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Vascular Anomalies Causing Tracheoesophageal Compression: A 20-Year Experience in Diagnosis and Management

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ABSTRACT

Background: Vascular rings and other congenital aortic arch anomalies may be major causes of tracheoesophageal obstruction in children. In this report, our diagnostic approach, surgical treatment, and early and late results for 30 patients are reported.

Methods: During a 20-year period (1982-2002), 30 children underwent surgery for tracheoesophageal compression caused by aortic arch anomalies. The median age at operation was 8 months (range, 36 days to 94 months), and the median patient weight was 8 kg (range, 2.4-16 kg). At 53.3% of cases, double aortic arch was by far the most common encountered cause of compression. Patients were admitted with respiratory distress, stridor, apnea, dysphagia, or recurrent respiratory tract infections. Diagnosis was established by barium esophagogram, computed tomography, magnetic resonance imaging, and angiography. The operative approaches were through a left thoracotomy or a median sternotomy.

Results: Operative mortality rate was 3.3%. Follow-up data from 2 months to 10 years (mean follow-up, 34 weeks) were available for all 30 patients. Twenty-six patients (86.7%) were essentially free of symptoms, 3 patients (10%) had residual respiratory problems, and 1 patient (3.3 %) had a gastroesophageal reflux problem.

Conclusion: These results suggest that surgical correction of symptomatic vascular rings can be performed with low mortality and morbidity rates.

INTRODUCTION

Anomalies of the aortic arch complex (vascular rings) occur in multiple forms, often as rings that cause tracheal obstruction and respiratory distress [Ruckman 1989]. Since the original description by Gross in 1945, the goal of surgery has primarily been the relief of airway symptoms [Gross 1945]. Clinical presentation and surgical approaches to aortic arch complex anomalies vary, depending on the specific lesion and the presence of associated congenital cardiac disease. The type of operation depends on the vascular anomaly and whether associated conditions such as cardiac defects or tracheomalacia are present.

The purpose of this report is to summarize our experience with the surgical correction of vascular rings and to report the patients' intermediate-to-late results.

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MATERIALS AND METHODS

Thirty patients underwent surgical treatment from 1982 to 2002 for the relief of tracheoesophageal compression by vascular anomalies. The clinical records were retrospectively reviewed to obtain specific data. In particular, sex, age, type of vascular rings, symptoms, results of physical examinations, diagnostic tests, type of operation, postoperative complications, long-term results, and associated anomalies were recorded for each patient. Patient age at the time of operation ranged from 36 days to 94 months (mean, 8 months). Seventeen patients (56.7%) were female, and 13 (43.3%) were male. The weight range was 2.4 kg to 16 kg (median, 8 kg). The following anomalies were present in our series: double aortic arch in 16 patients (53.3%), right aortic arch with left ligamentum arteriosum or patent ductus arteriosus in 7 patients (23.3%), right aortic arch with aberrant left subclavian artery in 3 patients (10%), left aortic arch with aberrant right subclavian artery in 2 patients (6.7%), right aortic arch and left ligamentum arteriosum with retroesophageal left innominate artery in 1 patient (3.3%), and left pulmonary artery sling in 1 patient (3.3%) (Table). Also occurring were Kommerell's aortic diverticulum in 4 patients (13.3%), patent ductus arteriosus in 7 patients (23.3%), ventricular septal defect in 2 patients (6.7%), atrial septal defect in 1 patient (3.3%), and dilated cardiomyopathy in 1 patient (3.3%).

Clinical Manifestations

The most common symptom was stridor or noisy respiration. Respiratory distress was precipitated by crying or feeding. There were recurrent respiratory tract infections. Physical examination revealed significant findings in more than 80% of patients. These findings included rhonchi, stridor, retractions, tachypnea, wheezing, and rales.

Diagnostic Procedures

A chest roentgenograph was obtained for all patients and showed abnormalities in more than 70% of these children.

