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Vein of Galen Aneurysms: A Review and Current Perspective

Michael Bruce Horowitz, Charles A. Jungreis, Ronald G. Quisling, and Ian Pollack

The term *vein of Galen aneurysm* encompasses a diverse group of vascular anomalies sharing a common feature, dilatation of the vein of Galen. The name, therefore, is a misnomer. Although some investigators speculate that vein of Galen aneurysms comprise up to 33% of giant arteriovenous malformations in infancy and childhood (1), the true incidence of this anomaly remains uncertain. A review of the literature reveals fewer than 300 reported cases since Jaeger et al's clinical description in 1937 (2). As we will outline below, our understanding of the embryology, anatomy, clinical presentation, and management of these difficult vascular malformations has progressed significantly over the past 50 years.

Embryology and Vascular Anatomy of the Vein of Galen

The development of the human cerebral vascular system is complex; a thorough analysis has been conducted by Padget (3). Cerebral vascularization begins during gestational week 4, at the time of neural tube closure. By the end of week 5, the choroidal and quadrigeminal arteries, the main afferents to the vein of Galen, are well developed (4). During week 6 the circle of Willis is completed. The anterior cerebral artery supplies the choroid plexus of the lateral ventricles, and the middle cerebral artery supplies the striatum (4). Meanwhile, at the roof of the diencephalon, the median prosencephalic vein, or primitive internal cerebral vein, develops as the main draining structure for the telencephalic choroid plexus. By week 10 the median prosencephalic vein is largely replaced by the paired internal cerebral veins, which then become the predominant means of choroidal drainage (4). Although the median prosencephalic regresses, its most caudal portion joins

the internal cerebral vein to form the vein of Galen (4).

Differentiation of the venous sinuses occurs concurrently with development of arterial and venous drainage systems. By week 4, a primitive capillary network is drained by anterior, middle, and posterior meningeal plexi (3, 4). Each plexus has a stem that drains into one of the paired longitudinal head sinuses, which in turn drain into the jugular veins (3, 4). Atresia of the longitudinal sinuses leads to the development of the transverse and sigmoid sinuses by week 7 (3, 4). At birth only the superior and inferior sagittal, straight, transverse, occipital, and sigmoid sinuses remain, along with a still plexiform torcula (3, 4). On occasion, a transient falcine sinus extending from the vein of Galen to the superior sagittal sinus is seen (4). Such sinuses represent persistent intradural channels located within the falx cerebri.

Under normal circumstances, the mature vein of Galen persists as a bridge between the deep parenchymal venous system and the venous sinuses. As such, it serves as a conduit between the internal cerebral veins, basal veins of Rosenthal, precentral cerebellar vein, vermillion veins, and straight sinus (5).

Lying within the subarachnoid space in an area known as the great transverse cleft, the vein of Galen is bordered superiorly by the free margin of the falx, posteriorly by the tentorium cerebelli, anteriorly and inferiorly by the roof of the third ventricle, and laterally by the choroidal fissures of the lateral ventricles (4) (Fig 1).

Arterial Supply

In a series of 23 patients with vein of Galen aneurysms, Raybaud et al found the posterior choroidal arteries to be the primary feeders (4). The anterior cerebral arteries were the second

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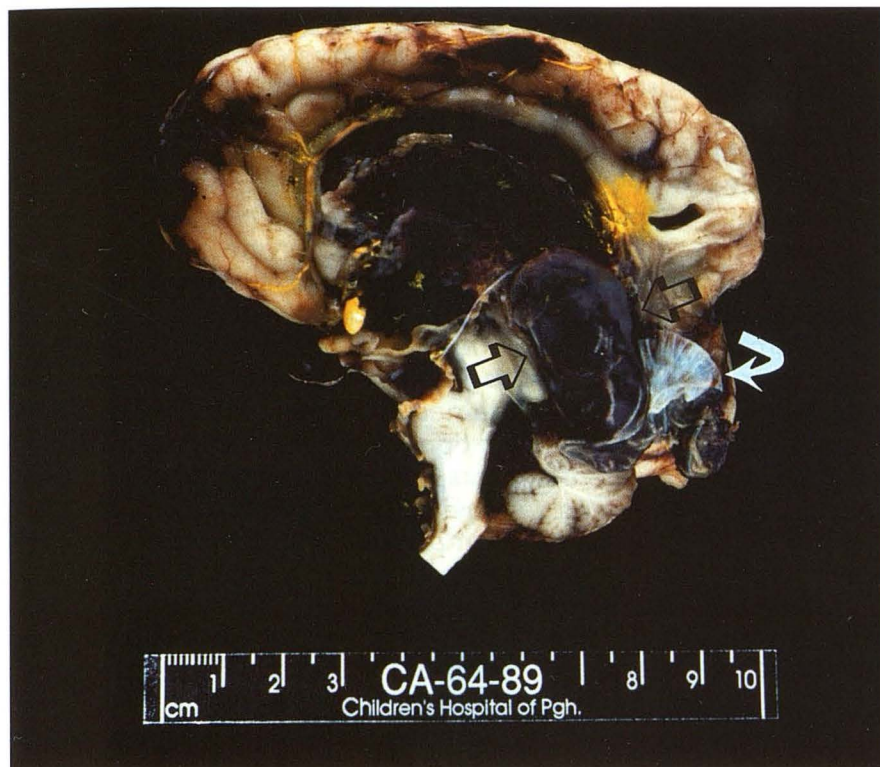


