

# Ruptured abdominal aortic aneurysm in an 11-year-old with multiple peripheral artery aneurysms

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## ABSTRACT

Pediatric abdominal aortic aneurysms (AAAs) are rarely encountered in clinical practice. The combination of a pediatric AAA in a patient with multiple peripheral artery aneurysms is even more rare. We report the management of an 11-year-old boy who presented with a ruptured AAA who also had multiple peripheral arterial aneurysms. Infectious, genetic, and inflammatory workup was negative, classifying this aneurysm as congenital. (*J Vasc Surg Cases and Innovative Techniques* 2020;6:539-42.)

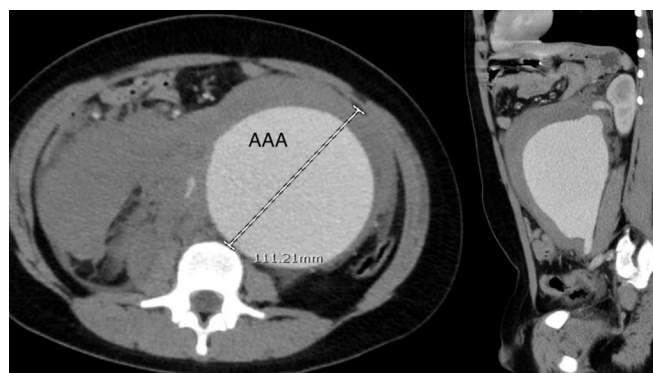
**Keywords:** Pediatric aneurysms; Ruptured aneurysm; Congenital aneurysm

## CASE REPORT

An 11-year-old boy initially presented to an outside facility with 2 days of severe abdominal pain. In the emergency department, he became unstable with profound hypotension and hypoxia and was intubated. Computed tomography angiography (CTA) was obtained demonstrating a ruptured 11 cm abdominal aortic aneurysm (AAA) (Fig 1). Past medical history obtained from the guardian revealed congenital cataracts and left-sided hearing loss since birth. Genetic testing was performed at that time, demonstrating no specific genetic disorder. The cataracts were attributed to prescription drug use of the mother. He was otherwise healthy, had an uncomplicated gestation, and was delivered at term with no intensive care unit requirements. There was no known previous suspicion for any abdominal pathology. There was no family history of connective tissue disease. There had been no recent infections, illnesses, or exposures. His paternal grandfather, maternal great grandmother, and grandfather all had degenerative AAAs with a normal age of presentation.

At our institution, he was evaluated in the emergency department and was anemic, hypotensive, and tachycardic. Physical examination revealed a distended and tense abdomen. He was taken immediately to the operating room allowing for permissive hypotension with expectation of using massive transfusion.

A laparotomy was performed revealing a large retroperitoneal hematoma but otherwise normal abdominal contents. The hematoma was evacuated while the supraceliac aorta was



**Fig 1.** Computed tomography angiography (CTA) of ruptured 11-cm abdominal aortic aneurysm (AAA).

compressed. The proximal aortic neck was dissected and supra-renal aortic control was obtained and the aorta clamped. No heparin was administered. Both distal common iliac arteries were dissected and clamped. A 12-mm Dacron tube graft was anastomosed end-to-end with pledgets to the proximal aorta. The distal aorta was inspected and it was clear the aortic bifurcation was significantly degenerated. The distal anastomosis was performed end-to-end to the left iliac artery. A 12-mm graft was interposed between the aortic graft and the right iliac artery, recreating the aortic bifurcation. The larger graft to the iliac arteries will allow growth of the patient and avoids the smaller limbs of a bifurcated 12-mm graft. A temporary abdominal closure was placed and the patient was transferred to the pediatric intensive care unit. The abdomen was closed on postoperative day (POD) 4, was extubated on POD 5, and discharged on POD 40. He suffered a prolonged ileus requiring nasogastric decompression and parenteral nutrition. Samples from the native aorta were tested for infection and connective tissue disorders, and were negative. The aortic tissue samples were negative for inflammation. Rheumatology evaluated the patient and were unable to make a diagnosis of vasculitis. The patient was tested for systemic infection and genetic disorders of 24 genes associated with aortic pathology (Invitae Aortopathy

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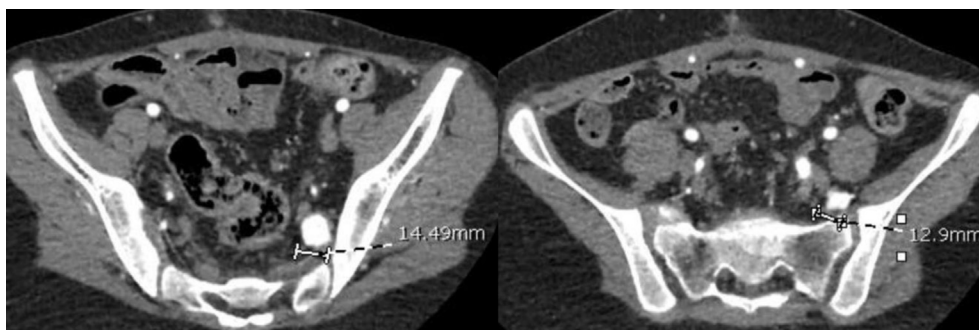
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**Fig 2.** Computed tomography angiography (CTA) of left internal iliac artery aneurysms (IIAAs).



