

Arteriovenous Malformation of the Vein of Galen

Treatment in a Neonate

David G. Watson, MD; Robert R. Smith, MD;
Alfred W. Brann, Jr, MD

• Heart failure in a 2-day-old infant was not readily explained by clinical examination. Cardiac catheterization suggested an intracranial arteriovenous (AV) fistula, and cerebral arteriography showed a malformation of the vein of Galen. The major feeding arteries were surgically obliterated. At age 27 months, the boy has normal mentation but moderate left hemiparesis. Review of the literature disclosed 39 other infants with AV malformations of the vein of Galen producing heart failure before age 3 months. Most of them were boys, and had cyanosis, a systolic murmur, cranial bruit, cardiomegaly, and right ventricular hypertrophy. Only three of the 13 who had surgery for their malformation survived. Removal of the malformation is difficult; obliteration of the nutrient vessels, using the operating microscope, is the currently accepted treatment.

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There have only been a small number of reports of infants with heart failure due to an arteriovenous (AV) malformation of the vein of Galen, and successful surgical treatment is exceptional.

In 1952, Pétranca¹ described an infant with such a malformation. Review of a subsequent description² of this patient indicates that she had heart failure. In 1955, Silverman et al³ reported two infants with fatal congestive heart failure resulting from intracranial AV fistulas. In 1964, Gold et al⁴ published a comprehensive review of the vein of Galen malformation, noting the differing presentation at various age periods. Symptomatic patients in the neonatal period tend to have cyanosis or dyspnea or both, and heart failure. The latter often does not respond to digitalis therapy. Older infants usually have hydrocephalus or convulsions or both as presenting symptoms; while the symptomatic older child, adolescent, or adult has headaches or indications of subarachnoid hemorrhage. There are excellent recent reviews of intra-

cranial AV malformations causing heart failure or hydrocephalus in infancy.⁵⁻⁷ Less than half were diagnosed during life; very few of these patients are living and well.

REPORT OF A CASE

A full-term male infant weighing 3.4 kg was born March 27, 1972, at Jefferson Davis Hospital, Natchez, Miss. Status at birth was normal but he fed poorly, and by age 2 days was listless, hypothermic, and tachypneic (respirations, 100/min). A chest x-ray film showed cardiomegaly. He was referred to the University Medical Center. On admission he was alert but tachypneic, with good peripheral pulses and no cyanosis or edema. There was a grade 1-2/6 medium-pitched, midsystolic murmur along the left sternal border. There was a gallop rhythm, and the liver was 4 cm below the right costal margin. The electrocardiogram showed a normal axis, with right atrial and right ventricular hypertrophy and abnormal T waves. The chest x-ray film showed cardiomegaly with increased pulmonary vascularity (Fig 1). After digitalization, the PR interval and heart rate varied. He developed 1+ pedal edema and received furosemide with added potassium. Cardiac catheterization was done at 7 days of age (Table 1). The right ventricular angiogram was not diagnostic, but the left ventricular angiogram showed contrast material returning very rapidly to the superior vena cava. Subsequent blood sampling of the superior vena cava showed hyperoxygenation. At this time, auscultation of the head revealed a cranial bruit. Cerebral arteriograms

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From the departments of pediatrics (Drs Watson and Brann) and neurosurgery (Dr Smith), University of Mississippi School of Medicine, Jackson.

Reprint requests to Department of Pediatrics, University Medical Center, 2500 N State St, Jackson, MS 39216 (Dr Watson).

