AORTIC ARCH ANOMALIES: TREATABLE CAUSES OF DYSPHAGIA AND RESPIRATORY DISTRESS

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INTRODUCTION:

VASCULAR RINGS, either complete or incomplete, formed by abnormalities in the aortic arch vessels, can compress the trachea and oesophagus to such an extent as to cause respiratory distress or dysphagia. Symptoms may appear at any age. Although surgical management may be complicated by residual respiratory distress and a stormy post-operative course, these are benign conditions and attempts should be made to relieve the obstruction. A high index of suspicion is imperative, so that appropriate investigations and treatment can be embarked upon early in the clinical course.

This paper reports on 3 patients presenting with symptoms indicative of aortic arch anomalies.

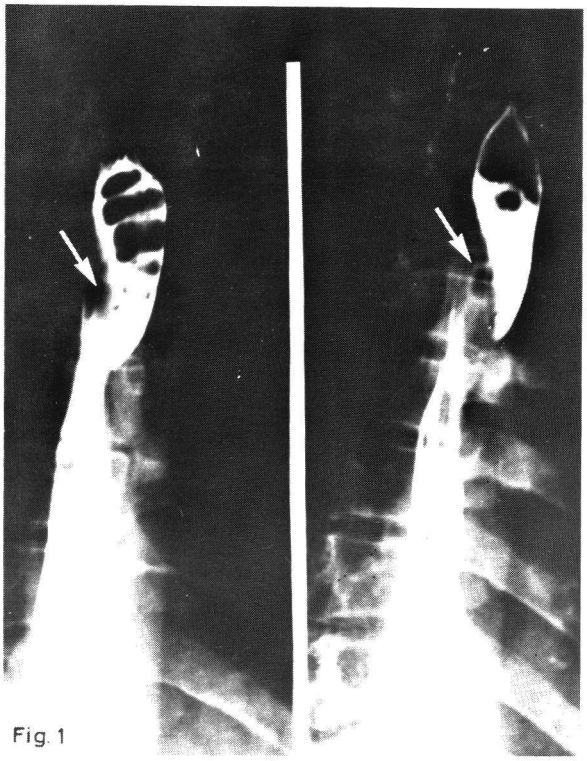

Fig. 1

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Case 1:

C.A.M. an 18 year old female, presented with a 2-year history of mild dysphagia to solid food. There was no history to suggest previous ingestion of corrosives. Clinically the patient was well nourished and no obvious abnormalities were detected on physical examination. The barium swallow examination performed revealed on oblique posterior identation of the oesophagus (Fig. 1) suspicious of an aberrant right subclavian artery. This was subsequently confirmed on arch aortography (Fig. 2). In view of her mild symptoms, no surgical intervention was contemplated. After 2 years of medical surveillance, she defaulted.

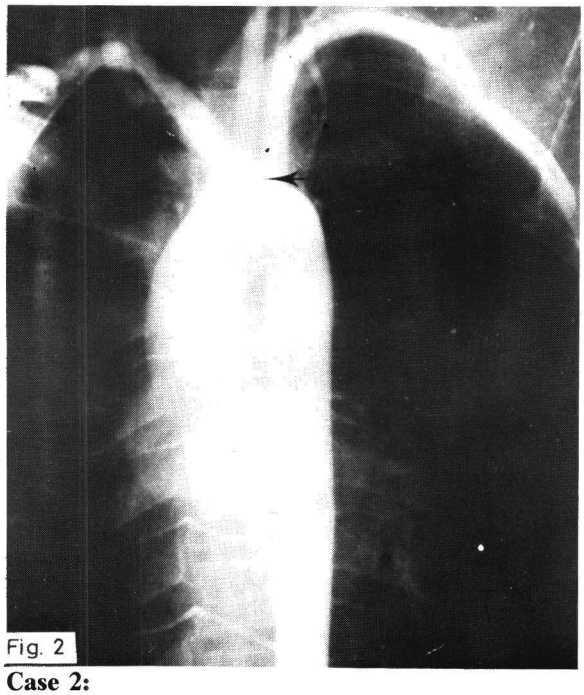
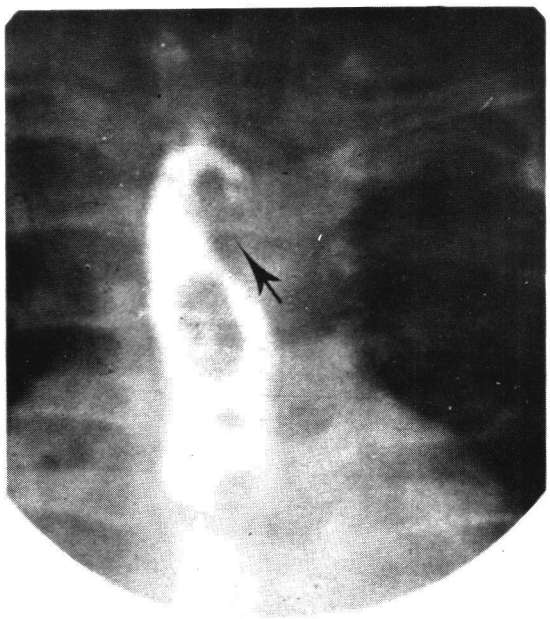


