Arterial Malformations which Cause Compression of the Trachea or Esophagus

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There are many vascular malformations in the superior mediasitum which have little or no clinical significance, but some arterial anomalies in this region assume importance because an abnormal or a displaced vessel can compress the trachea or the esophagus (or both) and partly obstruct these vital passages. Since surgical methods are now available for thoracic exploration of even the youngest subjects, it is important to recognize those malformations which threaten health or life; certain of these can now be brought under surgical attack, thus affording relief of the esophageal or tracheal obstruction.

Arterial derangements within the chest are common, can be complex, and can assume many forms. These facts are confirmed by a very extensive literature. A presentation of monographic size would be necessary to list and describe all of the anomalies which have been encountered. It is intended here to deal only with those, which so far have been found at the operating table when attempting to relieve an existing tracheal or esophageal compression. The following sections are based on the study and surgical care of 70 babies and children with such vascular anomalies. Occasionally, one of these vascular malformations might come to light in an adult, but almost certainly the vast majority of malformations, which are serious enough to cause important troubles, will come to the attention of a clinician or a surgeon in the early years of life, as was the experience in the series of cases described here.

The arterial malformations for which surgery has some therapeutic value include double aortic arch, right aortic arch with a left ligamentum arteriosum, anomalous innominate artery, anomalous left common carotid artery, and aberrant right subclavian artery.

Double Aortic Arch

Pathologic Anatomy. The fundamental pathologic change is concerned with the fact that the ascending aorta bifurcates into two branches, one passes in front of and to the left of the trachea, while the other progresses to the right of the trachea and esophagus; both limbs then join to form a descending aorta (figs. 1 and 2). In the majority of cases, the left (anterior) arch is the smaller of the two; in a minority, the right (posterior) arch is the smaller in size. The descending aorta is generally on the left side of the spinal column, but in an occasional case it is to the right of the midline (fig. 3). The space between the two aortic arches is insufficient for accommodation of a trachea and esophagus of normal size. Therefore, both of these structures become compressed in the crowded region between the two aortic segments.

Clinical Picture. In rare cases, the space provided between the two aortic arch limbs is relatively large; the encircled trachea and esophagus have a fair amount of room and there is little or no attendant difficulty from interference with the functions of these two pathways. However, most subjects with a
Fig. 1. Double aortic arch producing compression of the trachea and esophagus. Left. Preoperative state. There is a small anterior arch and a large posterior limb, which join and form the descending aorta. Right. Surgical therapy by dividing the ligamentum arteriosum and also the anterior arch. The left common carotid artery is tacked forward to the sternum, thus keeping it from pressing on the trachea. These various steps give sufficient room for the trachea and the esophagus to bulge to the patient's left and be relieved of constriction.

double aortic arch have very little room between the two arterial limbs, so that the esophagus and particularly, the trachea, are greatly compressed. While a double aortic arch is compatible with a long life and relatively minor symptoms, the anomaly is usually a serious one and has often led to fatality within the first year or two of life; the marked tracheal obstruction has led to superimposed pulmonary infection which has overwhelmed many of these babies.

Most human subjects with a double aortic arch have sufficient symptoms to come to the attention of a physician during infancy. There may be mild or moderate hesitation in swallowing. Of greater significance are the alarming symptoms which come from tracheal narrowing. The respiratory rate is generally increased. The baby struggles to obtain an adequate exchange of air, an effort which often requires the use of the accessory muscles of respiration. During inspiration there may be intercostal and suprasternal retraction. A loud wheeze can be heard by stethoscopic auscultation, but usually the noise is great enough to be heard with the unaided ear many feet or yards away. There is apt to be a “crowing” type of breathing, with a marked inspiratory and expiratory stridor. Respiratory distress is very apt to be made worse during or immediately after the swallow-
CLINICAL PROGRESS

Fig. 2. Double aortic arch with large anterior limb and a smaller posterior limb, producing compression of the trachea and esophagus. Above. The two drawings show the anterior and posterior views of the anomaly. Below. Treatment of the condition by division of the small posterior arch, thus returning all structures to normal.

Ining of milk or food. In severe cases, symptoms are serious enough to demand oxygen therapy. These babies are prone to lie with their heads in hyperextension, a position which tends to attenuate the trachea and push away from its anterior surface any structure which is impinging upon it. If the examiner forceably straightens the head, or flexes it toward the sternum, the exchange of air is reduced or can even be completely shut off, though the respiratory movements of the thorax continue.

