ANALYSIS OF ENDOVASCULARLY TREATED VEIN OF GALEN MALFORMATIONS, AN INSTITUTIONAL EXPERIENCE

THESIS

SUBMITTED IN PARTIAL FULFILLMENT FOR DEGREE OF **DM (NEUROIMAGING AND INTERVENTIONAL NEURORADIOLOGY)**

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OCTOBER2010 **Dr. SOMENATH CHATTERJEE**

DECLARATION

I hereby declare that this thesis entitled **"ANALYSIS OF ENDOVASCULARLY TREATED VEIN OF GALEN MALFORMATIONS, AN INSTITUTIONAL EXPERIENCE"**, has been prepared by me under the supervision and guidance of Dr. Arun Kumar Gupta, Professor and Head, Department of Imaging Sciences and Interventional Radiology, Sree Chitra Tirunal Institute for Medical Sciences & Technology, Thiruvananthapuram.

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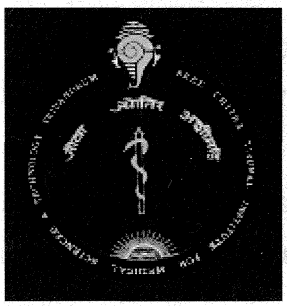

CERTIFICATE

This is to certify that the work incorporated in this thesis entitled, **"ANALYSIS OF ENDOVASCULARLY TREATED VEIN OF GALEN MALFORMATIONS, AN INSTITUTIONAL EXPERIENCE"** for the degree of **DM (NEUROIMAGING AND INTERVENTIONAL NEURORADIOLOGY)** has been carried out by **Dr Somenath Chatterjee** under my direct supervision and guidance. The work done in connection with this thesis has been carried out by the candidate himself and is genuine.

Guide

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INTRODUCTION

Vein of galen malformations are rare intracranial vascular anomalies. It constitutes approx 1% of all intracranial vascular malformations. They represent 30% of intracranial vascular malformations presenting in pediatric age group (1, 2).

Steihill et al made the first reference to a galenic malformation- referring it as a varyx aneurysm (3).

These lesions are characterized by aneurysmally dilated midline venous structure representing median prosencephalic vein of Markowski fed by abnormal arteriovenous communication (4, 5).

NOMENCLATURE-

There has been several different names in the literature referring to this entity. Aneurysms of vein of galen, arteriovenous aneurysms of the vein of galen, vein of galen aneurismal malformations and vein of galen malformation are the terms used by different groups and authors. All those nomenclatures are imprecise because the dilated midline venous structure represents the persistent embryonic median prosencephalic vein and not vein of galen (6).

EMBRYOLOGY-

VOGMs represent embryonic vascular malformations. Hence, they are associated with the persistence of vascular arrangements that are characteristic of a particular period of development. 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 (7).

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 (7, 8).

The primary abnormality that is responsible for the development of vein of Galen malformations occurs after the stage of the 21-23 mm embryo (8.6 -8.8 weeks). By that time, the primary internal carotid artery and its terminal branches-the anterior cerebral and anterior choroidal arteries have formed. 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 (7).

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 (7).

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 (approximately 4-50mm embryo) (7). 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) (2, 7).

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 (7, 8).

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. 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.

In type 1, there is pure cisternal fistula between pericallosal or posterior cerebral arteries and the vein of galen.

Type 2 shows fistulous connections between the thalamoperforators and the vein of galen.

Type 3 is mixed form with characteristics of both type1 and 2.

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 (9).

The angioarchitecture of VOGM was classified into the mural or choroidal type, depending on the characteristic of the fistula by Lasjaunias.

The mural type consists of high-flow shunts, supplied by the collicular and posterior choroidal arteries, ending directly within the aneurysmal wall of the median prosencephalic vein.

The choroidal type involves the interposition of an extensive arterial network between the arterial feeder, supplied by the choroidal, subfornical, or pericallosal arteries or thalamoperforator arteries, and the venous aneurysm. The choroidal form usually leads to more severe symptoms whereas the mural form is better tolerated clinically. Mixed forms combining direct shunts and arterial networks may also occur (8, 10).

Vein of Galen malformations should be differentiated from the VGAD, which represents a normally formed vein of Galen that dilates as a result of outflow obstruction. The dilated venous structure drains an arteriovenous malformation located in the subarachnoid space, either supra- or infratentorially, in addition to the adjacent normal

cerebral parenchyma (10, 11). The degree of dilation is variable and depends on the extent of stenoses or thrombosis. The frequency of VGADs in neonates and infants is low and patients with VGAD often present in later age during childhood, with intracranial hemorrhage, focal neurological deficits, and in the very young, with delayed psychomotor development (8). With VGAD, epilepsy is not common, because of the deep location of the lesions, and heart failure is also uncommon because of its presentation in older children (12).

PATHOPHYSIOLOGY-

The pathophysiological consequences of VOGM most often manifests as high-output cardiac failure and neurological symptoms secondary to cerebral venous congestion and abnormal CSF flow (12). The severity of the symptoms is variable and dependent on the underlying angioarchitecture of the VOGM as well as the age of the child. Typically neonates present with congestive heart failure, infants present with hydrocephalus, and the older children or adults present with headache and, frequently, with signs and symptoms of subarachnoid hemorrhage.

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 after birth. 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 ventricles working in parallel.

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. 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 (6).

Cerebral low-resistance arteriovenous shunts in VOGM lead to increased venous return to the right atrium, subsequent pulmonary hypertension due to increased pulmonary blood flow, and ultimately congestive heart failure as a result of the increased preload (12, 13). Although the VOGM develops in utero, it does not produce severe cardiac failure until after the child is born. The low resistance in the placenta competes with the VOGM, and therefore the flow through the malformation is not as great in the fetus as it is after birth. After birth and removal of the placenta, flow through the fistula suddenly increases. Large arteriovenous shunts reduce the diastolic pressure within the aorta, leading to reduced coronary blood flow, which, coupled with the high ventricular pressure, can promote myocardial ischemia (6, 14). Flow reversal in the aorta can lead to

renal hypoperfusion and subsequently to renal failure. Congestive heart failure developed prenatally can be detected on ultrasound and in severe cases can lead to multiorgan failure and irreversible brain damage (12, 13). While antenatal diagnosis is not an indication for abortion or early or cesarian delivery, in utero cardiac failure and brain damage represent indications for abortion (15). In the neonatal period, cardiac symptoms may range from mild volume overload to severe cardiogenic shock depending on the maturity of the cardiopulmonary system (10, 12, 16, 17). Mild symptoms can consist of feeding difficulties, tachycardia, and signs of cardiomegaly on chest radiography (12). In the mild condition, diuretics can be used to improve symptoms, permit normal feeding, and allow the patient to return in 5 months for embolization procedure (12). In severe cases, the patient may require mechanical ventilations, and in refractory cases, may require immediate embolization of the cerebral arteriovenous shunts (12). In contrast to prenatal patients and neonates, infants rarely present with symptoms of congestive heart failure. They can be treated with diuretics until embolization and can be frequently weaned from these medications after the first embolization procedure (12).

Because of the tremendous flow of blood to the fistula, the blood is stolen from the cerebral parenchyma, leading to ischemic damage. High blood flow in the arteriovenous shunt and reduction in venous drainage secondary to poorly developed venous system or venous stenosis, coupled with altered hemodynamics due to cardiac manifestations, can result in high cerebral venous pressure and cerebral edema (6). Together these manifestations can lead to a rapid loss of brain tissue in the neonatal period, the most severe form referred to as the melting brain (12). The high venous pressure, coupled with

the lack of maturation of the arachnoid granulations, can lead to impaired CSF resorption, resulting in hydrocephalus (6, 18). Obstructive hydrocephalus can also occur due to VOGM induced compression of the aqueduct. When hydrocephalus is present before 5 months, embolization should be the first line of treatment leading to a reduction in venous pressure and improvement of CSF resorption. A CSF shunt or ventriculostomy does not correct the underlying problem (12) and can potentially lead to intracerebral hemorrhage from the congested and immature venous system. Rerouting of blood flow into the facial veins or basilar or pterygoid plexus can lead to prominent facial veins and epistaxis (6, 19).

CLINICAL PRESENTATION-

The clinical pictures are correlated with the age of presentation and the underlying pathophysiology. Gold et al first presented this age wise presentation in 1964 (20).

Neonates. Symptomatic newborns present with severe cardiorespiratory failure including hydrops and renal failure secondary to flow reversal in the aorta at or shortly after birth (4). In one series, in the majority of cases (94%) diagnosed in the neonatal period, the patients presented with high-output cardiac failure (17). Severe pulmonary hypertension may also complicate management. Cyanosis may be present and may thus be mistaken for congenital heart disease (21). An electrocardiogram can detect some features of myocardial infarction (10). In any infant born with high-output cardiac failure, a VOGM should be ruled out. While the mortality rate in the past was close to 100%, recent advances in endovascular techniques have greatly improved the survival rate.

Infancy. In contrast to the neonates, patients presenting during infancy usually have smaller shunts and only mild cardiac manifestations. These patients most frequently present with increased head circumference or seizures (4, 6, 20). Those with longstanding cerebral venous hypertension may present with developmental delay (6). Some also have failure to thrive resulting from cardiac decompensation or even hypothalamic and hypophyseal dysfunction as a result of venous congestion (8, 22). Other signs and symptoms often include cranial bruit, dilated scalp veins, proptosis, and occasionally recurrent epistaxis (4).

Older Children and Adults. Older children most often present with headache and seizures caused by intraparenchymal or subarachnoid hemorrhage (4, 6, 12). The VOGM is usually small with a limited degree of arteriovenous shunt, but the angiomatous network supplying the vein of Galen can harbor microaneurysms. A vein of Galen dilation secondary to outflow obstruction should also be considered in the differential diagnosis.

IMAGING-

Ultrasound- It is a noninvasive technique for bedside evaluation of the cerebral vascular system that is quick and convenient, although strongly operator dependent (4, 23). Antenatal ultrasound can show the venous sac located posterior to the third ventricle and can use pulsatile flow within the lesion to differentiate a VOGM from other midline cystic structures (6, 24, 25). It can also demonstrate associated venous anomalies, the

presence of hydrocephalus, and cardiac dysfunction (6). In the postnatal period, ultrasound can be used to assess progressive thrombosis of the venous sac and quantify residual postembolization flow after endovascular therapy (26, 27). The use of ultrasound and Doppler imaging is also critical to assess flow reversal in the aorta and changes after treatment. Ultrasound is preferred over CT and MRI because of MRI artifacts due to metal embolization coils, but it may not be sensitive enough to detect slow flow in areas of embolization (5).

Computed Tomography. Contrast-enhanced axial CT scanning of the brain can demonstrate a multilobulated, intensely enhancing lesion, ventricular dilation, periventricular white matter hypodensities, diffuse cerebral atrophy, and thrombosis within the aneurysm sac (6). Computed tomography scanning is also an important modality for demonstrating intracerebral calcifications secondary to ischemic brain damage. Advances in CT angiography and multidetector technology have allowed for greater resolution of the vascular bed (28, 29). Although not as precise as angiography, CT angiography is a noninvasive technique like that of ultrasound or MRI and can be performed in awake or slightly sedated patients (4). In addition, multislice spiral CT angiography has been found to be very useful for detailed mapping of both arteries and veins, with the use of a single scan and a single dose of contrast medium (30, 31). It requires fewer than 5 minutes for processing and is faster and with better imaging quality than MR angiography or single-slice CT angiography. When multislice spiral CT angiography is not available, CT angiography is preferred over MR angiography because of greater image quality for cerebral vein identification (5).

Magnetic Resonance Imaging. Ultrasound now frequently detects VOGMs in utero, although MRI is increasingly used, not only for the characterization of the lesion but also the documentation of brain atrophy and cardiac abnormalities (32-34). Magnetic resonance imaging can identify the fistula location, presence of any nidus, venous drainage, venous thrombosis, and it can estimate the number and type of arterial feeders (4, 6, 35). Importantly, because of its exquisite soft-tissue contrast, MRI is the modality of choice to evaluate the ventricular system and cerebral parenchymal damage, which is important for therapeutic decision making and prognosis (4, 32).