Fig. 2

Case 2:

Z.B.Z., male, aged 8 months was first known to us soon after delivery when he was found to

have an imperforate anus, bilateral preauricular sinuses and a submucous cleft palate. An anal cutback was performed and this was followed by regular anal dilatation on an outpatient basis. During these visits, he had often been noted to be growing very poorly. One week prior to his present admission, he developed a fever associated with a cough and respiratory difficulty. Direct questioning revealed that he fed poorly — although he was able to suck well, he had difficulty swallowing and was often noted to be drooling with saliva.



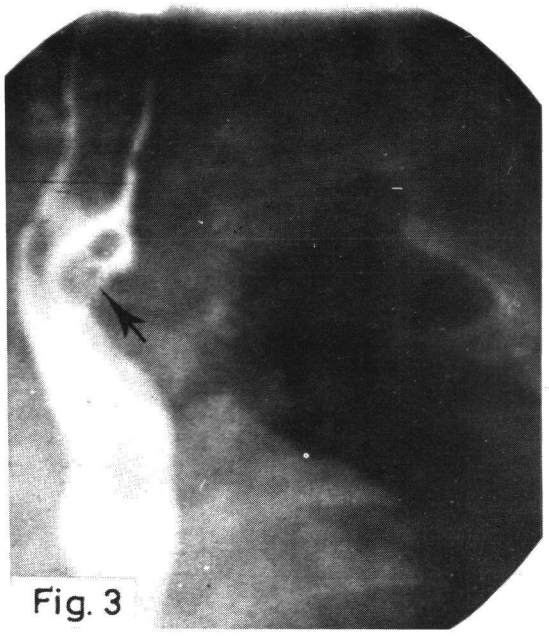


Fig. 3

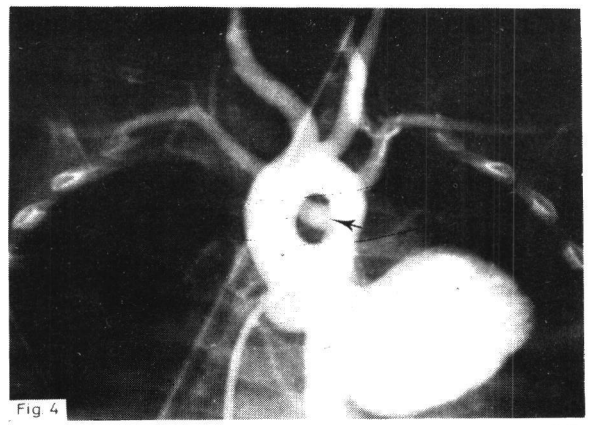


Fig 4

Clinical examination revealed a small, wasted infant. He was febrile, tachypnoeic, anaemic and dehydrated. There was mild intercostal recession and crepitations were heard over both lungs. Chest x'rays suggested bilateral bronchopneumonia and the child was treated accordingly. Because of the history of swallowing difficulty, a barium swallow was performed and this showed a posterior indentation of the oesophagus (Fig. 3). Aortography confirmed the presence of an aberrant right subclavian artery (Fig. 4). On the 18th hospital day, the chest was explored through a left thoracotomy. At operation, the anomalous subclavian artery was mobilised and divided at its origin. The post operative course was uneventful and at discharge the child was noticed to be able to swallow with no difficulty.

Case 3:

P. a 12 year old female was admitted with a one day history of productive cough and fever. Going back into her medical history it was noted that she was the product of a normal full-term pregnancy. However, at the age of 21 days, she was hospitalised for a period of 2 months for fever, cough and stridor. Since then she has had recurring episodes of cough productive of yellowish sputum. There was no history of dysphagia.

On examination, she was found to be febrile. Diffuse rhonchi and crepitations were heard over both lung bases. A chest radiograph done at this stage showed a right-sided aortic arch and evidence of bilateral basal bronchopneumonia. She responded well to antibiotic treatment and vigorous chest physiotherapy.

