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CASE REPORT

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# Aneurysm and subacute type a aortic dissection, in a pediatric patient with aortopathy

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**Abstract**

Aortic dissection in pediatrics is an extremely rare condition, which is generally related to predisposing factors such as connective tissue disorders, congenital heart disease and systemic arterial hypertension. A 3-year-old girl, with a history of bicuspid aortic valve, hypoplasia of the aortic arch and repaired aortic coarctation at one month of age. She was admitted 2 months of atypical chest pain, dysphonia, and low tone of voice. The echocardiogram and CT angiography showed an image corresponding to a Stanford A aortic dissection, with false lumen perfusion that generated a aneurysmal dilation with a saccular morphology of 53×40×70 mm dimensions. The patient was taken to surgery, exposure of the ascending aorta, aneurysmal dissection, and replacement with a 22 mm supracoronary tube were performed. We present a case of a 3-year-old pediatric patient with Stanford A aortic dissection, subacute evolution, with successful repair.

**Keywords** Ascending aortic dissection, Aneurysm, Aortic valve disease

## Background

The presentation of aortic dissection in pediatric patients is extremely rare [1] and is generally associated with predisposing conditions [2], those that weaken the middle layer of the aorta are the most frequently described. Congenital heart diseases such as coarctation of the aorta, critical aortic stenosis and bicuspid aortic valve are representative [3]. On the other hand, collagenopathies such as Marfan, Ehlers-Danlos, Turner and Loeys-Dietz syndromes, as well as tuberous sclerosis, and familial aortic dissection, among others, are also related to the presentation of aneurysms and aortic dissection [4]. In a 1-year follow-up study in New York including 5658 cases of aortic dissection, only 2 deaths of individuals under 19 years of age were reported [5].

The objective of this case report is to share the experience in the management of a 3-year-old patient with bicuspid aortic valve, ascending aortic aneurysm and subacute type A aortic dissection. The informed consent was obtained and it was approved by the ethics committee of the CardioVID Clinic.

## Case presentation

We report the case of a female patient, 3-years-old, with a history of bicuspid aortic valve, critical aortic valve stenosis, hypoplasia of the aortic arch, coarctation of the aorta, perimembranous ventricular septal defect and a patent ductus arteriosus. At 23 days, aortic valvuloplasty

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was performed with a 6 mm x 20 mm mini Tyshak balloon, with a residual gradient of 30 mmHg; 6 days later, reconstruction of the ascending aorta and aortic arch with a bovine pericardium patch, aortic valvuloplasty, and pulmonary artery banding were performed to manage defects with left-to-right shunting.

1 year later, echocardiographic follow-up showed evidence of aortic recoarctation at the level of the aortic arch, so it was decided to reconstruct the aortic arch with a bovine pericardial patch, remove the pulmonary banding, and close the interventricular communication with a pericardial patch.

After a period of 12 months without institutional follow-up, she presented once more to our institution with symptoms of atypical chest pain, dysphonia, hoarseness and worsening functional class. Due to her history, she was admitted for studies and a chest x-ray showed significant mediastinal widening, a cardiothoracic index of 0.65 (Fig. 1).

An echocardiogram was performed with findings of dysplastic aortic valve with bicuspid opening and severe dilation of the ascending aorta. Measurements included an aortic root of 16 mm (Z score + 0.13), a sinotubular junction of 20 mm, a proximal ascending aorta of 15 mm (Z score + 0.63) and an apparent ascending aorta aneurysm of 40 × 70 mm with an inner-thrombosed area and spontaneous contrast. (Fig. 2).

A chest angiotomography was performed where an intimal tear hole measuring 16 mm in diameter (type A aortic dissection) was found 40 mm above the valvular plane, with false lumen perfusion that generated a saccular aneurysmal dilation measuring 53 × 40 × 70 mm. The aneurysm produced compression of the pulmonary artery, left main bronchus and left upper lobe bronchus with signs of air trapping (Fig. 3).

