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Vein of Galen Malformation: Correlation of Clinical Presentation, Arteriography, and MR Imaging

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To test the value of currently proposed angiographic categorizations of vein of Galen malformations and a hypothesis regarding the causes of vein of Galen malformations and of accompanying hydrocephalus, as well as to assess the relative utility of MR imaging and CT in clinical evaluation, we reviewed the clinical and radiologic records of 34 patients with vein of Galen malformations. Patients were divided into two groups on the basis of the angiographic demonstration of either an arteriovenous malformation nidus or a direct arteriovenous fistula to the wall of the vein of Galen or one of its tributaries. Patients with such a nidus ($n = 17$) could be distinguished from those with arteriovenous fistulas alone ($n = 17$) on the basis of age at presentation ($p < .01$) and presenting symptoms. Venous constraints, thought to be etiologically important, were identified in 31 of 34 patients. The presence or absence of hydrocephalus was explainable by mass effect in only 24 of 32 patients. In seven of 32 cases, no obvious mass effect was seen in the presence of hydrocephalus, but arteriographic evidence of venous hypertension was present in all patients with hydrocephalus. MR provided improved depiction of both arterial and venous anatomy as compared with CT. Parenchymal abnormalities were uncommon. No patients had subarachnoid hemorrhages.

We conclude that MR is superior to CT in the clinical evaluation of vein of Galen malformations, that the angiographic finding of a nidus separates patients with vein of Galen malformations into clinical and therapeutically relevant groups, and that simple mass effect on the aqueduct is not an adequate explanation for all cases of hydrocephalus in patients with this disease.

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Vein of Galen malformation is a term used to describe a heterogeneous group of patients with an uncommon congenital anomaly of the cerebral circulation. In vein of Galen malformations, enlarged deep venous structures of the galenic system are fed by abnormal arteriovenous communications in the midline [1-4]. These may be caused by single or multiple arteriovenous fistulas (AVFs) or may be part of a more complex pial arteriovenous malformation (AVM) draining into the deep venous system [5]. In virtually all of these cases some type of venous outflow restriction, thought to be etiologically important, has been present [3].

Recent advances in endovascular therapy have produced a resurgence of interest in this lesion [3, 6-9]. Parallel developments in noninvasive imaging techniques including CT [10] and more recently MR imaging have prompted a reassessment of the optimum methods for diagnostic evaluation before and after endovascular or surgical management of these patients.

We report our experience with the diagnostic evaluation of vein of Galen malformations in order to assess the validity of the venous constraint hypothesis and the clinical relevance of the morphologic categorization of patients by angiography. In addition, we analyze the ability of noninvasive cross-sectional imaging techniques to predict those angiographic findings most crucial in the prognosis and treatment planning in these patients.

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Materials and Methods

Clinical and radiologic records of 34 consecutive patients with angiographically diagnosed vein of Galen malformations were examined. Analysis was retrospective and was performed by two observers whose consensus opinion was arrived at under nonblinded conditions with respect to other imaging findings. Clinical records were subsequently studied for age at clinical presentation, presenting complaints, and clinical course. Imaging studies at the time of initial presentation were used for analysis when available; when these were not available, studies prior to surgical or endovascular intervention were analyzed.

Each angiogram was analyzed for the following features: vessels injected, presence or absence of a nidus, presence and location of fistulas, identity of feeding vessels and draining veins, presence or absence of anomalous venous drainage or venous outflow constraints, presence of arterial or venous aneurysm, mass effect, and embolic material. Associated cerebral anomalies were recorded. Heterogeneous angiographic techniques were used, ranging from aortic arch injection to superselective cerebral vessel catheterization.

A "nidus" was defined angiographically as a tangle of vessels of varying caliber distinct from the feeding arterial and draining venous structures (Fig. 1B). Usually, the presence of a nidus was associated with a slight delay in venous opacification, in contrast with the immediate venous opacification seen in direct fistulas (Fig. 1A). Any vessel larger than our expectation of normal was considered to participate in feeding or draining the malformation, unless it was clearly determined to supply or drain an unusually large territory of normal brain or dura and other vessels supplied the lesion.

