

Intracranial Arteriovenous Anomalies of Infancy: Modern Concepts

John C. Godersky, Arnold H. Menezes

Department of Surgery (Division of Neurosurgery), University of Iowa Hospitals and Clinics, Iowa City, Iowa, USA

Key Words. Arteriovenous malformation · Arteriovenous fistula · Vein of Galen aneurysm · High-output cardiac failure · Hydrocephalus · Arterial digital subtraction angiography · Computed tomography

Abstract. A variety of cerebrovascular abnormalities may become manifest in the first years of life. We present our recent experience with 7 children, ages 1 week to 7 months of age, who presented with congestive heart failure and progressive head enlargement (3), heart failure alone (1) and head enlargement alone (2). The anomalies include vein of Galen aneurysm (3), complex arteriovenous fistula (2) and posterior fossa dural arteriovenous malformations (2). Computed tomographic scanning was invaluable in delineating the site of the lesion while arterial digital subtraction angiography was utilized to obtain the high-quality images, reduced dye volume and rapid filming. Direct surgical intervention was accomplished in 5 cases, while embolization was utilized in 2. One child underwent direct partial occlusion and embolization. Congestive heart failure resolved in 2 of 3 operated cases and improved in those undergoing embolization. Hydrocephalus improved in 2 cases with the elimination of the fistula. Morbidity included seizures and a hemorrhagic infarction in 1 case. Neurologic outcome was satisfactory and there was no surgical mortality.

Introduction

A variety of intracranial vascular abnormalities become manifest in the first days to months after birth. Some may be very dramatic in their clinical and radiographic presentation, while others are more subtle in appearance. None of these anomalies are seen commonly, while all present challenges in evaluation and management. These include obtaining an accurate definition of the vascular anatomy, while attending to the concomitant problems of congestive heart failure (CHF), hydrocephalus and cerebral ischemia. Computed tomography (CT) has provided valuable information regarding the nature and location of these abnormalities, as well as identifying any associated alteration in ventricular size or the presence of ischemic infarction. However, angiography continues to be required to define the specifics of the abnormal vascular anatomy.

This poses significant problems for these infants in terms of vascular access, volume of contrast agent administered and the potential for aggravation of CHF. The purpose of this report is to describe our recent experience in the evaluation and management of 7 infants with intracranial vascular lesions, including 3 vein of Galen aneurysms (VGA), 2 complex arteriovenous fistula (AVF) and 2 posterior fossa arteriovenous malformations (AVMs).

Patient Population and Illustrative Cases

Patient Population

Charts were reviewed on all infants with intracranial arteriovenous lesions cared for by the authors over the past 10 years. This included 7 individuals, 6 males and 1 female, ranging in age from 1 week to 7 months at the time of initial neurosurgical evaluation. Presenting symptoms included CHF in 5, in 4 of these infants progressive

Table I. Presenting symptoms in relation to age and type of anomaly

Case No.	Lesion	Age at presentation	CHF	Head enlargement	Cranial bruit
1	VGA	2 months	–	+ (shunted)	–
2	VGA	5 months	–	+	–
3	VGA	1 week	+ (severe)	+	+
4	AVF	6 months	+ (moderate)	–	+
5	AVF	7 months	+ (mild)	+	+
6	AVM(D)	6 months	+ (moderate)	+	+
7	AVM(P-D)	2 months	+ (severe)	+	+

D = Dural; P-D = mixed pial-dural.

head enlargement was also noted and all 5 had a demonstrable cranial bruit. The remaining 2 patients presented with head enlargement, without heart failure or a cranial bruit. In table I, the presenting symptoms in relation to age and type of anomaly are detailed. All patients were evaluated with cranial CT, with and without contrast enhancement. Angiography, either by conventional technique or arterial digital subtraction (ADSA), was also obtained. Perioperative management of volume status in those infants with CHF was aided by placement of a Swan-Ganz catheter. A pediatric cardiologist and neuroanesthesiologists functioned as integral members of the management team.

Case 3

This male child was seen during the 1st week of life with severe CHF, progressive head enlargement and a loud cranial bruit, heard best at the parietal vertex. He was somnolent and had a weak cry and suck reflex. A conventional arch aortogram was obtained prior to arrival at our institution, demonstrating a large VGA. Following initial management of the CHF by diuresis and digitalization, CT scanning (fig. 1) and ADSA were obtained on separate days to delineate the vascular supply of the malformation (fig. 2). Both carotids and one vertebral artery were studied by selective injection.

A planned, staged occlusion was undertaken when he was 8 days old. A right parietal craniotomy and parafalcine approach was utilized to expose the vessels entering the anterior-superior aspect of the fistula. The first two major feeding vessels were occluded with aneurysm clips as they entered the vein of Galen. When a third vessel was similarly occluded the brain began to swell and a subpial hemorrhage developed in the right parietal lobe. This perfusion pressure breakthrough was successfully managed by administration of mannitol and this therapy was continued postoperatively.

