Case Reports

Proximal Descending Thoracic Aortic Pseudoaneurysm

in a 24-Year-Old Man after 2 Childhood Repairs of Interrupted Aortic Arch

Saleh A. Alnasser, MD
Angela H. Martin, MD
Ourania A. Preventza, MD
Joseph S. Coselli, MD
Kim I. de la Cruz, MD

Improved management of interrupted aortic arch has increased long-term survival rates. Longer life expectancies in neonates and children surgically treated for interrupted aortic arch may necessitate complex reinterventions when sequelae develop in adulthood. We report the case of a 24-year-old man who had undergone initial repair of interrupted aortic arch type B at one week and reintervention at 6 years of age. He presented with a 5.5 × 9-cm pseudoaneurysm of the proximal descending thoracic aorta. He underwent surgical replacement of his distal aortic arch and proximal descending thoracic aorta, with a bypass to his left subclavian artery. In addition to our patient's case, we discuss considerations in treating recipients of early interrupted aortic arch repairs as they live longer and undergo multiple reinterventions. ***(Tex Heart Inst J 2020;47(1):27-9)***

Key words: *Aorta, thoracic/abnormalities/diagnostic imaging/surgery; aortic arch syndromes/surgery; cardiac surgical procedures; heart defects, congenital/complications/pathology; reoperation; treatment outcome*

From: *Divisions of Cardiothoracic Surgery (Drs. Alnasser, Coselli, de la Cruz, and Preventza) and Vascular Surgery (Dr. Martin), Michael E. DeBakey Department of Surgery, Baylor College of Medicine; and Division of Cardiovascular Surgery (Drs. Alnasser, Coselli, de la Cruz, and Preventza), Texas Heart Institute and Baylor–St. Luke's Medical Center; Houston, Texas 77030*

Address for reprints: *Saleh A. Alnasser, MD, Thoracic Surgery Unit, Surgery Department, King Saud University Medical City, 12372 6864 Riyadh, Saudi Arabia*

E-mail: *sal.alnasser@gmail.com*

Interrupted aortic arch (IAA), a rare congenital cardiac anomaly, is a separation between the ascending and descending aorta. It usually manifests itself as severe congestive heart failure in neonates. If uncorrected, IAA has a very poor prognosis; survival time can be as short as a few days. Typically, patients undergo early surgical reconstruction of the aortic arch, which is increasingly effective. However, sequelae can be expected as patients grow and live longer.

We present the case of a 24-year-old man with a twice-repaired IAA type B who presented with a large pseudoaneurysm of the proximal descending thoracic aorta. We discuss his treatment and present considerations for treating similar long-term survivors of childhood IAA repair.

Case Report

In October 2014, a 24-year-old man with a history of IAA type B presented with a pulsating bulge in his right neck, hoarseness, and a 20-mmHg blood pressure difference between arms. The patient had undergone repairs of the IAA at one week and at 6 years of age. The first repair involved placement of an 8-mm Dacron graft through a left thoracotomy, pulmonary artery banding, and closure of atrial and ventricular septal defects through a median sternotomy. During the second repair, for graft stenosis, a 16-mm Dacron interpositional graft was placed through a left thoracotomy.

At the current presentation, carotid duplex ultrasonograms showed normal results. A computed tomographic angiogram (CTA) revealed a descending thoracic aneurysm, a 5.5 × 9-cm pseudoaneurysm from the proximal to the distal anastomotic sites of the patient's 16-mm Dacron graft, and compression of the graft within the pseudoaneurysm (Fig. 1). We scheduled the patient for surgery.

We started total cardiopulmonary bypass and induced hypothermic circulatory arrest (body temperature, 18 °C). Arterial cannulation was done through the lower descending thoracic aorta; venous cannulation, through the left femoral vein with use of a long cannula. During circulatory arrest (duration, 27 min), the bypass pump flow rate was reduced to 2 L/min; cerebral perfusion was monitored with use of near-infrared spectroscopy and remained at baseline level throughout the period of low flow. We performed a left thoracotomy, removed the previous grafts, and replaced the distal aortic arch and proximal descending thoracic aorta with a 20-mm Dacron tube-graft. We used a separate 8-mm Dacron graft to bypass the left subclavian artery.

