

Vascular anomalies compressing the oesophagus and trachea

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Vascular rings formed by anomalies of major arteries can compress the trachea and oesophagus so much as to cause respiratory distress and dysphagia. Twenty-nine patients with this condition are reviewed and discussed in five groups. The symptoms and signs are noted. Radiological examination by barium swallow is the most useful diagnostic aid. Symptoms can only be relieved by operation. The trachea is often deformed at the site of the constricting ring. Only infrequently is there immediate relief from the pre-operative symptoms. Two babies were successfully treated for an aberrant left pulmonary artery.

The diagnosis and treatment of major arterial anomalies which cause compression of the oesophagus and trachea are now well established. Although an aberrant right subclavian artery was described in 1794 by Bayford (Fig. 1), who gave the detailed post-mortem finding in a woman who had died from starvation secondary to this anomaly, and a double aortic arch was described by von Siebold (1837), it was not until 1945 that Gross successfully divided a double aortic arch in a 4-month-old infant. Following this success the diagnosis and treatment of this and similar anomalies became firmly established.

MATERIAL

The surgical experience will be discussed of 29 patients treated consecutively during a 15-year period from 1952 to 1967 at the Hospital for Sick Children, Great Ormond Street, London. The embryology and classification of such anomalies are not discussed because we have nothing to add to previous papers (Edwards, 1959; Stewart, Kincaid, and Edwards, 1964; Gross and Ware, 1946).

The anomalies are discussed under the following headings:

Double aortic arch

Right aortic arch in conjunction with a left ligamentum arteriosum or persistent ductus arteriosus

Aberrant subclavian artery

Aberrant left pulmonary artery

Abnormally placed innominate artery

DOUBLE AORTIC ARCH Nineteen children were treated for some form of double aortic arch. Their age at operation ranged from 1 week to 11 months, but the majority were treated at about the age of 5 to 6 months (Fig. 2). Frequently the presenting symptoms had been noticed since birth but for varying reasons there was delay in making the correct diagnosis. Stridor, dysphagia or poor feeding, cyanotic attacks, and recurrent respiratory tract infections were the four main presenting symptoms. All but two patients had respiratory stridor and in seven of these children there was concomitant dysphagia or poor feeding. Six children had frequent respiratory tract infections. Initially, some children had bronchoscopy performed in the mistaken belief that they had primary laryngeal or tracheal disease. Plain radiographs of the chest were always taken and were usually non-contributory. Barium swallow enabled the correct diagnosis to be made in all patients in whom this was performed (Fig. 3a,b). In one patient with suspected heart disease angiography was also performed.

A right descending thoracic aorta was found in 13 patients, and in 11 of these the major arch was posterior, in one the major arch was anterior, and in one the arches were equal in diameter (Fig. 4). In a 9-day-old baby with a major posterior arch, the small anterior arch joined not the descending aorta but the middle of a patent ductus arteriosus (Fig. 5).

A left descending thoracic aorta was found in six patients, the main arch being posterior in three and anterior in three (Fig. 6). Eight patients had

