Neonatal Congestive Heart Failure as the Presenting Symptom of Cerebral Arteriovenous Malformation

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Although peripheral fistulas between major vessels elsewhere in the body may give rise to cardiac enlargement and cardiac failure, it has been generally assumed that even large arteriovenous malformations of the brain do not result in congestive heart failure. It was therefore surprising to be consulted with regard to a newborn girl in congestive heart failure, the basis of which was thought to be a cerebral arteriovenous malformation as deduced from studies of oxygen concentration obtained during heart catheterization. We verified the diagnosis of arteriovenous malformation by cerebral angiography and believe that this lesion was the cause of the cardiac failure.

Curiously, when Steinheil first described an arteriovenous malformation, he noted cardiomegaly. However, in a review of several large series, a total of 219 patients, no cardiac symptoms were noted, while one series described cardiomegaly in 9 of 110 cases. The relationship between cerebral arteriovenous malformation and the cardiovascular system was attacked more directly by Höök et al. who evaluated 13 such patients from this point of view. In no case were symptoms present to indicate an increased load on the heart or the circulation. The histories of an additional 123 patients were analyzed, again without demonstration of cardiovascular symptoms. It is pertinent that the youngest patient in that report was 14 years old and, indeed, the calculation has been made that only 35% of arteriovenous malformations become symptomatic before the age of 20. In a series dealing with 11 children between 4 and 15 years of age, there was no evidence for congestive heart failure accompanying such a cerebral lesion.

A search of the literature reveals 11 other patients who had a cerebral arteriovenous malformation associated with signs of congestive heart failure and without any other lesion to explain the cardiac symptoms. It is particularly interesting that all the patients were infants, 75% of whom were found to have cardiac failure within the first 3 days of life.

With increasing sophistication in interpreting the results of cardiac catheterization, we can expect that congestive failure secondary to cerebral arteriovenous malformation in infancy, will become a more frequent diagnostic entity. This report is an account of our findings in one patient and a review of the 11 other cases which can be ascribed to the same syndrome.

Case Report

An 8-day-old girl, referred by Dr. M. K. Sartwell, was transferred to the Children's Hospital of the District of Columbia for cardiac evaluation because of dyspnea and cardiomegaly. The baby weighed 6 lb. 10 oz. at birth. Delivery at full term had been uncomplicated although she was noted to be "dusky." Because she had taken only 1 oz. of formula per feeding, intravenous therapy was begun on the 5th day of life. Jaundice noted between the 3rd and 5th days, was interpreted as being "physiological." Because of persistent dyspnea, a chest x-ray was obtained on the 5th day, revealing cardiomegaly.

Examination. On admission, on the 8th day of life, the blood pressure was 90/60 with a pulse rate of 120 per minute and 110 respirations per minute. Head and chest circumference were 33 cm. and 29 cm., respectively. The patient exhibited peripheral cyanosis. The heart beat was forceful and the heart appeared to be enlarged to the left on percussion, with a hyperactive right ventricular impulse. No murmurs were heard. The liver was 6 cm. below the right costal margin. The rest of the physical examination was normal.

Laboratory findings included the following: hemoglobin, 19.4 gm./100 ml.; hematocrit, 62%; leukocytes, 9800/cubic mm.; serum bilirubin, 8.6 mg. % (indirect, 8.1 mg. %); normal serum electrolytes; electrocardiogram suggestive of right atrial hypertrophy.

Course. After admission the patient was treated
with oxygen and digitalis. On the 12th day of life, right heart catheterization was performed by Dr. L. Scott and Dr. A. Sparrow. This procedure (Table 1) revealed highly oxygenated blood in the pulmonary artery, the right ventricle, the right atrium, the superior vena cava and both internal jugular veins up to the angle of the jaw. This high degree of oxygen saturation from the venous system indicated a mass of arterialized blood within the skull. The finding of low saturation in the inferior vena cava was of crucial significance. On the 13th day a cranial bruit was heard for the first time; there were no other abnormal neurological findings. The patient's condition gradually deteriorated.

On the 15th day a right brachial arteriogram was performed. The views obtained (Fig. 1) outlined the lesion which consisted of a large arteriovenous malformation, apparently fed by the posterior, middle, and anterior cerebral arteries by way of the right internal carotid and vertebral-basilar systems, and draining into a dilated vein of Galen. Immediately following arteriography, the baby's heart stopped beating and she died despite intubation and external cardiac massage.

At autopsy, reported by Dr. S. C. Woodward, the cerebral hemispheres were sectioned. A large vascular channel, completely obliterated by a recent antemortem thrombus, was found in the inferior portion of the right temporal lobe. The thrombosed channel was in close proximity to the right posterior cerebral artery and appeared to be connected to it. It also passed into the greatly enlarged and elongated vein of Galen by way of several smaller vessels. The vein of Galen extended from the moderately dilated straight sinus as far anteriorly as the base of the thalamic pulvinar. Examination of the heart disclosed a mild coarctation of the aorta (6 mm.) and a patent ductus arteriosus (3 mm.) which were interpreted as incidental findings, being too small to represent the cause of the intractable heart failure. Both carotid arteries were hyperplastic, measuring 4 mm. in diameter at their point of origin and enlarging to 8 mm. at the cervical level. The heart together with the lungs weighed 113 gm.; (the normal value was considered to be 74 gm.). The right ventricular wall was 5 mm. in thickness and the left was 6 mm. On microscopic examination, the left ventricle disclosed a linear streak of degeneration of the myocardial fibers with congestion of the vessels, whereas the right ventricle contained only small focal areas of degeneration.

