Isolation of a Subclavian Artery

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Eight patients with isolation of a subclavian artery are described. Isolation of subclavian artery is defined as a loss of continuity between the subclavian artery and aorta but with persistent connection to the homolateral pulmonary artery through a ductus arteriosus. The connecting ductus arteriosus may or may not be patent. As the pulmonary vascular resistance is usually lower than the systemic resistance, the isolated subclavian artery is perfused by subclavian steal, and if the connecting ductus arteriosus is patent, flow occurs from the subclavian artery into the pulmonary artery. In this material and in all reported cases, isolation of subclavian artery has always been associated with other anomalies of the aortic arch, such as right aortic arch or bilateral ductus. Its frequent association with intracardiac defects, usually the tetralogy of Fallot, makes its recognition clinically important. Thoracic aortography with delayed filming is the only procedure that allows consistent diagnosis of this rare anomaly during life.

Isolation of a subclavian artery is an uncommon anomaly of the aortic arch system in which one subclavian artery loses its connection with the aorta and arises from the homolateral pulmonary artery by way of a ductus arteriosus. The term *isolation* was introduced in 1964 by Stewart, Kincaid, and Edwards [1] and has been adopted by others, including Victorica and associates [2] and Shuford and associates [3]. The condition, although not so named, was first described by Ghon [4] in 1908 as involving the left subclavian artery. Additional examples were reported by Becu and associates [5] in 1955 and by Barger and associates [6] in 1956. To our knowledge, the first report of isolation of the right subclavian artery was that of Barger and associates [7] in 1954.

When a subclavian artery is isolated, as defined, it is on the side contralateral to the position of the aortic arch. Radiologic and hemodynamic manifestations of subclavian artery isolation depend on the state of the associated ductus arteriosus and to some extent on associated intracardiac anomalies. It is often associated with the tetralogy of Fallot. Under those circumstances, clinical recognition is imperative, since a Blalock-Taussig shunt is usually carried out on the side opposite the aortic arch and an isolated subclavian artery cannot be used for this purpose. Eight cases of isolation are reported with emphasis on the pathologic findings and radiologic correlations.

Case Reports

Case 1

A 5-year old boy was diagnosed as having tetralogy of Fallot with a right aortic arch, scimitar syndrome, and sequestration of the right lower lobe (figs. 1A and 1B). By palpation, both radial pulses were described as identical. After the aorta was opacified during right ventriculography, isolation of the left subclavian artery was identified (figs. 1C and 1D). Reexamination of the radial pulses by palpation did not reveal any differences. Complete