Venous constraints were identified in several ways. Either an absolute anatomic narrowing of a normal-sized venous structure was present or a focal narrowing in an enlarged venous structure was observed. In the latter case, relative venous constraint was said to be present. In other cases, expected venous structures were absent and embryonic or collateral pathways of drainage were seen. In ambiguous cases, in which an abnormal venous drainage pattern was suspected but could not be absolutely confirmed, we labeled the vascular structure as normal and identified it according to its anatomic location. Aneurysms were defined as focal dilatations of vascular structures on the arterial side or within the malformation. Venous hypertension was diagnosed when we identified reflux into venous structures not typically opacified with a given injection, or when retrograde venous flow was observed.

Because patients were referred from a variety of institutions over

a long period of time, CT scans were obtained with a variety of instruments. Slice thicknesses ranged from 3 to 10 mm; scans with and without IV contrast enhancement were reviewed.

Each CT scan was analyzed for the following features: presence or absence of abnormal lucency of cerebral white matter or cortex, infarct, cortical or white matter calcifications, cerebral atrophy or hydrocephalus, presence and location of the nidus or fistulas, identity of arterial feeders and draining veins, presence or absence of anomalous venous drainage and venous outflow constraint, arterial or venous aneurysms, mass effect, and embolic material. Associated cerebral anomalies were recorded. Bifrontal white matter lucencies were considered normal in the neonate. Cerebral atrophy was diagnosed when the peripheral subarachnoid space and/or cortical sulci were prominent. Hydrocephalus was defined as enlargement of lateral ventricles in the presence of enlarged head size or enlarged ventricles and normal head size with normal or diminished sulcal pattern.

A nidus was defined as a cluster or collection of dense serpentine structures on a contrast-enhanced CT scan or occasionally on an unenhanced CT scan. Enlarged vessels were presumed to represent feeding arteries or draining veins. The identity of vascular structures was determined by their anatomic location. Any vessel situated between the nidus or fistula and an enlarged major arterial trunk or primary branch was presumed to represent a feeding artery whose identity was determined by anatomic location. Venous constraint was diagnosed when there was an abrupt decrease in caliber of a venous structure between CT slices or when a focal constriction was identified within a single slice and both the proximal and distal portions of the vein were within the same slice. Focal dilatations of vascular structures were noted. Mass effect due to vascular structures was diagnosed when the normal anatomic configuration of a neighboring structure was altered by an enlarged vessel in direct contact with it.

Because patients were referred from a large number of institutions over a long period of time, MR scans were obtained at field strengths ranging from 0.3 to 1.5 T with a variety of T1- and T2-weighted spin-echo techniques. Gradient-echo techniques were used occasionally.

MR scans were evaluated for the presence of abnormally high or low signal in the white matter or cortex, cerebral atrophy, and hydrocephalus. The presence and location of an AVM nidus or AVFs, presence and identity of feeding arteries and draining veins, and presence of a venous anomaly or venous constraint and arterial or venous aneurysms were analyzed. Mass effect, embolization material, and associated anomalies were recorded. A nidus was defined as a cluster or tangle of signal-void areas connected with tubular

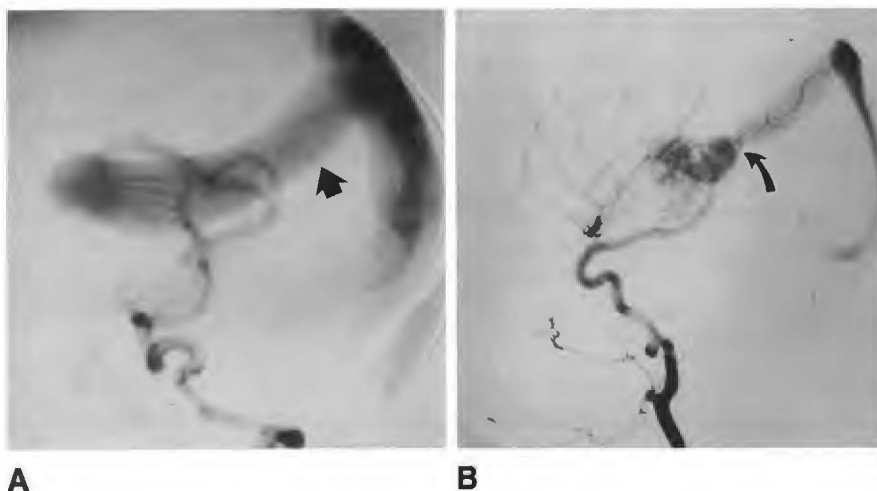


Fig. 1.—Arteriographic distinction between nidus and fistula patients.

