Giant arteriovenous malformations of infancy and childhood

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Twelve patients are presented with giant arteriovenous malformations of the brain requiring therapy in the neonatal or infant period. Eight patients were operated on, and six survived with no neurological deficit. Four patients were not operated on and all died. The diagnostic clinical triad of enlarged head, cranial bruit, and cardiac failure or enlargement is described, and angiographic characteristics and surgical therapy discussed.

Key Words arteriovenous malformation □9 infancy □9 surgical therapy

Giant arteriovenous malformations of the cerebral vessels are spectacular lesions, and those which become symptomatic in the neonatal period or in infancy are often life-threatening. Very few of these lesions have been reported, and only a small number have been successfully obliterated. The prognosis for these malformations is serious, many of these children are in desperate cardiac straits, and even the older children who are not in overt failure are often in tenuous cardiac balance.

In the past 6 years we have had experience with 12 of these lesions. Eight of the patients were operated on, and in all eight the malformation was completely obliterated. This paper describes these malformations in detail and elaborates upon the surgical techniques which have allowed six of these eight children to survive as normal individuals.

Clinical Material

In all of the cases diagnosed immediately after birth, the true nature of the problem became apparent during cardiac angiography. In each case no cardiac defect was found, but the angiographer astutely recognized unusually large vessels arising from the aortic arch destined for the brain. The diagnosis was confirmed with a single injection of the contrast medium and skull films demonstrating the malformation.

In the older children the diagnosis was suspected and the patients were referred because of the combination of cardiac enlargement, intracranial bruit, and an enlarged head; only one patient had cardiac failure. Definitive angiographic studies were carried out by a combination of direct carotid and brachial techniques or by femoral angiography with selective catheterization of each of the major vessels.

Seven of the 12 patients in this series presented shortly after birth in intractable cardiac failure. Four of the remaining five patients were operated on before the first year. One was in overt cardiac failure and had been so since birth, and another had a...
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markedly enlarged heart. The twelfth child was 3 years old at the time of surgery.

The four older children were all hydrocephalic, but none of these was investigated extensively for the etiology of the hydrocephalus. Head enlargement and angiographic evidence for hydrocephalus were common in several of the neonates; air encephalography was not carried out. Intracranial bruits were present in all patients. The four patients not operated on had arteriovenous malformations in the region of the great vein of Galen. At the time one patient was deemed not to be a candidate for surgery; later review suggests that this patient would now be considered for surgical procedure. In two patients surgery was declined by the parents or attending pediatricians; in one case the diagnosis was not suspected in spite of the classical clinical triad of symptomatology until after the child had died from cardiac failure.

The eight patients who underwent surgery will be described in detail.

Case Reports

Case 1

This 3-week-old child was admitted because of intractable cardiac failure present since birth and remarkable cardiac enlargement. Cardiac angiography demonstrated large carotid and vertebral vessels on the right side. Cerebral angiography revealed a large arteriovenous malformation feeding primarily from the right middle cerebral artery and draining into the transverse sinus (Fig. 1). The lesion was totally excised in the fourth week of life. There was prompt relief of cardiac symptoms, and the child is now developing normally.

Case 2

This child was referred to the University Hospitals at the age of 10 months but since birth had been in chronic congestive failure refractory to all medical therapy. The heart

Fig. 1. Case 1. Arteriograms demonstrating the malformation in the distribution of the middle cerebral artery. Left: Lateral view showing large draining vein through the sphenoparietal to the transverse sinus. The contribution from the vertebral artery is also apparent. Right: Anteroposterior view. The bulk of the malformation is seen located quite peripherally with a large dilated middle cerebral vessel supplying the major portion of the malformation medially and an enlarged vertebral artery feeding the complex. This malformation was totally excised from its location within the fissure without removal of brain substance.
FIG. 2. Case 2. Angiograms revealing a huge arteriovenous malformation in the distribution of the left middle cerebral artery. **Left:** Lateral view, showing large vessels coursing over most of the cortex. Some of these are arterial and some are venous. **Right:** The anteroposterior projection demonstrates the peripheral location of this malformation within the Sylvian fissure.

was markedly enlarged, the head was enlarged, and there was a cranial bruit. During cardiac angiography a huge left carotid artery was discovered. Intracranial studies revealed an arteriovenous malformation involving the entire left hemisphere, feeding primarily from the middle cerebral circulation with immediate filling of huge cortical draining veins (Fig. 2). At operation the malformation was obliterated and excised without interrupting the intrinsic supply of the left hemisphere. In spite of a hypoplastic left middle cerebral artery the child is developing well and is neurologically normal.

Case 3

This patient was admitted at the age of 6 months because of a loud cranial bruit and an enlarged head. The child had a pulsatile mass behind the left ear. There was no overt cardiac failure, but the heart was markedly enlarged. Carotid angiography revealed a major communication between the external carotid artery and the lateral and sigmoid sinuses. There were many smaller branches from the meningeal circulation and lepto-meningeal anastomoses from the cerebral circulation. The major extracranial vessels were ligated in one operation, and at a planned second stage all intracranial and extracranial feeding vessels that could be identified were ligated.

