The transtorcular embolization of vein of Galen aneurysms

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Vein of Galen malformations are a diverse group of deeply located high-flow vascular lesions that are difficult to eradicate with standard surgical techniques. New nonsurgical interventional techniques offer encouraging results as alternative therapeutic choices in these complicated vascular shunts. Use of a new procedure of transtorcular embolization with Gianturco embolic coils is described in three patients harboring vein of Galen aneurysms. Two of the three patients had a satisfactory outcome. This technique is simple and quick, and can produce progressive thrombosis in these high-flow vascular fistulas.

KEY WORDS • vein of Galen • aneurysm • embolization • vascular shunt

SURGICAL excision remains the best mode of therapy for most arteriovenous malformations of the brain. Vein of Galen malformations do not lend themselves to excision because they are deep midline shunts, usually with many feeding arteries that are difficult to reach. The excellent review by Hoffman, et al., detailed the clinical spectrum of this entity, the poor prognosis of the untreated malformation, and the poor surgical outcome in the neonate and older child. Since surgical excision of vein of Galen aneurysms is often impossible, it seems reasonable to attempt an interventional method of treatment with or without surgery in an effort to improve the outcome for these patients. Berenstein and Epstein demonstrated encouraging results with combined therapy. We report here the technique and early results with a new procedure known as the “transtorcular embolization of vein of Galen aneurysms.”

Rationale and Techniques

Since the high-flow shunt in vein of Galen aneurysms produces many of the pathophysiological changes observed in these patients, it is reasonable to assume that a reduction of this shunt would be of benefit to the patient. This can be accomplished by directly attacking the feeding vessels surgically, or by nonsurgical interventional techniques using a transarterial or (theoretically) transvenous approach. Encouraged by a report by Laws (unpublished data, 1982), we developed the following technique to reduce the shunt in the vein of Galen aneurysm by transtorcular embolization (Fig. 1).

We initially considered using a detachable balloon system for the embolic unit, but discarded this option because of the potential for acute venous outlet obstruction in the brain. We elected to use Gianturco coil-spring emboli since in theory these coils could be placed in such a way as to produce reduction in flow in a graded fashion.

The patients are operated on with the head in the full lateral position. A small craniectomy is made over the area of the torcular, and the torcular, straight sinus, or vein of Galen aneurysm are identified with a real-time Neurosector ultrasound system. The torcular is punctured with a No. 16 Angiocath, and a soft-ended guide wire is maneuvered into the vein of Galen aneurysm under fluoroscopic control. A standard 8- to 10-cm angiography catheter is threaded over the wire and positioned in the lumen of the vein of Galen aneurysm. All manipulations including the initial catheter placement and the deposition of embolic coils are carefully monitored fluoroscopically. At various intervals, venography is used to monitor blood flow rates through the fistula. Once the angiography catheter is placed within the center of the aneurysm, four to eight 15-cm Gianturco coils are inserted (Fig. 1). We have developed a special tethering plunger system for this part of the procedure which allows retrieval of the coil if it is inadvertently placed in an undesirable position or migrates distally into the straight sinus. We believe it is essential to be able to retrieve a misplaced coil prior to permanent placement, and the tethering plunger system is very efficient in allowing this control.
**Fig. 1.** Illustration of the embolization technique. The polyethylene embolization catheter (c) is fluoroscopically directed through the torcula and straight sinus into the fundus of the vein of Galen aneurysm. A Touhy-Borst adapter (a) fixed proximally to the catheter minimizes blood loss during the procedure. The Gianturco-coil loader is first introduced into the embolization catheter and the plunger is advanced, displacing the embolization wire toward the target. Once positioned satisfactorily, the tethering monofilament is severed by slipping the outer cannula with its distal cutting edge over the inner cannula, which has a distal cutting block. A twisting motion is then made to cut the monofilament. After the monofilament is detached, the entire coaxial system is removed and the next wire can be introduced through the embolization catheter which has remained in place.