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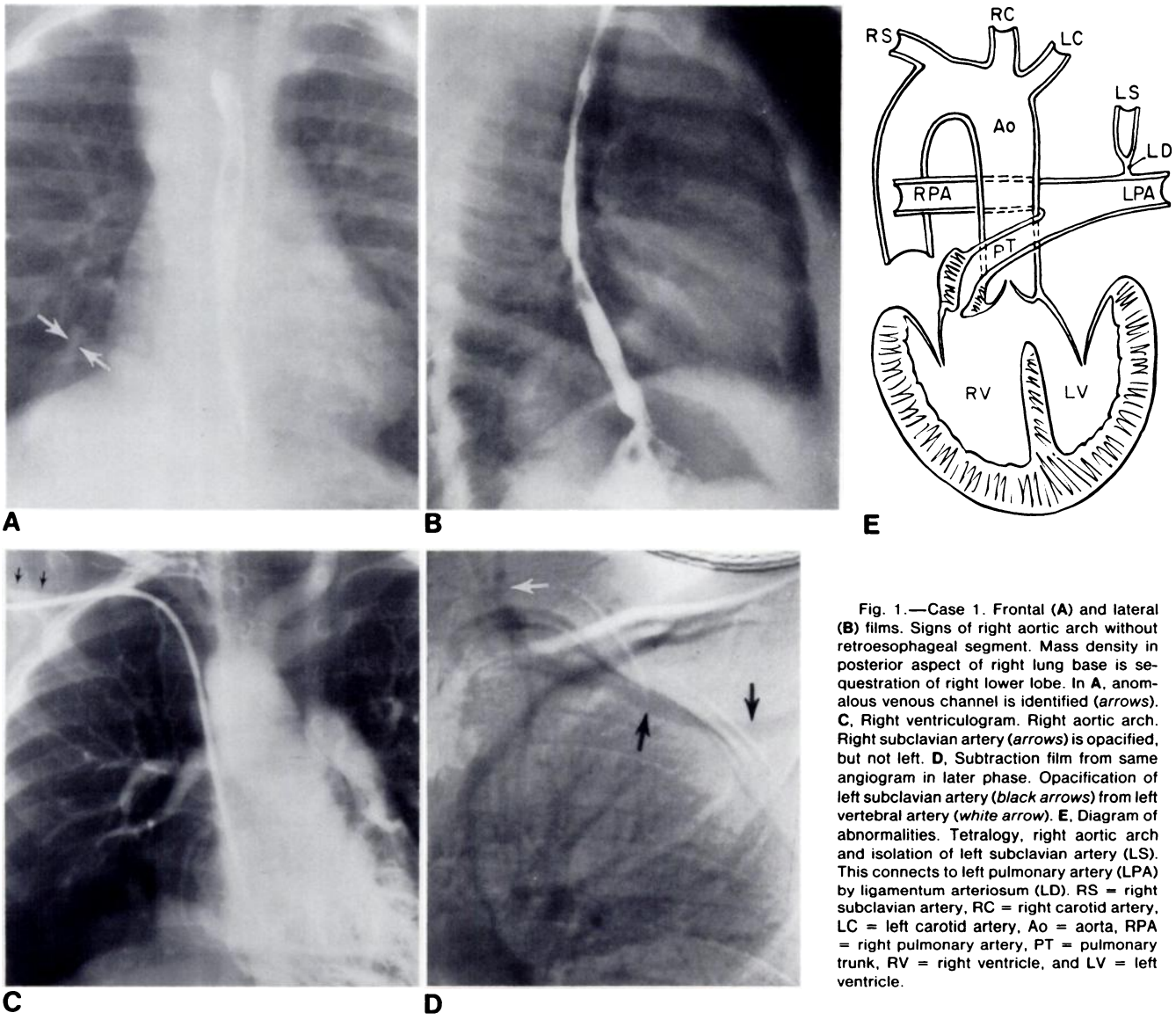

Fig. 1.—Case 1. Frontal (**A**) and lateral (**B**) films. Signs of right aortic arch without retroesophageal segment. Mass density in posterior aspect of right lung base is sequestration of right lower lobe. In **A**, anomalous venous channel is identified (*arrows*). **C**, Right ventriculogram. Right aortic arch. Right subclavian artery (*arrows*) is opacified, but not left. **D**, Subtraction film from same angiogram in later phase. Opacification of left subclavian artery (*black arrows*) from left vertebral artery (*white arrow*). **E**, Diagram of abnormalities. Tetralogy, right aortic arch and isolation of left subclavian artery (LS). This connects to left pulmonary artery (LPA) by ligamentum arteriosum (LD). RS = right subclavian artery, RC = right carotid artery, LC = left carotid artery, Ao = aorta, RPA = right pulmonary artery, PT = pulmonary trunk, RV = right ventricle, and LV = left ventricle.

repair of the tetralogy of Fallot was performed. At operation, isolation of the left subclavian artery was confirmed (fig. 1E). The child was well 2 years postoperatively.

Case 2

In this girl, the tetralogy of Fallot was diagnosed at age 4 months. Cardiac catheterization was performed at age 2 years when increasing cyanosis was evident. A right ventriculogram confirmed the clinical diagnosis of tetralogy of Fallot and right aortic arch. An aortogram was not performed. A left Blalock-Taussig shunt was planned. At the time of operation, isolation of the left subclavian artery was recognized. The radial pulses had been described as identical by clinical examination. A subsequent aortogram confirmed isolation of the left subclavian artery which opacified through the left vertebral artery ("subclavian steal").