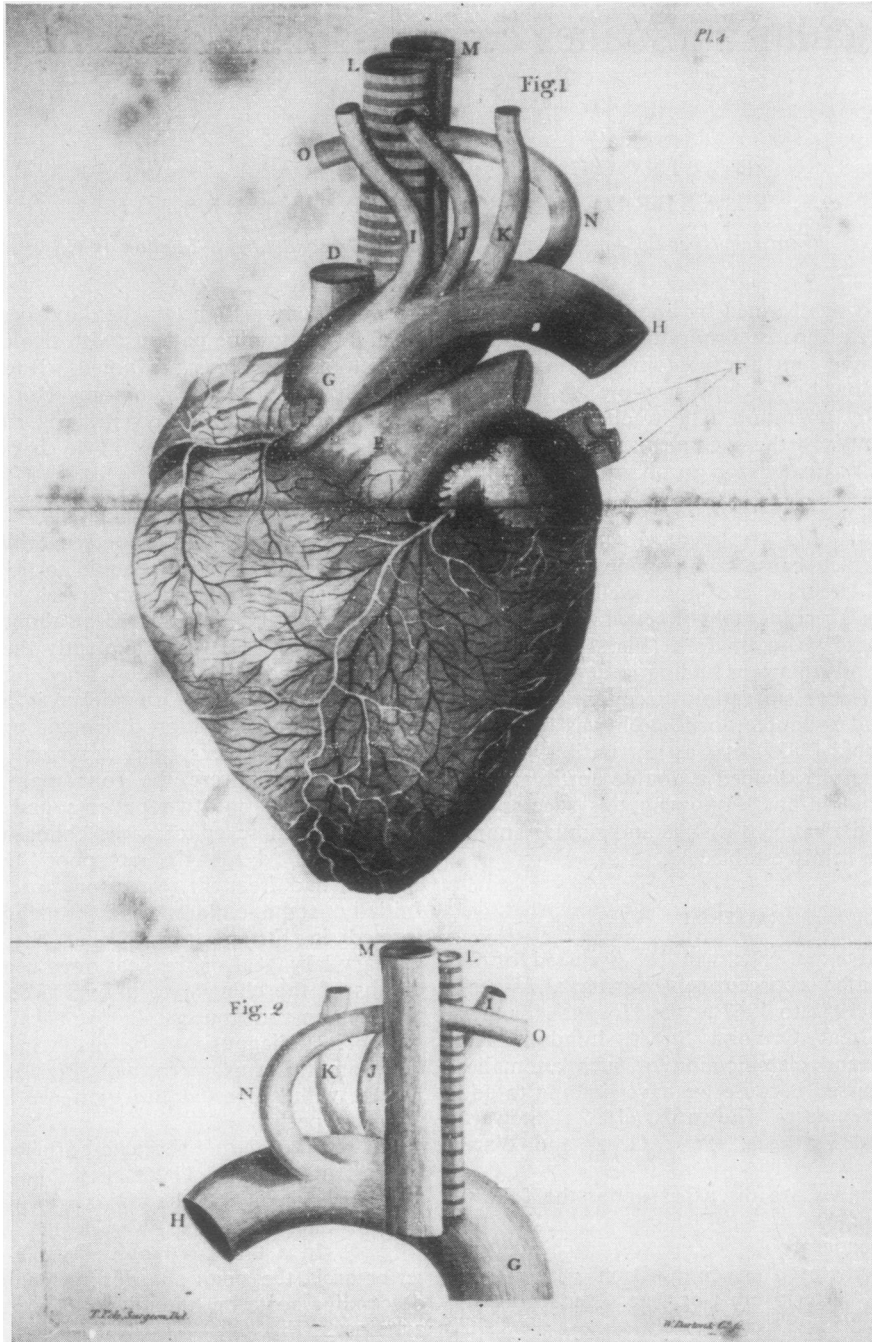


FIG. 1. An aberrant right subclavian artery. A drawing of the necropsy specimen described by David Bayford at a meeting of the Medical Society of London, 2 July 1787. In this patient the aberrant artery passed between the oesophagus and the trachea.

a patent ductus arteriosus and 11 a ligamentum arteriosum, all being on the left side.

Associated cardiovascular abnormalities were found in three patients. In one there was a ventricular septal defect and a left superior vena cava ;

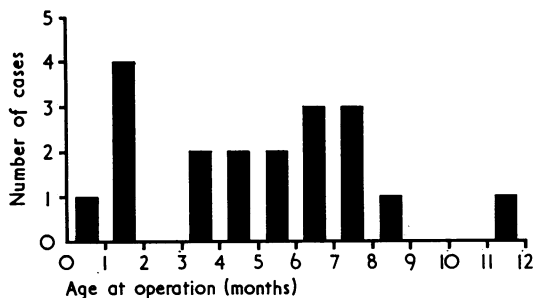


FIG. 2. Age at operation of 19 patients with double aortic arch.

in another an aberrant left subclavian artery associated with a right arch ; and in a third there was a persistent truncus arteriosus (Figs 7 and 8). The baby with the truncus arteriosus and double aortic arch had an exploratory thoracotomy but the vascular ring was very loose and was not divided. This child died four months after the operation in congestive cardiac failure. Another

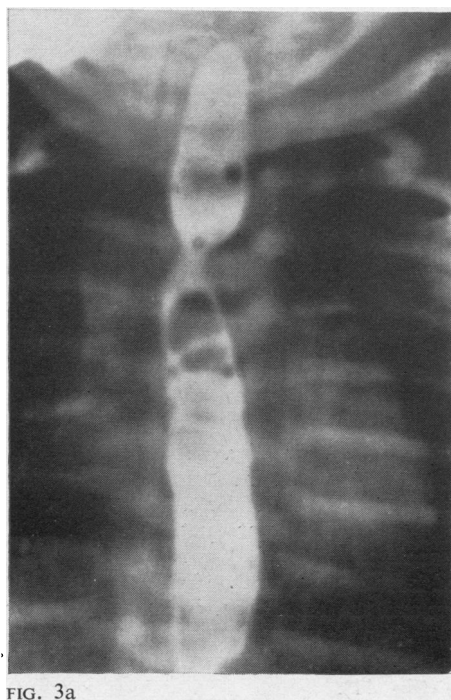
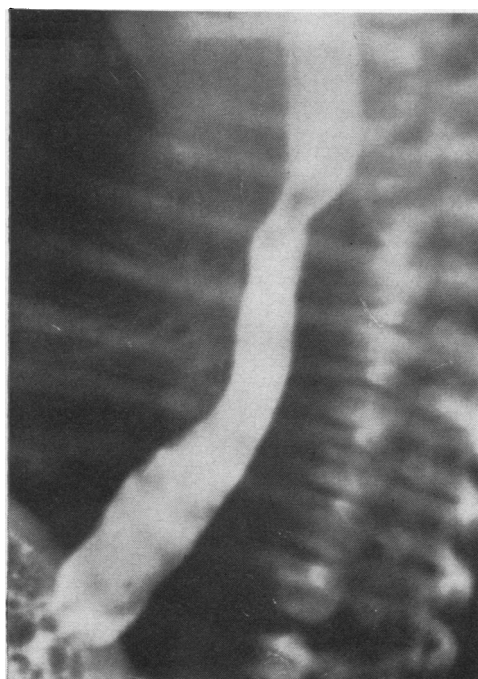


FIG. 3a

Z

patient died when ventricular fibrillation occurred as the chest was opened.

The anterior arch was divided distal to the origin of the left subclavian artery and, as this was frequently the smaller of the two arches, it was usually divided. A duct or ligament was always looked for and divided, and the trachea and oesophagus were dissected out to free these structures from constricting fibrous tissue.



(b)

FIG. 3. Double aortic arch. Postero-anterior (a) and lateral (b) views of a barium swallow. There are bilateral concave indentations of the oesophagus at the level of the aortic arch. In the lateral view the oesophagus is indented from behind.

Post-operative follow-up showed that in only two patients was there immediate complete relief from the preoperative signs, some degree of stridor being nearly always present in the first postoperative week, but then resolving. Respiratory signs became worse after the operation in five patients within 10 weeks, one requiring a tracheostomy and the other nasotracheal intubation.

