Surgical strategy for Kommerell's diverticulum: Total arch replacement

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Objective: Kommerell's diverticulum is a rare congenital aortic arch anomaly. Various surgical techniques have been reported; however, the surgical strategy is still controversial. In our institute, total arch replacement (TAR) and anatomic reconstruction of the subclavian artery (SCA) has been selected for the treatment of Kommerell's diverticulum to release the vascular ring completely and prevent postoperative complications, including dissection, rupture, hand ischemia, and subclavian steal syndrome.

Methods: From 2000 to 2012, 4 patients (aged 38-72 years) underwent TAR and anatomic reconstruction of the SCA for Kommerell's diverticulum. All patients had a right aortic arch with an aberrant left SCA. The indications for surgery were dysphagia and dilatation of Kommerell's diverticulum. TAR, using hypothermic cardiopulmonary bypass and circulatory arrest, was performed through a median sternotomy and right anterolateral thoracotomy. After resection of Kommerell's diverticulum, the SCA was reconstructed with a graft.

Results: No hospital deaths or major complications occurred, but a 72-year-old patient required prolonged hospitalization for respiratory failure. All patients were discharged from the hospital and were free of symptoms, rupture, dissection, hand ischemia, and subclavian steal syndrome at 5 months to 11 years postoperatively.

Conclusions: TAR is a reasonable surgical technique for Kommerell's diverticulum, because it enables the vascular ring to be completely released, preventing recurrence, rupture, and dissection. Anatomic reconstruction of the SCA was effective to prevent hand ischemia and subclavian steal syndrome. (*J Thorac Cardiovasc Surg* 2014;148:1423-7)

In 1936, Kommerell¹ reported a case of an aberrant right subclavian artery (ARSCA) originating from the descending thoracic aorta of a left aortic arch. Kommerell's diverticulum is an aneurysm of the origin of an aberrant subclavian artery (ASCA) from the descending aorta. An ARSCA with a left aortic arch occurs in 0.4% to 2.0%; however, an aberrant left subclavian artery (ALSCA) with a right aortic arch (RAA) has been less common, occurring in approximately 0.1%.² In patients with an ARSCA with a left aortic arch, the diverticulum of the aorta is an embryologic remnant of the dorsal right fourth aortic arch, but in patients with ALSCA and a RAA, the diverticulum is an embryologic remnant of the dorsal left fourth aortic arch. The ASCA will be located behind the esophagus (80%), between the esophagus and the trachea (15%), or in front of the trachea (5%)³ and can cause symptoms that include dysphagia, dyspnea, stridor, wheezing, cough, choking spells, recurrent pneumonia, obstructive emphysema, and chest pain.⁴ Most

patients with ASCA will be asymptomatic; however, 6% of patients with a RAA will present with rupture and 53% with either rupture or dissection.⁵

A number of surgical techniques have been reported for Kommerell's diverticulum. These have included graft replacement of the ASCA and descending thoracic aneurysms with carotid-to-ASCA bypass and ligation of the SCA proximal to the origin of the vertebral artery,⁶ graft replacement of the descending aorta with reconstruction in situ or ligation of the ASCA,⁷ endoaneurysmorrhaphy with reconstruction of the SCA,⁴ and total arch replacement (TAR) with reconstruction of the SCA.^{8,9} Recently, endovascular treatment has been used as an alternative to surgical repair in selected patients.¹⁰⁻¹³

At our institute, TAR with hypothermic cardiopulmonary bypass (CPB) and circulatory arrest and reconstruction of the SCA through a median sternotomy and right thoracotomy have been used as standard treatment of Kommerell's diverticulum. This technique enables the vascular ring to be completely released, which resolves the symptoms, decreases the risk of dissection or rupture, and prevents the development of hand ischemia and subclavian steal syndrome.

