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RIGHT AORTIC ARCH WITH ISOLATION OF THE LEFT SUBCLAVIAN ARTERY*

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RIGHT aortic arch with the left subclavian artery no longer attached to the aorta (isolation of the left subclavian artery) is a rare congenital malformation.[9] Knowledge concerning this anomaly has resulted primarily from study of anatomic specimens.[2,4,8] For this reason it was considered to be of interest to describe the aortographic features and routes of collateral blood supply to the left upper extremity in 2 patients with this malformation.

REPORT OF CASES

Case 1. L.D., W-36177, a 23 year old soldier, at the age of 7 was found to have a right aortic arch for which he was followed in the Cardiac Clinic. Blood pressure in the right arm was 98/60 mm. Hg, and blood pressure was unobtainable in the left arm. He had been asymptomatic with normal growth and development. At age 12 he first noticed that his left arm tired more easily, however, he participated in all sports without difficulty. At age 18, he gave a 2 month history of numbness in the left upper extremity brought on by raising his left arm above his shoulder. Exercise tolerance in the left arm was less than in the right. There were no symptoms to suggest congenital heart disease or cerebral or vertebrobasilar insufficiency. The patient was right-handed.

Admission for aortography revealed a well developed young man in no distress. The blood pressure in the right arm was 118/50 mm. Hg, raised to shoulder level 110/50 mm. Hg; left arm 80/56 mm. Hg, raised 70/60 mm. Hg; right leg 140/80 mm. Hg; left leg 138/70 mm. Hg. The left radial and antecubital pulses were markedly diminished when the extremity was dependent and disappeared when the extremity was raised above the shoulder. A feeling of numbness was present in the left upper extremity during the examination. The skin of the left arm was of normal color without ischemic changes. The diameter of the left arm measured 2 cm. less than the right arm. A continuous murmur was heard over the posterior triangle of the neck on the left side. Examination of the heart was within normal limits and no murmurs were noted. The electrocardiogram showed no abnormal findings.

Roentgenography revealed a right aortic arch (Fig. 1, *A* and *B*). The heart was of normal size and configuration and the pulmonary vasculature was within normal limits. Esophagograms in the right and left anterior oblique positions showed no abnormal defect of the esophagus (Fig. 1, *C* and *D*).

Percutaneous catheter aortography with injection of contrast material into the ascending aorta confirmed the presence of a right aortic arch. The left common carotid artery arose as the first branch and was followed by the right common carotid and right subclavian arteries. There was no opacification of the left subclavian artery from the arch. The descending aorta was on the right (Fig. 2). Figure 3*A* shows a selective injection of the left common carotid artery. Approximately 2 seconds following injection (Fig. 3, *B* and *C*), the left subclavian artery was opacified by collateral channels from the occipital branch of the left external carotid artery to branches of the thyrocervical and costocervical trunks of the left subclavian artery. In addition, there was abnormal opacification of the left vertebral artery by way of muscular branches of the occipital artery with retrograde flow down the vertebral artery, contributing blood to the left subclavian vessel. The left inferior thyroid artery of the thyrocervical trunk received blood directly from the left superior thyroid branch of the external carotid artery.

A second injection of the left common carotid artery revealed no blood reaching the left vertebral artery through the circle of Willis.

