Subclavian Steal in Patients with Blalock-Taussig Anastomosis

By Gordon M. Folger, Jr., M.D., and Kishor D. Shah, M.D.

Although the collateral anastomotic channels that occur secondary to obstruction of the first segment of the subclavian artery have received relatively little comment by anatomists, the participation by the vertebral artery in such instances has been mentioned. Recently, the finding of retrograde flow from the vertebral artery on the side of an obstruction in the first part of the subclavian artery to the distal segment of that subclavian artery has been noted by several investigators. Anatomically, obstruction may be complete or, if partial, of sufficient severity to lower the pressure in the subclavian artery to below that in the vertebral artery. The reversal of flow, demonstrable angiographically, may cause basilar artery insufficiency giving rise to symptoms of headache, visual disturbance, paresthesia, hemiparesis, dizziness, and syncope. This condition, first reported by Reivich and co-workers, has been termed the "subclavian steal syndrome." With the exception of one report, the subclavian obstruction in all the cases of the entity recorded to date has been on the basis of atherosclerotic disease within the vessel.

Anastomosis of subclavian artery to pulmonary artery has been performed since 1945 on a large number of patients suffering from cardiac anomalies associated with pulmonary stenosis or atresia. On the assumption that patients treated in this manner might exhibit the subclavian steal angiograms performed at the Johns Hopkins Hospital on such patients between 1961 and 1964 were reviewed with specific attention directed to the manner of opacification of the subclavian artery distal to the anastomotic site. The following report concerns the findings in 12 patients with Blalock-Taussig anastomoses who have shown evidence of the subclavian steal. To our knowledge this condition in patients so treated has not previously been reported.

Materials and Methods

A total of 123 biplane angiograms done by the selective injection of contrast material into either the right or left ventricular cavity was reviewed. Only frontal projections were examined. Exposure speeds varied somewhat, with the majority of studies performed at four or six frames per second.

Care was taken to include only angiograms of those patients having a subclavian artery to pulmonary artery anastomosis and only angiograms that were satisfactory from all technical aspects. After this selection 114 angiograms were acceptable for analysis.

The acceptable angiograms were then subdivided into four groups. Group 1 consisted of 12 studies that were considered to have distinct evidence of late filling of the distal subclavian artery from the vertebral artery on the side of the Blalock-Taussig anastomosis. Group 2 consisted of eight angiograms that revealed late filling of the distal subclavian artery from an anastomotic channel other than the vertebral artery on the side of the Blalock-Taussig anastomosis. Interestingly, one patient with bilateral anastomoses demonstrated findings of group 1 on one side and group 2 on the other and is included in both groups. In group 3 were 91 angiograms in which late filling of the distal subclavian artery on the side of the Blalock-Taussig anastomosis was apparent but the contributing anastomotic channels could not be positively identified. Group 4 was comprised of four angiograms in which the distal subclavian artery on the side of the Blalock-Taussig anastomosis never opacified. This was an unexpected occurrence and the number of studies in this group is understandably small.

