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CASE REPORTS

Kommerell's diverticulum and aneurysmal right-sided aortic arch: A case report and review of the literature

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Right-sided aortic arch is a rare variant of the thoracic vascular anatomy that may be accompanied by an aberrant origin of the left subclavian artery. We report a true aneurysm of the distal arch and descending thoracic aorta in a patient with right-sided arch and review previous descriptions of aneurysms of anomalous right-sided aortas. In our patient, the left subclavian artery originated at the junction between the distal arch and the descending thoracic aorta located in the right chest and was aneurysmal (Kommerell's diverticulum); the thoracic aorta was also aneurysmal. Extra-anatomic left subclavian-to-carotid transposition was performed before the intrathoracic procedure. Subsequently, a right thoracotomy provided adequate exposure for repairing the aortic aneurysm and oversewing the aneurysmal origin of the subclavian artery. Because the distal aortic arch was involved, deep hypothermia and circulatory arrest were used. Only five previous instances of true aneurysms of a right-sided aortic arch have been reported; four of these patients underwent operative repair (via bilateral thoracotomy, median sternotomy, or right thoracotomy). We believe that a right thoracotomy provides good exposure and avoids the morbidity associated with bilateral thoracotomy. The reconstruction of the subclavian artery has not previously been reported in this setting. Performing subclavian reconstruction as an extrathoracic procedure before the intrathoracic repair would be expected to reduce the subsequent risk of distal ischemia or subclavian steal without increasing the overall morbidity associated with the procedure. (J Vasc Surg 2000;32:1208-14.)

A right-sided aortic arch is an anatomic variant occurring in approximately 0.1% of the population,[1] and in half of these cases the left subclavian artery is also aberrant.[2,3] These anomalies may be isolated or occur in combination with congenital heart defects.[4] We conducted a review of the literature and report one case of a Kommerell's diverticulum of an aberrant left subclavian artery in a patient with a right-sided aortic arch and a descending thoracic aortic aneurysm.

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Competition of interest: nil.

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CASE REPORT

A 61-year-old man was referred because of a dry cough of 3 years' duration and recent symptoms of dysphagia and hoarseness. His blood pressure was 130/90 mm Hg in both arms, and his heart rate was regular at 80 beats per minute. A prominent S4 and a faint, grade 2 left parasternal systolic murmur were audible. There were no carotid bruits; jugular venous pressure was 2 cm. Chest auscultation revealed stridulous inspiratory sounds, a few basal crackles, and expiratory wheezing. The remainder of the physical examination was unremarkable. A chest radiograph showed a large, well-circumscribed lesion in the superior mediastinum extending into both right and left sides but more pronounced on the right. The mass displaced the trachea anteriorly (Fig 1). Bronchoscopy revealed extrinsic compression of the trachea and of the right main stem bronchus. Computerized axial tomography with 3-mm sections demonstrated a right-sided