Roentgenographic Findings. Roentgenograms of the chest may show some pneumonitis, if they happen to be taken at a time when there is superimposed lung infection. Films taken during inspiration may indicate poor or irregular aeration of the lungs, whereas during expiration there is apt to be an appearance of hyperaeration. Lateral films, if made at the correct exposure, can frequently outline the trachea by the air which it contains. Thus, without the use of a contrast medium, a fair appraisal can be made of the anteroposterior diameter of the trachea. While its upper portion is of normal caliber, the lower part has a markedly reduced lumen and the structure is apt to be displaced forward. A swallow of barium shows indentation of the posterior wall of the esophagus at the level of the third or fourth thoracic vertebra, this defect being nearly a horizontal one, and occurring at a level close to that where the tracheal compression is found. If further evidence regarding the status of the trachea appears to be desirable,
Fig. 3. Double aortic arch with the aorta descending on the right, constricting the trachea and esophagus. Above. Anterior and posterior drawings of the anomaly. The left arch is the smaller of the two. The ligamentum arteriosum between the pulmonary artery and the aorta also forms a part of the constricting mechanism. Below. Method of surgical therapy. The smaller left arch has been divided and cut away from the descending aorta; the left subclavian artery has been divided at its origin. The left common carotid artery has been tacked forward to keep it off of the trachea. The ligamentum arteriosum has also been cut.
this can be gained by lipiodol injection and delineation; for children under a year of age this can generally be done without anesthesia, but for older subjects it is best carried out under some form of general narcosis. Anteroposterior films show a definite compression of both sides of the trachea in its lower third; lateral views shows a constriction which is striking. This narrowing is at a level approximating that where the defect was found on the posterior wall of the esophagus. The combination of a posterior esophageal compression and an anterior tracheal defect (when found in the absence of a demonstrable mediastinal mass) is almost certain proof of some type of an encircling “vascular ring”. The size of the posterior esophageal indentation is usually a fair indication of whether the posterior arch is the larger or the smaller of the two.

Surgical therapy. Surgery has much to offer these patients, because division of the smaller arch provides more room for the esophagus and particularly for the trachea. Without discussing the details of operative steps, the surgical solution of the problem is indicated in figs. 1, 2, and 3. Most commonly, it is the anterior arch which is the smaller of the two and thus is the one which must be severed. In a minority of cases, the posterior arch is the smaller and is the one which must be divided. In some cases the pulmonary artery has been held in a retro-displaced position by virtue of its attachment to the aorta through the ligamentum arteriosum. This ligament should be divided, to allow the pulmonary artery to fall forward. While this additional step seems to have little value for some patients, it is certainly an important one for others.

Results of therapy. Of the 26 patients with double aortic arches who have come to operation, 16 had the aorta descending on the left and 10 on the right. Of the 16 with a left descending aorta, there were 11 in whom the left (anterior) arch was the smaller of the two and was accordingly divided, and there were five in whom the right (posterior) limb was the smaller one and was therefore sectioned. Of the 10 patients who had double arches and a right descending aorta, it was always the left (posterior) one which was severed. These 26 sub-

jects ranged in age from one month to three years. There have been five deaths; two from hemorrhage at the operating table, one from cerebral edema the day following surgery (the child had been dangerously ill and in an oxygen tent for several months prior to surgical therapy), and two from pneumonia 3 and 10 days after operation. The 21 surviving patients have been followed for varying lengths of time, up to nine years, since operation. They have had extraordinary relief of symptoms. Usually, a marked improvement in the airway has been noted immediately after operation. In all instances, the exchange of air has obviously been more free, and the subjects have not had to use the accessory muscles of respiration for ordinary activity. Stridor has usually vanished, but in a few instances a trace of it remains. Postoperative tracheograms have usually shown marked improvement in size of the tracheal lumen after operation, but generally some deformity persists in the cartilages. It is reasonable to believe that the removal of all external pressure will give these tracheas a better chance to develop in future years as the children progress in body growth.

The overall results postoperatively have been exceedingly dramatic and have proved beyond a doubt that an operative attack on this vascular abnormality has great merit. This does not imply that all humans with a double aortic arch should necessarily be operated upon, because there are doubtless a few who tolerate the condition in a reasonably satisfactory way through a long life. However, it appears that most infants and young children with this abnormality are extremely apt to develop serious or even fatal complications at some subsequent time, and, hence, we feel that it is generally highly desirable to undertake surgical correction of the vascular anomaly whenever it is discovered.