Percutaneous catheterization of the left

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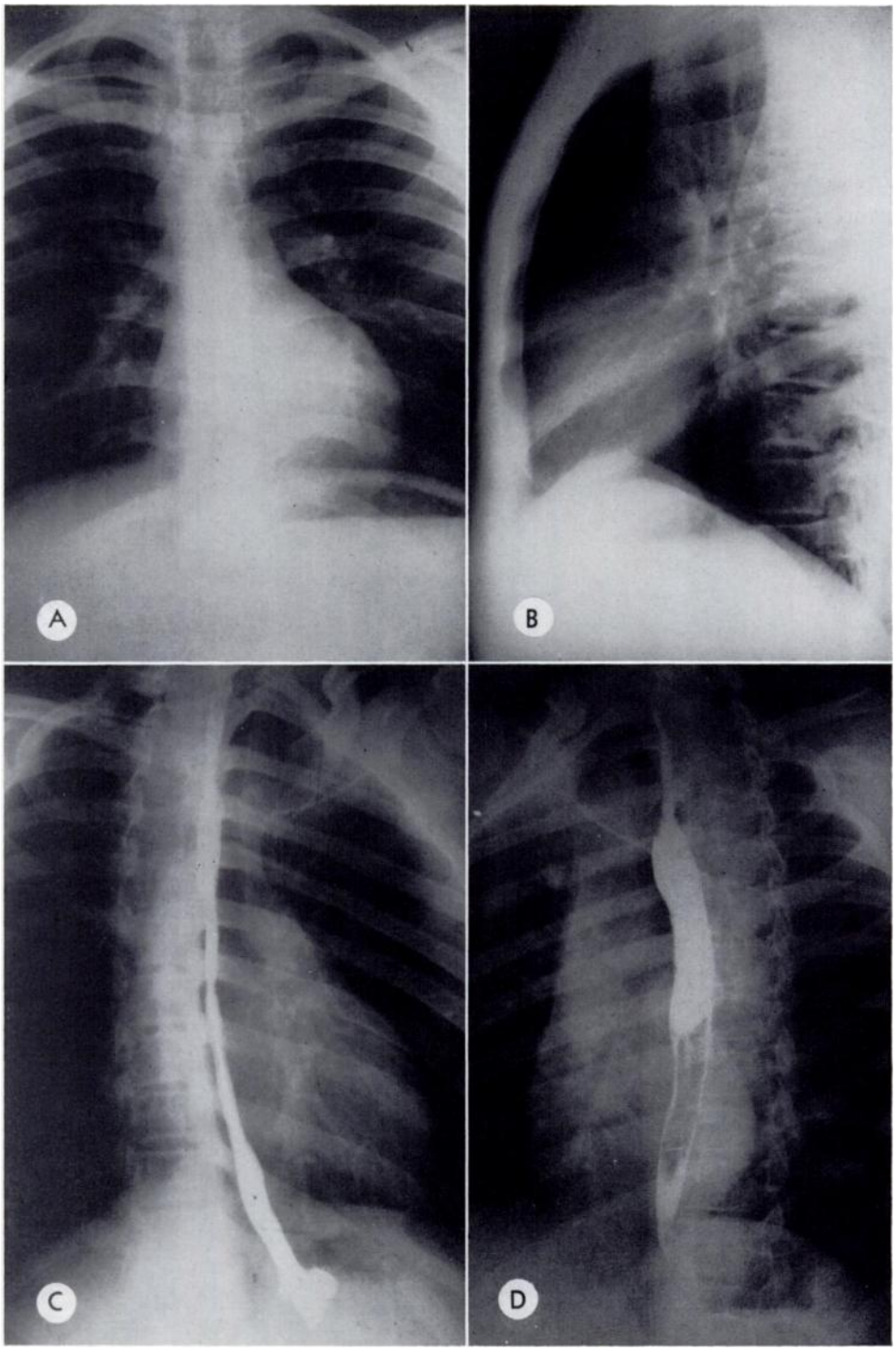


FIG. 1. Case 1. (*A* and *B*) Right aortic arch and isolation of the left subclavian artery. The heart is not enlarged. In the lateral view the trachea shows no posterior compression. (*C* and *D*) Right and left anterior oblique esophagograms. There is no defect on the posterior aspect of the esophagus.

brachial artery was then performed. Upon advancement, the catheter met resistance in the first or thoracic portion of the left subclavian artery. Hand injection of contrast material showed this segment of the left subclavian artery to end blindly with no visible connection with either the aortic arch or with the left pulmonary artery. There was filling of the vertebral, costocervical and thyrocervical trunks of the left subclavian artery (Fig. 4, *A* and *B*).

Case II. B.B., C-125926, a 34 year old man, was admitted with the diagnosis of chronic schizophrenia. Except for mental illness, the patient had been in good health without symptoms of heart disease. Apparently he experienced no difficulty with the use of his left arm.

The patient was well developed and appeared in good physical condition. The pulse rate was 104 in the right arm, and the blood pressure was 140/86 mm. Hg. Blood pressure and pulse were unobtainable in the left arm. Examination of the heart revealed no abnormal findings and no murmurs were heard. Several electrocardiograms were within normal limits.

Chest roentgenograms and a barium swallow examination showed a right aortic arch and heart of normal size and configuration. The pulmonary artery segment and peripheral pulmonary vasculature were unremarkable. There was no defect visible on the posterior esophageal wall (Fig. 5, *A* and *B*).