Angiography. Angiography is the gold standard for precise evaluation of VOGM angioarchitecture, including detailing the anatomy of arterial feeders and the hemodynamics of venous drainage, and it provides access for endovascular management (4). If a VOGM is diagnosed in utero, it is imperative to preserve the umbilical artery because this will give the best endovascular access in the neonatal period. Catheterization of the umbilical artery is most convenient and can be done up to the 3rd day of life. Femoral access is possible, but the vessel is smaller and it carries the risk of possible limb ischemia (4). In newborns with severe VOGM and cardiac failure concomitant with renal insufficiency, angiography can be complicated by the limited amount of contrast agent that can be used. In general if the patient is clinically stable, angiography should be delayed until the 5th month of life (36).

MANAGEMENT-

With a comprehensive multidisciplinary approach in the intensive care units, has significantly improved the poor prognosis of patients with VOGM (4, 15, 37). Despite advances in microneurosurgery, complete removal of the lesion in newborns is rarely achieved or advised, because of the hemodynamic instability and location of the lesion, poor myelination of the brain parenchyma, and cerebral venous hypertension. As a result, surgical treatment is now reserved for the evacuation of intracranial hematomas and the treatment of hydrocephalus, or in cases of embolization failure. Medical treatment with diuretics, inotropes, and other cardiovascular agents is used only to relieve symptoms of cardiovascular instability and renal insufficiency until the patient can undergo embolization (38). Most of the newborns with VOGM need immediate treatment. The main goal in the newborn with high-output cardiac failure is to decrease the flow in the VOGM and therefore reduce the cardiac output failure, improve the coronary perfusion, and increase the blood flow to the renal arteries. If a patient has persistent or recurrent heart failure and cyanosis after initial successful endovascular treatment, a persistent patent ductus arteriosus has to be ruled out. The pathological hemodynamic situation with increased venous return to the right side of the heart and right-to-left shunting through a patent foramen ovale can lead to a failure of the ductus arteriosus to close requiring treatment.

Endovascular Treatment. Endovascular access can be gained via the transarterial or transvenous route (4). In the newborn arterial access can be achieved through the umbilical artery if patent or through the femoral artery. The embolic glue of choice to occlude the arteriovenous fistula on the arterial side is *N*-butyl-cyanoacrylate, and more

recently Onyx has been used as well (4, 15, 39). Detachable microcoils can also be used, although their application may take longer and they may have a higher risk of vessel rupture (4). The transvenous route can be approached via the femoral or jugular vein or alternatively, through direct puncture of the torcula (4, 40, 41). Although some groups have used the venous route with success, (2) others reserve it for patients in whom the arterial route cannot be approached successfully (4, 15). In addition, occlusion of the venous aneurysm may hinder deep venous drainage or result in perforation of the venous aneurysm (42, 43). A patient's clinical presentation dictates the timing of endovascular management (6). Congestive heart failure refractory to medical management in a newborn necessitates emergency embolization to relieve the hemodynamic load on the heart. In this case, partial, rather than complete, obliteration of the shunt is the goal to allow for redistribution of blood flow to the heart and brain to allow normal cardiac and neurological development (37). The residual shunt can then be occluded at a later time to reduce complications. In a child who does not present with cardiac failure, the goal is to prevent the development of cerebral venous hypertension (44). Treatment often involves multiple successive procedures that target different pedicles to avoid parenchymal bleeding and venous thrombosis (4). Ideally, if the patient is not in heart failure, treatment should be deferred until 5–6 months of age, when fluid hemodynamics are still amenable to change and there is a low risk of affecting brain maturation (4-6). However, excessive delay may lead to permanent neurological and cardiac impairment unsalvageable by intervention. Spontaneous thrombosis is a rare event and should not be relied on to delay intervention (4, 15). Recently, Lasjaunias et al. described a 21-point scale score based on cardiac, cerebral, hepatic, respiratory, and renal function to guide

therapeutic decisions. A score of < 8 often signifies a poor prognosis that does not warrant treatment, a score from 8 to 12 is an indication for emergency embolization, and a score of > 12 indicates medical management until the child is at least 5 months of age (15).

The following table shows the neonatal score calculation.

Score	Cardiac	Cerebral	Respiratory	Hepatic	Renal
5	Normal	Normal	Normal		
4	Overload & no medical Rx	Subclinical, isolated EEG abnormalities	Tachypnea, finishes bottle		
3	Failure, stable with medical Rx	Nonconvulsive, intermittent neurological signs	Tachypnea, does not finish bottle	No hepatomegaly,normal hepatic function	Normal
2	Failure, not stable with medical Rx	Isolated convulsions	Assisted ventilation, normal saturation FiO2<25%	Hepatomegaly,normal hepatic function	Transient anuria
1	Ventilation needed	Seizures	Assisted ventilation, normal saturation FiO2>25%	Moderate or transient hepatic insufficiency	Unstable diuresis with Tx
0	Resistant to medical Tx	Permanent neurological signs	Assisted ventilation, desaturation	Abnormal coagulation,elevated enzyme levels	Anuria

REVIEW OF LITERATURE

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. 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.

There has been extensive literature regarding vein of galen malformation. Endovascular as well as surgical series as well as case reports have been plenty. Primary surgical treatment for this vascular malformation is not practiced currently; few old reports are there in the literature with discouraging results.

Watson et al described the case of a 2-day-old infant whose heart failure was not readily explained by clinical examination. Cardiac catheterization suggested an intracranial arteriovenous (AV) fistula, and cerebral arteriography showed a malformation of the vein of Galen. The major feeding arteries were surgically obliterated. At age 27 months, the boy has normal mentation but moderate left hemiparesis. Review of the literature disclosed 39 other infants with AV malformations of the vein of Galen producing heart failure before age 3 months. Most of them were boys, and had cyanosis, a systolic murmur, cranial bruit, cardiomegaly, and right ventricular hypertrophy. Only three of the

13 who had surgery for their malformation survived. They concluded that removal of the malformation is difficult; obliteration of the nutrient vessels, using the operating microscope, was the then currently accepted treatment (45).

A case of aneurysm of the great vein of Galen was reported by Hashi et al in which craniotomy and clipping of a feeding artery arising from the left posterior cerebral artery was successfully performed. This 5-month-old girl developed progressive hydrocephalus from 2 months after birth. At 5 months the head circumference was 50 cm with tense enlarged fontanelle. Both eye balls were deviated downward. The deep tendon reflexes were hyperactive with bilateral ankle clonus and positive Babinski's sign. Mild cardiomegaly and hepatomegaly were also noted. Cerebral angiograms showed a large aneurysm of the great vein of Galen fed by a single enlarged arterial branch from the left posterior cerebral artery. After the evaluation of systemic circulatory status and under strict control of fluid transfusion craniotomy was performed. The feeding artery was clipped at its entrance to the aneurysm via left parieto-occipital interhemispheric approach. A rise in the mean arterial blood pressure of 20 mmHg was observed immediately following clipping. The postoperative course was satisfactory except for a left subdural fluid collection which required subduro-peritoneal shunt. The aneurysm was completely disappeared on the postoperative angiograms and the child is regaining the normal development. This was the first case of aneurysm of the great vein of Galen successfully operated in Japan (46).

A giant arteriovenous fistula in a newborn infant was treated by surgical occlusion of the feeding vessels at 20 days of life. Congestive heart failure responded favorably to

operative treatment. Because of persistent hydrocephalus, a shunt was inserted at 2 months of age. At 9 months of age, the child remained without signs of cardiac failure. Cerebral damage was manifested by a mild left hemiparesis. Successful surgical treatment of this unusual lesion in a neonate was considered exceptional (47).

Menezes et al reported two cases where parieto-occipital craniotomy with an interhemispheric approach was performed to clip the numerous feeders. The procedure was terminated when the sac collapsed and blood aspirated from the lesion was venous in oxygen saturation. A Doppler probe over the aneurysm then revealed a venous flow. Serial postoperative CT scans demonstrated that the mass had shrunk in size. Follow-up angiography was not thought necessary (48).

Eiras et al reported a case of primary aneurysm of the great vein of Galen treated successfully by clipping the afferent arteries and reduction of the bulk of the aneurysm by bipolar coagulation. Five months after the operation the child's development was normal (49).

Moriarity et al reported staged surgical treatment of a vein of Galen aneurysm in a neonate who presented in congestive heart failure. Cerebral angiography 6 months following staged surgical treatment revealed complete obliteration of the aneurysm. The patient tolerated surgery well, and at 6 years of age remained free of neurologic or cardiovascular impairment (50).

Another report describes a newborn who developed severe cardiac failure in the second week of life. An aneurysm of the vein of Galen was noted on the computerized

tomography scan, confirming the clinical diagnosis of cerebral arteriovenous fistula. The vascular abnormality was well shown by cerebral arteriography. At the age of four weeks, surgery was carried out by clipping the afferent arteries. The patient was followed up to three years age with moderate developmental delay and hypotonia (51).

The clinical details of diagnosis and treatment in 13 patients with such lesions together with a review of 232 cases collected from the literature were presented in the report by Johnston et al. There were 132 males, 77 females, and 36 cases in which the sex was not stated. Eighty patients presented as neonates, 82 were 1 to 12 months old, 39 were 1 to 5 years old, 22 were 6 to 20 years old, and 22 were over the age of 20. The most common presenting symptoms were congestive cardiac failure (110 cases), raised intracranial pressure secondary to hydrocephalus (94 cases), cranial bruit (57 cases), focal neurological deficit (37), seizures (26 cases), and hemorrhage (25 cases). The most characteristic vascular supply to the midline fistula involved multiple bilateral vessels, although bilateral posterior cerebral and unilateral posterior cerebral supply was relatively common. The overall figures for treatment and outcome showed that 91 patients (37.1%) were treated by direct operation and 29 patients (11.3%) were treated by other forms of operation, predominantly shunting or remote vessel ligation. Forty-six patients (18.8%) were treated by medical means (digoxin, diuretics, and ventilatory support). In 79 patients (22.2%), there was no treatment or no details of treatment were available. There was an overall series mortality of 55.6% (no details were available in 33 cases) and a 37.4% mortality for surgically treated cases. After operation, there was a 46.3% incidence of significant morbidity in surviving patients. Neonatal patients fared worst, with an overall mortality of 64 of 70 cases (91.4%) where details were available.

The outcome was equally bad for surgically and conservatively treated cases. Operation in the 1- to 12-month age group was more successful, but still carried a mortality of 31.7%, with a significant morbidity in approximately half of the surviving patients. Over the age of 1 year, the surgically treated patients had 25.6% mortality and a 42.3% major morbidity in survivors. Consideration is given to some of the ways in which these figures may be improved, in particular a staged approach during the neonatal period, with the use of selective embolization or occlusion of vessels to reduce the volume of the arteriovenous shunt until the patient is older and better able to tolerate major operation (52).

Three cases are described by Morgan et al of infants who developed malignant brain swelling (and in one case hemorrhage) after surgery for vein of Galen malformations. The cause for the brain swelling was felt to be due to hyperperfusion, or the "normal perfusion pressure breakthrough" syndrome. Although well-described for cerebral parenchymal arteriovenous malformations, cases of this complication occurring in vein of Galen malformations have not previously been reported. It is concluded from these cases that infants with large arteriovenous shunts, as attested by cardiac failure and cerebral atrophy, have an increased risk of developing this complication (53).

There is a series with direct comparison of surgical as well as endovascular experience from the same hospital by Circillo et al. Since 1978, the authors have seen 14 neonates with vein of Galen malformations who were born with severe congestive heart failure. The 5 infants treated before 1983 underwent craniotomy and clipping of feeding vessels; all died in the perioperative period. Since 1983, 8 neonates have been treated with

combined arterial and venous interventional neuroradiological techniques; 6 infants survived. Two-dimensional echocardiography, color Doppler flow imaging, and pulsed Doppler ultrasound were used to assess blood flow within the malformation before and after staged transluminal embolic procedures were performed. The results of the diagnostic studies and the clinical status of the infants were used to evaluate the success of embolic therapy and the need for further neuroradiological intervention (3).