The proximal anastomosis was immediately distal to the left common carotid artery (LCCA), and the distal anastomosis was to the descending thoracic aorta just beyond the resected pseudoaneurysm.

Postoperatively, we reintubated the patient. He could not clear secretions because of left vocal cord palsy, and he had hypoxemia because of pulmonary contusions from difficult adhesiolysis during his third left thoracotomy. After left vocal cord injection, the patient recovered uneventfully. Before his discharge from the hospital on postoperative day 18, a chest CTA showed no residual aneurysms, leaks, or atypical findings (Fig. 2). Five years later, the patient was well and needed no further aortic reintervention.

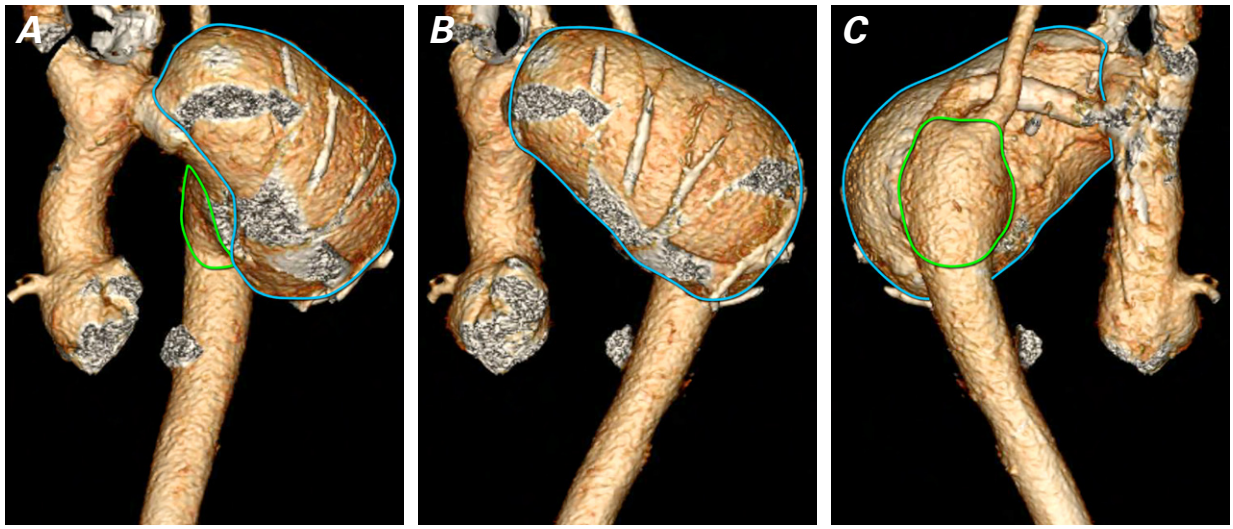

Fig. 1 *Computed tomograms (3-dimensional reconstruction) in **A**) anterior, **B**) lateral, and **C**) posterior views show a pseudoaneurysm (blue outline) and descending thoracic aneurysm (green outline).*