His heart failure improved temporarily and he developed seizures which were controlled with phenobarbital. Further evaluation disclosed a patent ductus arteriosus which was surgically closed when it failed to respond to medical therapy. Because of persistent CHF at age 3 months, a second attempt at fistula occlusion was undertaken by insertion of 12 Gianturco coils, each measuring 15 cm [23]. These were inserted in a retrograde fashion through the torcula and straight sinus into the aneurysm, under fluoroscopic control. He has remained stable with medically controlled CHF and at 1 year of age his seizures are controlled but he has shown moderate developmental delay. The cranial bruit is no longer audible.

Case 4

This child was seen at age 6 months because of mild heart failure. He had experienced a number of episodes of coughing and low-grade fever which had been diagnosed as bronchitis. In retrospect, these were caused by his CHF. On examination, he had a cranial bruit but his head size and neurologic examination were normal. CT demonstrated enlarged vascular channels entering a dilated sphenoparietal sinus (fig. 3). ADSA confirmed the presence of an AVF involving the right middle cerebral artery and draining into the sphenoparietal sinus, which was greatly enlarged (fig. 4).

Treatment consisted of a frontoparietal approach with exposure of the malformation. The sylvian fissure was opened and the proximal middle cerebral artery identified. The middle cerebral artery branches were followed to their entrance into the venous varix where they were clipped. The varix changed from red to blue in color and its turgor was reduced dramatically. Excision of the varix was not per-



Fig. 1. Contrast-enhanced cranial CT. Multiple enlarged vascular structures are seen entering a central mass, which is the greatly enlarged vein of Galen. Posteriorly, the dilated straight sinus and torcula are evident. Ventricular enlargement is also present.

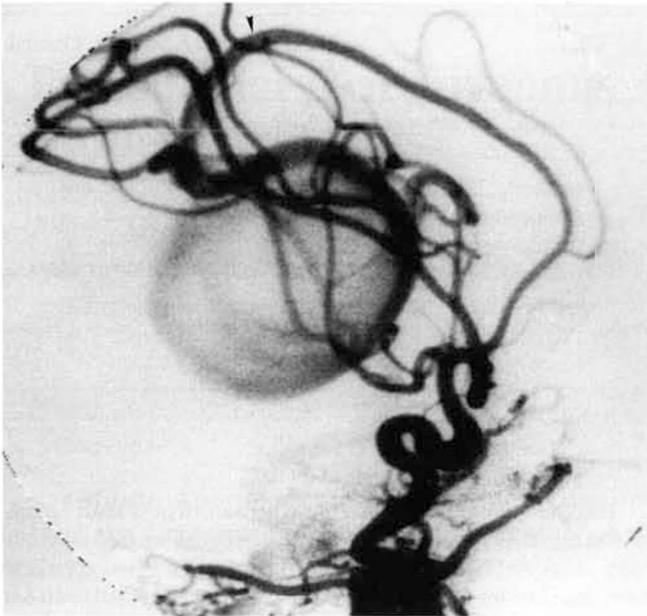


Fig. 2. AD SA of the right common carotid artery 1.5 s after injection. The enlarged anterior cerebral artery (arrowhead) and multiple middle cerebral branches can be seen entering the VGA. The paucity of normal vessels to the brain is evident.

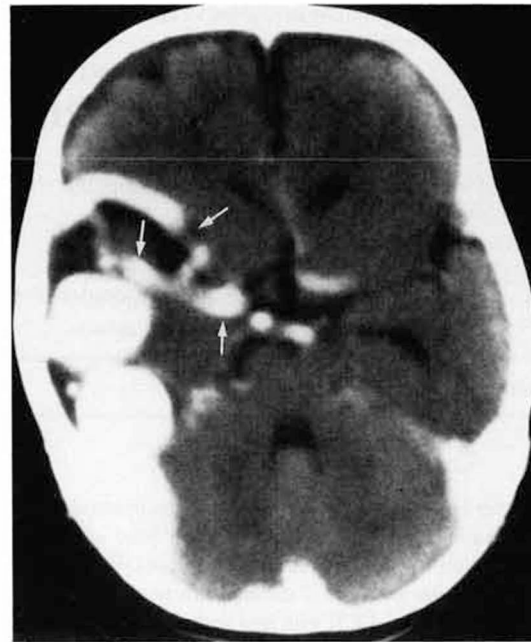


Fig. 3. Contrast-enhanced CT. Marked enlargement of the middle cerebral artery and its branches (arrows) is evident. The two globular masses are the dilated sphenoparietal sinus.



Fig. 4. (a) Anterior-posterior view of the left carotid AD SA. The dilated middle cerebral artery branches enter the venous varix, lying in the lateral sylvian fissure. There is a relative lack of normal middle cerebral artery branches to the parenchyma. (b) Lateral right carotid DSA showing the rapid shunting from middle cerebral artery into the venous varix and sphenoparietal sinus.

