ANEURYSMAL MALFORMATION OF THE GREAT VEIN OF GALEN CAUSING HEART FAILURE IN EARLY INFANCY

Report of Five Cases

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Unlike the adult, the infant develops congestive heart failure as an important manifestation of intracranial arteriovenous aneurysm. In the past, no fewer than five infants have been reported who developed congestive heart failure as a complication of an aneurysm of the vein of Galen. A review of other reported cases of this malformation indicates that three additional infants were in heart failure, although this diagnosis was not made. A fourth patient who had an aneurysm of the vein of Galen had, in addition, transposition of the great vessels, patent foramen ovale, and ductus arteriosus. Thus, of 24 infants with aneurysm of the vein of Galen, 9 had developed congestive heart failure.

In this paper we describe five additional cases of aneurysm of the vein of Galen in which congestive heart failure occurred. This is followed by discussion of the pathogenesis of congestive heart failure and cerebral ischemic lesions, by considerations on the embryogenesis of the vascular anomaly, and by recommendations on the treatment of this lesion. In the four infants who were examined at autopsy (Cases 1 to 4), the aneurysm of the vein of Galen conformed to the following definition: a singular dilatation of the great cerebral vein with evidence of direct arterial-venous communication.

CASE REPORTS

Case 1

A male Negro infant, born prematurely at home (weight 4 lb, or 1,814 gm), was well according to the mother, until the age of 3½ months, when he developed a cold. One week after the onset of the cold he was brought to Detroit Receiving Hospital because of respiratory difficulty and cyanosis. On admission he was critically ill; the body weight was 10 lb (4,536 gm); temperature, 100.8°F (38.2°C); heart rate, 140/minute; and respiratory rate, 64/minute. Subcostal retractions were present, and moist rales could be heard throughout both lung fields. No heart murmurs were heard, and the liver was not enlarged. The head was slightly asymmetrical and measured 16 in. (41 cm). A roentgenogram of the chest demonstrated pneumonia in the left and right upper lobes. The heart was not enlarged.

His general status and the roentgenographic appearance of the chest improved following therapy with penicillin, chloramphenicol, and oxygen. However, mild dyspnea, an expiratory wheeze, cough, and scattered rales persisted up to the time when heart failure was recognized. Twelve days after admission, he had a generalized convolution. At the time his temperature was 102°F (38.8°C). On the twenty-seventh day after admission he was found to be in congestive heart failure. The heart rate was 148/minute. No murmurs or thrills were found in the precordial area, and no intracranial bruit was heard. The respiratory rate was 60/minute, and there were scattered crepitant rales to auscultation of the thorax. The liver edge was palpable 3 cm below the right costal margin. The heart was enlarged, a finding not present in the earlier roentgenogram (Fig. 1). The electro-

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cardiogram showed biventricular hypertrophy. The child was digitalized and received mercaptopurine (Thiomerin) sodium and oxygen. After approximately 3 weeks of this therapy, he improved. A grade II systolic murmur could be heard in the left third intercostal space. Examination of the cerebrospinal fluid disclosed 958 erythrocytes and 1 leukocyte per cubic millimeter, and a total protein content of 65 mg/100 ml. The blood pressure was 95/50 mm Hg. The anterior fontanelle bulged and pulsed; the veins of the scalp were dilated, and a systolic thrill and bruit were noted in the suprasternal notch and over both carotid arteries. A systolic bruit was then heard for the first time in the occipital region. A retrograde left brachial aortogram was performed, and later a retrograde right-sided brachialvertebral arteriogram was done. Both studies demonstrated enormously dilated vertebral-basilar and ascending cervical arteries (Fig. 2). The posterior cerebral arteries were dilated and communicated directly with an enormously dilated great vein of Galen (Fig. 3). The straight sinus, the caudal part of the superior sagittal sinus, and the lateral sinuses were also dilated.

He had five additional generalized seizures. Because of the rapid deterioration of the patient's condition, with signs of both cerebral circulatory impairment and cardiac failure, neurosurgical intervention was not deemed advisable. During the last 2 weeks prior to death, a loud bruit could be heard over both temporal bones and the occipital bone. The patient died 2½ months after admission, at the age of 6 months.

