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CONGENITAL SUBCLAVIAN STEAL*

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WHILE Contorni in 1960 was the first to demonstrate by angiography retrograde collateral flow in the vertebral artery of man, it was Reivich *et al.*,[12] in 1961, who showed the relationship of this phenomenon to symptoms of cerebral ischemia. They showed that a block of the proximal portion of a subclavian artery resulted in blood being stolen from the cerebral circulation to supply the affected upper limb. In such cases, some of the blood delivered to the brain through the internal carotid arteries and the contralateral vertebral artery flowed through the Circle of Willis and down the vertebral artery to the patent distal portion of the subclavian artery beyond the obstruction. This type of collateral circulation is termed a subclavian steal. When it results in symptoms of cerebral ischemia it is called the subclavian steal syndrome.

The subclavian steal syndrome occurs most often in adults who have atherosclerotic obstruction of the proximal subclavian artery. In children, subclavian steal is infrequent, usually asymptomatic, and is caused by a congenital anomaly of the thoracic aorta. We know of reports of 29 cases of congenital subclavian steal demonstrated by angiography.[9,11,17] Our experience with an additional 5 cases forms the basis of this communication.

CASE MATERIAL

Since 1963 congenital subclavian steal has been demonstrated in 7 patients by angiography at The Johns Hopkins Hospital. Three of these patients were reported previously.[1,11] One of these cases[11] and the 4 other cases are presented in this article.

REPORT OF CASES

CASE I. P.M. (JHH# 142 98 60) was a healthy 4 year old girl referred for evaluation because of a murmur noted on routine physical examination. Blood pressure was 110/94 mm. Hg in the right arm and 130/68 in the left arm. Neither pulse nor blood pressure was detected in the lower limbs. She had a Grade 3/6 continuous murmur best heard beneath the left scapula. It was also present in the left second intercostal space and radiated to the neck. The electrocardiogram (ECG) was normal. A cardiac series showed left ventricular enlargement and a posterior indentation in the barium filled esophagus that ran obliquely cephalad from left to right. At cardiac catheterization left ventricular pressure was 134/9 mm. Hg and the femoral artery pressure was significantly lower (106/74). Left ventricular angiography (Fig. 1, *A–C*) showed coarctation of the aorta and an aberrant right subclavian artery that arose distal to the coarctation. The aberrant right subclavian artery filled from the vertebral artery and other collateral vessels.

CASE II. B.A. (JHH# 90 76 44), an 18 year old girl, was first seen in 1954 following discovery of a murmur. Since then she has had no symptoms except occasional cold feet. Blood pressure was 150/110 mm. Hg in the left arm and 120/105 in the right arm. Femoral pulses were barely palpable and the blood pressure was unobtainable in the lower limbs. A thrill was palpable over the right carotid artery. The first heart sound was decreased and an early ejection sound was present. She had an apical Grade 2/4 blowing pansystolic murmur that radiated into the axilla. The chest roentgenogram showed a normal heart size with a right aortic arch and a ring like calcification in the most proximal portion of the descending thoracic aorta. Angiography showed a high right aortic arch with a long coarctation and a

