Review Article

Vein of Galen malformations: Review

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Vein of Galen malformations are unique congenital malformations of the cerebral vasculature that result in persistence and 'aneurysmal' dilatation of the venous structures. The varied clinical presentations and their distinctive and complex angioarchitecture make it important for the caring physician to understand their embryological and pathophysiological aspects. Management of these lesions – both in the neonatal period and at the time of definitive intervention, is challenging. Considering the rarity of these lesions, there are very few studies that have been able to compare the results of different techniques in the management. Continuing developments in the diagnostic as well as interventional aspects during the last two decades have radically changed the management of these lesions. Antenatal diagnosis and referral to a center with facilities for advanced neonatal cardiac care as well as for interventional neuroradiological therapy can go a long way in improving the prognosis in these children.

Key Words: Vein of Galen, Endovascular therapy, Aneurysm.

Introduction

Vein of Galen malformations (VOGMs) are rare anomalies of intracranial circulation that constitute 1% of all intracranial vascular malformations. However, they represent 30% of vascular malformations presenting in the pediatric age group.[1] These lesions are characterized by the presence of an aneurysmally dilated midline deep venous structure, fed by abnormal arteriovenous communications.

Steinheil in 1895, made the first reference to a Galenic malformation—referring to it as a 'varix aneurysm'.[2] Since then, these lesions have been variably referred to as 'aneurysms of the vein of Galen', 'arteriovenous aneurysms of the vein of Galen', 'vein of Galen aneurysmal malformations' and 'vein of Galen malformations'. The nomenclature is imprecise because the dilated venous structure that is characteristic of these malformations has been demonstrated to represent the embryonic median prosencephalic vein, and not the vein of Galen.

Though these lesions are extremely uncommon, they are of special interest to the interventional neuroradiologist because endovascular therapy has proved itself to be an effective, and often the only safe therapeutic modality available to treat these patients.

Embryology

Since VOGMs represent embryonic vascular malformations, they are associated with the persistence of vascular arrangements that are characteristic of a particular period of development. A sound knowledge of the embryology of the cerebral vasculature is essential to understand the angioarchitecture and pathophysiological features that are unique to these lesions. Raybaud and co-workers were the first to recognize that the ectatic venous structure that is characteristically seen in these lesions represented the median prosencephalic vein and not the vein of Galen itself.[3]

The development of cerebral vasculature can be divided into three stages. During the first phase of *'extraembryonal supply'*, the open neural tube is nurtured by the amniotic fluid that surrounds it. The phase of *'extrinsic vascularization'* is characterized by the presence of a highly vascularized neural crest derivative known as 'meninx primitiva,' which surrounds the neural tube. Nutrients are transported from this cellular connective tissue to the neural tube by diffusion. Small capillaries form within this tissue, which unite in the more superficial layers and form a network of arteries and veins. The third phase of '*intrinsic vascularization*' is characterized by the development of blood vessels within the cerebral parenchyma.[3,4]

The primary abnormality that is responsible for the development of vein of Galen malformations occurs after the stage of the 21-23 mm embryo (Figure 1). By that time, the primary internal carotid artery and its terminal branches—the anterior cerebral and anterior choroidal arteries have formed.

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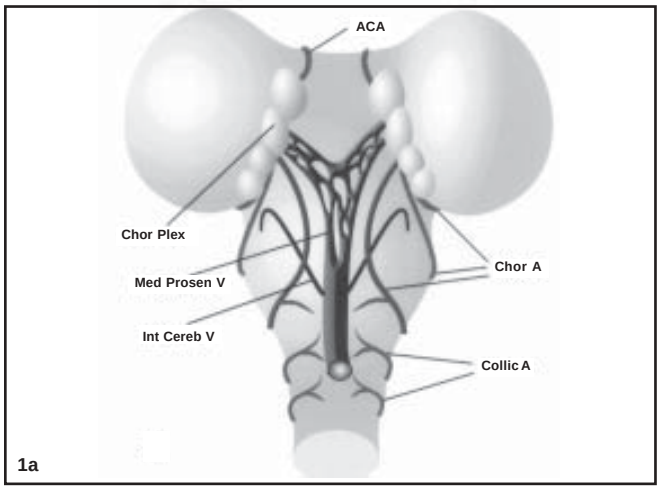


Figure 1a: Embryology of vein of Galen malformations: Dorsal aspect of the developing cerebral vasculature: The choroid plexus (Chor Plex) is supplied by the anterior cerebral (ACA) and the choroidal arteries (Chor A) and drains into the median prosencephalic vein (Med Prosen V). Later, development of the internal cerebral veins (Int Cereb V) results in the regression of the median prosencephalic vein

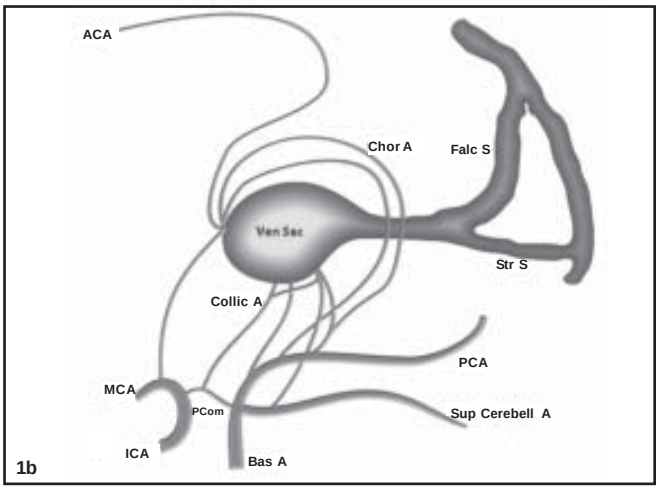


Figure 1b: Lateral aspect of vein of Galen malformation: Failure of regression caused by arteriovenous fistulous communications results in aneurysmal dilatation of the median prosencephalic vein. These fistulous communications can be supplied by the feeders from the anterior cerebral (ACA), anterior choroidal and posterolateral choroidal (Chor A), middle cerebral (MCA), the posteromedial choroidal, posterior thalamoperforating, quadrigeminal and superior cerebellar arteries (Collic A). Predominant venous drainage into the Falcine sinus (Falc S) and hypoplasia of the Straight sinus (Str S) are also depicted

Apart from supplying the developing telencephalic vesicles, these arteries also supply the area epithelialis in the region of the roof of the third ventricle, which later evolves into the choroid plexus. The internal carotid artery also supplies the hindbrain through the posterior communicating artery, from which originate the early posterior choroidal and mesencephalic arteries. Simultaneous growth of the collicular plate results in the development of the quadrigeminal arteries.[3]