**Autopsy Findings:** At autopsy the dural sinuses and the great vein of Galen were greatly dilated,
ANEURYSM

the latter measuring 3.0 by 1.8 by 1.3 cm (Fig. 4). The basilar, posterior communicating, posterior cerebral, and left anterior cerebral arteries were markedly dilated. An actual arteriovenous fistula was not identified. The brain tissue adjacent to the aneurysm was slightly compressed, and the ventricular system was slightly dilated. Microscopic examination did not, however, disclose any abnormalities of brain tissue. The heart was enlarged, weighing 98 gm (normal expected weight, 27 gm). The left ventricular myocardium measured 1 cm in thickness and the right, 0.5 cm. The foramen ovale was anatomically patent. The ductus arteriosus was represented by the ligamentum arteriosum. The pulmonary artery was dilated. The branches of the arch of the aorta were also dilated, but the continuation of the arch towards the descending portion of the aorta was markedly stenosed. Microscopic examination of the lungs revealed a generalized pulmonary congestion and edema. There was no evidence of an inflammatory process.

Case 2

A male infant was admitted to the Children's Hospital of Michigan one hour after his birth at home. Gestation had lasted 7½ months. The mother had felt well until one month previously, when she complained of abdominal pain. She was a healthy woman and had borne two older children without difficulty. On admission the patient weighed 1,854 gm. His length was 41 cm, and the head circumference measured 30 cm. His face and extremities were slightly cyanotic. Respirations were shallow at a rate of 60/minute. At the age of 12 hours there were respiratory retractions of the lower chest area, and the cyanosis was more marked. A grade II systolic murmur and a gallop rhythm could be heard at the cardiac apex. The pulse was 160/minute. The liver edge could be palpated 4 or 5 cm below the right costal margin. Roentgenograms of the chest demonstrated a markedly enlarged cardiac shadow. The electrocardiogram disclosed flattened T-waves in all leads. Despite treatment with digitalis, the infant died 36 hours after birth.

Autopsy Findings: The brain was small and atrophic, weighing 130 gm (normal expected weight, 226 gm). The great vein of Galen and the straight sinus were ectatic, the former measuring 3 and the latter 2 cm in diameter (Fig. 5). The basilar and posterior cerebral arteries were dilated, but a gross arteriovenous fistula was not identified. The left cerebral hemisphere was extremely atrophic, and on both sides there were ischemic necrosis, hemorrhage, cavitation, and focal calcification (Fig. 6). The meningeal vessels over the parietal and occipital portions of the cerebrum were extremely dilated. The meningeal veins were extremely sclerotic in addition to being ectatic, there being medial hypertrophy and patchy intimal fibrosis, the latter suggesting previous thrombosis. Other findings included moderate cardiac enlargement and slight hypertrophy of both ventricles. There was slight congestion of the lungs and liver. An atypical atrial septal defect was located in the superior portion of the septum and measured 0.8 cm in diameter.

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Case 3

A male Negro infant was admitted to the Children's Hospital, at the age of one day because of cyanosis and grunting respirations since birth. His mother had had three other children; the first died on the second day of life of unknown cause, the second died at 10 years with leukemia, and the third had sickle-cell anemia. No history of pregnancy and delivery was available. On admission the infant weighed 3,360 gm. His length measured 50.8 cm, head circumference 36 cm, and chest circumference 33 cm. The anterior fontanelle was flat. He was extremely cyanotic and slightly jaundiced. His temperature was 98.6°F (37.0°C); respiratory rate, 68/minute; and radial pulse, 190/minute and of good quality. There was good respiratory exchange. A coarse blowing systolic murmur could be heard over the entire precordium and back. The lower liver edge could be palpated 5 cm below the right costal margin. The femoral pulses could not be felt.

Laboratory studies revealed a hemoglobin concentration of 13.7 gm/100 ml. The total leukocyte count was 22,000/mm³, with 58% neutrophils, 30% lymphocytes, and 2% monocytes.

