The Congenital Variety of the "Subclavian Steal" Syndrome

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THROMBOTIC occlusion of the subclavian artery in its intrathoracic portion is common in atherosclerosis, but gangrene of the ipsilateral extremity is rare owing to the extensive collateral network connecting with the branches of the subclavian artery. The vertebral artery, which is the subclavian's first and largest branch, is perhaps the most important channel directing collateral blood to the subclavian artery beyond the point of occlusion. Such retrograde flow through the vertebral artery would siphon blood away from the basilar system and even from the circle of Willis, hence the descriptive term "subclavian steal." Altogether, reports of 14 cases have appeared in the literature during the past 2 years. In addition, a recent case published under the unrelated title "gustatory sweating" was discovered in which occlusion of the left subclavian artery led to the flow of blood from the right to the left vertebral artery and from there to the left subclavian distal to the occlusion, as illustrated clearly by the published angiogram.

The syndrome represents a unique physiologic phenomenon in which loss of blood supply to one region brings about diversion of blood from a distant, unrelated region and causes symptoms not readily explicable in the light of the outward manifestations of ischemia.

All the cases reported heretofore have been in adults with demonstrable atherosclerotic disease of the major cerebral arteries and symptoms of cerebral ischemia resulting from diversion of blood from the basilar system or from the circle of Willis. An example is recorded here of the subclavian steal syndrome caused by congenital atresia of the intrathoracic segment of the left subclavian artery. The child was free from atherosclerosis and exhibited no symptoms of cerebral ischemia.

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Case Report

R.W., a 6-year-old boy was admitted to the Pediatric Department of the D.C. General Hospital for tonsillectomy and adenoidectomy. History also revealed symptoms of a vascular ring. A pulsatile mass was observed in the right lower anterior cervical region, and angiographic studies led to the diagnosis of a cervical aortic arch. It was demonstrated on retrograde aortograms that the proximal segment of the left subclavian artery did not opacify while its distal segment opacified some time after the aortic arch and left common carotid. At the time of the original report the significance of this finding was not appreciated and the delayed opacification was attributed to atresia of the left subclavian trunk and opacification of its distal segment through local collateral vessels.

Subsequent thoracotomy for correction of the vascular ring proved the existence of right common carotid and left subclavian atresia. Recent review of these aortograms with films taken at 1/2-second intervals revealed a situation that has been typical of all the other cases of the "subclavian steal" heretofore reported. Thus, the aortic arch and the right subclavian and the left common carotid arteries opacified promptly and simultaneously. The right common carotid and left subclavian in its proximal segment were known to be atretic and did not opacify (fig. 1). The distal left subclavian and vertebral arteries were not seen on this nor the subsequent 3 films, representing 1 1/2 seconds in time. In film 6, however, opacification of the left vertebral and left subclavian beyond the vertebral junction occurred (fig. 2). At this time the arch, the left common carotid, and the right subclavian had been cleared of dye. It was evident that opacification of the left vertebral artery was retrograde via the arteries within the skull or up in the neck.

With this observation the patient was recalled to the laboratory for further examination. The left axillary pulse was found to be delayed and weak and the left radial was unobtainable. Blood pressure was 110/70 in the right arm and 70/50 in the left. Auscultation over the region of the left subclavian revealed a systolic bruit. Indirect pulse tracings were recorded from all the major arteries, including a very weak left axillary artery (fig. 3). When timed against the Q waves of the simultaneously recorded electrocardiograms, the arrival time, that is, the onset of the upstroke of the left axillary pulse was on the average 0.03
second later than its counterpart on the right; the upstroke was considerably slower, with lower amplitude, and the peak of the pulse wave was on the average 0.12 second later than its counterpart on the right. The pulse tracings from the left carotid and both femoral arteries had normal contour and timing. These findings were interpreted as an indication that the flow to the left axillary artery was indirect and through the long, retrograde path down the vertebral artery.

Neurologic examination was entirely negative as was the electroencephalogram. In contrast to adults with this syndrome, this patient has been free from symptoms and signs of basilar artery insufficiency. Exercising the left arm failed to provoke symptoms. A carotid angiogram was deemed unsafe because the left common carotid, being the only vessel and constituting the major arterial supply to his brain, would have been punctured.

Discussion

The interesting phenomenon of subclavian steal has received considerable attention since Reivich’s paper encompassing both clinical and experimental material.2 Actually, Contorni8 was the first to call attention to this phenomenon in March 1960, when he reported retrograde opacification of the left vertebral artery after injection of contrast material into the right subclavian. It was shown that a vertebro-vertebral flow directed blood from the right to the left vertebral artery. In January 1961, at the Third Conference on Cerebrovascular Diseases under the auspices of the American Neurological and American Heart Associations, Toole and Fields9 described the phenomenon under the term “siphoning,” and Rob10 mentioned a case of ischemia associated with occlusion of the right subclavian artery and relieved by subsequent ligation of the right vertebral artery.