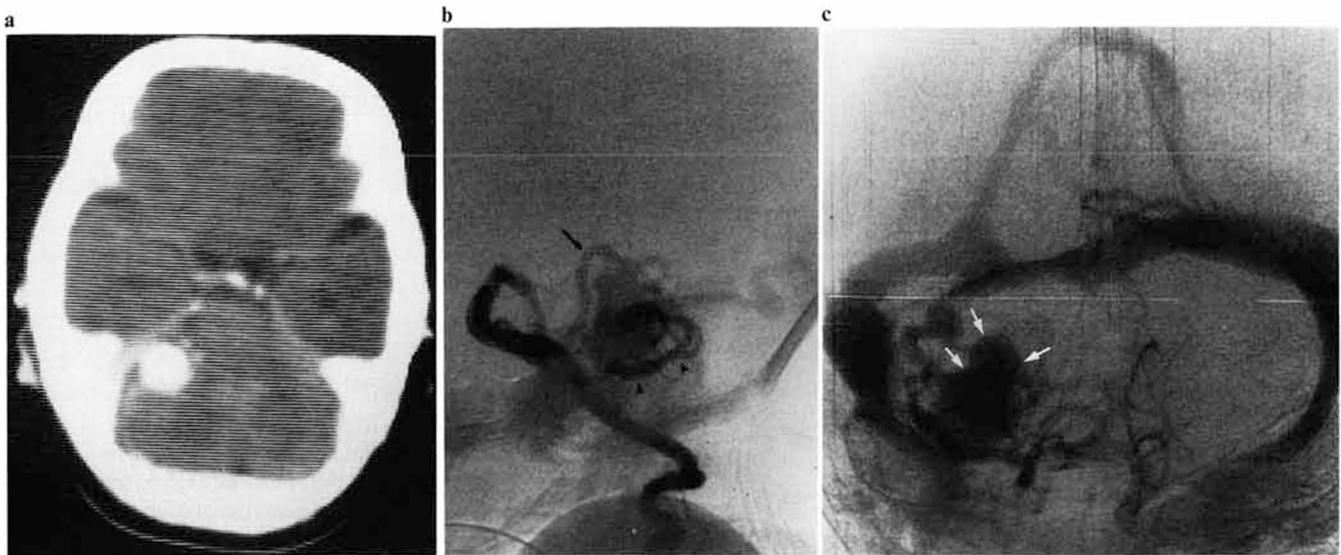


Fig. 5. Contrast-enhanced CT (a) with enhancing mass adjacent to the tentorium, two vessels are seen anterior and medial to the main lesion. Lateral vertebral artery injection (b) after $2/3$ s, demonstrating enlarged superior cerebellar artery (arrow) and anterior-inferior cerebellar artery (arrowheads) supplying the fistula. Anterior-posterior view at 3 postinjection of the vertebral artery (c). The venous system is opacified while contrast remains in small arteries. The venous varix is evident in the region of the right cerebellopontine angle (arrows).

formed. Progressive collapse of the aneurysmal sac was evident on the postoperative CT scans 2 weeks and 3 months after surgery. He recovered uneventfully.

Case 5

This 7-month-old male was evaluated for intermittent episodes of tachypnea, tachycardia and poor feeding. A bruit was audible in the right parietal-occipital area. Mild CHF and cardiomegaly were evident on examination and chest radiograph. The head was enlarged and the anterior fontanelle full. Radiographic evaluation included a CT which demonstrated an enhancing 1×1.5 cm mass in the right cerebellopontine angle. ADSA defined the lesion as an AVF involving the anterior-inferior cerebellar artery and marginal branches of the superior cerebellar artery (fig. 5). In addition, the ipsilateral anterior circulation filled the fistula by way of an enlarged posterior communicating artery. Surgical correction was undertaken through a right subtemporal transtentorial approach. The enlarged superior and anterior-inferior cerebellar artery branches were followed to the fistula and occluded. The aneurysmal sac was not excised. Subsequent CT scans showed normalization of the ventricular size and resolution of the aneurysmal mass.

Case 6

This 6-month-old child presented with high-output heart failure, head enlargement and a cranial bruit. On CT scan, an enhancing posterior fossa mass (fig. 6) was evident. Angiography done prior to our routine use of digital subtraction techniques demonstrates contributions to the malformation from posterior meningeal, middle meningeal, tentorial and occipital artery branches (fig. 7). This AVM was not considered to be resectable by a direct approach. Staged embolization was chosen as a means of reducing the flow through the

malformation and thereby treating the heart failure. Particulate embolization was carried out on 2 separate occasions. Potembolization CT demonstrated reduced flow through the AVM. His heart failure became medically controllable and his head size stabilized. He is 2 years postembolization and has not required further intervention.