RIGHT AORTIC ARCH WITH LEFT LIGAMENTUM ARTERIOSUM OR PATENT DUCTUS ARTERIOSUS
 Compression of the oesophagus and trachea by a vascular ring comprising a right aortic arch and

either a patent ductus arteriosus or a ligamentum arteriosum occurred in four patients. These children tended to be older than those with a double aortic arch, the youngest being two months and the oldest three years of age. All had stridor, and in three there was concomitant difficulty with feeding; this was so in the 3-year-old child. Recurrent respiratory tract infection occurred in all but one of the children. Plain radiographs of the chest were useful; evidence of a right arch was noted in all, and in the 3-year-old child the trachea at the site of constriction was seen to be narrowed

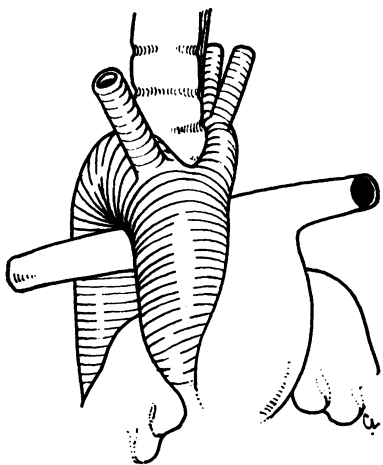


FIG. 4. A double aortic arch with a right descending thoracic aorta. The major arch is posterior.

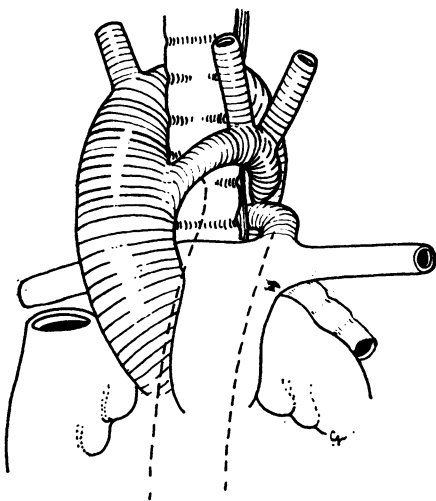


FIG. 5. A double aortic arch. The minor arch is anterior and terminates by joining a patent ductus arteriosus.

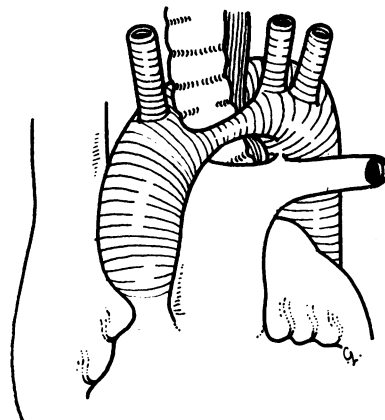


FIG. 6. A double aortic arch with a left descending thoracic aorta. The major arch is posterior. The anterior arch is waisted between the innominate and left common carotid arteries.

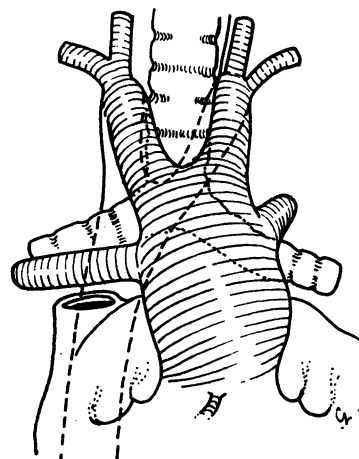


FIG. 7. A persistent truncus arteriosus with a double aortic arch. There is a right descending thoracic aorta. The innominate artery arises from the posterior arch, and the left common carotid and the left subclavian arteries arise from the anterior arch. Only the right pulmonary artery is seen.

in an oblique view (Fig. 9). Barium swallow was helpful in all cases; the oesophagus was indented by the right arch of the aorta and on the left side by the ligament or duct (Fig. 10a, b).

Associated congenital cardiac anomalies were noted in two patients. One had a ventricular septal defect and the other a persistent left superior vena cava and an aberrant left subclavian artery, which arose from a partly patent ligamentum arteriosum (Fig. 11).

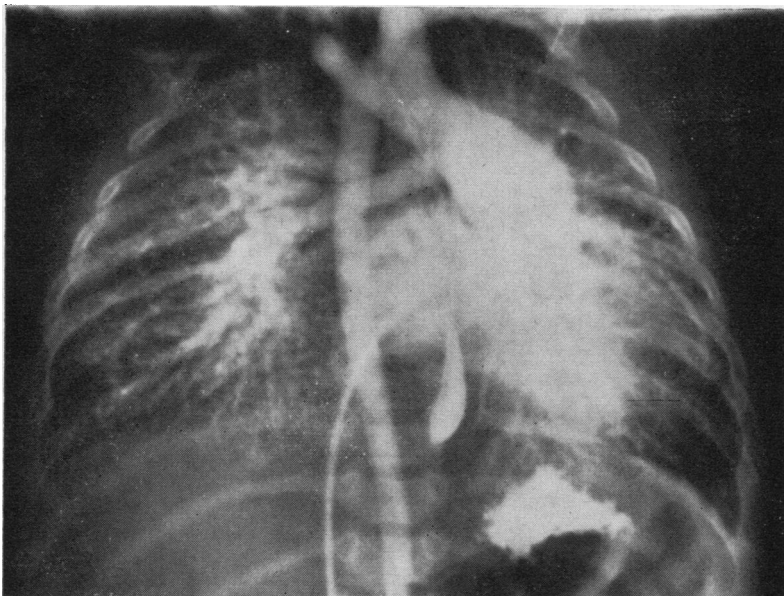


FIG. 8. *Persistent truncus arteriosus with double aortic arch. Angiogram shows a right pulmonary artery emerging from the main aortic trunk. The two aortic arches join to form a common right descending thoracic aorta.*