In groups 1 and 2 the charts of the patients

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Table 1

Data on Patients with Subclavian Steal

<table>
<thead>
<tr>
<th>Pt.</th>
<th>Sex, age (yr.)</th>
<th>Cardiac diagnosis</th>
<th>Age first visit</th>
<th>Hb. (Gm. %)</th>
<th>Type of anastomosis age performed</th>
<th>Radial pulse</th>
<th>Rt. arm</th>
<th>Lt. arm</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. MW</td>
<td>F; 37</td>
<td>T/F,* corrected Jan. 1964</td>
<td>8% (yr.)</td>
<td>23/79</td>
<td>RBT† 9 yr.</td>
<td>Weak</td>
<td>Normal</td>
<td></td>
</tr>
<tr>
<td>2. MF</td>
<td>M; 11</td>
<td>T/F, corrected April 1962</td>
<td>2 yr.</td>
<td>18.8/72</td>
<td>RBT 2 yr.</td>
<td>0</td>
<td>Normal</td>
<td></td>
</tr>
<tr>
<td>3. MM</td>
<td>M; 11</td>
<td>T/F</td>
<td>18 mo.</td>
<td>17.1/52</td>
<td>RBT 18 mo. LBT‡ 3½ yr.</td>
<td>0</td>
<td>Normal</td>
<td></td>
</tr>
<tr>
<td>4. AB</td>
<td>F; 21</td>
<td>T/F, corrected Aug. 1961</td>
<td>5% (mo.)</td>
<td>21/65</td>
<td>RBT 5 yr.</td>
<td>Weak</td>
<td>Normal</td>
<td></td>
</tr>
<tr>
<td>5. SB</td>
<td>M; 13</td>
<td>T/F</td>
<td>5 yr.</td>
<td>19.3/64.5</td>
<td>LBT 5% yr.</td>
<td>Normal</td>
<td>Weak</td>
<td></td>
</tr>
<tr>
<td>7. EZ</td>
<td>M; 15</td>
<td>T/F</td>
<td>12 mo.</td>
<td>16.5/55</td>
<td>LBT 12 mo. Pott’s/6 yr.</td>
<td>Normal</td>
<td>0</td>
<td></td>
</tr>
<tr>
<td>8. JF</td>
<td>M; 12</td>
<td>T/F</td>
<td>2 9/12 (yr.)</td>
<td>19.9/72</td>
<td>RBT 3½ yr.</td>
<td>Weak</td>
<td>Normal</td>
<td></td>
</tr>
<tr>
<td>9. CC</td>
<td>F; 16%</td>
<td>T/F, corrected Nov. 1962</td>
<td>13 mo.</td>
<td>17.5/50</td>
<td>LBT 3 yr.</td>
<td>Normal</td>
<td>Weak</td>
<td></td>
</tr>
<tr>
<td>10. CH</td>
<td>M; 15</td>
<td>T/F</td>
<td>9 yr.</td>
<td>13.2/41</td>
<td>LBT 9 yr.</td>
<td>Normal</td>
<td>0</td>
<td></td>
</tr>
<tr>
<td>11. AD</td>
<td>F; 19</td>
<td>T/F</td>
<td>3 yr.</td>
<td>–/60</td>
<td>LBT 5% yr.</td>
<td>Normal</td>
<td>0</td>
<td></td>
</tr>
<tr>
<td>12. RG</td>
<td>F; 17</td>
<td>T/F, corrected Nov. 1963</td>
<td>4 yr.</td>
<td>26.5/78.5</td>
<td>LBT 4 yr.</td>
<td>Normal</td>
<td>Weak</td>
<td></td>
</tr>
</tbody>
</table>

