VASCULAR COMPRESSION OF THE TRACHEA AND OESOPHAGUS

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(RECEIVED FOR PUBLICATION FEBRUARY 22, 1959)

In the developing embryo the foregut is surrounded by a group of vascular structures of bilateral distribution connected with the branchial arches. Portions of these arches are obliterated to form the normally described aorta and great vessels. The ultimate persistence of only one main vascular arch, the left, as the aorta, permits the trachea and oesophagus to lie freely to its right side. Where, however, the vascular obliteration in the embryo stage is abnormal, the residual vessels may so closely surround the trachea and oesophagus that either or both become compressed and symptoms are produced (Fig. 1).

The first description of this anatomical abnormality appears to have been made by Hommel in 1737. Quain in his surgical anatomy (1844) gives the following account:

"Hommel, who has been the assistant of Haller, described the very remarkable preparation represented in this figure (Fig. 2), and gave a delineation, from which this is copied, with the omission of the heart and right lung, which are shown in the original drawing. 'The aorta,' says the author, 'divided at the beginning of the arch and was again united at its end. Through the sort of island constructed by the bifurcation pass the trachea and oesophagus; a disposition,' he correctly adds, 'perhaps never previously observed.'"

In 1837 von Siebold noted the compression of these structures when he reported the death of a 10-day-old infant from respiratory obstruction due to an aortic ring. In this case other abnormalities were present, an atrial septal defect, and transposition of the great vessels.

It is difficult to estimate the incidence of this condition. Maude Abbott (1936) in her 1,000 cases of congenital heart disease coming to necropsy found five cases of double aorta, of which two had symptoms and three were without. The oldest was 87. In the same series there were 14 cases of primary right aortic arch and seven cases of aberrant right subclavian artery. At the Royal Liverpool Children’s Hospital, out of 1,874 cases of congenital heart disease investigated in the heart clinic, 12 cases of aortic ring have been found. Of these 10 were investigated because of symptoms and two were found as incidental observations. There appears to be no predominant sex incidence. In the same series five cases of aberrant right subclavian artery with symptoms were recognized.

Blincoe, Lowance, and Venable (1936) have given a review of earlier reports of the condition, and it is manifest from the ages of the described cases that many succumb in infancy and childhood. Wolman (1939) clearly defined the syndrome and paved the way for its diagnosis and treatment. Relief of the constriction can only be obtained by surgical means, and Gross and his colleagues (Gross, 1945, 1953; Gross and Ware, 1946; Gross and Neuhauser, 1951) have described their very considerable experience from the Boston Children’s Hospital.

EMBRYOLOGY

In the early stage of the development of the cardiovascular system, a single stem arises from the arterial end of the heart (aortic bulb) and divides into two ventral aortae which pass forward along the ventral wall of the pharynx. Opposite each branchial arch, a lateral branch is given off from each ventral aorta and it passes backwards around the pharynx, joining with its fellows of the same side to form a dorsal longitudinal vessel, resulting in two dorsal aortae lying upon the dorsal wall of the pharynx (Fig. 3). The need for two dorsal aortae is due to the presence of the central mass of the foregut, neural tube, and notochord at this level, but just below the fifth dorsal segment they unite to form a single vessel. At the twelfth week (6 mm. embryo) this system has reached its full development, and subsequently various sections are
Fig. 1.—Pathological specimen in an infant of the trachea and oesophagus, surrounded and compressed by an aortic ring. A large patent ductus arteriosus is also seen producing a secondary constriction below the double aortic arch.
absorbed leaving behind the structures which persist as the normal human aorta and its branches, together with the pulmonary artery.

The parts concerned with the development of the aorta and its main branches are the third to the sixth branchial arterial arches. Basically the third arch remains as the carotid arteries, and the fourth arch becomes the two aortic arches, of which the right usually disappears. The fifth arch disappears and has no known residuum in the adult human form; in fact in foetal life only the most evanescent signs of this arch have been demonstrated (Congdon, 1922). The sixth arch forms the ductus arteriosus and gives off branches to the developing lung buds, which become the pulmonary arteries. The subclavian arteries arise from the dorsal aortae before they fuse to become the common aorta. These changes are well shown in the standard diagram (Fig. 4), which is a plan view of Fig. 3.

