

# Stanford A aortic dissection with involvement of supraaortic vessels associated with bicuspid aortic valve, ascending aortic aneurysm and coarctation of the proximal descending aorta: A case report

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## Abstract

**Background:** Of all acute aortic syndromes, more than 85% correspond to aortic dissection, reaching a mortality rate in the first 24 hours up to 50% in its natural evolution (1). Although this entity has some specific signs and symptoms for early diagnosis, in most cases they do not present as in the literature, so when acute aortic pathology is suspected, computed axial tomography is the most widely used imaging study due to its speed, low cost, availability, high sensitivity, risk stratification and the possibility of determining the management (2).

**Case Description:** This paper reports the case of a patient who presented in the emergency service with precordial pain radiating to the jaw and back, diffuse changes in the electrocardiogram and showing in the tomography type A aortic dissection associated with aneurysm of the aorta and coarctation of the proximal descending aorta. In addition, the echocardiogram showed severe early diastolic aortic regurgitation and bicuspid aortic valve. He underwent surgery immediately, achieving hospital discharge after a few weeks.

**Conclusion:** The early suspect, the diagnosis with early imaging of the heart and aorta and prompt intervention may improve the prognosis in a pathology as serious and with such a high mortality rate as aortic dissection.

## Introduction

Aortic dissection is characterized by a tear in the intima and media layer of the aorta, producing a dissection flap that separates the true lumen from the extraluminal channel called the false lumen, with the possibility of anterograde and/or retrograde extension of the lesion and limiting the distal flow (3). Determining the exact prevalence of aortic dissection is a challenging due to its high mortality rate, particularly before reaching the hospital. However, it was reported and age- and sex-adjusted incidence of 4.4 per 100,000 person-year from 1995 to 2015 (4). This condition occurs more frequently in older men, typically between their fifth and seventh decade of life, who have a history of high blood pressure and atherosclerosis. In younger patients, aortic dissection is often associated with factors such

as a bicuspid aortic valve, collagen disorders, history of cardiac surgery, arterial hypertension and smoking (1).

## Case report

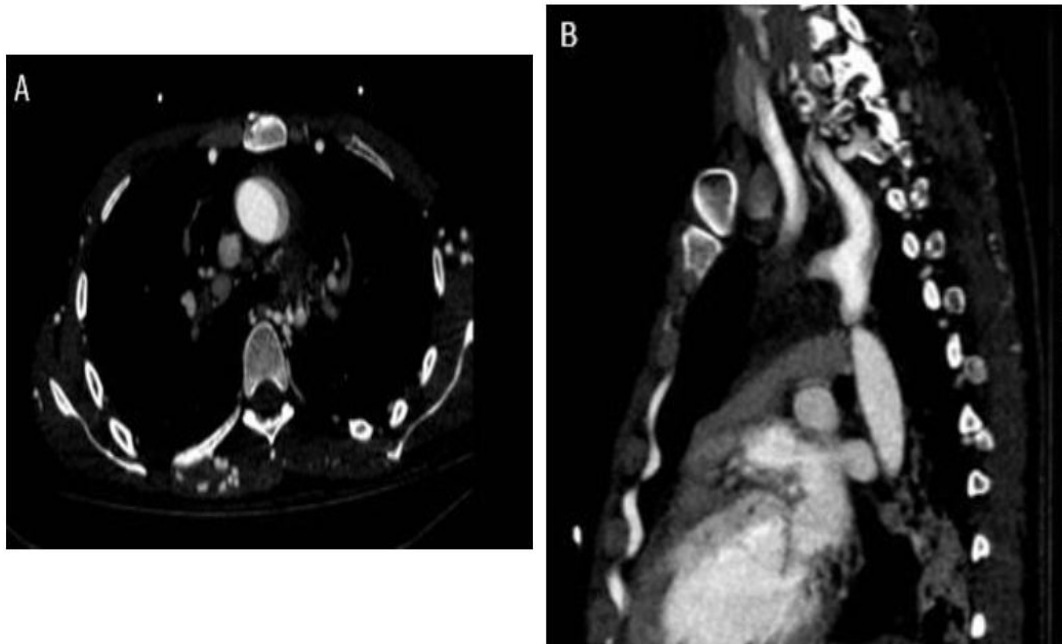
### History and presentation

A 30 year-old male patient, with a history of smoking, unstudied arterial hypertension, and dislocated lens of the left eye, consulted in emergency service due to precordial pain lasting one hour, pain intensity 10/10, of sudden onset, radiating to the jaw and back.