At age 5 years, the patient died 2 days after intracardiac repair of the tetralogy of Fallot. The autopsy revealed tetralogy of Fallot with right aortic arch. The left subclavian artery was connected to the left pulmonary artery by an atretic ductus arteriosus. As in case 1 (fig. 1E), there was no right ductus arteriosus.

Case 3

Our primary observations in this case were through referral of the specimen of the thoracic organs obtained at autopsy. Clinical data from another institution were as follows.

When seen at age 2 months, this girl was studied for cyanotic congenital heart disease. No appreciable difference in radial pulses was detected by clinical examination. The barium-filled esophagus was unremarkable except for evidence of a right aortic arch. A diagnosis of single ventricle and mitral atresia was made from a ventriculogram. Isolation of the left subclavian artery had not been recognized. Aortography had not been performed. The child died during an attempt to band the pulmonary artery.

Autopsy revealed that there were two ventricles with malposition of the great vessels, both originating from the right ventricle (fig. 2A). A supracristal ventricular septal defect was present. There was

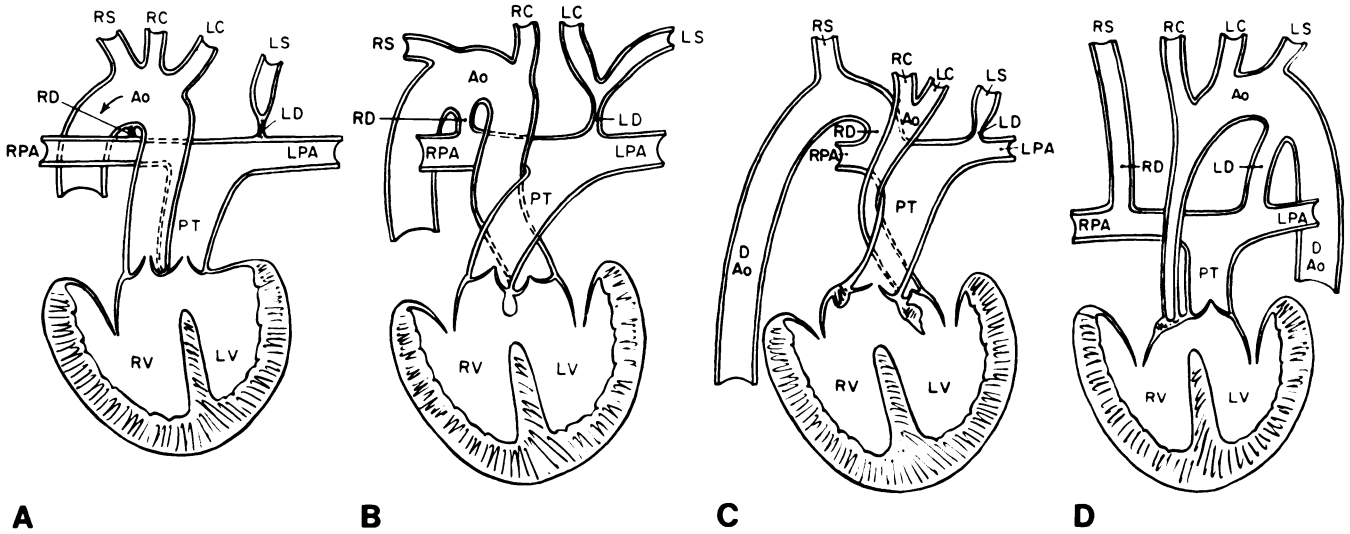

Fig. 2.—**A**, Case 3. Right aortic arch with right ductus and isolation of left subclavian artery (LS). Intracardiac malformations include mitral atresia, ventricular septal defect, and double outlet right ventricle (RV). **B**, Case 4. Isolation of left innominate artery from which arise left common carotid (LC) and left subclavian (LS) arteries. Right aortic arch, right ductus (RD), and left ductus (LD). The latter connects isolated left innominate artery to left pulmonary artery (LPA). Associated intracardiac malformation was ventricular septal defect. **C**, Case 5. Interruption of right aortic arch with patent right ductus (RD) and right-sided descending aorta (D Ao). Isolated left subclavian artery. Intracardiac malformations include ventricular septal defect, subaortic stenosis, and biventricular origin of pulmonary trunk (PT). **D**, Case 6. Left aortic arch with patent left ductus (LD). Isolation of right subclavian artery (RS) as it connects by way of patent right ductus (RD) with right pulmonary artery (RPA). Other malformations include malposition of great vessels with ventricular septal defect and aortic atresia. RC = right carotid artery, LV = left ventricle.