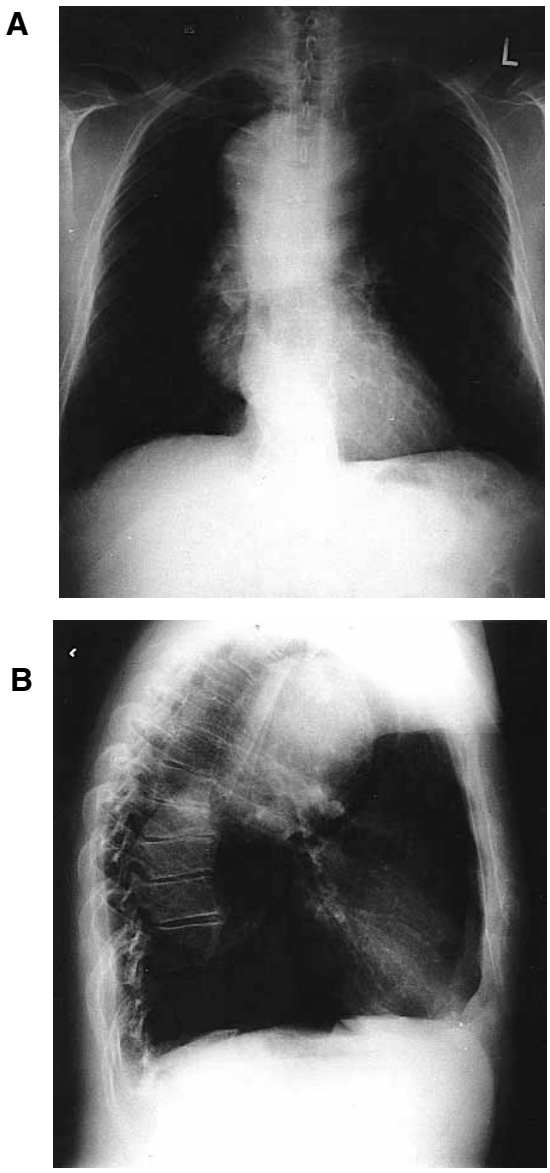

Fig 1. Chest radiograph. Large circumscribed mass projects on right and left hemithorax **(A)** and in posterior-superior mediastinum, with compression of trachea **(B)**.

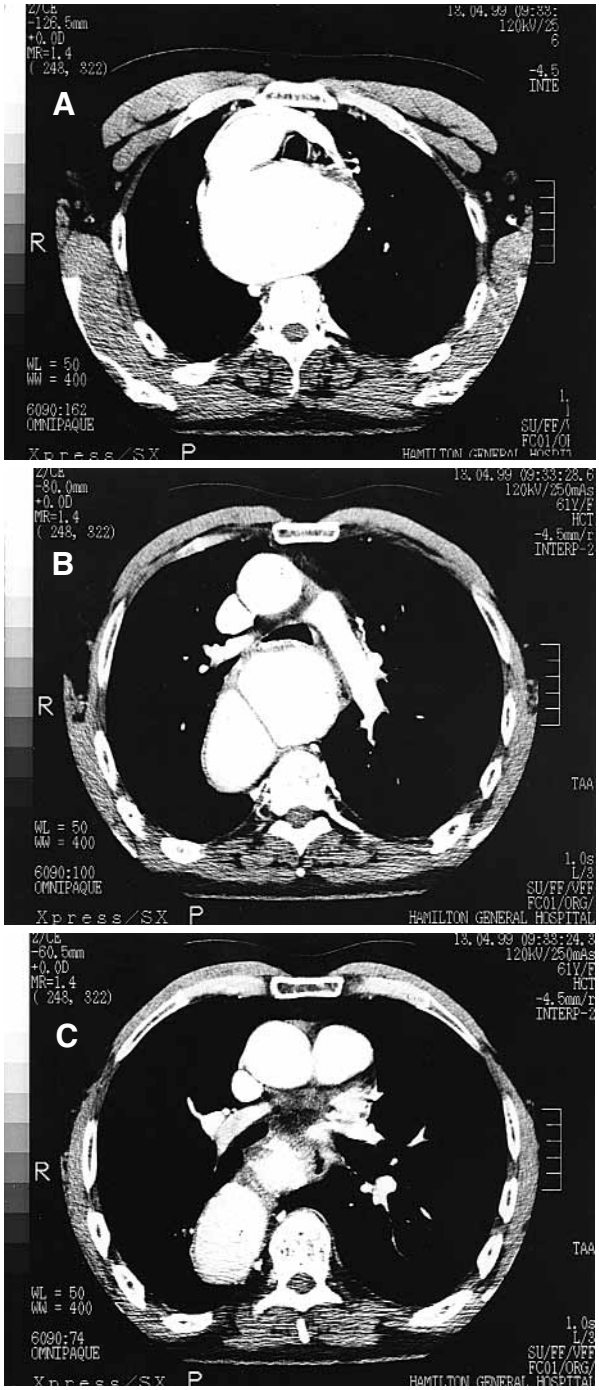

Fig 2. Computerized axial tomography shows large aneurysm in posterior mediastinum **(A)** with compression of right main stem bronchus **(B)** and aneurysm of descending thoracic aorta 4 × 6 cm in diameter **(C)**.

aortic arch and a right descending thoracic aorta with an aberrant left subclavian artery originating in the right chest. A 9.3-cm aneurysm involved the distal arch and the origin of the aberrant left subclavian artery (Fig 2). Angiography defined the ordering of the aortic arch branches as left common carotid, right common carotid, right subclavian, and left subclavian arteries from proximal to distal (Fig 3). Surgery was performed in two stages during the same operative procedure. With the patient in the supine position, a left subclavian-to-carotid transposition was done through a J-stick incision extending down the anterior border of the left sternocleidomastoid muscle and laterally 2 cm over the clavicle. In the second stage, the patient was positioned in the left lateral decubitus with the chest at 45° and the hips nearly horizontal. A right posterolateral thoracotomy was performed through the fifth intercostal space. A large

