Abdominal aortic aneurysms (AAAs) are rare in children. Although AAAs in adults are typically acquired from connective tissue disorders, cardiovascular pathologies, infection, or trauma, pediatric AAAs will usually be idiopathic, with <30 cases reported since 1967. Regardless, the risk of rupture carries high mortality, making a prompt and accurate diagnosis crucial. Although rare, AAAs have a known association with tuberous sclerosis (TS), and we have presented the case of a child who underwent successful surgery to repair an AAA.

CASE REPORT
A 3-year, 8-month-old girl with known TS and cardiac rhabdomyomas was admitted because of breakthrough seizures. On examination, a painless pulsatile abdominal mass was noted. Abdominal ultrasound (US) identified a large saccular AAA, 45 mm at its widest and 27 mm long, above the aortic bifurcation. The diameter of the normal aorta proximal to the aneurysm was 7 mm. Computed tomography (CT) showed the inferior mesenteric artery arising from the left of the AAA (Fig 1). Because of the risk of rupture, urgent surgical repair was arranged.

A midline incision was made to enter the peritoneal cavity. A large saccular infrarenal AAA was identified. Key vessels to the gonadal and renal system were identified and protected. The proximal neck of the AAA was controlled (Fig 2, A). After heparinization, the aorta and common iliac arteries were clamped. The aneurysm was opened longitudinally, revealing a thin layer of clots. The proximal aorta was beveled and anastomosed to a 12-mm Gelweave woven Dacron graft (Vascutek Ltd, Renfrewshire, UK), which was deliberately oversized and curved to allow for growth (Fig 2, B). We used 6-0 Prolene continuous suture, with reinforcement using pledgeted interrupted stitches. Care was taken to ensure the absence of unnecessary tension on the suture lines. The distal graft was anastomosed to the aorta above the bifurcation with slits to match the size.
The graft was covered by the partially original saccular AAA and partially raised peritoneum. An additional omental flap was not required. The inferior mesenteric artery was harvested and anastomosed to the side of the graft. Good colonic perfusion and pulsation of the common iliac arteries were ensured before closure. A polytetrafluoroethylene graft was not used for two reasons: (1) its porosity and tendency to bleed from the suture lines; and (2) the possibility of kinking due to bowing. Histologic examination showed a focal degeneration of media, disorganized collagen fibers, and chronic inflammation of media and adventitia.

Her postoperative recovery was unremarkable. Ultrasound was performed on the fifth postoperative day, before discharge, which demonstrated a successful repair. The child was examined in the clinic at 7 months after surgery with no concerning findings.

**Table.** Summary of case reports of tuberous sclerosis (TS)-associated abdominal aortic aneurysm (AAA) from 1985 onward

| Investigator            | Age at diagnosis | AAA location                        | Treatment                       | Complications and outcome                                                                 |
|-------------------------|------------------|-------------------------------------|---------------------------------|-------------------------------------------------------------------------------------------|
| Rolfes et al.\(^\text{8}\) 1985 | 9 months         | Extending from pelvis to near diaphragm | Synthetic graft                 | Proximal suture aneurysmal rupture resulting in death                                      |
| Van Reedt et al.\(^\text{9}\) 1991 | 5 years          | Infrarenal                          | Dacron Y-graft                   | No complications                                                                          |
| Tsukui et al.\(^\text{10}\) 1995 | 4 years          | Infrarenal                          | Tube graft                       | No complications                                                                          |
| Tamisier et al.\(^\text{11}\) 1997 | 2 years, 6 months | Infrarenal                          | PTFE graft                       | No complications                                                                          |
| Baker et al.\(^\text{12}\) 2000  | 1 year           | Infrarenal                          | AAA repair                       | No complications                                                                          |
| Jost et al.\(^\text{13}\) 2001  | 9 years          | Infrarenal                          | Dacron tube                      | No complications                                                                          |
| Kimura et al.\(^\text{14}\) 2005 | 2 years          | Thoracic to abdominal               | Artificial vessel replacement of aorta | No complications                                                                        |
| Wong et al.\(^\text{15}\) 2006  | 1 year           | Suprarenal and infrarenal           | Tube graft                       | Distal aortic suture aneurysm requiring open repair                                       |
| Moon et al.\(^\text{16}\) 2009  | 8 months         | Infrarenal                          | PTFE graft                       | No complications                                                                          |
| Salerno et al.\(^\text{17}\) 2010 | 2 years, 9 months | Mid-abdominal aorta, juxta renal   | Cadaveric graft                  | No complications                                                                          |
| Ye et al.\(^\text{18}\) 2012   | 1 year, 5 months | Infrarenal                          | Dacron Y-graft                   | No complications                                                                          |
| Dueppers et al.\(^\text{19}\) 2016 | 9 years          | Infrarenal                          | PTFE graft                       | No complications                                                                          |

PTFE: Polytetrafluoroethylene.
DISCUSSION

TS is a condition that typically presents with a triad of epilepsy, intellectual disability, and sebaceous adenoma. First described in 1880, TS has been noted to manifest across multiple systems including the cardiovascular system. TS has been associated with mutations in TS complex 1 or 2, which, in turn, affects smooth muscle cell proliferation and contractility. TS-related aortic aneurysms have been described in patients from a few months old to adulthood. A review by Salerno et al highlighted that most patients had presented before the age of 5 years and, more worryingly, that the mortality associated with ruptured aortic aneurysms was 29%.

The mainstay of treatment is surgery. Studies of the nonoperative management of children with aortic aneurysms, who were monitored closely with scans and had received antiplatelet or antihypertensive therapy, showed worse outcomes. Of the conservatively treated pediatric patients, 75% had died, with aneurysmal rupture causing 42% of these deaths. In contrast, 84% of the surgically managed patients had survived.