no pulmonary stenosis. The mitral valve was atretic. The aortic arch was on the right, and the left subclavian artery was isolated, being connected to the left pulmonary artery by the left ligamentum arteriosum. A right ligamentum arteriosum was also present. Reexamination of the ventriculograms revealed delayed filling of the left subclavian artery through the left vertebral artery.

Case 4

A newborn boy with Goldenhar syndrome [8] died during the second day of life after repair of a tracheoesophageal fistula. The infant was in congestive heart failure, and the clinical diagnosis had been ventricular septal defect. No angiographic procedures were performed.

At autopsy, an infracristal ventricular septal defect was found. The aortic arch was on the right side, and the left innominate artery was isolated from the aortic arch. The left ductus arteriosus leading to the isolated innominate artery was patent but stenotic. A widely patent right ductus arteriosus was also present (fig. 2B).

Case 5

A newborn girl showed central cyanosis and signs of congestive heart failure and died in the newborn period. The clinical diagnosis was interruption of the aortic arch. No angiograms were obtained.

At autopsy, a ventricular septal defect was present associated with biventricular origin of the pulmonary trunk and subaortic stenosis. The aortic arch was interrupted. A patent right ductus arteriosus led to the descending aorta, the latter lying to the right of the esophagus. The aortic arch terminated after giving rise to the two common carotid arteries. The left subclavian artery was isolated, being connected to the left pulmonary artery by a patent left ductus arteriosus, while the right subclavian artery arose from the upper descending aorta (fig. 2C).

Case 6

A newborn cyanotic boy was operated on for repair of a meningoencephalocele. Associated cardiac abnormalities were diagnosed as part of the polysplenic syndrome. Thoracic radiographs with barium swallow did not reveal any abnormal impressions on the esophagus. The aortic arch was on the left side. The child died before planned cardiac catheterization.

At autopsy, dextrocardia was present. The aorta originated from the right ventricle, and the aortic valve was atretic. The pulmonary artery exhibited biventricular origin. The aortic arch was on the left and received a patent left ductus arteriosus. The right subclavian artery was isolated, communicating with the right pulmonary artery through a patent right ductus arteriosus (fig. 2D).

Case 7

A 1½-year-old girl was referred because of congestive heart failure and increasing cyanosis of the right arm. The infant had respiratory problems soon after birth and the mother noticed that the right arm, but no other part of the body, was slightly blue. Both radial pulses were equal and blood pressures, measured by the cuff method, were identical in each extremity. At age 1 month there were no audible murmurs and chest radiographs appeared normal. The child was lost to follow-up until age 1½ years. At that time cyanosis was still evident in the right arm, more apparent on crying. Both radial pulses and upper extremity blood pressures were identical. She was in congestive heart failure and a typical murmur of patent ductus arteriosus was audible along the left sternal border. Chest radiographs revealed cardiomegaly, shunt vascularity, a left aortic arch and no abnormal impressions on the esophagus. Electrocardiography revealed right ventricular hypertrophy and right atrial enlargement. Blood gases were measured from each radial artery; Po_2 was 51% in the left arm and 40% in the right arm. Cardiac catheterization demonstrated systemic pressure in the right

