AORTIC VASCULAR RINGS ENCOUNTERED IN THE SURGICAL TREATMENT OF CONGENITAL PULMONIC STENOSIS*

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RECENT ADVANCES in the surgical treatment of patients with congenital heart disease have been responsible for the visualization of many anomalies of the great vessels previously not reported or seen only rarely in the dissecting room.[1] Gross and Ware[2] described the surgical treatment of anomalies of the aortic arch which may cause tracheal or esophageal obstruction. In such instances all or part of a vascular ring encircles the trachea and esophagus. It is the purpose of this report to describe the variations of such a vascular ring encountered in the surgical exploration of 841 patients for suspected congenital pulmonic stenosis.

A large and unexpected number of anatomic variations have been encountered in the treatment of congenital pulmonic stenosis, an example of the known fact that congenital defects are apt to be multiple. Variations of an aortic vascular ring have been encountered in 40 instances and other cases have probably escaped our notice. The observed cases have taken one of three forms: (1) A subclavian artery which arises from the medial side of the aortic arch as the last branch and passes behind the esophagus to the arm contralateral to the side of the aortic arch; (2) a retro-esophageal innominate artery; and (3) the persistence of both right and left embryologic fourth aortic arches.

EMBRYOLOGY†

During the first three weeks of embryonic life six aortic arches join the ventral aortic sac and the dorsal paired aortas around the interposed pharynx. The aorta caudal to the branchial arches becomes fused as an unpaired dorsal aorta. Figure 1-A shows diagrammatically the approximate configuration of the aortic arches in the 12 mm. embryo. The embryonic subclavian artery is the seventh cervical segmental artery which supplies the limb bud. As the heart and aortic sac move caudad and the cranial portion of the embryo elongates, the subclavian artery moves craniad on the aorta. Normally the caudalmost segment of the right fourth arch becomes obliterated as the left fourth arch takes over the majority of the cardiac output. Most of the right fourth arch becomes the first portion of the subclavian artery. The right third arch persists as the proximal part of the carotid artery. It ultimately arises from the fourth arch, which persists as the innominate and subclavian arteries. The left fourth arch persists as the definitive aorta, and the carotid and sub-

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clavian arteries arise independently. In rare instances the left subclavian artery may ascend to the third arch, and a left innominate artery results.

If the left instead of the right arch becomes obliterated, a right aortic arch results, an anomaly which has been present in about 20 per cent of patients with congenital pulmonic stenosis. If the proximal rather than the distal part of the right fourth arch becomes obliterated, the right subclavian artery arises from the unpaired aorta and courses behind the esophagus to reach the right arm (Fig. 1B). In rare instances the vessel may go between the trachea and esophagus or anterior to the trachea.[4] A short segment of the distal end of
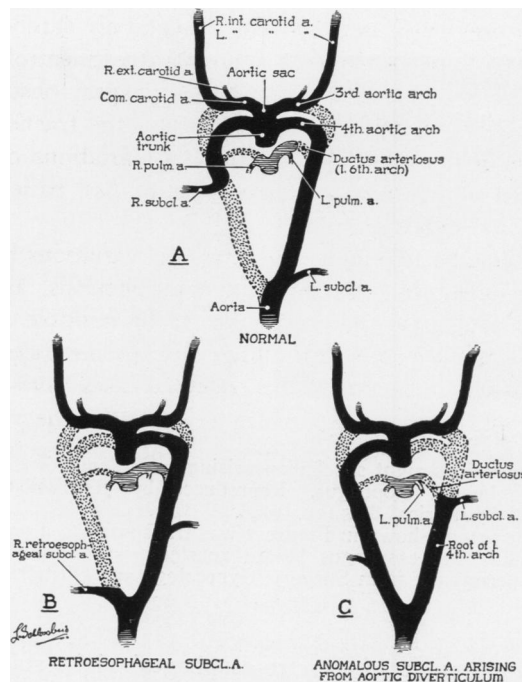

FIG. 1.—Development of aortic arch and anomalies. Diagram of aortic arches in 12 mm. embryo. Stippled segments disappear. See text.

the right fourth arch may persist as an aortic diverticulum from which the subclavian arises. If the right aortic arch persists and the left arch becomes obliterated, the mirror image of the above described arrangement may occur.