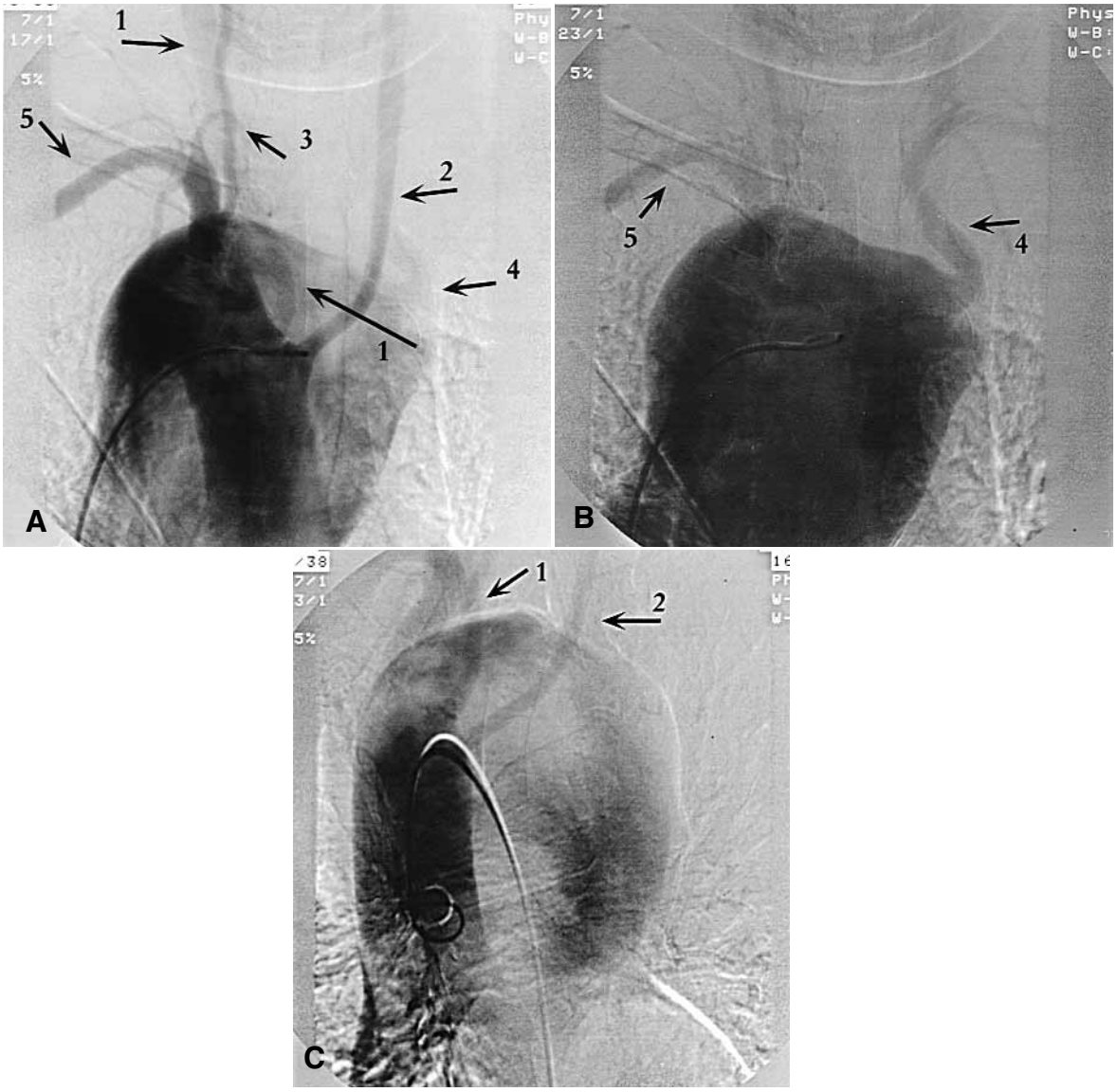

Fig 3. Angiography shows aneurysm of distal arch and origin of main aortic arch vessels. **A** and **B,** Anteroposterior projections of aortic arch at different times of contrast injection. **C,** Left anterior oblique projection. *1,* Right common carotid artery; *2,* left common carotid artery; *3,* right vertebral artery; *4,* left subclavian artery; *5,* right subclavian artery.