The initial stage of the embolization procedure is intended to reduce by approximately one-half the original flow through the shunt. This has required the use of four to eight large coils in each of our three patients. The flows in the torcular and vein of Galen aneurysms are monitored postoperatively with Doppler ultrasonic studies. In the second stage of treatment, the patient again undergoes embolization 3 to 21 days after the initial procedure, if needed. In all of our patients we have elected to eliminate completely the flow through the vein of Galen aneurysm during the second procedure. An average of 14 to 17 coils was required to accomplish this goal in each patient. Shorter coils can be deposited in the center of the aneurysm during the later phases of embolization.

**Case Reports**

**Case 1**

This baby girl was born with mild heart failure and a loud murmur over the cranium, neck, and chest. There was a palpable thrill in the neck. Computerized tomography of the head showed what appeared to be a vein of Galen aneurysm and moderate hydrocephalus. A standard ventriculoperitoneal (VP) shunt was placed in the right frontal horn of the lateral ventricle and, at the time of that operation, the intraventricular and superior sagittal sinus pressures were recorded (Table 1). At 4 months of age the patient developed focal motor and generalized seizures resistant to medical control. Angiography disclosed a large shunt into the dilated vein of Galen (Fig. 2 upper left).

In June, 1984, the patient underwent placement of four long Gianturco spring emboli into the vein of Galen aneurysm via a transtorcular approach. Angiography showed reduced but persistent flow through the malformation (Fig. 2 upper right) and her seizures continued. Therefore, 2 weeks after initial operation she underwent repeat wire embolization. A total of 17 emboli were deposited. Flow within the vein of Galen aneurysm was reduced to zero, as revealed by venography and Doppler ultrasonic testing. The child remains well with an occasional seizure. Doppler ultrasonic studies showed persistent lack of flow through the vein of Galen aneurysm, and the last angiogram obtained in April, 1985, showed no shunting through the fistula (Fig. 2 lower).

**Case 2**

This 4-year-old girl was referred to our institution with a known vein of Galen malformation following a subarachnoid hemorrhage (SAH) (Fig. 3). She had been treated at 6 weeks of age with a VP shunt for hydrocephalus, and was placed on a course of phenobarbital for a generalized seizure disorder. She had had a presumed SAH at the age of 2 years and a bout of status epilepticus at 3 years old. She recovered from the most recent SAH, and cerebral angiography demonstrated a deep midline angiomatous malformation draining into a large vein of Galen. The patient underwent transtorcular wire embolization on two occasions in June, 1984. Doppler ultrasonic studies showed absence of flow in the vein of Galen, and angiography demonstrated a persistent angioma, but with marked changes in the rate and pattern of flow. Clinically, the child is well with only an occasional left temporal-lobe seizure.

**Table 1**

<table>
<thead>
<tr>
<th>Case No.</th>
<th>IVP/SSSP* (mm H2O)</th>
<th>Blood Flow† (cm/sec)</th>
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<tr>
<td></td>
<td>Aneurysm</td>
<td>Torcular</td>
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<tr>
<td>1</td>
<td>1: 90/450</td>
<td>1: 205</td>
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<tr>
<td></td>
<td>2: 80/20</td>
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<td>2</td>
<td>1: 110/500</td>
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<td></td>
<td>2: 100/0</td>
<td>3: 0</td>
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* Superior sagittal sinus pressures (SSSP) in both patients were very high preembolization (1) but fell to nearly zero after the first procedure (2). In contrast, intraventricular pressures (IVP) remained within the normal range.
† Blood flows was measured by Doppler ultrasonic studies preembolization (1), 1 week after the last embolization (2), and 6 months after the last embolization (3).
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**Fig. 2.** Case 1. *Upper Left:* Preembolization carotid angiogram, lateral projection, demonstrating filling of the vein of Galen aneurysm (An), ectasia of the straight sinus (s), and a high position of the torcular Herophili (T). The major feeding vessels were the posterior cerebral arteries bilaterally. The right posterior cerebral artery (pca) is opacified in this instance. The angiographic architecture demonstrates a minimal arteriolar-capillary barrier and little or no vascular matrix, characterizing this lesion as mainly an arteriovenous fistula. *Upper Right:* Vertebral angiogram, lateral projection, 2 weeks postembolization demonstrating residual flow through the vein of Galen aneurysm, the straight sinus (s), and the torcular (t). The blood flow through the shunt is markedly reduced in volume. The aneurysm, the straight sinus, and the torcular are smaller in size. The inhomogeneous opacification of the torcular is related to partial thrombosis, which was further corroborated by computerized tomography. *Lower:* Six months postembolization, lateral angiograms of the carotid (left) and vertebral (center) arteries demonstrate complete obliteration of the fistula, the vein of Galen aneurysm, the straight sinus, and the torcular. The cerebral and cerebellar arterial filling is normal with no evidence of branch occlusions, although visualization of distal posterior cerebral vessels through the wires is difficult. The venous phase (right) of the carotid angiogram demonstrates no filling of the straight sinus (s) or torcular (t). The distal third of the superior sagittal sinus is only partially opacified. The filling defects (arrow) represent residual intraluminal clot. The major route of cerebral venous drainage is through large superficial cerebral veins (v) emptying into the cavernous sinuses.