Hernesniemi reported three cases of arteriovenous malformations of the vein of Galen operated on without mortality and morbidity. Postoperative angiography confirmed total occlusion of the fistulas in each case. Microsurgical operation in the parieto-occipital region with interhemispheric approach was performed to cut the numerous feeders. The procedure was terminated when the sac was diminished in size with bipolar coagulation and clipped with encircling clips to preserve the flow through the vein of Galen. In spite of the good surgical results, the long-term outcome in the two pediatric patients was discouraging because of mental retardation caused by the arteriovenous malformation itself (54).

A consecutive series of nine VOGMs in eight children aged 4 to 14 years and in one adult were treated with gamma radiation. Six of the patients were male, including the adult, and three were female. Among these patients there were three Yasargil Type I, one Type II, two Type III, and three Type IV malformations. Previous embolization had failed in four cases. Three VOGMs were treated with gamma radiation twice. An additional patient with a Type III VOGM underwent stereotactic angiography in preparation for gamma radiation but was judged to be suitable for direct embolization. Follow-up

angiograms were obtained in eight of the VOGMs treated. Four no longer filled; one has probably been obliterated, but this cannot be confirmed because the patient refused to undergo final angiography; one patient has residual fistulas not included in the initial treatment field, which were retreated recently; and two other patients have marked reduction of flow through their VOGMs. Authors concluded that Gamma radiation is a viable option in the treatment of VOGMs in clinically stable patients. Combined endovascular therapy and gamma radiation is of benefit in complex malformations (55).

Watban et al presented three cases of vein of Galen aneurysmal malformations diagnosed in infancy and submitted by the referring teams for stereotactic radiotherapy as the initial therapy (therapeutic doses ranging between 20-25 Gy and 40-50 Gy to the peak dose). After the conventional follow-up of 18-24 months, no change could be detected in the angioarchitecture of the lesions. All three cases were then referred for endovascular treatment and underwent embolization by the transarterial route using liquid adhesives (N-butyl cyanoacrylate). This resulted in complete anatomical exclusion of the lesion. Regardless of the theoretical efficiency of radiotherapy in the management of brain arteriovenous malformations, the present authors believe that transarterial embolization remains the treatment of choice in VOGMs. It offers a high rate of morphological cure and the best chances for normal neurocognitive development. The time required by stereotactic radiotherapy to achieve a significant result is too long for developing and maturing brain and may not prevent the negative effects of the lesion, mainly in regard to hemo- and hydrodynamic disorders (atrophy, subcortical calcifications etc.) created by the VOGM, thus leading to irreversible mental retardation (56).

Takemoto et al reported the surgical experience of two cases of vein of Galen aneurysmal malformation in the newborn, whose congestive high-output cardiac failure was intractable. Along with the intensive care to clinical manifestations of the heart failure, multi-staged feeder clipping was carried out to decrease the high-flow shunt of the malformation. As stages going on, heart failure was relieved gradually and catecholamine was weaned. Although certain retardation became apparent in both cases, they are showing satisfactory development in the long-term follow up. By the recent advancement of the embolization technique, the embolization appears to have already taken place the treatment of choice for this malformation. According to the neonatal evaluation score of Lasjaunias, the embolization would no longer be recommendation in neonates, whose general condition scored less than eight points. The authors believe, based on our two cases, that multi-staged feeder clipping is one of the effective modality of treatment in neonates of the vein of Galen aneurysmal malformation with severe multiorgan failure (57).

Since 1985, five patients with VOGM have been referred to the neurosurgical unit of the University of Cologne, two neonates, one infant and two adults. Four patients underwent direct operation and two patients received a shunt. The treatment was performed without mortality. Their review of the literature at that time reflects no substantial difference between neurosurgical treatment during the last 15 years (mortality 10%) and endovascular treatment (best series mortality 6%) (58).

Heuer et al collected retrospective clinical data for patients evaluated with a diagnosis of VOGM from 1994 to 2007. Thirteen patients with VOGM were evaluated from 1994 to

2007. Seven patients presented emergently with medically intractable cardiac failure, and six were treated in the first 2 weeks of life. Five children treated after this period (1.5-31 months of age) manifested enlarging head circumference, abnormal development, or subarachnoid hemorrhage. Eleven patients were managed endovascularly. Four disease or procedure-related complications occurred. Two complications were associated with poor outcome, both of which occurred in patients treated at less than 2 weeks of age. Two other patients experienced transient neurological deficits with no evidence of permanent sequelae. Outcome in the six patients treated emergently in the first 2 weeks of life included two patients who developed normally, one with mild to moderate neurological deficits, one with severe neurological deficits, and two deaths. Outcome in the five older patients (treated between 1.5 and 31 months) was considerably better than in the group treated early and included three with normal outcome and two with mild neurological deficits (59).

Between December 2005 and January 2008, Jankowitz et al treated 6 children with cerebral vascular malformations using Onyx-18. The ages of the patients ranged from 1 day to 12 years. Pathological types of the vascular malformations included 4 arteriovenous malformations and 2 vein of Galen malformations. Clinical presentations included intracranial hemorrhage in 2 patients, papilledema in 1 patient, and high-output heart failure in 3 patients. In 6 pediatric patients, 21 embolization procedures were performed utilizing a combination of Onyx-18, platinum coils, and Embosphere microspheres. The average estimated size reduction for the arteriovenous malformations was 60%. Total obliteration of a malformation was achieved in 1 patient. Two patients received adjuvant stereotactic radiotherapy. Of the 2 vein of Galen malformations, one

was completely embolized and the other had an approximately 50% reduction in flow. No open surgical intervention was used. Clinical follow-up ranged from 7 to 12 months. Angiographic follow-up data were obtained at 1, 6, and 7 months in 3 patients, whereas 1 patient awaits repeat angiography. Complications included a transient monoparesis with complete resolution in 1 patient. Two patients died within 24 hours of an embolization procedure due to intracranial hemorrhages (39).

Twelve children with intracranial AV shunts were treated by Ishiguro T et al with endovascular embolization between December 1993 and March 2008. These comprised two cases of vein of Galen aneurysmal malformation, three of dural sinus malformation, two of infantile dural AV shunt, five of pial AV fistula including two of vein of Galen aneurysmal dilatation. There were eleven boys and one girl. The age at the first embolization ranged from day 0 to 9 years. We reviewed their clinical features and outcomes. Six patients including four neonates presented with congestive heart failure, one infant with macrocrania and three children with headache, seizure or ataxia. The number of endovascular embolization ranged from one to five per patient. These included eighteen transarterial embolizations and ten transvenous embolizations. All patients except for one who died eventually from pulmonary hemorrhage showed improvement in their symptoms. Although only five patients achieved complete occlusion of AV shunts, six patients including them developed normally (60).

Germanwala et al reported for the first time in the literature, a case of an infant with VOGM treated initially with staged coil embolizations followed 1 year later by the transarterial and transvenous catheter based injection of Onyx-18 (ethylenevinylalcohol

copolymer) in a single treatment session. The fistula was eliminated, and the infant's cardiopulmonary symptoms were improved (61).

Li et al reported a 12-week-old baby with a vein of Galen aneurysmal malformation (VOGM) successfully treated with performing transarterial microcatheter-directed embolization with Berenstein Liquid Coils and n-butyl cyanoacrylate in the feeding arteries. Post-procedure angiography showed a marked decrease of the blood flow into the dilated vein of Galen. Three months later, follow-up angiography showed that the vein of Galen aneurysmal malformation had totally disappeared, and the baby recovered very well without any sequelae (62).

Lasjaunias et al reported 14 cases of vein of Galen dilatation explored and/or treated between 1983 and 1986. Three anatomic types have been individualized: the vein of Galen arterio-venous malformation (AVM) (7 cases); the cerebral AVM with vein of Galen ectasia (6 cases); the varix of the vein of Galen without AV shunt (1 case). When an AVM is present the shunt is located either in the venous wall (vein of Galen AVM) or in the brain parenchyma; in the latter the AVM drains into a tributary of the vein of Galen (cerebral AVM with vein of Galen ectasia). However in all the 14 cases downstream to the draining vein, a venous (dural) anomaly could be demonstrated. This anomaly suggests the secondary nature of the dilatation proximal to a developmental obstacle. Clinically the vein of Galen AVM reveals early in neonates by cardiac complications; the other types are usually seen later following hydrocephalic or bleeding episodes. As far as therapeutic aspect, we can technically stabilize the hemodynamic problem, by occluding most if not all the shunts by endovascular approach, and make most of the neonates

survive. However the quality of survivance and the future neurological development is impossible to predict yet. Nevertheless the short term follow-up is very encouraging. The combined per operative embolization through the vein must be exclusively reserved to vein of Galen AVM. The torcular approach is contraindicated in brain AVM with Vein of Galen ectasia. The Vein of Galen varix do not require any type of morphological correction (11).

They again reported a series of 36 vein of Galen aneurysmal malformations diagnosed in the paediatric (78%) and adult (22%) populations that were referred to them for therapeutic management between 1982 and 1988. The clinical signs leading to the diagnosis were variable: 36% of systemic manifestations, 22% of neurological symptoms, 17% of hydrocephaly and 11% of intracranial haemorrhage. Thirty angioarchitectural analyses could be obtained and allowed to classify these VOGMs into 5 different types: 44% parenchymatous AVMs, 20% mural AVFs, 30% choroidal arteriovenous fistulas, 3% dural AVFs, 7% vein of Galen varices. This series demonstrates that the paediatric population is most sensitive to shunt effect whatever its type. Systemic manifestations and hydrocephaly are the most common signs encountered in the newborn and infants; whereas neurological signs and symptoms and haemorrhage belong mostly to the adult symptomatology. Because of the poor outcome of VOGMs, all authors believe that these malformations have to be treated aggressively. However, they found contra-indications to be represented by pretherapeutic demonstration of cerebral tissue damage, or uncontrollable systemic failure, thus treatment is indicated to compensate for cardiac failure previously responding (even partially) to medical treatment. Secondly, appearance of sub-cortical calcifications, resistance to medication or clinical deterioration will also

lead to urgent treatment. The endovascular method represents at present the best treatment with an overall low mortality (13%) and a 0% technical morbidity in children compared to the surgical one of (91% mortality in newborns and 38% in infants). The results achieved by embolization in this series were as follows: 27% satisfactory results with complete or almost complete occlusion of AV Shunt, 53% significant clinical improvement, 7% of patients were unchanged. The authors believe fundamentally that these patients (specially those belonging to the paediatric population) have to be treated in a centre where a paediatric intensive care unit, neurological, neurosurgical and surgical neuro-angiographic departments coexist, in order to assure the best possible management of these children (63).

In a later series 43 patients with true vein of Galen aneurysmal malformations were reported by Lasjaunias et al. Thirty-four were embolized transarterially with isobutyl cyanoacrylate or N-butyl cyanoacrylate embolization. No cutdown or hypotension during or after the embolization was used and no balloon catheter was employed. Forty-seven percent of the children had a completely occluded lesion which was confirmed when the child was at least 6 months of age at the follow-up angiographic examination; 52.9% were found to be completely normal or only to have mild cardiac failure that could be treated medically or moderate macrocephaly without neurological symptoms or mental retardation. In the embolized group 5.8% died as a result of the wrong treatment (1 case) or poor timing of embolization 3 days after ventricular shunting (1 case). The overall mortality (embolized and non-embolized groups) in the neonatal children was 27.7% with a total of 18.6% for all ages. Complete morphological exclusion of the arteriovenous malformation was accomplished in 41.9%; 74.4% of all children referred are now

clinically normal or present moderate mental retardation which is diminishing. There was 3% neurological morbidity in the embolized group (only following the venous approach) in 78 sessions and more than 100 arteries embolized. These results compare favorably with surgical or other techniques of arterial embolization (balloon or particles), as well as transvenous (transtorcular or transfemoral) embolization, where the morbidity and mortality are significantly higher and the late clinical evaluation is seldom satisfactory (64).