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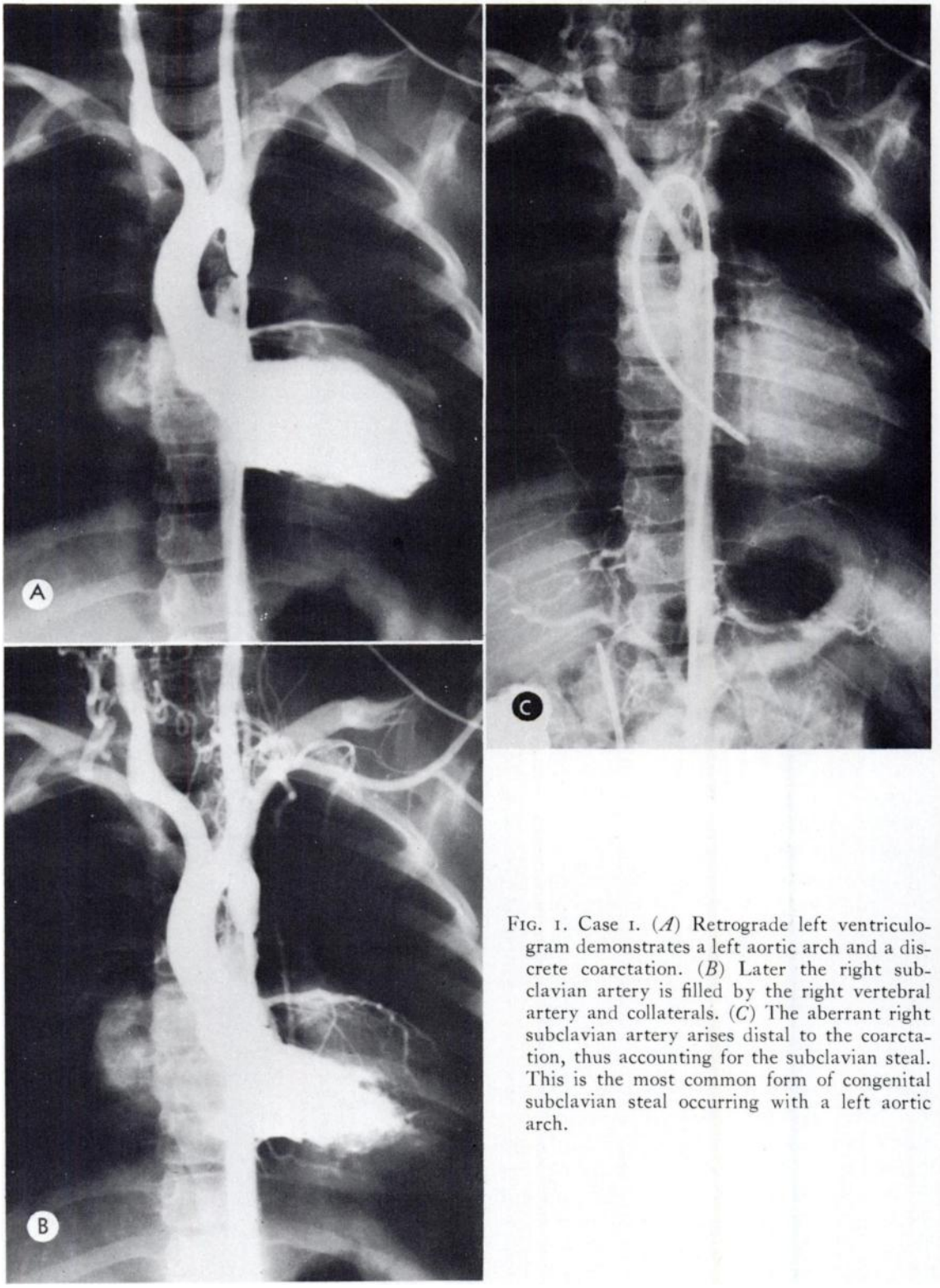

FIG. 1. Case 1. (*A*) Retrograde left ventriculogram demonstrates a left aortic arch and a discrete coarctation. (*B*) Later the right subclavian artery is filled by the right vertebral artery and collaterals. (*C*) The aberrant right subclavian artery arises distal to the coarctation, thus accounting for the subclavian steal. This is the most common form of congenital subclavian steal occurring with a left aortic arch.