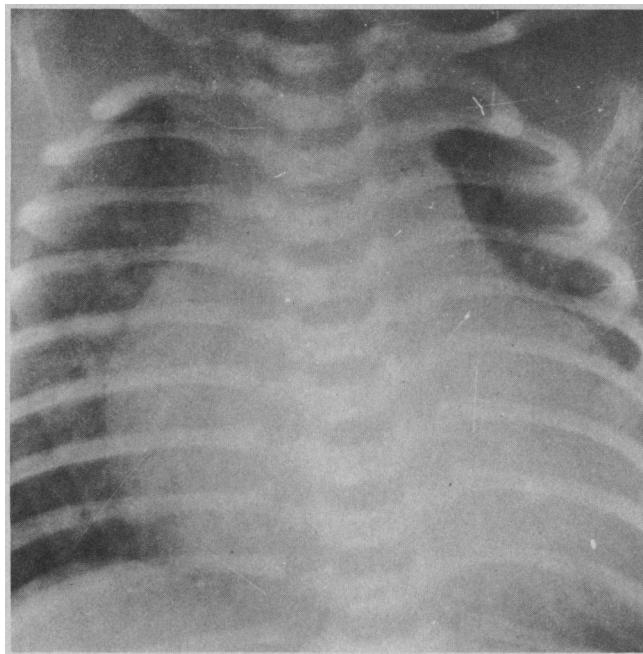


Fig 1.—Initial chest x-ray film showing gross cardiomegaly.

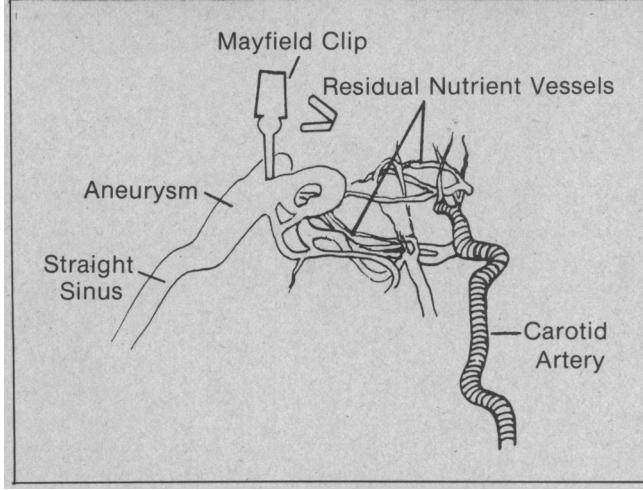
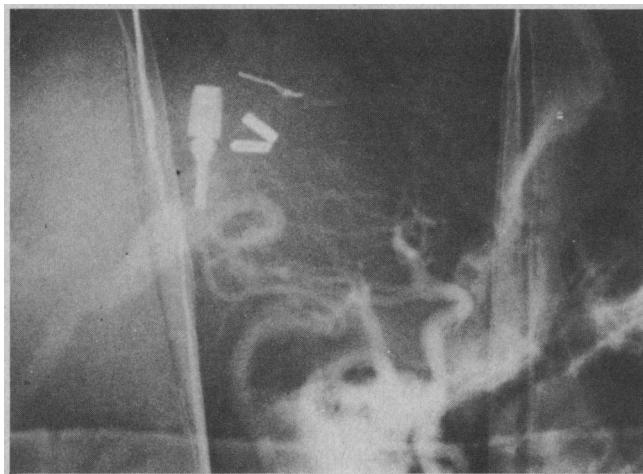


Fig 2.—Top, Arteriogram. Bottom, Explanatory line drawing. Large aneurysm of vein of Galen is fed primarily through posterior choroidal arteries.