Lasjaunias et al again came up with increased number of patients and described the management of 179 cerebral arteriovenous malformations (CAVMs) in children and infants. Seventy-seven were true vein of Galen malformations and 102 were pial AVMs (PAVMs), i.e., developed in the subpial space. Hemorrhage occurred as the first symptom in 50% of the children with pial AVMs, but was present in none of the VOGM cases. Only 31 children were found to be unsuitable for endovascular treatment, and in 124 cases embolization was indicated as the primary treatment (104 embolization performed). Only 21 children underwent a direct surgical approach (none in the VOGM group). In the embolized group in whom treatment has been completed (n = 56), 8 children died, 39 have an anatomical cure, and 34 are clinically normal. In the group under treatment (n = 48), 16 are not normal. The problems are timing and the aims (total or partial treatment) of the therapeutic procedures. In the nonembolized group (n = 31), 8/13 of the pial lesions were operated on (no mortality, 2 patients with moderate neurological deficits). In the VOGM group 13/18 died and 4 had spontaneous thrombosis (only 1 is neurologically normal). In the nonembolized group 13 lesions have been completely excluded, but only 5 patients are neurologically normal. This fact again

stresses the need for prognostic evaluation before treatment and a clear definition of the treatment aims. Analysis of a large number of published series on the management of children with AVMs (1017 cases) reveals inconsistencies that hamper proper evaluation and comparison. In their experience, endovascular treatment always seems to be the best primary treatment in both VOGMs and PAVMs. However, management of children with these lesions requires a large multidisciplinary team, which is the only way of offering the most suitable and effective treatment, the sole guarantee of a good result (65).

Later experience of Lasjaunias et al, based on 317 patients with VGAM who were studied in Bicerte Hospital between October 1981 and October 2002, allows to describe the angioarchitecture, natural history, and management of VOGM in neonates, infants, and children. Of the cohort of 317 patients, 233 patients were treated with endovascular embolization; of these, 216 patients were treated in their hospital. The treatment method of choice was a transfemoral arterial approach to deliver glue at the fistulous zone. Of 216 patients, 23 died despite or because of the embolization (10.6%). Twenty out of the 193 (10.4%) surviving patients were severely retarded, 30 (15.6%) were moderately retarded, and 143 (74%) were neurologically normal on follow-up (15).

Nakano et al reported two neonatal patients with high-flow intracranial or cervical arteriovenous (AV) shunts who presented with congestive heart failure and were successfully managed with endovascular treatment. One with vein of Galen malformation with body weight of only 2.0 kg is, to our knowledge, the smallest neonate successfully treated with endovascular treatment. Even in neonates, endovascular treatment is the

procedure of choice for high-flow AV shunts presenting with progressive congestive heart failure resistant to medical management (66).

In the series of Fullerton et al, outcome of patients who underwent endovascular treatment for VOGM between 1983 and 2002 was assessed by chart review and parental questionnaires. Development was classified as normal, minor delay (slow initial acquisition of milestones but no permanent disability), or significant delay (slow or incomplete acquisition of milestones with some permanent disability) using an adaptation of the Denver Developmental Questionnaire. Twenty-seven patients were identified: five presented prenatally (by ultrasound), 16 as neonates, and 6 after the neonatal period. The most common presenting features were congestive heart failure (CHF; 16/27) and hydrocephalus (8/27). The 16 patients with CHF all presented either prenatally or neonatally; 4 died acutely, 6 had significant delay, and 6 had no or minor developmental delay. Of those presenting in the perinatal period without CHF, all survived, two of five were significantly delayed, and three of five had no delay. Of those presenting after the neonatal period, all survived and only one of six had delay. By angiographic classification, outcome was worse for those with choroidal VOGM (3/13 died; 5/13 had significant delay) than for those with mural VOGM (2/10 had significant delay; none died). For the entire series, 52% of all cases (61% of survivors) had no or minor delay. Fourteen of 27 children who received treatment for VOGM had a favorable outcome. Features associated with worse outcome were perinatal presentation, presence of CHF, and choroidal angioarchitecture (67).

Jones et al retrospectively reviewed the radiology studies, hospital charts, and outpatient clinic chart notes (when applicable) of 13 children evaluated and treated for VOGM at a single tertiary care pediatric hospital. Clinical presentation, diagnostic methods, treatment strategies, and outcome were documented for each child. The present neurologic status and level of function of each patient was determined by review of the outpatient charts and direct contact with the clinicians who were conducting the follow-up. Outcome was graded on a 5-point scale, ranging from 0 (death) to 4 (normal), taking into account only neurologic and developmental characteristics. Eight of 13 patients presented as neonates with congestive heart failure. The other five patients ranged in age from 4 months to 13 years at the time of presentation. The five patients presenting outside of the neonatal period achieved normal or near-normal outcomes. Two of the eight patients presenting during the neonatal period achieved normal or near-normal outcomes, one experienced significant impairment, and the other five died. They were unable to identify significant differences in outcome on the basis of differences in treatment strategies. Their experience confirms that children with VOGM presenting during the neonatal period have a generally much worse prognosis than do those presenting later in childhood (33).

Rodesch et al described their experience of 168 consecutive cerebral arteriovenous shunts, all antenatally diagnosed lesions were vein of Galen aneurysmal malformations. This series consists of 18 cases of VOGMs detected by ultrasound during the third trimester of pregnancy. There were 12 normal vaginal deliveries, 5 deliveries by cesarean section, and 1 induced abortion. Sixteen newborns (94%) presented with systemic cardiac manifestations as the first clinical symptoms; 12/16 were managed effectively by digitalo-diuretic treatment, while 4 (25%) died shortly after birth from acute heart and/or

multiorgan failure with extensive brain damage. Twelve babies underwent embolization via the arterial route in infancy (2 at 2 months of age). Total exclusion was obtained in 8 babies (67%, 3 with 6 months follow-up). Furthermore, 67% of the newborns managed by our team are neurologically normal (Denver and Brunet-Leizine tests). These results emphasize that the pessimism that follows antenatal discovery of these lesions and the previous assumption of a bad prognosis for VOGMs can nowadays be reviewed in the light of transarterial endovascular therapy applied according to a strict clinicoradiological protocol. Interventions in the neonatal period are rarely required. Generally, poorly timed mechanical therapy should be discouraged (17).

In another series, twenty-four newborns with cardiac failure requiring mechanical ventilation were consecutively admitted from 1986 to 2000. Cardiovascular evaluation including echocardiogram was performed in all cases. Eighteen transarterial shunt occlusions with glue were applied by the same team of three physicians. Twelve babies survived and underwent one endovascular session at least (median age 20 days) with a mean 63 months follow-up. Embolization was not performed in 6 of the 12 nonsurvivors because of severe brain damage or profound hypotension Cardiogenic shock occurred in all nonsurvivors, but also in one long-term survivor ($p<0.0001$). Echocardiogram showed signs of right ventricular failure, most often in the babies who did not survive ($p=0.005$). The pulmonary systemic arterial pressure ratio was significantly higher in the nonsurvivor group ($p=0.031$), and it decreased significantly after the first embolization only in patients who survived ($p=0.01$). Patent ductus arteriosus and a diastolic aortic reversed-flow were present in all nonsurvivors in contrast to 30% of the long-term survivors ($p= 0.003$). There was no difference in the left ventricular contractility and

mean cardiac output between the two groups. The outcomes of vein of Galen malformation complicated by severe cardiac failure requiring mechanical ventilation remains poor. Neonatal embolization seems to be beneficial only in babies without suprasystemic pulmonary hypertension (13).

In Mitchell et al series, Between 1996 and 1998, five infants (three male, two female) were diagnosed with symptomatic VOGMs in the first week of life, four of whom had intractable, high-output cardiac failure and underwent initial endovascular treatment. There were 15 endovascular procedures and one neurosurgical clipping in these five patients. Transarterial and transvenous routes were required, using multiple embolic agents. We emphasized the use of sonographically guided, percutaneous transtorcular-venous-access, moveable-core guidewire as an embolic agent; routine MR imaging; and MR angiography. Immediate outcomes included control of cardiac failure with normal neurologic function in four (80%) patients and one (20%) death from intractable cardiac failure. On follow-up examination, three (60%) infants showed no evidence of neurologic abnormality or cardiac failure; one (20%) infant showed moderate developmental delay. Two have had no further shunting on angiography, one has minimal flow, and one is awaiting follow-up imaging. Endovascular therapy with modern neuroanesthetic and neurointensive care can provide good outcomes even in the highest-risk neonates with VOGMs and cardiac failure. If medical management of cardiac failure fails, and there is no evidence of gross cerebral parenchymal damage on imaging, urgent endovascular treatment is feasible and can reduce the almost-100% mortality otherwise expected, without invariably severe morbidity. Use of multiple embolization strategies in multiple

stages usually is necessary in these patients, and novel approaches and embolic agents may be necessary (41).

From September, 1986, to March, 1990, the Lylyk et al treated 28 children harboring a vein of Galen vascular malformation. Eleven (39.3%) of the patients were neonates, 13 (46.4%) were 1 to 2 years old, and four (14.3%) were more than 2 years old. Fifteen patients (53.6%) presented with severe congestive heart failure, six (21.4%) had seizures, four (14.3%) had hydrocephalus, and three (10.7%) presented with intraventricular hemorrhage. Based on the Yasargil classification of malformations, 10 lesions (35.7%) were Type I, seven (25%) were Type II, eight (28.6%) were Type III, and three (10.7%) were Type IV. In 11 patients (39.3%), a combined transfemoral, transarterial, and transvenous embolization of the vein of Galen malformation was performed. A pure transtorcular approach was utilized in eight patients (28.6%), and postembolization surgical clipping of arterial feeders was performed in two cases with intractable congestive heart failure. Complete anatomical occlusion of the galenic malformation was achieved in 13 patients (46.4%). An immediate postembolization improvement in the patient's clinical status was obtained in 23 (82.1%) of 28 patients and a good long-term clinical outcome was seen in 17 patients (60.7%). Five deaths (17.9%) occurred in this series of 28 patients; three (10.7%) were related to a transtorcular embolization and two (7.1%) to the unchanged natural history of the disease (68).

Shen et al treated 11 cases of the vein of Galen aneurysmal malformation, one of which was diagnosed by MRI only, and 10 underwent CAG diagnostic procedure. Among the 10 CAG diagnosed cases, 5 were classified as the vein of Galen aneurysmal

malformation with the AV shunt directing to the vein of Galen. The other 5 were classified as the vein of Galen aneurysmal dilitation (VGAD) secondary to parenchymal AVM or dural AVF. Eight cases underwent endovascular treatment. For VGAM, the shunts in the wall of the vein were embolized. For VGAD, the primary AVM or AVF were embolized (69).

Friedman et al described their multidisciplinary team approach using a staged transcatheter embolization and neurosurgical protocol was applied to 22 patients with neonatal presentation of vein of Galen malformations over a 12 year period. Aggressive medical therapy was combined with interventions including: ventricular shunting, transcatheter embolization, retrograde transtorcular embolization, and neurosurgical obliteration. There was a high frequency of high output cardiac failure, multiple organ system dysfunction, seizures, hydrocephalus, visual, developmental and neurological disability. Of the first 11 patients, five survived; four with seizures and three with marked retardation. Of the last 11 patients, six survived; five with seizures but only one with retardation. Despite persistently high morbidity and mortality, their continuously evolving protocol offered these otherwise hopeless patients some chance of survival (70).

Later, Friedman et al described improvement of endovascular technique. Early studies using transcatheter embolization techniques, in a series of 22 patients, had a 50% mortality rate and a 37% incidence of severe mental retardation in survivors. Modifications of embolization techniques and neonatal care have improved the outlook in a more recent series of 11 patients. The diagnosis was established within 3 days of life in 91% of the cases. No mortality occurred, and 6 of the patients were functionally normal

at up to 30 months' follow-up. Although two patients had severe neurologic deficits and/or a seizure disorder, only one case was possibly temporally associated with the embolization procedure. Some developmental delay was noted in one other patient. These improvements result, in part, from modifications of the treatment protocol, including earlier diagnosis, avoidance of digoxin, improvement in the application of newer microcatheters and acrylic polymers (n-butylcyanoacrylate), avoidance of overly aggressive neurosurgical procedures, and the use of stable central vascular access for total parenteral nutrition accompanying other general improvements in neonatal care (71).

