

An idiopathic congenital abdominal aortic aneurysm with impending rupture in a 23-month-old boy

Yong-Pil Cho, MD, PhD,^a Seong Chul Kim, MD, PhD,^b Sun A Kim, MD, PhD,^c Heungman Jun, MD,^a and Tae-Won Kwon, MD, PhD,^a *Seoul, Korea*

Abdominal aortic aneurysms are distinctly uncommon in infants and children. These aneurysms, which are idiopathic in nature without any definite predisposing factors, are exceedingly rare. We present the case of a giant idiopathic congenital infrarenal abdominal aortic aneurysm with impending rupture in a 23-month-old boy, which was successfully treated with surgical repair using a cryopreserved cadaveric allograft. To the best of our knowledge, this is the oldest case and the third successful treatment of an idiopathic congenital abdominal aortic aneurysm repaired with a cryopreserved allograft in infants and children. Continued follow-up with multimodality imaging is required. (J Vasc Surg 2013;57:508-10.)

Abdominal aortic aneurysms (AAAs) are extremely rare in infants and children, and are mostly associated with congenital cardiac or aortic malformations, systemic diseases, connective tissue disorders, and umbilical artery catheterization in newborns.¹⁻⁸ Less common etiologies include infection, vasculitis, autoimmune diseases, and trauma. The least common cause of AAAs is not associated with any definitive predisposing factors. These aneurysms, which are idiopathic in nature, involve all layers of the aortic wall.⁵ We report the case of an idiopathic congenital AAA with clinical manifestations caused by impending rupture in a 23-month-old boy, which was treated successfully with surgical repair using a cryopreserved cadaveric allograft.

CASE REPORT

A 23-month-old boy who had an uneventful antenatal and neonatal history was admitted to the hospital with a 1-day history of irritability, vomiting, and poor oral intake. He was born by normal, full-term, spontaneous vaginal delivery to a 32-year-old mother. His birth weight was 3.22 kg, and the umbilical artery was never cannulated. He had no family history of connective tissue disorders or aneurysmal disease.

On admission, clinical examination revealed diffuse tenderness, more intense on the left side, and palpable pulsating abdominal mass.

From the Division of Vascular Surgery^a and Division of Pediatric Surgery,^b Department of Surgery, and the Department of Pathology,^c University of Ulsan College of Medicine and Asan Medical Center.

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Reprint requests: Yong-Pil Cho, MD, PhD, Division of Vascular Surgery, Department of Surgery, University of Ulsan College of Medicine and Asan Medical Center, Asanbyeongwon-gil 86, Songpa-gu, Seoul 138-736, Korea (e-mail: ypcho@amc.seoul.kr).

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His initial blood chemistry values were within normal limits, except for a decreased hemoglobin level (8.3 g/dL). Contrast-enhanced axial computed tomographic scan revealed a giant infrarenal AAA with impending rupture. The aneurysm measured 8.4 cm in maximal diameter and 8.0 cm in length. It was located 1.1 cm below the origin of the renal artery and extended 2.3 cm proximal to the aortic bifurcation (Fig 1). In addition, small amounts of hemoperitoneum and hemoretroperitoneum and mild hydronephrosis of the left kidney compressed by the aneurysm were observed. Small punctuate calcifications were noted in the aortic wall. No other arterial abnormalities were identified.

An emergency exploratory laparotomy revealed a giant infrarenal AAA with impending rupture, extending about 2.0 cm proximal to the aortic bifurcation (Fig 2, A). The diameter of the proximal, normal infrarenal aorta was 0.7 cm. After the aorta and both common iliac arteries were isolated and cross-clamped, the aneurysm was opened and repaired with an interposition 0.7-cm cryopreserved adult cadaveric iliac artery from our tissue bank (Fig 2, B). A straight end-to-end anastomosis was performed using interrupted sutures with polypropylene 7-0. Histologic evaluation of the aorta revealed diffuse myxoid degeneration in the media without any evidence of vasculitis, suggestive of an uncharacterized congenital aortic degenerative change. A focus of clear cell aggregation was present in the adventitia. The clear cells showed no nuclear atypia and were negative for smooth muscle actin and S-100 protein, making the possibility of pericyte or melanocyte low. The biological significance of these cells was not clear. Further characterization of these clear cells was difficult based on the morphology and immunophenotype. Gene mutation analysis revealed normal sequence at the coding region of transforming growth factor- β receptor 1 and receptor 2 genes. Tissue culture did not grow any organisms.

The patient's postoperative course was uneventful. Follow-up contrast-enhanced axial computed tomographic scan confirmed complete exclusion of the diseased aorta and patency of both iliac and femoral arteries (Fig 3). Ten-month follow-up revealed patency to both leg arteries and no other complications.