Subtypes of Aortic Arch Anomalies

Anomaly	No. of Patients
Double aortic arch	16 (53.3%)
Right aortic arch with left ligamentum arteriosum or patent ductus arteriosus	7 (23.3%)
Right aortic arch with aberrant left subclavian artery	3 (10%)
Left aortic arch with aberrant right subclavian artery	2 (6.7%)
Right aortic arch and left ligamentum arteriosum with retroesophageal left innominate artery	1 (3.3%)
Left pulmonary artery sling	1 (3.3%)

Hyperaeration of the lung fields was seen and was sometimes unilateral, especially in the patient with pulmonary vascular sling. Varying degrees of atelectasis and pneumonic infiltration were present.

The most reliable procedure for the diagnosis of vascular rings was the barium esophagogram (Figure 1), which showed indentations on the esophagus. A bronchoscopic study was performed in 15% of the patients. Bronchoscopy demonstrated varying degrees of tracheal narrowing. Angiography was performed in all patients (Figure 2). For 3 patients with intracardiac defects (2 with ventriculoseptal defect, 1 with atrial septal defect), hemodynamic studies indicated no surgical intracardiac correction. Angiographic assessment clearly defined the vascular anatomy with the origins and respective courses of the great arteries.

Operative Findings and Management

The approach to this lesion in all but 1 patient was a left thoracotomy. The vascular ring caused by the double aortic arch was released by dividing the lesser of the 2 arches, usually at the atretic area; when the arches were patent, the lesser arch was divided between clamps at a site selected to preserve the brachiocephalic blood flow. Aberrant left or right subclavian arteries were divided. The left ligamentum arteriosum or the patent ductus arteriosus was always divided, and careful dissection around the esophagus and the trachea was performed to remove any residual adhesive bands. The recurrent laryngeal and phrenic nerves were identified and protected. Kommerell's diverticulum was resected to avoid the recurrence of dysphagia by compression. In the case of pulmonary vascular sling, the repair was performed through a midline sternotomy with the use of extracorporeal circulation (Figure 3). The entire course of the left pulmonary artery was dissected out. After the left pulmonary artery was divided at its origin, it was passed anteriorly to the trachea and reimplanted to the left lateral aspect of the main pulmonary artery.

RESULTS

All but 1 patient survived, and recovery was straightforward in most cases. Some patients, however, needed several weeks for complete remission of their initial symptoms. One patient who had dilated cardiomyopathy died on the third postoperative day.

The early mortality rate was 3.3%. Follow-up data from 2 months to 10 years (mean follow-up duration, 34 months) were available for 30 patients. Twenty-six patients (86.7%) were essentially free of symptoms, 3 patients (10%) had residual respiratory problems, and 1 patient (3.3%) had a gastroesophageal reflux problem. There was no late mortality. Three patients who had been operated on for a vascular ring with intracardiac defect were still in follow-up treatment at the time of this report.

DISCUSSION

The incidence of vascular anomalies of the aortic arches and pulmonary artery system has been estimated to be as high as 3% of all cardiovascular malformations [Bertolini 1987]. Only one third of patients with this anomaly become symptomatic and require surgical treatment [Edwards 1953, Keith 1967]. One of the first clinical reports of an aortic arch complex anomaly was published in 1974 [Bayford 1974]. In 1945, Gross reported the first surgical repair of double aortic arch [Gross 1945]. In 1946, Neuhauser reported a radiographic description of aortic arch anomalies [Neuhauser 1946]. Patients present a multitude of signs and symptoms at the time of diagnosis, but symptoms arising from the respiratory system are the most prevalent. Wheezing was the most common respiratory symptom in our patients and was followed in frequency by stridor, upper respiratory tract infection,