aneurysm of the aberrant origin of the left subclavian artery, involving the distal transverse arch and developing posteriorly into the left hemithorax, was found. The descending thoracic aorta was also aneurysmal to the level of T6. Femorofemoral partial cardiopulmonary bypass grafting with an oxygenator was initiated through a 25 French venous cannula and a 28 French arterial cannula. Hypothermia was induced and circulation arrested at a nasopharyngeal temperature of 25°C for 13 minutes. After the aneurysm was entered, the origin of the left subclavian artery (which had previously been ligated more distally at the time of the subclavian-to-carotid transposition) was oversewn. A 26-mm double velour woven Dacron (DuPont, Wilmington, Del) graft was cut with an appropriate bevel and anastomosed to the distal arch. With the patient in the Trendelenburg position, the arch and the prosthesis were filled with blood and a clamp was applied to the prosthesis distal to the anastomosis. The patient was then rewarmed to a temperature of 30°C and defibrillated to normal sinus rhythm. The intercostal arteries from T3 to T5 were suture ligated, and the graft was then anastomosed to the distal thoracic aorta (Fig 4). A follow-up angiogram at 6 months disclosed an intact repair with revascularization of all aortic arch branches (Fig 5).

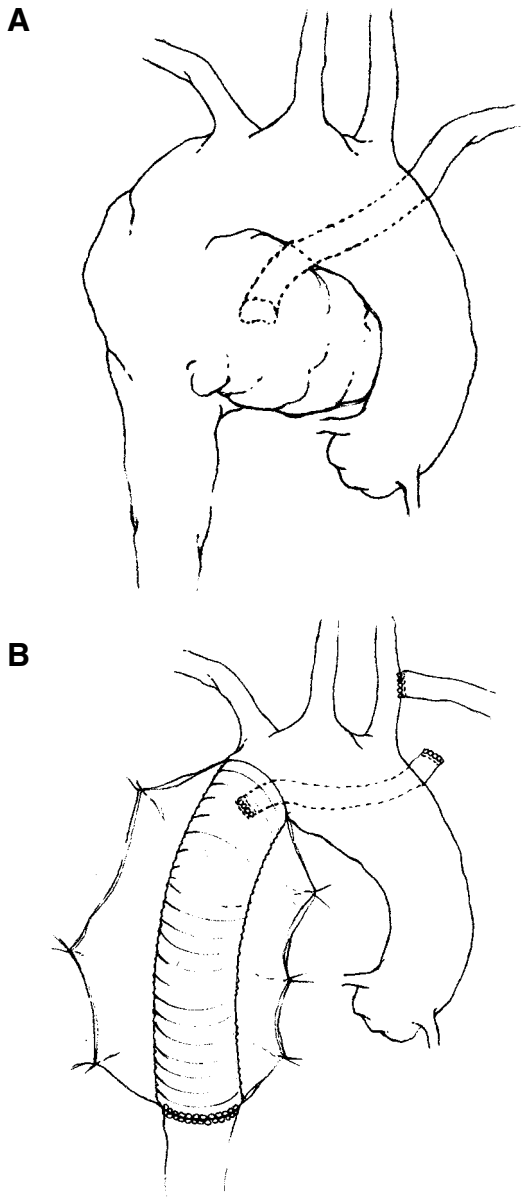

Fig 4. Drawing shows arch anomaly with aneurysm of aberrant left subclavian artery and aneurysm of descending thoracic aorta **(A)** and reconstruction after surgery **(B)**.