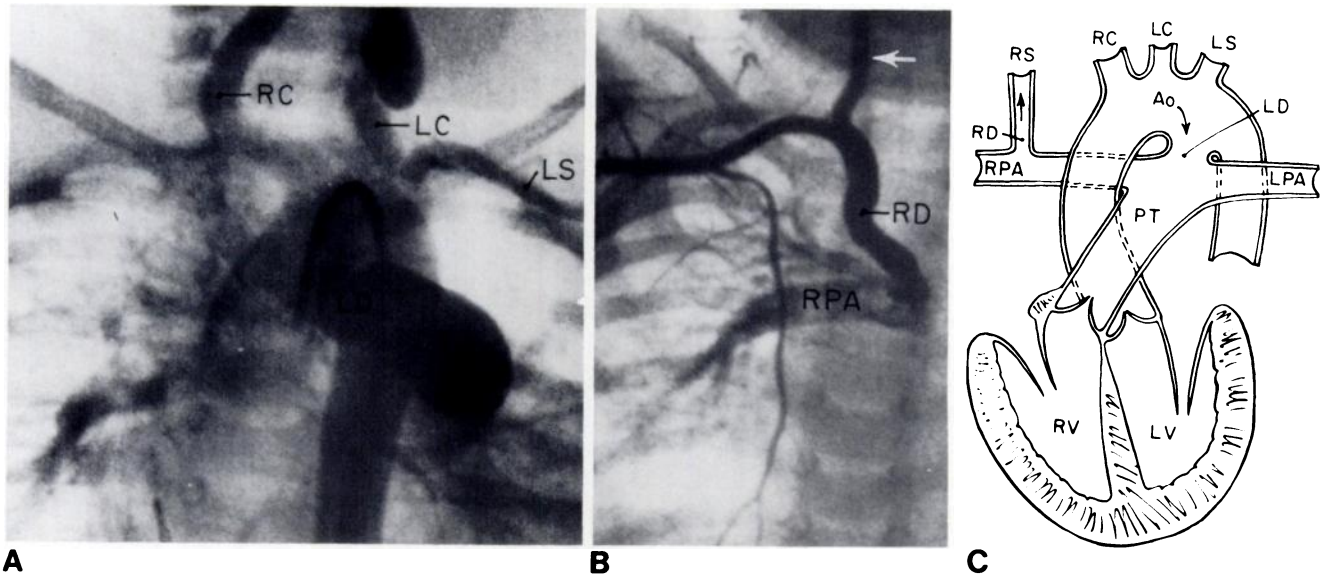

Fig. 3.—Case 7. **A**, Frontal view of thoracic aortogram. Patent left ductus arteriosus (LD) and opacification of pulmonary arterial system. Right carotid (RC), left carotid (LC), and left subclavian (LS) arteries arise from left aortic arch. Right subclavian artery not opacified. **B**, Flush right brachial arteriogram, frontal view. Opacification of right pulmonary artery (RPA) by way of patent right ductus (RD) connecting with isolated right subclavian artery. Wide right vertebral artery (*arrow*). **C**, Essentials of malformations. Left aortic arch and patent left ductus arteriosus (LD). Patent right ductus (RD) runs between right pulmonary artery (RPA) and right subclavian artery (RS). No intracardiac malformations. Left-to-right shunt occurred through left ductus arteriosus and right-to-left shunt through right ductus arteriosus. LPA = left pulmonary artery, PT = pulmonary trunk, RV = right ventricle, LV = left ventricle.

ventricle and pulmonary artery with elevated pulmonary vascular resistance. A thoracic aortogram demonstrated a patent left ductus arteriosus shunting left to right. The right carotid artery was the first branch of the aortic arch and the right subclavian artery was not visualized (fig. 3A). There was no delayed opacification of the right subclavian artery. A pulmonary angiogram demonstrated transient opacification of a systemic artery from the right pulmonary artery. A flush right brachial angiogram opacified the right subclavian artery connected to the right pulmonary artery by a right ductus arteriosus (fig. 3B). There was slight reduction in the pulmonary vascular resistance following administration of Priscoline. Both ductus arteriosus were surgically divided. Following the operation, the right radial pulse was markedly diminished in amplitude, but the cyanosis disappeared. The patient continued to do poorly and a repeat cardiac catheterization 9 months after surgery revealed persistence of systemic pressure in the pulmonary circulation and further elevation of pulmonary vascular resistance. The patient did not improve on medical management and died 1½ years after the surgery. At autopsy stumps of ductus arteriosus were seen bilaterally. The right subclavian artery did not originate from the aortic arch. There was severe right ventricular hypertrophy. Left heart chambers were normal. Histologic examination of the lungs revealed signs of grade IV pulmonary vascular disease. A summary of the vascular connections is shown in figure 3C.