After these diagnostic studies, it was decided to undergo surgical correction. Cervical arterial cannulation is performed at the level of the carotid artery and right jugular vein. Extracorporeal circulation is started and cooled to 20ºC. A new opening is made with rupture of the aneurysmal sac, the ascending aorta is exposed, antegrade cardioplegia of the nest is performed, antegrade cerebral perfusion is started, the aneurysm and dissection area are dissected and excluded. The aortic wall is resected and sent for pathological study, the bicuspid aortic valve is calibrated, the ascending aorta is reconstructed with a 22 supracoronary Dacron tube, anastomosis to the proximal and distal arch with the island technique. Cerebral circulation is gradually restarted and total flow is restarted. Extracorporeal circulation

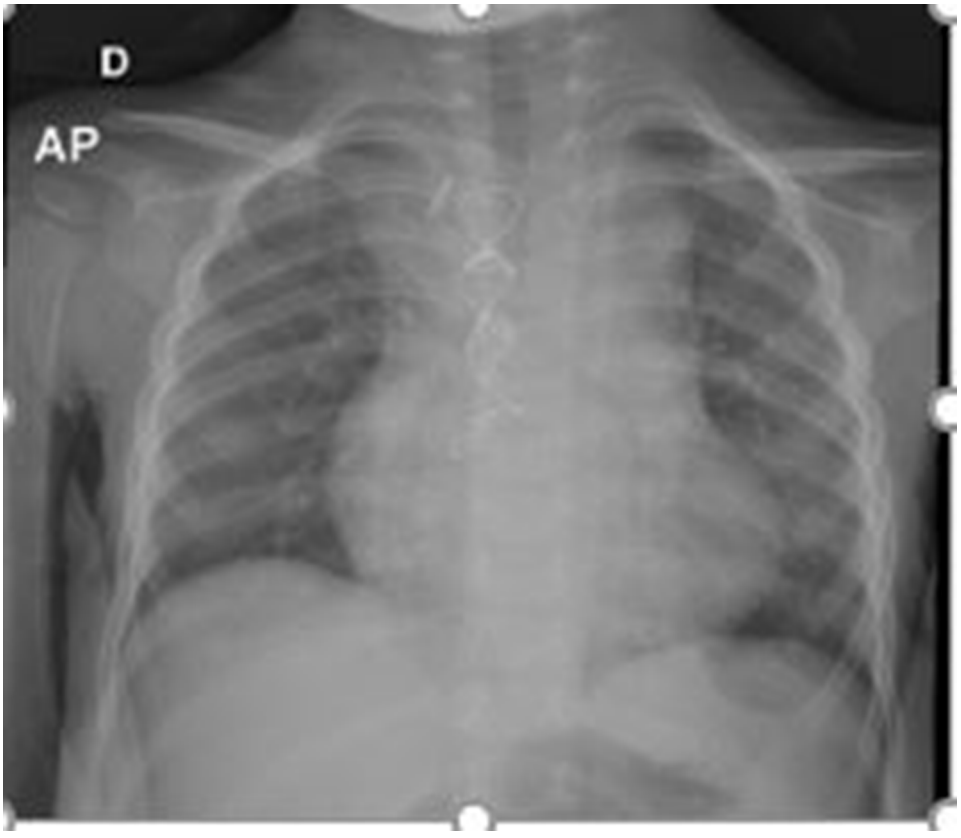


**Fig. 1** Findings on chest x-ray. ** Mediastinal widening. Poorly visualized aortic arch. Magnified global cardiomegaly. Pulmonary hila without lesions. Adequately expanded lungs with no evidence of focal lesions, pleural effusion, or pneumothorax

