Tracheal Compression With "Hairpin" Right Aortic Arch: Management by Aortic Division and Aortopexy by Right Thoracotomy Guided by Intraoperative Bronchoscopy

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Background. **Four patients with severe tracheal obstruction due to right aortic arch, aberrant left subclavian artery, diverticulum of Kommerell, ligamentum or ductus arteriosum, and, additionally, right descending aorta, mild pectus excavatum, and high aortic arch apex, with narrow space between the ascending and descending aortic limbs, underwent division of ligamentum, excision of diverticulum and division (and reimplantation) of aberrant subclavian, either in multiple or single operations, but failed to achieve relief of obstruction.**

Methods. **In addition to the procedures noted, fabric sling aortopexy of ascending and descending aortic limbs around adjacent ribs, with or without aortic division after prosthetic graft between ascending and descending aortic limbs was required, all performed through a right thoracotomy and adjunctive cervical incision, and with flexible bronchoscopic monitoring of each step.**

Results. **Three patients obtained full relief of airway obstruction, which has persisted in follow-up from eight to over 12 years. One who had persistent severe tracheal malacia after prior tracheal resection and resultant chronic pulmonary sepsis died from these complications.**

Conclusions. **In this unusual subset of a rare vascular ring anomaly, radical methods were necessary for correction of airway obstruction after failure of prior conventional procedures.**

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Obstruction of the trachea and esophagus in the usual patient with right aortic arch and aberrant left subclavian artery (ALSA) responds to division of the vascular ring (left ligamentum arteriosum and ALSA), and further to excision of a diverticulum of Kommerell when present [1]. The aberrant subclavian artery is best anastomosed to the side of the left carotid artery (LCC). If an atretic left arch is present it is divided.

Three patients were treated, who failed to respond to varied prior divisions of ligamentum arteriosum or ductus, atretic arch, ALSA, and excision of aortic diverticulum (Kommerell). Two patients, with very severe airway obstruction, had also undergone subsequent tracheal resection in futile attempts to obtain relief. In one additional patient, who had not previously been operated upon, the vascular steps listed were carried out serially under bronchoscopic monitoring until demonstrable airway relief was achieved by aortopexy. Aortopexy was required in every case, and division of the aortic arch in three of the four. In the three patients operated upon previously, their obstructive symptoms had worsened after all prior procedures. Both patients who had previously undergone tracheal resection arrived with T-Y silicone stents in their airways.

All four patients had right aortic arch, ligamentum arteriosum, ALSA, and diverticulum of Kommerell (Fig 1A). In addition, all showed a degree of pectus excavatum with anteroposterior narrowing of the thorax, high apex of the aortic arch, and right descending aorta. This gave the aortic arch a "hairpin"-like configuration with little space between the two aortic limbs (Fig 2). Airway obstruction hence persisted there despite division of the vascular ring, including ALSA, and excision of the diverticulum. Sling aortopexy was necessary in all four patients, with aortic division in the three who had been operated upon previously.

Patients and Methods

Patient details are listed in Table 1. All four had suffered respiratory symptoms, including dyspnea on effort, low exercise tolerance, wheezing, and frequent respiratory infections; three also reported dysphagia initially. Two (patients 1 and 4) had severe respiratory limitation and two (patients 2 and 3) were completely incapacitated. As noted, three had undergone multiple operations previously without relief. These procedures are listed in Table 1.

Anatomical configurations were carefully outlined with computed tomographic scans in all patients (Fig 3).