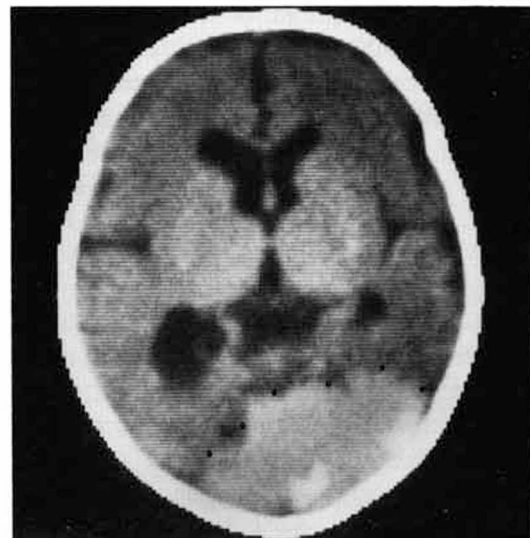


Fig. 6. Noncontrast head CT. Mild ventricular enlargement is present. A mass is seen posteriorly (●) which is comprised of enlarged vascular channels in the tentorium and the venous outflow through the dilated torcula.

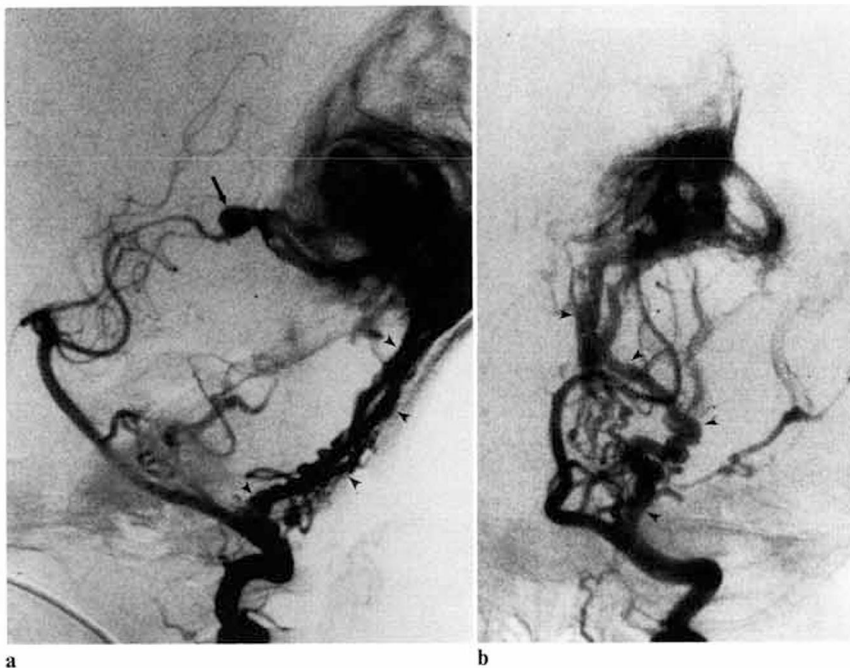


Fig. 7. Lateral (a) and anterior-posterior (b) views of the left vertebral angiogram 1.5 s after injection. a An aneurysmal dilatation is seen on the superior cerebellar artery (arrow) before it enters the AVM. Enlarged posterior meningeal vessels constitute the main supply to the malformation (arrowheads in a and b).

Treatment and Results

A summary of the therapies employed for control of these malformations is listed in table II. In the first 5 cases, a direct surgical approach was used with occlusion of the arterial branches at the site of the fistula. A single operation was required in 4 cases. In case 3, a second procedure, consisting of transvenous placement of Gianturco coils, was required for control of his symptoms. In both dural AVMs (cases 6 and 7) staged embolization with particulate material was chosen to reduce the flow through the malformation. A direct surgical approach was not felt to be feasible in these patients. Although the AVMs were incompletely occluded, the

patients CHF was controlled in both cases with regression of the AVM over subsequent follow-up examinations by CT.

The outcome was favorable in the group as a whole (table III). Hydrocephalus was present preoperatively in 6 of 7. One underwent ventriculoperitoneal shunt placement at another institution prior to our evaluation. In the remaining 5 cases, the ventricular enlargement was mild in 3 and moderate in 2. The ventricular size improved in all 5 cases and none of these have required shunt placement. CHF, present in 5 cases, resolved completely in 2 and was controlled medically in the remaining 3. The neurologic status was considered normal in 5, while 2 showed moderate developmental delay.

Table II. Summary of therapies

Case No.	Lesion	Treatment
1	VGA	complete occlusion
2	VGA	complete occlusion
3	VGA	partial occlusion/venous embolization
4	AVF	complete occlusion
5	AVG	complete occlusion
6	AVM(D)	staged arterial embolization (partial)
7	AVM(P-D)	staged arterial embolization (partial)

D = Dural; P-D = mixed pial-dural.