In the production of the normal human arch an area of the right dorsal aorta disappears, the resulting components being as demonstrated in Fig. 5.
Usually the involuting portions of the arches disappear without trace, but occasionally remnants persist as fibrous bands, or as a narrow “co-arcted” area with or without a lumen.

When only the left arch persists, as in the normal human pattern, the trachea and oesophagus lie to the right and are unimpeded by the vascular structures. If, however, both arches persist, or the ductus arteriosus of the opposite side to the main arch remains patent or obliterates to form a short ligamentum arteriosum, or if residual bands exist, then the trachea and oesophagus may be compressed. Furthermore, aberrant vessels may run anteriorly or posteriorly to the trachea and oesophagus and may produce compression of one or the other.

Although there are many minor variations of the residual anatomy in man, compression of the trachea and oesophagus are produced by certain defined patterns of vascular abnormality.

For a clear understanding of the various abnormalities of the aortic arch that may occur, the developmental state at the sixth week (12 mm.) embryo must be used as the basis, and this is demonstrated in the succeeding diagrams.
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Fig. 5.—Involution of a portion of the right dorsal aorta (blacked out) results in the formation of the normal aortic arch and branches. The right subclavian artery swings across to lie lateral to the right common carotid artery.

Fig. 6.—Persistence of the 12 mm. embryo stage will result in a double aortic arch. The arches may be of equal size, but usually the right or left is dominant.

former more commonly than the latter (Fig. 6). With the rotation of the heart, its apex running to the left and its base to the right, and the laevo-position of the aorta, the right and left branchial arterial arches with their corresponding portions of the dorsal aortae take up a posterior and anterior position relative to the developing trachea and oesophagus. Thus in the double aorta the right fourth branchial arterial arch and its connexions become the posterior aortic arch and the left fourth branchial arterial arch and its connexions give rise to the anterior arch. The embryological right and left arches and the anatomical posterior and anterior arches are respectively synonymous. It is usual for the right common carotid and the subclavian arteries to arise from the posterior arch and the left common carotid and subclavian arteries from the anterior arch, but the branches of the lesser arch may have arisen so peripherally that they appear to arise from the dominant arch as the first and last branches respectively. Absorptive processes may take place in either arch, causing areas of coarctation and fibrous bands, but the whole follows a standard pattern. In a double aortic arch the developing trachea and oesophagus may be compressed between the two arches.

If the portion of the left arch between the origin of the left subclavian artery and the left ductus arteriosus is absorbed, then a right aortic arch is left with a left innominate artery arising as the first branch from the aorta (Fig. 7a). If a remnant of the absorbed area persists as a band, then the left arch is completed around the foregut, but even without this band the location of the left innominate artery is such that pressure on the trachea can occur. In a right aortic arch the ductus arteriosus is longer than normal and winds around the side of the left main bronchus and oesophagus and may thus produce a second level of constriction of a mild degree. If the right aortic arch continues as a right descending aorta,
then the ductus may have a potentially longer course and be even more constricting.

If the portion of the left arch between the ductus arteriosus and the combined dorsal aorta obliterates, then the ductus arteriosus will run into the left innominate artery (Fig. 7b), the ring being completed on occasion by a band from the left subclavian artery to the right aortic arch.

As a variable form of right aortic arch, obliteration of the anterior arch between the left common carotid and the left subclavian artery produces a left common carotid arising as a first branch from the aorta, which crosses over the front of the trachea to ascend its left side (Fig. 7c). A left subclavian artery may have the ductus arteriosus attached to it. A band connecting the left common carotid and the left subclavian artery may complete the ring.

Obliteration of the portion of the right aortic arch between the right common carotid and the right subclavian arteries results in an aberrant right subclavian artery arising as the last branch from the normal left aortic arch (Fig. 7d). The branch will cross from the left to the right side behind the oesophagus. Under exceptional circumstances the artery may cross the midline between the trachea and oesophagus, or even in front of the trachea, which would appear to be due to an abnormal development of the foregut in relation to the right aortic arch. Holzapfel (1899), in a study of 133 examples of aberrant right subclavian artery, found that 107 passed behind the oesophagus, 20 between the oesophagus and trachea, and six in front of the trachea.