Development of the telencephalic choroid plexus is accompanied by simultaneous differentiation of a transient venous structure on the roof of the diencephalon. This venous structure drains the choroid plexuses and has been designated as the median prosencephalic vein or the primitive internal cerebral vein. By the 11th week (50-mm embryo), the development of the basal ganglia results in the formation of the paired internal cerebral veins, which annex the venous drainage of the choroid plexuses. This results in the regression of the median prosencephalic vein, except for its most caudal part, which joins the internal cerebral veins to form the vein of Galen.[3]

Vein of Galen malformations arise as a result of direct arteriovenous communications between the arterial network and the median prosencephalic vein. Based on the angioarchitecture of these lesions, Raybaud and co-workers concluded that the insult causing this abnormal development occurs between the 6th and 11th week of intrauterine life.[3] The arteriovenous communications occur within the cistern of velum interpositum and the quadrigeminal cistern. The principal feeders of the malformation are those that normally supply the tela choroidea and the quadrigeminal plate. These include the anterior or prosencephalic group (the anterior cerebral, anterior choroidal, middle cerebral and the posterolateral choroidal arteries), and the posterior or mesencephalic group (the posteromedial choroidal, posterior thalamoperforating, quadrigeminal and superior cerebellar arteries).[1,3] The median prosencephalic vein, which drains the shunt, lacks a fibrous wall and is largely unsupported. It lies free in the subarachnoid space within the cistern of velum interpositum and therefore it balloons out to a large size. The high flow across the arteriovenous fistula may result in the retention of fetal patterns of venous drainage. Persistence of the falcine sinus, which is a transient embryonic structure that connects the straight sinus to the superior sagittal sinus, is one such association. Retention of fetal patterns of venous drainage could prevent development of other sinuses such as the straight sinus. Retention of the embryonic pattern of vasculature can explain the presence of several vascular anomalies that are associated with these lesions.[3,4]

Classification

There have been several attempts to classify VOGMs. The two most widely used classification systems have been provided by Yasargil and Lasjaunias. Yasargil classified VOGMs into four categories (Table 1).[5] Type 1, 2 and 3 lesions in Yasargil's classification involve a direct fistulous communication with the vein of Galen. There is no other proximal nidus. Type 4 lesions represent parenchymal arteriovenous malformations (AVMs), which drain into the vein of Galen. Yasargil described that angiographic differentiation of Type 4 lesions from the other types is based on the appearance of veins draining the AVM (internal cerebral vein, median atrial vein or basilar vein) during the arterial phase of the angiogram.

VOGMs have been classified by Lasjaunias and colleagues

into choroidal and mural types depending on the location of the fistula (Figure 2).[4,6] Multiple fistulas communicating with the anterior end of the median prosencephalic vein characterize choroidal type malformations. This type of malformations is supplied by the choroidal, subforniceal or pericallosal arteries or by subependymal branches of thalamoperforators. In mural type malformations, the fistula is located in the wall (usually in the inferolateral margin) of the median prosencephalic vein. The collicular and posterior choroidal arteries usually supply the shunt.

These types should be differentiated from the vein of Galen aneurysmal dilatation (Figure 3), which represents dilatation of a normally formed vein of Galen, secondary to outflow obstruction. In this form, the dilated venous structure drains a parenchymal AVM as well as the normal cerebral parenchyma.[6,7] These patients present in childhood or early adulthood, with features of hemorrhage, seizures and focal neurological deficits.[4]

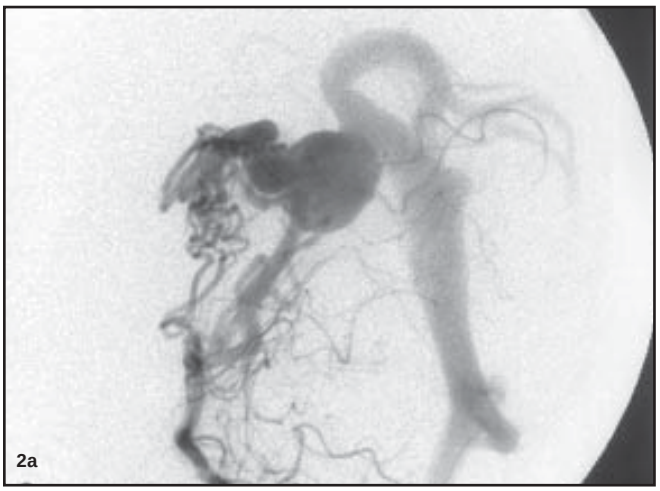
Table 1: Yasargil's classification of vein of Galen malformations

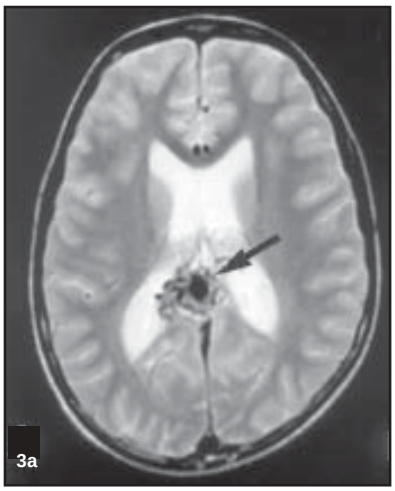
Pure cisternal fistula between pericallosal arteries (anterior or posterior), posterior cerebral artery (P4 and its branches) and the vein of Galen
Fistulous connections between the thalamoperforators (basilar and P1 segment) and the vein of Galen.
Mixed form with characteristics of both Type 1 and Type 2 lesions
Plexiform AVM with one or more intrinsic niduses within the mesencephalon or thalamus with draining veins emptying into the vein of Galen
a. Pure plexiform nidus in the parenchyma of mesencephalon or thalamus
b. Nidus within the parenchyma combined with fistulous cisternal nidus (Type 1)