The ductus arteriosus usually connects the main or the left pulmonary artery with the left aortic arch. When the proximal left fourth arch becomes obliterated, the ductus arteriosus is connected with the distalmost portion of the left dorsal aorta along with the subclavian artery (Fig. 1C). If the arch becomes obliterated distal to the ductus arteriosus, this relationship is lost.

When both aortic arches persist, a complete aortic ring is formed about the trachea and esophagus. In such instances the brachiocephalic vessels usually

arise from the arches independently, although double aortic arches with an innominate artery have been described.[5, 6]

REPORT OF CASES

A retro-esophageal subclavian artery has been encountered in 36 patients in our series. The anomaly was equally distributed between the right and left subclavian arteries. In no case was there a history of tracheal or esophageal obstruction, although attacks of cyanosis and dyspnea attributed to the congenital cardiac deformity may have been partially caused by the anomalous vessel.
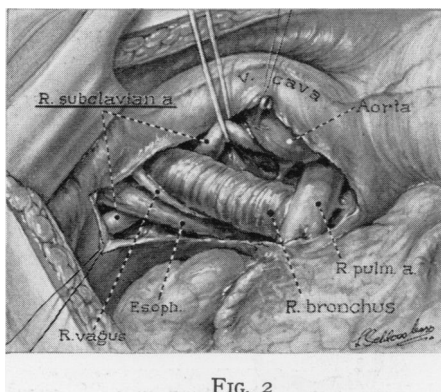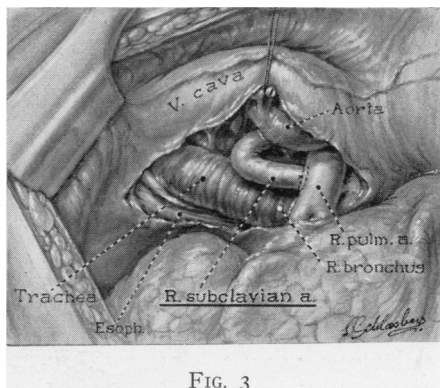

FIG. 2.—Anomalous right subclavian arising from descending aorta and entering right chest behind esophagus. Reproduced by permission from Surgery, Gynecology and Obstetrics, 87: 100, 1948.

FIG. 3.—The artery shown in Figure 2 was transposed to a position anterior to the right bronchus and esophagus before anastomosis to the pulmonary artery. Reproduced by permission from Surgery, Gynecology and Obstetrics, 87: 100, 1948.

The usual appearance of a retro-esophageal right subclavian artery is shown in Figure 2. The vessel arises from the medial side of the left descending aorta and enters the right chest behind the esophagus. In no case in this series has such an anomalous vessel passed between the trachea and esophagus or anterior to the trachea. An anastomosis may be performed without altering the position of the subclavian artery or it may be brought anterior to the bronchus and esophagus as shown in Figure 3. A retro-esophageal subclavian artery is well adapted for the creation of an artificial ductus arteriosus.

An unusual type of anomalous left subclavian artery is shown in Figure 4. There is an aortic diverticulum from which the subclavian artery and obliterated ductus arteriosus arise. The anastomosis of the subclavian artery to the pulmonary artery is shown in Figure 5.

In one instance the innominate artery arose from the aorta and passed to the left behind the esophagus.[1] Its subclavian branch was suitable for an anastomosis.

The presence and course of the ductus arteriosus has been variable. In only several instances has it been connected to a retro-esophageal vessel. When present, the recurrent or inferior laryngeal nerve loops around it. If there is no ductus arteriosus, the inferior laryngeal nerve may enter the larynx directly,
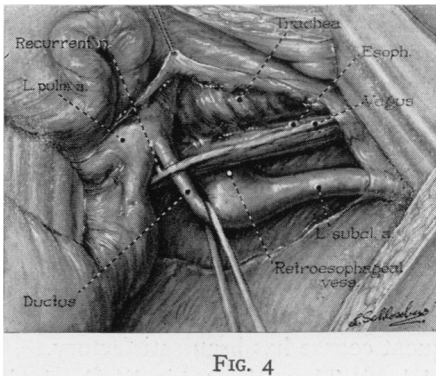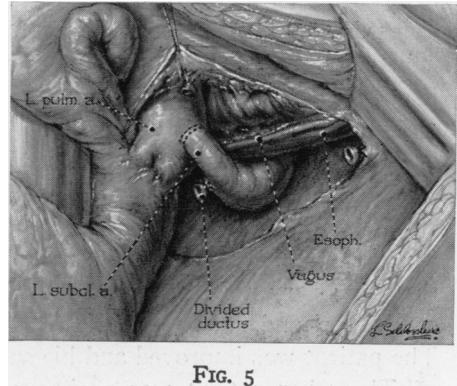

FIG. 4.