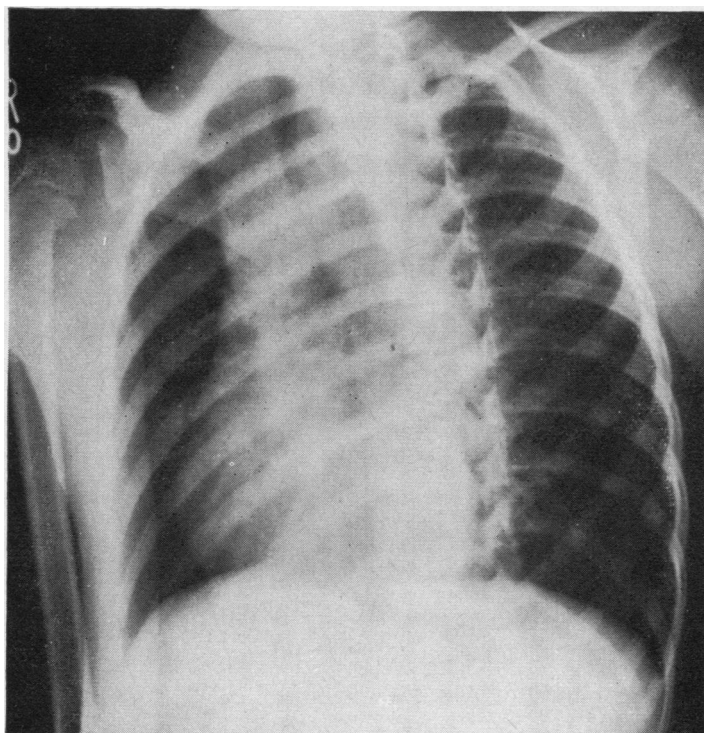
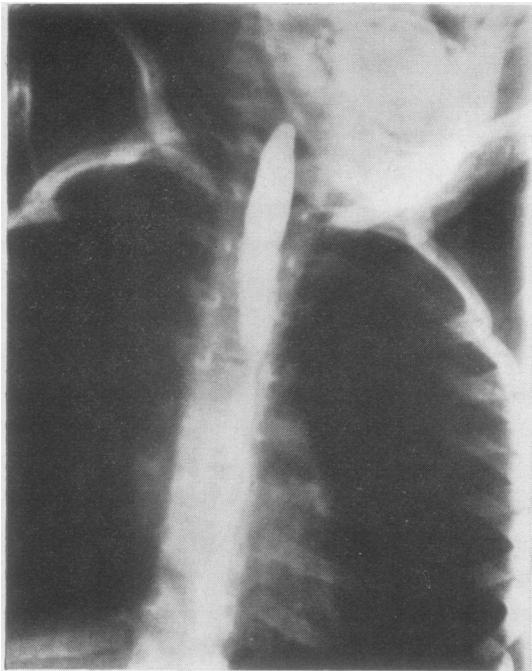
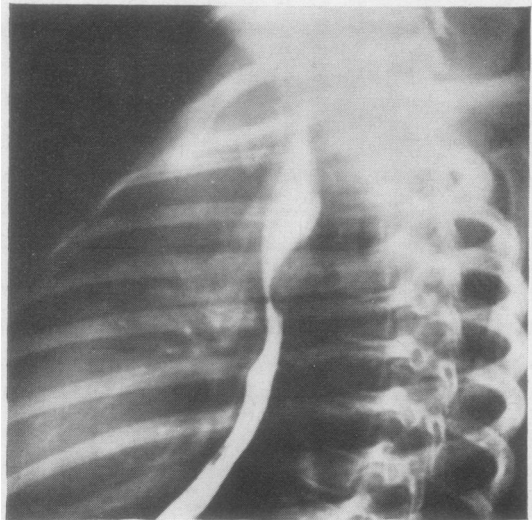


FIG. 9. *Right aortic arch with persistent ligamentum arteriosum. Oblique, plain radiograph of chest. The trachea is narrowed in its middle third.*



(a)



(b)

FIG. 10. *Right aortic arch with persistent ligamentum arteriosum. Postero-anterior (a) and lateral (b) radiographs of a barium swallow. The main arch of the aorta is seen to the right and indenting the oesophagus. The posterior defect seen in the lateral view is produced mainly by the ligamentum arteriosum and partly by the arch of the aorta.*

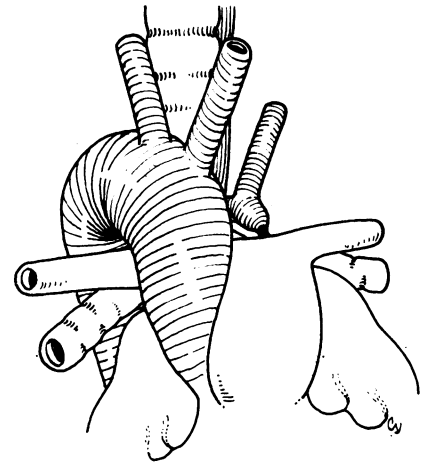


FIG. 11. *Right aortic arch with part ligamentum arteriosum and part patent ductus arteriosus. The left subclavian artery arises from the patent ductus arteriosus.*

At operation a patent ductus arteriosus was present in two patients and a ligamentum arteriosum in the other two. All four had a right descending thoracic aorta (Fig. 12). The ligamentum or duct was divided and the tissues surrounding the trachea and oesophagus were dissected out as previously described. One patient had acute airway obstruction at operation and had a cardiac arrest. During resuscitation procedures the aortic end of the patent ductus arteriosus slipped from the vascular clamp and receded into the right chest, allowing fatal haemorrhage.