### Table 1

<table>
<thead>
<tr>
<th>Site</th>
<th>Percentage Oxygen Saturation</th>
</tr>
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<tr>
<td>Left atrium</td>
<td>82.5</td>
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<tr>
<td>Superior vena cava (low)</td>
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<tr>
<td>Pulmonary artery</td>
<td>84.5</td>
</tr>
<tr>
<td>Right atrium</td>
<td>84.5</td>
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<tr>
<td>Superior vena cava (high)</td>
<td>84.0</td>
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<td>Left jugular vein</td>
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<tr>
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<tr>
<td>Inferior vena cava (high)</td>
<td>63.0</td>
</tr>
<tr>
<td>Inferior vena cava (low)</td>
<td>53.0</td>
</tr>
</tbody>
</table>

**Fig. 1.** Lateral and anterior-posterior views after injection of the right brachial artery showing aneurysmal dilatation of the vein of Galen with arterial feeding vessels from the anterior, middle, and posterior cerebral arteries.
The anatomic diagnosis was congestive heart failure secondary to an arteriovenous malformation involving the vein of Galen.

Review of Cases

In reviewing reports of cerebral arteriovenous malformations in association with congestive heart failure, those patients with basic cardiac anomalies and those in whom only cardiomegaly without frank failure were detected, have been stringently eliminated. For the same reason, 2 cases with concomitant subdural hematoma are not discussed. The 12 remaining patients (Table 2), including the one reported here, were seen in cardiac failure with a cerebral arteriovenous malformation as the apparent cause.

In 8 patients the diagnosis was made after an autopsy; in 4 it was established by cardiac or cerebral angiography during life. All 12 required medical care because of frank congestive heart failure during the first 12 weeks of life and 8 of these during the first 3 days of life. Their course was one of rapid, progressive deterioration including the development of cardiomegaly, hepatomegaly and peripheral edema leading to death, usually within 2 weeks of onset. Two patients had surgery. One died four days after ligation of the common carotid artery and the jugular vein on the same side. The patient who survived had a ligation of the common carotid on one side and the external carotid on the other.

The malformation directly involved a major venous sinus in all patients and in 8 of the 12 took the form of a vein of Galen anomaly. Although the observed neurological signs and symptoms presented by this group are limited, it is known that such malformations commonly act as a mass lesion, often resulting in hydrocephalus because of obstruction of the aqueduct of Sylvius. A bruit over the head is common. Seizures and subarachnoid hemorrhage have been noted with malformations involving the vein of Galen.

The relationship between arteriovenous malformation and the cardiovascular system has been demonstrated by digital occlusion of the fistulous communications. The results were as follows. 1. Bradycardia was observed and the diastolic and systolic pressures increased immediately. 2. The systemic venous pressure increased. 3. When followed fluoroscopically, a slight decrease in the size of the heart shadow was noted. In 1 patient...
the cranial bruit could be eliminated by pressure over the carotid artery.\textsuperscript{38} We must, however, inject a note of caution with regard to excess manipulation in the area of the carotid sinus in a patient with a badly compromised circulation; the resulting bradycardia may be sufficient to precipitate the patient’s death. Indeed, this may have been a cause of death in our patient.

**Discussion**

In the absence of other pathological findings, most notably the lack of significant congenital cardiac anomalies, one is led to conclude that the congestive heart failure in the 12 patients under discussion was due to the cerebral arteriovenous malformation. The characteristic compensatory increase in cardiac output\textsuperscript{5,19} accompanying the flow of arterial blood directly into the venous system would be expected to result in cardiomegaly and, if sufficiently severe, in congestive heart failure. Although this summarizes the situation, we must take account of the absence of similar findings in adults with equally large arteriovenous anomalies.

For an arteriovenous shunt of a given size, the proportion of cardiac output entering the shunt is much greater in an infant than in an adult. Thus, an adult brain representing 2% of the body weight\textsuperscript{9} receives 17% of the cardiac output and accounts for 20% of the total oxygen requirement.\textsuperscript{9} However, the brain at birth constitutes 14% of body weight\textsuperscript{11} and utilizes more than 50% of the body’s oxygen consumption.\textsuperscript{7,18} Furthermore, the neonatal cardiac output, which may be approximated at 500 cc. per minute,\textsuperscript{38} is twice as great on the basis of heart weight as that of an adult, even though the heart size at this age is larger. Although the absolute volume of arterial blood “lost” as the result of a cerebral shunt of a given size would be expected to be equal in both infant and adult, it is obvious that the relative loss is much greater for the infant. Since the size of the arteriovenous shunts under discussion are quite large (Fig. 1), and since the correlation between the size of systemic arteriovenous shunts and cardiac enlargement has been reported,\textsuperscript{20} it appears reasonable that a cause and effect relationship exists between the cerebral lesion and congestive heart failure.

**Summary**

We have reported a case of a newborn with concurrent cerebral arteriovenous malformation and congestive heart failure, and have reviewed 11 other reported cases. In each instance the patient had congestive heart failure during the first few days or weeks of life and in no case was there any evidence of other significant cerebral or cardiac lesions. We suggest that the cause of congestive failure in these infants is the high proportion of the cardiac output shunted directly from arterial to venous vessels, thereby resulting in cardiomegaly and, subsequently, heart failure. This relationship appears to be a peculiarity of the newborn because of the unique cardiovascular dynamics of the neonatal period.

**Acknowledgment**

It is with pleasure that we acknowledge the persistence of Dr. L. Scott in obtaining the diagnostic studies involved. We are grateful for permission to use his results obtained on heart catheterization and his discussion of them.

**References**

Heart Failure and Arteriovenous Malformation