At the age of 5 years the child returned because of increasing irritability and recurrence of the bruit in the neck. Reconstitution of the malformation by enlargement of multiple feeding channels was evident. In a third operation these intracranial feeding vessels were ligated at their entrance to the transverse sinus and the sinus ligated above and below the malformation. Postoperative angiography revealed obliteration of the fistula on the third operative attempt, and the child has remained normal to date.

Case 4

This child was referred to the University Hospitals because of cardiac symptomatology and a cranial bruit; examination at the time of admission showed that the heart was enlarged. Angiography revealed a huge arteriovenous malformation involving the posterior circulation, fed primarily from the
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Case 4

Left: Lateral angiogram of isolated injection of the vertebral artery demonstrating filling from greatly enlarged superior cerebellar, anterior inferior and posterior inferior cerebellar arteries. The draining veins in the region of the torcular are well seen. Right: Anteroposterior projection showing the greatly dilated venous drainage into the transverse sinus and into the region of the torcular. The greatly enlarged arterial supplies are demonstrated but individual feeding vessels cannot be defined in this view.

superior cerebellar, anterior inferior cerebellar, and posterior inferior cerebellar arteries on the left side (Fig. 3). This malformation was entirely excised, and the child has done well since. A shunting procedure has been required for control of hydrocephalus.

Case 5

This patient was transferred to the University Hospitals in intractable cardiac failure at the age of 1 day. After beginning digitalis therapy, angiocardiography was carried out, and a large malformation involving midline vessels was discovered. Selective catheterization of vessels demonstrated a malformation fed from both anterior cerebral and both posterior cerebral vessels, with an aneurysm of the great vein of Galen and a dilatation of adjacent venous structures draining directly into the torcular and transverse sinuses (Fig. 4). Because of the progressive cardiac failure surgery was undertaken on an emergency basis, with a standby cardiopulmonary bypass. The malformation was successfully obliterated without difficulty, and the child's precarious cardiac status appeared to be responding to therapy until 12 hours after surgery when sudden fatal cardiac arrest occurred.

Case 6

This patient was admitted because of rapidly progressive cardiac failure and the presence of a cranial bruit. Cardiography revealed that the heart was enlarged but normal in configuration and cerebral angiography demonstrated a large arteriovenous malformation of the great vein of Galen with feeding vessels from all major supertentorial arteries bilaterally (Fig. 5). Emergency surgery was carried out, and the malformation obliterated without difficulty. The draining veins were left in place, and the arterial communications ligated. The patient did not respond to cardiac therapy, and died 12 hours after the operation.

Case 7

This child was admitted at the age of 9 months because of an intracranial bruit and a progressively enlarging head. There was no history of cardiac problems, but at the time of admission the heart was found to be markedly enlarged and the liver palpable beyond the costal margin. Angiocardiography revealed a large arteriovenous malformation with an aneurysm of the great vein of Galen. The feeding vessels came...
from both anterior, both middle, and both posterior cerebral vessels with multiple connections between these vessels (Fig. 6). The magnitude of the shunt was demonstrated by the precarious cardiac status and cardiac enlargement. When the first operation was performed at the age of 11 months, it was not possible to completely obliterate
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FIG. 6. Case 7. Left: Lateral angiogram showing a relatively small aneurysm of the great vein of Galen with the arterial supply primarily from the posterior cerebral arteries bilaterally. Right: Anteroposterior view demonstrating the midline location of the aneurysm, and the superior entrance of the large feeding vessel from the posterior circulation.

the malformation. One year later the patient demonstrated a steady increase in head size and in size of the malformation (Fig. 7). At a second operation the malformation was completely obliterated. Cardiac arrest occurred during the procedure, and during the period of cardiac standstill the remainder of the feeding vessels were obliterated. The child was successfully resuscitated and 3 years later is entirely normal.

Case 8

A 6-month-old boy was admitted with a history of an enlarging head. Because of maternal blood incompatibility, exchange transfusion had been necessary in the first 6 hours of life. Head enlargement was noted at 2½ months of age; a cranial bruit was discovered after the child had had a focal seizure in the left leg. Angiography revealed a huge malformation involving the anterior and middle cerebral arteries and both the superior and inferior sagittal sinuses bilaterally (Fig. 8). The heart was enlarged, and the child was in chronic congestive failure. At operation the malformation was totally excised. The child is now 6 years of age and appears to be completely normal.