Because of the absent pulse in the left arm, percutaneous catheter aortography was performed. Injection of contrast material into the ascending aorta showed a right arch with filling of the left common carotid, right common carotid and right subclavian arteries, in that order. The left subclavian artery did not opacify from the aorta (Fig. 6*A*). Serial studies at 2–3 seconds after injection revealed faint opacification of a small, tortuous left subclavian artery which filled by retrograde flow down the left vertebral artery (Fig. 6, *B* and *C*). The descending aorta was on the right. On the lateral aortogram a diverticulum-like structure was present at the junction of the arch and descending aorta (Fig. 7).

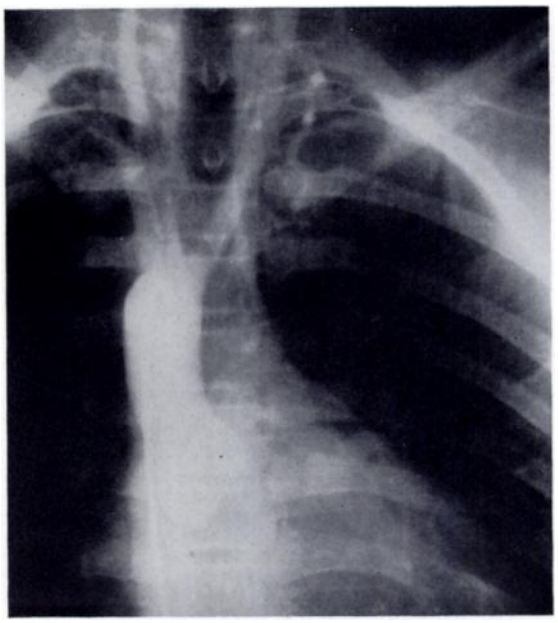
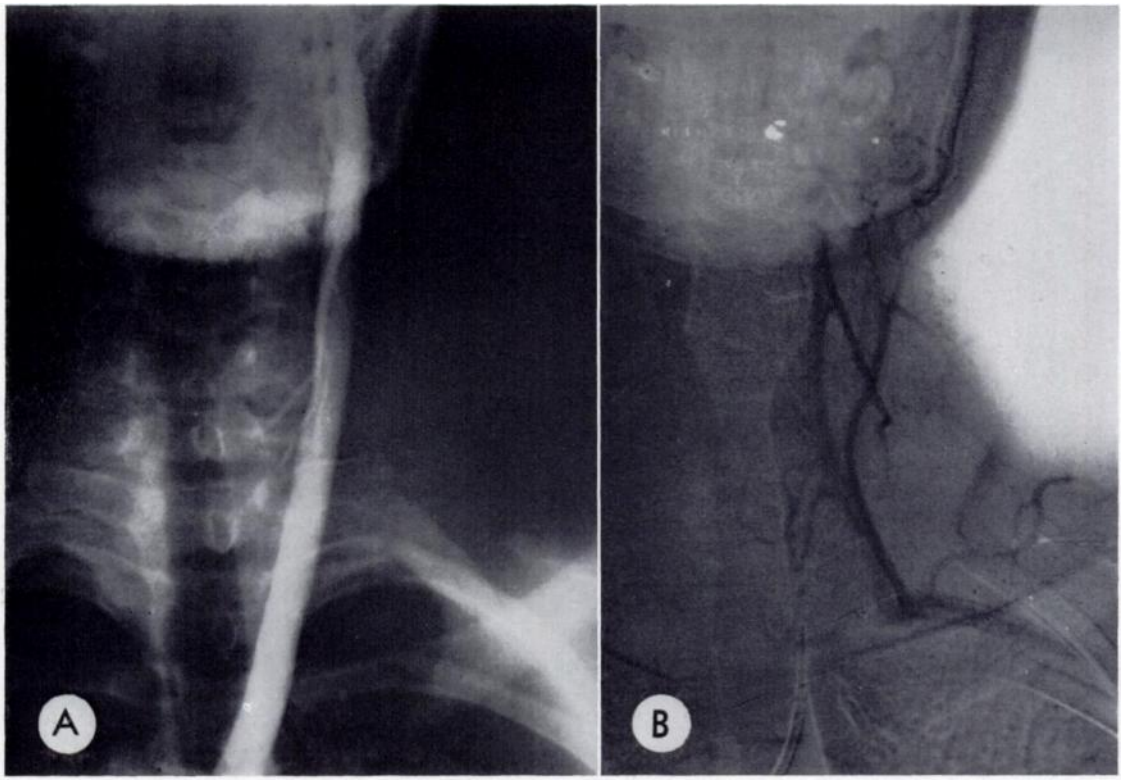


Fig. 2. Case 1. Right aortic arch and isolation of the left subclavian artery. The descending aorta is on the right. There is opacification of the left common carotid, right common carotid and right subclavian arteries only from the arch.