Roentgenographic and fluoroscopic examination of the heart disclosed an enlarged cardiac shadow. The transverse diameter measured 8.1 cm, compared with a measurement of 10.7 cm for the internal diameter of the chest. The lower trachea and esophagus were posteriorly displaced, and there was narrowing of the thoracic portion of the trachea in its anteroposterior diameter. There was little evidence of any vascular congestion in the lungs. He was placed in an incubator and given oxygen, meralluride (Mercuhydrin), mor-
phine, digitoxin, and sulfadiazine. The infant died 18 hours after admission on the second day of life.

**Autopsy Findings:** The brain weighed 316 gm (normal expected weight 335 gm). A large saccular aneurysm of the vein of Galen was present and communicated freely with the straight sinus (Fig. 7). The aneurysm was partially filled with clotted blood. By dissection and injection of fluid a communication could be demonstrated between this aneurysm and a branch of the right posterior cerebral artery. The right occipital lobe was atrophic, and only a thin lamina of tissue surrounded the dilated ventricle. Microscopic examination of this area disclosed marked ischemic necrosis with hemorrhage and cavitation. There was also softening and hemorrhage in the right thalamus. Numerous veins in the thalamus were extremely ectatic, and medial hypertrophy of the walls was present. The meningeal veins were also greatly dilated and moderately sclerotic. The heart was dilated and hypertrophied, weighing 55 gm (normal expected weight, 17 gm). No structural abnormalities of the heart or great vessels were seen, but the myocardium had a striking yellow color suggestive of fatty infiltration. There was moderate congestion of the liver.

**Case 4**

A 2-day-old Negro boy was admitted to Children's Hospital because of difficult respirations and refusal to eat. He was the first child of healthy young parents and was born after a full-term pregnancy and 8 hours of labor. Birth weight was 3,030 gm. At birth it was necessary for him to be placed in oxygen because of difficult respiration and cyanosis. His cry was feeble. His temperature was 97° F (36.1° C); length, 47 cm; head circumference, 33 cm; and chest circumference, 30.5 cm. The anterior fontanelle was flat. He had intercostal and suprasternal retraction during inspiration. The respiratory rate was 56/minute. Breath sounds were decreased over the right lower lung field posteriorly. There were rales over the same area. A loud systolic murmur was heard over the entire precordium. The liver edge was palpable 4 cm below the right costal margin.

Laboratory studies revealed a hemoglobin concentration of 17.9 gm/100 ml. The total leucocyte count was 16,800/mm³, with 77% polymorphonuclear neutrophils, 21% lymphocytes, 1% monocytes, and 1% basophils.

Roentgenographic and fluoroscopic examination of the chest showed an enormously enlarged heart approximating the lateral chest wall on the left and almost reaching the right lateral chest wall. The transverse diameter of the heart measured 7.8 cm, compared with 9.3 cm for the internal diameter of the chest. There was no consolidation in the lungs. He was treated with oxygen, penicillin, and digitalis. He died a few hours after admission on the second day of life.

**Autopsy Finding:** The brain weighed 298 gm (expected normal weight approximately 300 gm). The vein of Galen and straight sinuses were greatly enlarged and were surrounded by a congeries of vessels which appeared to be both arterial and venous (Fig. 8). The vertebral, basilar, and posterior cerebral arteries were dilated and tortuous, and the last appeared to communicate directly with the abnormal vascular plexus. Microscopic examination of the cerebrum showed focal areas of hemorrhage, ischemic necrosis, and calcification. The heart weighed 40 gm (expected normal weight, approximately 16 gm), and there was marked dilatation of the right atrium and ventricle. Scattered areas of the myocardium had a yellow color, indicating fatty infiltration. A mild coarctation of the aorta was present just distal to the ductus arteriosus, but no intracardiac defects were observed. An incidental finding was severe hypoplasia of the left kidney, which was grossly deformed and contained cystic and dysplastic structures.