A series of 14 vein of Galen vascular malformations diagnosed in the pediatric populations and treated at the Hospital for Sick Children-Necker, Paris, between 1988 and 1994 was repoted by Borthne et al. Five of the patients were diagnosed in the neonatal period, of whom 4 presented with life-threatening, intractable cardiac decompensation and high-flow arteriovenous fistulae. Embolization was performed on vital indications in 4 patients during the first week after birth. One embolization failed with fatal outcome. Of the 3 who were embolized, 2 succumbed within 1 week and 1 survived with marked improvement of cardiac symptoms. The older children presented with hydrocephalus and neurologic symptoms. The 10 patients older than 1 year were embolized. These procedures were successful in 90 %, with hemodynamic stabilization and improvement of clinical symptoms. In this group the mortality rate was 10 %. The total mortality rate was 29 %. Hydrocephalus was secondary to a compression of the Sylvian aqueduct in 44 % of cases. Five patients had ventricular drainage before embolization followed by a staged elective embolization. Transarterial embolizations

were performed in 11 patients, whereas 2 patients were embolized via the transvenous route (72).

Halbach et al reported eight children (six infants and two neonates) who suffered from symptoms caused by a mural-type VGM were treated by means of endovascular therapy. Their age at the time of treatment ranged from 13 days to 19 months (mean 7.6 months). Two neonates and three infants who presented with hydrocephalus and increased head circumference, one of whom was stabilized with a shunt, underwent elective closure of the malformations 3, 4, 6, 6, and 13 months later, respectively. Two patients presented with hemorrhage; one had an intraventricular hemorrhage (IVH) on the 1st day of life and one, a 5-month-old infant, suffered a large parenchymal hemorrhage and an IVH; both patients were immediately cured by means of endovascular techniques. One child presented with a seizure and cortical venous drainage that were treated immediately. Eleven separate treatment sessions were conducted; eight via transarterial femoral access and the remaining three via a transvenous approach. Two patients were treated by using transfemoral transvenous embolization with fibered coils, and one patient required a transtorcular transvenous approach to permit complete closure of the fistula with electrolytically detachable coils. The embolic devices used included silk suture emboli (three patients), electrolytically detachable coils (three patients), and fibered platinum coils (seven patients). In seven patients, complete closure was demonstrated on postembolization arteriographic studies. The eighth patient had stagnant flow in a giant 6-cm varix treated with arterial and venous coils but has not yet undergone follow-up studies. Late follow-up arteriography was performed in four patients at times ranging from 11 to 24 months postprocedure. In one patient, thrombosis of the malformation and

shrinkage of the varix were confirmed on follow-up computerized tomography scanning. The remaining three patients have not yet undergone follow-up angiographic examination. Two asymptomatic complications occurred, including separation of the distal catheter, which was removed with a snare device, and a single platinum coil that embolized to the lung, producing no symptoms in 101 months of clinical follow up. The follow-up period ranged from 3 to 105 months, with a mean of 52 months (73).

.Use of a new procedure of transtorcular embolization with Gianturco embolic coils is described by Mickle et al in three patients harboring vein of Galen aneurysms. Two of the three patients had a satisfactory outcome. This technique is simple and quick, and can produce progressive thrombosis in these high-flow vascular fistulas (40).

Two children presented with congestive heart failure in the neonatal period due to malformations of the vein of Galen. Transtorcular coil embolisation produced rapid relief of heart failure. Post-mortem examination showed occlusion of the malformation in one child. The other child is clinically normal at seven months (74).

King et al described experience in managing two neonates and a 10-year-old boy with vein of Galen aneurysms. The neonates underwent transtorcular embolization, while the boy was treated by standard transarterial balloon embolization. All patients had satisfactory outcomes. The combined surgical and radiologic approaches were felt to be simple when compared to previous surgical techniques, and early results suggested that they are preferable alternatives (75).

Three infants with vein of Galen malformations, all presenting with congestive heart failure, underwent a total of five embolization procedures that employed a percutaneous transfemoral venous approach to catheterize the vein of Galen by Dowd et al. In one instance, direct retrograde catheterization of feeding arterial pedicles to the vein of Galen and embolization of the fistulous connections was achieved via this route. The indications for transfemoral venous treatment included persistent symptoms despite transarterial and transtorcular embolization in one patient, an unsuccessful transarterial embolization attempt (complicated by catheter fracture) in another and the inadvisability of transarterial embolization because of an excessive number of feeding arteries in a third. Complete obliteration of the malformation was achieved in one patient and significant flow reduction in the other two. Vein of Galen perforation with the catheter tip complicated one procedure. All three patients were stable after clinical follow-ups (9-12 months). The transvenous route to the vein of Galen can be undertaken from a transfemoral approach, obviating surgical exposure of the torcular Herophili. In addition, they introduce the concept of direct retrograde catheterization of the feeding arteries to the vein of Galen malformation by a transfemoral venous approach, a procedure that has not been reported previously (42).

Seven cases of vein of Galen aneurysms treated by percutaneous transvenous endovascular occlusion of the aneurysmal vein were presented by Casasco et al. In one case, the approach was via the femoral vein, and in the other six cases, by the jugular vein. All of the malformations were multipedicular and, additionally, in six of the seven there was an intervening arterial-arterial network between the posterior thalamoperforating arteries and the wall of the venous aneurysm. This fistulous network

was interpreted as purely arterial and not as an associated arteriovenous malformation. For this reason, the transvenous approach was considered justified, and was performed without risk of hemorrhage caused by retrograde venous hypertension. Measurement of intra-aneurysmal pressure during the course of treatment allowed better understanding of the hemodynamics of the lesions, guided the amount of occlusion to be accomplished during each treatment session, and thus may have prevented the phenomenon of normal perfusion pressure breakthrough. The percutaneous transvenous approach offers all the advantages of the transtorcular approach but avoids surgery. Because of our excellent angiographic and clinical results--five complete and two partial occlusions, with favorable outcomes and no major complications--they believe that this technique is better for the treatment of multipedicular vein of Galen aneurysms than transarterial embolization or surgery (2).

Rao et al described a series from our Institute. Angioarchitecture of the aneurysmal malformation of the vein of Galen in 9 patients is analysed. An arterial maze arborised directly into the vein of Galen in 6 patients. Direct fistulous communication from one or two pedicles was noted in three children. Unilateral choroidal arterial contribution via an eccentric cul-de-sac in two infants suggested embryonic involvement of the ipsilateral internal cerebral vein along with the median prosencephalic vein of Markowski. Unusually, a sylvian branch of the middle cerebral artery drained directly into the basal vein of Rosenthal which in turn communicated to the aneurysm of vein of Galen in another patient. Two adult patients had curvilinear rim calcification of the venous sac with rapid circulation. All but two showed venous sinus anomalies. Two children had prior shunt surgery at the referring hospitals. The pathophysiology of hydrocephalus and

the possible consequences of shunt, question the need for CSF diversion as a routine in these patients. Percutaneous trans-arterial embolotherapy, in five patients achieved complete obliteration of the malformation in four patients and partial reduction of flow in another. While transvenous/trans-torcular approach is reserved for selected patients and direct surgery carries high morbidity, this report emphasises the efficacy of trans-arterial embolotherapy (76).

Gupta et al reported another series in 2006. Between October 1983 and June 2003, 25 patients with VOGMs were referred to them for evaluation and management. Ten children younger than 2 years of age presented with rapidly increasing head size as the chief complaint. Among 11 children 2 years of age or older, the most common presenting symptom was chronic headache. Four patients who presented during adulthood had chronic headache for many years before presentation. Angiographic evaluation of the lesion was performed in 21 patients. Fifteen patients were treated using endovascular techniques. Injection of the embolic material was performed after induction of systemic hypotension when the flow in the fistula was high. Complete occlusion of the arteriovenous shunt could be achieved in two patients with vein of Galen aneurysmal dilation (100% of patients with this type of malformation) and in five of the six patients with the mural type of malformation (83%). Among patients with the choroidal type of malformation, complete obliteration of the shunt could be achieved in three patients. In three patients with high-flow choroidal malformations, embolization carried out in a single sitting resulted in shunt reduction of nearly 90%. These patients received clinical follow up (37).

AIMS AND OBJECTIVES

1. To review, **prospectively and retrospectively,** all types of VOGMs that were treated in the Department of Imaging Sciences & Interventional Radiology (IS&IR), Sree Chitra Tirunal Institute of Medical Sciences and Technology (SCTIMST), over a period of 26 years.
2. To analyze age of presentation of VOGMs.
3. To analyze different clinical presentations of VOGMs, in relation to different age of presentation.
4. To study the relative frequency and angioarchitecture of both types of VOGMs.
5. To study the management protocol, details of the interventional procedure for VOGM treatment.
6. To analyze the outcome of the endovascular management of VOGMs, in relation to clinical presentation, different routes of approach, various embolic materials used and types of VOGMs treated.
7. To analyze the complications.
8. To revaluate and restandardize the diagnostic and therapeutic protocol for future.

MATERIALS AND METHODS

The design of this study carried out in the department of IS & IR, at SCTIMST, Trivandrum was retrospective as well as prospective. For the purpose of this study, patients who had undergone endovascular management for VOGM in the department of IS & IR, SCTIMST, dating from 1st October 1983 to 31th August 2010 were included. There were a total of 24 cases.

The clinical data of the patients was obtained by reviewing their case sheets obtained from the Medical Records Department & the imaging data was obtained from the DSA lab archive. From these, the data regarding clinical presentation, imaging, angiographic characteristics, details of the embolization procedure, post procedural status and the follow up was collected. For the purpose of this study, post procedure follow-ups were further carried out by sending letter to the patients inviting them to participate in the follow up study or else to participate by responding to questionnaires.

A total of 24 cases (age range; newborn- 28years, sex ratio; male: female =17:7) of VOGM were treated. All patients underwent a complete cardiac and neurological evaluation prior to the procedure. The patients had a baseline imaging done – either a USG or CT or MRI of the brain. Subsequently, the patients underwent a complete four vessel diagnostic cerebral angiogram. All the studies were performed on Advantx digital substraction angiography unit (GE Milwaukee, USA) or Innova biplane flat panel digital substraction angiography unit (GE Milwaukee, USA). A wide variety of catheters and embolic material was used for the procedures as was best suited for the individual patient depending on the angioarchitecture.

Diagnostic angiographies were done under local anesthesia when they were performed separate from interventional procedure or performed as a check angiography for follow up. The interventional procedure was done under general anesthesia. Premedication (Inj. Pethidine 25-50mg i.m & Inj. Phenergan12.5- 25mg i.m) was given before the local procedures. Post procedure these patients were managed in the wards when angiography was performed under local anesthesia. For procedures carried out under general anesthesia, the patient was monitored in the neurological intensive care unit.

After discharge, the patients were followed up with clinical evaluation for improvement in their symptoms. Follow up imaging or angiograms were performed when required.

Patients who were not regular in follow up were contacted by sending letter to their home address as per hospital record and were asked to participate in the follow up. Patients who were unwilling to visit the hospital were requested to respond to the questionnaire send along with the letter.

A complete evaluation of the patient including demographic profiles (age, sex), clinical presentation, imaging features, treatment method and outcome including complications and follow up was carried out as per the proforma attached.

MATERIAL USED FOR ANGIOGRAM AND EMBOLIZATION

1.	Sheath – Radiofocus, Cordis, Impulse	4,5, 6, 7 French size
2.	Diagnostic catheters – Vertebral glide (Terumo), Right coronary (Cordis), Mani cerebral (Cordis), Simmons (Cordis), Multipurpose (Cordis)	5, 4 French size
3.	Guide wire – Terumo exchange length, Terumo standard,	150 cm – Standard 260 cms - Exchange
4.	Guiding catheters – Vistabrite, Launcher (Cordis)	6, 7 French size
5.	Microcatheters –Tracker, Spinneker, Magic, Echelon	1.5 to 1.8 French
6.	Microguidewires – Transcend, Agility, Dasher, Mirage.	.008, .010, .014 of varying length (190-205cm)
7.	Embolic materials –Glue (Isobutyl or N-Butyl Cyanacrylate), Coils – GDC, matrix, Berenstein liquid coil, Vein of Galen coil; autologous muscle	Coils varying no-1 to 18 Glue 17%-95%

TECHNIQUES OF EMBOLISATION-

The procedures were performed under general anesthesia. A guiding catheter was placed in the main feeding arteries In anterior feeders, guiding catheter was placed in vertical portion of petrous ICA and then feeders were selectively cannulated with microcather, positioned near the

fistula and were embolized with coil or N-Butyl cyanacrylate. In posterior feeders, guiding catheter was placed in V2 segment and then feeders were cannulated and embolized using coil or glue. In some cases, the fistula was accessed via the venous route. A femoral vein puncture was followed by placement of the guiding catheter into the internal jugular vein (IJV). The microcatheter was then negotiated into the sinus and coils were placed to occlude the fistula. The coils were released after confirming their position in the sac. A complete check angiogram was performed after all embolization procedures.