DISCUSSION

Right aortic arch has been classified into three major sub-groups: (1) with mirror-image branching of the major arteries; (2) with an aberrant left subclavian artery; and (3) with the left subclavian artery isolated from the aorta.[9] Right arch with isolation of the left subclavian artery is the least common of these types.[7] In this malformation the left common carotid artery arises as the first branch of the right arch and is followed by the right common carotid and right subclavian arteries. The left subclavian artery no longer has a connection with the aorta, but is connected to the left pulmonary artery by a left ductus arteriosus (Fig. 8). The right arch is anterior to the trachea and esophagus, and produces no posterior defect on the lateral esophagogram.[7]

Most reports of isolation of the left subclavian artery have been in patients with a right aortic arch and tetralogy of Fallot. Ghon's case[4] showed a ventricular septal defect and right ventricular infundibular stenosis. The right as well as the left ductus arteriosus was closed. In the patient described by Stewart, Kincaid and Edwards,[8] tetralogy of Fallot was present with a patent left ductus arteriosus. In the patient



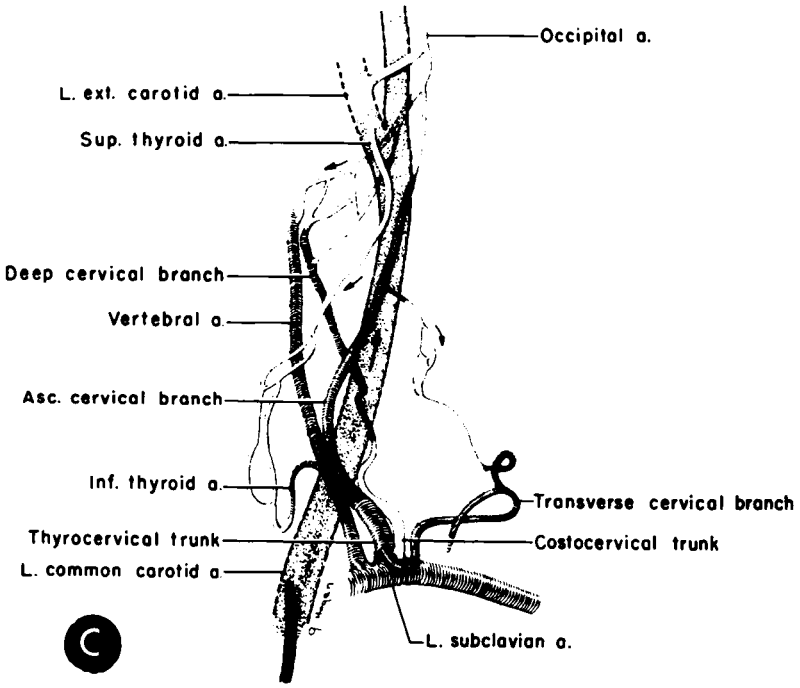


FIG. 3. Case 1. (*A*) Selective injection of the left common carotid artery. (*B*) Subtraction arteriogram 2 seconds following injection. The left external carotid artery supplies the vertebral artery, the inferior thyroid artery, the thyrocervical and costocervical trunks with retrograde flow opacifying the left subclavian artery distal to the vertebral origin. The thoracic portion of the left subclavian artery is not opacified. (*C*) Composite drawing of *A* and *B*.