**Case 5**

A white female infant was found to have a harsh systolic murmur during a routine physical examination in the third day of her life. On admission to Henry Ford Hospital she was found to be in heart failure, her respirations were 45/minute; she had no cyanosis. The liver was en-
larged; its lower edge was palpable 2 cm below the right costal margin. A loud systolic murmur was heard over the precordium. The head was not enlarged. Fluoroscopy disclosed cardiomegaly. The electrocardiogram suggested right ventricular hypertrophy. She was treated with mercaptopurine and digitoxin. Cardiac catheterization demonstrated the following pressures and oxygen saturation values:

<table>
<thead>
<tr>
<th>Pressure (cm H₂O) (mm Hg)</th>
<th>Saturation (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Inf. vena cava</td>
<td>10.2</td>
</tr>
<tr>
<td>Sup. vena cava</td>
<td>13.3</td>
</tr>
<tr>
<td>Right atrium</td>
<td>19</td>
</tr>
<tr>
<td>10</td>
<td>15.5</td>
</tr>
<tr>
<td>Right ventricle</td>
<td>48</td>
</tr>
<tr>
<td></td>
<td>15.4</td>
</tr>
</tbody>
</table>

(Oxygen capacity 19.4 vol/100 ml).

Cineangiograms showed rapid disappearance of the dye from the pulmonary artery into the lung and rapid return to the left atrium. The ascending aorta and aortic arch filled well, and the innominate artery was extremely tortuous. Immediately following disappearance of the dye from the aorta there was opacification of the superior vena cava and right atrium.

As a result of these findings the head was auscultated, and a loud, blowing, continuous bruit was heard over the right temporal region. She remained in heart failure. Cerebral arteriograms demonstrated a large arteriovenous malformation with a saccular dilatation of the vein of Galen (Fig. 9).

A few days later the right common carotid artery was ligated. The bruit over the left temporal area decreased in intensity. The patient remained in congestive heart failure for a few days, but gradually improved. The left external carotid artery was then ligated, and she was discharged very much improved and out of failure.

**COMMENT**

It is well established that arteriovenous fistulas can produce an increase in cardiac output and cardiac work, which may result in cardiac dilatation, hypertrophy, and decompensation. Reid and Holman produced cardiac hypertrophy in dogs by means of artificial arteriovenous fistulas, and in some instances cardiac decompensation ensued. Laplace reported enormous cardiac enlargement secondary to a traumatic fistula between the left femoral artery and vein. After surgical correction of the fistula, the transverse diameter of the heart was reduced by 45%.

According to Dandy, Steinheil in 1895 reported for the first time an intracranial arteriovenous aneurysm found at autopsy. The great vein of Galen drained the anomalous dilatation. The patient, who died at the age of 49 years, also had a slight hypertrophy of the right side of the heart. Dandy found no instances of cardiac hypertrophy in eight patients with intracranial arteriovenous aneurysms, but cited that Emanuel, Isenschmid, and Laves had separately reported such cases. Ray found that two of five adults and one 3-year-old child with cerebral arteriovenous aneurysms had electrocardiographic evidence of left ventricular preponderance and roentgenographic signs of left ventricular hypertrophy. Silverman et al., who described two infants with cerebral arteriovenous fistulas and fatal congestive heart failure first pro-
posed that these anomalies could cause heart failure in the newborn period.

Hook et al.20 investigated the cardiac function of 14 adult patients with arteriovenous aneurysm and reviewed the histories of another 133 cases, including 10 with carotid cavernous fistula. These authors found no “symptoms indicating an increased load of the heart and circulation that could be regarded as caused by the arteriovenous aneurysm.” It is interesting that the youngest of the 14 fully investigated patients was 16 years old.

Analysis of the cases of aneurysms of the great vein of Galen (Tables I, II, III) reveals a striking relationship between the age of the patients and the occurrence of congestive heart failure. All 14 patients who developed this complication were less than 4 months old at the onset of symptoms (Table I) and 11 were less than 5 days old. Congestive heart failure was not reported