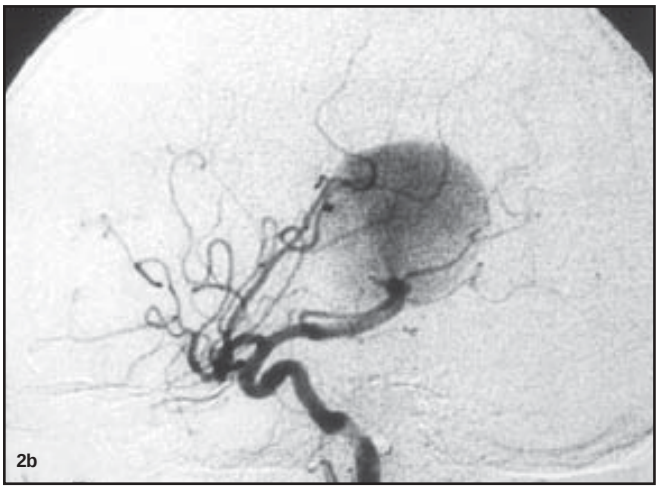
Pathophysiology

Cardiac manifestations

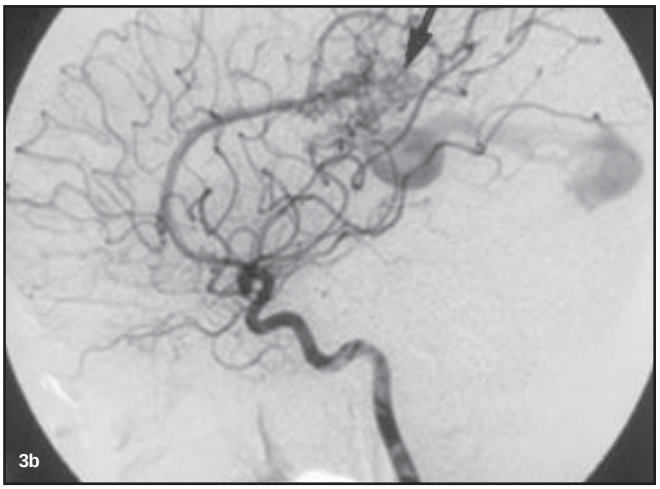
During intrauterine life, the low resistance of the placental circulation competes with the cerebral arteriovenous shunt, thereby blood flow through the shunt is not as great as it is







Figures 2a and b: Lasjaunias classification of vein of Galen malformations
a: Choroidal type of malformation (Yasargil Type II and Type III)
b: Mural type of malformation (Yasargil Type I)

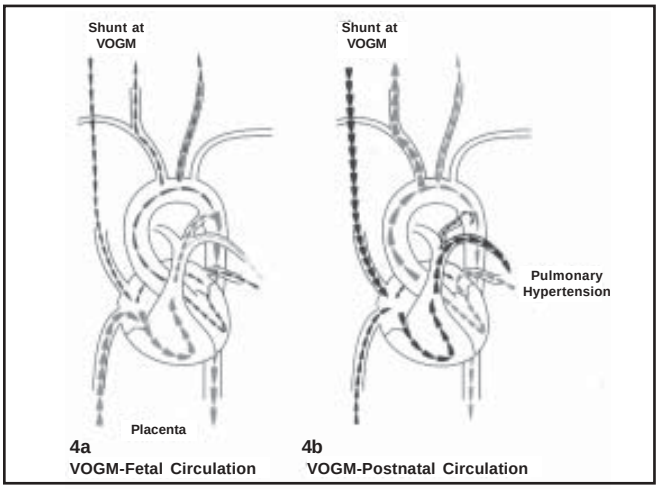


Figures 3a and 3b: Vein of Galen aneurysmal dilatation : MRI (a) and Carotid angiogram (b) in a patient with vein of Galen aneurysmal dilatation showing an arteriovenous malformation in the splenium of the corpus callosum draining into a dilated venous sac.

after birth (Figure 4a). The left ventricle supplies the fistula while the right ventricle supplies the placenta and the rest of the body. Thus, the circulatory overload is shared between the two ventricles, which work in parallel.[8]

After birth, each ventricle supplies the entire circulation in series. Thus, the burden on each ventricle increases and cardiac failure ensues. Exclusion of the low resistance placental circulation results in an abrupt increase in the flow across the fistula (Figure 4b). As much as 80% of the left ventricular output may be supplied to the brain in severe cases. This necessitates a compensatory increase in the cardiac output and blood volume to maintain perfusion of the systemic vasculature. This excessive flow across the pulmonary vasculature results in pulmonary hypertension. Increased venous return to the right atrium promotes right-to-left shunting through the patent foramen ovale. Right-to-left shunting also occurs at the level of the ductus arteriosus, which remains patent due to the rise of pulmonary arterial pressure above the systemic pressure. These right-to-left shunts are responsible for the cyanosis that may occur in these patients. Large arteriovenous shunts significantly reduce the diastolic pressure within the aorta, causing reduced coronary artery flow. The increased cardiac output results in high ventricular intracavitary pressure. Both these factors are responsible for the reduction of the subendocardial blood flow, thereby promoting myocardial ischemia.[8-10]

Thus, the cardiac failure in neonates with VOGMs is multifactorial in origin and is usually refractory to medical management.



Figures 4a and 4b: Pathophysiology of vein of Galen malformations
a: Fetal circulation in a case of vein of Galen malformation: Low resistance of the placental circulation prevents high flow though the intracranial shunt
b: Early Post natal circulation : Cessation of placental flow suddenly increases the forward flow within the intracranial shunt and across the pulmonary vasculature and may result in congestive cardiac failure. Flow across the ductus arteriosus and foramen ovale may persist. Size of the arrowhead indicates volume of flow. Large arrowhead represents high flow and Small arrowhead represents low flow. Color of arrowheads indicates oxygen saturation. Grey arrowhead represents high oxygen saturation and Black arrowhead represents low oxygen

Neurological manifestations

Cerebral venous hypertension is the etio-pathogenetic factor that is responsible for most neurological manifestations of VOGMs. These lesions are usually associated with venous anomalies in the form of poorly developed venous drainage or secondary venous stenosis and occlusion. The high flow within the arteriovenous shunt and restriction of venous drainage results in high cerebral venous pressure. In infants, as the arachnoid granulations have not yet fully matured, most of the ventricular CSF is reabsorbed across the ventricular ependyma, into the brain parenchyma, for subsequent drainage by the medullary veins. In infants with VOGMS, the high venous pressure transmitted to the medullary veins prevents resorption of fluid and thus results in hydrocephalus, cerebral edema, and hypoxia.[11] Thus, hydrocephalus is secondary to impaired resorption of CSF due to venous hypertension and not due to aqueductal compression.[12,13] The chronic hypoxia produced by the venous hypertension results in progressive cerebral parenchymal damage resulting in cognitive impairment, which can range from delayed milestones to mental retardation.[4,14]

The fistula may be drained by rerouting its blood flow into the cavernous sinus and further into the facial veins or basilar or pterygoid plexus. These collateral pathways of venous drainage account for the prominence of facial venous channels, which is commonly seen in infants with VOGMs and also for the occasional case that presents with epistaxis.[4,15]

Clinical presentation

Gold et al in 1964 provided a clinical classification system for VOGMs that remains valid today.[16] They correlated the age at presentation with the clinical presentation and pathophysiology and described three characteristic groups of patients.