Fig 1. Midsagittal section of the brain illustrating the large thrombosed vein of Galen aneurysm (*open arrows*) overlying the cerebellum and the collicular plate. The ventricles are distended with recent hemorrhage. Multifocal subarachnoid hemorrhage is also seen. *Yellow streaks* represent a postmortem attempt at perfusing the arteries with yellow silicone. Note the small size of the brain and cerebellum. White fibrotic meninges cover the medial occipital lobe adjacent to the malformation (*curved arrow*). (Courtesy of Dr M. A. Barmada, Children's Hospital of Pittsburgh.)

most commonly involved vessels, generally providing bilateral blood supply. The anterior thalamoperforators were common secondary tributaries, joining primary afferents at the level of the choroidal fissure. Perimesencephalic vessels were constantly involved in neonates and frequently involved in older children. Distal branches of the posterior cerebral arteries and posterior thalamoperforators supplied the malformations in a moderate number of neonates. In approximately 50% of patients, the meningeal arteries were significant.

Venous Drainage

In healthy persons the vein of Galen drains the internal cerebral veins, basal vein of Rosenthal, posterior mesencephalic vein, superior vermian vein, precentral cerebellar vein, and superior cerebellar veins (5). Sixty-eight percent of Raybaud's patients had major venous anomalies including absent or interrupted straight sinuses, straight sinuses divided into two segments, and straight sinuses judged too small in relationship to the sacs (4). A small number of patients demonstrated both falcine and patent straight sinuses. An equally small number presented with falcine loops. This arrangement consisted of a falcine sinus draining the sac into the superior sagittal sinus. From the

superior sagittal sinus, blood flowed posteriorly into the torcula and then into one transverse sinus. Some blood, however, flowed anteriorly within the superior sagittal sinus, entered a second falcine sinus, which angiographically seemed to cross the first (in a separate dural sheet), and finally discharged into the torcular or other transverse sinus. In these situations blood may drain via petrosal and tentorial venous channels into the cavernous sinus. Falcine sinuses were generally associated with high-flow shunts. Five of Raybaud's cases revealed angiographic absence of straight, falcine, transverse, and sigmoid sinuses with stasis of contrast within the sacs and lack of jugular vein opacification. In terms of nonsinus venous drainage, the authors at no time demonstrated dilatation of the internal cerebral veins, although they did see retrograde flow in these structures. Large choroidal veins and an engorged subependymal system drained into the basal veins, uncus veins, and cavernous sinuses. Lateral mesencephalic venous drainage entered the transverse sinus.

Categorization

Yasargil divides vein of Galen aneurysms into four categories (6). Type 1 contains pure fistu-

las between arteries and the vein of Galen with the nidus of the lesion being the ampulla of the vein. This entire lesion is extrinsic to the brain parenchyma. Type 2 is composed of thalamoperforators that travel through normal parenchyma and supply both brain tissue and give branches to the vein of Galen. These lesions are both intrinsic and extrinsic to the central nervous system. Type 3 malformations are mixed lesions with characteristics of both type 1 and type 2 lesions. Type 4 lesions have malformations proximal to the vein of Galen aneurysms that drain into veins that then empty into the veins of Galen.

A further modification of these classifications has been provided by Quisling and Mickle, who categorized Galenic vascular malformations on the basis of several factors: nidus complexity, afferent supply, and efferent drainage patterns (7). Type 1 consists of "true" Galenic fistulas with direct arteriovenous communication via a unilateral, choroidal arterial trunk. As it approaches the vein of Galen aneurysm this trunk can be divided into as many as five smaller distal branches. When an angiomatous matrix is present it is usually less than 1 cm in greatest diameter and is typically in direct continuity with the Galenic aneurysm. Type 2 Galenic fistulas are actually "ordinary" deep arteriovenous malformations located within the thalamus and/or hypothalamus. They are supplied by the thalamoperforating arteries and drain via the superior thalamic veins and Galenic system. Arteriovenous malformation (AVM) matrix size in such cases ranges from 1 to 2 cm in greatest diameter. Type 3 Galenic vascular malformations are similar to type 1 fistulas but have significantly more afferent vascular complexity. Instead of single feeding arteries, type 3 malformations are supplied by both anterior and posterior choroidal arteries and the persistent embryonic remnants of the distal anterior cerebral arteries. The authors observed that their angiographic distinctions correlated with the clinical presentation of the lesion. For example, no patients in categories 1 or 2 had overt cardiac decompensation (7), whereas the type 3 Galenic fistulas, which as a group exhibited the most shunting, were more likely to present with high-output cardiac failure in the newborn period.