Right Aortic Arch With Left Ligamentum Arteriosum

Pathologic anatomy. The first part of the ascending aorta lies in a normal position, but, instead of being directed upward and to the left in front of the trachea, it ascends and passes to the right of the trachea and the esophagus,
and then continues as a descending aorta which may lie either to the left or to the right of the vertebral column. A right aortic arch in itself causes little or no disturbance, though at times it has been known to press on the right main bronchus or one of its branches and has led to pathologic changes in the lung. If the ligamentum arteriosum, or a patent ductus, runs from the pulmonary artery to the left of the trachea and the posterior aspect of the esophagus to join the descending aorta, this completes a constricting ring which encircles the esophagus and trachea (fig. 4). If this ring is sufficiently large, the functions of the esophagus and the trachea are not altered to any important degree, but if the ligamentum arteriosum is taut, the
space within the encircling structures is so small that the compressed esophagus, and especially the trachea, are greatly disturbed.

Clinical picture. With this anomaly, symptoms can develop quite similar to those which are produced by a double aortic arch, but generally are apt not to be quite so severe. As a rule, the onset of complaints is delayed until a bit later in childhood; the patients that we have encountered were on an average a few years older than those with a double aortic arch, the latter usually being seen in infancy. There may be a crowing type of respiration, some intercostal and suprasternal retraction, possibly recurrent pulmonary infection, and occasionally even some hesitancy in swallowing. The respiratory symptoms are generally aggravated during and following deglutition.

Roentgenographic findings. There may be evidence of pulmonary infection. There is a prominent shadow (aortic arch) projecting from the right side of the superior mediastinum. During inspiration there may be incomplete aeration of the lungs, and during expiration there may be hyperaeration, indicative of an obstruction somewhere above the carina. Lateral films of the chest, if appropriately exposed, can outline the air-filled trachea, showing the upper portion to be normal, whereas the lower segment is distinctly narrowed. Instillation of lipiodol into the trachea gives better visualization, shows a slight and rather elongated indentation of the right wall of the trachea, imposed by the adjacent aortic arch, indentation of the anterior surface of the trachea where the pulmonary artery is pulled against it, and a depression on the left side of the trachea from the ligamentum arteriosum. With a swallow of barium, one finds, at the same level as the tracheal defects were observed, that the esophagus has a narrow but very deep constriction in its left-lateral and posterior surfaces. Above this posterior notch of the esophagus, it is sometimes possible to identify a separate defect, passing obliquely upward and to the left, which is caused by the left subclavian artery which arises from the aorta and passes behind the esophagus to reach the left apex of the chest. By fluoroscopic and film studies, it is usually possible to recognize accurately this combination of vascular anomalies, but in some instances it is almost impossible to differentiate it from a double aortic arch which is combined with a right descending aorta.

Surgical therapy. Treatment of this condition is obviously one of alleviation and not of cure; it is impossible to shift the aortic arch from its right-sided bed, but we can divide the ligamentum arteriosum, which thus breaks the constricting "ring". The ligamentum arteriosum should also be divided. When these steps are properly performed, the pulmonary artery falls forward into a more normal position and sufficient room is given for the esophagus, and particularly the trachea, to drop backward and to the left. Whenever the left subclavian artery is found behind the esophagus, it, too, should be severed.

Results of therapy. Eighteen patients with this anomaly have been operated upon, the youngest two months and the oldest 12 years of age. There have been no deaths. The result in all subjects has been striking improvement. There has always been complete disappearance of any pre-existing dysphagia. There has been marked alleviation of respiratory distress which all subjects had prior to operation. In one of the older children, repeat tracheograms at the end of one year showed only disappointing growth of the tracheal rings in the involved area; this case emphasizes the desirability of carrying out these operations at an earlier time in life so that removal of all external pressure will give the trachea the best possible chance to enlarge during the growth period of the child.

It has long been known that a considerable number of humans possess a right aortic arch, but have few symptoms therefrom. Presumably, these have had a ligamentum arteriosum in front of the trachea or else this structure, even though it passes to the left and posterior aspects of the trachea and esophagus, has been very long and lax. In such instances operative intervention is not required. However, it is important to recognize those occasional individuals who do have a high degree of tracheal narrowing from one of the encirclings under discussion, because it is in this group that surgical therapy is a procedure of great value, particularly when it is performed early in life.
before the tracheal wall has become permanently deformed.

**Anomalous Innominate Artery**

*Pathologic anatomy.* The innominate artery can originate at a point farther along on the arch than is normal; when it does so, it must wind around the anterior surface of the trachea as it courses upward and to the right, to reach the right apex of the chest. If this vessel is large and taut, it can compress the trachea to a serious degree (fig. 5). While the pathologic anatomy is seemingly only a minor variation from normal and would appear to be of little consequence, the severity of symptoms which can accompany it has been emphasized by several patients who have been studied and found to have this anomaly.

