Aortic Arch Anomalies: Simplified Classification

Surgical Experience with Ten Cases*

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Between the years 1965 and 1970, ten patients were operated on for congenital anomalies of the aortic arch which caused symptoms of tracheoesophageal compression. The classic Edwards' diagram was modified for a more simple interpretation of these anomalies. Accordingly, all our patients were found to belong to four distinct variations: 1) double aortic arch, which was divided in two subgroups, a) with functioning double aortic arch, b) with partial involution of one of the arches; 2) right aortic arch, mirror image branching of the arch vessels and retroesophageal aortic diverticulum with left arterial duct or ligament connected to the diverticulum; 3) right aortic arch with left aberrant retroesophageal subclavian artery; 4) Left aortic arch with right aberrant retroesophageal subclavian artery. All patients were operated on through a left thoracotomy; the technique used varied according to the anomaly found. There was no surgical mortality. One patient's death four months following operation was caused by a technical error which allowed obstruction of tracheal tube during emergency treatment for tracheomalacia. The remaining nine patients have been asymptomatic in an observation period of six months to five years.

Congenital anomalies of the aortic arch and its main branches may determine the formation of vascular rings around the trachea and esophagus, with varying degree of compression upon these organs. Hommel, in 1737, was the first to describe such an anomaly of the aortic arch. Bayford reported the symptoms and pathologic findings in a patient with dysphagia lusoria produced by an aberrant subclavian artery. Von Siebold, in 1836, described the symptoms of a double aortic arch. In 1932, Abbot pointed out the possible surgical solution for these anomalies.

Two centuries after the first description of this malformation, Gross in 1945 performed successful division of a double aortic arch. The exact diagnosis of these malformations acquired real importance when surgical treatment became feasible.

This report is based on experience gained from ten patients undergoing corrective operation for congenital symptomatic aortic arch anomalies in our hospital in the last five years.

Clinical Material

Between the years 1965 and 1970, ten patients underwent surgery in our department for congenital anomalies of the aortic arch that caused symptoms of compression on the trachea or esophagus or both.

The age of the patients ranged between two days and 11 years; eight were less than one year old, and two were seven and 11 years old. Seven were boys and three girls. One of the patients was a two-day-old baby, whose operation was urgent because of asphyxia immediately following birth which required tracheal intubation until operation.

Symptoms were stridor and intercostal retraction in all cases. In seven of the patients there were repeated infections of the respiratory tract. There was dysphagia in two patients. In patients of up to one year of age, there was a delay in weight and height development (Table 1). Symptoms appeared in four patients immediately after birth, and between two and nine months in the rest.

Confirmation of the clinical diagnosis was achieved by

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means of an esophagogram, tracheal laminogram or aortic arch angiogram. Esophagograms were performed in all patients and were all abnormal. Laminograms of two patients showed tracheal compression above the bifurcation in both. Aortic arch angiography was done in all patients and helped to outline the precise vascular anomaly. These procedures aided in the planning of surgical technique. Right brachial counter current (noncatheter) aortic arch angiography was done in eight patients, left brachial in one, and selective percutaneous transfemoral in one.

On the basis of the angiographic findings and confirmation at operation, all the patients were classified into four groups. (See additional details in discussion.)

1. Double aortic arch, which was divided into two subgroups:
   a) with functioning double aortic arch in one patient (Fig 1).
   b) with partial involution of one of the arches in three patients (Fig 2).

2. Right aortic arch, with mirror image branching of the arch vessels and retroesophageal aortic diverticulum and left ductus arteriosus connected to the aortic diverticulum. Three patients belonged to this entity (Figs 3 and 4).

3. Right aortic arch with aberrant left subclavian artery in two patients (Fig 5 and 6).

4. Left aortic arch with aberrant right subclavian artery in one patient (Fig 7 and 8).

**Surgical Treatment**

The lesion in each patient was approached through a left thoracotomy. In the patient with a functioning double aortic arch, the posterior arch was divided between the right subclavian artery and the descending aorta. In three patients in whom there was partial involution of the arch, the anterior was found to be the hypoplastic and therefore divided. In the three patients with right aortic arch, mirror image branching of the aortic arch vessels and retroesophageal aortic diverticulum with left ductus arteriosus, treatment consisted of division of the arterial ligament or duct, combined by dissection of peritracheal and periesophageal adhesions, which included the remnant of the arch.

The two patients with right aortic arch and aberrant retroesophageal left subclavian artery and arterial duct, underwent division of both. In the patient with left aortic arch and right aberrant retroesophageal subclavian artery, the latter was transected as well as the ductus arteriosus.
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RESULTS

There was no surgical mortality. Symptoms disappeared in eight of the patients immediately after surgery, and in these patients no symptoms have been recorded during the six-month to five-year observation period. In two cases, stridor persisted during the first months following surgery; it was attributed to a softening and deformity of tracheal cartilages. One patient became asymptomatic later on, and the second, a ten month old infant, continued to have some stridor. He was admitted to another hospital four months after operation with severe respiratory distress. Bronchoscopy and tracheal intubation were performed under anesthesia, but he died during transportation to our hospital because of tracheal tube obstruction.

DISCUSSION

Anomalies in the development of the aortic arch ramifications may be considered of little importance, unless they produce compression on the trachea or esophagus or both.

There is considerable confusion in medical literature concerning the nomenclature and interpretation of the aortic arch anomalies.