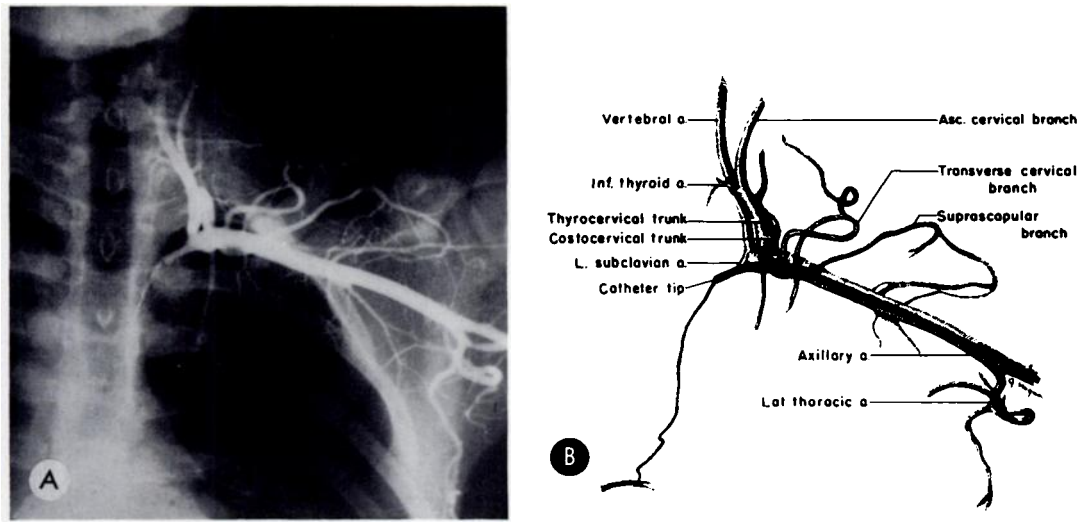


FIG. 4. Case I. (*A*) Retrograde injection of the left subclavian artery. Catheter tip is in the first portion of the left subclavian artery. (*B*) Drawing of *A*. No connection of the left subclavian artery with the aortic arch or left pulmonary artery can be seen.

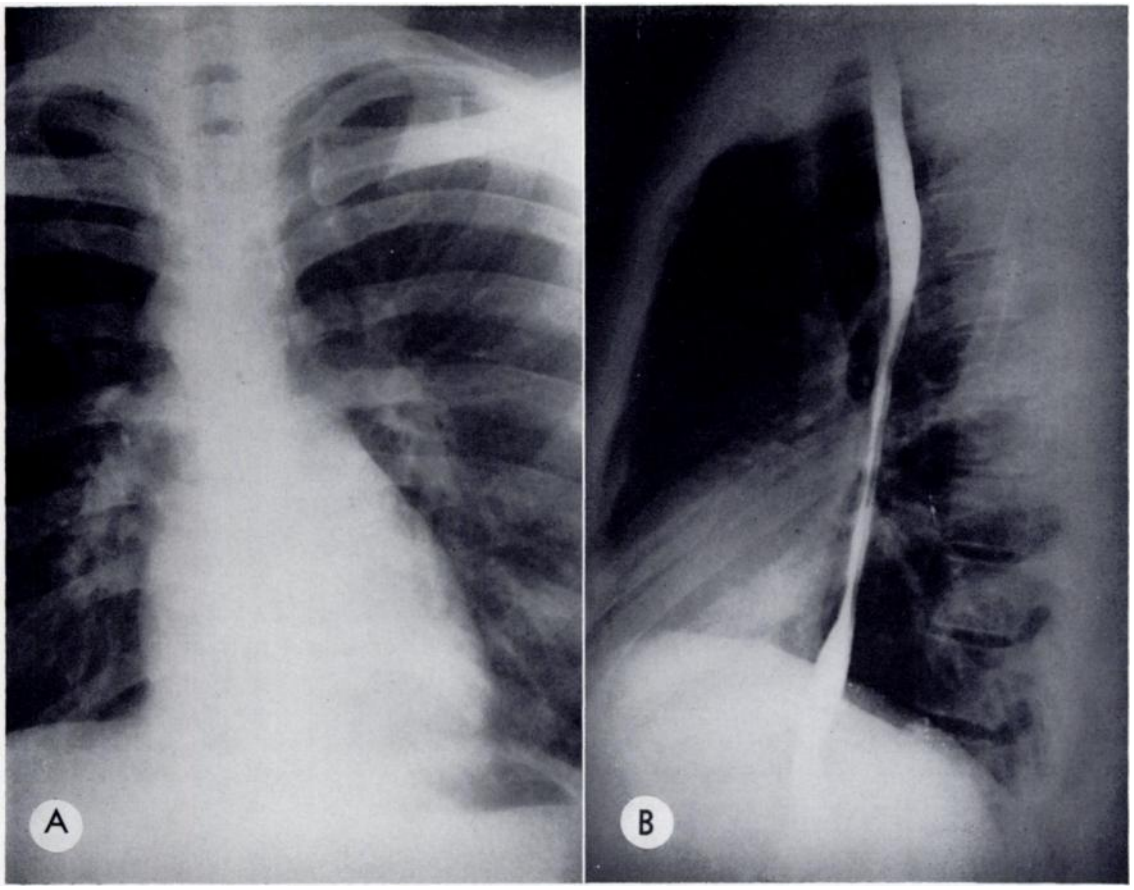


FIG. 5. Case II. (*A*) Right aortic arch with isolation of the left subclavian artery. The heart has a normal configuration with no abnormality of the pulmonary vasculature. (*B*) The barium-filled esophagus shows no retroesophageal compression.