Neonates

Neonates characteristically have multiple fistulas. Up to 25% of their cardiac output passes through the fistulas causing high-output congestive cardiac failure. Depending on the size of the shunt, adequacy of venous drainage, complexity of arterial supply and the host response, the cardiac manifestations can range from asymptomatic cardiomegaly to severe cardiac failure that is refractory to medical management. Cyanosis may be seen in these patients and the presentation may be mistaken for congenital cyanotic heart disease.[17] Features of myocardial ischemia may be detected on electrocardiography.[4,8,18,19]

Infants and children

Infants and children usually have a single fistula with a smaller shunt. Cardiac manifestations are absent or very mild. These patients present with macrocephaly or with hydrocepha-

lus. Patients with longstanding cerebral venous hypertension may also present with delayed milestones. A high proportion of these children present with failure to thrive. Though this could be due to cardiac decompensation, hypothalamic and hypophyseal dysfunction secondary to venous congestion must also be considered as a potential mechanism.[4,20]

Older children and adults

Older children and adults usually have low-flow fistulae. These patients usually present with headache and seizures. A small number of patients may also present with developmental delay, focal neurological deficits, proptosis and epistaxis. Subarachnoid hemorrhage and intracerebral hemorrhage can occur in this age group due to rerouting of blood into the pial veins.[4]

Imaging

The widespread use of routine antenatal ultrasonographic examinations has enabled the detection of several cases of VOGMs in the third trimester of pregnancy.[21-24] Apart from identification of the abnormality and its differentiation from other nonvascular space-occupying lesions, ultrasonography has also been used to assess the status of the fetal cardiovascular system.[25] Referral of such patients to centers where better facilities for delivery and immediate postnatal therapy are available has resulted in considerable improvement in the prognosis of infants born with this condition.

The role of imaging in a patient referred for endovascular therapy is to non-invasively demonstrate the number and site of arteriovenous fistulae, presence of thrombosis, sinus abnormalities and venous drainage patterns. This is of particular importance in neonates with VOGMs as the cardiac and other associated co-morbid conditions limit the acceptable contrast load during angiography. Obtaining as much information as possible from noninvasive imaging studies enables planning of diagnostic angiography and intervention in the same sitting. This eliminates the risk of repeated angiographic studies under anesthesia in such patients.[4,26,27,34]

The multiplanar imaging capabilities of MRI, its high intrinsic tissue contrast and its sensitivity to flow make it the modality of choice in the investigation of patients referred for endovascular therapy. Infants who have been subjected to interventional therapy can be followed up using neurosonography, while transcranial Doppler and CT or MRI can be used in the older children and adults.

Radiography

Plain radiography of the skull contributes little to the diagnosis of VOGMs beyond the demonstration of a rim of calcification within the wall of the aneurysmal sac (Figure 5). Calcification is seen in about half the patients with thrombosed VOGMS; compared to only 14% of patients without thrombosis.[27] Chest radiographs may reveal features of congestive heart failure such as cardiomegaly, widening of the superior mediastinum and retrosternal fullness. Retropharyngeal soft tissue prominence may be seen due to dilatation of the great vessels of the neck.[28]

Ultrasound

Antenatal ultrasound scans demonstrate the venous sac as a sonolucent mass located posterior to the third ventricle. Ultrasonic demonstration of pulsatile flow within it helps in differentiating VOGMs from other midline cystic lesions. Associated venous anomalies can often be visualized. Evidence of hydrocephalus and cardiac dysfunction can also be obtained on antenatal ultrasonography.[21-25,29]

In the postnatal period, Doppler ultrasonography can be used to demonstrate the hemodynamic changes associated with the malformation.[30] Ultrasound is of special significance in the follow-up of patients who have been treated with endovascular therapy (Figure 6), where progressive thrombosis of the venous sac can be demonstrated and the status of the shunt can be assessed on serial studies.[31,32]

Computed tomography (CT)

Contrast enhanced axial CT scan of the brain usually demonstrates a well-defined, multilobulated, intensely enhancing lesion, located within the cistern of velum interpositum. Dilatation of the ventricular system, periventricular white matter hypodensities, as well as diffuse cerebral atrophy are the commonly associated findings.

Features of cerebral parenchymal damage in the form of diffuse chronic ischemic changes, parenchymal calcifications, generalized cerebral atrophy and focal parenchymal infarcts are also demonstrated well on CT (Figure 7a).[4,14] Jayakumar et al described the de-novo development of cerebral parenchymal calcifications in a child with VOGM, in whom ventriculoperitoneal shunt placement was performed for hy-