A, Vertebral arteriogram shows multiple choroidal and thalamoperforate arteries with direct fistulas to vein of Galen malformation. Note absence of straight sinus and presence of accessory falx sinus (arrow).

B, Carotid injection. Multiple choroidal and thalamoperforate vessels contribute to nidus of thalamic arteriovenous malformation. Note venous narrowing (arrow), absence of straight sinus, and presence of falx sinus.

structures representing enlarged vessels. Criteria for identification of vessels, anomalies, and mass effect were similar to those used for CT. In evaluating parenchymal abnormalities, changes in the MR appearance of the maturing brain were taken into account.

A group of 16 patients examined by MR was separated for further comparative analysis of cross-sectional imaging studies. In each case angiographic findings were considered the reference and the CT and MR findings were judged accurate to the extent that they agreed with angiographic findings. For purposes of analysis major arterial trunks were defined as the carotid, vertebral, or basilar artery; primary arterial branches were the middle, anterior, and posterior cerebral arteries; and secondary branches included the lenticulostriate, choroidal, or thalamoperforating vessels. Angiographic reference was not used for hydrocephalus, parenchymal abnormality, or mass effect produced by enlarged vascular structures even when these diagnoses could be suspected angiographically, since cerebral parenchyma and ventricles were not directly visualized by angiography.

Results

Clinical records and angiograms were available in all 34 patients (21 boys and 13 girls). CT was performed in 32 cases and MR in 16. Both CT and MR were performed in 14 patients. The sex distribution was similar for the fistula and AVM groups. Respectively, five of 17 and eight of 17 patients in the two groups were female (not significant by binomial distribution).

Patient Groups

Patients were divided angiographically into two groups: those in whom only a direct AVF (Fig. 1A) was present and those in whom a nidus (Fig. 1B) was identified. There were 17 patients in each group. The nidus was located in one or both thalami in 16 of 17 cases. In one case part of the nidus lay in the retropulvinar cistern.

Angiographic differentiation of these two groups had clinical significance. Both the presenting symptoms and age of onset differed between the groups. In the fistula group, the age of presentation was skewed heavily to the neonatal period, with a mean age of 4.2 months and median age of 1 month. In the nidus group, the patient ages averaged 15.3 months at presentation with a median age of 9 months. The skew of the two age distributions differed significantly ($p < .01$) (Fig. 2A).

The presenting symptoms also varied between the two groups. Among the patients with fistulas alone the predominant presenting symptoms were referable to congestive heart failure in 13 patients and to hydrocephalus in the other four. In the nidus group the presenting symptoms were evenly distributed among those referable to congestive heart failure, hydrocephalus, ophthalmologic abnormalities, and less specific causes (e.g., developmental delay). Five patients had congestive heart failure (all responsive to medical treatment), four had increased head circumference and hydrocephalus, two had ocular findings, and six had nonspecific symptoms (Fig. 2B).