In addition to categorizing the arterial supply to the vein of Galen aneurysm, these authors also favored using a grading system for the

venous aspect of the malformation. In grade 1 venous structure, the degree of ectasia of the straight sinus is proportional to that of the vein of Galen, with both being only minimally enlarged. Grade 2 structure occurs when the vein of Galen is more dilated than the straight sinus, with both structures moderately increased in size. Grade 3 lesions demonstrate marked dilatation of both structures, and grade 4 have significant enlargement of the veins of Galen with normal, stenotic, or absent straight sinuses (7). These angiographic findings correlated well with measurements of venous pressure within the malformation. Whereas the mean venous pressure within Galenic aneurysms in individuals with venous restrictions was 40 cm of water, the pressure in patients without venous restrictions averaged 25 cm of water (3). These angiographic findings and pressure measurements were related in a logical way to a number of clinical features. For example, no patient with obstructed drainage was in cardiac failure at the time of presentation, thus confirming that some measure of cardiac protection is provided by restriction of venous outflow from the malformation. Conversely, no case in which the efferent venous pressure was less than 20 cm of water had brain calcifications, implicating elevated venous pressure in the development of the finding. Finally, grade 4 patients with severe outflow restriction were more likely to undergo spontaneous thrombosis of their malformations, which suggests that a lower flow rate is present in those fistulas that have high-grade efferent stenosis. Taken together, these observations provided the rationale for initial attempts to treat Galenic malformations via the venous route particularly in patients with high-output cardiac failure.

Developmental Theory

Raybaud proposes that vein of Galen aneurysms are not a result of dilatation of the veins of Galen, but rather a consequence of dilatation of persistent median prosencephalic veins. The evidence he cites supporting this statement includes the following: (a) the vein of Galen develops late and lacks connections to the choroidal branch of the anterior cerebral artery, which is a primary feeder in most vein of Galen aneurysms; (b) the typical vein of Galen aneurysm directly drains both prosencephalic and mesencephalic arteries in a pattern typical of the me-

dian prosencephalic vein, but the normal mature vein of Galen does not (4). Anomalous venous drainage as described in previous sections probably represents persistent fetal drainage that remains intact because it effectively deals with the high-flow system (4). Such persistent fetal drainage may prevent the development of the normal sinus system (4). Alternatively, persistent falcine sinuses may be a consequence of straight sinus occlusion early in the developmental period (4).

Lasjuanias et al have proposed additional theories concerning the development of vein of Galen aneurysms (8). They agree that these aneurysms may develop secondary to proximal angiomatous malformations. However, they also point out the frequency of sinus obstruction (especially the straight sinus) associated with vein of Galen malformations, thus speculating that increased resistance to outflow at an early stage may lead to proximal venous ectasia. These authors felt that venous agenesis was a more likely mechanism of outflow obstruction than was sinus thrombosis, because acquired thrombosis usually fails to produce subsequent vein of Galen aneurysms. Mayberg and Zimmerman, however, have reported a case of a 64-year-old man with a dural AVM, vein of Galen aneurysm, and straight sinus thrombosis (9), and contend that the straight sinus thrombosis was the primary event leading to the development of both vascular abnormalities.

Clinical Presentation

The association in neonates of intractable congestive heart failure and a cranial bruit provides the most striking manifestation of vein of Galen aneurysm; however, less fulminant modes of presentation are the norm in older infants, children, and adults. Although attempts have been made to subdivide clinical presentation on a strictly age-related basis, it is readily apparent that, between age groups, signs and symptoms overlap (10, 11). For example, in Amacher and Shillito's classification schema, group 1 consists of neonates with cranial bruits and severe congestive heart failure; group 2 consists of neonates and infants with mild heart failure who develop craniomegaly and cranial bruits within 1 to 6 months. Group 3 comprises children 1 to 12 months old with craniomegaly and cranial bruits, but no heart failure; and group 4 consists of persons 3.5 to 27 years of

age who present with headaches, exercise syncope, and subarachnoid hemorrhage (10). In addition, certain presenting signs such as visual deterioration, proptosis, seizures, hemiparesis, developmental retardation, facial vein enlargement, epistaxis, and vertigo do not necessarily coincide with age-related groupings. Nonetheless, the above classification schema provides a useful basis for categorizing patients with vein of Galen aneurysms.