Case 8

A full term male neonate was referred for evaluation of respiratory distress and cyanosis. There was no record of asymmetry in radial pulses. A clinical diagnosis of complete transposition of great vessels with a ventricular septal defect was made and cardiac catheterization was performed. Systemic pressures were found in both ventricles. Pulmonary artery was not entered. Right and left ventriculograms were obtained. Right ventriculogram demonstrated the anatomy of a complete transposition of the great vessels. The ventricular septum appeared intact. A left aortic arch was found along with an apparently closing left ductus arteriosus. Four months later the patient developed symptoms and signs of left ventricular outflow tract obstruction and a right Blalock-Taussig shunt was recommended. At that time the right radial pulse was found to be markedly diminished. Review of the angiograms demonstrated isolation of the right subclavian artery which was opacified by subclavian steal (figs. 4A–4C). A right ductus diverticulum was identified on the right pulmonary artery (fig. 4D). The aortic arch anatomy was interpreted as isolation of the right subclavian artery, bilateral ductus arteriosus, and left aortic arch (fig. 4E). A central shunt was created between the ascending aorta and the main pulmonary artery using a Gor-Tex graft.

Discussion

Isolation of a subclavian artery is an uncommon malformation of the aortic arch system. Its true incidence is unknown, probably because some cases are not recognized during life, as it may not cause symptoms.

There is no uniformity of opinion as to what constitutes this lesion. Several reports [9–11] describe patients with a congenital subclavian steal syndrome, in whom an atretic segment of subclavian artery is connected to the aortic arch, as examples of subclavian isolation. Other reports of congenital subclavian steal syndrome do not contain a pathologic description [3, 12, 13]. In addition, some of the patients described had supravalvular aortic stenosis [13]. In those patients, the subclavian steal syndrome could conceivably be due to obstruction of a subclavian artery as it arises normally from the aorta. We believe that the term *isolation* should be confined to a subclavian artery not connecting to the aorta but rather to the homolateral pul-