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**TABLE I**

**Patients with Aneurysm of the Great Vein of Galen Who Developed Symptoms of Heart Failure**

<table>
<thead>
<tr>
<th>Source</th>
<th>Age at Onset of Symptoms of Failure</th>
<th>Cardiac Findings</th>
<th>Outcome</th>
<th>Autopsy Findings*</th>
</tr>
</thead>
<tbody>
<tr>
<td>Clement et al.,* (Case 1)</td>
<td>4 wk (approx.)</td>
<td>.</td>
<td>Died, 38 da</td>
<td>Heart normal size</td>
</tr>
<tr>
<td>Clement et al.,* (Case 2)</td>
<td>7 da</td>
<td>Heart radiogr. enlarged</td>
<td>Died, 8 mo</td>
<td>Heart weight 130 gm (55 gm); hypertrophy of right ventricle</td>
</tr>
<tr>
<td>Pollock &amp; Laslett¹</td>
<td>51 hr</td>
<td>Heart radiogr. enlarged</td>
<td>Died, 83 hr</td>
<td>Cardiac hypertrophy and dilatation</td>
</tr>
<tr>
<td>Corrin,² (Case 1)</td>
<td>42 hr</td>
<td>.</td>
<td>Died, 4 da</td>
<td>Heart weight 41 gm (18 gm); biventricular hypertrophy and dilatation</td>
</tr>
<tr>
<td>Claireaux &amp; Newman,² (Case 1)</td>
<td>16 hr</td>
<td>Soft syst. murmur</td>
<td>Died, 4 da</td>
<td>Dilatation of right atrium and ventricle</td>
</tr>
<tr>
<td>Claireaux &amp; Newman,² (Case 2)</td>
<td>1 wk</td>
<td>Heart radiogr. enlarged</td>
<td>Died, 16 da</td>
<td>Hypertension and dilatation of right atrium and ventricle</td>
</tr>
<tr>
<td>Glatt &amp; Rowe,² (Case 1)</td>
<td>3½ hr</td>
<td>.</td>
<td>Died, 5 mo</td>
<td>Heart weight 39.9 gm (29 gm); right ventricle dilated, wall measured 6 mm in thickness, left ventricle 4 mm</td>
</tr>
<tr>
<td>Glatt &amp; Rowe,² (Case 2)</td>
<td>43 hr</td>
<td>Heart radiogr. enlarged</td>
<td>Died, 70 hr</td>
<td>Heart weight 26.5 gm (17 gm)</td>
</tr>
<tr>
<td>Hirano &amp; Solomon*</td>
<td>Birth</td>
<td>.</td>
<td>Died, 7 wk</td>
<td>Transposition of great vessels, patent ductus, &amp; foramen ovale</td>
</tr>
<tr>
<td>Present series, (Case I)</td>
<td>4 mo</td>
<td>Heart radiogr. enlarged</td>
<td>Died, 19 wk</td>
<td>Heart weight 98 gm (29 gm); stenosis of descending aorta</td>
</tr>
<tr>
<td>Present series (Case 2)</td>
<td>12 hr</td>
<td>Heart radiogr. enlarged</td>
<td>Died, 36 hr</td>
<td>Right atrium and ventricle moderately dilated; right ventricle wall 2 mm, left 4 mm in thickness</td>
</tr>
<tr>
<td>Present series, (Case 3)</td>
<td>Birth</td>
<td>Heart radiogr. enlarged</td>
<td>Died, 2 da</td>
<td>Heart weight 55 gm (17 gm); biventricular dilatation &amp; hypertension, right ventricle wall 5 mm; left 5 mm in thickness</td>
</tr>
<tr>
<td>Present series, (Case 4)</td>
<td>Birth</td>
<td>Loud syst. murmur, heart radiogr. enlarged</td>
<td>Died, 2 da</td>
<td>Heart weight 40 gm (17 gm); hypertension of right atrium; coarctation of aorta</td>
</tr>
<tr>
<td>Present series, (Case 5)</td>
<td>3 da</td>
<td>Cardiomegaly by fluoroscopy</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

* Figures in parentheses are the normal weights of the heart for that age.4
ARTICLES