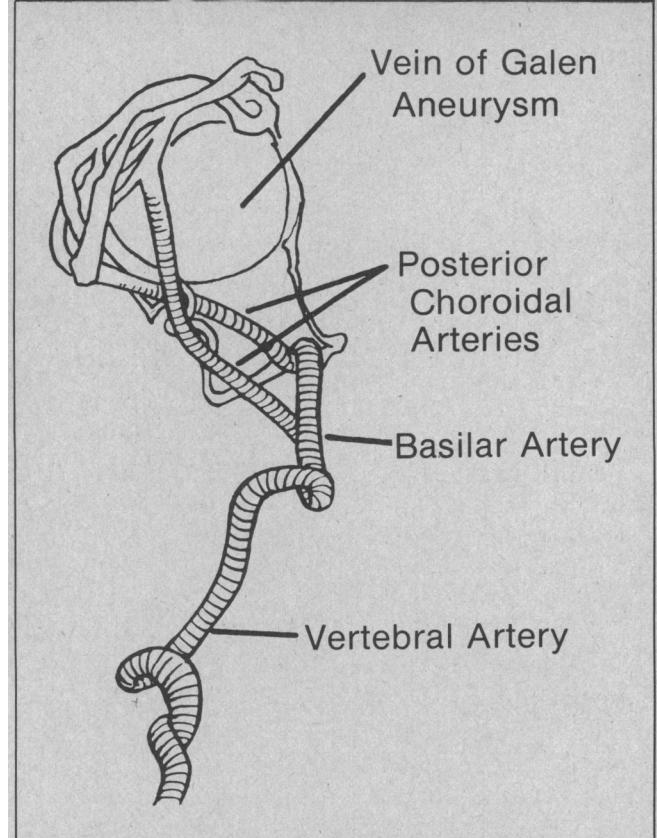
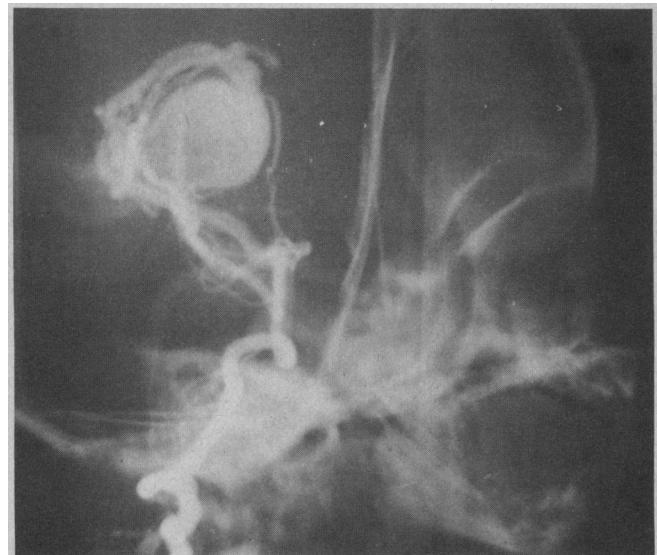


Fig 3.—Top, Arteriogram. Bottom, Explanatory line drawing. Postoperative arteriogram demonstrates absence of aneurysmal dilation, but continued early filling of vein of Galen and straight sinus.

Table 1.—Catheterization Data

	Age 7 Days		Age 24 Days		Age 11 Weeks	
	Oxygen Saturation, %	Pressure, mm Hg	Oxygen Saturation, %	Pressure, mm Hg	Oxygen Saturation, %	Pressure, mm Hg
Superior vena cava	95	71	...
Inferior vena cava	60	68	...
Right atrium	86	4 (mean)	70	0 (mean)
Right ventricle	84	105/8	71	26/-2
Pulmonary artery	89	85/30	72	24/4
Left atrium	99	7 (mean)	99	-2 (mean)
Left ventricle	99	80/5	100	80/10
Brachial artery	85	85/35

showed an AV malformation of the vein of Galen (Fig 2). On April 5, two days later, a right craniotomy was performed, and with the use of an operating microscope, anterior cerebral and posterior choroidal arteries entering the aneurysm were coagulated or clipped.

Prior to and temporarily following this surgery, seizure activity was noted. This was primarily manifested by opisthotonic posturing and sudden, unexplained outbursts of crying and nystagmus. An electroencephalogram showed slightly reduced voltage over the right cerebral hemisphere. Although the heart failure improved, the systolic murmur and diastolic gallop persisted. Repeat cerebral arteriography dem-