Associated with the normally developed left aortic arch, slightly abnormal origins of the great branches of the arch may so alter the course of the vessels that they come to overlie the trachea and so produce some compression of this structure. This is particularly seen with a laevoposed right innominate artery and a dextroposed common carotid artery (Fig. 8).

Further descriptions of the development of the aortic arch and its abnormalities will be found in the publications of Congdon (1922) and Edwards (1948).
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Fig. 8.—Slightly abnormal origins of the vessels from the aortic arch can result in tracheal compression.

PATHOLOGY

Vascular compression of the trachea and oesophagus is thus produced by:

(a) A vascular ring
   (1) Double aortic arch
   (2) Right-sided aorta with a left-sided ductus arteriosus

(b) A partial vascular ring which may be completed by a band
   (1) Right-sided aorta with a left innominate artery and the ductus arteriosus running to it
   (2) Right-sided aorta with a left common carotid as the first branch and the ductus arteriosus running to the subclavian artery or the aorta.

Vascular compression of the trachea alone may be produced by laevo-position of the innominate artery and dextro-position of the left common carotid artery.

Vascular compression of the oesophagus alone may be produced by an aberrant right subclavian artery arising as the last division of the normal aortic arch.

Compression of the trachea results in maldevelopment and softening of the cartilages and a narrowing of the lumen some 2 to 3 cm. above its bifurcation. The walls are unduly lax and the pressure changes of respiration produce a stridor usually more marked on expiration. The line of the chondromalacia is somewhat oblique, rising from right to left in conformity with the direction of the arch or vessels. The mucosa in the region of the stricture may be oedematous and reddened due to persistent irritation, and considerable mucoid secretion is frequently present. The inflamed stricture acts as a ciliary barrier and there is a tendency for this secretion to accumulate below it. The longer and the more severe the compression of the trachea, the greater the lack of development of the lumen, which will remain as a permanent feature if relief is not provided early.

Although the lumen of the oesophagus is compressed, it is unusual for a true stricture to develop. Even though there is some local hypoplasia the pliable walls rapidly stretch to normal size after relief of the compression.

Other congenital derangements are not infrequently found with abnormalities of the aortic arch, and these include hemivertebrae, spina bifida, abnormal rib formations in the upper thoracic region, pulmonary stenosis, and the tetrad of Fallot.

SYMPTOMS

Only a proportion of patients with an aortic ring develop constriction at an early age. Symptoms may develop in middle age due to increasing rigidity of the aorta, but not infrequently the condition is totally asymptomatic.

The principal feature is "stridor," usually from birth, which, if the obstruction is severe, is associated with dyspnoea. There may be difficulty in taking foods, with regurgitation into the trachea and associated coughing and spluttering. When studied radiologically, there is a "build up" of the oesophageal contents at the level of the constriction and the weight of the material then forces some through. The process repeats itself and an intermittent type of swallowing is characteristic of the condition with possible overflow into the air passages. This inhalation of
food will be responsible for attacks of bronchopneumonia, and with any form of respiratory infection the stridor becomes more marked and may even be associated with severe respiratory embarrassment. As the child grows older it becomes conscious of the food temporarily sticking at a high level in the thorax and requires quite marked deglutitional efforts to get it through.

Pressure on the trachea by the vascular ring is relieved somewhat by extension of the head, and the infant may persistently take up an attitude of lying on its front or side, in which position head extension can more easily be maintained. If the head is manually flexed complete respiratory obstruction due to tracheal kinking may be produced. If the compression is severe the condition may be lethal in early infancy, but not infrequently there is some slow improvement in the symptoms. It is unusual for them to disappear completely, and recurrence of the stridor on exercise or with a respiratory infection is frequent. With a mild degree of tracheal narrowing stridor may be absent until the mucosa thickens following a tracheo-bronchial infection.