Congestive Heart Failure

Congestive heart failure is the major cause of mortality and morbidity in neonates and infants harboring vein of Galen aneurysms. In severe cases, as much as 80% of left ventricular output may be delivered to the head as a consequence of the low vascular resistance within the malformation (12). Because this output returns directly to the right ventricle, right heart failure from volume and work overload may ensue. Myocardial ischemia is further promoted by decreased afterload induced by the AVM. Consequent reduction in diastolic pressures jeopardizes myocardial perfusion (8). Other cardiac anomalies reported in association with vein of Galen aneurysms include transposition of the great vessels and aortic stenosis (13, 14).

Heart failure generally presents not before, but shortly after birth. The explanation for this may reside in the fact that in utero, the placenta's low vascular resistance reduces the amount of blood, which is "stolen" by the abnormal, low-resistance cerebral shunt (12). On examination, the neonate or infant with cardiac decompensation from a vein of Galen aneurysm may manifest cyanosis, decreased peripheral pulses, and in some cases audible cranial bruits (12). In the absence of an obvious bruit, the correct diagnosis initially may be missed unless a high index of suspicion is maintained.

Hydrocephalus

The etiology of hydrocephalus is multifactorial and includes Sylvian aqueduct obstruction, resorptive blocks, hydrocephalus ex vacuo (a consequence of encephalomalacia and cerebral atrophy [15]), and abnormal transependymal cerebrospinal fluid resorption. Resorptive blocks may be attributable to increased pressure within the sagittal sinus (7, 8).

Developmental Retardation

Developmental retardation is common with vein of Galen aneurysms and is often used as an argument against offering treatment to severely affected infants. Mechanisms implicated for such damage are arterial steal, ischemia caused by compression from engorged draining veins and an enlarged vein of Galen, and increased venous pressure with subsequent venous infarction after spontaneous, partial, or complete aneurysm thrombosis (7, 8, 16). Grossman et al recognized the role of arterial steal after noting resolution of optic disc pallor in two patients undergoing vein of Galen aneurysm excision (17). They concluded that early surgery might abort ischemic brain damage in those fortunate enough to be born with normal parenchyma.

Failure to Thrive

Many neonates and infants with vein of Galen aneurysms fail to thrive. Cardiac decompensation undoubtedly plays a tremendous role in such failure. However, hypothalamic and hypophyseal dysfunction secondary to venous congestion within these structures also must be considered a potential mechanism (8).

Vein of Galen Aneurysms Thrombosis

Spontaneous thrombosis of vein of Galen aneurysms may be heralded by the development of obstructive hydrocephalus or by the onset of intraventricular hemorrhage (18–24). Heinz et al, the first to describe thrombosis of a vein of Galen aneurysm, felt it occurred in utero or during birth (20). However, later reports clearly indicated that thrombosis could take place throughout infancy and, in some cases, in adulthood (18, 25). There is no typical presentation of thrombosed vein of Galen aneurysm except the nearly ubiquitous presence of hydrocephalus and its attendant signs and symptoms.

Associated Illnesses

Both Turner syndrome and blue rubber bleb nevus syndrome have been reported to occur in conjunction with vein of Galen aneurysms (26, 27). The former involves the absence of a single X chromosome in female patients and has been associated with other vascular anomalies including coarctation of the aorta and pulmonic stenosis. The latter presents with blue, nipple-

like, compressible skin lesions composed of blood-filled venous and cavernous angiomas. Whether the association of these diseases with vein of Galen aneurysms is a chance occurrence is not known at this time. Other malformations reported in association with vein of Galen aneurysms include supernumerary digits, hypospadias, transposition of the great vessels (28), and aortic stenosis (13, 14).

Evaluation and Diagnostic Studies

Ultrasound

Ultrasound is an excellent method of screening for and evaluating vein of Galen aneurysms, both in utero and during the neonatal period (29–31). Ultrasonic demonstration of vascular pulsations helps differentiate vein of Galen aneurysms from other possible midline structures (30) (Figs 2 and 3). Color Doppler ultrasound permits the characterization of blood flow within the malformation and, although useful in delineating feeding and draining vessels, is especially valuable in evaluating the effectiveness of therapy (32, 33).

Chest Radiographs

Chest radiographs may reveal cardiomegaly with right-sided chamber enlargement, widening of the superior mediastinum, retrosternal fullness, posterior displacement of the upper trachea, and retropharyngeal soft tissue prominence caused by encroachment on this space by dilated carotid arteries and jugular veins (34).

Skull Radiographs

Radiographs of the skull are of minimal utility in the diagnosis and evaluation of suspected vein of Galen aneurysms. Occasionally they demonstrate rims of calcium corresponding to a calcified aneurysm sac (35).