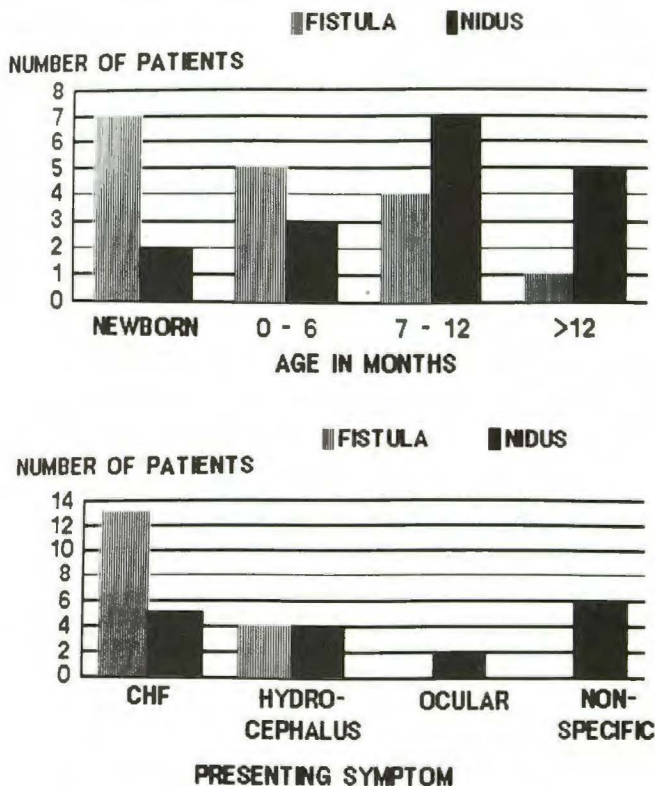


Fig. 2.—Clinical distinction between nidus and fistula patients.

A, Bar graph shows differences in age at presentation between patients with arteriographically demonstrated malformation nidus and patients with fistulas alone.

B, Bar graph shows differences in presenting symptoms between nidus and fistula patients. CHF = congestive heart failure.

Vascular Anatomy

Venous outflow constraint was identified angiographically in 31 of 34 patients. In the nidus group, venous outflow constraint was identified in 15 of 17 patients; in the fistula group, 16 of 17 patients exhibited this finding. The most frequent sites of venous outflow constraint were the distal vein of Galen and straight or inferior sagittal sinus (Fig. 3A). In each case in which a venous constraint was identified angiographically the outflow of the vein of Galen was involved. In the majority of these cases an accessory or inferior sagittal sinus was identified (Fig. 3). This was seen in 11 of 17 nidus patients and in 12 of 17 fistula patients. Complete absence of the straight sinus was noted in eight of the fistula patients and in eight of the nidus patients.

Arterial supply was generally via enlarged thalamoperforating and choroidal arteries. Lenticulostriate branches of the anterior and middle cerebral arteries occasionally contributed supply. Direct fistulas from the pericallosal artery to the vein of Galen were also observed occasionally.

The dominant supply to the fistula in all 17 patients was provided by the posterior choroidal vessels. Other vessels supplying one or more fistulas in the 17 patients included the thalamoperforating vessels in eight, the anterior cerebral ar-



Fig. 3.—Patterns of venous drainage.

A, Venous phase of vertebral artery angiogram. Pertinent findings here are vein of Galen varix, distal vein of Galen stenosis, inferior sagittal sinus, and absence of straight sinus. Additional constraints in venous drainage are seen at transverse sinus on one side (*curved arrow*) and jugular confluence on the other (*arrowhead*). Note retrograde venous flow to temporoatrial region (basal vein of Rosenthal and deep sylvian veins) (*straight arrow*).

B, Right common carotid artery angiogram shows dilatation of vein of Galen secondary to direct fistula. Note septated accessory inferior sagittal sinus within falx (*arrow*) and absence of straight sinus.

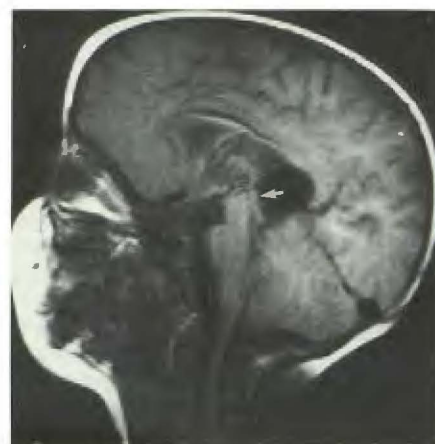


Fig. 4.—Mass effect without hydrocephalus. Sagittal T1-weighted MR image shows mass effect (*arrow*) on posterior third ventricle and tectum. Hydrocephalus was not seen on either MR or CT.

tery (including lenticulostriate and pericallosal vessels) in seven, the middle cerebral branches in two, and the anterior choroidal vessels in one. In no patients in the fistula group did the dominant supply involve lenticulostriate or thalamic vessels.