Severe symptoms in infancy are usually associated with the double arch, but children having a right aortic arch with a left ductus arteriosus or ligamentum arteriosum often carry on with mild and intermittent symptoms and may present for surgical treatment later on.

If the anomaly is an aberrant innominate or carotid artery arising from a normal left aortic arch, then no oesophageal obstruction or symptoms are present. If an aberrant right subclavian artery is the sole abnormality then dysphagia is the only symptom, dysphagia lusoria. This form of dysphagia was so entitled by Bayford (1794) from "lusus naturae," a trick of nature. His described case in a middle-aged woman was one of the less common varieties where the aberrant right subclavian artery runs between the trachea and oesophagus.

**Investigations**

A postero-anterior radiograph of the chest in the presence of a double aortic arch will show bilateral aortic knuckles, but these may not be easy to see, as they are usually somewhat smaller than a normal single arch. A right-sided aortic arch alone should suggest the possibility of an aortic ring completed by a ductus or ligamentum arteriosum. Aberrant vessels compressing the trachea may arise from either a right- or left-sided arch.

A barium swallow will show bilateral compression of the oesophagus at the level of D4 in the presence of a double arch (Fig. 9). A predominant posterior arch will give an oblique filling defect slightly higher on the right than on the left and the sub-aortic part of the oesophagus will lie a little to the right of the supra-aortic portion (Fig. 10). On the lateral view the size of the posterior arch or the posterior element of a double arch can be gauged by the magnitude of the defect (Fig. 11). A small defect suggests an aberrant right subclavian artery. An anterior oesophageal defect will indicate the rarer anomaly of a vascular channel running between the oesophagus and the trachea.

A little iodized oil run into the trachea may show similar bilateral compression in the presence of a double arch, or compression of the anterior surface on the lateral view when aberrant vessels are present.

An angiocardiogram will give considerable help in identifying the vascular channels and in particular the distribution of the branches of the arch or arches (Figs. 12, 13, and 14), but for practical purposes this is the least necessary investigation, as the anatomical state can be well examined at the time of operation and in any case bands will not be demonstrated. Nevertheless, unsuspected intracardiac defects may be shown which may be of prognostic importance and which may even be corrected at operation.

The observations obtained by direct endoscopy are, however, the most informative. Compression of the trachea by a pulsating vessel can clearly be seen, and it is important to note the changes in the cartilaginous structures in the region of the compression, in order to give a prognosis to the parents, as the degree of chondromalacia and local restricted development of the lumen may delay considerably the time when the stridor disappears after the operation.

Oesophagoscopy may demonstrate a pulsatile swelling elevating the posterior wall which is produced by a retro-oesophageal arch or branch of the arch or both. The vessel can be rolled under the tip of the oesophagoscope and the pulsations transmitted to it. Compression of the vessel against the front of the spine by the end of the oesophagoscope and simultaneous palpation of the pulse at the wrists will unequivocably diagnose an aberrant right subclavian artery by loss of the right radial pulse. If compression of a retro-oesophageal vascular structure produces no change in the two pulses then it is part of a ring of which there is a
Fig. 9.—Oesophagogram in a patient with an aortic ring.
FIG. 10.—Oesophagus with predominant posterior arch.

FIG. 11.—Lateral and oblique views of the oesophagus demonstrate the size of the posterior arch.

FIGS. 12, 13, and 14.—Angiograms showing double aortic arch (Fig. 12), dextroposed left common carotid artery (Fig. 13), and aberrant right subclavian artery (Fig. 14).
functioning anterior arch. If pressure produces weakening of the left radial or femoral vessels then there is a posterior arch with a non-functioning anterior arch, but very marked pressure has to be exerted completely to obliterate the vascular flow. The differential criteria may thus be tabulated:

<table>
<thead>
<tr>
<th>Type</th>
<th>Chest Radiograph</th>
<th>Retro-oesophageal Defect</th>
<th>Oesophagoscopy</th>
<th>Bronchoscopy</th>
</tr>
</thead>
<tbody>
<tr>
<td>Double aortic arch</td>
<td>Bilateral aortic knuckles</td>
<td>Yes</td>
<td>Large retro-oesophageal vessel. Compression gives no loss of pulse</td>
<td>Marked pulsation anterior wall of trachea with probable indentation</td>
</tr>
<tr>
<td>R. aortic arch with L. obliterated ductus</td>
<td>R.-sided aortic knuckle</td>
<td></td>
<td>Large retro-oesophageal vessel. Compression may give weakness of L. radial or femoral pulse</td>
<td>Constriction of trachea mainly from left</td>
</tr>
<tr>
<td>Aberrant L. common carotid or L. innominate</td>
<td>Normal</td>
<td>No</td>
<td>Nil</td>
<td>Pulsion across front of trachea with constriction</td>
</tr>
<tr>
<td>Aberrant R. subclavian</td>
<td>,,</td>
<td>Yes</td>
<td>Small retro-oesophageal vessel. Compression causes loss of R. radial pulse</td>
<td>Nil</td>
</tr>
</tbody>
</table>

Some conception can thus be obtained of what type of vascular anatomy is to be found. The presence, however, of residual bands, abnormal origins from the arches of secondary branches of the main trunks, and vessels running between the oesophagus and trachea make an exact diagnosis difficult. It might be considered that a too detailed investigation of the anatomy is an unnecessary refinement, as the surgical approach in all cases is the same and the vascular distribution can be studied after the operative exposure. All the investigations are relatively simple and may indicate abnormalities unsuspected on the clinical examination.

**Differential Diagnosis**

The main conditions from which vascular compression of the trachea and oesophagus must be distinguished in infancy are the various forms of congenital oesophageal stenosis and atresia, primary laryngeal and tracheal deformities, tracheo-bronchitis, and enlarged bronchial and tracheal lymph nodes.

**Indications for Surgical Treatment**

A chance finding of an aortic ring without symptoms in late childhood or adult life is no indication for operation. In any infant or young child, however, in whom a double aortic arch or right aortic arch is found radiologically, the question of stridor should be carefully inquired into, particularly in relation to exercise and respiratory infections. If there is a history of transient stridor under these circumstances bronchoscopy should be considered to see if the trachea is compressed. If no compression is seen then the aortic ring can be considered to provide adequate space for the trachea, or if it is a right aortic arch alone then the ligamentum arteriosum either runs to the right or if it is on the left then it is long and lax and no further action is called for. If endoscopic evidence of tracheal deformity is present then operation should be considered to prevent its becoming permanent and possibly increasing as the child gets older. As a principle it is better to undertake such an operation as early as possible in life to permit maximal growth of the trachea after relief of the constriction.

Where, in addition to the stridor, regurgitation or dysphagia is present, then operation is even more clearly indicated.

**Surgical Treatment**

**Pre-operative.**—In small infants bronchitis, bronchopneumonia, and tracheitis will require active treatment. Vapour tents, oxygen, and a course of antibiotics are frequently necessary in order to relieve the dyspnoea. (Elevation of the foot of the cot will permit gravitational drainage of the bronchial tree. Rarely a gastrostomy will be necessary if food cannot be taken without overflow into the respiratory system.) When the above methods do not produce any amelioration of the symptoms, then temporizing must be accepted as ineffective and a thoracotomy performed under circumstances which on occasion must be accepted as unfavourable.

**Operation.**—A thoracotomy from the left side through the fourth interspace is made. A wide exposure should be made as for a coarctation of the aorta, and, if necessary, the posterior ends of two or three ribs should be divided. The lung is retracted and the mediastinal pleura is divided. The thymus is dissected upwards and retracted with a stitch, and if it is voluminous a portion of it can be removed. The left vagus nerve is identified and the position of the left recurrent laryngeal nerve located. If a left innominate artery is present with a right-sided ductus
Fig. 15.—Anatomical details of 11 cases of double aortic arch submitted to operation. The site of division of the ring is shown by the arrow.
arteriosus then the nerve will probably be found in relation to the left subclavian artery. A clear definition of the aortic arches, ductus arteriosus or its remnants, the branches of the aorta, and any residual constricting bands is made. The presence of any areas of coarctation is noted and before any division is made the exact path of the circulation is established. The proposed line of section is then temporarily clamped and it must be clearly demonstrated that the carotid vessels and the descending aorta are receiving an adequate circulation before any final division is performed. As a principle the smaller limb of the ring is the one to be divided, but this may not be so, as the larger limb may have a coarcted area in it which in fact reduces the blood flow through it to a negligible amount. If the diameter of the site of proposed section of the constricting ring is small then division can be made by simple ligation and transfixion suture at each end, but if the vessel is large then clamping of the vessel with a formal suturing of the two ends with 0000 silk is preferable. Division of the ductus if patent or obliterated and any other constricting bands or aberrant vessels will be required. After relief of the constriction the anterior part of the ring may appear to lie in direct contact with the trachea. Gross advises slingling this portion of the ring to the back of the sternum or the neighbouring costal cartilage by picking up the adventitia of the vessel with sutures which pass to the peristium of the back of the sternum or through the muscles of an intercostal space.