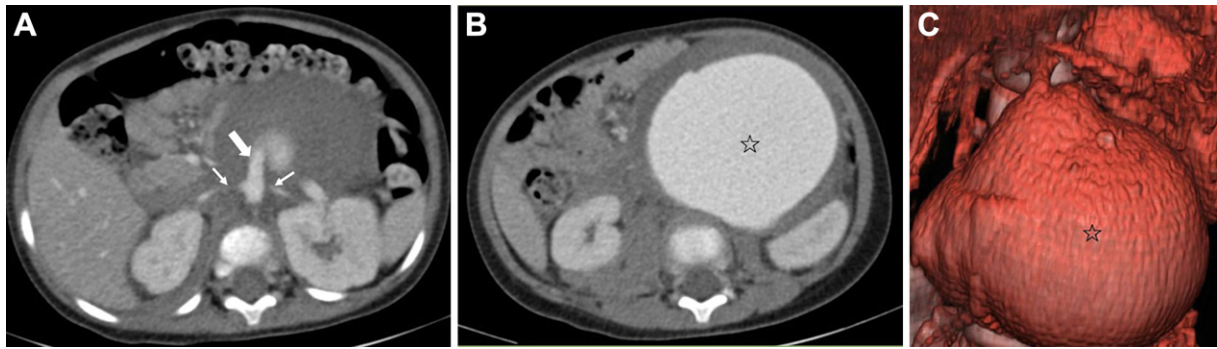


Fig 1. A and B, Initial contrast-enhanced axial computed tomographic image showing an 8.4 × 7.7 × 8.0-cm giant infrarenal abdominal aortic aneurysm (AAA) (*star*) with impending rupture. Note the neck of the aneurysm (*thick arrow*) and both renal arteries (*thin arrows*). C, Three-dimensional reformation with shaded surface display computed tomographic image showing the aneurysm (*star*).

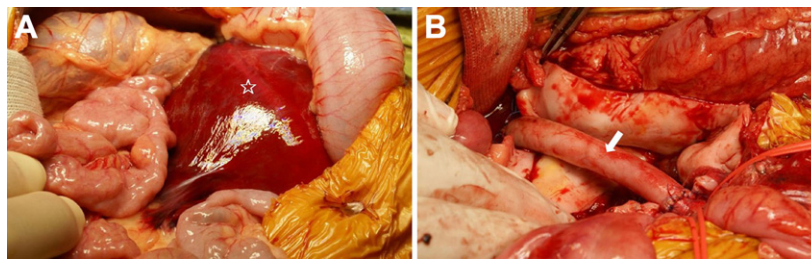


Fig 2. A, Intraoperative finding of a giant aneurysm (*star*) with impending rupture. B, Aorto-aortic I-graft interposition with a cryopreserved cadaveric allograft (*arrow*) was performed after aneurysmectomy.

DISCUSSION

Aneurysms of the abdominal aorta in infants and children are distinctly uncommon. They are mostly associated with definitive predisposing factors, such as infection, Ehlers-Danlos syndrome, Marfan syndrome, other connective tissue disorders, autoimmune diseases, Kawasaki disease, tuberous sclerosis, trauma, or umbilical artery catheterization.¹⁻⁸ An idiopathic congenital AAA is extremely rare. To date, only 16 cases have been reported in the English language literature; most were infrarenal (nine cases [56.3%]), prenatally diagnosed in seven and postnatally in nine.¹⁻⁸ An idiopathic congenital AAA is a distinct disease entity from acquired AAA. Although no clear conclusions have been drawn about related conditions or the etiology and prevalence of this condition because of the rarity of the disease, it is generally accepted that early elective repair is preferred to prevent rupture.⁷ Except for one unreported case, operative interventions were performed in nine cases (60%). Of these patients, seven with successful surgical repair showed uneventful postoperative recovery, and two with acute rupture did not survive operative intervention. Nonoperative management was performed in six cases (40%). Of these patients, three died during follow-up, one showed rapidly increased maximal diameter of the aneurysm (from 3.2 to 9.3 cm) at 8-month follow-up, one had progressive renal dysfunction,

and only one had stable aneurysm size at 6-month follow-up. Of the previous case reports, the patients were doing well after successful surgical repair at the time each case was reported, and early elective surgical repair was recommended to prevent rupture, although the investigators emphasized the future need for graft replacement as the patients grow older.⁴⁻⁶ No definite size criteria for surgical repair and no standard operative approach has been established.⁷ Furthermore, no studies have evaluated the long-term prognosis of idiopathic congenital AAA after surgical repair.¹⁻⁸