In the nidus group the distribution of vessels was somewhat different. There was a greater representation of thalamoperforating vessels, perhaps reflecting the thalamic location of the AVM nidus in these cases. In 16 cases thalamoperforators provided the dominant supply. Other vessels that contributed one or more branches to the nidus in this group of 17 patients included the posterior choroidal vessels in 16, the anterior cerebral artery in four, the middle cerebral artery in three, the anterior choroidal vessels in one, and the dural vessels in one. In contrast to the fistula group, thalamoperforating vessels dominated the supply in the nidus group, while the choroidal supply was generally less prominent.

Cause of Hydrocephalus

Of the 32 patients in whom CT was performed, 24 had hydrocephalus and eight did not. Of the 17 patients in whom only direct fistulas were identified, 11 patients had hydrocephalus and six did not. Of the 15 patients in whom a nidus was identified angiographically and in whom CT was available, 13 showed hydrocephalus and two did not. CT was not performed in two of the nidus patients. Mass effect on the posterior third ventricle was noted in 17 of the 24 patients with hydrocephalus; mass effect was not identified in the remaining seven. Of the eight patients without hydrocephalus,

mass effect on the aqueduct or posterior third ventricle was seen in one.

Thus, in 24 of the 32 patients evaluated with CT, mass effect seemed to play a role in the production of hydrocephalus. The findings in seven additional patients with hydrocephalus could not be attributed to mass effect. Furthermore, one patient with mass effect did not have hydrocephalus (Fig. 4). Therefore, another factor has to be proposed to explain the enlargement of CSF spaces. Venous hypertension was seen on angiography in all of these patients (Fig. 3A).

Cross-sectional Imaging

The ability of MR and CT to predict vascular anatomy noninvasively was assessed separately for the arterial and venous circulations. The 16 patients in whom MR was performed were used for this analysis. Cases in which only CT was performed were not considered. Of the 16 patients in whom MR was performed, eight had AVFs alone and eight had a nidus on angiography. MR correctly categorized 16 of 16 cases and CT correctly categorized 11 of 14 cases. In no case was a nidus incorrectly suspected on the cross-sectional imaging techniques and in no case was a fistula directly demonstrated. Therefore, it may be more meaningful to note that CT identified three of the six nidus patients as being distinct from the fistula group. MR identified eight of the eight patients in the nidus (Fig. 5) group correctly as well as correctly identifying the patients in the fistula group.

Identification of arterial vessels feeding the malformation was compared within the two morphologic classifications.

Fig. 5.—MR demonstration of nidus and feeding arteries.

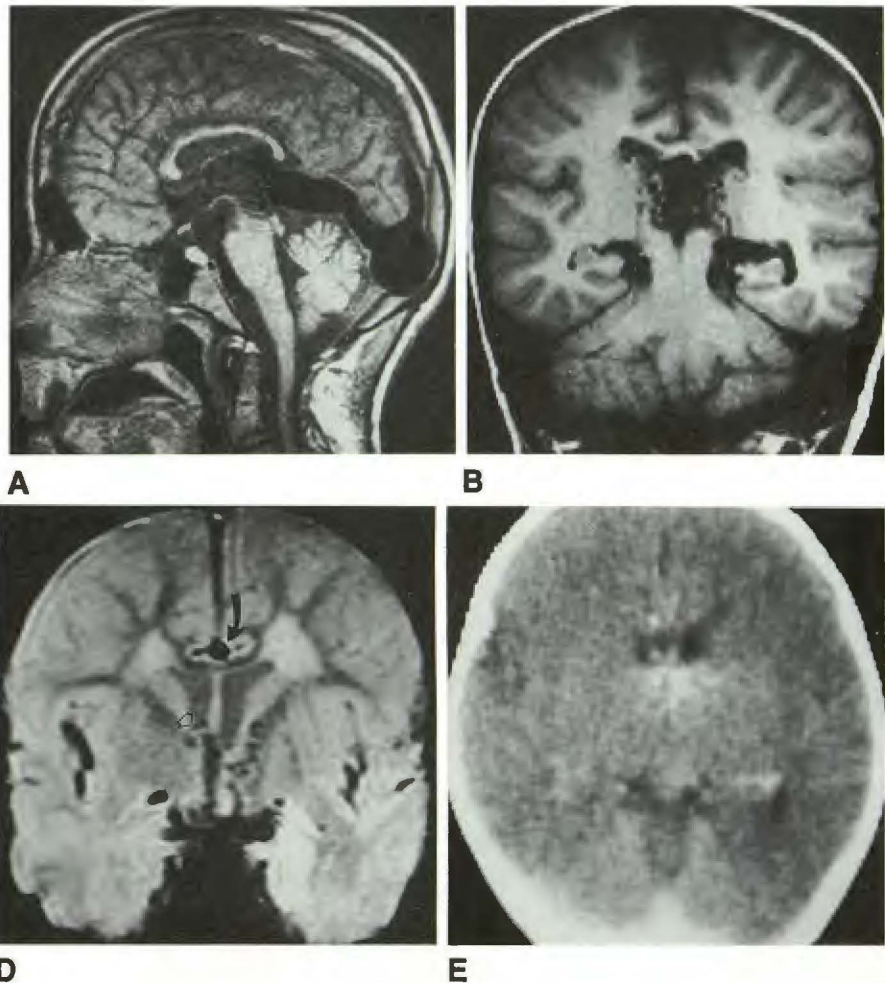
A, Sagittal T1-weighted MR image shows relative stenosis of distal midline dural venous sinus, prominent torcular confluence, and high signal in superior sagittal sinus.

