Vertebrobasilar Ischemia After Total Repair of Tetralogy of Fallot: Significance of Subclavian Steal Created by Blalock-Taussig Anastomosis

Vertebrobasilar Ischemia After Correction of Tetralogy of Fallot

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SUMMARY Patients who have undergone a Blalock-Taussig anastomosis for treatment of congenital heart disease may have the vascular anatomy of the subclavian steal syndrome. Cerebral ischemia has been reported in such patients, but not when total surgical correction has eliminated other predisposing factors. We report a patient who developed vertebrobasilar insufficiency 31 years after Blalock-Taussig anastomosis and 4 years after total intracardiac repair of tetralogy of Fallot. He had angiographically proven subclavian steal and no other known predisposing factor for cerebral ischemia. This case suggests that symptomatic subclavian steal may be a late risk of surgical treatment of congenital heart disease that leaves the vascular anatomy of subclavian steal intact. Vascular reconstructive surgery can be effective treatment for these patients and may be indicated prophylactically at the time of intracardiac repair if subclavian steal syndrome becomes a more frequently recognized sequela of prior Blalock-Taussig anastomosis.

SYMPTOMS OF CEREBRAL ISCHEMIA in the distribution of the posterior circulation may be caused by occlusion of the basilar artery or its branches, basilar artery stenosis, lacunar strokes, "top of the basilar" ischemia (presumably embolic), vertebral artery occlusion (either intracranial or extracranial) and the subclavian steal syndrome.1 Occlusion of the proximal subclavian artery that produces the subclavian steal syndrome is usually produced by atheromatous disease.2 Performance of a Blalock-Taussig (subclavian-pulmonary artery) anastomosis for treatment of congenital heart disease may also lead to the vascular anatomy of the subclavian steal syndrome. Cerebral ischemia has been reported in patients who have undergone the Blalock-Taussig procedure as palliative treatment of tetralogy of Fallot (pulmonary artery stenosis, ventricular septal defect, dextroposition of the aorta, right ventricular hypertrophy)3,4,5 but has not been reported as a late sequela for those in whom total intracardiac surgical correction has eliminated other predisposing factors for cerebral ischemia. We report such a case below.

**Case Report**

A 38-year-old man was admitted because of incoordination and vomiting. While working at his desk on the day of admission, he had the sudden onset of tinnitus in his right ear, incoordination of his right arm and leg, and vomiting. When he attempted to walk, he listed toward the right and noted a "rubbery feeling" of all four limbs. He developed bi-occipital throbbing headache. There were no associated palpitations, chest pain or fever. There was no history of migraine headaches or prior neurological symptoms.

At age seven, the patient had undergone a left subclavian-to-pulmonary artery anastomosis (Blalock-Taussig procedure) for treatment of tetralogy of Fallot. At age 34, because of progressive exertional dyspnea, he underwent total intracardiac corrective surgery which included pulmonary valvotomy, infundibulec- tomy and closure of the ventricular septal defect. He had no further cardiac symptomatology.

Physical examination revealed arm blood pressures of 140/90 right and 90/70 left; pulse, 60, without orthostatic change; temperature, 98.6°F. There was no cyanosis or evidence of systemic embolization. There were no bruits over the carotids, supraclavicular fossa or posterior neck. A right ventricular heave and a 3/6 holosystolic cardiac murmur were present. Distal arterial pulses were normal.

On neurological examination, he was awake and alert, and his mental status was normal. The neck was supple. There were no retinal emboli. Eye movements were full but saccadic, with ocular dysmetria. Pupils were 4 mm. Motor power and sensation were normal. The plantar responses were flexor. There was dysdiado- kinesis and dysmetria of the right arm and leg. The gait was wide-based and ataxic with a tendency to fall to the right on tandem gait. Romberg’s test was positive.

ECG showed a sinus rhythm with an incomplete right bundle branch block unchanged from previous tracings. Blood hemoglobin was 14.9 gm/dl, and hematocrit was 43%. Prothrombin and partial thromboplastin times, SMA-12, serum electrolytes, white
blood cell count and differential were all normal. Computerized tomography of the head was normal.

Intravenous heparin infusion was begun and within one hour, all symptoms and neurological deficits had resolved. The total duration of symptoms was approximately two hours. Two-dimensional echocardiography and computerized tomography of the thorax showed no evidence of intracardiac thrombus or right-to-left shunt. Twenty-four hour ambulatory ECG monitoring revealed sinus rhythm, rare supraventricular ectopic beats, 1,500 multiform ventricular premature contractions, rare bigeminy and coupling, all asymptomatic. Arch aortogram showed subclavian steal (fig. 1); the vertebral and carotid arteries appeared normal, but the basilar artery was not visualized.

On the sixth hospital day, heparin was discontinued and an anastomosis of left common carotid artery to left subclavian artery was performed (fig. 2C).

Postoperatively equal blood pressures were recorded in both arms. He has not been treated with anticoagulants and remains asymptomatic two years after surgery.