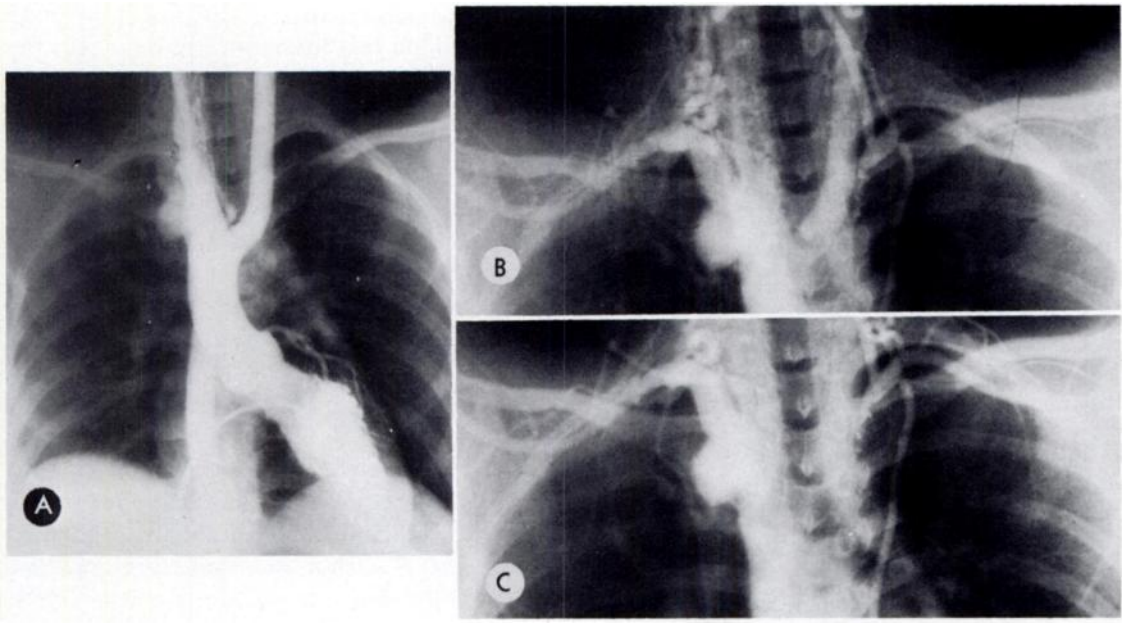

Fig. 2. Case II. (*A–C*) Transseptal left ventriculograms. A right aortic arch, coarctation, and a calcified aneurysm arising immediately distal to the coarctation are demonstrated. The left subclavian artery is isolated from the aortic arch and fills *via* a large left vertebral artery. The right subclavian artery arises at the level of the coarctation and also fills by the right vertebral artery and collaterals.

calcified aneurysm immediately distal to the coarctation (Fig. 2, *A–C*). The left subclavian artery did not fill directly from the aorta but instead filled from retrograde flow in the left vertebral artery and other collateral vessels. The right subclavian artery arose from the aortic aneurysm and was also filled in part by collateral vessels.

Case III. C.G. (JHH# 136 33 41), a 3,600 gm. boy, developed severe congestive heart failure at 15 days of age and was transferred to Johns Hopkins Hospital for evaluation. He was dusky with marked tachypnea. Respirations were 80 per minute and pulse 156. The blood pressure was 100/65 mm. Hg in the right arm and leg and 80/60 in the left arm. The left radial and brachial pulses were absent. He had a gallop rhythm and hepatomegaly caused by heart failure. The ECG showed biatrial enlargement, left ventricular hypertrophy and an indeterminate axis. A right aortic arch, cardiomegaly, increased pulmonary vascularity, and a small posterior esophageal indentation were noted on the roentgenograms. Cardiac angiography demonstrated a large ventricular septal defect with a left-to-right shunt. The left subclavian artery did not fill from the aortic arch but instead was opacified by retrograde flow in the left vertebral artery and other collaterals. A diverticulum was demonstrated in the distal aortic arch.