However, no uniform method has been determined for the surgical correction of AAAs. Younger pediatric patients will not be suitable for an endovascular approach, and only the open surgical approach has been reported. Dueppers et al analyzed the surgical outcomes of 11 pediatric patients, in addition to their own patients, with an average age of 3.5 years, similar to the age of our patient. They reported the use of various grafts, including Dacron grafts, with mostly good outcomes. They reported a postoperative complication due to dehiscence of a proximal anastomosis of an aortic graft at 15 months after surgery, highlighting the need for long-term follow-up.

It is also necessary to be cognizant of the growth potential of children. Grafts must be oversized and placed in a redundant curved shape to accommodate growth but could require subsequent surgical revisions as the child develops. Pediatric patients are smaller anatomically and their immature vascular system is more pliable to physiologic variations in pressure and flow. It is also necessary to be mindful of any comorbidities that could predispose the patient to complications of thrombosis, embolism, or stenosis. Thus, it is essential to consider the complexities of each case individually.

Aneurysmorrhaphy, a surgical technique in which the sac of the aneurysm is sutured closed to restore original luminal dimensions, has been considered as an alternative approach to grafts in the management of AAAs. However, using the defective vascular segment would predispose to AAA recurrence. Also, information on the use of aneurysmorrhaphy in the pediatric population is sparse. We found three published case reports, although only one had included long-term follow-up data. They had discussed the case of an 11-year-old boy who had undergone aneurysmorrhaphy for his AAA and had continued to have a good outcome 25 years later. However, the remaining two reports of aneurysmorrhaphy to treat an AAA in younger patients had not reported any long-term or subsequent follow-up findings. In addition, none of the three patients had had a TS-associated AAA.

We reviewed the data from other similar TS-associated AAA cases and found that grafts had provided good outcomes in most cases (Table).

CONCLUSIONS

TS-associated AAAs should be surgically corrected without delay to avoid the risk of aortic rupture and mortality. It is also necessary to allow for the child’s growth. Continued long-term follow-up is important to ensure the treatment’s suitability in the developing child.

REFERENCES

1. Le-Nguyen A, Johanfard S, Cote G, Borsuk D, Ghali R, Lallier M. Neonatal microsurgical repair of a congenital abdominal aortic aneurysm with a cadaveric graft. Eur J Pediatr Surg Rep 2021;9:e25-7.
2. Salerno AE, Marsenic O, Meyers KE, Kaplan BS, Hellinger JC. Vascular involvement in tuberous sclerosis. Pediatr Nephrol 2010;25:1555-61.
3. Cao J, Cong L, Guo DC, Mietzsch U, Kuang SQ, Kwartler CS, et al. Thoracic aortic disease in tuberous sclerosis complex: molecular pathogenesis and potential therapies in Tsc2+/− mice. Hum Mol Genet 2010;19:1908-20.
4. Criberi C, Meadors FA, Crawford ES, Coselli JS, Sati HJ, Svensson LG. Thoracoabdominal aortic aneurysm associated with umbilical artery catheterization: case report and review of the literature. J Vasc Surg 1992;16:75-86.
5. Dueppers P, Duran M, Grabitz K, Schelzig H. Open repair for abdominal aortic aneurysm in a young boy with tuberous sclerosis and review of the literature. Ann Vasc Surg 2017;39:286.e1-5.
6. Clark N, Brener BJ. Abdominal aortic aneurysm in a young child: a 25-year follow-up study. J Vasc Surg 1988;4:715-8.
7. Wang Y, Tao Y. Diagnosis and treatment of congenital abdominal aortic aneurysm: a systematic review of reported cases. Orphanet J Rare Dis 2015;10:4.
8. Rolfes DB, Towbin R, Bove KE. Vascular dysplasia in a child with tuberous sclerosis. Pediatr Pathol 1985;3:359-73.
9. van Reede Dortland RW, Bax NM, Huber J. Aortic aneurysm in a 5-year-old boy with tuberous sclerosis. J Pediatr Surg 1991;26:420-2.
10. Tsukui A, Noguchi R, Honda T, Tobita T, Fukuda S, Shimoji K. Aortic aneurysm in a four-year-old child with tuberous sclerosis. Paediatr Anaesth 1995;5:67-70.
11. Tamisier D, Coutière F, Sidi D, Vaksmann G, Bruneval P, Vouhe P, et al. Abdominal aortic aneurysm in a child with tuberous sclerosis. Ann Vasc Surg 1997;11:637-9.
12. Baker PC, Furrival RA. Tuberous sclerosis presenting with bowel obstruction and an aortic aneurysm. Pediatr Emerg Care 2000;16:255-7.
13. Jost C, Giovicikzi P, Edwards WD, Stanson AW, Joyce JW, Pairolero PC. Aortic aneurysms in children and young adults with tuberous sclerosis: report of two cases and review of the literature. J Vasc Surg 2001;33:659-42.
14. Kimura Y, Sugimura H, Toda M, Nakamura Y, Shibuya K, Murakami A, et al. A case of 2-year-old boy with tuberous sclerosis complicated with descending aortic aneurysm. Pediatr Int 2005;47:224-6.
15. Wong H, Hadi M, Khouny T, Geary D, Rubin B, Filler G. Management of severe hypertension in a child with tuberous sclerosis-related major vascular abnormalities. J Hypertens 2006;24:597-9.
16. Moon SB, Shin WY, Park YJ, Kim SJ. An abdominal aortic aneurysm in an 8-month-old girl with tuberous sclerosis. Eur J Vasc Endovasc Surg 2009;37:569-71.
17. Ye C, Yin H, Lin Y, Zhou L, Ye R, Li X, et al. Abdominal aortic aneurysms in children: single-center experience of six patients. Ann Thorac Surg 2012;93:201-5.