FIG. 5.

FIG. 4.—Unusual retro-esophageal left subclavian artery arising from an aortic diverticulum with obliterated ductus arteriosus.

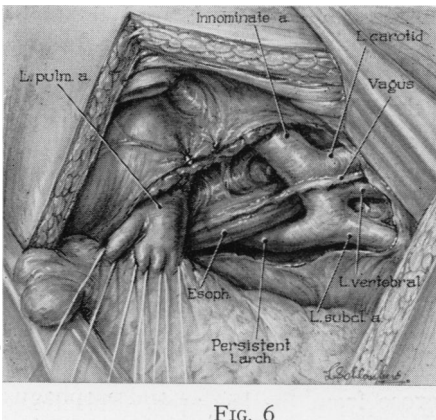FIG. 5.—Anastomosis of the subclavian artery shown in Figure 4 to the left pulmonary artery.
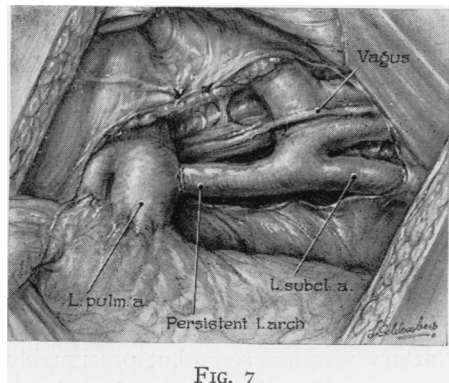

FIG. 6.

FIG. 7.

FIG. 6.—Left side of double aortic arch. The size of the vessel diminishes beyond the origin of the left subclavian artery.

FIG. 7.—Anastomosis performed using divided end of left aortic arch shown in Figure 6 and the left pulmonary artery.

loop around the vertebral artery, or loop around the inferior thyroid artery.[7] In such instances, of course, the nerve is not seen during thoracotomy.

There were three patients who had a complete aortic ring. The diagnosis in the first patient was mentioned at operation but not appreciated until she died of a ball embolus 14 days later. The patient had a marked scoliosis, and before operation there was considerable doubt about the side of the aortic arch. At

operation on the right side, deviation of the descending right aortic arch to the left was attributed to the scoliosis. This patient has been reported previously.[1]

The second patient was an eight year old boy with history and physical findings typical of the tetralogy of Fallot. Fluoroscopy with barium swallow demonstrated a right aortic arch with a retro-esophageal vessel. It was thought that this vessel might be a left descending aorta or more probably a small left subclavian artery. At operation (Figs. 6 and 7) the innominate artery was as usual the first branch of the right aortic arch. It gave rise to the left carotid and subclavian arteries and another vessel which was probably the left vertebral. In addition, there was a vessel which descended behind the esophagus. Upon occlusion of this vessel it was learned that a pulsating flow entered the vessel from either end and there was no diminution of pulses in the legs. The junction of this vessel with the aorta could be felt but not seen. It was thought that the artery represented a persistent left aortic arch. It was divided at its caudal end and the proximal end anastomosed to the left pulmonary artery. The patient was improved and his subsequent course has been satisfactory.

The other patient was a six-year-old boy. Throughout his life he had experienced frequent upper respiratory infections. He often had a loud barking cough. There were frequent fainting spells which had decreased with increasing age but occurred about twice a month at the time he was first seen. He was occasionally cyanotic at rest and definitely but not strikingly so on exertion. The physical findings were those of the tetralogy of Fallot with moderate incapacity. Oximeter studies showed a fall of his arterial oxygen saturation from 88 per cent to 84 per cent on exercise. Cardiac catheterization and angiocardiography were compatible with the diagnosis of tetralogy of Fallot but no information was obtained about the aortic arch. Roentgenography with barium swallow showed bilateral esophageal indentation in the anterior-posterior projection and indentation from behind in both the right and left anterior oblique positions. It was thought that the main aortic arch was on the left side, but that a double aortic arch had been responsible for the fainting spells in the absence of marked cyanosis and anoxemia. At operation on the left side a double aortic arch was found. The anterior and smaller arch was the left from which the subclavian artery arose. The origin of the left carotid artery was not seen, but presumably it arose from the left arch. Pulsations were noted at both ends when the arch was occluded. An obliterated ductus arteriosus joined the left aortic arch and the left pulmonary artery. Division of this structure partially relieved tracheal constriction. The arch was divided and the proximal end was closed proximal to the origin of the left subclavian artery. It was thought the subclavian artery could be used in an anastomosis but on attempting to close the distal end of the arch the subclavian artery was partially constricted. Consequently the subclavian artery was sacrificed and the distal end of the arch anastomosed to the left pulmonary artery without undue tension. Division of the left aortic arch seemed to relieve all respiratory distress. After operation he showed evidence of respiratory obstruction, and it was feared that manipulation and traction on the left recurrent nerve was