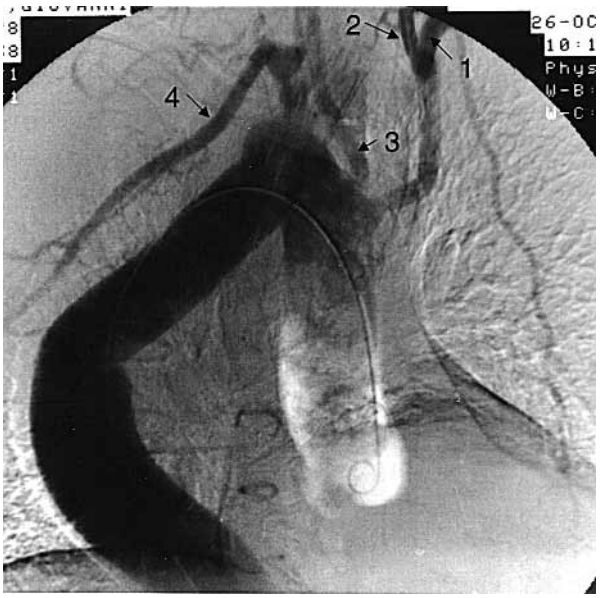

Fig 5. Follow-up angiogram at 6 months. *1,* Left subclavian artery; *2,* left carotid artery; *3,* right carotid artery; *4,* right subclavian artery.

DISCUSSION

A right-sided aortic arch is the result of an abnormal organogenesis of primitive aortic arches. Between the fourth and fifth weeks of embryonic life, blood leaves the heart by a single vessel, the truncus arteriosus, which divides into two branches, the ventral aortae. These are connected with the paired dorsal aortae by six branchial vessels, called *aortic arches*[5] (Fig 6). Segments of the first three arches, together with their dorsal and ventral aortic connections, form the carotid system. A segment of the right ventral aorta, the right fourth arch, and a portion of the right dorsal aorta develop into the right subclavian artery and the innominate artery. The left fourth arch persists as the adult aortic arch, and with the anlagen of the seventh dorsal intersegmental artery it forms the left subclavian artery. The fifth arches are both resorbed, and the sixth arches form the pulmonary artery and the truncus arteriosus (Fig 7).[4-7] The right-sided aortic arch results from persistence of the right fourth aortic arch and involution of the left (Figure 7, *B*). The right arch passes over the right main stem bronchus to the right of the trachea and esophagus. It is usually associated with involution of the left dorsal aorta and persistence of the right, causing the descending thoracic aorta to be located in the right hemithorax. If, instead, the right dorsal aorta disappears, the right-sided arch passes behind the esophagus to join the left dorsal aorta and the thoracic aorta descends in the left chest.[8]

Several classifications of these anomalies have been proposed.[4,6-10] The classification of Felson and Palayew[4] offers an anatomically sensible distinction between variants that are clinically different with respect to presentation, associations, and prognosis. Type 1, in which the left subclavian artery originates together with the left common carotid artery, or just proximal to it, is the result of the reabsorption of the left fourth aortic arch distal to the origin of the left

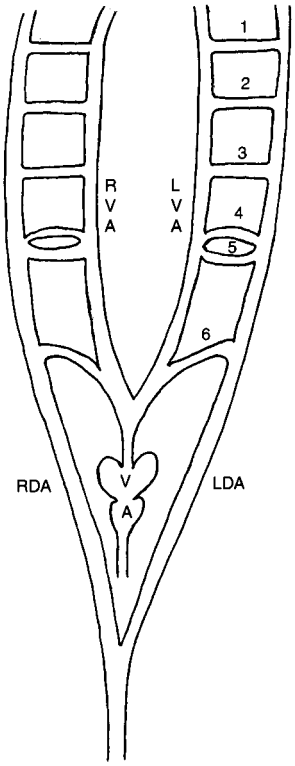

Fig 6. Schematic drawing shows circulation in 4-week human embryo. *1-6,* Primitive aortic arches; *RVA,* right ventral aorta; *LVA,* left ventral aorta; *RDA,* right dorsal aorta; *LDA,* left dorsal aorta; *V,* ventricle; *A,* atrium.

subclavian artery (brachiocephalic branches are the mirror image of normal anatomy). Cyanotic congenital heart disease (including tetralogy of Fallot, pulmonary stenosis with ventricular septal defects, tricuspidal atresia, and truncus arteriosus) occurs in approximately 75% of these patients. Type 2 is associated with an aberrant left subclavian artery arising either as the last branch of the right-sided aortic arch or from an aortic diverticulum, called *Kommerell's diverticulum*, that is a remnant of the left dorsal aortic arch (Fig 7, *B*). This occurs as a result of the reabsorption of the left fourth aortic arch proximal to the origin of the left subclavian artery.[10,11] Type 2 is more common than the type 1, and congenital heart anomalies are present in only 5% to 10%. The type 2 right-sided arch is the variant present in our patient. Because of the atherosclerotic changes that occur in the arterial wall during life, in adults it is generally not possible to distinguish a true diverticulum (Kommerell's diverticulum—ie, an embryonic remnant) from an acquired aneurysm of the origin of the aberrant subclavian artery.