Computed Tomography (CT)

CT images of vein of Galen aneurysms generally reveal round masses lying in the quadrigeminal cistern behind the posterior border of an anteriorly displaced third ventricle (Fig 4A). High density within the lesion may suggest vein of Galen aneurysm thrombosis (5, 22, 36). After administration of contrast, dense, homoge-

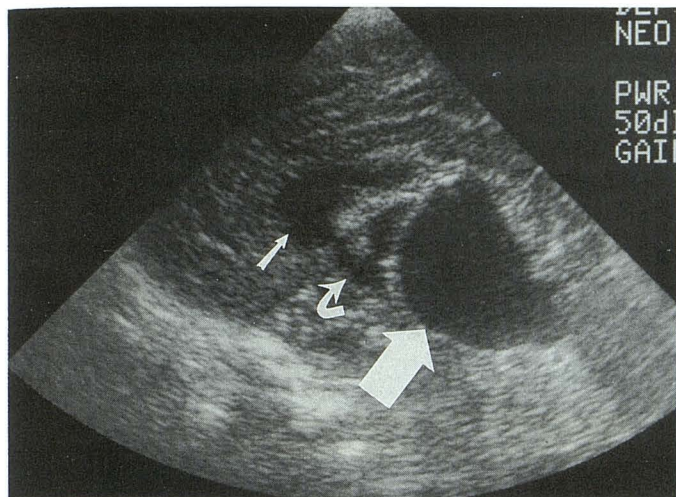


Fig 2. Sagittal-view head ultrasound revealing a dilated lateral ventricle (*small arrow*), a dilated and anteriorly displaced third ventricle (*curved arrow*), and a vein of Galen aneurysm (*large arrow*).

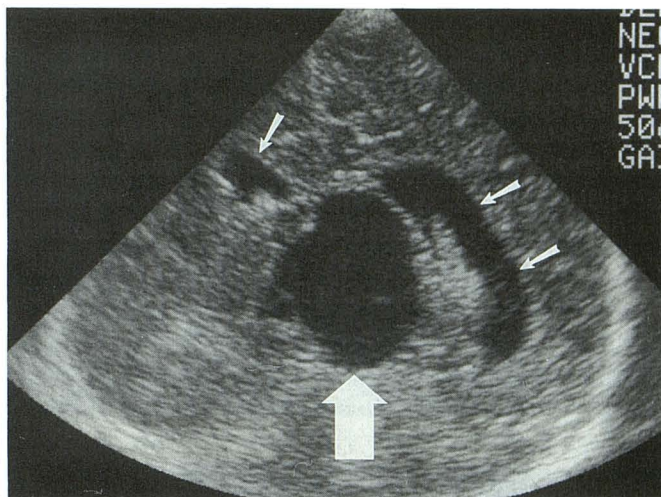


Fig 3. Axial-view head ultrasound with dilated lateral ventricles (*small arrows*) and a midline vein of Galen aneurysm (*large arrow*).

neous opacification of both the vein of Galen aneurysm and the adjacent tentorial vessels and draining sinuses is seen (5, 36) (Fig 4, B and C). When the malformation is thrombosed contrast tends to enhance the aneurysm wall and opacify small zones within the aneurysmal pouch (5, 36). A "target sign" has been described in this circumstance. Calcification of the malformation wall, seen in approximately 14% of patients, is rarely seen in patients younger than 15 years of age (18). Cerebral parenchymal calcifications are generally attributed to ischemia especially when located in watershed regions and, as such, may be an index of cerebral damage.

Magnetic Resonance (MR) and MR Angiography

The ability of MR to image noninvasively in sagittal, coronal, and axial planes makes it an invaluable tool in the characterization of arterial afferents, venous drainage, malformation position and size, and appearance of surrounding brain. MR allows the early identification of sinus abnormalities and venous drainage patterns, which not only facilitates therapeutic planning of transvenous or transtocular endovascular approaches, but also helps guide the angiographer toward the most important vessels for study (37). This is particularly critical in neonates in whom venous access is difficult, and total acceptable contrast loads are low. How-

ever, MR has not obviated the need for catheter angiography, especially when intervention is planned (38, 39). MR angiography, on the other hand, has begun to provide substantial insight into the anatomy of many vascular diseases and with improvement may become the primary tool for the evaluation of vein of Galen aneurysms.

Angiography

Angiography remains the standard of reference for the evaluation of vein of Galen aneurysms (Fig 4, D and E).

Treatment

The primary indication for treating neonates with vein of Galen aneurysms is congestive heart failure refractory to medical treatment. Surgery or endovascular treatment can in many cases be postponed by medical care until the child is older, at which point intervention is safer and easier. In those patients requiring invasive treatment, the goal of therapy is not necessarily the complete obliteration or extirpation of the aneurysms, but rather the arrest of congestive heart failure. Relative contraindications to treatment include medically controlled congestive heart failure or uncontrollable systemic failure. Finally, imaging evidence of brain damage has been considered a contraindication by some (40).