*Tetralogy of Fallot.
†Right Blalock-Taussig anastomosis (end-to-side).
‡Left Blalock-Taussig anastomosis.
<table>
<thead>
<tr>
<th>Blood pressure</th>
<th>Highest Hb. (Gm. %)</th>
<th>Time (yr.) from anast. to increase of Hb./Hcrt.</th>
<th>Type of cerebral disturbance and degree</th>
<th>Time (yr.) from anast. to symp.</th>
</tr>
</thead>
<tbody>
<tr>
<td>Rt. arm</td>
<td>Lt. arm</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>85</td>
<td>115</td>
<td>19.8</td>
<td>12</td>
<td>Severe headache 4-6 hr. duration; dizziness and faint feeling 2-4 times per week; blurred vision</td>
</tr>
<tr>
<td>70</td>
<td>70</td>
<td>67</td>
<td></td>
<td>14</td>
</tr>
<tr>
<td>95</td>
<td>125</td>
<td>16.4</td>
<td>5</td>
<td>Severe headaches and dizziness (From 2nd anast.)</td>
</tr>
<tr>
<td>70</td>
<td>72</td>
<td>53</td>
<td></td>
<td>6</td>
</tr>
<tr>
<td>105</td>
<td>75</td>
<td>18.7</td>
<td>5</td>
<td>Occasional severe headache; vomiting, eye pain</td>
</tr>
<tr>
<td>60</td>
<td>60</td>
<td>58</td>
<td></td>
<td>1</td>
</tr>
<tr>
<td>128</td>
<td>0</td>
<td>19.5</td>
<td>4</td>
<td>Frequent severe headache; episodes of weakness and faintness</td>
</tr>
<tr>
<td>70</td>
<td></td>
<td>64.5</td>
<td></td>
<td>2-3</td>
</tr>
<tr>
<td>142</td>
<td>0</td>
<td>23</td>
<td>4</td>
<td>Frequent headache and vomiting; dizziness; recent onset of diplopia (1 episode)</td>
</tr>
<tr>
<td>80</td>
<td></td>
<td>69</td>
<td></td>
<td>8</td>
</tr>
<tr>
<td>80</td>
<td>100</td>
<td>17.2</td>
<td>5-6</td>
<td>Headache usually following exercise; episodes of weakness</td>
</tr>
<tr>
<td>40</td>
<td>40</td>
<td>55</td>
<td></td>
<td>3-4</td>
</tr>
<tr>
<td>110</td>
<td>0</td>
<td>17.5</td>
<td>5</td>
<td>None</td>
</tr>
<tr>
<td>80</td>
<td></td>
<td>54</td>
<td></td>
<td></td>
</tr>
<tr>
<td>110</td>
<td>0</td>
<td>15.8</td>
<td>4</td>
<td>Four episodes of protracted headache and vomiting</td>
</tr>
<tr>
<td>70</td>
<td></td>
<td>52</td>
<td></td>
<td>2</td>
</tr>
<tr>
<td>110</td>
<td>0</td>
<td>17</td>
<td>–</td>
<td>None</td>
</tr>
<tr>
<td>80</td>
<td>52</td>
<td></td>
<td></td>
<td>None</td>
</tr>
<tr>
<td>110</td>
<td>0</td>
<td>19.5</td>
<td>Not known</td>
<td>None</td>
</tr>
<tr>
<td>70</td>
<td>60.5</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
were reviewed, questionnaires were sent to all surviving patients, and when possible the patients themselves were examined for findings suggesting basilar artery insufficiency. All the available material was evaluated with respect to neurologic symptoms and their temporal relationships to anastomotic procedures, degrees of peripheral oxygen desaturation, and, when pertinent, the effect of total correction of the cardiac malformation.

The patients in groups 3 and 4 were of necessity excluded from further analysis in view of the inability to demonstrate adequately the vascular pathways leading to the involved subclavian artery.

Case Analysis

The pertinent data on all 12 patients comprising group 1 are summarized in table 1. The cardiac malformation in all is the tetralogy of Fallot. All the patients are Caucasian. Four of these patients are briefly described as follows.

Case 1

M.W. (JHH no. 41 63 14) is a 37-year-old housewife who has been followed in the Cardiac Clinic since the age of 8 years with the diagnosis of tetralogy of Fallot. Because of increasing cyanosis in 1947, at the age of 10 years, she underwent anastomosis of the end of the right subclavian artery to the side of the right pulmonary artery with satisfactory improvement, and she was able to lead a normal life for 14 years. Throughout this period the physical findings were those of tetralogy of Fallot with a functioning Blalock-Taussig anastomosis. Moderate polycythemia persisted, which increased gradually although never reaching alarmingly high levels. Pulses were always obtainable, though weaker in the right arm than in the left; blood pressure in the right arm was 85/70 mm. Hg compared to 120/55 mm. Hg in the left arm.

The patient delivered a 4-pound stillborn infant in 1959, but in 1960 she gave birth to a 5-pound 4-ounce normal infant after a full-term pregnancy.

