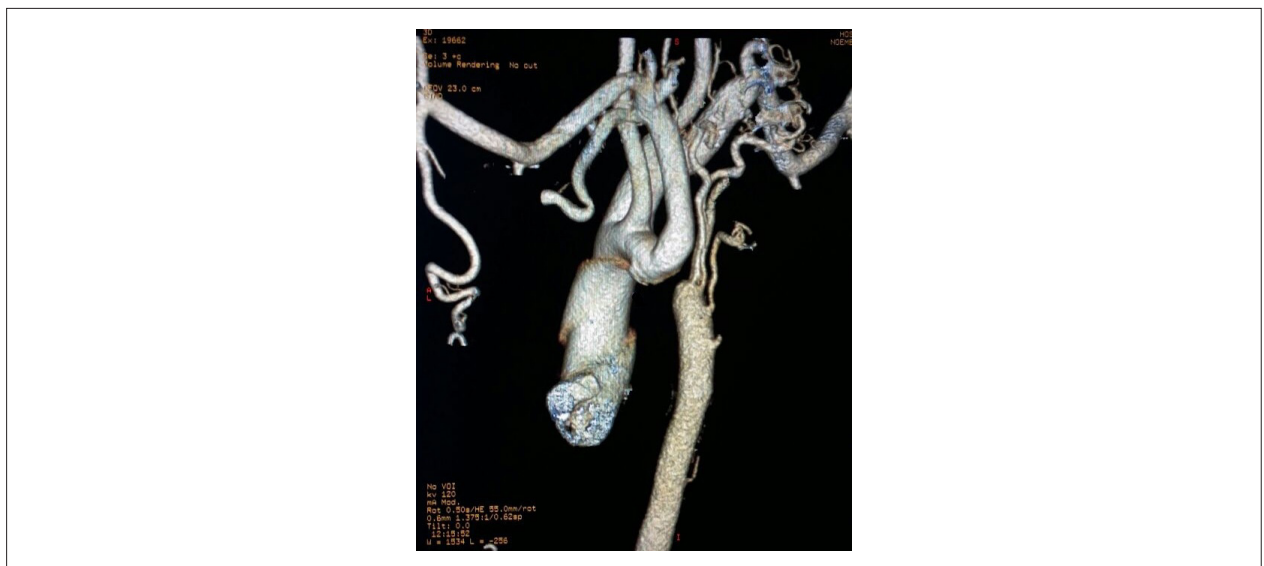
Postoperative two-dimensional Doppler echocardiography showed left atrial enlargement, left ventricular concentric hypertrophy, preserved left ventricular systolic function, left ventricular relaxation deficit, normally functioning metal aortic prosthesis, mild mitral regurgitation and absence of pericardial effusion.

In the postoperative period, the patient evolved with vasoplegia requiring Noradrenaline, myoclonus twitching in the upper limbs and eyelids, and upward gaze. Embolic phenomenon was ruled out by computed tomography of the skull without abnormalities.

After stabilization of the hemodynamic parameters, the patient remained in hospital and presented clinical improvement, followed by hospital discharge.



**Figure 1** – A: Femoral aortography showing stop in descending thoracic aorta. B: Radial aortography showing stop after the arch.



**Figure 2** – Tomography angiography showing type A aorta interruption in the plane of the posterior knee of the arch with extension of approximately 1 cm.

## Discussion

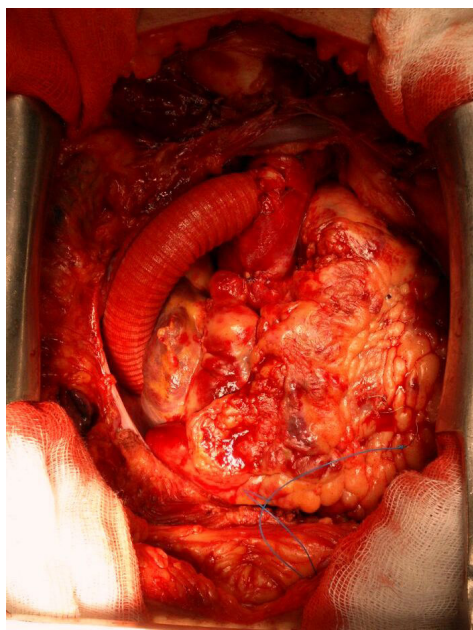
The age groups in which aortic interruption is diagnosed are usually childhood and young adults, but in some cases, such as in the patient of this report, the disease is diagnosed in older adults.<sup>7</sup> This is due to the rich network of collateral circulation formed over time to compensate for the hypoperfusion of some organs and tissues. This adaptation masks the symptoms and makes it difficult to provide early diagnosis for this condition,<sup>2</sup> whereas the patient may even be normotensive.<sup>8</sup>