Discussion

Angiographic Techniques

In newborn children the diagnosis has routinely been made at the time of angiocardiography. It is our opinion that the diagnosis of intracranial malformation
should be made with a single injection of medium, and skull films, simply to document that the malformation is present. Unless the cardioangiography unit is equipped for cerebral vascular demonstration it is rarely worthwhile undertaking these studies of the malformation; the definition is usually not adequate, and the angiogram must be repeated. Administration of 15 to 20 cc of contrast medium to small children in cardiac failure can be a significant problem, and the necessity for repeating the entire angiogram should be avoided if possible. Once the diagnosis is made, complete studies are not obtained, and the procedure is transferred to the cerebral angiography unit, usually at a later date. Direct carotid, brachial, and femoral angiography have all been used successfully to demonstrate these malformations. However, it is extremely important to keep the amount of contrast medium at a minimum in newborn children because of volume and electrolyte content; therefore, femoral techniques designed to do this are preferable. In older children the diagnosis is most often suspected, and definitive studies are carried out primarily in the cerebral unit.

Operative Techniques

The operative procedures were carried out under general anesthesia. Four of the eight patients were cooled by external hypothermia to 25° to 29°C, but the advantage of this is dubious. A central venous catheter was used, and a brachial or femoral arterial catheter was in place in each patient. In the most recent cases, computer monitoring of all vital functions, blood gases, electrolytes, and fluids have been used. A unilateral craniotomy approach was employed for the laterally-placed malformations: a bilateral exposure via the vertex approach was used for midline lesions. A standby cardiopulmonary bypass was ready in one case; this precaution may be valuable for the large midline lesions especially those involving the region of the great vein of Galen. Magnifica-
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tion was used by the surgeon in each instance, and microsurgical techniques in most of the cases. Bipolar coagulation is of great value in obliterating the smaller branches of the malformation without significant injury to surrounding brain structures.

Probably the most important factor is proper handling of the critical cardiac status of these small infants.\(^1\)\(^3\)\(^,\)\(^11\) These children have an expanded blood volume because of the huge arteriovenous shunt, and rapid obliteration of this shunt can lead to overwhelming cardiac failure and sudden cardiac arrest. The enlarged heart is simply not capable of pumping the expanded blood volume against a sudden increased peripheral resistance. We use two methods for reducing the frequency of this complication. The first is a judicious and measured reduction of blood volume during the exposure. This usually means no blood replacement during as much of the operative procedure as possible with careful monitoring of actual blood loss and comparisons with calculated and measured blood volumes. It is certainly desirable to make serial measurements of blood volume during surgery, and in the postoperative period.

The second factor which the surgeon can control is the rapidity with which the malformation is obliterated. We have found that it is an advantage to ligate the major feeding vessels individually and relatively slowly, giving the heart time to equilibrate with the change in intravascular volume and peripheral resistance, rather than presenting it with an overwhelming load to be pumped against the suddenly increased resistance. If there is any question whether or not an individual branch should be ligated immediately it is our practice to occlude the vessel with temporary clips so that it can be opened if cardiac irregularities occur. During the time that the feeding vessels are being clipped it is extremely important to have an accurate assessment of cardiac status and to keep the anesthesiologists aware at all times of the progress of the procedure. As with any type of arteriovenous malformation the venous drainage should not be attacked until the arterial supply is obliterated; then the draining veins can be ligated if total removal of the malformation is planned. Midline lesions which involve the anterior portion of the sinuses can be excised, but those in the posterior portion of the sagittal sinus or in the region of the great vein of Galen are better left in place, once the arterial communications have been obliterated.\(^7\)\(^,\)\(^10\)

**Diagnosis and Prognosis**

The clinical diagnosis of these malformations is relatively easy, and the triad of signs and symptoms quite distinctive. In the neonates cardiac failure has routinely been present and combined with an intracranial bruit certainly suggests the diagnosis.\(^2\)\(^,\)\(^4\)\(^,\)\(^6\) An enlarged head is often present as well.\(^8\) In older children the cranial bruit is present, head enlargement more striking, and cardiac failure less common, although cardiac enlargement is still the rule. The malformations are of two basic types. The first is midline involving the great sinuses with feeding vessels entering from both sides. The second type is lateral, usually in the distribution of one middle cerebral artery with drainage both laterally and medially.

One of our malformations was in the posterior circulation and filled from all branches of the basilar on the left side and the posterior inferior cerebellar artery of the right side, the draining venous channels emptying into the transverse sinus. One was placed laterally involving both external and internal circulation.

Because of the impressive appearance of these malformations when viewed angiographically and because of their prognosis when left untreated, an aura of pessimism pervades their discovery. In our opinion this pessimism is unwarranted for many of these lesions can be successfully extirpated or obliterated.\(^1\)\(^,\)\(^11\) In the past 6 years at the University of Minnesota 12 of these malformations have been discovered. Eight of the patients have been operated on with total removal or obliteration of the malformation, and six are alive and well without significant neurological deficits; two patients died following surgery. All of the patients who were not operated upon have subsequently died.

It is our belief that the surgical treatment of these lesions when they become sympto-
matic in the neonatal period or early infancy offers the best and almost the only chance for successful survival.

References


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