The symptoms are usually those of basilar artery insufficiency and are aggravated by exercising the arm on the side of the subclavian occlusion, for the exercise leads to an increased “steal” of blood from the basilar system. An obvious question is what happens to the many patients who undergo ligation of the subclavian artery for such procedures as Blalock-Taussig shunt for tetralogy of Fallot, for bypassing of a coarctation, and for the use of subclavian artery as the input channel during cardiopulmonary bypass. The Blalock-Taussig procedure as originally described13 involves use of either the subclavian artery coming off the aorta or that originating from the innominate trunk. In the former case, the vertebral artery is to be ligated before the subclavian trunk is pulled down for shunting. In the latter case the subclavian and common carotid arteries are ligated at their origin, allowing the innominate trunk to be pulled down for use in the shunt. This

Figure 1

Retrograde aortogram. Exposure made 1 second after injection of contrast medium into the ascending aorta, showing opacification of the right subclavian-vertebral system and of the left common carotid artery (arrows). The left vertebral-subclavian system has not been opacified as yet.

Figure 2

Exposure made 1½ seconds after that of figure 1, showing clearing of aortic arch, right subclavian-vertebral system, and left common carotid artery of contrast medium. The left vertebral-subclavian system is now opacified via retrograde flow down the vertebral artery (arrows).
latter procedure undoubtedly gives rise to the steal phenomenon in a certain percentage of cases; symptoms of basilar artery insufficiency are usually absent, however, perhaps due to the youthful state of cerebral arteries. In Lamb’s paper on the choice of the side for approach in Blalock-Taussig shunt operations, a case is described (case 4) in which, due to the short length of the right subclavian artery, the vessel was ligated before the origin of the vertebral artery and shunt procedure was completed. Several hours postoperatively the physicians were astounded to find a palpable radial pulse on the right, attesting to an already well-established, sufficient collateral circulation to the arm. There can be little doubt that the collateral circulation was via retrograde vertebral flow. Reivich’s acute experiments show the readiness with which retrograde vertebral flow can be established after ligation of the subclavian before vertebral take-off.

An unusual situation that might superficially resemble basilar insufficiency due to the subclavian steal phenomenon occurs when the vertebral artery on the side of subclavian ligation happens to be the major or the sole contributor to the basilar system. Ligation of the subclavian artery in such cases will deprive the brain from its basilar supply. According to these authors this anomaly exists in 3.1 per cent of the cases on the left, and 1.8 per cent on the right.

North, Fields, and DeBakey have suggested the term “brachial-basilar insufficiency” to replace the term “subclavian steal.” In light of the present case showing retrograde vertebral flow without any symptoms suggestive of basilar artery insufficiency, and the large number of juvenile cases of tetralogy not exhibiting basilar insufficiency after ligation of the subclavian proximal to the origin of the vertebral, it may be wise to retain the descriptive and picturesque term “steal.”

Summary
A case of the subclavian steal syndrome due to congenital atresia of the left subclavian artery is described. Attention is called to the alterations in contour and peak time of the pulse in the involved arm, making clinical diagnosis possible. Absence of signs and symptoms of basilar arterial insufficiency is pointed out and ascribed to the youthful state of the cerebral arteries. The continued use of the term “subclavian steal” is encouraged.

References
7. MUSSUMI, R. A., WIENER, L., AND SHARIFF, P.:

William Withering

Despite the demands of his increasing medical practice and the work required for the publication of his botanical treatise, Withering found time to devote to mineralogy and chemistry. He alternated these with his botanical pursuits, depending on the season. As he expressed it at the beginning of winter, "Botany now no longer presides at my board—her season is past, and chemistry overspreads the table."

Withering's study of the natural and medicinal properties of mineral waters of Great Britain was practically the first really scientific examination and analysis made. His mineralogical researches were important and his discovery of native barium carbonate, named in 1790 by Werner "witherite," has given him a definite place among mineralogists. This mineral, which occurs in large masses and in crystalline form near Hexham, Northumberland, is a white to grayish or yellowish material extensively used in sugar refining, plate glass making, and paint manufacture.—Louis H. Roddis, M.D. William Withering: The Introduction of Digitalis into Medical Practice. New York, Paul B. Hoeber, Inc., 1936, p. 37.
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