B, Coronal T1-weighted MR image shows bithalamic and basal ganglionic nidus and bilateral thalamoperforating supply.

C, Axial T2-weighted MR image shows bithalamic and basal ganglionic nidus and enlarged vein of Galen.

D, Coronal gradient-refocused T2*-weighted image shows medial lenticulostriate supply, increased signal in frontal white matter, prominent anterior cerebral artery (*solid arrow*), and subependymal venous drainage (*open arrow*).

E, Axial enhanced CT scan shows confluent enhancement in thalamus. Individual vessels cannot be resolved.



This analysis was based on the angiographic recognition of the vessels. Scans were analyzed separately with respect to major trunks (internal carotid artery and vertebral artery) and primary (anterior, middle, and posterior cerebral arteries) and secondary (choroidal or lenticulostriate arteries) branches. In the nidus group, MR identified the major trunks in seven of eight cases and CT in five of six. There was greater disparity between MR and CT in the identification of secondary branches. In the nidus group MR correctly identified secondary branch groups in six of eight cases while CT identified none of six. Therefore, MR supplied more information regarding feeding vessels in six of six cases in which both studies were performed.

In the fistula group, major trunks were identified by MR in seven of eight cases and by CT in five of eight. Primary branches were identified by MR in six of eight and by CT in three of eight; secondary branches were identified by MR in five of eight while no secondary branches were correctly identified by CT. More information about the position and identity of feeding arteries was provided by MR in five of eight cases; in three of eight cases the two procedures were equal in identifying fistula feeders. In no case did CT provide more

information than MR regarding arterial feeders. Overall, MR was superior to CT in identification of arterial feeders in 11 of 14 cases and MR was equal to CT in three of 14 cases in which both studies were available (Fig. 5).

Imaging features of the venous anatomy included identification of the draining veins, significant venous anomalies, and venous constraints. MR correctly identified all significant features of venous anatomy in the nidus group in seven of eight cases; CT was correct in three of six cases. MR identified all significant features of venous anatomy in the fistula group in eight of eight cases; CT was correct in four of eight cases. Overall, MR was superior to CT in six of 14 cases and MR was equal to CT in eight of 14 cases. In no case was CT superior to MR in demonstrating significant venous anatomy. Angiography, however, had the ability to assess dynamic aspects of the venous drainage of the normal brain and its hemodynamic relationship to the venous drainage of the abnormal arteriovenous shunt (Fig. 6).