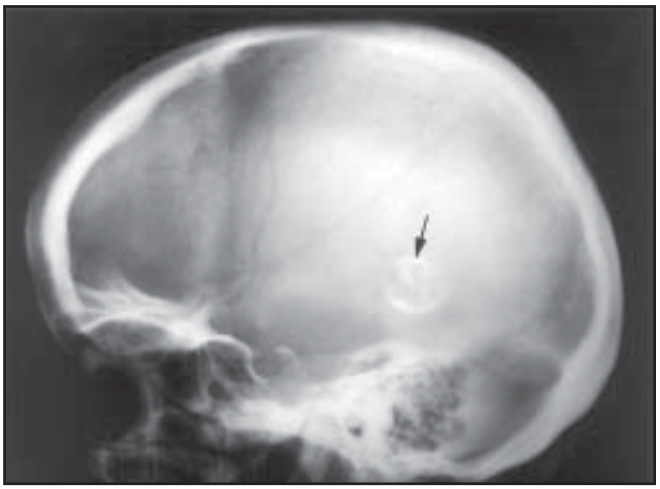
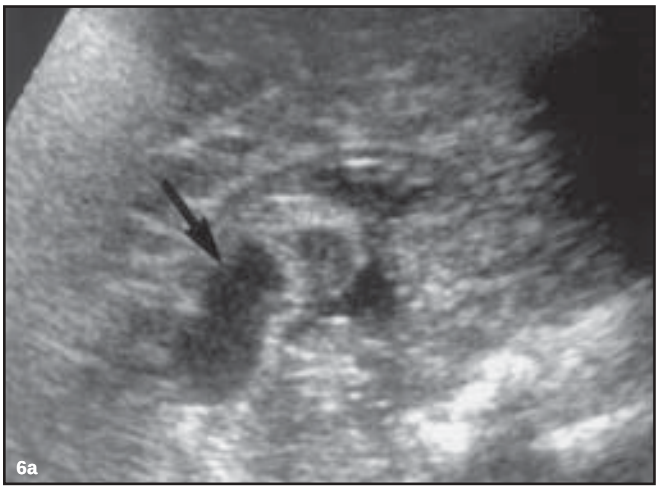
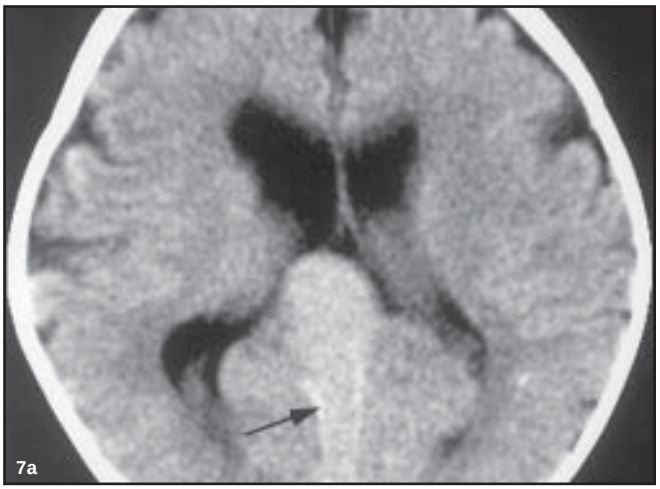
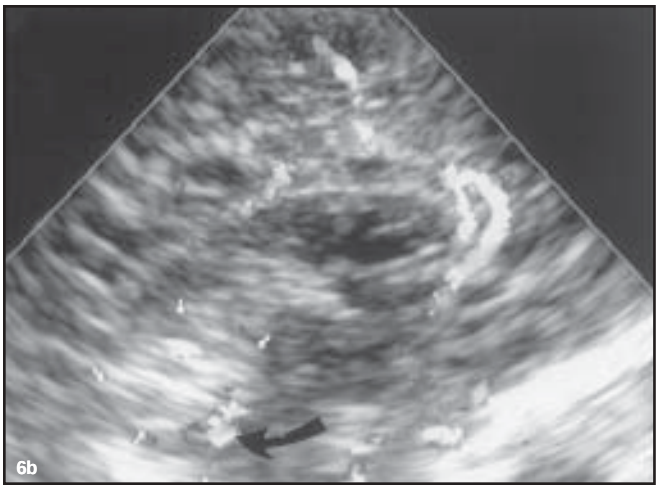
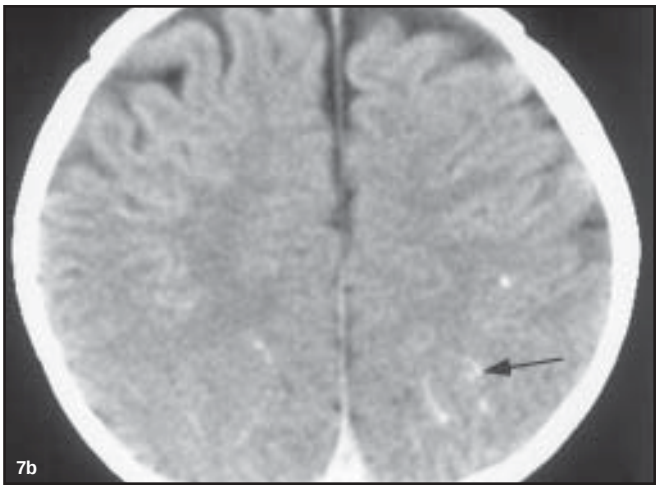


Figure 5: Plain radiograph of the skull showing calcification of the wall of the venous sac of a vein of Galen malformation









Figures 6a and 6b: Ultrasound and Transcranial colour duplex Doppler study in a child with vein of Galen malformation
a: Neurosonogram prior to embolization demonstrating the venous sac as an anechoic structure located posterior to the third ventricle.
b: Doppler study after embolization demonstrating the coils within the venous sac (white arrows) as echogenic structures. A small area of residual flow is noted in the anteroinferior part of the sac

Figures 7a and 7b: CT scan in a 3 month old child with vein of Galen malformation a: Plain axial CT scan of the brain showing a rim of calcification located along the wall of the venous sac
b: Axial section at a higher level demonstrating diffuse cerebral atrophy. Foci of parenchymal calcification are appreciated involving the grey-white matter junctions in both parietal regions.

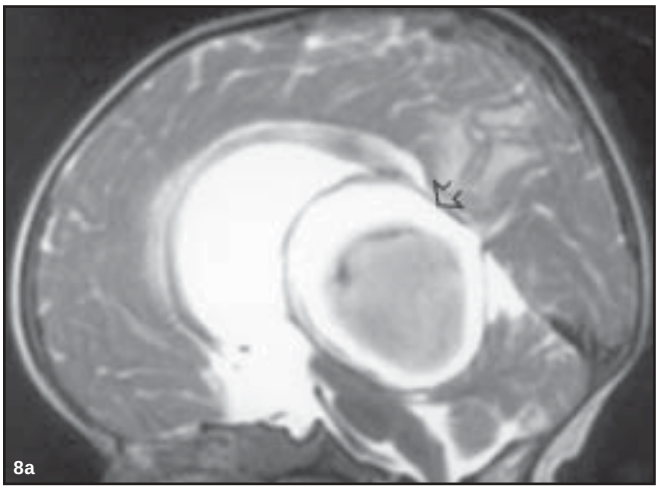
drocephalus.[33]

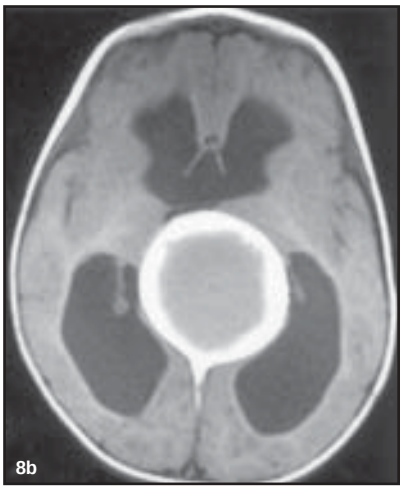
The presence of thrombosis within the aneurysmal sac can be demonstrated well on CT. Since the original description by Heinz et al in 1968, several reports of spontaneous thrombosis of VOGMs have appeared in medical literature.[35,27,36-38] The thrombus within the aneurysmal sac commonly appears as mixed hypodense, isodense and hyperdense areas due to variable maturation of the clot. On contrast enhanced CT, the presence of a central thrombus and peripheral circulating blood along the wall of the sac can produce the so-called 'Target sign'.[27] The dilatation of collateral parenchymal veins can usually be appreciated.