Case IV. D.B. (JHH# 106 06 33) was a normally developed 13 year old girl who had a murmur first noted at 3 years of age. On physical examination, the blood pressure was 150/0 mm. Hg in the right arm and 165/90/70 in the left arm. Femoral pulses were decreased and the right brachial pulse was present only when the head was turned to the right. She had a left ventricular heave, a thrill in the suprasternal notch and an aortic ejection click preceded by a harsh, Grade 2/4 aortic ejection murmur. The ECG showed left axis deviation and a right-sided conduction delay. The chest roentgenogram showed bilateral cervical ribs, normal heart size and a "figure of 3 sign" indicating coarctation of the aorta. At cardiac catheterization systolic pressures in the left ventricle and proximal aorta were 140 mm. Hg, while in the distal aorta, systolic pressure was 82 mm. Hg. Aortography demonstrated coarctation of the aorta with bicuspid aortic valve and an aberrant right subclavian artery that originated distal to the coarctation. The right subclavian artery filled by retrograde flow from numerous

collateral vessels, including the vertebral artery.

CASE V. R.K. (JHH# 12 00 93), a 53 year old male, suddenly lost the ability to speak following a coughing spell. His past medical history included longstanding congestive heart failure, hypertension and occasional headaches and dizziness. On physical examination, blood pressure in the right arm was 140/90 mm. Hg and in the left arm it was 120/90. The left carotid pulse was diminished. Left hemiparesis, left hemianesthesia, left homonymous hemianopsia, and a left central facial palsy were present. Lumbar puncture, echo encephalogram, brain scan, and ophthalmodynamometry were normal. He improved rapidly and a diagnosis of transient ischemia to the post central region of the right cerebral hemisphere was considered likely. A thoracic aortogram showed a right aortic arch with early bifurcation of the left common carotid artery. The left subclavian artery did not fill directly from the aortic arch but instead opacified from the left vertebral artery and collateral vessels from the left external carotid artery. A right carotid arteriogram showed very tortuous vessels without stenosis or recent occlusion.

DISCUSSION

Significant obstruction or absence of the proximal subclavian artery is associated with the development of collateral circulation to the ischemic upper limb. The most common pathways of collateral blood flow include: (1) from the external carotid artery to the superior thyroidal artery, then to the inferior thyroidal artery, the thyrocervical and costocervical arteries and finally the subclavian artery; (2) from the contralateral internal mammary artery to the ipsilateral internal mammary artery, thence to the subclavian artery; (3) from the aorta and posterior intercostal arteries to the superior intercostal artery, then to the costocervical artery and finally the subclavian artery; (4) from the external carotid artery to the occipital artery, then to the muscular branches of the vertebral artery and finally to the subclavian artery; and (5) from the contralateral vertebral artery to the basilar artery to the ipsilateral vertebral artery and finally the subclavian artery. This last anastomotic network, because of its size, is usually the major collateral pathway.

The degree of obstruction in the subclavian artery which is necessary for retrograde vertebral flow has been the subject of numerous investigations. Flow is reversed when the mean pressure in the basilar artery exceeds that in the vertebral-subclavian system. Reivich *et al.*[12] found that blood flow reversed in man with a drop in systemic artery pressure greater than 10 per cent across the stenotic segment of the subclavian artery. This corresponded to a 50 per cent reduction in the diameter of the lumen. Ekestrom and Retamal[4] noted subclavian steal with a mean pressure gradient of 30 mm. Hg across the stenosis while Sammartino and Toole[13] found that a range of 21–40 mm. Hg was necessary. The steal may be intermittent, fluctuating with changes in pressure and resistance. Because exercise of a limb increases flow and lowers vascular resistance, it probably increases the pressure gradient across the obstruction and facilitates retrograde flow. A compensatory increase in flow in the carotid arteries and contralateral vertebral artery has been observed in the experimental animal and man. It does not, however, compensate for flow lost *via* the ipsilateral vertebral artery and thus, there is a net decrease in cerebral flow.[6,7]