Parenchymal abnormalities and subarachnoid hemorrhage were uncommon in our series. MR and CT demonstrated findings compatible with cerebral immaturity in two cases. In one additional case, apparently pathologic cerebral immaturity



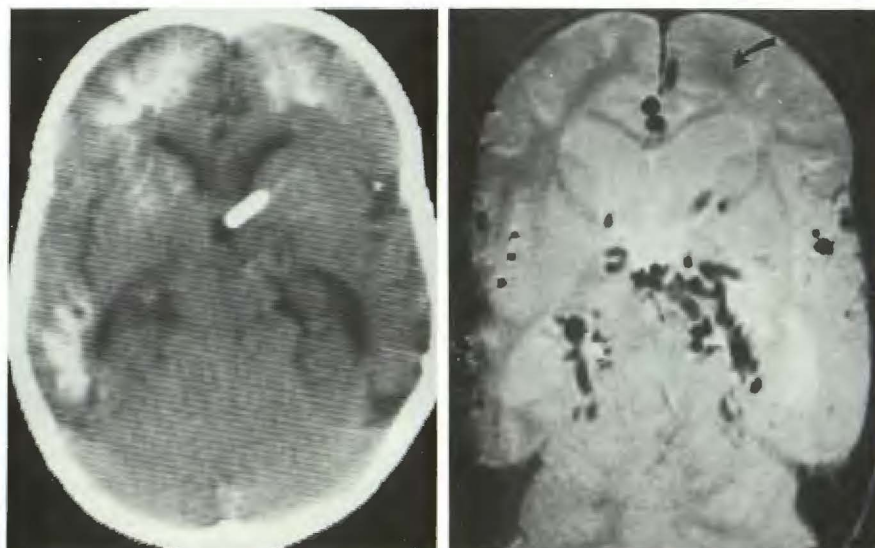
A

B

Fig. 6.—MR demonstration of venous constraint.

A, Sagittal T1-weighted MR image shows hypoplasia of corpus callosum, Chiari malformation, and stenotic straight sinus.

B, Vertebral artery angiogram shows concordance of venous findings.



A

B

Fig. 7.—CT demonstration of cerebral calcification.

A, Unenhanced CT scan shows bilateral cerebral calcifications.

B, Axial T2-weighted MR image shows no definite parenchymal abnormality. Subtle low signal corresponds to calcifications on CT (arrow).

was demonstrated on MR when CT appeared normal. Also in a single case, bifrontal white matter abnormalities evidently related to hydrocephalus were missed on CT. On the other hand cortical calcifications were missed on MR while clearly demonstrated on CT in one patient (Fig. 7). Subarachnoid hemorrhage occurred late in the illness of one of our patients and MR was not performed at that time. There were no parenchymal hemorrhages. Venous thrombosis occurred in one case and was documented on MR. CT was not performed in that case.

Discussion

Our data analysis is based on an angiographic system of patient classification that is derived from that originally proposed by Litvak et al. [1]. They divided patients into three groups: those in whom only direct AVFs were present, those in whom a racemose AVM was present, and those in whom elements of both were present. This classification was based

solely on the angiographic appearance of the lesion rather than on clinical or therapeutic considerations. We divided patients into two groups based on the presence of an AVM nidus. This separates patients into two groups for whom natural history and treatment are different.

Our classification is justified biologically by the differences in presenting symptoms and age at presentation. It is also justified therapeutically and therefore is a more useful classification. In addition, no differences in age of presentation or presenting symptoms were noted between the subcategories of our nidus group when those in whom direct fistulization plus a nidus were compared with those in whom a nidus alone was present.

We dispensed with the third, or mixed, category because the presence of nidus within the cerebral tissue precludes transcranial surgical cure with acceptable morbidity, although palliation by endovascular techniques may be effective [3, 6–8]. The therapeutic significance of the remaining two categories is then obvious: Although endovascular therapy may

be used for either group, transcranial surgery plays a major role only in the fistula group, either alone or as one element of a combined approach.

The pathophysiologic and biological significance of our patient categorization is evident from our data. The age at onset and symptoms leading to clinical presentation are clearly different between the two groups. Other proposed systems of patient classification do not seem to account adequately for the biological and therapeutic differences between the groups of patients we define [1, 2, 5, 11].