To exclude bronchiectasis, a bronchogram was arranged and this demonstrated a localised indentation of the left wall of the trachea just above its bifurcation (Fig. 5). At bronchoscopy, this tracheal narrowing was confirmed, in addition,

the posterior wall was observed to be pulsatile. Barium swallow examinations showed circumferential indentations of the oesophagus at the level of the aortic arch (Fig. 6). Subsequently, an arch aortogram was carried out and this revealed a double aortic arch with a right-sided descending aorta (Fig. 7). These findings were confirmed at exploratory thoracotomy. After freeing the anterior aortic arch, it was divided distal to the origin of the left subclavian artery. The ligamentum arteriosum was similarly mobilised, ligated and divided.

The post operative course was complicated by sputum retention which cleared with intensive physiotherapy. Tracheograms performed 3 months after the operation showed some persistence of narrowing. Regular follow-ups were gratifying in that the child remained symptom-free and had put on weight satisfactorily.

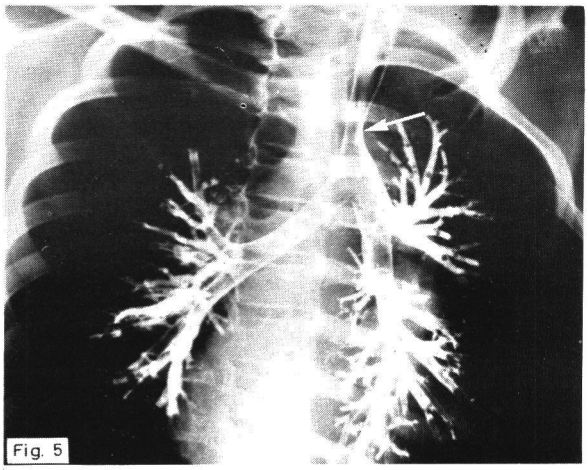


Fig. 5

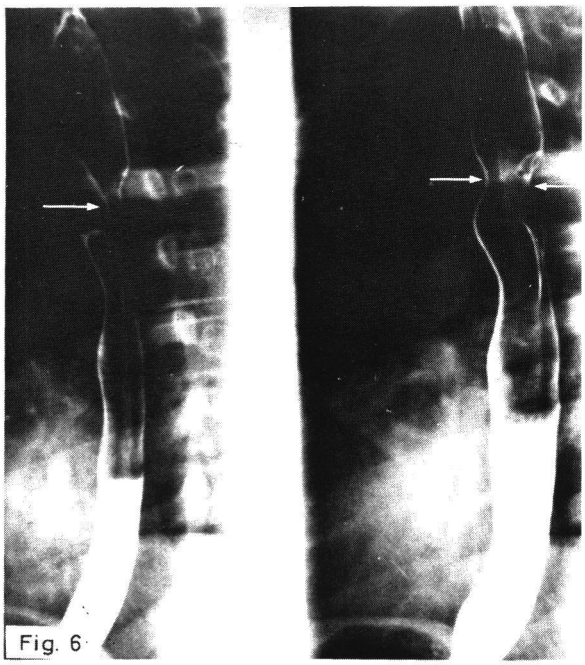


Fig. 6

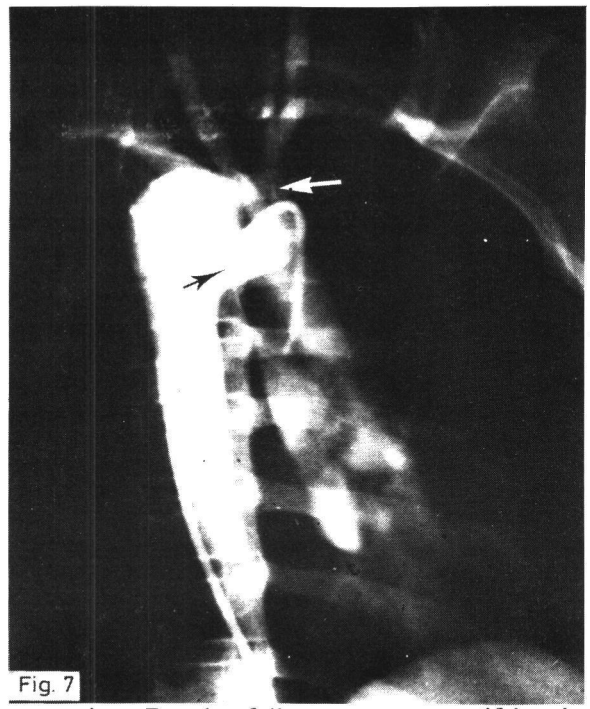


Fig. 7

legend to figures;

Fig. 1 Barium filled oesophagus showing a long length oblique indentation suggestive of an anomalous origin of right subclavian artery. (Arrows showing the oblique extrinsic impression).