Table 2.—Review of the Literature*

Source	Age at Onset of Heart Failure	Sex	Birth Weight, kg	Cra- nial Bruit	Cya- nosis	Heart Sounds	Cath- eter	Angi- ogra- phy	Diag- nosis Dur- ing Life	Sur- gery for Mal- forma- tion	Result	Associated Defects
Clement et al ²	1 mo	F	3.2	...	+	No murmur	—	—	—	—	Died	Ventricular septal defect
Fisher & Johnson ⁸	5 days	F	3.1	...	+	Systolic murmur	—	+	+	+	Died	Patent ductus arteriosus
Pollock & Laslett ⁹	2 days	M	3.6	...	—	Gallop	—	—	—	—	Died	None
Gagnon & Boileau ¹⁰	2 days	M	3.5	...	+	...	—	—	—	—	Died	Patent ductus arteriosus
Glatt & Rowe, ¹¹ Hope & Izukawa ²⁹	1 day	M	2.8	...	—	Systolic murmur	—	+	—	—	Died	Patent ductus arteriosus
Glatt & Rowe, ¹¹ Hope & Izukawa ²⁹	1 day	M	4.4	—	+	Gallop	—	—	—	—	Died	Patent ductus arteriosus
Claireaux & Newman ¹²	1 day	M	2.2	...	+	Systolic murmur	—	—	—	—	Died	Patent ductus arteriosus, extravascular defect
Claireaux & Newman ¹²	1 day	M	3.7	...	+	Systolic murmur	—	—	—	—	Died	Patent ductus arteriosus
Hirano & Solomon ¹³	6 wk	M	3.9	...	4+	Systolic murmur	—	—	—	—	Died	Transposition of great arteries, patent ductus arteriosus
Levine et al, ¹⁴ Gold et al ⁴	1 day	M	3.4	...	+	Systolic murmur	—	—	—	—	Died	Patent ductus arteriosus, extravascular defect
Gomez et al ¹⁵	1 day	M	1.9	...	+	Systolic murmur, gallop	—	—	—	—	Died	Atrial septal defect
Gomez et al ¹⁵	1 day	M	3.4	...	3+	Systolic murmur	—	—	—	—	Died	None
Gomez et al ¹⁵	Birth	M	3.0	...	+	Systolic murmur	—	—	—	—	Died	Coarctation of the aorta, extravascular defect
Gomez et al ¹⁵	3 days	F	...	+	—	Systolic murmur	+	+	+	+	Improved	...
Falcone et al ¹⁶	1 day	F	2.8	+	+	Systolic murmur	+	+	+	—	Died	Extravascular defect
Ford ¹⁷	10 days	M	+	Systolic murmur	+	+	+	+	Died	...
Sütterle ¹⁸	2 days	M	3.8	...	+	Systolic murmur	—	—	—	—	Died	None
Carroll & Jakoby ¹⁹	4 days	F	4.0	+	+	No murmur	—	—	—	—	Died	Patent ductus arteriosus, coarctation of the aorta
Brown & Alexander, ²⁰ Stern et al ²¹	2 days	F	2.6	...	+	No murmur	—	—	—	—	Died	Patent ductus arteriosus
Brown & Alexander ²⁰	Weeks	M	...	+	—	+	+	+	Died	...
Noetzel & Zorger ²²	1 day	M	3.2	...	+	...	—	—	—	—	Died	...
Mennicken & Rau ²³	8 days	F	+	Systolic murmur, gallop	+	+	+	—	Died	None

Table 2.—Review of the Literature* (Cont)

Source	Age at Onset of Heart Failure	Sex	Birth Weight, kg	Cranial Bruit	Cyanosis	Heart Sounds	Catheter	Angiography	Diagnosis During Life	Surgery for Malformation	Result	Associated Defects
Levy et al ²⁴	1 day	M	4.9	+	+	Diastolic murmur, systolic murmur	+	+	+	+	Well	...
Lagos & Riley ²⁵	1 mo	M	—	—	+	—	Hydrocephalic	...
Sunderland ²⁶	1 day	M	4.5	+	+	Systolic murmur	+	+	+	+	Well	...
Holden et al ⁶	1 day	F	2.3	+	+	+	+	Died	...
Holden et al ⁶	3 days	M	—	+	+	+	Hydrocephalic	...
Holden et al ⁶	1 day	M	3.4	+	+	+	+	Died	...
Boynton & Morgan ²⁷	1 day	F	3.8	+	+	Systolic murmur	+	+	+	+	Died	Patent ductus arteriosus
Stehbens et al ²⁸	4 days	F	3.6	—	+	Gallop	+	+	+	+	Died	Patent ductus arteriosus
Hope & Izukawa, ²⁹ Norman & Becker ³⁰	1 day	M	4.4	+	+	Systolic murmur	+	+	+	+	Died	Patent ductus arteriosus
Hope & Izukawa, ²⁹ Norman & Becker ³⁰	4 days	F	...	+	+	Systolic murmur	+	+	+	—	Died	Patent ductus arteriosus
Hope & Izukawa, ²⁹ Norman & Becker ³⁰	3 days	M	...	+	+	Systolic murmur	+	+	+	+	Died	Patent ductus arteriosus
Hope & Izukawa, ²⁹ Norman & Becker ³⁰	1 day	M	...	+	+	Systolic murmur	+	+	+	—	Died	Patent ductus arteriosus
Hope & Izukawa, ²⁹ Norman & Becker ³⁰	1 day	M	...	+	+	Systolic murmur	+	+	+	—	Died	Patent ductus arteriosus
Norman & Becker ³⁰	1 day	M	+	Systolic murmur	—	—	—	—	Died	...
Long et al ³¹	1 day	—	+	+	+	Died	...
Cunliffe ³²	9 days	F	2.6	+	+	Systolic murmur	—	+	+	—	Died	None
Cunliffe ³²	2 days	M	3.5	+	—	...	+	—	+	—	Improved	None
Present case	2 days	M	3.4	+	—	Systolic murmur, gallop	+	+	+	+	Well	None