A crescentic rim of calcification is more commonly seen in patients with thrombosed VOGMs (Figure 7b). However, the presence of calcification has proved to be a poor predictor of the tendency of the lesion to subsequently thrombose. Calcification is rarely seen before the age of 15 years and complete calcification of the sac is extremely rare.[39]

Magnetic resonance imaging (MRI)

MRI is gaining popularity as the modality of choice for initial assessment of VOGMs. It can demonstrate the location of fistula, presence of any nidus, the arterial components, the venous sac as well as the status of venous drainage. Thrombosis of the venous sac is also depicted well on MRI (Figure 8). The position and identity of major arterial trunks, primary branches as well as secondary branches feeding the fistula are better identified on MRI than on CT. Accurate identification of draining veins, venous anomalies and venous constraints is also possible with MRI. The exquisite soft tissue contrast of MRI makes it the modality of choice in the evaluation of the ventricular system and cerebral parenchymal changes. MR angiography is being increasingly used as a noninvasive alternative to diagnostic angiographic studies in





Figures 8a and 8b: MRI of a thrombosed vein of Galen mlaformation:
a: Plain T2 weighted sagittal scan of the brain revealing the characteristic location of the lesion
b: Plain T1 weighted axial scan of the brain revealing the presence of thrombus at various stages within the venous sac

the initial evaluation of these lesions.[26,34]

Angiography

Angiography remains the gold standard for the evaluation of VOGMs. It scores over noninvasive modalities such as CT angiography and MR angiography in demonstrating small feeders supplying the fistula, as well as the dynamic aspects of the venous drainage of the normal brain, and hemodynamic relationships with the venous drainage of the arteriovenous shunt.[26,34]

VOGMs are associated with several arterial and venous anomalies. These anomalies represent the persistence of embryological vascular patterns. The limbic ring represents a persistent arterial bridge between the anterior cerebral and anterior choroidal artery (Figure 9). The absence or interruption of the straight sinus is a common association with these anomalies. Persistence of venous channels such as the

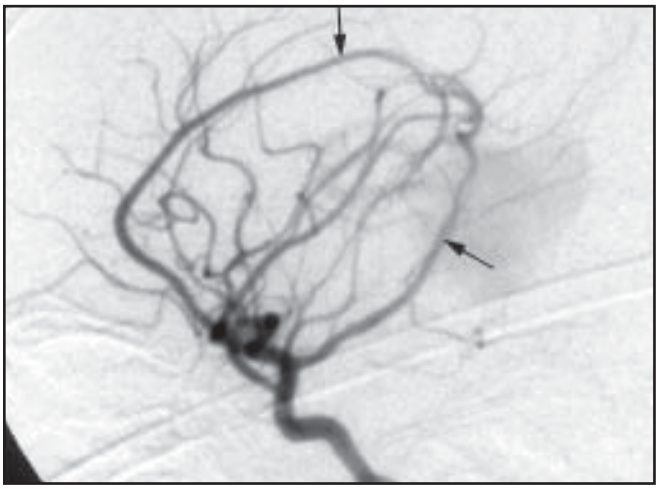
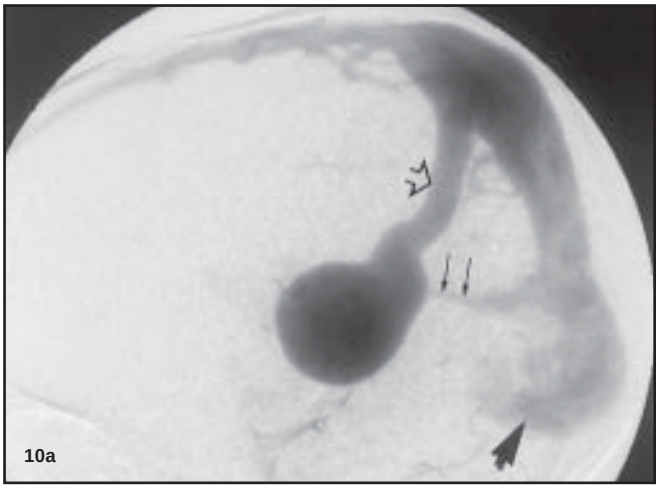
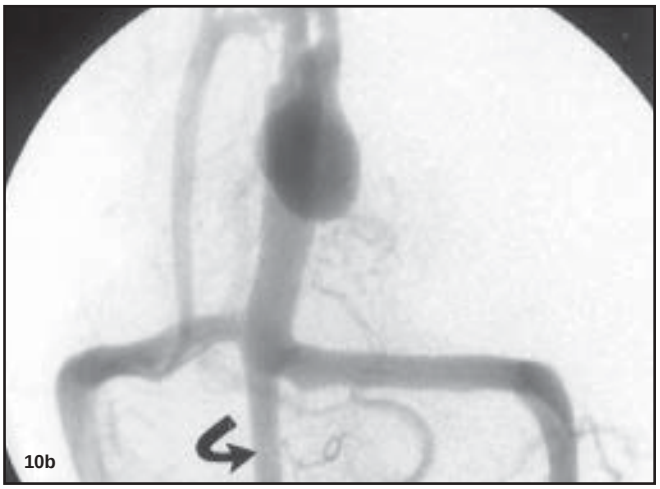


Figure 9: Intracranial arterial anomalies in vein of Galen malformations : Carotid angiogram in a patient with choroidal type of vein of Galen malformation (Lateral view) showing persistence of embryonic limbic arterial ring





Figures 10a and 10b: Venous anomalies in a patient with vein of Galen malformation: a: Venous phase of vertebral angiogram (Lateral view) revealing atresia of straight sinus and presence of falcine sinus. A large venous sac is seen at the torcular herophili. b: Venous phase of vertebral angiogram (Frontal view) in another patient demonstrating the persistent midline occipital sinus extending inferiorly from the torcular herophili

falcine sinus, occipital sinus and marginal sinus may also be seen. Atresia of transverse and sigmoid sinuses may also be associated (Figure 10).

VOGMs are also associated with the Turner syndrome and blue rubber bleb syndrome. Supernumerary digits, hypospadias, transposition of great vessels, aortic stenosis and right-sided aortic arch have been reported in association with VOGMs (Figure 11).[34,40,41]

Management

Untreated VOGMs have a very poor prognosis.[5,14,42,43] A high proportion of patients who present in the neonatal period rapidly deteriorate and succumb to congestive cardiac failure. Rapid and aggressive management of the cardiac failure is essential. Aggressive medical management can usually postpone the intervention until the child is aged about 5 – 6 months, at which point intervention is easier and safer. Emergency embolization of the malformation may be necessary to reduce the shunt in neonates with congestive cardiac failure that is refractory to medical therapy.[4,12,43]

The evolution of the management of VOGMs over time is summarized in Table 2.