Discussion

None of the vascular lesions described in this report are commonly seen [4, 13]. The VGA have been reported most frequently. In a review of the literature in 1982, Hoffman et al. [11] noted 128 previous reports of VGA. The actual incidence of this malformation is unknown. Reports of AVF [20, 31, 39] and dural AVM [1, 15, 21] have been infrequent.

It is evident that CHF has a prominent role in the pathophysiology of these infants [5, 8, 9, 18, 24, 25, 36].

Table III. General and neurologic outcome

Case No.	Lesion	General outcome	Neurologic outcome
1	VGA	hydrocephalus previously shunted	normal
2	VGA	hydrocephalus improved	normal
3	VGA	CHF improved, hydrocephalus improved	parietal infarct, moderate developmental delay
4	AVF	CHF resolved	normal
5	AVF	CHF resolved, hydrocephalus improved	normal
6	AVM(D)	CHF improved, hydrocephalus improved	normal
7	AVM(P-D)	CHF improved, hydrocephalus improved	moderate developmental delay

D = Dural; P-D = mixed pial-dural.

Recognition of heart failure is not difficult when overt but in milder cases it may be confused with other forms of respiratory disease, such as recurrent bronchitis, pneumonia or asthma. The heart failure occurs because of the increased volume of blood passing through the intracranial arteriovenous communication [7, 14]. This high output failure is difficult to manage prior to closure of the intracranial shunt. A pediatric cardiologist plays an integral role in the management of these patients pre- and postoperatively. These children were frequently referred to us by the cardiologist because of the suspected intracranial shunt. It must be remembered that associated congenital heart disease may also be present, for example a patent ductus arteriosus, and this may complicate the patient's management [7, 19].

Disordered embryogenesis has led the list of causative factors for the development of these malformations [6, 26]. These malformations are frequently characterized by massive enlargement of the venous outflow tract with the formation of a venous varix. AVMs encountered in adults rarely are associated with such venous dilatation, especially if the drainage is via a cortical vessel. The structure of the vein of Galen and the fact that it lies free in the subarachnoid space, as opposed to being encased in dural sheath, has been proposed to explain the massive enlargement in some VGAs. However, we and others [20, 39] have seen similar enlargement at other sites of venous drainage, associated with high-flow AVF (cases 4, 5). Viñuela et al. [38] have recently described alterations in the venous drainage of deep-seated AVMs including venous occlusions, vessel wall irregularities and stenosis. They pro-

pose that these changes are the result of intimal damage from the turbulence. It is most probable that these are secondary changes and not responsible for the malformation itself. However, Lasjaunias et al. [16] have proposed that enlargement of the vein of Galen only occurs if the intracranial fistula is associated with a distal dural sinus occlusion.

Previous reports dealing with VGA have detailed the presenting signs and symptoms of these lesions and related them to the age at the time of diagnosis [2, 10, 11]. It is generally accepted that the higher the flow through the malformation the earlier the age of presentation. Those with very high flows are seen in the first days to weeks of life with high-output cardiac failure. Malformations with lesser flows usually present for medical attention because of head enlargement, secondary to hydrocephalus. Headaches, hemorrhage or progressive neurologic dysfunction are seen in older children or adults.

The presence of head enlargement in these infants, with or without a cranial bruit, led to evaluation by CT scanning. These studies were routinely obtained both with and without contrast enhancement. In all instances, the presence of the vascular pathology was demonstrated [3, 34]. Additional information is also obtained with CT regarding ventricular size, the presence of existing ischemic infarction or associated extracerebral fluid collections. Angiography was performed in all cases [6]. General anesthesia was employed and a transfemoral route used for access. Early in our experience, standard angiography with selective injections of the feeding vessels was obtained. As our ex-

perience with ADSA widened, this technique was used routinely. In the evaluation of high-flow AVF and AVMs, ADSA offers the advantage of very rapid filming up to 10 frames per second, as compared to 3 per second with conventional angiography. Furthermore, ADSA uses less contrast per injection, allowing selective study of the multiple vessels contributing to the malformation. Conventional angiography often reaches the maximum contrast limit of 4–5 cm³/kg without obtaining full definition of the vascular malformation [3, 20]. This necessitates repeating the study at a subsequent procedure. The resolution of ADSA has been quite satisfactory for planning operative approaches in the 3 cases described. The reduced dye volume with ADSA also limits the risk of exacerbation of CHF and renal failure.