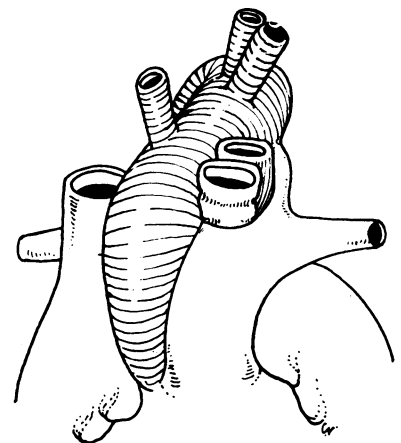


FIG. 12. *A right aortic arch with a right descending thoracic aorta. There is a persistent ligamentum arteriosum causing narrowing of the trachea and oesophagus.*

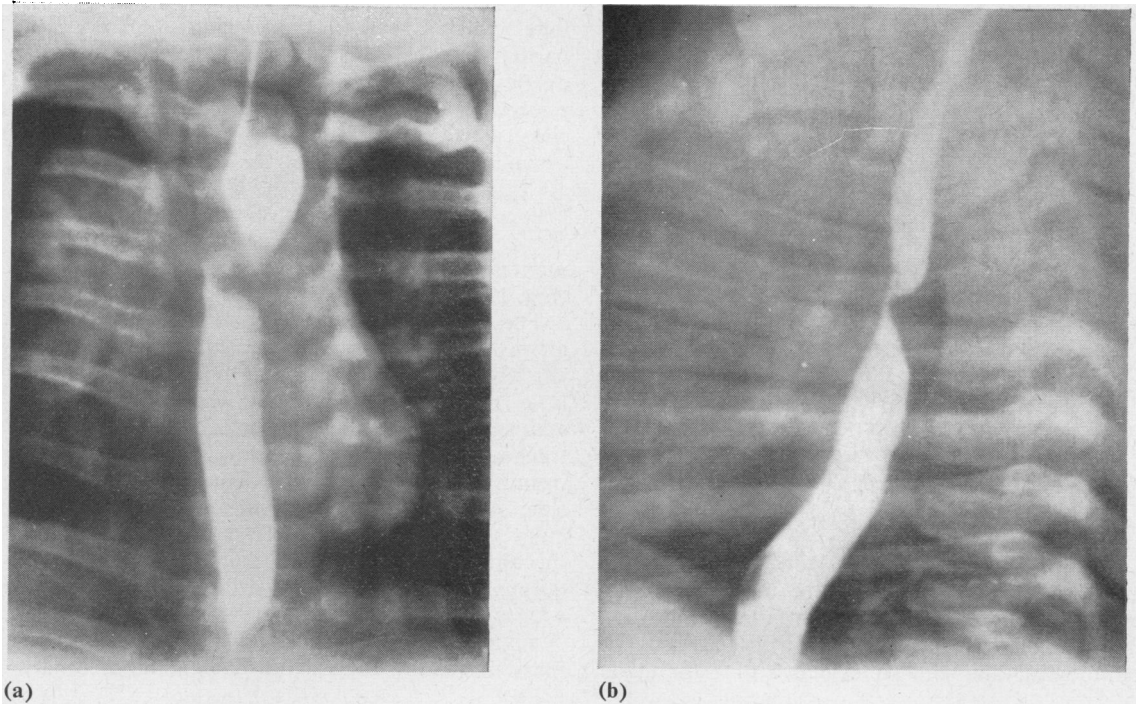


FIG. 13. *Aberrant right subclavian artery. Postero-anterior (a) and lateral (b) radiographs of a barium swallow, showing a concave posterior indentation of the upper third of the oesophagus by an aberrant right subclavian artery. This vessel passes upwards and obliquely at an angle of about 65°.*

Complete relief from stridor was achieved by the fifth post-operative day, and in three cases there was immediate relief.

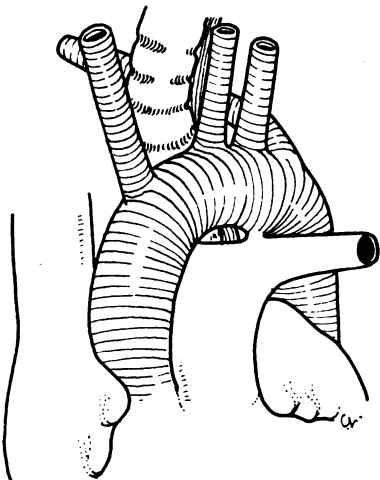


FIG. 14. *An aberrant right subclavian artery, passing obliquely upwards and posterior to the oesophagus.*

ABERRANT SUBCLAVIAN ARTERY Despite the frequency of aberrant subclavian arteries (Edwards, 1953 ; Stewart *et al.*, 1964) the number of patients seen with symptoms or signs was small—four patients (ages 2 to 8 months). In one child only was this the sole vascular anomaly, the others being found in association with a double aortic arch, a tetralogy of Fallot, and a right aortic arch and left patent ductus arteriosus. One child, a poor feeder, had a typical indentation of the posterior aspect of the upper third of the oesophagus demonstrated at barium swallow (Fig. 13a, b). This baby had the aberrant right subclavian artery divided at its origin and subsequently improved (Fig. 14). The other children had no symptoms. One infant had an aberrant left subclavian artery which passed posterior to the oesophagus, having arisen from a right aortic arch (Fig. 15). This was found when performing a left Blalock-Taussig shunt.

ANOMALOUS LEFT PULMONARY ARTERY This anomaly has been seen in three babies, their ages at operation being 1 month, 5 months, and 8 months. These patients presented with inspiratory

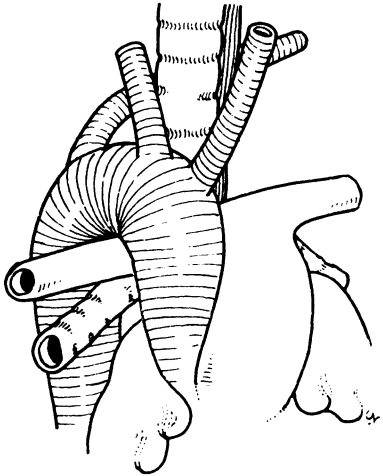


FIG. 15. An aberrant left subclavian artery arising from a right aortic arch.

and expiratory stridor, and in the 5-month-old child there was persistent right upper lobe collapse. There was no difficulty with feeding. The radiological and clinical evidence of right upper

lobe collapse caused some confusion until a barium swallow was performed and the correct diagnosis was made. The aberrant artery compressed the right upper lobe bronchus. The classical concave indentation of the anterior wall oesophagus at the level of the carina was seen in all three patients (Fig. 16). However, the significance of this sign was missed in the 8-month-old baby. Angiography was performed in the 1-month-old baby but was not clinically essential (Fig. 17).