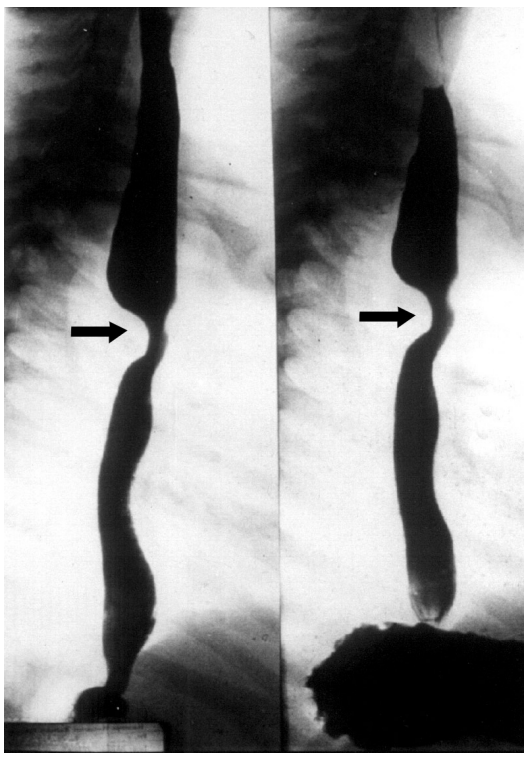

Figure 1. Barium esophagogram demonstrating indentation caused by double aortic arch. The arrow is pointing at the posterior indentation of the esophagus caused by external compression.

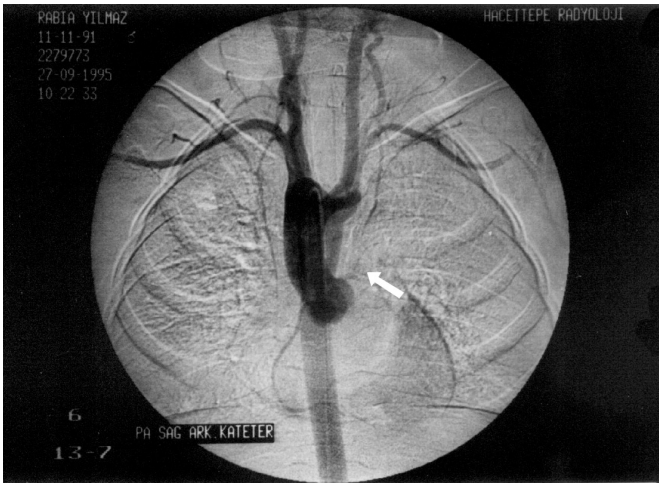

Figure 2. Digital subtraction angiogram showing the right dominant double aortic arch. The arrow is pointing at the two aortic arches.

pneumonia, and cough. Emesis and gastroesophageal reflux were the chief gastrointestinal findings in patients. When these common pediatric signs and symptoms are found in a newborn or young infant, either chronically or on a recurrent basis, the physician should become alerted to the possibility of an aortic arch complex anomaly [Kocis 1997]. In previously published reports, there was a predominance of male patients for all types of aortic arch complex anomalies, but in our series there was a predominance of female patients (female-male ratio, 56%:44%) [Hewitt 1970, Kocis 1997, Lincoln 1969, Richardson 1981].

A barium esophagogram is extremely reliable for diagnosing a vascular ring but does not delineate the precise vascular anatomy. Aortography has been the gold standard for providing a preoperative road map of the anomaly [Valette 1989]. Echocardiography and magnetic resonance imaging (MRI) are new modalities for diagnosing aortic arch anomalies [Azarow 1992, Han 1993]. Recent publications have especially favored MRI for the diagnosis of vascular anomalies causing compression [Beekman 1997, Soler 1998, Zachary 2001]. In our institution, MRI has not been the method of choice for the diagnosis of vascular rings. MRI requires as much as an hour for the acquisition of images, when the patient has to be anesthetized and usually intubated. This procedure in our clinical practice seems to put unnecessary stress on an already dyspneic patient.