**Fig 3.** Computed tomography angiography (CTA) of left axillary, brachial, and interosseus artery aneurysms.

Comprehensive Panel, Invitae Laboratory, San Francisco, Calif). These tests were negative for infectious or genetic conditions.

At the 2-month follow-up, a surveillance CTA of the torso was obtained demonstrating adequate repair of the AAA and absence of any aortic coarctation but multiple internal iliac artery aneurysms (IIAAs) were noted (Fig 2). The initial CTA was reviewed and only one of these aneurysms were present at that time and was markedly smaller. The patient also complained of a new painful swelling in the left antecubital fossa. A left arm CTA revealed a 9-mm axillary artery aneurysm, a 6-mm brachial artery aneurysm, and multiple aneurysms of a penetrating interosseus artery off of the ulnar artery (Fig 3). Review of the hospital record confirmed the arterial line was located in the right brachial artery. After undergoing total body imaging, he underwent repair of the IIAA, brachial artery aneurysm, and interosseus aneurysms.

The IIAAs were repaired via a left brachial artery approach. The left brachial artery was exposed proximal to the aneurysm and cannulated with a 5F sheath. A left iliac artery angiogram was obtained. Eight Penumbra Ruby coils (Penumbra Inc, Alameda,

Calif) were used to embolize the IIAAs. Completion arteriogram confirmed exclusion of the aneurysms (Fig 4).

The brachial artery aneurysm sac was dissected and involved the radial artery. The radial artery was ligated and the aneurysm sac resected. A saphenous vein patch angioplasty was performed with 7-0 polypropylene suture from the brachial artery onto the ulnar artery. The interosseus artery aneurysms were identified under duplex ultrasound and injected with 250 units of thrombin (Thrombin-JMI [Bovine], Pfizer, New York, NY). Color flow showed no flow confirming the aneurysms were fully thrombosed. The axillary aneurysm was not repaired as it was relatively small and asymptomatic. He was discharged the following day without complications.

A CTA was performed at 10 months showing no new aneurysm formation (Fig 5). He is scheduled for 3-month serial physical exams and magnetic resonance angiography with ultrasound surveillance of the axillary aneurysm. Should this aneurysm grow or become symptomatic, it will be repaired. Publication consent was obtained from the guardian during follow-up.



**Fig 4.** Left internal iliac artery angiogram with multiple aneurysms before and after coil embolization.

## DISCUSSION

AAA in children is rare and the initial presentation is often with rupture and death.<sup>1</sup> Most pediatric AAAs are associated with connective tissue disorders such as Marfan syndrome, Ehlers-Danlos syndrome, or tuberous sclerosis; vasculitis (ie, Henoch-Schonlein purpura); infection; or acquired from a trauma such as umbilical artery catheterization.<sup>2-5</sup> Congenital aneurysms are extremely rare with sizes reported between 1.6 and 11.0 cm.<sup>1,6-9</sup> The majority of reported congenital AAAs are diagnosed before the age of 2.<sup>1</sup>

Proposed etiologies of congenital AAA include defects in the transforming growth factor- $\beta$  signaling pathway and fibrillin-1. Transforming growth factor- $\beta$  controls cellular proliferation and differentiation and impacts connective tissue development. Fibrillin-1 is a glycoprotein component of microfibrils that provides structural support in elastic and nonelastic connective tissue. Defects in these connective tissues

can lead to aortic wall weakness and aneurysmal degeneration.<sup>10</sup>

Because pediatric AAA are so rare, there are no standardized treatment algorithms. Treatment options include conservative measures, surveillance with eventual repair, or immediate repair. If a pediatric AAA is associated with other congenital abnormalities resulting in a limited life expectancy, a conservative approach may be best. Conservative approaches can include steroids, cyclophosphamide, and antihypertensives. Other therapies include statins, antiplatelet drugs, and nonsteroidal anti-inflammatory drugs.<sup>1,11</sup> There are no currently published size criteria guidelines and the decision to surveil the aneurysm vs immediate repair must be made based on estimated risk of rupture.

Because the native pediatric aorta is so much smaller than the adult counterpart, there are no endovascular options. The majority of reported cases used nonautologous grafts.<sup>1,3-8</sup> Sacular diverticular aneurysms have



**Fig 5.** Ten-month follow-up of repaired abdominal aortic aneurysm (AAA) showing adequate repair without further aneurysmal degeneration.

been repaired with resection and patching.<sup>12</sup> When selecting the graft size, a considerable amount of oversizing should be encouraged to allow the graft to accommodate the growing native aorta. Additionally, as in our patient, there may be other nonaortic locations of aneurysmal degeneration. A full radiographic surveillance should be considered once a congenital aneurysm is discovered.

## CONCLUSIONS

We report an 11-year-old boy with no known genetic abnormalities who presented with a ruptured 11-cm AAA that was successfully repaired with an open bifurcated aortic graft.

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