TABLE II

INFANTS WITH ANEURYSM OF THE GREAT VEIN OF GALEN IN WHOM HEART FAILURE WAS NOT DISCOVERED

<table>
<thead>
<tr>
<th>Source</th>
<th>Symptoms</th>
<th>Age of Onset</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Wohak⁴</td>
<td>Birth</td>
<td>Died, 12 hr</td>
<td></td>
</tr>
<tr>
<td>Jaeger &amp; Forbes⁵</td>
<td>Epistaxis, fainting spells, &amp; hydrocephalus</td>
<td>8 mo after “flu” at 8 mo</td>
<td>Carotid ligation; died, 4 yr</td>
</tr>
<tr>
<td>Russell &amp; Nevin⁶</td>
<td>Prominent face &amp; neck veins</td>
<td>3 mo</td>
<td>Died, 17 mo</td>
</tr>
<tr>
<td>Turhan &amp; Rossler¹¹</td>
<td>Hydrocephalus</td>
<td>2 mo</td>
<td>Died, 17 mo</td>
</tr>
<tr>
<td>Lumsdon¹²</td>
<td>Convulsions; intraventricular hemorrhage</td>
<td>4 mo</td>
<td>Died, 7 mo</td>
</tr>
<tr>
<td>Boldrey &amp; Miller¹⁶</td>
<td>Hydrocephalus; subarachnoid hemorrhage</td>
<td>8 mo</td>
<td>Carotid ligation; clipping of feeding artery</td>
</tr>
<tr>
<td>Boldrey &amp; Miller¹⁶</td>
<td>Swollen face &amp; eye veins</td>
<td>Birth</td>
<td>Carotid ligation</td>
</tr>
<tr>
<td>Petit-Dutaillis et al.¹⁷</td>
<td>Hydrocephalus</td>
<td>4 mo</td>
<td>Alive</td>
</tr>
<tr>
<td>Hirano &amp; Terry¹⁹</td>
<td>Hydrocephalus</td>
<td>5 mo</td>
<td>Died, 18 mo</td>
</tr>
<tr>
<td>Gibson et al.²⁰</td>
<td>Hydrocephalus</td>
<td>6 mo</td>
<td>Died, 4 ½ yr</td>
</tr>
<tr>
<td>Litvak et al.¹¹ (Case 1)</td>
<td></td>
<td>4 mo</td>
<td>Clipping of feeding artery; ventriculopleural shunt</td>
</tr>
<tr>
<td>Litvak et al.¹¹ (Case 2)</td>
<td>Large head; unable to sit alone;</td>
<td>3½ mo</td>
<td>Died, 1½ mo</td>
</tr>
<tr>
<td>Schwartz²²</td>
<td>Birth</td>
<td>Died, 4 da</td>
<td></td>
</tr>
</tbody>
</table>

in 24 patients (Tables II & III). The actual incidence of cardiac hypertrophy or dilatation in these two groups is not known.

Heart failure might have been caused by structural defects in the heart or aorta in three of the five cases cited here, and in one described in the literature.⁶ Since one of our four patients had no cardiac or vascular defect and the fifth patient improved after carotid ligation, the cerebral arteriovenous fistula alone could be a cause of heart failure. Furthermore, in the three cases with associated structural anomalies of the heart or aorta, the clinical and pathological evidence led us to conclude that the cerebral arteriovenous shunt was the principal cause of heart failure.

Physiopathogenesis of Heart Failure

In 1948, Shenkin and associates³¹ studied cerebral blood flow and metabolism by the nitrous oxide technique, together with the cardiac output, total blood volumes, and mean arterial pressure in two adult cases