On the basis of our data, the hypothesis that mass effect on the aqueduct or posterior third ventricle is the main pathophysiologic cause of hydrocephalus may be valid in the majority of cases. However, in seven of the 32 cases in whom hydrocephalus was noted, no discernible mass effect from the enlarged draining veins was found. An alternative hypothesis must be proposed to account for these cases. The increased choroidal blood supply frequently present in these patients may contribute to an increase in CSF production. Venous hypertension may impair CSF reabsorption [10, 12–15] and thereby result in an enlargement of the subarachnoid space. This has been reversible after therapy in many of our patients [7, 8].

Increased intracranial venous pressure, with impaired CSF reabsorption and hydrocephalus, has been reported to occur in patients with superior vena cava obstruction [10] and following bilateral cervical vein ligation in dogs [23], as well as in the pediatric and adult types of dural AVFs [12] or in infants without fistulas [13]. Elevation of intracranial pressure, as well as of the pressure in the superior sagittal sinus prior to the surgical treatment of a dural AVM of the posterior fossa in an adult (which returned to normal after surgical resection), was noted by Lamas et al. [14]. Therefore, similar mechanisms can be proposed to explain the hydrocephalus in many of our patients. In addition, our series of direct AVFs of the brain without dilatation of midline structures [15] supports this hypothesis. In vein of Galen malformations, Quisling and Mickle [16] demonstrated the increased venous pressure in the inferior and superior sagittal sinuses.

The ability of MR to demonstrate vascular anatomy in a manner superior to CT is not surprising. Owing to the wide referral base and the sporadic appearance of patients with this uncommon condition and their often precarious clinical state, we were unable to perform any systematic analysis of the relative merits of particular MR pulse sequences or protocols. The MR studies reviewed here were obtained with a wide variety of instruments and techniques. Despite these problems, clear advantages of MR over CT were demonstrated.

Our recommended protocol for imaging patients with known or suspected vein of Galen malformations includes a sagittal T1-weighted spin-echo sequence and an axial T2-weighted spin-echo sequence. This may be supplemented by gradient-refocused T2-weighted sequences with good anatomic definition. If these techniques are not available, coronal T2-weighted spin-echo images are of value to supplement the axial images or may be substituted for them when the diagnosis is apparent from the sagittal T1-weighted study.

Prospective identification of venous drainage, venous anomaly, and venous constraint is clinically important. This is particularly true if a transvenous or transtocular [8] approach to the malformation is considered. Identification of an atretic straight sinus or anomalous inferior sagittal sinus will alter the operative approach in this case. The almost universal presence of venous outflow constraint in our patients suggests that venous anomalies may play a role in the formation of vein of Galen malformations.

The importance of prospective identification of the nidus by MR or CT lies in the fact that vascular access and contrast load limits the ability to study these patients angiographically. It is desirable to predict which injections will provide the maximum yield, especially when the patient's precarious clinical condition precludes complete angiographic evaluation prior to emergency endovascular therapy. This permits diagnostic as well as therapeutic angiography to be performed in the same sitting with minimum contrast load. This is especially important in the neonate with heart failure. Prospective identification of arterial supply is valuable for the same reasons.

The sole remaining advantage of CT in the evaluation of vein of Galen malformations lies in its superior ability to depict intracerebral calcifications in these patients. Since some centers rely on the appearance of calcification for some aspects of therapeutic planning, it may be reasonable to perform CT if this finding is considered clinically important. We have not found it to be a common finding and have not based therapeutic decisions on the presence of, absence of, or change in calcifications.

Angiography remains the definitive study in the complete evaluation of the patient with a vein of Galen malformation prior to transvascular or transcranial therapy. However, during the period of initial management, MR may be performed, sparing the patient a contrast load and preserving the femoral arteries for access during transvascular therapy. Angiography may be reserved as a direct therapeutic or preoperative technique. When both studies are available, follow-up may be facilitated by combined analysis.

Patients with vein of Galen malformations can be separated into two groups angiographically. This categorization is clinically relevant in terms of presenting symptoms, age of presentation, and therapeutic management and can be accomplished reliably with MR. In addition, MR demonstrates many of the features of arterial and venous anatomy important for precise endovascular therapy planning. A significant number of cases of hydrocephalus are not explained by simple mass effect. This finding may suggest future directions for management.

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