The pathogenesis of the hydrocephalus associated with these lesions has engendered a great deal of discussion. Such hydrocephalus occurs unusually in neonates, but frequently is seen in infants and older children [11] and has had its onset as late as 62 years of age [29]. In VGA it is felt that the mass created by the venous aneurysm compresses the aqueduct of Sylvius leading to the hydrocephalus [24]. Other cases of venous compression of CSF pathways have also been reported [30]. Hydrocephalus is frequently present when the VGA is thrombosed but the exact cause of the hydrocephalus in these cases is unclear [17, 32]. Measurement of sagittal sinus pressure, before and after occlusion of the AVF, has shown high pressures that become normal after occlusion [23]. This elevated sinus pressure may impede CSF absorption leading to ventricular enlargement. Repeated, subclinical episodes of subarachnoid hemorrhage have also been implicated as a potential cause of hydrocephalus [29]. It is also possible that a combination of factors may be involved. In 5 of our cases, the hydrocephalus improved following obliteration of the fistula or reduction in the flow through the malformation. To date, none of these patients have required a shunt. This observation does not help us answer the question as to the pathogenesis of hydrocephalus, in that it would fit with all the theories except that of recurrent subarachnoid hemorrhage. However, we feel the shunting is not uniformly required if the malformation can be treated. Other reports of the need for shunt placement following malformation treatment have been variable.

It is well accepted that in the neonate with high-output cardiac failure, management must include reduction or obliteration of flow through the fistula. How this end is best accomplished remains to be settled. The majority

of VGAs are composed of a fistulous connection between feeding arteries and the vein of Galen. Some authors have recommended ligation of the arterial supply along with resection of the aneurysmal vein of Galen [33]. A more widely held opinion is that the venous portion need not be resected (as it represents a normal channel for venous drainage) as long as the fistula is occluded [2, 40]. We ascribe to this approach [22] and have demonstrated gradual reduction in the size of the venous portion of the AVF (cases 4 and 5) following fistula interruption. The procedure is simplified by leaving the sac in place and avoids the risk of causing impairment in venous drainage. In instances where the varix is clotted, removal of the thrombus from the venous sac may be appropriate, but again the vein itself need not be removed [32].

A more critical question in management is a determination of whether the fistula may be occluded in a single procedure or whether staggered occlusion is more appropriate [19]. This must be individualized based on the patient's symptoms and cardiac status. Attempted complete occlusion of the fistula in neonates with high-output cardiac failure has uniformly led to volume overload and death from heart failure. Even partial occlusion may stress an already ischemic and maximally dilated heart [11, 12]. Furthermore, perfusion pressure breakthrough, as seen in our case 3, may occur. This problem has been discussed in a number of reports [28, 35]. Measurements of sagittal sinus pressure [23], cardiac outputs and flow through the fistula [27] may eventually serve as guides to determine the amount of occlusion that can be best tolerated. In older children, without significant cardiac failure, fistula occlusion can usually be accomplished in a single procedure, but volume status and cardiac function need to be monitored closely in these individuals.

High-flow dural and mixed dural-pial AVMs presenting in infancy are extremely difficult problems to eradicate. Attempts at embolization, resection and a combination of these procedures have been employed [1]. Lasjaunias et al. [15] have described symptoms ranging from cranial nerve dysfunction to impairment in cortical function, in adults with dural AVMs. It was our goal (cases 6 and 7) to reduce the flow through the AVM to a sufficient degree to control the CHF. This has been accomplished for the present. It is possible that further intervention will be needed should other symptoms arise.

Autopsy studies of neonates with VGA and severe heart failure have demonstrated the existence of antenatal ischemic infarction in water shed zones, as well as

hemorrhagic infarction occurring postdelivery [26, 37]. These findings and the poor prognosis with or without surgery have led some to fatalistically recommend that these infants should not be aggressively treated. Although the outcome for these gravely ill infants is poor, it does not appear hopeless. Further advances in medical and surgical management may lead to satisfactory outcomes for some. Those individuals presenting with hydrocephalus, seizures, subarachnoid hemorrhage or cerebral ischemia have a more favorable prognosis.

Summary

Seven infants are described with intracranial vascular anomalies. Evaluation with CT and ADSA angiography helped guide interventional therapy. CHF was resolved or became medically manageable following reduction or elimination of flow through the vascular malformation. Obliteration of the fistula without resection of the venous aneurysm improved the hydrocephalus in 2 cases. Reduction of flow through the malformation in 3 other cases also controlled the hydrocephalus. Neurologic outcome was considered satisfactory following successful treatment.