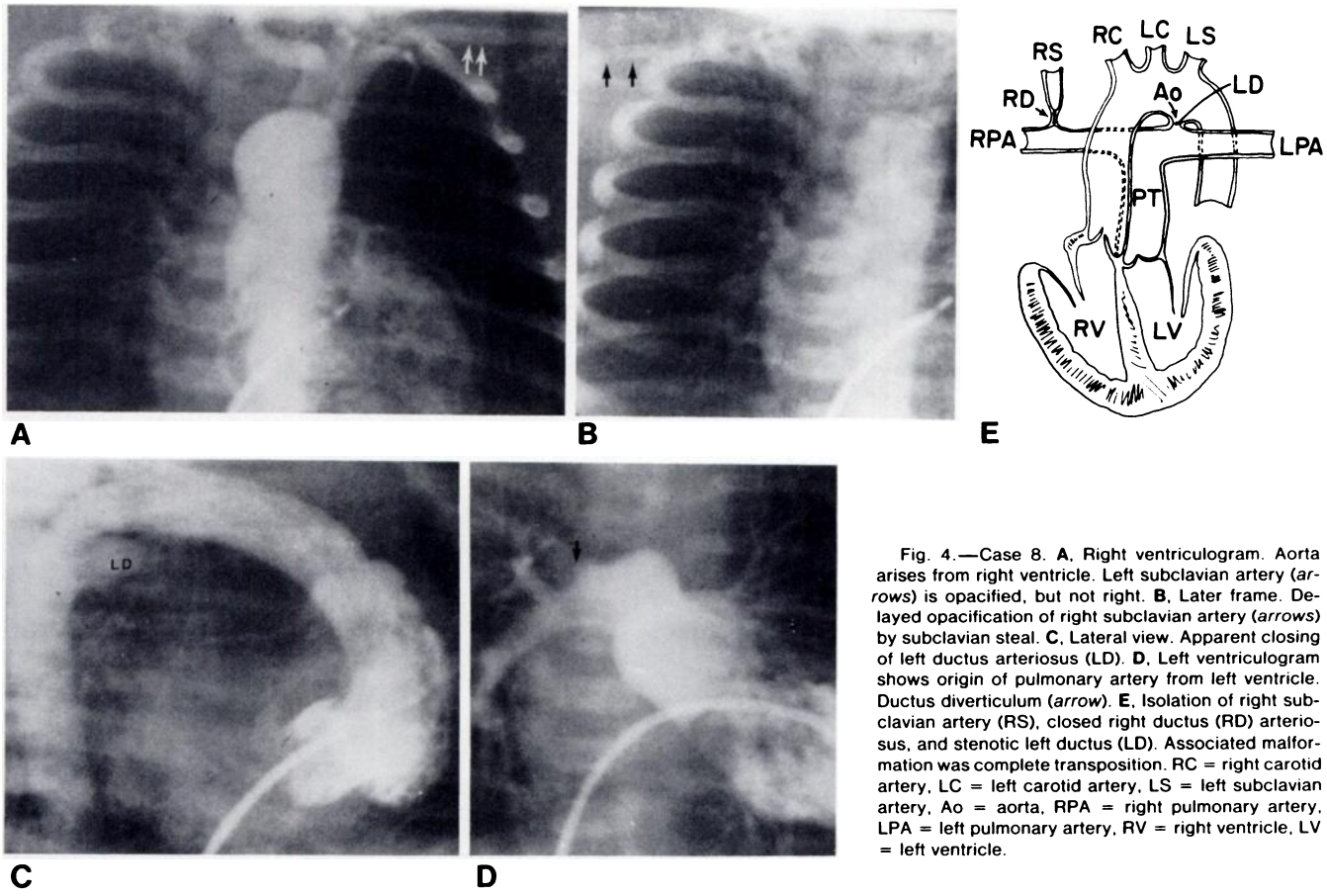

Fig. 4.—Case 8. **A**, Right ventriculogram. Aorta arises from right ventricle. Left subclavian artery (*arrows*) is opacified, but not right. **B**, Later frame. Delayed opacification of right subclavian artery (*arrows*) by subclavian steal. **C**, Lateral view. Apparent closing of left ductus arteriosus (LD). **D**, Left ventriculogram shows origin of pulmonary artery from left ventricle. Ductus diverticulum (*arrow*). **E**, Isolation of right subclavian artery (RS), closed right ductus (RD) arteriosus, and stenotic left ductus (LD). Associated malformation was complete transposition. RC = right carotid artery, LC = left carotid artery, LS = left subclavian artery, Ao = aorta, RPA = right pulmonary artery, LPA = left pulmonary artery, RV = right ventricle, LV = left ventricle.

monary artery by a ductus arteriosus, whether closed or patent.

In this lesion, the isolated artery is supplied by collaterals originating principally in the contralateral subclavian artery. Among the supplying branches are the vertebral arteries. While this arrangement sets the potential for the cerebral symptoms of the subclavian steal syndrome, such symptoms are, at most, uncommon as judged by reported cases and our material. There are reports claiming identification of the cerebral symptoms of the subclavian steal syndrome in instances of subclavian isolation. In none of these has there been autopsy confirmation of the anatomic state. Some or all of these cases may not be examples of isolation as defined in this paper.

Nevertheless, the potential for symptoms exists depending, in part, on whether the ductus leading to the isolated artery is patent or closed. If patent the pulmonary arterial pressure is also a factor.

In the case of the 3-month-old patient without pulmonary stenosis reported by Sunderland and associates [14], a left-to-right shunt was demonstrated from the isolated subclavian artery. Such a shunt would accentuate the degree of steal, yet no cerebral symptoms were apparent.