TABLE III

ANEURYSM OF THE GREAT VEIN OF GALEN WITH ONSET OF SYMPTOMS AFTER INFANCY

<table>
<thead>
<tr>
<th>Source</th>
<th>Symptoms</th>
<th>Age of Onset</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Steinheil⁴</td>
<td>Convulsions</td>
<td>Child</td>
<td>Died, 49 yr</td>
</tr>
<tr>
<td>Alpers &amp; Foster¹²</td>
<td>Mental retardation; hydrocephalus</td>
<td>6 yr</td>
<td>Died, 18 yr</td>
</tr>
<tr>
<td>Wolfe &amp; Franev⁴</td>
<td>Convulsions; headaches</td>
<td>3½ yr</td>
<td>Torkildsen’s operation; died</td>
</tr>
<tr>
<td>French &amp; Peyton¹⁸</td>
<td>Hydrocephalus</td>
<td>..</td>
<td>Alive at 11 mo</td>
</tr>
<tr>
<td>French &amp; Peyton¹⁸</td>
<td>Subarachnoid hemorrhage</td>
<td>..</td>
<td>Alive at 12 yr</td>
</tr>
<tr>
<td>French &amp; Peyton¹⁸</td>
<td>Headache</td>
<td>..</td>
<td>Alive at 12 yr</td>
</tr>
<tr>
<td>French &amp; Peyton¹⁸</td>
<td>Headache</td>
<td>..</td>
<td>Alive at 14 yr</td>
</tr>
<tr>
<td>French &amp; Peyton¹⁸</td>
<td>Headache</td>
<td>..</td>
<td>Alive at 14 yr</td>
</tr>
<tr>
<td>Cohen¹³</td>
<td>Headache; coma; vomiting, after hemiparesis</td>
<td>52 yr</td>
<td>Died, 10 days</td>
</tr>
</tbody>
</table>
of cerebral arteriovenous malformation. The patients were 43 and 28 years old. In both, cerebral blood flow was enormously increased. In one cerebral blood flow was 143 ml/100 gm of brain/minute, and in the other it was 185 ml/100 gm/minute (normal, 54-65 ml/100 gm/min.). Blood flow through the arteriovenous malformations was 1,250 to 1,830 ml of blood per minute. Cardiac output was increased in both cases, 20 to 39% while horizontal and 61 to 122% when standing. The mean arterial blood pressure was reduced, and the hearts were enlarged in both patients. Plasma volume was normal in one and slightly increased in the other. Other investigations have shown greatly increased cerebral blood flow and decreased circulation times in cerebral arteriovenous malformations.\textsuperscript{32-37} The abnormal arteriovenous malformation in physiological terms provides an arteriovenous fistula or shunt within the cranium. Cranial\textsuperscript{*} blood flow is greatly accelerated and increased because of decreased vascular resistance within the arteriovenous malformation.

Epstein et al.\textsuperscript{38} found that the closure of arteriovenous fistulas raised the peripheral resistance, and they inferred that the lowered peripheral resistance produced by a fistula would increase cardiac output by the following three factors acting singly or jointly: (1) a carotid sinus reflex producing an increase in heart rate; (2) a decrease in the pressure in the aorta against which the left ventricle must work; and (3) an increase in the inflow of blood to the right heart resulting in increased distension of the right atrium and ventricle during diastole. The third factor in our judgment appears to be the major determinant in the young infant. This view is supported by pathological findings in six cases of aneurysm of the great vein of Galen with heart failure (Table I, Cases 2, 5, 6, 7, 11, & 13). In those patients the cardiac enlargement was entirely the result of dilatation and/or hypertrophy of the right atrium and/or right ventricle. One feature of the normal fetal circulation is important in this regard. During the fetal state superior vena caval blood is felt to be preferentially directed to the right ventricle. The increase in blood flow through the cerebral circulation due to lowered resistance results in an increased amount of blood entering the superior vena cava which to a large extent enters the right ventricle causing increased diastolic filling. Although at any age, the size of the arteriovenous malformation, and hence the magnitude of the shunt, plays the major role in determining the degree of stress upon the heart, in the newborn period the adjustments in the circulatory systems which occur as the newborn adapts to extrauterine life are important. Glatt and Rowe\textsuperscript{3} have commented on Adams' finding that the cardiac output of the newborn is probably three times higher than during fetal period, and that the newborn's heart cannot withstand a further significant increase in cardiac output without decompensating.

Massive intracranial arteriovenous malformations, such as were present in our five patients, are incompatible with life for two reasons. (1) The massive increase in cardiac output leads to cardiac failure. (2) Arteriovenous malformations of the brain that are of sufficient size to cause heart failure are frequently associated with brain injury due to ischemia or hemorrhage. Death may occur from either cause or a combination of the two.

Thus, it would appear that individuals with an aneurysm sufficiently large to produce heart failure do not live to adult age. In four cases reported here, death was primarily due to heart failure, although cerebral damage also played a part.