*Clinical picture.* There can be inspiratory and expiratory crow of considerable degree, intercostal and suprasternal retraction, marked dyspnea, and even superimposed pneumonitis. Tracheal compression can be sufficiently severe to require the use of an oxygen tent. The head is apt to be held in hyperextension, which apparently improves the exchange of air for the child. There is no hesitancy in swallowing, nor are the respiratory symptoms made worse during feeding.

*Roentgenographic findings.* By roentgenologic study, the esophagus is entirely normal. The aortic arch is normal in size and in position. Visualization of the trachea may show little in the anteroposterior view, but lateral projections indicate a considerable narrowing in its lower third, the indentation appearing entirely on the anterior wall, whereas the posterior wall is straight and in normal alignment. It may be exceedingly difficult, or indeed impossible, to determine whether the base of the innominate artery or the base of the left common carotid artery (described in the next section) is at fault, but in some instances the obliquity of the anterior tracheal defect suggests which of these vessels is the offending one.

In the absence of any anterior mediastinal shadow (such as a cyst or neoplasm) a tracheal defect of this sort certainly suggests some abnormal blood vessel in the region as a cause of the trouble. Any narrowing of the trachea which hints of the presence of an abnormal blood vessel of this sort can raise the question of whether the diminished lumen of the trachea is dependent upon incomplete development of its tracheal cartilages. Data on this point can be gathered from direct tracheoscopy and also by serial observations made roentgenologically throughout the respiratory cycle. If the tracheal defect is due to some external pressure, the narrowing remains relatively constant throughout the respiratory cycle; if the narrowing is due to deficient cartilagenous rings, the caliber
of the trachea will increase during inspiration and decrease during expiration.

Surgical therapy. After dissecting off and discarding the thymus, it is possible to view the aortic arch and the vessels in question. The base of the innominate artery can be seen wrapped around the anterior surface of the trachea, causing a decided indentation. Because of possible consequent brain damage, the offending vessel should not be severed. However, the first part of this artery and the accompanying aortic arch can be drawn forward by sutures taken through their adventitias and running these silk sutures forward through the substance of the sternum. Appropriate tension on these silk sutures draws the vessels forward and thus carries them away from the trachea, thereby completely relieving the preexisting external pressure (fig. 5). These maneuvers are relatively easy to perform.

Results of therapy. Nine patients, all under one year of age, have been operated upon; there have been no deaths. The results of these undertakings have been very gratifying. The first patient had previously been hospitalized for four months because of severe respiratory distress; she was discharged eight days after operation, completely free of symptoms. The other patients have likewise been dramatically relieved of their respiratory complaints.

Anomalous Left Common Carotid Artery

Pathologic anatomy. This malformation is probably very rare. It is somewhat akin to the anomalous innominate artery described in the last section. The vessel is of normal size and distribution, but has an origin which permits it to branch off of the aortic arch more to the patient's right than is normal (fig. 6). It must therefore wind around the anterior surface of the trachea as it courses upward and to the left, to reach the left apex of the chest.

Clinical picture. The symptoms produced will obviously depend upon the degree of pressure which is exerted upon the front of the trachea. There may be moderate respiratory distress, some crowing during respiratory movements, dyspnea of mild to moderate degree, and possibly bouts of pulmonary infection. There are no disturbances in swallowing.

Roentgenologic findings. By roentgenographic examination, the findings are quite similar to,
and often indistinguishable from those described in the last section for anomalous innominate artery. The esophagus is normal by barium study. The trachea has a posterior wall which is sharp, distinct, and of normal contour and position, whereas the anterior surface of the trachea shows an indentation just at or above the level of the aortic arch, there being no anterior mediastinal cyst or tumor to account for these changes. Films taken at various angles may show that the tracheal defect is a grooved one, running upward and obliquely to the left, and strongly suggesting that the indentation is due to the unusual origin and pressure of the left common carotid artery. However, it is often difficult to see that such an obliquity exists, and hence the roentgenologist may not be able to determine which vessel is the offending one.

Surgical therapy. The surgical treatment for this condition is similar to that described in the last section for anomalous innominate artery. The first part of the left common carotid artery impinges against the trachea; this could be relieved by division of the vessel, but the chances of producing cerebral ischemia are so great that this maneuver should be avoided. The therapy consists of pulling the carotid artery and the adjacent portion of the aortic arch forward with mattress sutures which pierce the substance of the sternum. Appropriate tension applied through these silk sutures draws the common carotid artery and the arch forward so that they no longer are in contact with the trachea.