**Discussion**

Reivich et al established the subclavian steal syndrome as stenosis of the subclavian artery proximal to the origin of the vertebral with reversal of blood flow in the vertebral and diversion of blood from the basilar system. Although subclavian steal rarely results in permanent neurologic deficits, disabling and recurrent symptoms may occur. Atherosclerosis is the most frequent cause, but congenital atresia, trauma and surgical obstruction may also produce the syndrome.

Since 1945, anastomosis of the subclavian artery to the pulmonary artery (Blalock-Taussig procedure) has been performed as palliative treatment for patients with tetralogy of Fallot. This procedure may create a vascular anatomy identical to that of subclavian steal.
Figure 2. A: Blalock-Taussig anastomosis resulting in anatomy of subclavian steal. B: Ligature of Blalock-Taussig anastomosis performed at time of total repair of tetralogy of Fallot. C: Carotid-subclavian anastomosis graft correcting subclavian steal.

(fig. 2A), but depending on the development of cervical and intrathoracic collaterals to the subclavian there may not be reversed flow in the vertebral artery. Angiographic evidence of subclavian steal after Blalock-Taussig anastomosis has been demonstrated in 10 to 55% of cases.3,4

All reported patients with neurologic symptoms experienced these before total correction of their tetralogy,3,4,11 at a time when hypoxia, polycythemia,12 bacterial endocarditis,13 cardiac mural or valvular thrombosis, paradoxical embolism,14 cardiac arrhythmias and associated anomalies of the aortic arch or cerebral vessels predispose to cerebral ischemia.15-17 Most had symptoms which were non-localizing (headache, dizziness, blurred vision) and may have been secondary to polycythemia present in each. Although cerebral embolism from a cardiac source was not definitively excluded, our patient developed signs and symptoms of vertebrobasilar insufficiency, had angiographically proven subclavian steal and none of the
other factors predisposing to cerebral ischemia associated with congenital heart disease. No other such patient has been reported.

At the time he developed symptoms, our patient had the anatomy of subclavian steal for 31 years. It is impossible to know how long reversed flow was present in his left vertebral artery, but two possibilities seem likely. His subclavian steal may have been long-standing and asymptomatic. Symptoms may have appeared when a branch artery in the vertebrobasilar system or another major collateral supply to the distal subclavian occluded. Alternatively, the subclavian steal may have developed at the onset of his symptoms in response to loss of alternative collateral to the distal subclavian. These hypotheses infer that another process played a decisive role in producing the symptoms, the most likely candidate being atherosclerotic disease.

The clinical significance of subclavian steal syndrome complicating Blalock-Taussig anastomosis may increase as current surgical treatment improves longevity and allows patients to develop atherosclerosis. Garson et al estimate that 89% of patients with tetralogy of Fallot (2,400 cases per year in the United States) will survive to adulthood, 70% to age 30. Our case suggests that survivors who have had a Blalock-Taussig anastomosis are at risk for vertebrobasilar ischemia from subclavian steal. Thus, a history of corrected congenital heart disease or evidence on physical examination consistent with previous cardiac surgery should not be treated casually in the patient with vertebrobasilar ischemia.

When Blalock and Taussig described their operation for tetralogy of Fallot, they did not discuss management of the vertebral artery. Reivich recommended, if possible, to ligate the subclavian artery distal to the origin of the vertebral. If the subclavian must be ligated proximal to the vertebral artery, he suggested also ligating the vertebral. (In some centers, this may be current practice.) He cautioned that ligation of a vertebral artery is not without danger, however, since various vascular anomalies of the aortic arch and base of the brain may co-exist with the tetralogy of Fallot. The lack of connection between one vertebral and the basilar artery is one such anomaly. In these cases, ligation of the vertebral artery could have severe consequences.

Comprehensive cerebral angiography pre-operatively and intra-operative electroencephalography and/or cerebral blood flow monitoring may be useful in making a decision whether or not to ligate the vertebral artery. Management of the vertebral artery during Blalock-Taussig anastomosis remains an unsettled issue.

The vascular anatomy of the subclavian steal syndrome is generally not altered by intracardiac repair of the tetralogy of Fallot when ligature of the Blalock-Taussig anastomosis is performed (fig. 2B). When intracardiac repair is performed on an older child or adult, cerebral ischemia from subclavian steal could be avoided by simultaneous reconstruction of the distal subclavian artery blood supply using a procedure such as was employed in our patient (fig. 2C). This would leave patients at risk for vertebrobasilar ischemia during the interval between Blalock-Taussig anastomosis and total repair but would ultimately provide the best approximation of the normal blood flow both to arms and brain. Since vascular anastomosis may not be feasible in younger children, their appropriate management is uncertain.

We believe that subclavian steal must be considered a possible etiology for vertebrobasilar ischemia occurring after Blalock-Taussig anastomosis and intracardiac repair of tetralogy of Fallot. If subclavian steal is demonstrated and if symptoms are disabling or recurrent, consideration should be given to construction of a carotid-subclavian shunt. Whether or not prophylactic vascular reconstruction should be performed at the time of total intracardiac repair awaits further experience concerning the frequency of symptomatic subclavian steal in this group.

References
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