**Case 3**

This 2-hour-old baby boy was referred to our institution *in extremis* secondary to a huge vein of Galen malformation (Fig. 4). He was severely acidotic on admission, with a pO₂ of 30 torr and a cardiac rate of 320/min. The pupils were equal and reacted to light and he had a poor suck. He had been intubated shortly after birth. Following admission, he was taken to the operating room and four Gianturco coils were placed in the vein of Galen. The flow was markedly reduced during this manipulation, and he improved dramatically with an increase in arterial pO₂, a reduction in the cardiac rate, and normalization of cardiac size as demonstrated on chest x-ray films. The child was seen by a cardiology consultant, and it was considered that no cardiac lesion existed. Over the ensuing 48 hours, however, the child’s pO₂ began to drop, the cardiac rate increased, his acidosis became difficult to control, and urinary output began to fall. Therefore, he was returned to surgery for the placement of another 14 coils within the vein of Galen aneurysm. Flow through the fistula was totally eliminated, as determined by venography.

Postoperatively, the child failed to respond to the complete obliteration of the shunt and remained with what appeared to be a persistent fetal circulation. He died 3 days after admission. At postmortem examination it was apparent that he probably had total obliteration of the vein of Galen aneurysm, with thrombosis in the vein of Galen and straight sinus. Also, it was noted that he had chronic ischemic changes in the frontal and temporal lobes of the brain. There was a small amount of SAH around the midbrain and pons. The baby had a huge patent ductus arteriosus and

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Fig. 3. Case 2. Preembolization computerized tomography section through the vein of Galen aneurysm (An). The ventricle size is mildly enlarged despite the presence of a functioning ventriculoperitoneal shunt, indicating central cerebral atrophy. The cerebral parenchyma contains extensive dystrophic calcifications (arrows) in both the periventricular and basal ganglia regions.

essentially a persistent fetal circulation. Figure 5 demonstrates the pathological specimen of the vein of Galen containing the embolic wires and thrombus.

Discussion

The pathophysiological effects of vein of Galen malformations are age-dependent and range from severe cardiac failure and cerebral ischemia in the neonate to the relatively quiescent lesion in the older child and adult. In the latter group, the clinical presentation is usually that of mass effect or SAH. In the infant, the entity usually presents with progressive hydrocephalus or seizures. However, it should be pointed out that any of these constellations of symptoms can be present in any of the age groups. Prenatally it is documented that significant injury can occur to the brain in the form of ischemia. Hydrocephalus can also be present at birth.

As Hoffman, et al., pointed out, the vein of Galen malformation differs from the more common arteriovenous malformation in the cerebral hemispheres in that the fistulous tract of the former is the principal pathological entity and the shunt itself probably produces most of the clinical findings in the various syndromes. The rationale for any treatment, therefore, seems to be directed at attenuating significantly or completely eliminating the flow through this fistulous connection.

