

Endovascular repair of a Kommerell's diverticulum in a patient with a left-sided aortic arch and right-sided descending thoracic aorta

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Thoracic aortic anomalies are rare and may be associated with pathologic vascular conditions necessitating intervention. We present a case of a patient with a left aortic arch, right descending thoracic aorta, and a Kommerell's diverticulum giving rise to an aberrant right subclavian artery. The Kommerell's diverticulum was successfully managed with a right carotid to subclavian artery bypass and thoracic endograft exclusion. (*J Vasc Surg* 2009;49:1577-9.)

Several congenital anomalies of the aortic arch and great vessels have been extensively described.¹ Indications for surgical intervention may include symptoms manifesting from compression of adjacent structures or associated aneurysmal changes. The application of thoracic endografts to thoracic aortic anomalies is not commonly reported.

We present a case of a patient with a rare aortic anomaly. The patient was found to have a left aortic arch, right descending thoracic aorta, and an aberrant right subclavian artery (SCA) originating from a Kommerell's diverticulum. The diverticulum was successfully treated with a thoracic endograft following right subclavian artery revascularization and proximal ligation.

CASE REPORT

A 47-year-old white male with a history of hypertension and tobacco abuse presented to the Emergency Department with acute onset chest and interscapular back pain. The patient did not report a history of breathing or eating difficulties. Blood pressure was slightly elevated on initial assessment (145/96 mm Hg). He received oral nitroglycerine, oxygen, as well as intravenous metoprolol and morphine. Computed tomographic angiography (CTA) of the chest was performed to evaluate for aortic dissection.

Radiographic findings demonstrated anomalies of the thoracic aorta and great vessels consistent with a left aortic arch and right descending thoracic aorta. The ascending aorta arose from the left ventricular outflow tract and coursed to the left, anterior to the trachea in the usual anatomic fashion. In the upper thorax, the aorta turned abruptly to the patient's right and passed posterior to the esophagus (Fig 1). The thoracic aorta continued as a right-sided descending thoracic aorta. The first arch vessel originating from the aorta was the right common carotid artery (CCA), followed by the left CCA, and the left SCA. The last (fourth) aortic arch vessel was an aberrant right SCA arising from a Kommerell's

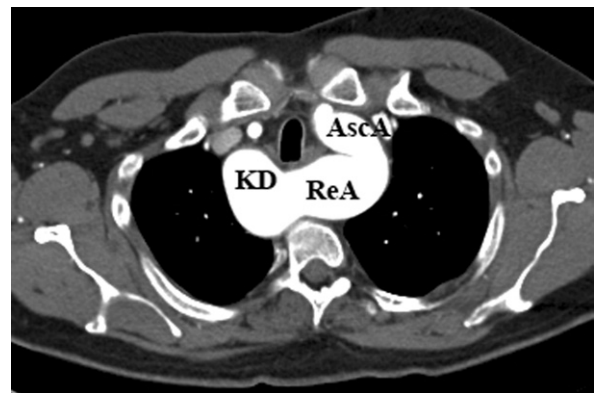


Fig 1. Computed tomographic angiography of the chest demonstrates the ascending aorta (AscA), retroesophageal (ReA) portion of the aortic arch, and Kommerell's diverticulum (KD).

diverticulum at the retroesophageal-descending thoracic aortic junction in the right chest (Fig 2). The transverse diameter of the Kommerell's diverticulum was 3.3 cm.

Cardiac enzymes and electrocardiogram (EKG) findings were normal. The patient's presenting symptoms completely resolved and were assessed to be unrelated to his aortic anomaly. He was discharged from the Emergency Department and scheduled for follow-up with our Vascular Surgery Division.

Due to the size of the Kommerell's diverticulum and potential for rupture, the patient elected to undergo thoracic aortic endovascular repair (TEVAR). A right CCA to right SCA bypass using an 8-mm polytetrafluoroethylene graft was performed immediately prior to the endovascular portion of the procedure due to the presence of a dominant right vertebral artery (VA) arising from the aberrant right SCA on preoperative cerebral arteriogram. Of note, magnetic resonance angiography was performed but inadequate in determining vertebral artery dominance. The proximal right SCA was ligated proximal to the origin of the VA.

The mid-portion of the patient's retroesophageal aorta measured 36 mm in diameter and was the planned location of the most proximal aspect of the proximal landing zone. The aorta tapered to 33 mm immediately prior to the origin of the diverticulum, and the aorta distal to the diverticulum measured 35 mm in diameter.