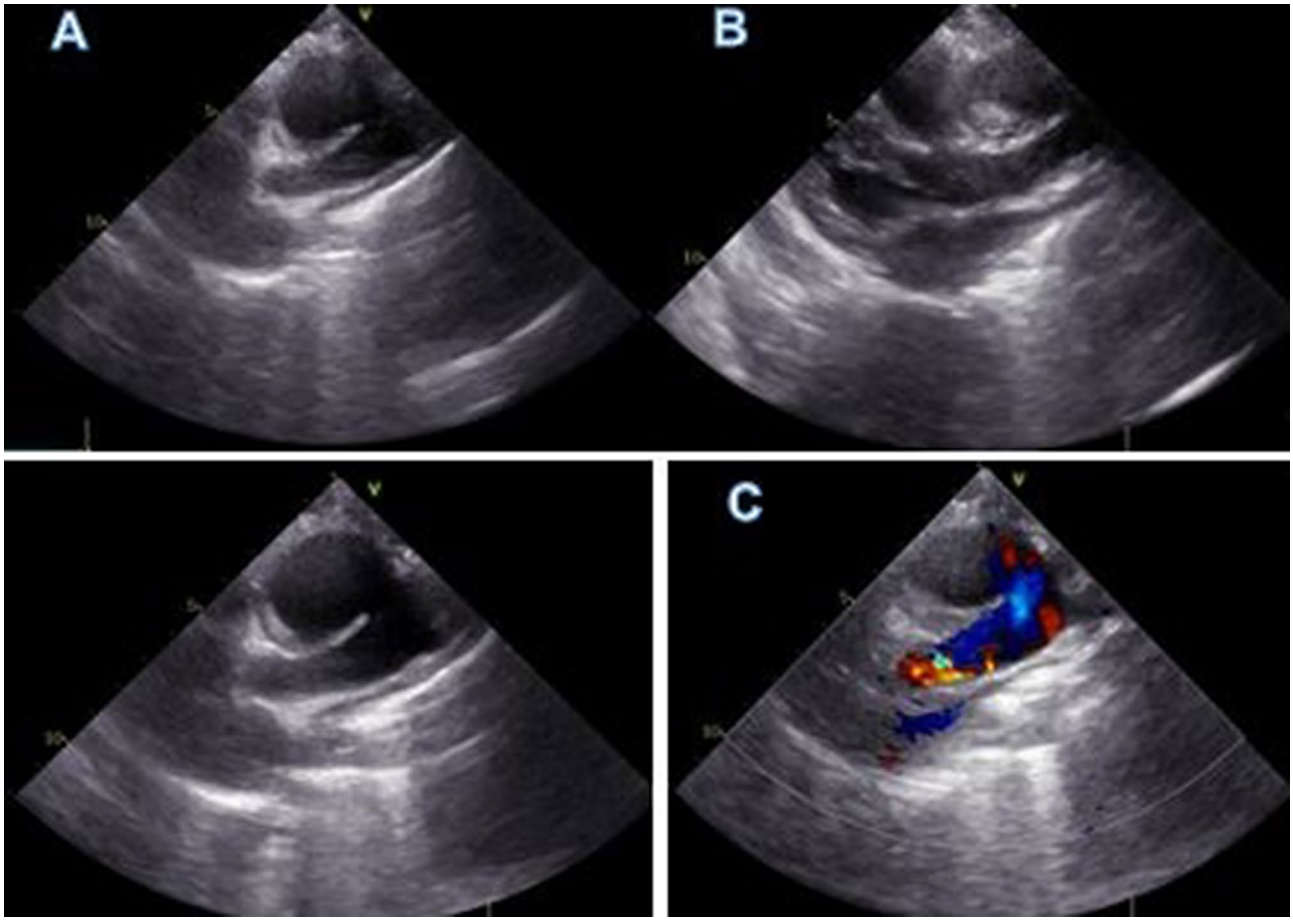


**Fig. 2** Echocardiographic findings in suprasternal section. **Severe dilatation of the ascending aorta that begins 20 mm from the sinotubular junction, where a 12 mm aneurysmal mouth is observed in the long axis sternal plane, the diameter of said aneurysm is 40 by 70 mm with a thrombosed area inside and spontaneous contrast. Diameter of the ascending aorta prior to the aneurysm is 15 mm and the aortic root is 16 mm