METHODS

From 2000 to 2012, 4 patients (all men, aged 38-72 years) underwent TAR for Kommerell's diverticulum. All patients had a RAA with an ALSCA. The clinical characteristics of the patients are summarized in Table 1. The preoperative symptoms were dysphagia in 2 patients and

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Disclosures: Authors have nothing to disclose with regard to commercial support. Received for publication Aug 6, 2013; revisions received Nov 5, 2013; accepted for publication Nov 13, 2013; available ahead of print Dec 22, 2013.

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0022-5223/\$36.00

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<http://dx.doi.org/10.1016/j.jtcvs.2013.11.026>

Abbreviations and Acronyms

ALSCA	= aberrant left subclavian artery
ARSCA	= aberrant right subclavian artery
ASCA	= aberrant subclavian artery
CPB	= cardiopulmonary bypass
RAA	= right aortic arch
RSCA	= right subclavian artery
SCA	= subclavian artery
TAR	= total arch replacement

dizziness in 1. The remaining patient was asymptomatic. The maximum size of the aneurysm was 40 × 80 mm. The Tokyo Women's Medical University institutional review board approved study, and the need for patient consent was waived.

Operative Technique

The patient was placed in the supine position. The right hand was slightly deviated, and a towel pad was placed under the right scapula to keep the hemithorax elevated. Through a median sternotomy, 3 arch vessels and the distal portion of the ALSCA were exposed and taped. CPB was established by femoral artery and/or ascending aorta cannulation and bicaval drainage with a pulmonary artery vent. With the patient under deep hypothermia at a core temperature of 18°C, circulatory arrest with retrograde cerebral perfusion was induced. The aortic arch was reconstructed in the following order: right SCA, right common carotid artery, and left common carotid artery, using the arch first technique with a quadrifurcated graft (Triplex, Terumo, Tokyo, Japan).¹⁴ After reconstruction of the arch vessels, antegrade cerebral perfusion was resumed from the side arm of the graft. A right anterolateral thoracotomy in the third, fourth, or fifth intercostal space was added, without connecting the median sternotomy, and the Kommerell's diverticulum was exposed. After crossclamping, the descending aorta was transected at the level of the azygos vein and anastomosed to the distal end of the graft. Reperfusion and rewarming were started. The distal portion of the ALSCA was

interposed with the graft and anastomosed to the graft through the median sternotomy. Finally, the graft was anastomosed to the ascending aorta (Figure 1).

RESULTS

No hospital deaths or major complications occurred; however, a 72-year-old patient required prolonged hospitalization because of respiratory failure. All patients had been discharged from hospital, and all were still alive without any symptoms 5 months to 11 years postoperatively. The mean CPB, aortic crossclamp, and circulatory arrest time was 254.3 minutes (range, 194-306), 122.8 minutes (range, 87-163), and 47.0 minutes (range, 26-68), respectively. No patient developed dissection or rupture after surgery. Also, no patient developed hand ischemia or subclavian steal syndrome. In a 38-year-old patient with dysphagia, a preoperative 3-dimensional computed tomography scan revealed Kommerell's diverticulum, 25 mm in size, that was compressing the esophagus. After TAR, the esophagus was not compressed and the dysphagia had disappeared (Figure 2). The left SCA was reconstructed with a graft.

DISCUSSION

The indications for surgical repair have not been established because the number of patients with Kommerell's diverticulum has been small. Most patients with Kommerell's diverticulum will have no symptoms; however, Austin and Wolfe⁵ reported that 19% of patients presented with rupture, all of whom died. Cinà and colleagues⁴ reviewed the published data, including 32 patients, and reported that 6% of the affected patients presented with rupture and 53% with either rupture or dissection. The size of Kommerell's diverticulum cannot be used to predict rupture because of the limited data. Cinà and colleagues⁴ suggested aggressive treatment of aneurysms with a diameter of ≥3 cm in low-risk patients. Ota and colleagues⁷ recommended surgical repair for symptomatic aneurysms with a diameter of ≥5 cm. Backer and colleagues¹⁵ suggested that