Right-sided aortic arch may be asymptomatic. In infancy, symptoms are related to congenital heart anomalies or to compression of mediastinal structures such as the trachea or the esophagus. In adulthood, symptoms are more often the result of early atherosclerotic changes of the anomalous vessels, dissection, or aneurysmal dilatation with compression of surrounding structures causing dysphagia (*dysphagia lusoria*—"dysphagia by a trick of nature"), dyspnea, stridor, wheezing, cough, choking spells, recurrent pneumonia, obstructive emphysema, or chest pain.[4,12,13]

Indirect evidence of vascular anomalies of the aortic arch and its branches may be obtained with barium swallow, esophagogastroscopy, and bronchoscopy showing indentation of the esophagus or compression of the upper airways. Diagnostic techniques include computerized axial tomography, angiography, and magnetic resonance angiography.[3,14-19]

Definition of the anatomy of the aortic arch vessels may prove difficult even with angiography; occasionally, an incorrect diagnosis of aortic dissection has been made.[14] Multiple projections are often required, and some authors suggest a left transaxillary route for angiography to reduce problems with interpretation of findings.[14] However, the combination of thin-section computerized axial tomography and angiography is usually able to define vascular anatomy.

Reports of thoracic aortic aneurysms associated with a right-sided aortic arch were identified through a Medline database search from 1966 to November 1999 using Ovid software (Ovid Technologies, Inc, New York, NY) and the search strategy outlined in the Appendix. Reference lists of all relevant articles and reference lists of review articles were also examined. Nineteen cases of thoracic aortic aneurysms with a right-sided aortic arch have previously been reported; of these, 14 were associated with dissection[2,11,20-30] and five were not.[12,31-34]

Of the 14 cases with aortic dissection, nine were treated surgically.[2,11,20,21,23-25,27] The approaches used were as follows: right thoracotomy (six patients),[2,23-25,27] bilateral thoracotomy (one patient),[11] left thoracotomy with sternotomy (one patient),[20] and left thoracotomy (one patient).[21] The left subclavian artery was reconstructed in only one case, in which unusual anatomy permitted this to be approached through the right thoracotomy used for the aortic repair.[24]

Of the five patients with aneurysms without dissection, four underwent surgery; a variety of approaches have been described.[12,32-34] We feel that a left subclavian-to-carotid transposition completed before the thoracic approach revascularizes the subclavian distribution without increasing the complexity of the intrathoracic procedure. Caus et al[34] used

a wide anterolateral right thoracotomy through the bed of the fifth rib with transection of the sternum. In addition, an anterior left thoracotomy through the bed of the third rib exposed the left subclavian artery, though this artery was not reconstructed.

Median sternotomy has been described in two patients whose aneurysms were limited to the distal arch; in both of these cases the left subclavian artery was simply oversewn.[32,33] For more extensive aneurysms, right thoracotomy has been the preferred approach.[12] The exposure of the ascending, transverse arch, and descending thoracic aorta is adequate for reconstruction of these arteries and excision of a Kommerell's diverticulum, if present. However, the origin of the aberrant left subclavian artery is not easily accessible, particularly in the presence of a large aneurysm displacing this structure posteriorly and to the left. As with the approaches described, reconstruction of the left subclavian artery was not attempted in the report of the use of right thoracotomy. Extra-anatomic subclavian-to-carotid transposition has been previously described in the repair of aberrant right subclavian artery with Kommerell's diverticulum arising from a normal left-sided arch.[35] We believe that reconstruction of the left subclavian artery should be attempted; in young individuals this may prevent arm claudication, and older patients may avoid the possibility of subclavian steal syndrome. Subclavian-to-carotid transposition before intrathoracic repair is a practical way to achieve this without increasing the overall morbidity of the procedure. A bilateral thoracotomy[34] allows access to the second portion of the left subclavian artery and offers the possibility of reconstruction, but it is a more invasive procedure.