Additional considerations and concern are required when treating aneurysmal disease in infants and children. Considering that the average diameter of the adult aorta is 16 to 20 mm, use of a size-matched, 5- to 10-mm synthetic graft material for infrarenal AAA repair in infants and children likely will necessitate future intervention or revision as the children grow. Use of a larger synthetic graft material may impair optimal performance of growing children because of the diverging size discrepancy between the native aorta and the graft. However, no reported studies have documented long-term follow-up of infants and children with size-matched or larger synthetic grafts implanted in any position. Cadaveric allografts have been used successfully for the treatment of problematic mycotic aneurysm and infected synthetic vascular grafts in adults for a long

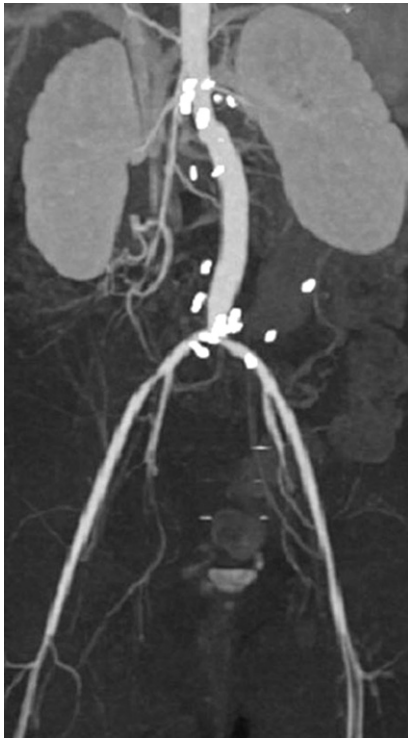


Fig 3. One-month follow-up contrast-enhanced axial computed tomographic image showing well-placed I-graft with no usual postoperative changes.

time, although controversy exists regarding the best durable cadaveric allografts, fresh or cryopreserved.^{4,8,9} Endothelial and fibroblast cells are considered likely sources of allograft antigenicity, which leads to gradual deterioration and degeneration of the graft used in vascular surgery. A decellularization process may reduce tissue antigen expression without reducing the strength of the allograft.^{4,8} Cryopreserved cadaveric allografts of the necessary length are readily available from tissue banks, and reduction of implant cellularity in these grafts may enable host recellularization of the matrix, which should have a favorable effect on long-term durability.^{4,8} For our patient, we chose a cryopreserved cadaveric allograft in the hope that its potential for cellular ingrowth might reduce late-term degeneration and have the potential for growth as the child grows. Only

careful, long-term follow-up will answer the question of these potential properties.

In our patient, the cause of the aneurysm is still unclear. Although we have not identified any genetic syndrome in this patient, the histologic findings were quite nonspecific. Given that our patient had no identifiable etiology for his aneurysm, we suspect that it was an idiopathic congenital AAA. The natural history of this AAA is not known. Although the extremely rare nature and lack of long-term follow-up reports of these lesions make the option of expectant treatment unclear in these children, it is generally accepted that early elective repair is preferred to prevent rupture. In our case, an idiopathic congenital AAA with impending rupture in a 23-month-old boy was successfully treated with surgical repair using a cryopreserved cadaveric allograft. Continued follow-up with multimodality imaging is required.

REFERENCES

1. 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.
2. Kim JI, Lee W, Kim SJ, Seo JW, Chung JW, Park JH. Primary congenital abdominal aortic aneurysm: a case report with perinatal serial follow-up imaging. *Pediatr Radiol* 2008;38:1249-52.
3. Cheung SCW, Khong PL, Chiu W, Metreweli C. Congenital abdominal aortic aneurysm and renal dysplasia. *Pediatr Radiol* 2004;34:827-30.
4. Bell P, Mantor C, Jacocks MA. Congenital abdominal aortic aneurysm: a case report. *J Vasc Surg* 2003;38:190-3.
5. Kim ES, Caiati JM, Tu J, Nowygrod R, Stolar CJ. Congenital abdominal aortic aneurysm causing renovascular hypertension, cardiomyopathy, and death in a 19-day-old neonate. *J Pediatr Surg* 2001;36:1445-9.
6. Mehall JR, Saltzman DA, Chandler JC, Kidd JN, Wells T, Smith SD. Congenital abdominal aortic aneurysm in the infant: case report and review of the literature. *J Pediatr Surg* 2001;36:657-8.
7. McAteer J, Ricca R, Johansen KH, Goldin AB. Extensive congenital abdominal aortic aneurysm and renovascular disease in neonate. *J Vasc Surg* 2012;55:1762-5.
8. Meyers RL, Lowichik A, Kraiss LW, Hawkins JA. Aortoiliac reconstruction in infants and toddlers: replacement with decellularized branched pulmonary artery allograft. *J Pediatr Surg* 2006;41:226-9.
9. Hüttel T, Balázs G, Entz L, Hüttel K, Bodor E, Szabolcs Z. Aortic coarctation reconstructed by resection and homograft replacement in the 1950s: long-term outcome 40 years after operation. *Ann Thorac Surg* 2002;73:1678-80.

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