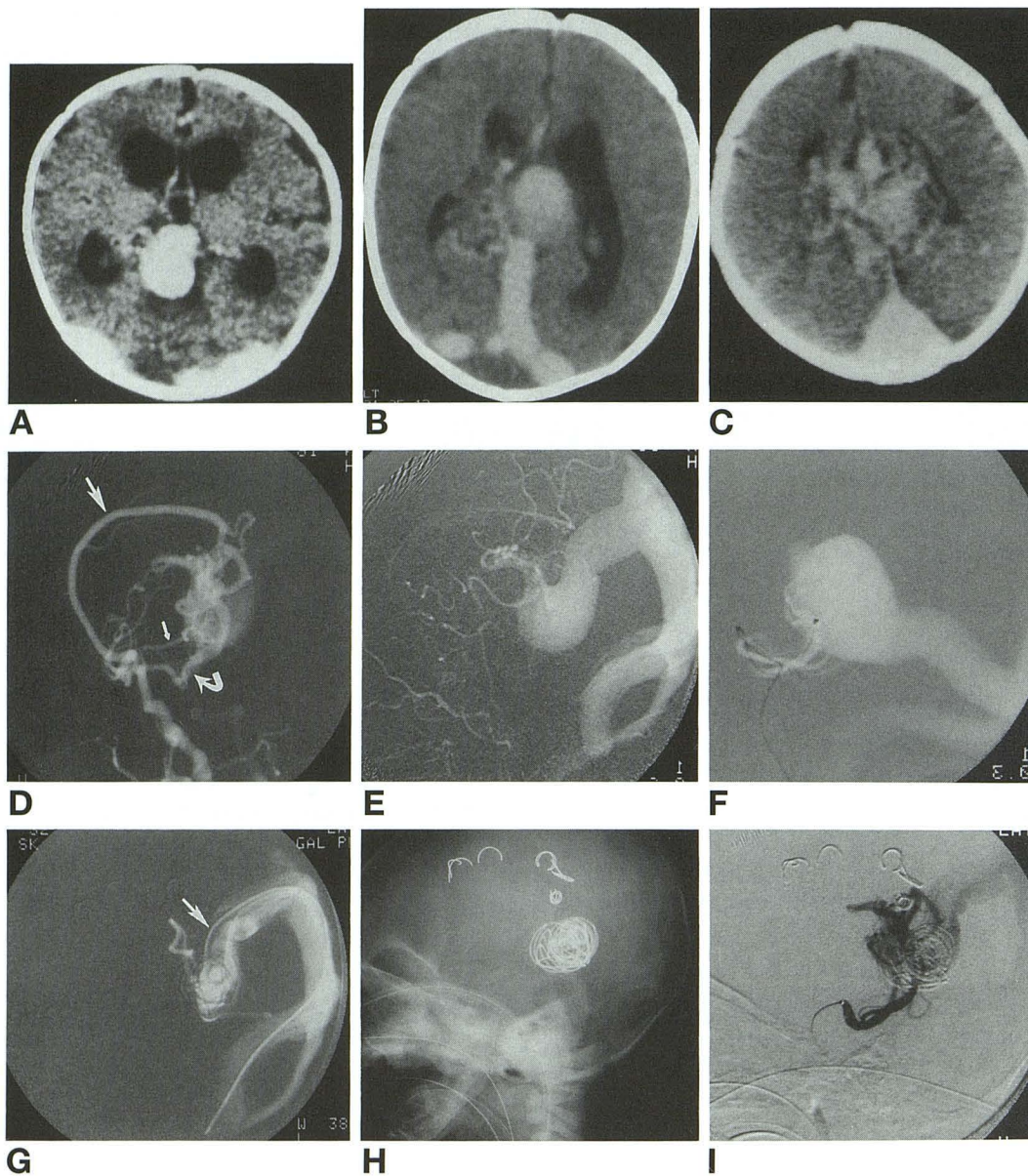


Fig 4. Neonate with a vein of Galen aneurysm.

A, Contrast-enhanced axial head CT scan revealing a 2.5-cm midline vein of Galen aneurysm anteriorly displacing the third ventricle with accompanying hydrocephalus.

B, Contrast-enhanced axial head CT in the same patient demonstrating an enlarged straight sinus.

C, Contrast-enhanced axial head CT more superiorly demonstrating the extensive surrounding vascular plexus.

D, Lateral internal carotid artery angiogram revealing the anterior cerebral artery (*large arrow*), anterior choroidal artery (*small arrow*), and posterior choroidal artery (*curved arrow*) supply to the vein of Galen aneurysm.

E, Lateral angiographic view showing aneurysm drainage via enlarged falcine, transverse, and sigmoid dural sinuses. Notice that the junction of the falcine sinus with the superior sagittal sinus is *not* at the torcula.

F, Lateral angiogram via superselective catheterization of posterior circulation. Posterior choroidal artery supply is opacified. Prominent venous drainage via the straight sinus is apparent.

G, Lateral angiogram displaying transvenous approach with the catheter (*arrow*) in the sinus system and the catheter tip in the vein of Galen aneurysm. Some coils have been deposited in the aneurysm.