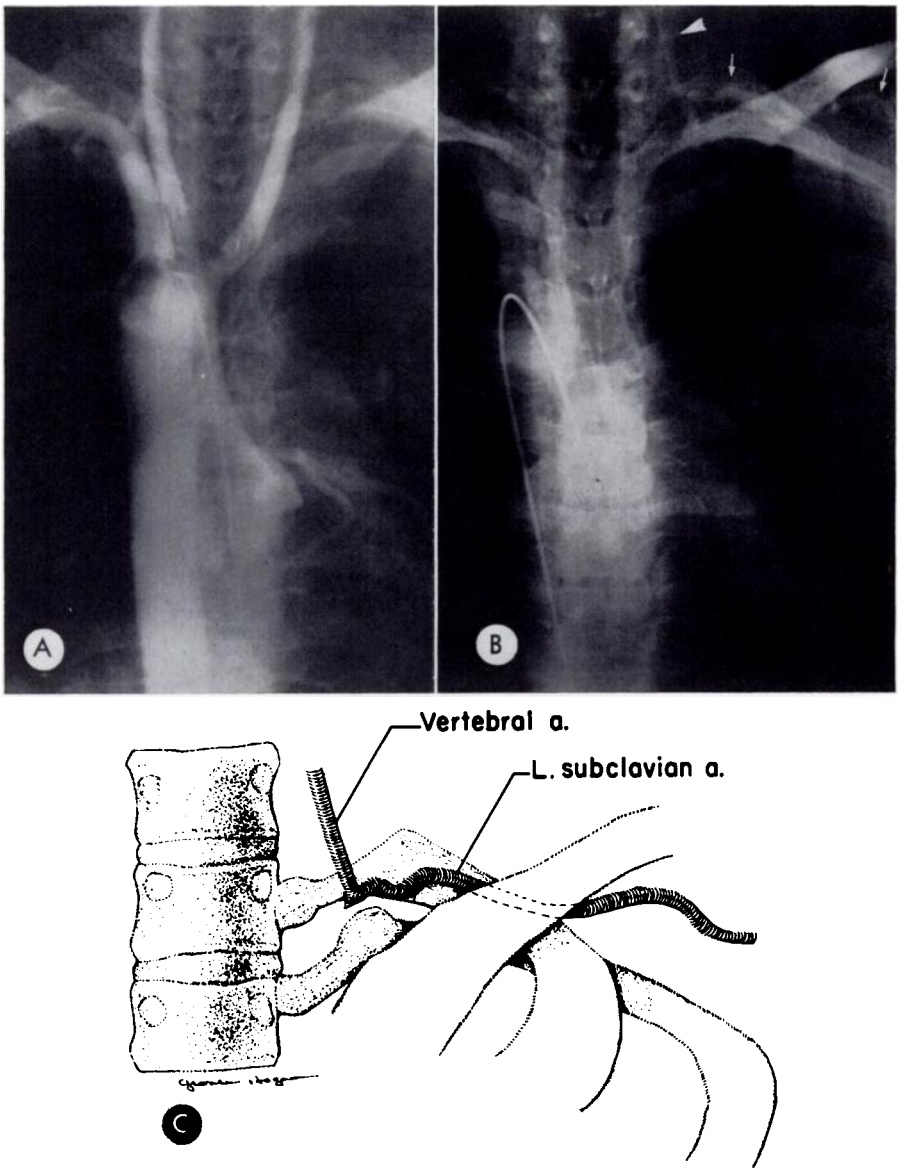


FIG. 6. Case 11. (*A*) Aortogram showing the aortic arch on the right, a right-sided descending aorta, and the left common carotid, right common carotid and right subclavian arteries arising from the arch in that order. There is no opacification of the left subclavian artery from the arch. (*B*) Two seconds after injection reversed flow in the left vertebral artery (upper arrow) opacifies the left subclavian artery distal to the vertebral origin (lower arrows). (*C*) Diagram of arteriogram in *B*.

of Barger and associates,[2] the heart showed no major malformations. Each ductus arteriosus was patent, the right being widely open. Right aortic arch with isolation of the left subclavian artery and only a right ductus has not been reported to our knowledge. Maranhao *et al.*[6] have recently described an asymptomatic patient with congenital subclavian steal syndrome associated with a right aortic arch. The aortographic findings in this case appear to be similar to the 2 patients in the present report. In all of these cases the descending aorta was on the right side.