Depending on the exact anatomy, deep hypothermia (as used in our case) will not always be necessary, but it should be readily available for the management of unexpected intraoperative findings in the anomalous anatomy. Cardiopulmonary bypass grafting may be performed by cannulation of the right atrium. We preferred an approach from the right femoral vein using a long cannula with multiple side holes (BioMedicus wire wand, Medtronic, Minneapolis, Minn) advanced to the level of the atrium. The high flow rate achievable with this cannula permits venous access comparable with that of atrial cannulation without cluttering of the limited surgical field. If adequate venous return is not achieved, vacuum-assisted venous drainage can be used as an adjunct. In the presence of aortic insufficiency or if ventricular distension occurs, the left ventricle can be vented through one of the right pulmonary veins.

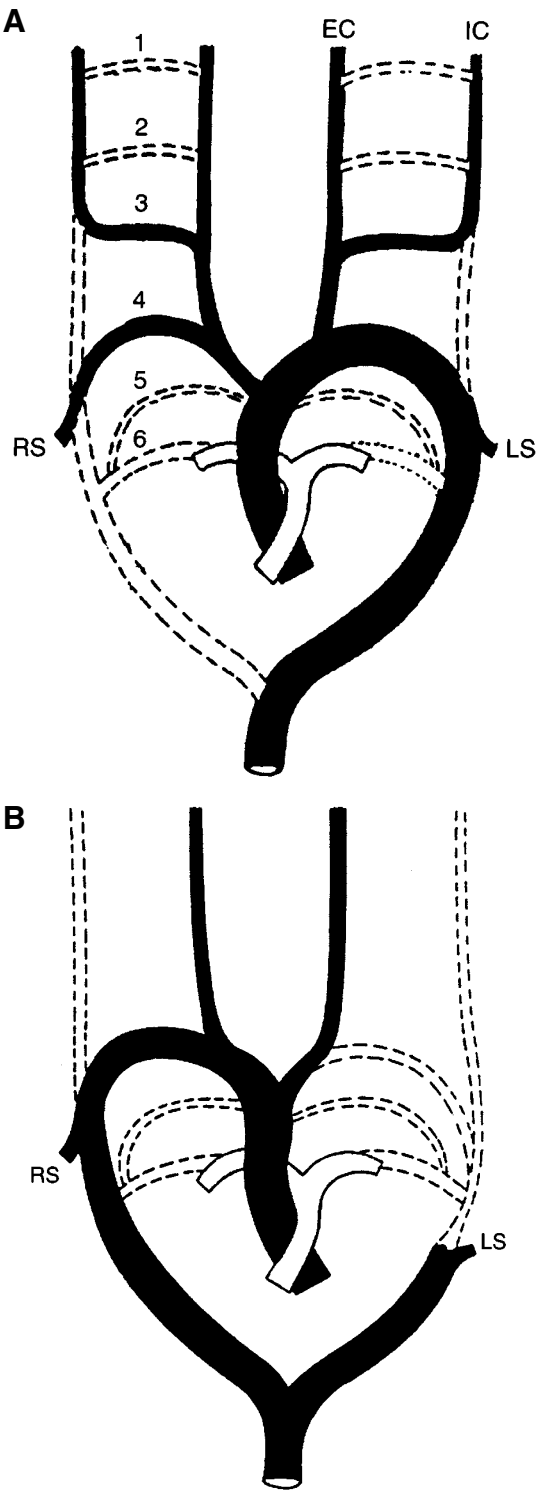

Fig 7. Development of left-sided aortic arch **(A)**. Development of right-sided aortic arch with aberrant left subclavian artery **(B)**. *1-6,* Primitive aortic arches; *RS,* right subclavian artery; *LS,* left subclavian artery; *EC,* external carotid artery; *IC,* internal carotid artery; *C,* common carotid artery.

CONCLUSION

Kommerell's diverticulum and right-sided aortic

arch are rare. An aneurysm involving the distal arch, the origin of the left subclavian artery, and the thoracic aorta may be repaired through a right thoracotomy. A left subclavian-to-carotid transposition before the thoracic repair allows a practical approach to reconstruction of the subclavian artery. Careful preoperative imaging and consideration of the individual anatomy in surgical planning are essential to a successful outcome.

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APPENDIX

Search strategy: [Aneurysm and thoracic aorta (Exploded MeSH headings)] and [right-sided arch or Kommerell or arch or aberrant left subclavian (text word) or thoracic aorta (abstract word)]