Where two arches of equal size are present, the point of division will depend upon the distribution of the vessels. Division of the posterior arch will probably be more difficult technically than division of the anterior arch. Division through a coarcted or narrowed area, if present, seems an obvious choice as the circulation will have already adapted itself to this state.

In all cases of double aortic arches the ligamentum arteriosum should be identified. It may, in fact, be producing a second constriction below the first. Aberrant right innominate and left common carotid vessels producing tracheal compression are treated by anterior elevation of the vessels and stitching them to the back of the anterior chest wall. The division of an aberrant right subclavian artery usually presents no difficulty.

Post-operatively those patients who have had tracheal compression may develop a temporary increase in respiratory difficulty. It is thus advisable that all such cases should be housed in a vapour tent with oxygen and that antibiotic treatment be continued.

Complete relief of the stridor may be delayed, and some residual symptoms may persist for many weeks until the trachea resumes its normal shape and cartilaginous consistency.

### Results

Sixteen cases of vascular obstruction have been operated upon for symptoms. Eleven had a double aortic arch and five an aberrant right subclavian artery. Gross (1953) in his series records 21 cases of double aortic arch, 15 cases of right aortic arch with a constricting left ligamentum arteriosum, and 12 cases of aberrant subclavian artery (11 right and one left). We have had no case of a simple right aortic arch with a left ligamentum arteriosum presented to us for surgical treatment. The age group is as follows:

<table>
<thead>
<tr>
<th>Total No.</th>
<th>Sex</th>
<th>Age Group</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td>0–1 Year</td>
</tr>
<tr>
<td>Double aortic arch</td>
<td>11</td>
<td>5</td>
</tr>
<tr>
<td>Aberrant right subclavian artery</td>
<td>5</td>
<td>3</td>
</tr>
</tbody>
</table>

The anatomical details and site of division of the ring in the 11 cases of double aortic arch is shown in Fig. 15.

All cases survived operation. Although dysphagia, when present, appeared to be relieved immediately, the stridor frequently took some months to disappear, and even then a mild stridor might be noticeable if a tracheo-bronchial infection occurred. It is confidently expected, however, that, in the course of time, as the trachea grows this symptom will completely disappear.

### Summary

Vascular compression of the trachea and oesophagus due to development abnormalities of the aortic arch and its branches is of clinical significance. It produces symptoms in some 1% of all cases of congenital abnormalities of the heart and great vessels.

Its embryology is discussed.

Cases of persistent stridor and dysphagia in infancy may be due to this condition. It can be cured by surgical division of the obstructing vessels and may prevent the frequent fatal outcome.
The type of constriction can be accurately diagnosed in most cases by radiographic study and endoscopy.

The results in 16 cases, treated surgically, are described.

I am grateful to my colleague, Mr. B. J. Bickford, for permitting me to include the cases operated upon by him, to Professor John Hay and Dr. Gordon Farquhar for referring the cases to us, and to Dr. G. Jackson Rees and Dr. Alan Stead for their handling of the anaesthetic problems.

The Oxford University Press have kindly given permission for the reproduction of Fig. 3.

My thanks are due to Miss Barbara Duckworth for preparing the diagrams.
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Thorax 1959 14: 187-200
doi: 10.1136/thx.14.3.187

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