Six months later she met with a minor auto accident following which she noted the onset of dizzy spells and severe headaches, although there was no evidence of central nervous system injury. The headaches occurred several times daily, often lasting for a number of hours, and were unrelated to exertion or posture. Hemoglobin and hematocrit values at this time were 18.2 Gm. per cent and 62 per cent, respectively.

Right ventricular selective angiocardiography in 1963 revealed typical findings of tetralogy of Fallot. The left vertebral and subclavian arteries were seen to opacify normally from the arch of the aorta 2½ seconds after the injection. Retrograde opacification of the right vertebral and subsequently the right subclavian arteries was observed 4½ seconds later.

The following year she underwent total correction of her cardiac defect with the use of cardiopulmonary bypass. Four months postoperatively she is leading a normal life and has had no recurrence of headache or dizziness.

Comment

This patient had symptoms suggestive of basilar artery insufficiency 14 years after creation of a Blalock-Taussig anastomosis. At this time there was no evidence of increasing hypoxemia and just prior to this she was capable of carrying a pregnancy to full term satisfactorily. Although the automobile accident in which she was involved may have contributed to this problem, she was not known to have suffered injury, and several examinations failed to uncover evidence of cerebral trauma. Selective angiocardiography confirmed retrograde filling of the right subclavian artery by way of the right vertebral artery.

Case 2

M.F. (JHH no. 69 15 65) is an 11-year-old boy who was first seen at the age of 9 months because of cyanosis and cyanotic spells. He was diagnosed as having tetralogy of Fallot. In view of increasing cyanosis, anastomosis of the end of the right subclavian to the side of the right pulmonary artery was performed at 2 years of age followed by marked improvement.

He was able to lead an active, unrestricted life over the following 4½ years. From this time, however, he gradually developed noticeable fatigability and when seen 5½ years postoperatively he had significant polycythemia with hemoglobin and hematocrit values of 18 Gm. per cent and 60 per cent, respectively. He was only slightly cyanotic and the continuous murmur originating from the anastomosis was satisfactory.

When seen 7½ years postoperatively he was markedly incapacitated. His cyanosis had deepened and his hemoglobin had risen to 21.3 Gm. per cent with a hematocrit value of 67 per cent. At no time were blood pressures or pulses recorded in the right arm.

A right ventricular selective angiocardiogram revealed findings typical of tetralogy of Fallot. The left subclavian and vertebral arteries were seen to opacify from the arch of the aorta 2 seconds following injection (fig. 1A); opacification of the right subclavian artery in a retrograde manner from the right vertebral artery was observed 8½ seconds from injection, a lag of 8½ seconds from opacification of the left vertebral artery (fig. 1B and C).

Total correction of his tetralogy of Fallot under
cardiopulmonary bypass was carried out without complication on April 25, 1962, and the child has remained well since that time.

Comment

At no time did this patient present any clinical evidence of basilar artery insufficiency. Yet, his angiocardiograms revealed retrograde filling of the right subclavian artery from the right vertebral artery after a delay of greater than 6 seconds from the time of left vertebral artery visualization.

Case 3

M.M. (JHH no. 78 61 49) is an 11-year-old boy who was initially seen in the Cardiac Clinic at the age of 6 months for a heart murmur and one episode of cyanosis. Although acyanotic on examination, he had clinical and laboratory findings indicating a mild tetralogy of Fallot.

By 18 months of age fatigue and dyspnea had appeared and the child squatted frequently. In addition cyanosis and polycythemia were prominent. An anastomosis was created between the end of the right subclavian and the side of the right pulmonary artery. This was followed by immediate improvement, but by 18 months after this procedure he had again begun to show signs of increasing fatigability and dyspnea and by 2½ years after operation cyanosis and polycythemia were prominent. At no time were pulses in the right arm detected.

Because of increasing difficulty an anastomosis of the end of the left subclavian artery to the side of the left pulmonary artery was carried out 3½ years after the first procedure. Although this was followed by marked improvement, he remained mildly cyanotic with moderate dyspnea and fatigability on exertion.