Results of therapy. Five patients have been subjected to this undertaking. In each instance, there has been complete relief of the preexisting respiratory symptoms.

Aberrant Right Subclavian Artery

Pathologic anatomy. As long ago as 1794, Bayford gave a fascinating description of the clinical picture and the relevant pathology in a case of an aberrant right subclavian artery which produced "dysphagia lusoria". The right subclavian artery, instead of arising in a normal fashion from the innominate artery, takes off independently from the distal part of the aortic arch. It then ascends and passes towards the right, to reach the right apex of the chest. It is rare for this artery to course in front of the trachea and it is uncommon for it to pass between the trachea and esophagus. In the majority of cases, it runs behind the esophagus in its upward path.

Clinical picture. An aberrant right subclavian artery is often recognized by radiologists who see a posterior indentation of the esophagus during barium studies of the alimentary tract. In a very high percentage of subjects there have been no important disturbances in the swallowing function. However, in a few cases, the subclavian artery can stretch sufficiently taut around the esophagus so that the peristaltic wave in the latter is altered and the patient experiences hesitation in swallowing which may be bothersome and indeed can interfere with the nutrition because of the diminished intake of food. In babies, delay in swallowing can be found in the early months of life. The subject is hungry and is eager for food and will start to swallow in an apparently normal way; he then has difficulty in getting the bolus started along the esophageal pathway. Some of the food or milk may pass downward, while the remainder stays in the pharynx for a considerable period of time, or is expectorated. Some of these patients have no delay in the swallowing of milk or other fluids, but do encounter difficulty when solid or semisolid food is later added to the diet. At no time is there respiratory distress, but some aspiration of material while attempting to swallow is not uncommon, so that gurgling or noisy respirations appear at such times.

Roentgenographic findings. By roentgenographic examination, the abnormality can be identified quickly and with certainty. The aortic arch is normal in size, position, and direction. The trachea shows no abnormality. With a swallow of barium, the lateral view shows a defect of rather small caliber on the posterior wall of the esophagus at the level of the third or fourth thoracic vertebra. By anteroposterior view, this indentation runs obliquely upward and towards the patient's right, the direction and position of this defect being in a line from a distal part of the aortic arch to the right apex of the chest. There is usually little or no ballooning of the esophagus above this zone, but a delay in the passage of
barium is often observed, due to an interference with progression of the peristaltic wave.

Surgical therapy. The surgical undertaking for the treatment of dysphagia lusoria is a rather simple one. It consists merely of exploration of the posterior mediastinum, freeing the anomalous artery from its bed, doubly ligating and dividing it. There are always sufficient collateral vessels to provide an adequate supply of blood to the right arm.

Results of therapy. In our series, 12 patients have been operated upon; these varied in age from three weeks to six years. All subjects survived this surgery and in each instance there was complete disappearance of the dysphagia which had existed prior to operation.

Summary

In recent years observations have shown that there are certain arterial malformations in the superior mediastinum which bring about significant compression of either the trachea or esophagus, or both of these structures. If the arterial derangements are of such a nature that symptoms are produced by obstruction of the esophagus and particularly by the impairment of the tracheal airway, it is of prime importance to recognize the underlying vascular malformation because surgical approach to the problem has a great deal to offer either by division of an offending vessel or by displacement of an artery in such a manner that more room is provided for the esophagus and trachea.

Roentgenographic examination of the esophagus and trachea with contrast media usually gives a fairly clear impression of the arterial derangement and leads to accurate diagnosis. Visualization of the aortic pathways by angiocardiography or by retrograde aortography is seldom necessary.

At operation, all these anomalies can be exposed through a left anterolateral transpleural approach to the superior mediastinum. Most of the thymus gland can be dissected off and discarded, allowing good visualization of the region.

Emphasis must be placed upon the fact that constrictions of the esophagus or trachea are not caused solely by vessels or ligaments, but that constriction is likewise produced by fibrous bands or sheaths which accompany these vessels and ligaments. Hence, it is important, not only to divide or displace the appropriate vessel or ligament, but also to cut any strands or bands of tissue which accompany these structures and which form a part of a constricting mechanism.

In a series of cases in which surgical therapy has been undertaken for alleviating compression of the esophagus or trachea for the various vascular anomalies under consideration, it has been shown that the risks of surgery can be kept relatively low and that the benefits which accrue from such treatment are real and well worth seeking.

REFERENCES

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