H, Lateral skull radiograph demonstrating minicoils within both the vein of Galen aneurysm and the right pericallosal artery. A transvenous approach had been used for coil deposition in the aneurysm, and a transarterial approach had been used for coil deposition in the pericallosal artery.

I, Lateral view of a right posterior cerebral artery angiogram demonstrating residual posterior choroidal artery supply to the vein of Galen aneurysm. Endovascular coils have been deposited in the aneurysm and the anterior pericallosal artery. The flow through the malformation has been decreased sufficiently by the coils to resolve the congestive heart failure in this infant.

Surgery

Jaeger et al performed carotid ligation in an attempt to control the degree of blood flow through a Galenic malformation (2). This procedure provided no clear protection against further cardiovascular compromise and eventual death. Subsequent to that initial case report, numerous other reports have described surgical approaches to Galenic malformations (6, 11, 15, 41, 42). Certain issues must be considered before undertaking any planned ablative procedure. The delicacy of the poorly myelinated neonatal brain makes the retraction necessary to visualize and treat the vein of Galen aneurysm adequately and its arterial supply potentially more dangerous than in older children. Moreover, the risk of life-threatening blood loss is accentuated in such patients because of their limited blood volumes. Accordingly, where feasible, every attempt should be made to treat such lesions nonsurgically in this age group with the thought that if surgery is required, it can be undertaken more safely in an older patient.

Theoretical risks of normal perfusion pressure breakthrough must be considered when planning any treatment of vascular malformation. After obliteration of some vascular malformations, previously hypoperfused peripheral brain tissue has increased blood flow. Sudden increases in blood flow to these regions has been reported to result in brain swelling, hemorrhage, and seizures (43, 44). Normal perfusion pressure breakthrough has been associated with malformations greater than 4 cm in largest dimension, angiograms showing steal, and individuals with possible steal-related neurologic deficits (45). Many surgeons and interventionalists, therefore, feel that staged treatment of complicated Galenic malformations should be considered, especially when the above-mentioned risk factors are present.

The need for and timing of ventricular drainage in conjunction with primary treatment of vein of Galen aneurysms remains controversial. Schneider et al have described a number of complications associated with ventriculoperitoneal shunts in children with hydrocephalus and vein of Galen aneurysms. These included status epilepticus, intraventricular hemorrhage, subdural collections, and new neurologic deficits (46). In Yasargil et al's series two postoperative deaths were heralded by intraventricular hematomas in hydrocephalic patients who had

shunts placed at surgery. The authors felt that all patients requiring shunts should have catheters placed before malformation ablation, because after ablation the subependymal veins may become distended and thus possibly prone to rupture if a catheter is passed through the ventricular wall (42). Distended ependymal veins, however, may be present before malformation ablation, and the risks of shunt placement may be high at this time as well.

In many patients, particularly those with obstructive hydrocephalus secondary to thrombosed vein of Galen aneurysms, cerebrospinal fluid shunting is the only therapy required (21). In such patients, attempts to resect the thrombosed aneurysm sacs have been associated with unacceptable morbidity (20–23, 25, 42).

Endovascular Approaches

Advances in catheter design and embolization materials have brought neuroradiologic endovascular approaches to the forefront of therapeutic options in treatment of vein of Galen aneurysms. Berenstein et al's indications for embolization include: (a) preoperative obliteration of less surgically accessible feeding vessels in the hope of reducing surgical morbidity; (b) postoperative obliteration of smaller feeding vessels after surgical ligation of major arterial suppliers; and (c) definite therapy of vein of Galen aneurysms using a staged technique (47). Lasjaunias et al have made an important distinction in their discussions of endovascular treatment of vein of Galen aneurysms. In those patients with vein of Galen aneurysms secondary to a parenchymal or choroidal AVM, they emphasize the necessity of avoiding venous embolization. Approaches to such malformations should be from the arterial side to avoid venous hypertension and congestion and their associated morbidity and mortality (40).

Various techniques have been described for obliteration of vein of Galen aneurysms, including transarterial, transvenous, and transtocular embolization (8, 40, 47–52) (Fig 4, F–I). The specific endovascular approach has depended on the specific anatomy of the given case. Arteriovenous fistulas can be occluded on the arterial side from either a transarterial or transvenous approach using embolic agents such as coils, acrylics (cyanoacrylates), and endovascular balloons. Fistulas pose the technical challenge of depositing emboli at the site of the

shunt in such a way as to maintain their positions. High-flow fistulas are notorious for transmitting even large emboli to the venous side and to the pulmonary circulation with fatal results. As a consequence of such pass-through phenomena, venous occlusion therapy has gained popularity. In cases of multiple fistulas converging on the vein of Galen, the transvenous and transthoracic approaches have even more appeal. The goal in these approaches is to occlude the outflow in the vein of Galen, thus inducing retrograde thrombosis and obliteration of the fistulas. Many embolic agents have been used, although coils are currently the primary agents used in this approach. Complete cure is not usually required, and partial occlusion of the dilated vein frequently reverses the cardiovascular compromise (Ron Quisling, personal communication).