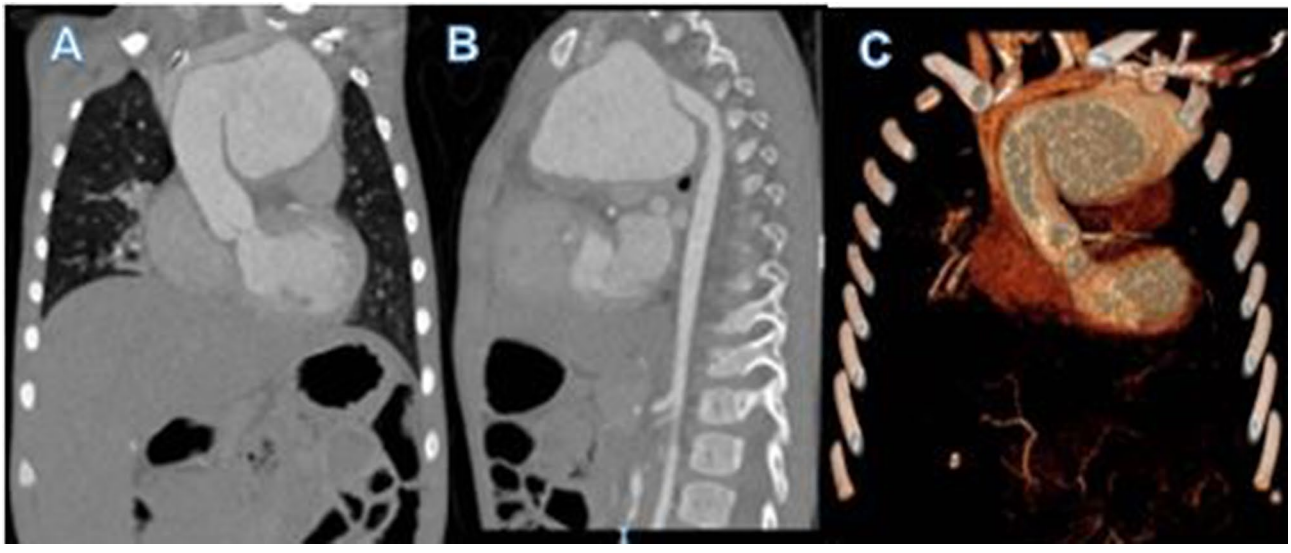


**Fig. 3** Findings on chest angiotomography. ** 40 mm above the valvular plane, in the proximal arch (just at the height and in front of the emergence of the supra-aortic trunk), an intimal tear orifice of 17 mm in diameter and a dissection flap with perfusion of the false lumen, which generates a saccular dilation measuring 53 mm x 40 mm x 70 mm, are observed. Mural thrombus towards its lower margin. The aortic arch after the emergence of its two branches measures 8 mm in diameter, the descending aorta after the aneurysm measures 8.6 mm in diameter