In the adult, intermittent symptoms of cerebral or limb ischemia or both frequently accompany subclavian steal, reflecting inadequate collateral supply to these regions. Exercise may precipitate both central nervous system and limb symptoms. Pain, easy fatigability, and numbness are most frequently noted in the arm, while dizziness, vertigo, headache, and visual symptoms are the most common central nervous system complaints.[8] The spectrum of reported symptoms, however, is great, particularly those related to brain stem ischemia. Since most cases of subclavian steal syndrome are due to atherosclerotic obstruction of the subclavian

artery, it is not surprising that coexistent occlusive disease is found in cerebral vessels in approximately 30 to 40 per cent of patients. In a collective review of the subclavian steal syndrome, only 10 per cent of adult patients were asymptomatic.[10] In infants and children, however, most patients are asymptomatic. Pieroni *et al.*[11] found only 5 of 26 children with congenital subclavian steal who had symptoms definitely ascribable to ischemia. These symptoms usually were parasthesia or exercise intolerance of the affected limb. Of our 5 cases, only Case v, a 53 year old man, had symptoms referable to the central nervous system. The degree to which his subclavian steal contributed to his transient ischemic event is open to speculation. However, 2 patients, 30 and 33 years old have been described with central nervous system symptoms attributable to congenital subclavian steal.[2] Although the natural history of congenital subclavian steal is not known, it appears likely that development of collateral channels does not always compensate fully for the anatomic abnormality.

Fifty-five potential causes for congenital subclavian steal have been described.[2] Many of these have not been observed in man. In the group studied by angiography, the following observations have been made: (1) congenital subclavian steal occurs approximately twice as frequently with a right than with a left aortic arch; (2) when a right aortic arch and subclavian steal coexist, the affected side is usually the left and is caused by hypoplasia, atresia or isolation of the left subclavian artery.[2] Three of our 5 patients had a right aortic arch; all had a left subclavian steal. Two of these patients had a barium esophagogram. Case II had no posterior esophageal indentation, indicating isolation of the left subclavian artery.[15] The posterior esophageal indentation in Case III could have been caused by an atretic band behind the esophagus that connected the aorta to the distal subclavian artery or, more rarely, an interrupted double aortic arch, Subtype III.[16]

Congenital subclavian steal in patients with left aortic arch is almost always associated with localized coarctation or complete interruption of the aortic arch. Eight of 9 patients reviewed by Pieroni *et al.*[11] had coarctation or aortic interruption. Our 2 patients had an aberrant right subclavian artery that arose distal to the coarctation. With a left aortic arch, the side of the subclavian steal is not predictable, since it depends on the relative positions of the subclavian artery and the site of the obstruction.

The incidence of congenital heart disease is increased in patients with congenital subclavian steal. An increased incidence of patent ductus arteriosus, ventricular septal defect or both are particularly common in patients with congenital subclavian steal and a left aortic arch. With a right aortic arch and mirror image branching of the aorta, cardiac malformations are much more common than with right aortic arch and aberrant right subclavian artery, regardless of the presence or absence of subclavian steal.

Cronquist[3] demonstrated retrograde opacification of the opposite vertebral artery in 20 per cent of normal patients with selective vertebral artery injections. Gonzalez *et al.*[5] showed retrograde vertebral artery flow when contrast material was injected in the contralateral brachial artery of a patient who subsequently had a normal aortic arch study; *i.e.*, false positives are possible with selective brachial or vertebral studies, and angiography of the aortic arch is essential for accurate diagnosis.

SUMMARY

Five cases of congenital subclavian steal demonstrated by angiography are reported.

The anatomy, hemodynamics, clinical findings and angiographic features are reviewed.

Ischemic symptoms are infrequently present in infants and children and most cases are discovered incidentally during evaluation of congenital heart disease and/or coarctation of the aorta. The relative lack of symptoms in infants and chil-

dren when compared to adults reflects in part more collateral development and the absence of associated occlusive disease of the cerebral vessels in children. While the natural history of this disorder is not known, there is some evidence that cerebral ischemic symptoms may arise as early as the fourth decade.

Subclavian steal associated with a right aortic arch usually occurs on the left side and is caused by hypoplasia, atresia or isolation of the proximal portion of the left subclavian artery. Subclavian steal associated with a left aortic arch is usually caused by coarctation of interruption of the aortic arch with the affected subclavian artery arising distal to the obstruction.

Arch aortography is the best method to evaluate possible congenital subclavian steal.

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