The roentgenologic findings in these 2 cases would seem to justify the diagnosis of right arch with an isolated left subclavian artery. As, however, anatomic proof is lacking, two other anomalies of the aortic arch should be considered. One theoretical possibility is double aortic arch with atresia in the left arch between the origins of the left common carotid and the left subclavian arteries, and a second area of atresia in the left arch between the origin of the left subclavian artery and the aortic insertion of the ductus arteriosus.[3] The most distal portion of the left arch may persist as a bulbous enlargement on the descending aorta, the so-called diverticulum of Kommerell.[5] In this condition the left ductus would be expected to run from the aortic diverticulum to the left pumonary artery and not be directly connected to the left subclavian anomaly (Fig. 9). To our knowledge, no such example of this type of double arch has been reported. The other possibility is right aortic arch and aberrant left subclavian artery with atresia or stenosis of the proximal portion of the subclavian artery as illustrated in the 2 patients reported by Antia and Ottesen[1] (Fig. 10).

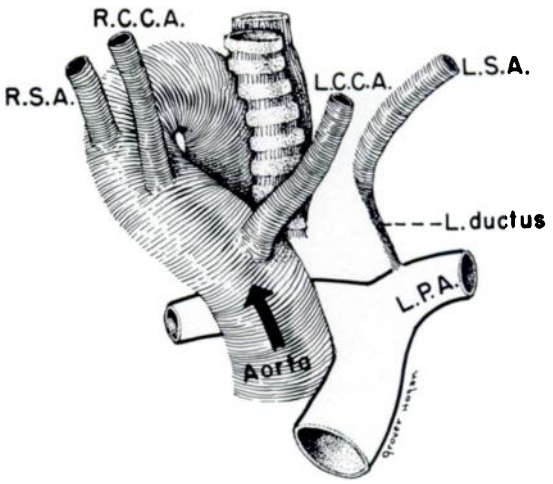


Fig. 8. Diagram of right aortic arch with the left subclavian artery isolated from the aorta. The left subclavian artery is connected to the left pulmonary artery by the left ductus arteriosus.

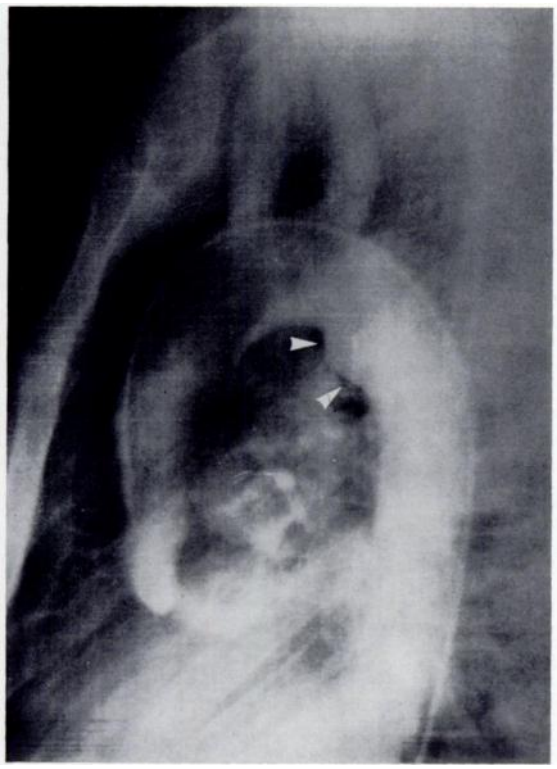


Fig. 7. Case II. Right aortic arch with isolation of the left subclavian artery. Lateral aortogram showing diverticulum-like structure (arrows) at the junction of the arch and descending aorta.