POST PROCEDURAL MANAGEMENT-

The patient was shifted to the ward or the neurological or cardiac ICU for 24 hours after the procedure as required. They were extubated in the cathlab itself or later in the ICU as decided by the anesthetist concerned and was decided as a case to case basis. Patient was kept nil orally for 6 hours or until the return of cough reflex and were gradually started with clear fluid. Lower limbs were immobilized for 12 hours to avoid puncture site bleeding. Vitals and peripheral pulses were monitored and neurological and cardiological status monitored frequently.They were kept on intravenous antibiotics as per body weight and was given antiedema measures with dexona (3-6mg, TDS) and mannitol (25-75 ml TDS), (for 3 days) when felt necessary. Blood pressure was controlled meticulously to avoid perfusion pressure breakthrough phenomenon.

RESULTS

A total of twenty four patients had undergone endovascular treatment for vein of galen malformation in the last twenty six years in the department of Imaging Science and Interventional Radiology at Sree Chitra Tirunal Institute for Medical Science and Technology, Trivandrum.

Among 24 patients seventeen (71 %) were males and the rest seven (29 %) were females with age at presentation ranging from newborn to twenty eight years (mean age 4.8 years). Table 1 shows the age distribution of the patients at presentation and at first sitting of embolisation.

TABLE 1

	Presentation (Number)	Presentation (Percentage)	Embolisation (Number)	Embolisation (Percentage)
Fetus	1	4.17	NIL	
Neonate (<1 month)	3	12.5	0	0
Infant (1 month-2 years)	11	45.83	12	50
Children (2-16 years)	7	29.17	10	41.67
Adult (>16 years)	2	8.33	2	8.33

Presenting complaints were increased head size, headache, prominent scalp vein, delayed milestone, poor scholastic performance, neurological deficit, vomiting, seizure, subarachnoid hemorrhage, irritability and cardiac failure. Presenting complaints are summarized in Table 2.

TABLE 2

Presenting complaints	Number	Percentage
Increased head size	9	37.5
Headache	8	33.33
Delayed milestone	3	12.5
Prominent scalp vein	1	4.17
Neurodeficit	8*	33.33
Poor scholastic performance	1	4.17
Seizure	0	0
Subarachnoid hemorrhage	1	4.17
Vomitting	6	25
Irritability	3	12.5
Cardiac failure	6	25
Antenatal	1	4.17

*- including 1 with paralytic polio.

Other than the abovementioned presenting complaints, patients had other unrelated manifestations, complaints and past history including facial dysmorphism, hernia, hydrocele, pigmented macules, hyperactivity, history of tuberculosis and history of meningitis and paralytic polio. They are summarized in Table 3.

TABLE 3

Past history or complaints	Number	Percentage
Facial dysmorphism	1	4.17
Hernia	1	4.17
Hydrocele	1	4.17
Pigmented macules	1	4.17
Hyperactivity	1	4.17
History of tuberculosis	1	4.17
History of meningitis	1	4.17
Paralytic polio	1	4.17

On clinical examination at our out patient clinic, patients had increased head circumference, neurological deficit, prominent scalp vein or signs of cardiac failure. They are summarized below in Table 4.

TABLE 4

Examination findings	Number	Percentage
Increased head circumference	8	33.33
Neurological deficit	8	33.33
Prominent scalp vein	2	8.33
Cardiac failure	5	20.83
Bruit	3	12.5
No signs	8	33.33

Analysis of echocardiography revealed associated cardiac manifestations including atrial septal defect (ASD), ventricular septal defect (VSD), patent ductas arteriosus (PDA), patent foramen ovale (PFO), transverse arch hypoplasia (TAH), upper pulmonary vein hypoplasia (PVH), cardiomegaly, pulmonary hypertension (PHT) and trivial MR. Table 5 summarises the echocardiography findings of the patients.

TABLE 5

Echocardiography features	Number	Percentage
ASD	4	16.67
VSD	1	4.17
PDA	1	4.17
PFO	1	4.17
TAH	1	4.17
PVH	1	4.17
Cardiomegaly/LVH	4	16.67
PHT	3	12.5
MR	1	4.17

Laboratory investigations revealed liver function abnormalities in three patients. They are summarized in Table 6.

TABLE 6

Liver function derangement	Number	Percentage
Deranged LFT	2	8.33
Transient isolated hyperbilirubinemia	1	4.17

One patient revealed rachitic changes in epiphyses in skeletal X Ray with elevated Alkaline phosphatase is in serum.

Cross sectional imaging data including transcranial ultrasonography, CT scan and MRI scan analysis revealed predominant findings as hydrocephalus, brain atrophy, calcification, infarct and partial thrombosis of venous sac. Features are detailed in Table 7 as below.

TABLE 7

Features on Imaging	Number	Percentage
Hydrocephalus	14	58.33
Cerebral atrophy	3	12.5
Calcification	3	12.5
Infarct	1	4.17
Hemorrhage	1	4.17
Partial thrombosis of venous sac	1	4.17

All the patients underwent angiography before endovascular treatment. One patient underwent conventional angiography and other 23 underwent digital subtraction angiography. Angiography images were analysed to determine the type of malformation

(choroidal or mural), to identify feeders (anterior choroidal, posterior choroidal, P1 perforators, pericallosal, dural branch or others), to analyse the draining venous sinuses and their anomalies (persistent falcine sinus, atretic straight sinus, atretic lateral sinus, persistent occipital and marginal sinus and others) and associated anomalies.

Table 8 shows the type of malformations.

TABLE 8

Type	Number	Percent
Mural	13	54.17
Choroidal	11	45.83

Table 9 depicts the distribution of arterial feeders.

TABLE 9

Feeding arteries	Number	Percent
Anterior choroidal	8	33.33
Posterior cerebral including Posterior choroidal	24	100
Pericallosal	4	16.67
Dural (occipital)	2	8.33
Others including SCA, PICA	5	20.83

Table 10 shows the result of analysis of venous drainage pathway.

TABLE 10

Venous features	Number	Percent
Persistent falcine sinus	8	33.33
Persistent occipital and marginal sinus	9	37.5
Atretic straight sinus	11	45.83
Atretic transverse sinus	3	12.5
Duplicated straight or transverse sinus	2	8.33
Atretic superior sagittal sinus	1	4.17

Table 11 shows the associated other vascular anomalies as below.

TABLE 11

Associated vascular anomalies	Number	Percent
Persistent proatlantal artery	3	12.5
Rt aortic arch	2	8.33
Aberrant left subclavian with Kommorels diverticulum	1	4.17
Hypoplastic aortic arch	1	4.17
Bovine CCA origin	1	4.17
Direct aortic origin of VA	2	8.33
Interrupted IVC	1	4.17

Persistence of Limbic arch was visualized in 2 patients (8.33%).

Embolisation was performed by arterial or combined (arterial and venous) approach. For embolisation material used were glue, coils, both glue and coils or autologous muscle. Number of embolisation sittings for one patient were variable between 1-6, a total of 34 sittings for 24 patients (average 1.42 sittings per patient). Number of coils used varied from 1-18. Concentration of glue used varied between 15%-98% (15, 17, 20, 25, 33, 40, 50, 60, 66,80,75,90, 95 and 98).

Table 12 summarises the routes of embolisation used.

TABLE 12

Routes of embolisation	Number	Percent
Arterial	15	62.5
Venous	0	
Combined	9	37.5

Table 13 summarises the type of embolising materials used.

TABLE 13

Embolising material	Number	Percent
Glue	11	45.83
Coil	2	8.33
Both Glue and Coil	10	41.67
Autologous muscle	1	4.17

Embolisation results were interpreted as absolutely complete occlusion when there was no residual contrast filling seen across the shunt. It was graded as near complete occlusion when approx 90% or above was embolised and partial when 50-90% was embolised. It was regarded as incomplete when less than 50% was embolised. Table 14 summarises the embolisation results.

TABLE 14

Therapeutic result	Immediate post procedure- Number (%)	Follow up imaging of surviving children- Number (%)
Absolutely Complete	9 (37.5)	11 (55)
Near complete	11 (45.83)	8 (45)
Partial	3 (12.5)	0
Incomplete	1, (4.17), to be staged	1 (5)

One patient with complete occlusion, 1 patient with near complete occlusion died, 2 patients with partial occlusion died. Three patients who showed near complete occlusion on immediate postprocedure angiography showed complete occlusion on subsequent imaging. One patient with partial occlusion on immediate post procedure angiogram showed near complete occlusion on subsequent imaging.

Complications after therapeutic embolisations were analysed and grouped as transient neurological complication (seizure or focal neurological deficit that subsequently improved), permanent neurological complication, nonneurological compication (access route complication, limb ischemia, systemic dysfunction) and hemorrhage after embolisation. They are summarized as below in Table 15.

TABLE 15

Complication	Number	Percent
Transient neurological complication	3	12.5
Permanent neurological complication	0	0
Nonneurological compication	2	8.33
Hemorrhage after embolisation	2	8.33

Intraprocedural technical complications were also analysed and migration of glue or coil across the venous side was found to be the most common technical complication. They are analysed in Table 16.

TABLE 16

Procedural complication	Number	Percent
Migration of glue	3	8.82
Prolapse or migration of coil	2	5.88
Perforation of artrery	2	5.88
Reflux of glue to feeder	1	2.94

Three patients underwent shunt surgery for CSF diversion and 1 underwent ventriculostomy. 1patient developed shunt related meningitis.

Two patients showed improvement immediately after embolisation but were lost to follow up and never turned up for a subsequent follow up or imaging. Clinical follow up for the rest of the 18 patients who survived initial postembolisation period was obtained for a duration ranging from 15 days to 11 year 10months (mean 4.3 years). One of them expired after 6months due to shunt related meningitis. Clinical follow up of the patients is as follows in Table 17.

TABLE 17

Clinical status	Immediate- Number (%)	Long term- Number (%)
Improvement	20 (83.33)	17 (77.27)
Death	4 (16.67)	5 (22.73)

DISCUSSION

Management of children with high flow arteriovenous shunts of the brain is among the most challenging areas in modern medicine. Intracranial arteriovenous shunts in children differ considerably from those seen in adults, in whom brain arteriovenous malformations and acquired dural arteriovenous fistulae predominate. These differences are seen both in the types of lesion and in their effects. In the neonatal and infantile age groups, the most common type of arteriovenous shunt is the vein of Galen aneurysmal malformation. Progressing further into childhood, dural malformations and brain arteriovenous malformations become more common. The consequences of a shunt in the developing brain are different from those in an adult, principally because of the immature cerebral venous system. The arachnoid granulations by which cerebrospinal fluid will be returned to the cerebral venous sinuses are not fully matured until 16–18 months of age. In infancy, cerebrospinal fluid is reabsorbed across the ventricular ependyma and brain parenchyma into the medullary veins. The presence of VOGM may raise venous sinus pressure, which is transmitted in turn to the cortical and finally the medullary veins. This will result in water congestion of the brain parenchyma, and impaired oxygenation leading to subependymal atrophy and in severe cases a progressive "melting brain Syndrome". The most common presentation of VOGM results from the size of the shunt itself, imposing elevated preload on the right side of the heart leading to cardiac failure. This may progress to multisystem failure. Haemorrhage in children with VGAMs is rare. These are rare lesions and experience in their management has been restricted generally to large paediatric centres where a close collaboration between neuroradiologists, neonatologists, paediatric cardiologists, and neurologists has been achieved. Foremost among these centres has been Bicetre Hospital in Paris where Professor Pierre

Lasjaunias' group has done much to clarify the nature of the disease and its appropriate management. A typical neurosciences unit serving a population of about three million could expect approximately one new VOGM patient a year. The aetiology of VOGMs is unknown; however, an early insult, perhaps resulting in a somatic mutation in neural crest and/or adjacent cephalic mesoderm in the early embryo, could be expected to cause such vascular abnormalities.