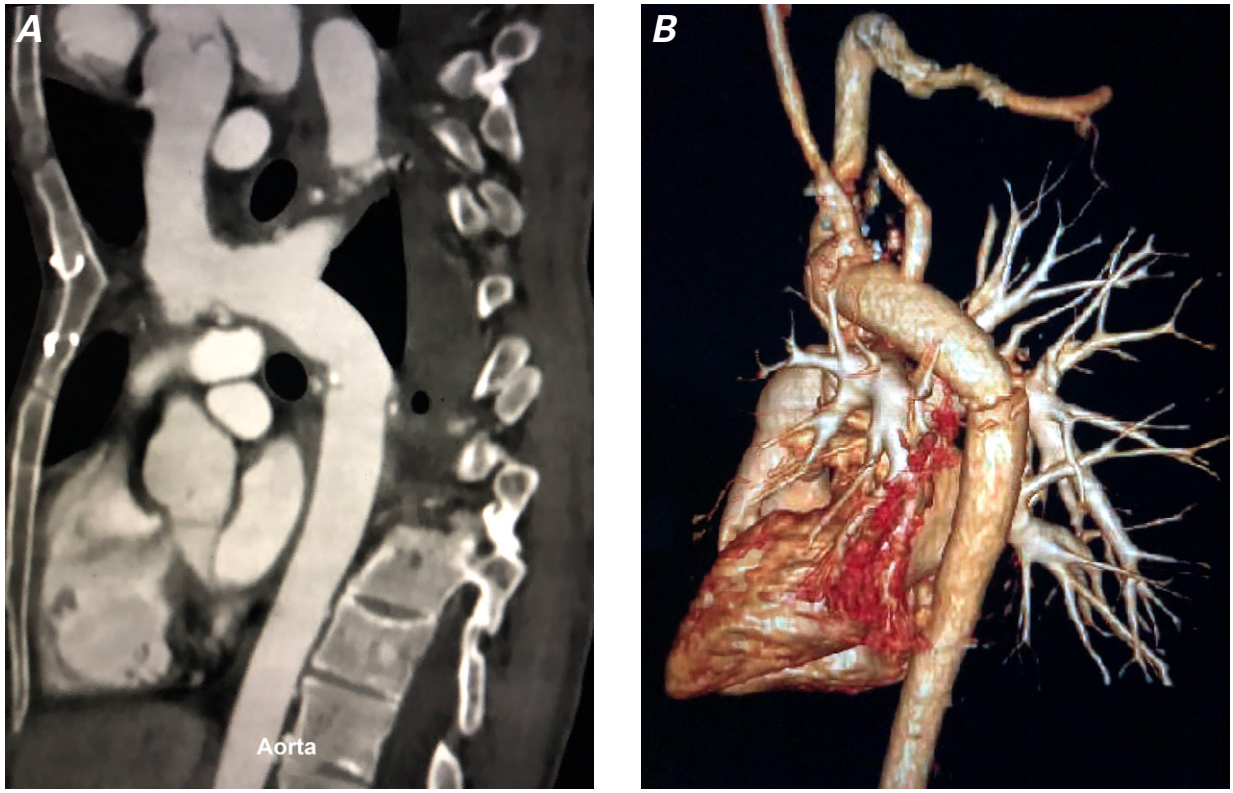

Fig. 2 *Computed tomograms in **A**) sagittal and **B**) 3-dimensional reconstruction views show the proximal descending thoracic aorta after pseudoaneurysm repair.*

Discussion

Interrupted aortic arch, which is usually associated with other congenital cardiac anomalies,[1] constitutes approximately 1% of congenital heart disease (incidence, approximately 3 per million live births).[2,3] The main pathologic finding is loss of anatomic communication between the ascending and descending aorta.

The classification of IAA depends on the site of aortic interruption. In type A, the arch interruption occurs distal to the origin of the left subclavian artery. In type B, the interruption occurs distal to the origin of the LCCA. In type C, the interruption occurs proximal to the origin of the LCCA. Interrupted aortic arch can occur with other congenital anomalies because of genetic mutations.[4,5] It usually manifests itself as severe congestive heart failure in the neonatal period, and 90% of affected neonates die at a median age of 4 days if the IAA is not treated.[6]

The prognosis for survival after surgical repair of IAA has improved. In an early patient series (1974–1987), the 10-year survival rate was 47%,[7] whereas in a more recent series (from 1993), the 5-year survival rate was 83%.[8] In 2009, authors of a review noted an early mortality rate of 5% to 10% in neonates undergoing IAA repair.[9] Variables associated with a higher mortality rate include IAA type B or C, female sex, low birth weight, truncus arteriosus, primary aortic anastomosis, concurrent complex anomalies, and earlier year of repair.[8,10-12] In addition, prolonged deep hypothermic circulatory arrest has been associated with higher mortality rates.[13]

More patients treated early for IAA are living longer, so more sequelae are seen. The Congenital Heart Surgeons' Society reports that risk factors for late reintervention include repair of truncus arteriosus, use of polytetrafluoroethylene material, and arch repair by any means other than direct anastomosis.[10] In our patient's previous repairs, 2 Dacron grafts of different sizes had been used. His first reintervention at 6 years of age was needed to treat graft stenosis; the current reintervention was needed to treat a large pseudoaneurysm.