The 8-month-old baby was admitted with acute airway obstruction, a tracheostomy having been carried out at the referring hospital—but with little relief. The baby was bronchoscoped on admission and a concentric narrowing of the trachea was seen 2–5 cm. below the tracheostomy opening; the trachea was indented from the right side. A small polythene catheter was with difficulty threaded into the left main bronchus and the airway was somewhat improved. Plain radiographs of the chest showed a large anterior mediastinal shadow.

With a presumptive diagnosis of a mediastinal mass narrowing the trachea a right thoracotomy

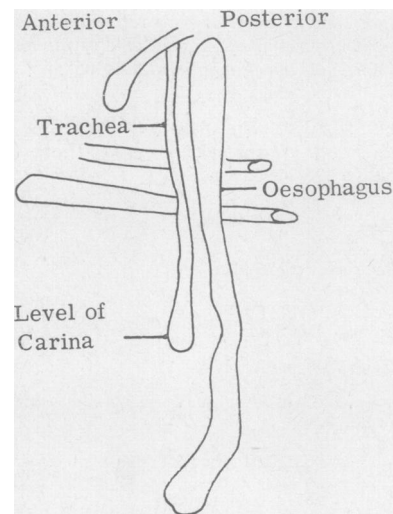
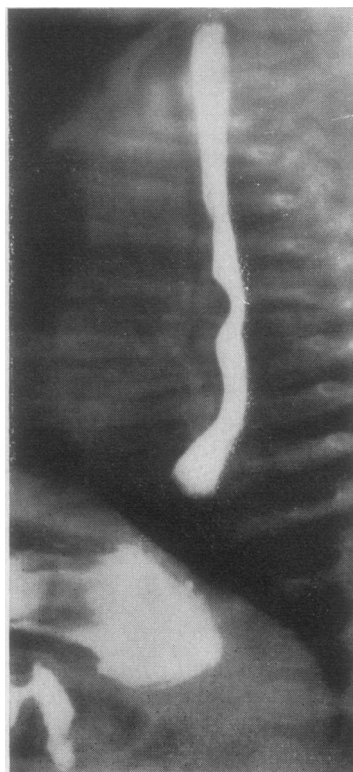


FIG. 16. Anomalous left pulmonary artery. Lateral radiograph of a barium swallow showing a concave indentation of the oesophagus at the level of the carina.

was performed and an aberrant left pulmonary artery was found. It passed posteriorly and then to the right of the superior tracheo-bronchial angle, and then passed posterior to the trachea and onward to the left pulmonary hilum. The airway obstruction increased and therefore the trachea was dissected out and found to be hypoplastic and solid with complete cartilagenous rings. The aberrant pulmonary artery was then dissected out and fascial bands were divided, but even when this artery was retracted away from the trachea there was no relief of the airway obstruction. An attempt was made to dilate the trachea from below through a second tracheostomy. As the baby's condition was perilous no attempt was made to re-implant the aberrant pulmonary artery and the

chest was closed. The baby died. Post-mortem examination showed a congenital abnormality of the trachea with complete ring cartilages.

The correct diagnosis was made pre-operatively in the other two patients and at left thoracotomy the route of the aberrant artery was identical in both. The artery arose from the terminal part of an elongated main pulmonary artery, passed posteriorly around the right superior tracheo-bronchial angle, and then onto the posterior surface of the trachea, being anterior to the oesophagus, and so to the left lung (Fig. 18). The artery was dissected out and divided from the main pulmonary artery. The artery was brought anterior to the trachea. In both patients the left pulmonary artery was anastomosed to the side of

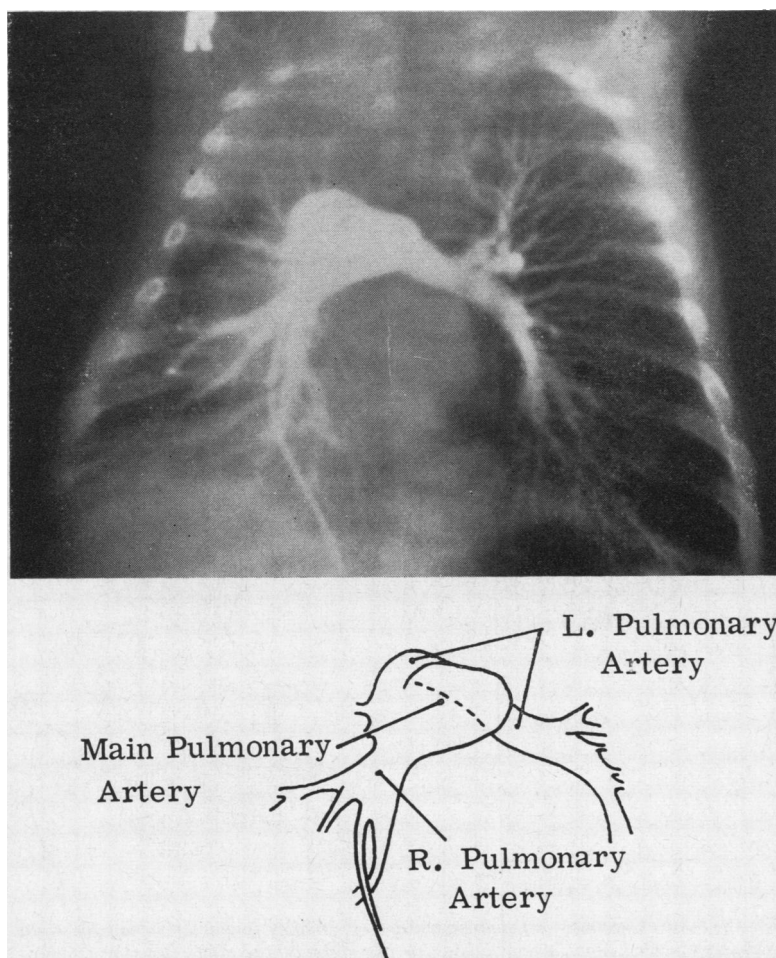


FIG. 17. Anomalous left pulmonary artery. A pulmonary angiogram showing an elongated and deviated left pulmonary artery.

the main pulmonary artery because when attempting to re-anastomose at the site of transection excessive kinking occurred.