**Mechanism of Brain Injury**

The liability of arteriovenous malformations to rupture and to cause brain damage is well known, but the encephalopathy in the cases presented here was due to

\* We use here the term "cranial" and not "cerebral" because the site of the malformation is actually outside the cerebral tissue.
ischemic anoxia. In three cases, the lesion was spongy necrosis of white matter in areas that presumably had diminished blood supply. There are five explanations for ischemic anoxia due to massive arteriovenous malformations. Arteriovenous malformations or arteriovenous communications, no matter what their cause,\textsuperscript{39} tend to shunt blood away from surrounding tissue since the peripheral vascular resistance is negligible in the malformation and blood tends to flow from high to lower pressure areas.\textsuperscript{40} A second factor is the development of heart failure, causing reduction of the cardiac output and hence further impairment of the cerebral circulation.\textsuperscript{41,42} A third factor, which did not appear to play an important part in the cases described here, is thrombosis of the abnormal vessels. A fourth one is the pressure caused by the aneurysmal sac on the neighboring structures of the brain. Finally, if the aqueduct of Sylvius is obliterated by this pressure, there is in addition production of internal hydrocephalus.

**Embryological Considerations**

Scharrer\textsuperscript{43} and Padget,\textsuperscript{44} in their studies on the development of veins in the cerebral venous system in mammals and in man, showed that the primitive arteries and veins are formed on the surface of the neural tube by two separate precapillary plexuses. The veins do not accompany the arteries. In the embryo, the thin walled pia-arachnoidal arteries (tubes of endothelium) are superficial to the pial veins and cross each other perpendicularly, sometimes lying in close approximation. The arteries are more numerous and anastamose more freely than the veins. The veins have more plentiful tributaries. With the growth of the forebrain, the distance increases between the medially located veins and the lateral venous channels which are the forerunners of the dural sinuses. The Galenic venous system is formed by a number of primitive veins of metencephalic, diencephalic, and telencephalic origin. The internal cerebral vein originally located in contact with the diencephalic roof receives the superior choroidal vein, draining the relatively large choroidal plexus. The basal cerebral vein receives telencephalic and diencephalic tributaries and joins the internal cerebral vein to form the great vein of Galen. It would appear that the close contact of the primitive veins with the choroidal, callosal, posterior cerebral, and superior cerebellar arteries is an important factor in the production of arteriovenous fistulae in this region.

**Diagnostic Considerations**

From a diagnostic standpoint, an intracranial arteriovenous fistula must be considered in the differential diagnosis of an infant with congestive heart failure. Auscultation of a cranial bruit might suggest the presence of an intracranial aneurysm but several factors must be kept in mind. First, intracranial bruits are not uncommon in congenital heart disease without intracranial vascular anomalies. Secondly, in some normal infants a physiologic bruit is heard over the anterior fontanelle. Finally, our experience with Case 1 confirms the observation of Claireaux et al.\textsuperscript{2} and Clement et al.\textsuperscript{4} that an early cranial bruit may be faint or absent in early age and apparent later on.

The only way to confirm the diagnosis during life is by arteriography. Since the vertebral-basilar system supplies these anomalies, either retrograde brachialvertebral arteriography or retrograde aortography is indicated. The roentgenographic exposure should include the skull as well as the chest. Since this appears to be a more common anomaly than previously suspected and is surgically remediable in some cases, we would advise such arteriographic investigation in cardiac enlargement with or without congestive failure in infants and children who show increased cardiac output and rapid cerebral circulation times.

**Treatment**

When diagnosis is established early, before the onset of heart failure, the arteriovenous malformation could be resected by
an intracranial approach.* If the patient is in heart failure he should be treated by digitalis, oxygen, and diuretics. As soon as possible the feeding arteries in the neck should be ligated. This will rapidly decrease the cardiac output and the work of the heart. Intracranial intervention is not recommended in the presence of heart failure because operative interference of this magnitude would almost certainly prove fatal.

SUMMARY

Five cases of heart failure, resulting from a cerebral arteriovenous fistula and aneurysm of the great vein of Galen, are reported. In two infants the diagnosis was made roentgenographically; one improved after arterial ligation. In four cases, autopsy demonstrated the anomalous vessels and dilatation or hypertrophy of the heart. In three cases there was spongy necrosis of the brain. The pathogenesis of heart failure seems to be related to an enormous increase of the intake of the right atrium and ventricle. The cerebral damage seems to be due to ischemia caused by shunting of blood toward the veins bypassing the capillary network. Early diagnosis can be made by auscultation of the head and cerebral arteriography in infants who develop heart failure. Treatment is discussed.

REFERENCES


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