At the age of 9 years he developed the onset of recurrent severe headaches and episodes of dizziness without visual disturbance. Additionally, increasing fatigability and cyanosis were noted. At age 10½ years physical examination revealed palpable pulses in the left arm with a blood pressure in this extremity of 90/70 mm. Hg. Moderate cyanosis and clubbing were present. A prominent continuous murmur was heard. The hemoglobin and hematocrit values were 15.3 Gm. per cent and 54.5 per cent, respectively.

A selective right ventricular angiocardiogram revealed findings typical of tetralogy of Fallot with bilateral functioning Blalock-Taussig anastomoses. Neither vertebral nor subclavian artery was seen to opacify from the arch of the aorta. Two seconds after the carotid arteries were visualized retrograde opacification of the left subclavian artery from the left vertebral artery was seen. Of further interest was the finding 2½ seconds later of opacification of the right subclavian

Figure 1

Case 2. A, top. Two seconds following injection of contrast material. Opacification of the left subclavian artery (arrows) and the right subclavian to right pulmonary artery anastomosis are seen. The distal right subclavian artery does not opacify. B, center. Eight and three-quarters second after injection. The right vertebral and right subclavian arteries are opacified (arrows). C, bottom. Magnified view of the right vertebral and right subclavian arteries (arrows). The presence of a second vessel in this area suggests the contribution of the thyrocervical trunk in this instance.
artery in a retrograde manner from an anastomotic channel in the region of the right axilla. The right vertebral artery was, however, never observed to opacify.

**Comment**

This patient with bilateral anastomoses had onset of cerebral symptoms at 9 years of age when he showed some increase in fatigability and cyanosis but without evident polycythemia. The blood pressure was recordable in the left arm only. The angiogram demonstrated filling of the left subclavian artery from the left vertebral artery and the right vertebral artery was not opacified. In view of these findings it is apparent that the vertebral artery on the left was the only functional vertebral artery. Therefore, it is conceivable that the runoff from the basilar system to the left subclavian artery may have been highly significant.

**Case 5**

S.B. (JHH no. 76 93 46) is a girl who was first seen at the age of 5 years in 1956 with the diagnosis of tetralogy of Fallot. Because of intense cyanosis, frequent cyanotic spells, and polycythemia, a left subclavian artery to left pulmonary artery anastomosis was performed, which resulted in considerable improvement. Ten months following surgery, however, she had had several episodes of severe headache generally localized to the parietal areas, more severe on the left. Severe pain in the eyes and vomiting occurred with these attacks, which usually lasted several hours.

Due to the persistent nature of this complaint an electroencephalogram was performed 3 years following surgery which was interpreted as normal. The physical findings during this time, although compatible with tetralogy of Fallot with a functioning Blalock-Taussig anastomosis, had at no time suggested a central nervous system abnormality. She had remained mildly cyanotic. The blood pressure was recorded in the left arm at 75/60 mm. Hg and in the right arm at 95/60 mm. Hg. Moderate polycythemia was present from 1960 without significant progression.

Right ventricular selective angiography in 1962 confirmed the clinical impression of tetralogy of Fallot. The right subclavian artery was seen to opacify normally 1 2/3 seconds after the injection. The left subclavian to pulmonary artery anastomosis was at this time readily visualized (fig. 2A). Two seconds later the distal left sub-

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**Figure 2**

Case 5. A, top. One and two-thirds second after injection of contrast material. The right subclavian artery is seen to opacify normally (arrow). The distal left subclavian artery is not seen. B, center. Three and two-thirds seconds after injection. The left subclavian artery is opacified and is seen to be continuous with the left vertebral artery (arrows). C, bottom. Magnified view of the left vertebral-left subclavian artery communication.
The clavian artery was noted to opacify in a retrograde manner from the left vertebral artery (fig. 2B and C).