These malformations would satisfactorily explain absence of opacification of the left subclavian artery from the right arch. Also, these anomalies would be a satisfactory explanation for the aortic diverticulum in Case II. In both these conditions, a vascular ring is present resulting in a posterior constriction on the barium-filled esophagus. In our 2 patients failure to demonstrate a retroesophageal indentation on the lateral esophagogram probably excludes these anatomic possibilities. The aortic diverticulum noted in Case II most likely represents a patent rudiment of the most distal portion of the embryonic left arch, but without anatomic significance.

The arteriographic studies in Case I (Fig. 3, *A*, *B* and *C*) clearly indicate that two major sources of blood to the left subclavian artery were from the occipital branches of the left external carotid artery to muscular branches of the left vertebral artery and from the occipital branches of the left ex-

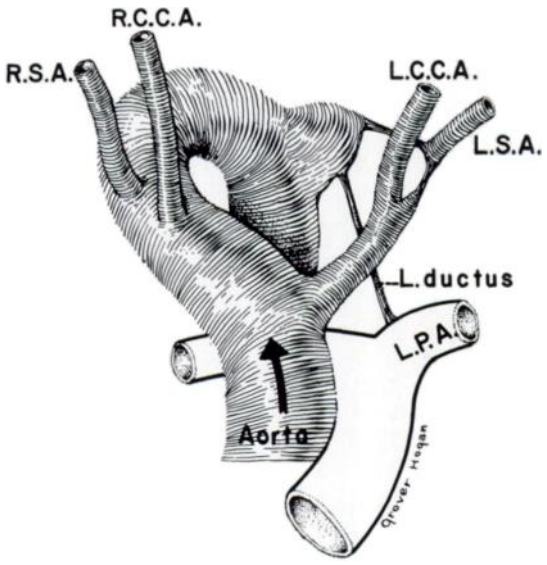


FIG. 9. Diagram of double aortic arch with atresia of portions of the left arch. A vascular ring is present.

ternal carotid artery to branches of the thyrocervical and costocervical trunks. In addition, the left superior thyroid artery contributed blood to the left thyrocervical trunk through its inferior thyroid artery. In this patient contrast material injected into the left common carotid artery revealed no opacification of the left vertebral artery through the circle of Willis. Since contrast studies of the right vertebral artery and right carotid arteries were not performed, a direct vertebrovertebral flow and/or cross-over filling by way of the circle of Willis cannot be excluded. In Case II aortography revealed the left subclavian artery to fill from the left vertebral artery but the collateral channels to the vertebrla artery were not demonstrated.

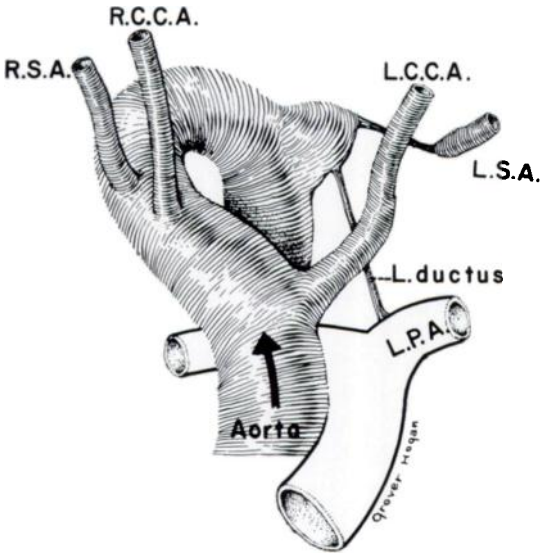


FIG. 10. Right aortic arch and aberrant left subclavian artery with partial atresia of the left subclavian artery.

There was no indication in these patients that blood reached the left subclavian artery through a left ductus arteriosus from the left pulmonary artery. Also, there was no evidence of blood flow from the left subclavian artery into the pulmonary arterial system. Retrograde injection of contrast material into the left subclavian artery in Case I (Fig. 4, *A* and *B*) showed no connection between the left subclavian artery and the left pulmonary artery. Presumably, the ductus arteriosus in these patients was closed.

SUMMARY

Two patients with a right aortic arch and isolation of the left subclavian artery are presented. The clinical findings resulted from relative ischemia of the left arm. There were no features to suggest cerebrovascular insufficiency or congenital heart disease. The esophagograms showed no posterior esophageal compression. In one patient the distal left subclavian artery was opacified by collateral channels from the left external carotid artery through the vertebal artery and branches of the thyrocervical and costocervical trunks. In the other, aortography demonstrated retrograde vertebral flow filling the distal left subclavian artery.

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