TABLE
TOTAL NUMBER OF ANOMALIES IN 29 PATIENTS

Type of Anomaly	Total No. of Anomalies	M	F	Age Group (yrs.)			Death in Hospital
				0-1	1-2	>2	
Double aortic arch...	19	10	9	19	—	—	1
Right arch + left ligamentum arteriosum or P.D.A. ...	4	2	2	2	1	1	—
Aberrant subclavian artery	4	2	2	4	—	—	1
Anomalous left pulmonary artery	3	3	—	3	—	—	1
Abnormally placed innominate artery...	1	—	1	—	—	1	—

In one infant excessive tracheo-bronchial secretions necessitated a tracheostomy. Both babies made an uninterrupted recovery with rapid relief of their symptoms.

ABNORMALLY PLACED INNOMINATE ARTERY In one 3-year-old child the innominate artery arose so far

to the left on the aortic arch that it compressed the trachea (Fig. 19). She fed poorly from birth and inspiratory and expiratory stridor with costal recession was marked. Plain radiographs of the chest and a barium swallow showed no abnormality.

Bronchoscopy showed indentation of the trachea anteriorly and proximal to the carina. On pressing the tip of the bronchoscope against the indentation, the right radial pulse diminished in volume (Mustard, Trimble, and Trusler, 1962).

At thoracotomy (through the third left intercostal space) the aortic arch was slung forward by sutures attached to the back of the sternum and the pericardial reflection. Following this the stridor greatly improved.

DISCUSSION

SIGNS AND SYMPTOMS The clinical picture of major vascular anomalies which compress the trachea and oesophagus varies according to the type of anomaly present. In the patients with a

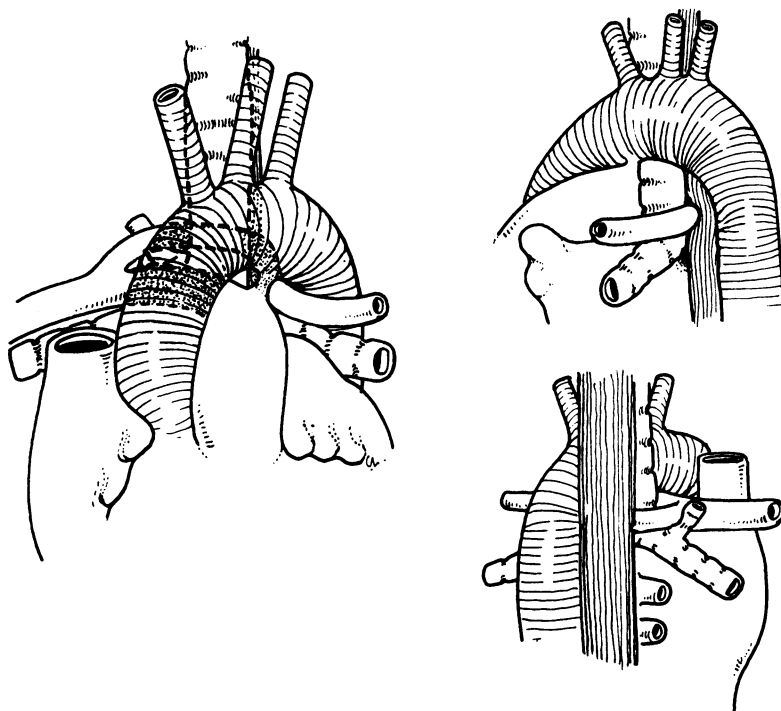


FIG. 18. An anomalous left pulmonary artery, smaller in diameter than the right pulmonary artery. This artery passes to the right of the right superior tracheo-bronchial angle and posterior to the trachea, and indents the anterior part of the oesophagus at the level of the carina.

double aortic arch, right arch with ligamentum arteriosum or persistent ductus arteriosus, aberrant left pulmonary artery and abnormally placed innominate artery, the signs were well marked, whereas many aberrant subclavian arteries often cause no signs. The main signs of a 'vascular ring' are stridor, respiratory embarrassment, dysphagia or poor feeding, and recurrent respiratory tract infection. These were present together or separately. The respiratory tract infections followed spill-over of food and subsequent inhalation.

Children with a right aortic arch, in conjunction with a left duct or ligament and aberrant subclavian artery, presented later in life although not conspicuously so in our experience. The other anomalies usually presented in the first year of life.

DIAGNOSIS Primary laryngeal and tracheal congenital deformities, pressure on the trachea from lymphadenopathy, and congenital oesophageal narrowing were considered in the differential diagnosis.

INVESTIGATIONS Accurate barium swallow was the most certain and, in many of the anomalies, the only method of making the diagnosis. Plain radiographs of the chest had only been informative in that group of patients with a right aortic arch in combination with a left duct or ligament. This anomaly should be suspected when on postero-anterior radiographs the aortic knuckle is absent from the usual site and the aortic arch is to the right of the mid line (Neuhauser, 1946). Oblique films showed a well-marked narrowing of the trachea in one child.

Barium swallow was the most useful radiological aid to the diagnosis. With the double aortic arch the oesophagus was narrowed by lateral pressure of the anterior and posterior aortic arches. The patients with a right arch and persistent left ligamentum arteriosum and patent ductus arteriosus showed a well-marked indentation of the right side of the oesophagus and some indentation of the left side of the oesophagus. This was most marked on screening when the right-sided indentation pulsed. An aberrant subclavian artery was demonstrated by indentation of the posterior aspect of the upper one-third of the barium-filled oesophagus when seen in the lateral view (only occasionally does the artery go between the oesophagus and trachea, and we have had no experience of this). When the left pulmonary artery was anomalous, a concave indentation was seen in the oesophagus at the level of the carina.