The vein of Galen shunt most amenable to surgical intervention exists in the infant with a small number of feeding vessels. The more common racemose malformation is extremely difficult to approach surgically, especially in the neonate. The largest surgical series available for review is that of Hoffman, et al.; they reported the best outcome in the infant group, with overall survival in the 80% range. However, it should be noted that the numbers included in their study were small: surgical treatment was attempted in 54 of the 128 patients reviewed, and 26 of these patients were subsequently thought to be normal. Most of these patients were infants. Of 74 untreated patients, 50 died, 12 were described as impaired, and 12 were lost to follow-up review. The overwhelming impression of the authors was that the treated group, as a whole, did better than the untreated group.

The rationale for developing a transtorcular embolic technique for interrupting flow in these deep central high-flow fistulous disorders arose from our knowledge of the experience of several authors. Laws (unpublished data) has described a case of vein of Galen aneurysm controlled via the deposition of phosphor-bronze wire directly into the aneurysm. That patient has remained well 21 years postembolization. Poppen and Avman reported success in excising a vein of Galen aneurysm by interrupting the vein first and proceeding with the excision from that point. Thus, it seemed rational that placing embolic materials directly via a venous approach might be feasible. The concept was developed to deposit embolic material directly into the vein of Galen aneurysm in an effort to diminish the flow in this high-flow condition. It was believed that the flow itself was detrimental, and any method that would simply interrupt this flow could help the patient. It was also reasoned that any acute interruption might produce a generalized venous thrombosis with venous infarction.
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and the clinical constellation associated with this entity. Therefore, occluding-balloon emboli, with the potential for total venous outlet obstruction and perhaps embolization of the balloon, were discarded as the embolic material. Experience described in the literature and the treatment of peripheral arteriovenous malformations' stimulated interest in the use of Gianturco coils as the embolic material for vein of Galen aneurysms. It was reasoned that, in this high-flow fistulous condition, the introduction of a number of wires to alter the flow in a graded fashion could be effected.

A major consideration and concern in developing this technique was the possibility of misplacement or misdirection of wire emboli at the time of deposition. This potential complication was addressed by developing a tethering system for the emboli, which could be used to retrieve them in the event of misplacement or movement. During several of our staged embolization procedures we have noted the misdirection of the tip of the wire into either venous or arterial structures, and we were able to retrieve this wire without complication.

As initially designed, the transtorial approach to vein of Galen aneurysms in the neonatal patient had as its main goal the ability to alter the flow acutely and rapidly in order to improve the survival rate in this poor-outcome group. The procedure itself is extremely quick and simple to perform and, as we have seen in our one neonate (Case 3), can rapidly produce dramatic changes. Unfortunately, we were forced to totally occlude the vein of Galen and straight sinus over a 2-day period in this neonate because of persistent high-flow shunt symptomatology. The clinical outcome is obscured by the fact that this patient had a persistent fetal circulation, which may have represented a major clinical response to our initial embolization. We think it is very important to consider this peripheral shunt as a potential problem when treating vein of Galen malformations in the neonate. Also, too rapid occlusion of the cranial shunt may lead to venous infarction in the brain. It is significant that, at the time of postmortem examination in this child, major gliotic and atrophic changes were apparent in both frontal lobes and temporal lobes. This occurrence may portend a poor prognosis in the neonate even if the shunt is treated early.

Of the two long-term survivors of this technique, one has been cured radiographically and the other demonstrates the persistence of a very small shunt around the midbrain. Seizures persist in both of these patients, but are controlled medically. Both children have demonstrated progressive development at an increased rate from their preembolization course.

It is too early to compare the results of this new technique with those obtained with surgery or with the newer combined approach of Berenstein and Epstein. However, as demonstrated in this report, the transtorial deposition of embolic wires is technically feasible, can result in progressive thrombosis of a high-flow fistulous condition, and can produce rapid metabolic improvement in the gravely ill neonate harboring a high-flow vein of Galen shunt.

References


Manuscript received August 8, 1985.