Fig. 2 Arch arteriogram showing anomalous origin of right subclavian artery (arrow) from the left side as the last major branch of the aortic arch.

Fig. 3 Barium filled oesophagus showing an oblique extrinsic impression (arrow) in its middle one third suggestive of an aberrant right subclavian artery.

Fig. 4 Left ventricular angiogram showing the ascending aorta and its branches. The arrow indicates the presence of an anomalous right subclavian artery arising on the left side as the last major branch of the aortic arch.

Fig. 5 Brochogram demonstrating a localised indentation (arrow) on the left wall of the trachea just above the bifurcation.

Fig. 6 Oesophagogram showing extrinsic compressions on both sides of the oesophagus (arrows) at the level of the aortic arch.

Fig. 7 Arch arteriogram showing a double aortic arch with right sided descending aorta. The small arch (arrow black) lies anteriorly with the left subclavian arising at the origin of the left arch (arrow white).

Discussion:

Since Bayford's (1794) first reported case of an aberrant right subclavian artery causing dysphagia, many different varieties of aortic arch anomalies have been reported. From a clinician's view point, the simplified clinical classification put forth by Bradham *et al.* (1968) seems useful. In brief, aortic arch anomalies fall into 3 groups. (Table I).

They manifest clinically by causing compression of the oesophagus and/or trachea. The four main presenting symptoms are stridor, cyanotic attacks, poor feeding or dysphagia and recurrent respiratory tract infections. In children, respiratory symptoms without dysphagia are more common. In adults, on the other hand, the trachea is more rigid and therefore more resistant to compression, so that dysphagia is the principle complaint. Symptomatology may be severe and be evident shortly after birth or it may be altogether absent so that some patients with arch anomalies may remain asymptomatic throughout life. The question frequently asked is, why do some patients remain symptom-free? Klinkhamer (1966) is of the opinion that an artery pressing on the oesophagus from behind, is not in itself sufficient explanation for the occurrence of compression. It is his belief that symptoms arise only if the flexible trachea and oesophagus are hindered from being displaced forward at the crossing of the retro-oesophageal vessel. This can occur, for example when both carotids arise together or close to each other on the aortic arch. (Normally, the innominate artery and the left carotid artery are separated by a distance of about 4cm.). Other workers (Bailey, 1955; Ekstrom, 1959) believe that respiratory distress is caused by aspiration as a consequence of the dysphagia. It is worth noting that both theories cannot explain why symptoms persist post-operatively in some patients.

In the management of these patients, an accurate anatomical diagnosis is imperative. This should include an arch aortogram to delineate the incriminating vessels. In aberrant right subclavian arteries, complete relief of symptoms can be obtained by surgical division of the first part of the artery as described by Gross (1946). The arm will have an adequate blood supply through collateral vessels communicating with the second and third parts of the subclavian artery. Vascular rings are best managed by division of the minor arch together with division of the patent ductus arteriosus or ligamentum arteriosum. To prevent residual compression by a large posterior arch, it has been recommended that the anterior arch should be suspended to the back of the sternum.

Post-operatively, these patients should be closely observed as the respiratory symptoms may take some time to regress. Numerous theories have been put forward to explain this residual respiratory compression. Nuboer (1951) was the first to suggest that persistent stridor is caused by softening and distortion of the cartilage rings of the trachea. Two types of tracheal defects have been observed. Firstly, there may be a stenotic or hypoplastic segment localised to the area compressed by the vascular encirclement. Growth here may be inhibited in utero by compression and this continues till the trachea is liberated. It is for this reason that some surgeons feel very strongly that surgery should be done as soon as the diagnosis is made. Secondly, in some the lumen of the trachea is not greatly diminished but the cartilages may be softened by ring pressure so that they collapse on inspiration. Whether this tracheomalacia is related to compression or whether it is a separate intrinsic entity is open to conjecture.

TABLE I: Clinical Classification of Aortic Arch Anomalies

Group I:	**Complete trachea and oesophageal encirclement**
	— double aortic arch.
	— right arch with P.D.A. or ligamentum arteriosum.
Group II:	**Compression of anterior portion of trachea without oesophageal involvement**
	— aberrant origin of innominate artery.
	— aberrant origin of left carotid artery.
Group III:	**Compression of posterior portion of oesophagus with or without tracheal involvement**
	— aberrant right subclavian artery.

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