Surgery

Considering the many problems associated with the management, these lesions have been termed as the 'Gordian knot' of cerebrovascular surgery. Despite technological advances in microneurosurgery, complete elimination of the lesion by surgery is rarely achieved. The problems of major cranial surgery involving a deep-seated, high-flow shunt in an infant with multiorgan failure are compounded by the poor myelination of the brain parenchyma, which tends to tear easily on retraction.[34] Similarly, ventricular shunting may worsen the cerebral venous hypertension, and should be avoided before elimination of the arteriovenous shunt. This procedure is not tolerated by infants and must be preceded by emergency embolization.[12,14]

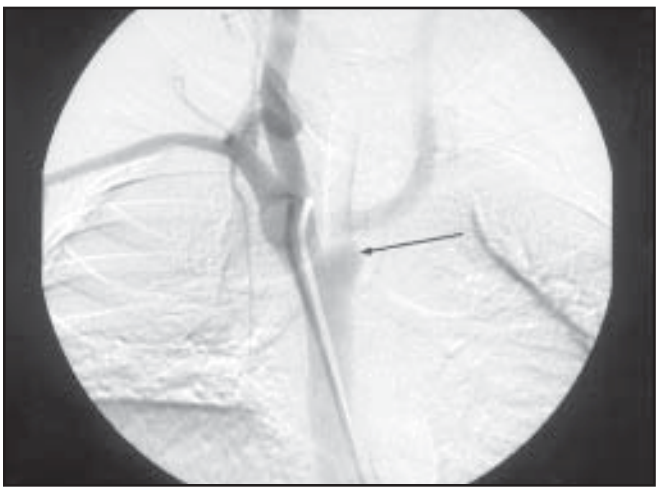


Figure 11: Extracranial vascular anomalies in vein of Galen malformations: Right sided aortic arch with mirror image branching pattern of the aortic arch vessels in a patient with vein of Galen malformation. Kommerel's diverticulum is also seen on the left side of the arch with atresia of the origin of left subclavian artery

Endovascular management

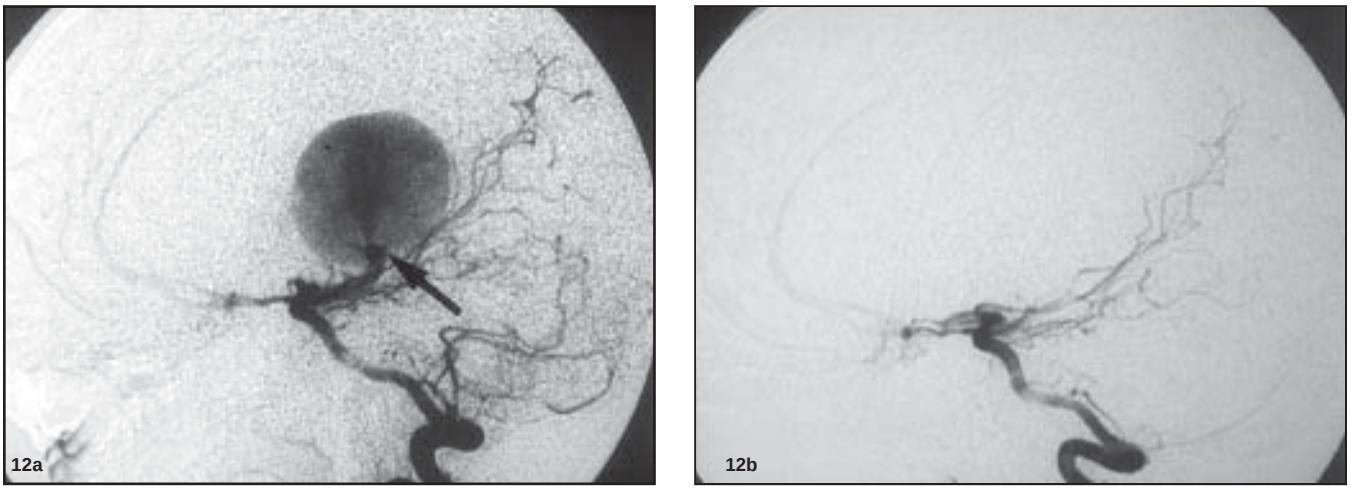
Advances in the field of interventional neuroradiology have ensured significant improvements in outcome in these patients. Several studies have documented the efficacy and safety of endovascular treatment in these patients.[46-52] The timing of endovascular management is determined by the clinical presentation. Congestive cardiac failure in a neonate that is refractory to medical treatment is an indication for emergency embolization. The goal of therapy in such patients would be to arrest the congestive cardiac failure rather than to achieve complete obliteration of the shunt. In such children, it may be acceptable to perform partial embolization to reduce the arteriovenous shunt and facilitate normal systemic and neurological development, even with the presence of a residual shunt. The procedure can be performed in a staged manner to minimize complications. In a child who has not presented with cardiac failure, the aim of endovascular therapy would be to prevent consequences of chronic cerebral venous hypertension and to promote normal cerebral development.[4,53] Treatment at the age of 5 months balances the benefits of safe embolization against the risk of cerebral damage. Imaging evidence of encephalomalacia is thus considered a relative contraindication to endovascular therapy.[11,46]

Lasjaunias and co-workers, who have the largest experience in managing these lesions, have discussed therapeutic decisions, based on the clinical expression in the neonatal period. They described a 21-point scale based on cardiac function, cerebral function, hepatic function, respiratory function and renal function. A score of less than 8 usually indicates a poor prognosis and does not warrant emergency management. A score of 8–12 is an indication for emergency endovascular management. A score of > 12 indicates a well-preserved neonate and attempts are made to delay the endovascular procedure, by medical management. The presence of failure to thrive, unstable cardiac failure or macrocrania are indications to advance the embolization.[54]

The choice of the specific endovascular approach depends on the angioarchitecture of the malformation. Arteriovenous

Table 2: Timeline for management of vein of Galen malformations[7,44,45,47]

1895	Steinhill	First description of vein of Galen malformation
1905	Ballance	Bilateral internal carotid artery occlusion
1947	Oscherwitz and Davidoff	First intracranial surgical management
1949	Boldrey and Miller	Clipping of posterior cerebral arteries supplying the fistula
1987	Lasjaunias	Transarterial embolization
1986	Mickle and Quisling	Retrograde transtorcular approach
1990	Dowd	Transfemoral venous approach



Figures 12a and 12b: Transarterial embolization of mural type of vein of Galen malformation:
a: Diagnostic angiogram reveals a single-hole fistula along the inferolateral aspect of the venous sac.
b: The feeder was superselectively catheterized and embolized with cyanoacrylate "glue" resulting in complete obliteration of the shunt