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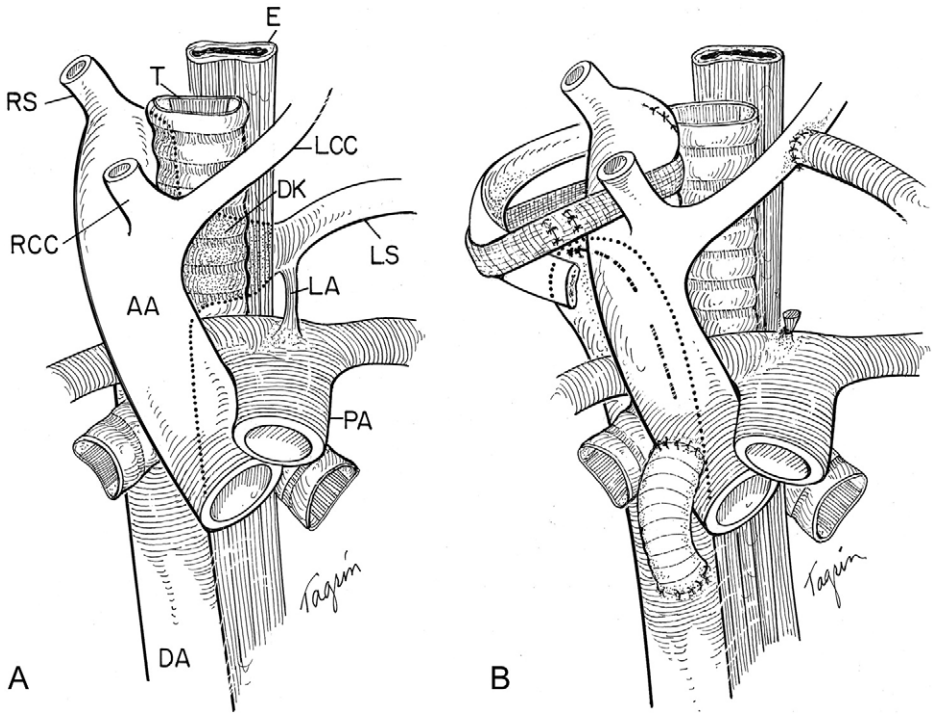

Fig 1. (A) Anatomic features. (AA = ascending aorta, right arch; DA = descending aorta, right side behind AA; DK = diverticulum of Kommerell; E = esophagus; LA = ligamentum arteriosum; LCC = left common carotid; LS = anomalous left subclavian; PA = pulmonary artery; RCC = right common carotid; RS = right subclavian; T = trachea.) (B) Surgical correction. Aortic arch divided beyond RS; lower suture line on DA at site of excision of DK; alternatively DK excision encompassed with excision of short segment of DA; Gore-Tex sling around AA limb and second rib; LS implanted into LCC; LA divided; Dacron graft from AA to DA below hilum.

Three had aortograms (Fig 2). Bronchoscopy showed pulsatile compression of the lower third of the trachea on the right anterior wall of up to 80% to 90% of the cross-sectional area by the ascending right aortic arch. Nearer the carina and at the takeoff of the right main bronchus the airway was also compressed posterolaterally on the right, with strong pulsations visible, by the diverticulum of Kommerell in the descending arch. The lumen between the two points of compression was "teardrop" shaped, with the wide portion to the left (Fig 4). The airway narrowed even further on expiration. Cartilaginous rings, although deformed, were identified in the affected tracheal segment in all except the eight-year-old patient who had undergone prior tracheal resection. Access for tracheal resection in this patient had been obtained by temporary division of the aortic arch. His lower trachea was now malacic and required stenting throughout his subsequent course.

Operations

Ventilatory anesthesia was provided by a single lumen flexible armored endotracheal tube placed bronchoscopically in the left main bronchus. A bronchial blocker was positioned in the proximal right main bronchus. Changes in the degree of airway occlusion were directly observed serially at each stage of operation by withdrawing the endotracheal tube into the upper trachea and inserting a flexible bronchoscope through a sealing adaptor. The endotracheal tube was then readvanced into the left main bronchus after each bronchoscopic observation.

The major portion of the procedure was performed through right thoracotomy by the bed of the shingled

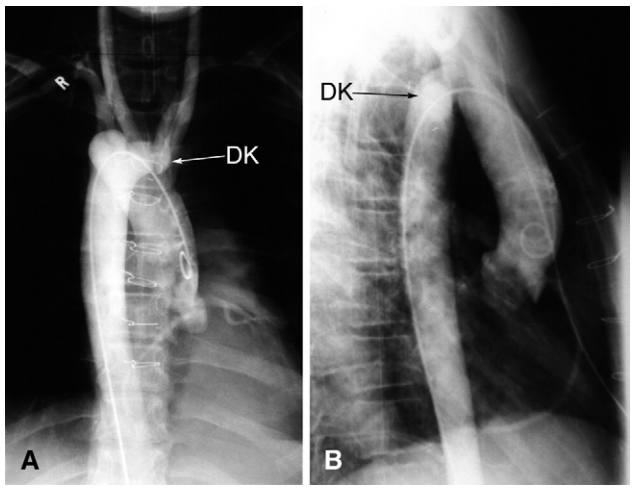

Fig 2. (A) Angiogram (anterior-posterior view) of patient 2 demonstrating right aortic arch, diverticulum of Kommerell (DK) and aberrant left subclavian artery. (B) Angiogram (lateral view) of patient (2) demonstrating "hairpin" aorta at apex of chest and location of DK.