In one study of 109 reinterventions, 57 were surgical and 52 were transcatheter.[10] We decided to treat our patient surgically because of his youth, the need for durable repair, and anatomic factors: the compression of the small Dacron graft by the pseudoaneurysm was better corrected openly. In conclusion, we think that expertise in both open and endovascular reintervention techniques will be necessary to deal with the range of sequelae likely to occur in long-term survivors of early IAA repair.

References

1. Gruber PJ, Epstein JA. Development gone awry: congenital heart disease. Circ Res 2004;94(3):273-83.
2. Kosucu P, Kosucu M, Dinc H, Korkmaz L. Interrupted aortic arch in a adult: diagnosis with MSCT. Int J Cardiovasc Imaging 2006;22(5):735-9.
3. Bayraktutan U, Kantarci M, Ceviz N, Yuce I, Ogul H, Sagsoz ME, Kaya I. Interrupted aortic arch associated with AP window and complex cardiac anomalies: multi detector computed tomography findings. Eurasian J Med 2013;45(1):62-4.
4. Jongmans MC, Admiraal RJ, van der Donk KP, Vissers LE, Baas AF, Kapusta L, et al. CHARGE syndrome: the phenotypic spectrum of mutations in the CHD7 gene. J Med Genet 2006;43(4):306-14.
5. Corsten-Janssen N, Kerstjens-Frederikse WS, du Marchie Sarvaas GJ, Baardman ME, Bakker MK, Bergman JE, et al. The cardiac phenotype in patients with a CHD7 mutation. Circ Cardiovasc Genet 2013;6(3):248-54.
6. Collins-Nakai RL, Dick M, Parisi-Buckley L, Fyler DC, Castaneda AR. Interrupted aortic arch in infancy. J Pediatr 1976;88(6):959-62.
7. Sell JE, Jonas RA, Mayer JE, Blackstone EH, Kirklin JW, Castaneda AR. The results of a surgical program for interrupted aortic arch. J Thorac Cardiovasc Surg 1988;96(6):864-77.
8. Oosterhof T, Azakie A, Freedom RM, Williams WG, McCrindle BW. Associated factors and trends in outcomes of interrupted aortic arch. Ann Thorac Surg 2004;78(5):1696-702.
9. Tlaskal T, Vojtovic P, Reich O, Hucin B, Gebauer R, Kucera V. Improved results after the primary repair of interrupted aortic arch: impact of a new management protocol with isolated cerebral perfusion. Eur J Cardiothorac Surg 2010;38(1):52-8.
10. McCrindle BW, Tchervenkov CI, Konstantinov IE, Williams WG, Neirotti RA, Jacobs ML, et al. Risk factors associated with mortality and interventions in 472 neonates with interrupted aortic arch: a Congenital Heart Surgeons Society study. J Thorac Cardiovasc Surg 2005;129(2):343-50.
11. Brown JW, Ruzmetov M, Okada Y, Vijay P, Rodefeld MD, Turrentine MW. Outcomes in patients with interrupted aortic arch and associated anomalies: a 20-year experience. Eur J Cardiothorac Surg 2006;29(5):666-74.
12. Shinkawa T, Jaquiss R, Imamura M. Single institutional experience of interrupted aortic arch repair over 28 years. Interact Cardiovasc Thorac Surg 2012;14(5):551-5.
13. Fulton JO, Mas C, Brizard CP, Cochrane AD, Karl TR. Does left ventricular outflow tract obstruction influence outcome of interrupted aortic arch repair? Ann Thorac Surg 1999;67(1):177-81.