The types of aortic arch anomalies encountered at our institution were similar to those reported in previously published series [Chun 1992, Han 1993, Horvath 1992, Kocis 1997, Woods 2001]. Double aortic arch was the most common anomaly, followed by right aortic arch with left ligamentum arteriosum and right aortic arch with aberrant left subclavian artery. Of the 16 cases of double aortic arches, 11 were right dominant, 3 were left dominant, and 2 were left atretic arches. Surgery was clearly indicated in all symptomatic patients with vascular compression. When performed early, surgery reduces morbidity and secondary long-segment tracheomalacia, which is an unfavorable prognostic risk factor [Arciniegas 1979, Blair 1986, Chun 1992, Greenholz 1986, Hagl 1997, Sebening 2000].

Exposure through a left thoracotomy incision is generally accepted as the operation of choice for division of a vascular ring [Chun 1992, Han 1993, Horvath 1992]. Jung and associates once recommended a right thoracotomy, and Sebening and associates preferred the median sternotomy in vascular rings [Jung 1978, Sebening 2000], but we found that a standard left posterolateral thoracotomy provided excellent exposure in all but 1 case. Median sternotomy is the approach of choice for division and anastomosis of a pulmonary vascular sling. We also preferred median sternotomy in a case of pulmonary vascular sling in which cardiopulmonary bypass was used for resection. Intensive postoperative care is required for these patients, including intensive respiratory therapy, oxygen when needed, humidity, chest physiotherapy, and repeated suctioning of oral and nasal secretions. Attention to these details also has subjectively improved results [Backer 1989]. Occasionally, intubation and assisted ventilation may be necessary, but prolonged endotracheal intubation should be avoided so as not to aggravate an already narrowed airway. Normally, the symptoms and signs of tracheobronchial and esophageal obstruction are relieved immediately but not completely following the division of the vascular ring. A slower regression of respiratory symptoms is expected in some patients because of preexisting pulmonary disease and mild-to-moderate tracheomalacia. Concomitant major congenital anomalies are the causes of prolonged hospital stay, rehospitalization, a poorer outcome, and death. In our series, 13% of children had residual respiratory or gastroesophageal symptoms, and 3.3% of children died in the early postoperative period because of dilated cardiomyopathy.

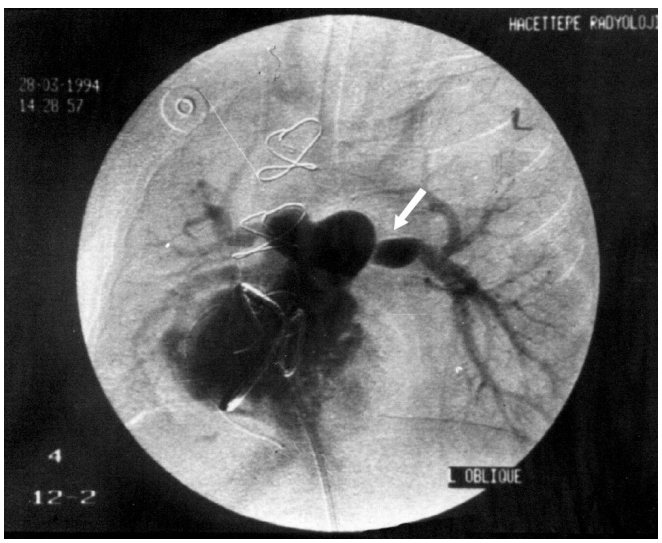

Figure 3. Cineangiogram of the patient with left pulmonary artery sling who was operated on through a median sternotomy with cardiopulmonary bypass. The arrow is pointing at the left pulmonary artery after reimplantation to the main pulmonary trunk and assessing normal pulmonary blood flow.

Vascular compression may cause complex intrathoracic tracheoesophageal obstruction. The clinical suspicion of a vascular ring as the cause of serious tracheoesophageal obstruction in a young child should lead to an early diagnosis and referral for surgical treatment. Surgical repair is associated with low or no mortality in patients with uncomplicated aortic arch anomalies.

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