References

- Albright, A.L.; Latchaw, R.E.; Price, R.A.: Posterior dural arteriovenous malformation in infancy. *Neurosurgery* 13: 129-135 (1983).
- Amacher, A.L.; Shillito, J.: The syndromes and surgical treatment of aneurysms of the great vein of Galen. *J. Neurosurg.* 39: 89-98 (1973).
- Brunelle, F.O.S.; Harwood-Nash, D.C.F.; Fitz, C.R.; Chuang, S.H.: Intracranial vascular malformations in children: computed tomographic and angiographic evaluation. *Radiology* 149: 455-461 (1983).
- Celli, P.; Ferante, L.; Palma, L.; Cavedon, G.: Cerebral arteriovenous malformation in children: clinical features and outcome of treatment in children and in adults. *Surg. Neurol.* 22: 43-49 (1984).
- Carroll, C.P.H.; Jakoby, R.K.: Neonatal congestive heart failure as the presenting symptom of cerebral arteriovenous malformation. *J. Neurosurg.* 25: 159-163 (1966).
- Clarisse, J.; Dobbelaere, P.; Rey, C.; D'Hellemmes, P.; Hassan, M.: Aneurysms of the great vein of Galen. Radiological-anatomical study of 22 cases. *J. Neuroradiol.* 5: 91-102 (1978).
- Cumming, G.R.: Circulation in neonates with intracranial arteriovenous fistula and cardiac failure. *Am. J. Cardiol.* 45: 1019-1024 (1980).
- Cunliffe, P.N.: Cerebral arteriovenous aneurysm presenting with heart failure, report of 3 cases. *Br. Heart J.* 36: 919-923 (1974).
- Eide, J.; Følling, M.: Malformation of the great vein of Galen with neonatal heart failure: report of 2 cases. *Acta paediat. scand.* 67: 529-532 (1978).
- Gold, A.P.; Ransohoff, J.; Carter, S.: Vein of Galen malformation. *Acta neur. scand.* 40: suppl. 11, pp. 1-31 (1964).
- Hoffman, H.J.; Chuang, S.; Hendrick, E.B.; Humphreys, R.P.: Aneurysms of the vein of Galen: experience at the hospital for sick children Toronto. *J. Neurosurg.* 57: 316-322 (1982).
- Jedeikin, R.; Rowe, R.D.; Freedom, R.W.; Olley, P.M.; Gihan, J.E.: Cerebral arteriovenous malformation in neonates: the role of myocardial ischemia. *Pediatr. Cardiol.* 4: 29-35 (1983).
- Kelly, J.J.; Mellinger, J.F.; Sundt, T.M.: Intracranial arteriovenous malformations in childhood. *Ann. Neurol.* 3: 338-343 (1978).
- Lakier, J.B.; Milner, S.; Cohen, M.; Levin, S.E.: Intracranial arteriovenous fistulas in infancy - hemodynamic considerations. *S. Afr. med. J.* 61: 242-245 (1982).
- Lasjaunias, P.; Chui, M.; Brugge, K.T.; Tolia, A.; Hurth, M.; Bernstein, M.: Neurological manifestations of intracranial dural arteriovenous malformations. *J. Neurosurg.* 64: 724-730 (1986).
- Lasjaunias, P.; Brugge, K.T.; Lopezibor, L.; Chui, M.; Flodmark, O.; Chuang, S.; Goasquen, J.: The role of dural anomalies in vein of Galen aneurysms: report of six cases and review of the literature. *Am. J. Neuroradiol.* 8: 185-192 (1987).
- Lazar, M.L.: Vein of Galen aneurysm: successful excision of a completely thrombosed aneurysm in an infant. *Surg. Neurol.* 2: 22-24 (1974).
- Levy, A.M.; Hanson, J.S.; Tabakin, B.S.: Congestive heart failure in the newborn infant in the absence of primary cardiac disease. *Am. J. Cardiol.* 26: 409-415 (1970).
- Lillquist, K.; Haase, J.; Thaysen, P.: Operative treatment of cerebral arteriovenous aneurysm of the vein of Galen complicated by congestive heart failure. *Br. Heart J.* 42: 738-741 (1979).
- Long, D.L.; Seljeskog, E.L.; Chou, S.N.; French, L.A.: Giant arteriovenous malformation of infancy and childhood. *J. Neurosurg.* 40: 304-312 (1974).
- Mazza, C.; Pasqualin, A.; Scienza, R.; Bazzan, A.; DaPian, R.: Intracranial arteriovenous malformations in the pediatric age: experience with 24 cases. *Child's Brain* 10: 369-380 (1983).
- Menczes, A.H.; Graf, C.J.; Jacoby, C.G.; Cornell, S.H.: Management of vein of Galen aneurysms: report of two cases. *J. Neurosurg.* 55: 457-462 (1981).
- Mickle, J.P.; Quisling, R.G.: The transtorcular embolization of vein of Galen aneurysms. *J. Neurosurg.* 64: 731-735 (1986).
- Montoya, G.; Dohn, D.F.; Merca, R.D.: Arteriovenous malformation of the vein of Galen as a course of heart failure and hydrocephalus in infants. *Neurology* 21: 1054-1058 (1971).
- Nielsen, G.: Arteriovenous malformations as a cause of congestive heart failure in the newborn and infant. *Eur. J. Pediatr.* 142: 298-300 (1984).
- Norman, M.G.; Becker, L.E.: Cerebral damage in neonates resulting from arteriovenous malformations of the vein of Galen. *J. Neurol. Neurosurg. Psychiat.* 37: 252-258 (1974).
- Nornes, H.; Grip, A.; Wikeby, P.: Intraoperative evaluation of cerebral hemodynamics using directional Doppler technique. I. Arteriovenous malformations. *J. Neurosurg.* 50: 145-151 (1979).
- Nornes, H.; Grip, A.: Hemodynamic aspects of cerebral arteriovenous malformations. *J. Neurosurg.* 53: 456-464 (1980).
- Rosenfeld, J.V.; Fabinyi, G.C.A.: Acute hydrocephalus in an elderly woman with an aneurysm of the vein of Galen. *Neurosurgery* 15: 852-854 (1984).
- Sang, H.; Kerber, C.: Ventricular obstruction secondary to vascular malformations. *Neurosurgery* 12: 572-574 (1983).