DEMOGRAPHIC PROFILE-

We had 1 antenatally diagnosed patient (4.17%), 3 (12.5%) were diagnosed at neonatal age, 11(45.83%) in infancy, 7(29.17%) in childhood and 2(8.33%) were diagnosed as adult. In Bicetre hospital series they had 29.3% of cases diagnosed as fetus, 37.5% in neonatal age, 25.9% as infant and 7.3% after 2 years of age (77). This is quite in contrast to our patients' demographic profile. Lesser availability of antenatal ultrasonography in our country may be responsible for so less number of antenatal detection of this easily identifiable ultrasonographic substrate. Presentation in adults is also rare but has been reported in literature. There is report of presentation upto 36 years of age (8, 78-80).

This proportion is in sharp contrast with the findings of Johnston, et al., who found that, of 232 cases collected from literature, only 18% of the patients presented at an age older than 5 years (52). In our series 33% patients were older than 5 years at presentation.

Sex ratio in our series was in accordance with literature. 71% of our patients were mlaes and 29% females with a male:female ratio of 2.5:1. Bhattacharyya et al reported it to be 3:1 in a review, a figure similar to ours (81).

PRESENTATION-

Presentation of vein of galen malformation patients can be varied. Our series had 9 (37.5%) with increased head size, 8 (33.33%) with headache, 8 (33.33%) with neurological deficit, and 6 (25%) with different manifestations of mild to severe cardiac failure. Other manifestations were prominent scalp vein in 1 (4.17%), delayed milestone in 3 (12.5%), poor scholastic performance in 1 (4.17%). 6 (25%) had recurrent vomiting, 3 (12.5%) had irritability and 1(4.17%) presented with subarachnoid hemorrhage.

Nine among 15 children who were younger than 2 years of age presented with rapidly increasing head size as the chief complaint. Increasing head circumference as presenting complaints were not at all observed in those presenting after 2 years. Three children in this group also had delay in achieving developmental milestones. Neurological deficits were present in 4 patients in this age group. Although six children had a history of respiratory distress at birth or symptoms related to cardiac cause, on examination 5 showed varying degrees of cardiac signs. All five had choroidal type of malformation. This is in accordance to the evidence from literature which states that choroidal type is usually associated with cardiac failure (80). Only one child had clinically significant severe congestive heart failure at the time of presentation. Four children had recurrent vomiting. 2 had increased irritability. One child in this group was having dilated facial vein as observed by the parents and another revealed same in clinical examination.

Among 9 patients who were 2 years of age or older, the most common presenting symptom was chronic headache. Headache was present in 8 among those 9 patients and interestingly headache was not present at all in patients who presented before 2 years of age. Delay in achieving neurological developmental milestones was also not observed in this group; however poor scholastic performance were seen in 1 child. Four patients also had neurological deficits. Presentation with subarachnoid hemorrhage was seen in one patient. Two patients in this group had recurrent vomiting. One patient had increased irritability who also had history of TB, TB meningitis and recurrent cough and fever.

Both patients who presented during adulthood had experienced chronic headache from many years before presentation. Among these, one patient presented to the hospital with intraventricular hemorrhage and subarachnoid hemorrhage. In the other, the presence of the malformation was detected during investigations for the cause of chronic headache, recurrent vomiting and neurodeficit.

Other than the above mentioned presenting complaints, patients had other unrelated manifestations, complaints and past history including facial dysmorphism, hernia and hydrocele, pigmented macules, hyperactivity, history of tuberculosis and history of meningitis and paralytic polio. An extensive pubmed search failed to show any correlation of these features with either type of vein of galen malformation and are better accepted as incidental findings in the patient group.

On physical examination 3 patients showed bruit, 1 had a neck-carotid bruit and 2 had bruit heard over head posteriorly as well as over mastoid. All were less than 2 years of age, 1 had mural and 2 choroidal type of malformation.

CARDIAC FINDINGS-

Analysis of echocardiography revealed associated cardiac manifestations in total 11 patients (45.83%), 6 with choroidal and 5 with mural malformation. Among those with Echocardiographic abnormality, clinical cardiac failure was present in only 5, all with choroidal type. None of those with mural type of malformation, even with structural defect in heart had cardiac failure, stressing the role of extracardiac shunt in pathogenesis of congestive cardiac failure in vein of galen patients. Atrial septal defect (ASD), cardiomegaly or left ventricular hypertrophy was the most frequent ECHO finding, being present in 4 patients each (16.67%). Pulmonary hypertension (PHT) happened to be the next most frequent manifestation present in 3 patients (12.5%). Pulmonary hypertension was always associated with left to right shunt, either ASD in 2 patients or PDA in 1 patient. Ventricular septal defect (VSD), patent ductas arteriosus (PDA), patent foramen ovale (PFO), transverse arch hypoplasia (TAH), upper pulmonary vein hypoplasia (PVH), and trivial MR were present in 1 patient each (4.17%). Among the 6 patients who had complaints related to cardiac failure, five showed presence of varying grades of failure on clinical examination. One neonate had severe cardiac failure at birth necessisating intensive care monitoring and early intervention. This unfortunate male newborn had complex congenital cardiac malformation including ASD, Pulmonary

hypertension, Transverse arch hypoplasia, Right upper pulmonary vein hypoplasia and interrupted IVC. Other patients with mild to moderate clinical failure had PDA and PHT, PFO and PHT, tiny muscular VSD and PFO, and isolated cardiomegaly respectively.

Cardiac manifestations are known in vein of galen malformation, constitutes an important component in the pathophysiology of the disease process and also has been prenatally diagnosed. In contrast to the cardiac failure observed in large hemangiomas, where they occur in infancy at the proliferative phase of the disease, the congestive cardiac failure in VOGM can be present during the neonatal period.

In his series of 18 antenatally diagnosed VOGM patients, Rodesch noted that 17 were born with cardiac failure. During prenatal ultrasound examination, some cadiac enlargement was noted in four out of 17 patients. In all the four patients the neonatal score was low (<8/21) either because of the significant peripheral effect of systemic failure or because of an already demonstrable encephalomalacia. Traetment was withheld in those four patients and they soon died. The others were medically managed, followed, embolised between 2 and 13 months transarterially. A total of 30% of them had slight retardation (less than 20%), which resolved in a few months after embolisation.

Therapeutic termination of pregnancy can be discussed in cases in which cardiac failure and or brain damage is demonstrated in utero (17).

With regards to the spontaneous evolution of CCF, the following has been suggested by Lasjaunias et al (77). After a brief period of stabilization, in most patients the CCF worsens during the first 3 days of life, and then stabilizes again to then improve with appropriate medical management. They noticed that in none of the babies referred to them with the diagnosis of VOGM did cardiac failure develop de novo after the 2nd week of life. However, it can decompensate at 3weeks or recur later following lung infection or other concomitant diseases. In infants CCF never constituted the presenting complaint nor did it worsen at that age if already present. We also observed similar pattern with none of the patients developing denovo failure at later age nor did it worsen after neonatal period.

Cardiac manifestations and degree of failure seems to be variable from one child to the other and seems independent of the characteristics of the shunt. Some obvious high flow lesions are well tolerated, while conversely some apparently smaller ones may lead to multiorgan failure. Hence the intracranial hemodynamic parameters available do not provide us with any prognostic or therapeutic information.

Renal and hepatic damage may further aggravate CCF, and their function can be transiently impaired (oliguria, increase of enzyme) or become rapidly unstable despite intensive medical care. One of our patients had transient neonatal hyperbilirubinemia and subsided on its own and was considered as physiological neonatal hyperbilirubinemia regarding its temporal evolution pattern. Two patients (8.33%) had deranged liver function test. One of them had grossly elevated serum alkaline phophatase and

Radiographic features of Ricket. This patient had undergone embolisation at 6months of age, showed improvement clinically and biochemically and doing well on long term follow up with total obliteration of the fistula. Another patient who had complex congenital cardiac anomalies as stated above with ASD, Pulmonary hypertension, Transverse arch hypoplasia, Right upper pulmonary vein hypoplasia and interrupted IVC also had deranged liver function test. He underwent early partial embolisation at 2 months of age, followed by right lower limb gangrene, cardiac arrest, ventricular fibrillation, renal dysfunction, lactic acidosis, consolidation and rapidly progressive downhill course and death. Experience with this unfortunate neonate reemphasizes the inevitable natural history in presence of severe failure and associated systemic organic dysfunction in neonatal presentation. If we retrospectively calculate the Bicerte neonatal score for this neonate, it is obviously less than 8. This neonatal score being a recent development in assessment of patients with vein of galen malformation, patient with a score less than 8 is destined for a poor prognosis and need not be treated. We hope to apply this score in future in all our patients scheduled for management which can avoid unnecessary procedures except when we are hard pressed for social indications to treat a child.

The cause of CCF is not fully understood. In fetal life, the effect of heart rate on combined ventricular output suggests that heart is functioning near its maximum performance. It seems that volume loading increases output to a limited extent. The fetal myocardium has less contractile tissue, as shown by its less myofibrillar contents. Several major events change the fetal circulation at birth: removal of low pressure placental

circuit, reversal of relative pressure between two atria with closure of foramen ovale, muscular contraction of ductus, and decrease in pulmonary vascular resistance.

CCF is mainly seen with choroidal type of malformation as seen in all cases of our series. Lasjaunias et al shared same experience. They stated that severe forms of CCF are associated with persistence of fetal type of circulation. Septal communications and ductus arteriosus are often noted and they should not be considered as associated cardiac malformations, even if they increase the systemic insufficiency (77). Like most of the disorders encountered in these circumstances, they either disappear spontaneously or following endovascular management of the AV shunt itself. They should be followed with special attention if embolisation is not planned early, and they may induce a failure to thrive condition.

In the series by Lasjaunias et al, two neonates presented with an associated cardiac malformation and an aortic coarctation for which they were first operated on: embolisation was then carried out at 1 and 2 month of age. Five and ten years follow up revealed satisfactory clinical outcome. In two other patients they decided to clip a patent ductus arteriosus before embolising the VOGM in neonates with severe CCF (77). None of our patients had undergone any cardiac procedure. In strict sense, only the neonate with complex cardiac malformation described above had true associated cardiac malformation. But in presence of systemic decompensation with organ failure it was doubtful if it could survive cardiac intervention pre embolisation.

Among four deaths in our series that happened post embolisation three had features of cardiac failure at presentation. One death was ascribed to SAH and IVH that occurred peri procedure. One case with complex congenital cardiac malformation as described above was definitely related to cardiac decompensation state. The third patient had PFO, PHT and associated right aortic arch with mirror image branching pattern. There was gross hydrocephalus with cerebral atrophy. Post embolisation he expired due to cardiac failure and pulmonary aspiration and infection.

NEUROLOGICAL FEATURES-

All the patients irrespective of the presenting complaints had undergone cross sectional imaging in the form of CT, MRI or Transcranial USG. They revealed predominant findings as hydrocephalus in 14 (58.33%), followed by brain atrophy and calcification in 3 patients each (12.5%). Infarct, hemorrhage and partial thrombosis of venous sac were less frequent and seen only in 1 patient each (4.17%).

Among patients with hydrocephalus 7 were having mural type and the rest 7 choroidal type of malformation. Eight were below 2 years of age and the rest above. Lowest age of the patient with hydrocephalus was newborn and the highest age of the patient with hydrocephalus at presentation was 7 years. None of the 2 adult presentations had hydrocephalus. Seven among 9 patients with increasing head size as presenting complaint had hydrocephalus. Among patients with headache 4 had hydrocephalus and 4 did not. Among patients with neurodeficit 5 had it and 3 did not. Among those with recurrent

vomiting 4 had hydrocephalus and 2 did not have. Among 6 patients with cardiac complaints 4 had hydrocephalus. However, all 4 patients with delayed development of milestone and poor scholastic performance had ventriculomegaly. Hence delayed socioscholastic development had consistent association with hydrocephalus on imaging.