Edwards7 was the first to explain the composition and development of the aortic arch and its ramifications as a system with a double aortic arch from each of which originated two brachiocephalic vessels and an arterial duct. Based on Edwards’ diagram, four possible areas of involution can be listed in each arch, which will determine the type of
vascular anomaly (Fig 9). Accordingly, the normal aortic arch is the result of the involution of area A or B or both on the right arch. The reason for breaking down areas A and B is that B lies proximal to the right ductus arteriosus and A distal to it. There is, however, some difference of opinion with regard to the existence of the right duct, and therefore, we elected to identify this region as proximal or distal to the right duct.²

It is, therefore, evident that by numbering the classic Edwards' diagram with the possible involutional area one may derive to three main clinical groups of tracheoesophageal compression:

1. Double aortic arch, when the two arches are present and connected one to another without interruption. It can be divided in two subgroups:
   a) with functioning double aortic arch,
   b) with partial involution of one of the arches.

2. Right aortic arch, with mirror image branching of the arch vessels and retroesophageal aortic diverticulum and left ductus arteriosus connected to the aortic diverticulum. In this case the involution occurs in area B of the left aortic arch. Theoretically, this may occur with the right arch although this anomaly has not been encountered in our material or reported by others.

3. Aberrant retroesophageal innominate or subclavian arteries. Here also one could identify several variations: namely, right innominate or subclavian artery with left aortic arch, or left innominate or subclavian artery with right aortic arch.

   Each of these groups can be again subdivided depending upon the position of the descending aorta, on the left or on the right of the spine.

   The main symptoms caused by vascular rings are of compression of the trachea or esophagus or both. They generally appear before the first year of life, though there are patients with aortic arch malformations who may remain asymptomatic.

   Common symptoms are dyspnea, stridor, repeated respiratory infections and vomiting. Notable early signs in babies are intercostal retraction and cyanosis. During such a bout of dyspnea, babies are usually found with cephalic hyperextension which helps to straighten the deformed stenotic trachea.¹

   As a consequence of the repeated respiratory infections and difficulties in nutrition, these babies in addition usually develop anemia and are slow in physical development.

   The aberrant right retroesophageal subclavian artery rarely will exhibit symptoms in childhood. On the other hand it may produce symptoms in
adolescence or adult age, when it manifests itself mainly by dysphagia, the so-called dysphagia lusoria. Clinical diagnosis is easy if this anomaly is taken into consideration.

In our series, eight of the ten patients were younger than one year. All of them had symptoms of tracheal compression, and dysphagia was the leading symptom only in two, one of which represented the right aberrant retroesophageal subclavian artery. In none of our patients were associated cardiac or other malformations found.

Esophagogram usually confirms the diagnosis by showing a deformation of the esophagus at the aortic arch level. Aortic arch angiography is essential to outline the exact anatomic variation, thus aiding the surgeon with the planning of an adequate operation.

In most cases surgical treatment in a symptomatic patient must take place at a very early age of childhood, sometimes even as an urgent operation, as was the case with one of our patients. All the cases in this group were approached through a left thoracotomy. In the patients with a functioning double aortic arch, it is preferable to divide the arch which does not give rise to arch vessels. In the group of patients with partial involution the division is done in the smaller arch. Among patients with atretic arch but having aortic diverticulum and ductus arteriosus, the latter should be divided as well as a wide dissection of the peritracheal and periesophageal tissue. In those cases with aberrant left subclavian artery and ductus arteriosus, both should be divided.

As far as the right aberrant subclavian artery anomaly is concerned, the most acceptable surgical approach is by separating the vessels from the aortic arch and suspending its distal portion to the right side of the spine. This was done in one of our patients. According to the literature, this operation may bring occasionally, in patients with disturbed collateral circulation, ischemia and even gangrene of the upper limb. It may also produce the subclavian steal syndrome. Attempts have been made to obviate these complications by anastomosis of the right subclavian artery to the ascending aorta or carotid artery, in which case the access is through a right thoracotomy.

Prolonged compression of the trachea may lead to a softening of its rings, which may delay the disappearance of the symptoms in the postoperative period in spite of an adequate relief of compression.

From the experience in the literature and from our own, it is obvious that surgery is the single effective treatment of aortic arch anomalies causing symptoms. Surgery should be done early in life and occasionally these congenital anomalies may represent an emergency problem.

REFERENCES

Editorial Expression

This is an excellent report of these congenital lesions which are recognized more often lately. It should be emphasized that although the method of treatment proposed by the authors for aberrant subclavian arteries has been generally accepted in the past, it should be abandoned at the present time. A definitive method of treatment to re-establish flow to the distal artery, should be used to prevent the development later of a subclavian steal syndrome.

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The One-Hundredth Element of Mendeleev's Periodic Table

The one-hundredth element of the Periodic Table, together with the ninety-ninth, were first unsuspectedly found in the clouds of hydrogen-bomb explosion set off in 1952 at Eniwetok atoll in the Pacific. Element 100 was named fermium after Enrico Fermi (1901-1954), Nobel laureate in 1938 for his identification of new radioactive elements produced by neutron bombardment and his discovery of nuclear reactions effected by slow neutrons. He left his native Italy where he was full Professor of Physics at the University of Rome and at the age of 29 the youngest member of the Royal Academy. He reached the United States in 1939 and became Professor of Physics at Columbia University. Fermi suggested the possibility that nuclear fission might be the key to the release of colossal energy by the mechanism of chain reaction. He speculated that the fission of the uranium atom might liberate additional neutrons which might be made to fission other atoms of uranium, just as one firecracker on a string sets off another firecracker until the whole string seems to go up like a torpedoed munition ship. Subatomic energy could thus be released, producing from even a 0.1 percent conversion of a single pound of uranium equivalent to that produced by several million pounds of TNT.

Jaffe B: Men of Science in America.
New York, Simon & Schuster, 1958

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