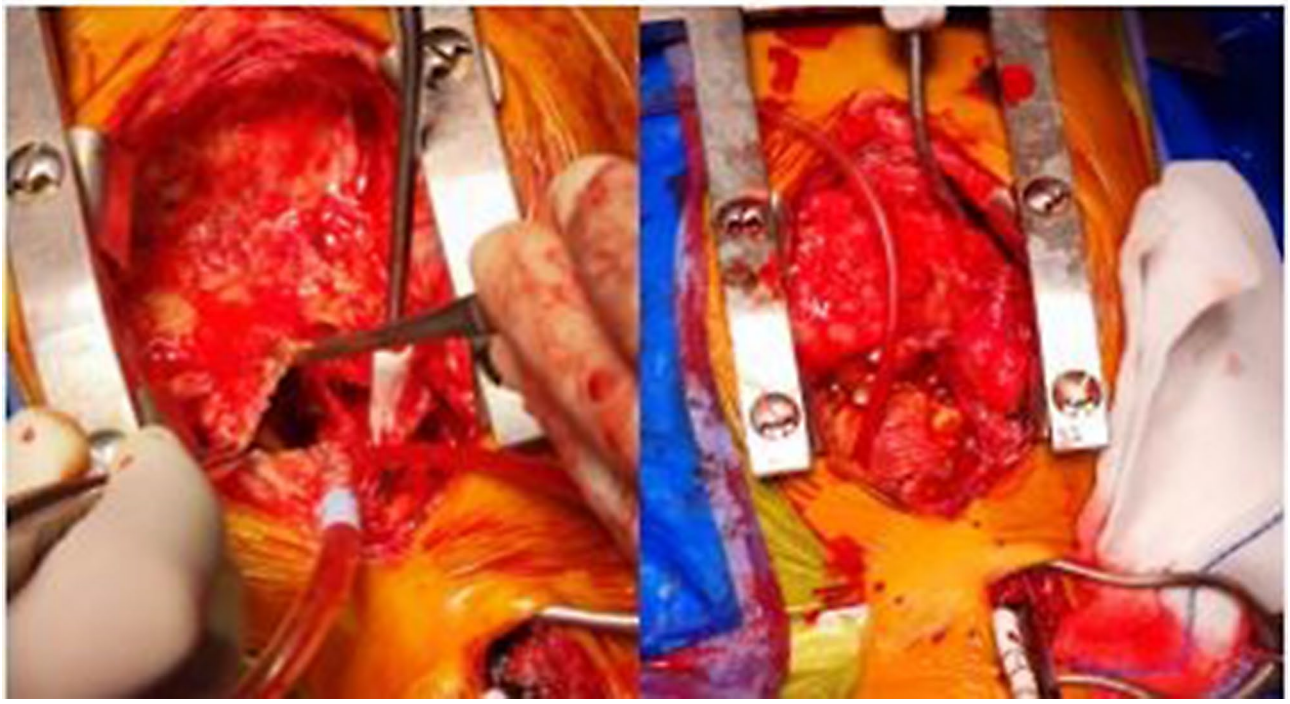

**Fig. 4** Surgical findings (supra-coronary tube placement)

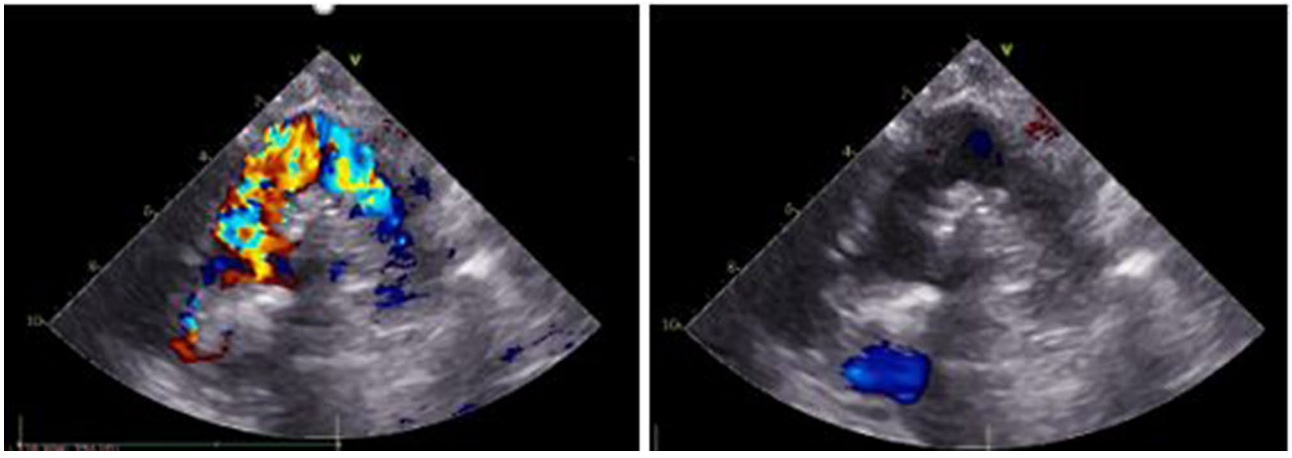

**Fig. 5** Control echocardiography 3 months after surgical procedure. **Reconstruction of the ascending aorta with a 22 mm Dacron tube. Maximum gradient in the ascending aorta was 16 mmHg and mean of 8 mmHg. Pulsatile flow in the descending aorta without stenosis