TABLE 1. Patient characteristics

Characteristic	Pt. no.			
	1	2	3	4
Age (y)	57	55	72	38
Height (cm)	171	172	163	168
Weight (kg)	69	69	52	63
BSA (m ²)	1.81	1.55	1.55	1.72
Symptoms	Dizziness	None	Dysphagia	Dysphagia
Hypertension	Yes	No	Yes	No
Hyperlipidemia	Yes	No	Yes	No
Diabetes	No	No	No	No
Cerebrovascular disease	No	No	No	No
Renal insufficiency	No	No	Yes	No
Dialysis	No	No	No	No
Coronary artery disease	No	No	Yes	No
Maximum aneurysm size (mm)	50	80	20	25
Thoracotomy approach intercostal space	4th	5th	4th	3rd
Length of hospital stay (d)	50	55	291	26

Pt. no., Patient number; BSA, body surface area.

FIGURE 1. Image after repair.

FIGURE 2. A, Preoperative 3-dimensional computed tomography (3D-CT) scan from front showing right aortic arch. B, Preoperative 3D-CT scan from rear showing Kommerell's diverticulum, 25 mm in size. The aberrant right subclavian artery originated from the descending aorta. C, Preoperative 3D-CT reconstruction of aorta, esophagus, and trachea demonstrating compression of the esophagus by Kommerell's diverticulum. D, Postoperative 3D-CT scan showing the absence of compression of the esophagus after surgery.

Kommerell's diverticulum in children should be resected if the diverticulum is a widening of the base of the SCA that is >1.5 times the diameter of the distal SCA.

A number of surgical approaches to Kommerell's diverticulum have been proposed, including median sternotomy alone,⁸ right thoracotomy alone,^{4,7} left thoracotomy alone,⁶ bilateral thoracotomy,¹⁶ and median sternotomy plus thoracotomy, such as we have reported. Kai and colleagues⁸ reported TAR for an ALSA with a RAA through a median sternotomy alone. Kommerell's diverticulum was completely transected, and open distal anastomosis was performed with an elephant trunk technique under limited exposure. However, a distal anastomosis might be difficult to perform if Kommerell's diverticulum is located in the lower portion of the descending aorta or involves a large segment of the aorta. Our approach for the RAA using a median sternotomy plus right thoracotomy provided excellent exposure for distal anastomosis of the descending aorta through the right thoracotomy and of the aortic arch and the proximal portion of the ALSA through the median sternotomy. However, the approach using median sternotomy plus right thoracotomy could have disadvantages, including infection, postoperative pain, and respiratory failure. One of our patients, a 72-year-old man, required prolonged hospitalization because of respiratory failure.

The management of Kommerell's diverticulum depends on its anatomy and size. Cinà and colleagues⁴ used endoaortic aneurysmorrhaphy for a small Kommerell's diverticulum

with a normal descending thoracic aorta and graft replacement for a large Kommerell's diverticulum or Kommerell's diverticulum associated with an aneurysm of the descending thoracic aorta. In children with a vascular ring with a RAA and ALSA, division of the ligamentum arteriosum has been a standard operation; however, some of those patients had residual symptoms postoperatively and required reintervention.^{17,18} Primary resection of Kommerell's diverticulum and transfer of the ALSA to the left carotid artery and division of the ligamentum have been recommended to eliminate residual symptoms.^{15,17-19} Backer and associates¹⁵ hypothesized 3 mechanisms that create tracheal and esophageal compression in patients with Kommerell's diverticulum. The first mechanism is the space-occupying effect of Kommerell's diverticulum in the mediastinum. The second mechanism is the sling-like effect of the ALSA pulling the RAA toward the left side. This pulls the RAA against the trachea and esophagus. The third mechanism is the ALSA acting as a "bowstring" between the descending thoracic aorta on the right side and the arm on the left side, and this alone can be enough to compress the esophagus posteriorly. The resection technique for Kommerell's diverticulum reported by Backer and colleagues¹⁷ is to place a side-biting vascular clamp on the descending aorta at the origin of the Kommerell's diverticulum and perform primary closure with sutures or apply a polytetrafluoroethylene patch if the base of the Kommerell's diverticulum is friable. However, we have hesitated to perform this technique