### Physical examination

As relevant, a tall patient is evident (6'3") with unusually long arms, presenting tachycardia, diaphoresis, blood pressure of 140/30 mmHg, imperceptible pulses in lower limbs and a holodiastolic murmur in mesocardium.

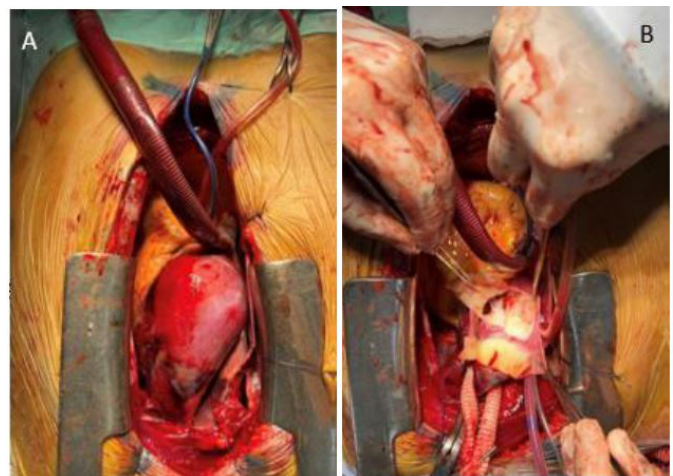
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**Figure 1:** A) In this computed axial tomography, the dissection flap can be seen at the level of the ascending aorta, at the same time that the coarctation point of the proximal descending aorta is seen (blue arrow). B) In another section of the same study, it can also be observe the involvement of the supra-aortic vessels, with the dissection flap ending inside the coarctation.



**Figure 2:** In this 3D reconstruction, it can be objectified the ascending aortic aneurysm, the coarctation of the proximal descending aorta generating the staining difference before and after coarctation, and the evidence of significant collateral circulation and increased permeability at the mammary, mediastinal, dorsal, axillary, abdominal and lumbar levels.



**Figure 3:** A) When performing the sternotomy, the aneurysmal dilatation of the ascending aorta is evident, observing a hematoma on its wall. B) Continued surgery, the intimal dissection of the same aortic zone was observed.

## Investigations

ECG showed signs of diffuse subendocardial injury. Given de suspicion of aortic dissection, an emergency computed axial tomography was performed evidencing ascending aortic aneurysm (diameter 62 mm) (Figure 2) with the presence of a dissection flap that extended to the proximal descending aorta, compromising the arch, brachiocephalic trunk, right and left carotids up to skull base and right and left subclavians at their birth, concluding inside a coarctation in the proximal descending aorta (8 mm diameter)(Figure 1). The transthoracic echocardiogram also revealed bicuspid aortic valve with severe early diastolic aortic regurgitation.

## Operation Procedure

Emergency surgery was performed on the same day of admission, revealing aneurysmal dilatation of the ascending aorta with a hematoma in its wall, wich when sectioned reveals the dissected intima (Figure 3). Aortic valve replacement with mechanical valve Number 27, replacement of the ascending aorta with a Dacron tube, and reimplantation of both coronary arteries was performed (Figure 3). Valve biopsy showed degerative valve disease with fibrosis and myxoid changes.

## Postoperative Course

From de hemodynamic point of view, he evolves with cardiogenic shock and requirements for high doses of inotropes and vasoactive agents, intervening with reanimated cardiorespiratory arrest. He also experienced massive medical bleeding, without response to the medical treatment established, with reoperation requirements, multiorgan failure requiring dialysis and sepsis due to nosocomial pneumonia. In the following days, the doses of inotropes were decreased until their closure was achieved and the patient could be extubated, without neurological sequelae. Four weeks later, hospital discharge was granted and percutaneous stent implantation was performed as an outpatient treatment for coarctation of the aorta without complication.

## Conclusion

Beyond low prevalence of acute aortic syndromes, clinical suspicion and request for adequate complementary studies is important because the prompt and targeted intervention can change the prognosis of a disease with such a high rate of mortality and complications.

Eventually, in patients in whom collagenopathy is suspected, try to look for aortic pathology, look for some type of aortic pathology, since with low-cost and highly available studies, early diagnosis and treatment can be made without the need to go through a potentially fatal acute event.

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