responsible. A temporary tracheotomy was done, and his subsequent course has been without complications. The cyanosis has disappeared, and the difficulty in breathing has been relieved. The findings in this case are illustrated in Figures 8 and 9.

DISCUSSION

The diagnosis of a retro-esophageal artery can usually be made by roentgenography with barium swallow.[8] The anomalous vessel causes an abnormal indentation in the posterior wall of the esophagus seen in the customary oblique and lateral projections. To determine the type of vessel is not always so easy. The size and level of the indentation are important. The most
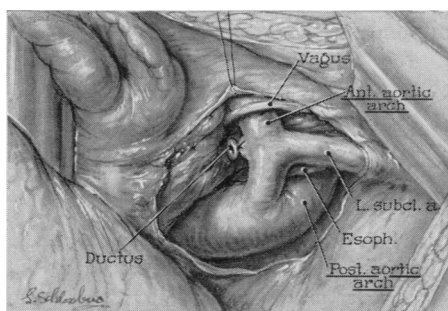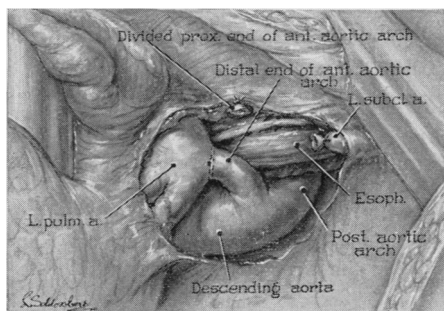

FIG. 8

FIG. 9

FIG. 8.—Left side of double aortic arch causing tracheal obstruction. Ductus arteriosus divided.

FIG. 9.—Anastomosis performed between distal end of left aortic arch as shown in Figure 8 and left pulmonary artery.

common retro-esophageal vessel has been an anomalous subclavian artery. A retro-esophageal subclavian artery arising from an aortic diverticulum may give the same roentgenographic picture as a double aortic arch since the size and location of the aortic diverticulum may be similar to the distal end of a persistent double aorta. Large collateral arteries from the aorta to the lungs in the presence of pulmonic atresia are a fairly common cause of indentation of the esophagus. Angiocardiography may demonstrate the anomalous vessel but this diagnostic aid has been of little help in this series. It has been of great value in demonstrating the side of the aortic arch in several unusual patients.

It is important to recognize the presence of a retro-esophageal subclavian artery at the time of operation for pulmonic stenosis and appreciate that it may be used in creating an artificial ductus arteriosus. In three instances early in this series of patients the anomaly was not recognized and the carotid artery with its greater operative risk was used.

In cases of double aortic arch the operative approach probably should be made on the side of the smaller arch if the two are of unequal size. If this is

done, the subclavian branch may be used for the anastomosis or the arch may be divided and one of the ends employed for this purpose. If the ring is responsible for tracheal or esophageal obstruction, it is, of course, important that the ring be divided and the obstruction relieved.

SUMMARY

Variations of a vascular ring due to anomalies of the aortic arch encountered in the surgical treatment of patients with congenital pulmonic stenosis are described. A brief description of the embryologic development of these anomalies is given. Suggestions are made in regard to the use of a retro-esophageal subclavian artery for the creation of an artificial ductus arteriosus.

Three cases of double aortic arch are described. In one of these patients the diagnosis was suspected at operation but not proved until later. In the other two patients the smaller arch was divided and one end was anastomosed to a pulmonary artery.

Marked respiratory difficulty as a result of tracheal constriction was present in only one patient in this group and was corrected by division of the smaller of the two arches.

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