in elderly patients, because they often have calcification of the aorta and have a risk of dissection with the use of a side-biting vascular clamp. Also, a small, but existent, risk of rupture owing to aneurysmal formation after surgery is present in the case of incomplete resection of Kommerell's diverticulum. Our strategy, TAR, with the patient under circulatory arrest with deep hypothermia, might be aggressive for treating Kommerell's diverticulum; however, the outcomes of TAR have improved during the past decade, even in elderly patients, with a low risk of perioperative stroke.²⁰ Circulatory arrest with deep hypothermia eliminates the need to clamp Kommerell's diverticulum, preventing perioperative stroke and dissection. In addition, TAR enables complete resection of Kommerell's diverticulum, completely correcting the 3 mechanisms proposed by Backer and colleagues.¹⁵

Various strategies for treating ASCA have been reported. In situ reconstruction of the SCA using a graft is a simple and useful technique⁷; however, a risk exists of dilatation of the residual ASCA in cases of incomplete aneurysm resection due to limited exposure. Aortoesophageal fistula caused by the graft contacting the esophagus is a rare complication, but its mortality has been quite high.^{21,22} Carotid-to-SCA bypass^{6,23} or transposition of the SCA on the carotid artery^{4,23} through a supraclavicular incision are alternative options for patients in whom direct continuity between the descending thoracic aorta and distal SCA cannot be easily established. Kamiya and colleagues²³ reported 3 different methods for ARSCA: complete resection of the ARSCA aneurysm through a left thoracotomy after reconstruction of the RSCA through a supraclavicular approach (2-staged operation, group 1), exclusion of the ARSCA aneurysm through a left thoracotomy without reconstruction of the RSCA (group 2), and closure of the distal site of the ARSCA aneurysm through a median sternotomy plus reconstruction of the RSCA. The ARSCA aneurysm was left as a blind sack (group 3). The patients in groups 1 and 3 had excellent outcomes after surgery; however, 1 patient in group 2 developed subclavian steal syndrome because the RSCA had not been reconstructed. In group 3, resection of the ARSCA aneurysm was attempted through a median sternotomy; however, the position of the ARSCA aneurysm made it too deep to resect. A risk exists of persisting symptoms and future rupture, because the aneurysm was left as a blind sac, which was not depressurized. Other investigators reported that patients with Kommerell's diverticulum developed subclavian steal syndrome after ligation of SCA and division of the ligamentum and required carotid-to-SCA bypass later.^{24,25} Our surgical strategy for ASCA has been anatomic reconstruction of the SCA with a graft placed to prevent the development of ischemia and subclavian steal syndrome.

Endovascular repair has been reported for selected patients with Kommerell's diverticulum.¹⁰⁻¹³ Attmann and

colleagues¹⁰ reported 2-stage surgical and endovascular treatment in a frail elderly patient with an ARSCA. Before endovascular treatment, left-sided carotid-to-SCA bypass was performed to secure the proximal landing zone. Endovascular repair is a valuable alternative to surgical repair using CPB, especially for ruptured Kommerell's diverticulum, because of the unstable hemodynamics.¹¹⁻¹³ However, the long-term outcomes are still unknown.

CONCLUSIONS

On the basis of our experience, TAR through a median sternotomy and right thoracotomy for Kommerell's diverticulum remains the preferred method of treatment, even though it requires deep hypothermic CPB and circulatory arrest. It provides excellent exposure and enables complete resection of Kommerell's diverticulum and anatomic repair of the ASCA. This resolves the symptoms without recurrence, decreases the risk of dissection and future rupture, and prevents the development of hand ischemia and subclavian steal syndrome.

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