When approaching these lesions from a transarterial route with primary arterial embolizations, the interventionalist must be aware of the risks of normal perfusion pressure breakthrough. Mickle and Quisling tried to avoid normal perfusion pressure breakthrough by reducing blood flow 50% in stage 1 and then returning 3 to 21 days later, if necessary, to eliminate completely all residual flow (51). If total obliteration could not be achieved, embolization was aimed at stabilizing the patient's cardiovascular status in the hope of converting a neonate or infant into an older-age-group member in whom surgery or embolization has a better outcome (47).

An unusual but potentially fatal complication of endovascular therapy is a consumptive coagulopathy manifested by a precipitous postembolization thrombocytopenia that is followed by intracranial hemorrhage. The cause of such coagulation abnormalities may rest in the rapid deposition of large amounts of thrombus within the recently treated vascular anomaly. As in the related Kasabach-Merritt syndrome (53), treatment consists of clotting factors and platelet replacement.

Outcome

Johnston et al reviewed 232 cases of vein of Galen aneurysms reported in the literature before 1987 and recorded cases involving 80 neonates, 82 infants (1 to 12 months of age), 39 children (1 to 5 years), 22 children and young adults (6 to 20 years), and 22 adults (older than

20 years). Of these patients, 110 presented with congestive heart failure, 94 with increased intracranial pressure, 57 with cranial bruits, 37 with focal neurologic deficits, 26 with seizures, and 25 with hemorrhage (41). Ninety-one patients underwent direct surgical treatment; 29 received shunts and remote vessel ligation; 46 had medical treatment alone; and 79 had no therapy or no details of therapy in their reports. Overall mortality was 55.6% with 37.4% mortality for surgical cases and a 46.3% incidence of significant morbidity in postoperative survivors. Neonates had 91.4% surgical mortality, which approximated the nonsurgical outcome. The 1-to-12-month age group suffered 31.7% surgical mortality, and the older-than-1-year age group had 25.6% surgical mortality and 42.3% major morbidity. Yasargil's series consisting of 70 patients showed 67% neonatal (younger than 1 month old) postoperative mortality, 40% infant (1 to 24 months old) postoperative mortality, and 27% child and adult postoperative mortality (6). Total survival in this series of patients operated on and not operated on was 10% for neonates, 47% for infants, and 56% for children and adults. An inescapable conclusion from these studies, which include cases spanning many eras of neurosurgical, medical, and radiologic advances, is that neonates have profoundly worse outcomes than other age groups, most likely as a consequence of cardiac decompensation with resultant multisystem failure.

Fortunately, in the current era of endovascular therapy, morbidity and mortality rates have improved. Lasjaunias et al have reported 36 cases treated by endovascular approaches (78% pediatric, 22% adult) with a mortality of 13% (40). Casasco et al reported 100% survival in seven infants treated via transvenous embolization (54). Dowd et al reported transvenous approaches to the venous and/or arterial sides of vein of Galen aneurysms in three neonates who survived and were stable at 9 to 12 months follow-up (52). Mickle's experience with transthoracic embolization in 15 infants and older children and 9 neonates was favorable. Forty-four percent of the latter and 93% of the former survived (55). Lylyk et al used endovascular therapy in 28 children. Forty-five percent of those younger than 1 year of age had good outcomes; 61.5% of those ages 1 to 2 years had good outcomes, and 100% of those older than 2 years had good outcomes (56).

Summary and Conclusions

Our approach to treating a patient with a vein of Galen aneurysm is, of course, influenced greatly by the age of the patient, the clinical symptoms, and the angiographic architecture of the malformation. Therapeutic options are primarily based on whether a true AVM is present or if the malformation represents an arteriovenous fistula involving the vein of Galen. Arterial endovascular approaches, microneurosurgery, and/or radiosurgery are preferred for management of the former; the transvenous endovascular approach has become the cornerstone of treatment in the latter. The most critical group, however, is the neonates in extreme cardiovascular distress. In this case our therapeutic intervention is initially endovascular from the venous side, either transfemoral or transthoracic. The immediate goal is to increase resistance to right ventricular output. Advantages of this approach over a transarterial approach include a shorter anesthesia time, minimal fluid and/or contrast administration, and creation of a wire "basket" or "bird's nest" on the venous side that helps prevent emboli that may be deposited on the arterial side in subsequent embolizations from passing through the malformation. The transvenous approach can be easily repeated multiple times and may be supplemented by transarterial embolizations. Endovascular coils have been the mainstay for such venous embolizations. The end point of treatment is not complete occlusion of the fistula but improvement in cardiac function. Often, more than one stage is required to reach our goal.

The results in recent years have been encouraging and are to a large degree attributable to the advances in endovascular approaches. With future improved tools for diagnosis and treatment, perhaps the prognosis for this difficult malady also will continue to improve.

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