Comment
This child developed symptoms indicating a central nervous system abnormality shortly following a Blalock-Taussig anastomosis. Although her attacks have had features suggestive of migraine, the angiographic demonstration of reversal of flow from the left vertebral artery to the distal left clavian artery and the return of recordable blood pressure to the left arm favors the diagnosis of clavian steal. Because of her otherwise satisfactory course to the present time, corrective surgery has not yet been performed.

Discussion
The diagnosis of the clavian steal rests with the angiographic demonstration of delayed retrograde filling of the vertebral-clavian system on the side of an obstruction of the first part of the clavian artery. This is due to the pressure differential between the vertebral artery and the obstructed clavian artery, flow being in the direction of the lower pressure. To make the diagnosis, one must ascertain that the clavian opacification has not occurred from any other possible anastomotic channel.

In none of the angiograms reviewed for the present study was a complete view of the neck available, as the angiograms were made for the analysis of the cardiac malformation and not primarily for the determination of the existence of a vertebral-clavian communication. For this reason our diagnostic criteria were identification of a cervical vessel in the position of the vertebral artery from which the clavian artery opacified and the presence of opacification sufficiently late that direct anastomosis with an artery immediately communicating with the aorta or carotid arteries seemed unlikely.

Symptoms suggesting basilar artery insufficiency were present in seven of the 12 patients comprising group 1. Most prominent among these was severe headache, which was present in all these seven patients. Dizziness was found in three patients. Feeling of faintness and weakness was noted in three patients. Two patients had noticed visual disturbances: one complained of diplopia and the other of blurred vision. Although all these complaints are recognized symptoms of basilar artery insufficiency, headache was unusually prominent in our group. Because of this, hypoxia was considered as a cause; it is evident however from table 1 that four of the symptomatic patients had developed cerebral symptoms prior to the development of significant polycythemia, and, of the remaining three, in only one (case 7) was there greater than a 2-year period from this initial elevation of hemoglobin and hematocrit levels to the development of symptoms. The time interval between the creation of the anastomosis and the appearance of symptoms varied from 1 to 14 years. It is of interest that only one of the seven patients in the second group, i.e., those in which clavian opacification occurred through channels other than the vertebral artery, exhibited any of the above-described symptoms, although these patients were comparable in age, hemoglobin and hematocrit levels, and in periods of time from the creation of the anastomosis.

It is apparent that a clavian steal phenomenon should be considered in all patients with Blalock-Taussig anastomosis who manifest symptoms of cerebral disturbance.

The recent report of Massumi et al. of an asymptomatic child with angiographic evidence of clavian steal is worthy of note. Although our patients form a younger group than those previously reported with symptoms, it is possible that in our group the degree of hypoxia present, together with the clavian steal, contributed significantly to cerebral ischemia, which may be comparable to that caused by the cerebral vascular changes in the older patients.

Only two of the patients of the symptomatic group have undergone total correction of their cardiac defects. Although the period of observation since surgery is short, both are symptom-free 6 months and 2 years postoperatively. It is impossible, however, to state at present whether our patients who are totally corrected will develop symptoms of clavian artery insufficiency with the degenerative vascular changes of advancing age.
Conclusion

Twelve patients with subclavian to pulmonary anastomoses who have angiographic evidence of the "subclavian steal" are reported. In seven of these patients symptoms commonly associated with basilar artery insufficiency were present. It is our belief that the reduction of cerebral blood flow caused by the runoff from the vertebral-basilar system to the distal portion of the ligated subclavian artery, together with significant degrees of hypoxemia from which these patients suffered is the cause of their symptoms.

Acknowledgment

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References


Principles of Medical Sciences and Animal Vivisection

Animal experiment, which we have ventured to call collateral experiment, must always be the chief instrument for the solution of fundamental medical problems. It has to be recognized, however, that the functional gap between man and other animals, and especially those commonly available in laboratories, is larger even than the morphological gap. . . .

The need for such caution is, however, fundamental and general, if medicine is to get the fullest benefit from what has been, and still is, its most fruitful resource.—The Collected Papers of Wilfred Trotter, F.R.S. London, Oxford University Press, 1946, p. 125.
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