- 31 Schijman, E.; Monges, J.A.: Giant arteriovenous aneurysm of the posterior fossa in a three-month-old infant. *Child's Brain* 10: 121–129 (1983).
- 32 Six, E.G.; Cowley, A.R.; Kelly, D.L., Jr.; Laster, D.W.: Thrombosed aneurysm of the vein of Galen. *Neurosurgery* 7: 274–278 (1980).
- 33 Smith, D.R.; Donat, J.F.: Giant arteriovenous malformations of the vein of Galen: total surgical removal. *Neurosurgery* 8: 378–382 (1981).
- 34 Spallone, A.: Computed tomography in aneurysms of the Vein of Galen. *J. Comput. assist. Tomogr.* 3: 797–782 (1979).
- 35 Spetzler, R.F.; Wilson, C.B.; Weinstein, P.; Mehdorn, M.; Townsend, J.; Telles, D.: Normal perfusion pressure break through theory. *Clin. Neurosurg.* 25: 651–672 (1978).
- 36 Stanbridge, R.D.L.; Westaby, S.; Smallhorn, J.; Taylor, J.F.N.: Intracranial arteriovenous malformation with aneurysm of the vein of Galen as a cause of heart failure in infancy: echocardiographic diagnosis and results of treatment. *Br. Heart J.* 49: 157–162 (1983).
- 37 Takashima, S.; Becker, L.E.: Neuropathology of cerebral arteriovenous malformations in children. *J. Neurol. Neurosurg. Psychiatr.* 43: 380–385 (1980).
- 38 Viñuela, F.; Nombela, L.; Roach, M.R.; Fox, A.J.; Pely, D.M.: Stenotic and occlusive disease of the venous drainage system of deep brain AVM's. *J. Neurosurg.* 63: 180–184 (1985).
- 39 Viñuela, F.; Drake, C.G.; Fox, A.J.; Pely, D.M.: Giant intracranial varices secondary to high flow arteriovenous fistulae. *J. Neurosurg.* 66: 198–203 (1987).
- 40 Yasargil, M.G.; Antic, J.; Laciga, R.; Jain, K.K.; Boone, S.C.: Arteriovenous malformations of the vein of Galen: microsurgical treatment. *Surg. Neurol.* 6: 195–200 (1976).

John C. Godersky, MD
 Division of Neurosurgery
 C42 E, GH
 University of Iowa Hospitals
 Iowa City, IA 52241 (USA)

Editorial Comment

This paper nicely outlines the therapeutic options that are available for the treatment of infants with high flow arteriovenous anomalies of the brain. The authors demonstrate that the prognosis for these infants with heart failure as a result of one of these lesions is no longer hopeless. With the exception of the newborn with severe uncontrollable failure who dies in the first 24–48 h of life, children with heart failure as a result of AVMs can be stabilized and, in many cases, have the lesion decrease in size or cured. The operative morbidity and mortality have diminished markedly and make the procedures well worth considering.

We still are not certain of the prognosis for this group of children if they go untreated. It does appear, however, that the majority of these children will develop neurological problems related to their vascular lesion: headache, bleeding, or intellectual deterioration, steal, even although this may be delayed for 10–20 years. The availability of CT and MRI scanning and the use of digital

subtraction angiography have made the definition of these lesions much easier. The ability to define the feeders has allowed intervention, whether surgical or radiological, with relatively low morbidity. The goals of surgery are not yet clarified. The authors rightly observe that in these lesions with heart failure, the initial goal of the therapy is to reduce the heart failure by diminishing flow through the lesion rather than to make efforts to completely abolish the arteriovenous fistula. It is not at all clear whether complete obliteration at some later time is necessary or not. Long-term follow-up of this group of patients will be necessary to obtain the answer.

This review points out the importance of selecting therapy based on the individual child and his or her problems. Many of these children will indeed survive the first few months of life without requiring surgical intervention, others may require only a shunting procedure in the early phases but will require some other surgical intervention through childhood.

Derek A. Bruce, MD