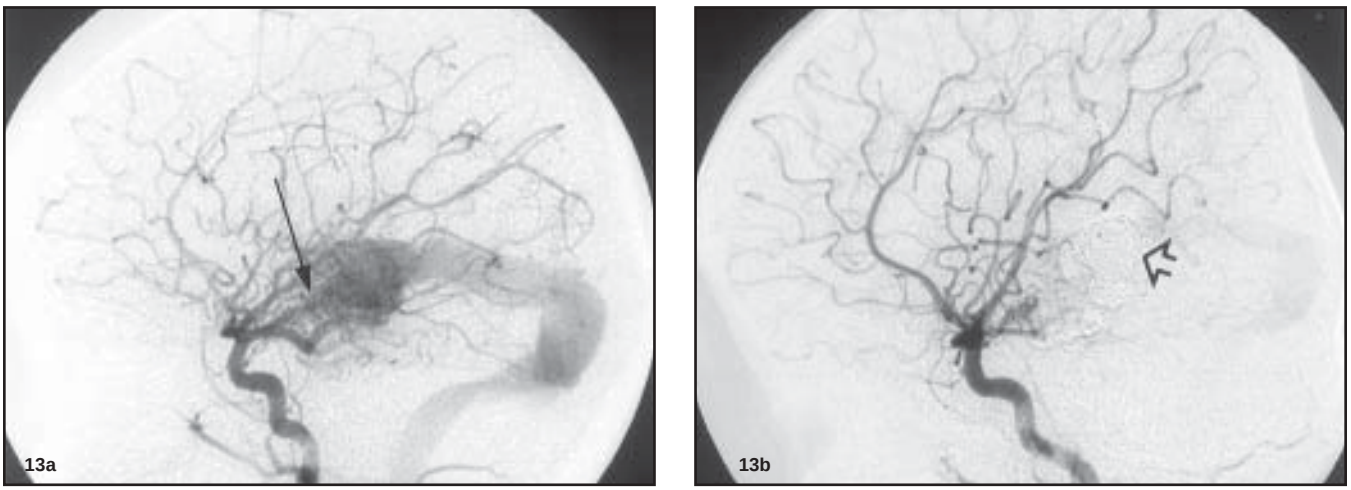
fistulas are occluded on the arterial side, using embolic agents such as coils, cyanoacrylates and detachable balloons (Figure 12). This route is preferred for embolization by most authors. Transvenous and transtorcular coil embolization of the venous sac have been used to achieve flow reduction in selected cases with high-flow fistulas. Transvenous embolization has been described as the technique of choice in patients with multiple fistulas, as it results in retrograde thrombosis and obliterates the fistulas (Figure 13). However, Lasjaunias and co-workers have recommended that venous embolization be reserved for patients in whom arterial route embolization is impossible or unsuccessful.[14] Venous embolization is also avoided in patients with parenchymal or choroidal arteriovenous malformations. These lesions are embolized from the arterial side to avoid venous hypertension (Figure 14).[46]

Complications

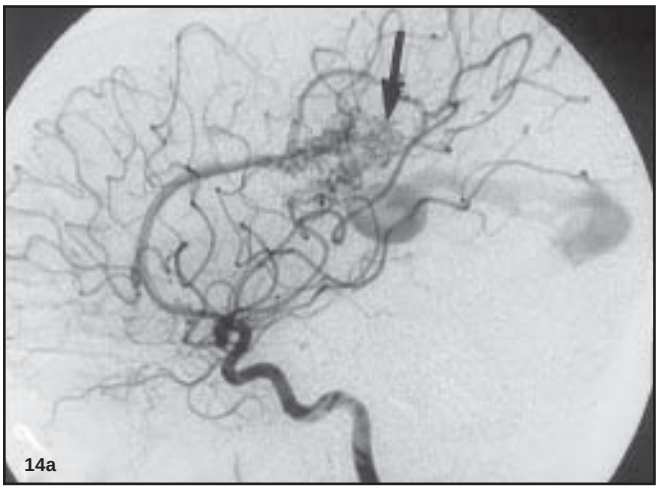
Potentially fatal complications of endovascular management include normal perfusion pressure breakthrough and intracerebral hemorrhage due to venous hypertension.[55] These can be largely avoided by staging the embolization procedure. Perforation of the venous sac has been reported to occur during positioning of the microcatheter during coil embolization, and can usually be managed by reversal of anticoagulation and continuation of coil embolization. Ischemic neurological deficits can occasionally be encountered after embolization. Pulmonary embolization with embolic agents is common considering the high flow across the intracranial shunt.

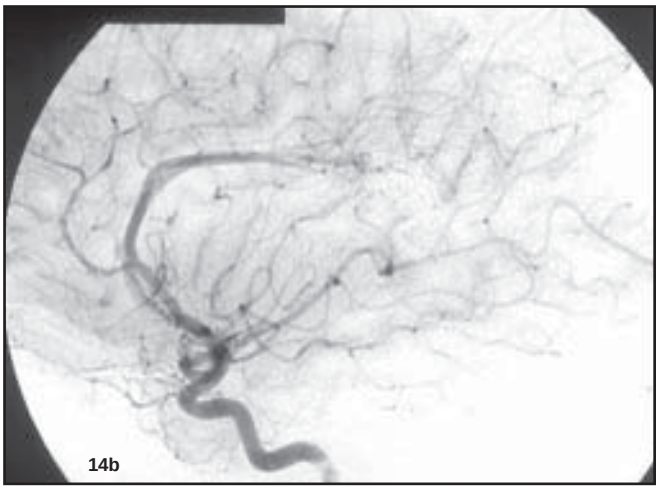
Conclusion

Thus, with advances in imaging technology, cardiac care,



Figures 13a and 13b:Combined transarterial and transvenous embolization of choroidal type of vein of Galen malformation :
a: Choroidal type of vein of Galen malformation fed by multiple perforators
b: The venous sac was initially accessed via transfemoral venous route and packed with GDC coils to achieve flow stasis. Subsequently, embolization of the choroidal feeders was performed from the arterial side, resulting in complete obliteration of the fistula





Figures 14a and 14b: Transarterial embolization in a case of vein of Galen aneurysmal dilatation:
a: Arteriovenous malformation involving the splenium of the corpus callosum draining into an aneurysmally dilated vein of Galen
b: Transarterial embolization of the pericallosal arteries was performed to resulting in complete obliteration of the arteriovenous malformation

developments in the field of interventional neuroradiology and availability of better post-procedure intensive care, these once non-treatable conditions with a very high mortality rate, are now potentially curable using interventional neuroradiological techniques, with excellent clinical results, low complication rate and very low morbidity and mortality. In the future, with the establishment of more and more centers with such facilities in the country, the treatment of this condition will be within the reach of all patients.

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