* Where two references are listed, the case has been reported twice.

onstrated a large left posterior choroidal artery still feeding the now smaller vein of Galen aneurysm. On April 26, this artery was clipped. Subsequently, the central venous pressure fell from 9.5 to 4.0 cm H₂O, and the jugular venous oxygenation was reduced. Seizures temporarily followed this surgery too.

The tachypnea and hepatomegaly diminished. The infant was discharged at 2 months of age on a regimen of phenobarbital. He was readmitted 19 days later, June 15, for repeat cardiac catheterization (Table 1) and cerebral arteriography (Fig 3). The hyperoxygenation of the superior vena caval blood and the pulmonary hypertension were no longer present. The vein of Galen aneurysm was much smaller, but still filled earlier than normal.

Examination at 9 months of age revealed a mild left hemiparesis and ankle clonus. Neither a cardiac murmur nor an intracranial bruit were heard. By 15 months of

age, the left hemiparesis was clearly evident. Cardiovascular examination, including ECG and chest x-ray film, was entirely normal. At age 27 months, the left hemiparesis was moderately severe, manifest more in the leg than arm. His personal-social adaptation and language development were progressing normally. He had just begun to walk alone. He could make two- to three-word sentences, point to five body parts, and follow verbal commands.

REVIEW OF THE LITERATURE

In Table 2, this case, together with all other reported cases of malformation of the vein of Galen with heart failure developing under 3 months of age,^{2,4,6-32} are summarized. The patients were considered to have heart failure if the authors so stated, or if the case report contained sufficient

evidence thereof. Only those cases in which the vein of Galen was the primary drainage of the AV malformation are included.

Most patients were boys (27 of 39). The birth weight ranged from 1.9 to 4.9 kg, and averaged 3.4 kg. Heart failure was usually noted in the first week of life (33 of 40); in 20 it was present by age 1 day. Twenty-nine had cyanosis, often slight. Cutaneous abnormalities (hemangiomas and telangiectases) have been noted.^{6,27} The scalp veins may be dilated. In the 31 patients in whom cardiac auscultation was described, 25 had a systolic murmur and six had a gallop. Cranial auscultation revealed a bruit in 16 of 18 patients. Several had strong peripheral pulses or a wide pulse pressure; a few had unremarkable or even

diminished peripheral pulses. In all 31 cases with mention of the chest x-ray film, the heart was enlarged, often to considerable degree. The pulmonary vascularity was said to be increased or congested in 11, and normal in five. Of the 25 cases in which an ECG was described, there were 12 patients with right atrial hypertrophy, one with left atrial hypertrophy, two with combined atrial hypertrophy, 16 with right ventricular hypertrophy, one with left ventricular hypertrophy, two with combined ventricular hypertrophy, and six with abnormal ST segments or T waves.

Although 18 patients had cardiac catheterization and 23 had angiography, only 16 of the 40 were diagnosed during life. Contrast medium injected into the left side of the heart or proximal aorta showed enlarged arteries supplying the head and a rapid return of contrast to the superior vena cava and right atrium. Blood samples from the superior vena cava were highly oxygenated. As in other large systemic AV fistulas, there was increased systemic and pulmonary blood flow, right ventricular and pulmonary hypertension, and sometimes a right-to-left atrial shunt.