is successfully started and sternal closure is achieved (Fig. 4). The biopsy revealed diffuse interstitial degenerative changes in the tunica media. Medical therapy was adjusted and she was discharged with strict follow-up by Pediatric Cardiology, Genetics and Pediatrics.

The patient was evaluated by medical genetics who reported that she did not present a syndromic phenotype. Exomic sequencing with copy number variation was negative. During the follow-up by pediatric cardiology, echocardiography was performed with evidence of reconstruction of the ascending aorta with a 22 mm Dacron tube. Maximum gradient in the ascending aorta was 16 mmHg and mean of 8 mmHg. Pulsatile flow in the descending aorta without stenosis (Fig. 5). Good biventricular function, with no evidence of cavity dilation and mild ventricular hypertrophy. Follow-up continued for 6 months with the specialty and medical management.

## Discussion

Aortic dissection and aneurysms are rare in pediatric patients, being generally associated with predisposing conditions such as structural cardiac lesions at the left ventricular outflow tract and aorta, as shown in the natural history of this patient. Follow-up studies of bicuspid

aortic valve in pediatric patients have estimated a higher risk than the general population of progressive aortic dilation and Stanford A aortic dissection [6–8].

The typical presentation of aortic dissection is acute, which is manifested by the onset of intense, tearing chest or abdominal pain, with changes in pulse and/or blood pressure. In imaging studies such as chest x-ray, mediastinal widening may be evident, which increases the index of suspicion and this must be confirmed with echocardiography and/or angiotomography. In the case presented, the temporal evolution of the condition was subacute, latent and with atypical symptoms, so the index of suspicion was essential to reach the diagnosis. Furthermore, the possibility of redissection, as well as the formation of new aneurysms in predisposed patients, is relevant to defining routine clinical follow-up [5].

In major pediatric aortic dissection series such as the KID aortic dissection cohort and the study by Fikar et al.; the average age of presentation was 15 to 17 years with a male predominance [6]. There are few cases that occur in children under 5 years of age and in females like the present case. A multicenter study by Luehr et al. from 139 cases with type A aortic dissection, 36% were related to connective tissue diseases and only 6 cases occurred in children under 18 years of age. A case of acute aortic dissection and aneurysm is considered a sentinel event for other family members, since there may be a familial genetic predisposition to present pathology of the middle aorta [7, 8].

Stanford A aortic dissection requires mandatory and urgent surgical management due to the risk of lethal complications such as rupture, cardiac tamponade and myocardial ischemia. Yoneyama et al. report 2 cases of acute aortic dissection in Stanford A in patients with ACTA2 mutation, requiring emergent management who died after the surgical event [9]. In recent years, with the improvement of the surgical technique, in addition to selective perfusion and hypothermia techniques, morbidity and mortality in adult and pediatric patients have decreased [5].

## Conclusions

We present the case of a 3-year-old girl with an ascending aorta aneurysm, Stanford A aortic dissection with a subacute course and complete and successful repair of her pathology, with good postoperative evolution.

#### Author contributions

Andres Aranzazu Ceballos (A), Margarita Maria Zapata (B), Sharoon Suarez (C), Ivan Mendieta (D), Ubaldo Rivas Aguilar (E) Authors A, B, C, D, E initiated the project and were present during the writing. A and D performed and revised the English translation. C and D took the images and organized them into the project. All authors approved the final version.

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#### Data availability

No datasets were generated or analysed during the current study.

## Declarations

#### Informed consent

The patient's mother signed informed consent to authorize the publication of the case.

#### Competing interests

The authors declare no competing interests.

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