Aortic interruption is frequently associated with other cardiac malformations such as ventricular septal defect and left obstructive lesions due to aortic constriction. Among these lesions, the bicuspid aortic valve is a very prevalent abnormality, accompanying 50 to 70% of AoCo cases. Double obstruction

leads more easily to dilation of the aorta, thus increasing the risk of dissection in these patients.<sup>9</sup> Despite its lower incidence, coarctation of the aorta has been associated with many diseases, including dextrocardia, complete atrioventricular septal defect, Turner syndrome (20%), Shone's syndrome, dilated cardiomyopathy, and others.<sup>8</sup>

Refractory arterial hypertension may be the guiding symptom for the diagnosis of coarctation of the aorta, since this is often the first clinical sign. Complications such as aortic aneurysmal dilation, dissection, and coronary artery diseases may be secondary to arterial hypertension caused by coarctation, and develop especially in asymptomatic patients.<sup>4</sup> Some patients may report headache, chest pain, lower limb pain, or even claudication during exercise.<sup>3</sup>

## Case Report



**Figure 3** – Bypass surgery with 18 mm graft in the supra-diaphragmatic descending aorta to the ascending aorta.

Patients with refractory arterial hypertension should ideally be screened for a thorough diagnostic investigation to identify this anomaly before more severe impairments. In this context, in addition to detailed anamnesis and thorough physical examination, imaging tests play an essential role in proving aortic constriction, and are important preoperative tools.<sup>1,4</sup>

As for diagnostic methods, two-dimensional Doppler echocardiography shows the coarctation zone, the calculation of the transcoarctation gradient and the presence of other associated disorders, such as valvulopathies and congenital malformations.<sup>10,11</sup> Angiography is of fundamental importance to define more precisely the location and extent of the obstructed segment, presence and degree of collateral circulation, thus helping to estimate the severity of the case and the type of intervention required. In addition, catheterization is also of great importance as evaluates the coronary arteries, looking for any abnormality or malformation that may interfere in the choice of the most appropriate procedure.<sup>5,10</sup>

The tests are fundamental for the diagnosis and are also important tools in preoperative preparations, aiming to provide information such as degrees of ventricular impairment and associated valvulopathies, besides the intensity and location of the constriction and the extent of the collateral arteries near the affected site. Such information is of fundamental importance for choosing the most appropriate treatment for the patient and for their long-term follow-up after surgery.<sup>5,12</sup>

Definitive treatment is surgical and increases the life expectancy of the patients, since without surgery, 90% of those affected die before age 58 and 50% before the age of 32.<sup>7,11</sup> There is more than one surgical technique described in the literature, and its choice is made after evaluating the coarctation site, the collateral circulation extent and, in case of recurrence, the degree of mediastinal adhesion.<sup>6</sup>

Balloon aortoplasty produced satisfactory immediate results; however, long-term follow-ups demonstrated vascular lesion causing high rates of restenosis and formation of aneurysms.<sup>7</sup> In order to improve this technique, stents were introduced to minimize adverse effects.<sup>13</sup>

The extra anatomical technique, initially introduced by Oliveira et al.,<sup>14</sup> consists of placing a Dacron tube between the ascending aorta and the descending thoracic aorta via sternotomy and with a posterior pericardial approach, to make the surgery simpler. Using this technique in adult patients with atypical variants such as coarctation of the aortic arch or with many associated comorbidities, as in the case reported, was proven better than anatomical surgeries, with lower morbidity and mortality rates and lower rate of long-term recurrence.<sup>7</sup>

### Conclusion

This case demonstrates the importance of investigating nonspecific clinical signs that develop suddenly with clinical worsening. Imaging studies in the department of cardiology are of critical importance, especially cardiac catheterization, echocardiography and computerized tomography angiography.

### Authors' contributions

Data acquisition: Valente AS; Data analysis and interpretation: Lino DOC, Belém LS; Manuscript drafting: Alcantara ACB, Brilhante MA; Critical revision of the manuscript for important intellectual content: Lima CJM.

### Potential Conflicts of Interest

There are no relevant conflicts of interest.

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**Academic Association**

This study is not associated with any graduate program.

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