Our case 7 is particularly unusual in that a right-to-left shunt occurred through the ductus into an isolated right subclavian artery because of pulmonary hypertension. There was no angiographic evidence of subclavian steal. A widely patent left ductus through which a left-to-right shunt occurred was present. To the best of our knowledge this is the only case in which right-to-left shunt into the isolated subclavian artery through the connecting ductus arteriosus was documented. The apparent paradox of a left-to-right shunt through the left ductus and a right-to-left shunt through the other may be explained as follows. In the postnatal period, normally there would be a fall in the pulmonary vascular resistance and pressure setting up a potential for perfusion of the isolated subclavian artery by way of collaterals. Consequent to the presence of the large left ductus arteriosus the pulmonary artery pressure in this patient must have been always systemic and this systemic pressure was transmitted to the right ductus and the right subclavian artery, thus precluding the development of collaterals. Once the pulmonary vascular resistance decreased to some degree, left-to-right shunt occurred through the left ductus arteriosus, but the systemic pressures remained in the pulmonary arteries, right ductus arteriosus, and the right subclavian artery. Probably the constant presence of the systemic pressure in the pulmonary arterial tree prevented collateral formation to the right subclavian artery. Similar physiologic findings were reported by Garcia et al. [15] and Tikoff and Bloom [16]. In previously reported cases and in ours the pressure in the pulmonary artery has been normal or, if elevated, less than systemic even in the presence of collateral flow to the isolated artery.

Several patients were investigated because of a difference in amplitude of radial pulses in the upper extremities [2, 12] and Victorica and associates [2] emphasized that this clinical feature, when associated with a right aortic arch, should strongly suggest the diagnosis of isolated subclavian artery. In our patients, palpation of radial pulses showed no recognizable difference in amplitude except in case 8. Because inequality of the pulses may not be present, Rodriguez and associates [17] suggested that isolation should be excluded by aortography in every patient with tetralogy of Fallot in whom a Blalock-Taussig anastomosis is contemplated.

It is not possible to diagnose subclavian isolation from radiographs of the chest with barium swallow because the isolated artery and its connecting ductus arteriosus or ligamentum arteriosum course anterior to the esophagus; hence, there will be no abnormal impression on the esophageal barium column in contrast to an aberrant subclavian artery which would pass behind the esophagus.

Aortography remains the investigation of choice in the identification of an isolated subclavian artery. Delayed films should be obtained to visualize opacification of the isolated subclavian artery through collaterals. Opacification of the isolated subclavian artery occurs almost invariably through the vertebral artery. If the ductus arteriosus is patent, the pulmonary arterial system may also be opacified, as the pulmonary vascular resistance is normally lower than the systemic vascular resistance [14, 18]. Opacification of an isolated subclavian artery during pulmonary angiography is rare but it may occur due to an elevated pulmonary vascular resistance and pressure secondary to associated lesions as in our case 7.

With subclavian isolation, other anomalies of the aortic arch system are almost always present. A right aortic arch is very common and consequently the left subclavian artery is more often involved [2, 17]. The frequency of bilateral ductus arteriosus in our series (six of eight patients) is striking. This high incidence was not mentioned in earlier reports on isolation but this condition was identified in the first known report of subclavian isolation reported by Ghon [4].

The association of intracardiac malformations is very common and was present in seven of eight patients in our series. Tetralogy of Fallot is the most commonly associated condition [2, 17, 19]. This association is clinically more significant, particularly when a Blalock-Taussig shunt is considered, because this anastomosis is almost always created on the opposite side of the aortic arch.

Subclavian isolation as an isolated anomaly is extremely unusual. We are not aware of any well documented case according to our definition. The embryology of subclavian isolation has been described [20] and is beyond the scope of this communication.

Although we define isolation as a subclavian artery which has lost its attachment to the aortic arch and retains its connection to the homolateral pulmonary artery through a ductus arteriosus, identical collateral flow into the involved subclavian artery may result when the proximal subclavian artery is atretic and connected to the aorta. Examples of the latter state include supravalvular aortic stenosis [13], atresia of the proximal subclavian or innominate artery [3, 9, 12], or in hypercalcemic syndromes [10].

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