Of 14 patients who had surgery, only four (including our patient) survive, one with hydrocephalus. Only three patients survived without surgery, one with hydrocephalus. Sixteen of the 33 patients who died were stated to have had a patent ductus. Because of the early age of death, this frequently noted patency of the ductus arteriosus may not be significant. However, all but one of these patients were cyanotic, and arterial oxygen desaturation in the neonatal period tends to keep the ductus patent. There were six patients with other cardiac defects: two with atrial septal defects, one with ventricular septal defect, two with mild coarctation of the aorta, and one with transposition of the great arteries. Coarctation of the aorta frequently accompanies defects with increased flow through the pulmonary artery and ductus arteriosus during fetal life. These infants would have such an increased flow because of increased superior vena caval blood return.

Otherwise, no causal relationship or common etiologic factors between the vein of Galen malformation and other congenital vascular defects is apparent. Only four patients had associated extravascular congenital defects.

COMMENT

If an AV malformation of the vein of Galen becomes evident in the neonatal period, it usually produces heart failure within the first few days. The circulatory changes at birth result in increased blood flow through the fistula and increased systemic venous return. Glatt and Rowe¹¹ have discussed the evidence that cardiac output normally increases considerably at birth, with the result that the heart in the newborn works nearly at maximum capacity. In this circumstance the heart can readily fail when additionally strained. The greatly increased cardiac output resulting from an AV fistula can produce the pulmonary hypertension noted on cardiac catheterization, and the typical electrocardiographic and x-ray film changes. Elevated right atrial pressure may cause a right-to-left shunt through a patent foramen ovale,²¹ thus explaining the cyanosis that often occurs. Most likely, the increased cardiac output produces the systolic murmur. The reduced systemic vascular resistance associated with the large systemic AV shunts tends to widen the pulse pressure. Since these cardiovascular effects dominate the clinical picture, the infants are first thought to have congenital heart disease. However, the findings are not typical of any one congenital heart defect.

When dealing with heart failure of obscure origin in a neonate, one must consider an intracranial AV fistula. One should listen for cranial bruits in such infants and in infants with enlarging heads. Although cranial bruits may be heard in normal children,^{33,34} those associated with intracranial AV fistulas tend to be continuous, and may be loud. Newborns with intractable heart failure need cardiac catheterization or angiography, unless an inoperable situation can be diagnosed confidently on clinical grounds. The major positive clues from such

procedures with regard to intracranial AV fistulas are hyperoxygenation in the superior vena cava and rapid return of contrast medium from the left side of the heart to the superior vena cava. The oxygen data may suggest total anomalous pulmonary venous return, but the pulmonary arteriogram will be normal.

The foregoing comments apply to all large intracranial AV malformations, whether or not they involve the vein of Galen. Those that do represent an especially difficult neurosurgical problem. Early diagnosis, with adequate arteriographic demonstration of the malformation and its nutrient vessels, will allow the surgeon to plan the best procedure. A direct approach, with total excision of the malformation, represents ideal management. If the ventricles are enlarged because of hydrocephalus, removal of ventricular fluid would facilitate exposure for such a direct approach. However, in most infants with early heart failure (such as ours), exposure will be difficult because the ventricles are normal in size, the malformation is large, and considerable blood flow is diverted. In these circumstances, it is preferable to clip or ligate nutrient vessels. The operating microscope makes it easier to visualize the operative field and minimizes the need for retraction of surrounding structures. An alternative approach is to pass emboli into the malformation percutaneously from extracranial arteries.³⁵ This procedure carries low risk, and although it rarely obliterates the malformation completely, it has substantially benefited some older patients, and might also be tried in infants.

Adequate reduction of blood flow into the vein of Galen alleviates the cardiac overload, and the heart failure clears. The aneurysm shrinks, and subsequent hydrocephalus and cerebral damage should be avoided. Cerebral parenchymal damage due to the AV malformation may have already occurred, as reported by Norman and Becker.³⁰ Even so, we believe that good results such as the one reported here justify vigorous medical and surgical attention to this otherwise generally fatal condition.

Dr Louis Lehmann referred this patient.

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