It was rarely necessary to perform angiography to confirm the diagnosis and this was carried out on two occasions when congenital heart disease was present. Tracheograms were not performed in any instance.

Endoscopic examination of the larynx, trachea, and oesophagus did not give sufficient positive or negative information to warrant its frequent use. Bronchoscopy could be dangerous in a small baby with an already critically narrowed trachea.

OPERATIVE TREATMENT A full left lateral thoracotomy was found to give the best exposure. In the only case in which a right thoracotomy was performed the anatomy could not be adequately defined and a left thoracotomy had to be performed.

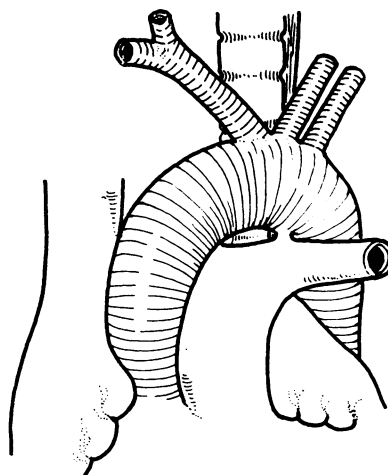


FIG. 19. An innominate artery arising from the aortic arch further to the left than is usual, thus pressing on the trachea.

A period of trial occlusion was carried out on each arch separately prior to division of the arch. In this series the anterior arch was the arch most frequently divided. When the posterior arch was the smaller of the two it was divided at its junction with the descending thoracic aorta, and when the anterior arch was the smaller of the arches it was usually divided distal to the origin of the left subclavian artery or sometimes between the left common carotid artery and the left subclavian artery if a definite narrowing was present at this point (Fig. 6). The combination of a persistent truncus arteriosus and a double aortic arch is uncommon, and only one other example was found in the literature (Kerwin, 1936). Both arches were equal in diameter and neither was divided as they

appeared to be very loose and not constricting the trachea or oesophagus.

A patent ductus arteriosus or ligamentum arteriosum was always divided whenever present.

The experience of aberrant subclavian arteries was small. It appears that this artery can be divided with impunity but the possibility of precipitating a subclavian steal syndrome should be remembered (Reivich, Holling, Roberts, and Toole, 1961).

When correcting an aberrant left pulmonary artery, rerouting this artery so that it lies in front of the left bronchus gave no problems, but its reanastomosis to the site of transection proved difficult because of kinking. It was necessary to reimplant the pulmonary artery into the side wall of the main pulmonary artery.

Dissection around the trachea and oesophagus was made in all cases to divide all constricting fascial bands.

Patients were nursed in maximum humidity in an oxygen tent and frequent oropharyngeal toilet was performed. Tracheostomy was performed postoperatively on two occasions for respiratory difficulty, but this was (and should be) avoided in nearly all cases. The tracheal rings are deformed and still partially obstruct the airway at the previous site of constriction for some weeks or even months after operation.

RESULTS

The ages, the variety of anomalies, and the deaths that occurred in hospital are shown in the Table. The immediate response to surgical correction was variable, particularly in those patients with a

double aortic arch. In only 6 of 29 patients was there immediate postoperative relief from stridor, and, of these, three had relief of a right arch and left ligamentum or duct, one an aberrant subclavian artery, and two were treated for double aortic arch. Occasionally the symptoms became worse during the first week after operation. From one to 12 weeks elapsed before freedom from stridor was noted.

We wish to thank our clinical colleagues for help in managing these patients. We also wish to thank the Department of Medical Illustration, Institute of Child Health.

REFERENCES

- Bayford, D. (1794). An account of a singular case of obstructed deglutition. *Mem. med. Soc. Lond.*, **2**, 275.
- Edwards, F. R. (1959). Vascular compression of the trachea and oesophagus. *Thorax*, **14**, 187.
- Edwards, J. E. (1953). Malformations of the aortic arch system manifested as "vascular rings". *Lab. Invest.*, **2**, 56.
- Gross, R. E. (1945). Surgical relief for tracheal obstruction from a vascular ring. *New Engl. J. Med.*, **233**, 586.
- (1955). Arterial malformations which cause compression of the trachea or esophagus. *Circulation*, **11**, 124.
- and Ware, P. F. (1946). The surgical significance of aortic arch anomalies. *Surg. Gynec. Obstet.*, **83**, 435.
- Kerwin, A. J. (1936). Persistent (partial) truncus arteriosus associated with double aortic arch. *J. techn. Meth.*, **15**, 142.
- Mustard, W. T., Trimble, A. W., and Trusler, G. A. (1962). Mediastinal vascular anomalies causing tracheal and esophageal compression and obstruction in childhood. *J. Canad. med. Ass.*, **87**, 1301.
- Neuhauser, E. B. D. (1946). The roentgen diagnosis of double aortic arch and other anomalies of the great vessels. *Amer. J. Roentgenol.*, **56**, 1.
- Reivich, M., Holling, H. E., Roberts, B., and Toole, J. F. (1961). Reversal of blood flow through vertebral artery and its effect on cerebral circulation. *New Engl. J. Med.*, **265**, 878.
- von Siebold, C. T. (1837). Ringförmiger Aorten-Bogen bei einem neugeborenen blausüchtigen Kinde. *J. Geburtsh. Frauenzimmer-Kinderkrank.*, **16**, 294.
- Stewart, J. R., Kincaid, O. W., and Edwards, J. E. (1964). *An Atlas of Vascular Rings and Related Malformations of the Aortic Arch System*. Charles C. Thomas, Springfield, Illinois, U.S.A.