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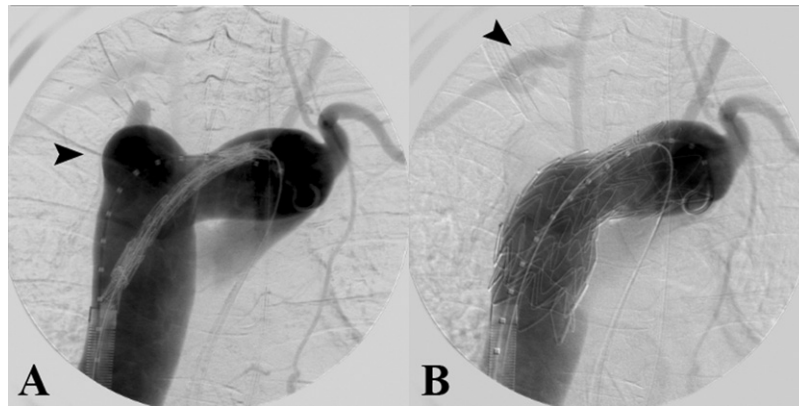


Fig 2. Intraoperative angiography during endovascular repair. **A**, The Kommerell's diverticulum (*arrowhead*) is illustrated during endograft advancement. **B**, Completion angiography demonstrates successful exclusion of the diverticulum and patency of the right common carotid to right subclavian artery bypass graft (*arrowhead*).

Vascular access was obtained through the left common femoral artery. A 40 mm × 10 cm Gore TAG Thoracic Endoprosthesis (W. L. Gore and Associates Inc, Flagstaff, Ariz) was advanced over a 0.035 inch Amplatz Super Stiff guidewire (Boston Scientific, Natick, Mass), but was unable to be advanced beyond the right SCA due to the abrupt angulation in the region of the retroesophageal-descending thoracic aortic junction. The device was withdrawn and the Amplatz guidewire was exchanged for a 0.035 inch Lunderquist Extra Stiff wire guide (Cook Inc, Bloomington, Ind), over which a 24F sheath was advanced into the mid-portion of the retroesophageal aorta. The endoprosthesis was then advanced through the sheath and deployed, excluding the diverticulum. Although the tapering of the aorta at the proximal landing zone created an oversizing of the TAG device at this location by 1 mm, a 40-mm device was utilized in order to obtain adequate device apposition to the aortic wall in the mid-portion of the retroesophageal arch. No device infolding was noted on completion angiography and follow-up imaging studies (Fig 2, B).

The patient's postoperative course was prolonged by persistent nausea and abdominal pain of unknown etiology. These symptoms slowly resolved permitting discharge on postoperative day 3. Six months after endovascular repair, CTA of the chest revealed no evidence of endoleak and the maximal transverse diameter of the Kommerell's diverticulum had decreased to 1.0 cm (Fig 3).

DISCUSSION

An aberrant right SCA is the most common congenital aortic arch anomaly (approximately 1% of the population).² Of patients with an aberrant right SCA, 60% have been reported to have an associated Kommerell's diverticulum.³ Aneurysmal dilatation of this diverticulum has been associated with rupture resulting in patient death,^{4,5} although definitive anatomic indications for intervention are unknown given the rarity of this anomaly. In a review of the literature by Cina et al,⁴ the mean size at which these aneurysms rupture was reported to be 5.8 cm, although this was observed to have occurred over a wide range of aneurysm diameters (2-10 cm).⁴ Due to the complexity associ-

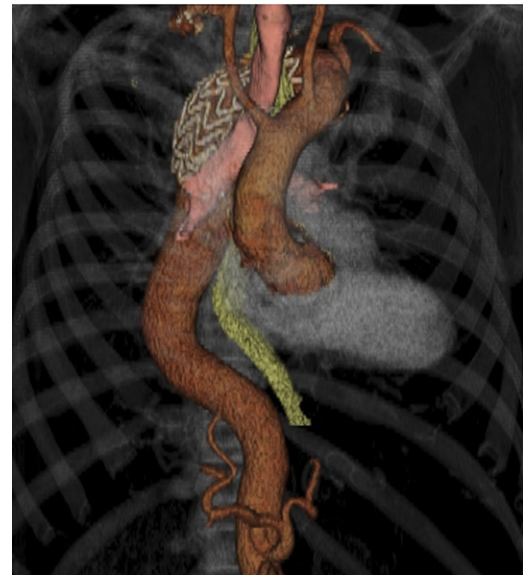


Fig 3. Computed tomographic angiography with three-dimensional reconstruction 6 months after thoracic endovascular aneurysm repair (TEVAR). Imaging demonstrates a left-sided aortic arch, retroesophageal aorta, and right-sided descending thoracic aorta in relation to the trachea/bronchi (*pink*) and esophagus (*yellow*).

ated with surgical treatment of larger lesions, the authors advocated treatment for these aneurysms when ≥ 3 cm. Given the poorly defined history of these lesions, it may be more appropriate to observe smaller lesions and base the decision for intervention on the presence or rate of growth over time.

A left-sided aortic arch with a right-sided descending thoracic aorta is a rare anomaly first reported by Paul in 1948.⁶ This aortic anomaly is a result of regression of the right aortic arch between the right CCA and the right SCA, with persistence of the distal end of the primitive right dorsal aorta as represented by the diverticulum.⁷ When a

right-sided ligamentum arteriosum or patent ductus arteriosus is present, transversing from the diverticulum to the right pulmonary artery, a vascular ring is formed. These patients may present with respiratory or swallowing difficulties early in life due to compression of the esophagus, trachea, and/or right mainstream bronchus.⁸⁻¹⁰ Division of the ligamentum arteriosum or patent ductus arteriosus through a right thoracotomy is necessary to relieve these symptoms when they develop. Our patient's aortic anomaly was detected relatively late in life compared to previously reported cases of this condition⁸⁻¹⁰ due to his absence of compressive symptoms.

Elective open surgical repair of these lesions has been reported to carry an operative mortality of 8-16%.^{4,5} TEVAR offers a minimally invasive intervention which may reduce the morbidity and mortality associated with the treatment of a wide range of thoracic aortic pathologies. The application of thoracic endografts for the treatment of congenital anomalies is less commonly reported. Chiesa et al¹¹ and Naoum et al¹² have both reported cases of TEVAR performed in patients with right-sided aortic arches and right-sided descending thoracic aortas. In both cases, an aneurysm appears to have involved a Kommerell's diverticulum from which an aberrant left SCA arose to course in a retroesophageal fashion. The applicability of TEVAR to complications arising from these and other congenital aortic anomalies appears to be limited to aneurysm exclusion as patients who present with compressive symptoms may necessitate conventional surgical resection. The durability of endografts in patients with thoracic aortic anomalies, who may present for intervention at a much earlier age than

patients with atherosclerotic aneurysms remains to be defined.

REFERENCES

1. Edwards JE. Anomalies of the derivatives of the aortic arch system. *Med Clin North Am* 1948;32:925-49.
2. Freed K, Low VH. The aberrant subclavian artery. *AJR Am J Roentgenol* 1997;168:481-4.
3. Davidian M, Kee ST, Kato N, Semba CP, Razavi MK, Mitchell RS, Dake MD. Aneurysm of an aberrant right subclavian artery: treatment with PTFE covered stentgraft. *J Vasc Surg* 1998;28:335-9.
4. Cinà CS, Althani H, Pasenau J, Abouzahr L. Kommerell's diverticulum and right-sided aortic arch: a cohort study and review of the literature. *J Vasc Surg* 2004;39:131-9.
5. Austin EH, Wolfe WG. Aneurysm of aberrant subclavian artery with a review of the literature. *J Vasc Surg* 1985;2:571-7.
6. Paul RN. A new anomaly of the aorta: left aortic arch with right descending aorta. *J Pediatr* 1948;32:19-29.
7. van Son JA, Konstantinov IE, Burckhard F. Kommerell and Kommerell's diverticulum. *Tex Heart Inst J* 2002;29:109-12.
8. Berman W Jr, Yabek SM, Dillon T, Neal JF, Akl B, Burstein J. Vascular ring due to left aortic arch and right descending aorta. *Circulation* 1981;63:458-60.
9. McFaul R, Millard P, Nowicki E. Vascular rings necessitating right thoracotomy. *J Thorac Cardiovasc Surg* 1981;82:306-9.
10. Whitman G, Stephenson LW, Weinberg P. Vascular ring: left cervical aortic arch, right descending aorta, and right ligamentum arteriosum. *J Thorac Cardiovasc Surg* 1982;83:311-5.
11. Chiesa R, Melissano G, Bertoglio L, Calliari F. Hybrid repair of an aortic arch aneurysm with complex anatomy: right aortic arch and anomalous origin of supra-aortic vessels. *J Vasc Surg* 2007;46:128-30.
12. Naoum JJ, Parenti JL, LeMaire SA, Coselli JS. Endovascular repair of a right-sided descending thoracic aortic aneurysm with a right-sided aortic arch and aberrant left subclavian artery. *Ann Thorac Surg* 2008; 85:1074-6.

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