As opposed to CCF, hydrodynamic disorders can manifest themselves in fetuses, neonates and infants. Choroidal and mural types almost equally give rise to this type of manfestations (77). They constitute the primary revealing factor at infant age if the diagnosis has not been made before. They result from the abnormal hemodynamic conditions present at the torcular venous sinus confluence, the posterior convergence of the venous drainage of the brain, and the immaturity of the granulation system of arachnoid villi. For many years, the mechanical compression of the nmesencaphalic aqueduct was considered to be the primary factor behind hydrocephalus. Actually aqueduct is usually present in all cases and this observation was consistently made in our patients as well. Macrocrania, while resulting in an increasing head circumference, is associated with slightly enlarged ventriclesd and a generous increase in perivascular spaces. The water dysfunction combines intracebral retention with an increase in cerebrospinal fluid volume. Both phenomena have little or no effect on the brain itself as long as the suture enlarges, since they tend to continually adapt to intracranial pressure vs the resistance by the cranial vault. On the other hand, in VOGM infants the lack of macrocrania is even more worrisome than its presence. The cerebrofugal medullary veins constitute a gradient that will induce absorption of most of the intracerebral water. If the suture stop growing, if the medullary vein resorption decreases (or if the pial venous

pressure increase), or if for any other reason the compliance of the venous system fails, hydrocephalus and intracranial hypertension occur.

At infant stage, persistence of the situation leads to clinical manifestations in the form of irritability, altered level of consciousness and neurological status, developmental delay, decreased brain volume with increased fluid space. Before ventricular enlargement occurs, intracranial pressure is not as high because of macrocrania, and therefore shunting is not required. Spontaneous stabilization of enlarging head phenomena can occur with cavernous capture of the sylvian veins. The progression of macrocrania to hydrocephalus is thus not inevitable. Going with this explanation all our patients with enlarged head size did not have ventriculomegaly.

Developmental delay is part of the natural history of untreated VOGM. Careful evaluation of neurocognitive performance shows that most children with macrocrania present some degree of mental retardation. In view of the poor prognosis of the disease, specialists and parents tend to accept as normal a child with mild retardation (up to 20% of normal for chronological age). This level of delay allows the child to attend a normal school albeit with some support.

Although there is no direct relationship between the degree of macrocrania and the severity of developmental delay (the head enlargement actually protects the childs brain), there is an obvious link between the hydrodynamic disorder and the delay. Any event that creates a loss in compliance has an impact on brain maturation, eg, intracranial

hypertension, spontaneous decrease in head circumference and ventricular shunting. All the four children with either developmental delay or poor scholastic performance had hydrocephalus in our series, this attests to the association.

Cerebral morphological sequelae express themselves in calcifications, subepebdymal atrophy (pseudoventriculomegaly), and eventually the stigmata of previous acute accidents with cortical and subcortical atrophy. The insult to the brain is a slow and permanent one.

Calcifications are three types. Mural, in lesion itself where the calcification is a result of partial or complete thrombosis, at the subcortical level in the white matter where they reflect hydrovenous failure. The latter occurs where the medullary vein loses its ventriculocortical gradient and its activity is shifted from subpial to medullary level. These calcifications are usually bilateral and symmetrical, located preferentially in the frontal region. The occipital lobe region is often affected earlier with atrophy and thinning of splenium of corpus callosusm. Calcifications can be asymmetrically located, mostly in unilaterally shunted children and often on the side opposite to the shunt. They are not caused by arterial steal. Any transient episode of hydrocephalus may give rise to calcifications, since it expresses the loss of compliance of the fragile hydrovenous system functioning in infant. A third type of calcification is located in the striatum and caudate and putamen bilaterally and symmetrically. They express subacute ischemia in the region of the prominent transcerebral collateral circulation system for telencephalic veins. Striate vein congestion occurs after the cortical veins can no longer drain the cerebral

white substance or when the persisting thalamic pathway are overloaded with the drainage of the parietooccipital system. The calcifications indicate both the mechanism and specific vulnerability of this area at the infant stage. The clinical manifestations do not parallel the intensity of these calcifications. Some of them demonstrated during infancy after a brief episode of increased intracranial pressure may be absent or remarkably reduced on follow up. It happened like this with one of our patient with parenchymal calcification. Therefore, although indicative of the previous ischemic insult, the calcifications do not have the predictive value for neurological outcome in a treated VOGM. They rarely produce abnormal movement disorders that are most often seen with more posteriorly located damage. We had two patients with parenchymal calcification and 1 with mural calcification in the wall of the sac. One calcification in the parenchyma showed gradual fading trend after embolisation.

Spontaneous thrombosis of the VOGM is rare. In the experience of Lasjaunias, 2.5% patients showed spontaneous thrombosis, but only half of them were neurologically normal, which is less than what is achieved by proper treatment (77). In addition, this thrombosis is mostly unpredictable, although the tentorial edge compression of the arterial feeder stogether with the secondary intraluminal thrombosis in the stenosed draining vein might be an indication of such a development. In any event this thrombosis tends to occur late, when cerebral damage is already irreversible. It is possible to make a retrospective diagnosis of a completely excluded VOGM. One of our patients had partial thrombosis of the venous sac, however he was treated appropriately and none of our patients had spontaneous exclusion of shunt by thrombosis.

Cerebral angiography showed predominance of feeders from posterior cerebral artery in our patients. All patients had posterior cerebral contribution in their malformation. Of interest, 2 patients had dural supply apparently supplying the sac from meningeal branch of occipital artery and 5 patients had apparent contribution from cerebellar arteries. Cerebellar ateries do not usually contribute to the supply of the VOGM, except indirectly through their dural branches, which can be enlarged, as they may participate in the supply of the vasa vasorum at the venodural junction. (77). Other dural contributions can be seen in true VOGM and may be located at a distance from the choroid fissure shunting zone. They often represent secondary dural AV shunts after sinus thrombosis or AV dural communications caused by sump effect from an otherwise patent sinus (77). Analysis of angiographic results also showed associated venous anomalies in the form of persistent fetal type of circulation. There was persistent occipital and marginal sinus in 33.33% and 37.5% patients respectively. Atretic straight sinus was present in 45.83% and atretic lateral sinus in12.5% patients. 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. Persistent occipital sinus connecting torcula to sigmoid-jugular by marginal sinus is another connection. Retention of fetal patterns of venous drainage could prevent development of other sinuses such as the straight sinus and transverse sinus. Retention of the embryonic pattern of vasculature can explain the presence of several vascular anomalies that are associated with these lesions (7, 8).

MANAGEMENT-

Twenty of the 24 patients treated by embolization had complete or near complete obliteration of the fistula, as demonstrated by angiographic studies immediately after the procedure. Four patients underwent partial or incomplete embolization (all four patients with choroidal malformation).

In seven of eleven patients with choroidal malformations, complete or near complete obliteration of the shunt was achieved. In three patients with high-flow choroidal malformations, embolization carried out resulted in partial shunt reduction of nearly 90%. These patients received clinical follow up. Two of them expired; the third patients improved and follow up imaging showed near total obliteration. One patient with incomplete embolisation which was done recently has been called for a second sitting. Among 13 patients with mural type of malformation, all showed total or near total obliteration on immediate post embolisation check angiogram.

Lasjaunias et al reported 100% occlusion in 82 of 193 patients, 95% in 8, 90% in 16, approx 50% in 75 and less than 50% in 12 patients (among surviving children). So in their series among surviving children approx 42.49% had complete, 12.44% had near total, 38.86% partial and 6.22% had incomplete embolisation (77). In our series, 55% had complete, 45% near complete and 5% incomplete embolisation. Results are similar.

Results were better for mural malformation with all malformations (100%) of mural type undergoing total or near total occlusion demonstrated in immediate post procedure angiogram. Hence mural malformations with limited number of feeders opening directly into venous sac are comparatively easy to treat with high predictability for cure. Only 1 patient with mural malformation expired where as 4 patients with choroidal malformation died.

Complications after therapeutic embolisations were analysed and grouped as transient neurological complication (seizure or focal neurological deficit that subsequently improved) (in 3 patients, 12.5%), permanent neurological complication (0 patients), nonneurological compication (access route complication, limb ischemia, systemic dysfunction) (in 2 patients, 8.33%) and hemorrhage after embolisation (2 patients, 8.33%). In the study by Lasjaunias et al, in surviving children, transient neurological complication was reported in 1.55%, permanent neurological complication in 2%, non neurological complication in 6.7%, hemorrhage after embolisation in 5.7%. There was death in 23 out of 216 embolised patients (10.6%) (77). In the series by Fullerton et al, embolization in 27 patients resulted in 61% having no or mild developmental delay and a 15% mortality rate during hospitalization (67). In our series death was in 5 out of 24 patients, among which 1 death was post shunt meningitis sequelae. Hence 4 deaths after embolisation (16.67%) is comparable figure. If we consider only surviving children, then the complication rates, namely transient neurological complication and nonneurological complication are 2 in19 (10.5%) and 1 in 19 (5.26%) patients respectively. So we had marginally more mortality and more transient neurological complication but less of

permanent neurological complication, hemorrhage and nonneurological complication in our series.

In two patients, intraventricular hemorrhage occurred during the periprocedural period. The first patient was a 28-year-old man who had a history of recurrent headaches since childhood and presented with intraventricular hemorrhage. Angiographic evaluation revealed a choroidal type of malformation. The venous sac was partially coiled using a detachable platinum coil (Guglielmi Detachable Coil; Boston Scientific/Target Therapeutics, Inc., Fremont, CA) to achieve flow reduction and trap the escaping glue thus preventing venous glue migration. Subsequently, as the arterial feeders were being catheterized for embolization using cyanoacrylate, extravasation of contrast from one of the feeders was detected. This feeder was embolized immediately proximal to the leak site to prevent progression of hemorrhage. A postprocedure CT scan was performed, which revealed intraventricular and subarachnoid hemorrhage. External ventricular drainage was performed immediately, but the patient died of bleeding 3 days later.

The second patient was a boy who presented with hydrocephalus. He had undergone VP shunt placement at 1 year of age. Angiographic evaluation revealed a mural type of malformation fed by the posterior thalamoperforator and thalamogeniculate branches of the posterior cerebral artery. Initially, using transarterial access, one of the feeders was cannulated, and a liquid coil (Berenstein Liquid Coil; Boston Scientific/Target Therapeutics, Inc.) was released to reduce the flow through the fistula. Subsequently, through a femoral venous access, the venous sac was cannulated, and seven vein of Galen

coils (Boston Scientific/Target Therapeutics, Inc.) were deployed. Then the thalamogeniculate branch feeding the fistula was cannulated and embolized using 15% NBCA (Histoacryl; B. Braun). The patient underwent elective ventilation after the procedure. About 3 hours after the procedure, the child developed pupillary asymmetry. A CT scan of the brain revealed intraventricular and subarachnoid bleeding and ventriculomegaly. Despite immediate placement of external ventricular drainage and institution of barbiturate coma, the patient died of bleeding 2 days later.

One infant with a choroidal VOGM underwent endovascular therapy in two sessions, resulting in 70% reduction and total occlusion of the shunt, respectively. The infant had no symptoms immediately after the procedure. However, 7 days later the infant developed irritability with neck rigidity. The clinical diagnosis of meningitis was confirmed by subsequent imaging and laboratory investigations. Despite aggressive management with antibiotics, the infant had a prolonged downhill course characterized by ventriculitis and hydrocephalus and died 5 months later.

Another male newborn presented immediately after birth with congestive cardiac failure. He had deranged LFT on biochemical examination and complex cardiac malformation including ASD, Pulmonary hypertension, Transverse arch hypoplasia, Rt upper pulmonary vein hypoplasia and interrupted IVC. He was taken for embolisation at an age of 2 months for shunt reduction to improve the cardiac failure. Angiography revealed choroidal type of malformation and embolisation was attempted through arterial route using 25 and 66% NBCA (Histoacryl; B. Braun). We could achieve partial embolisation

with >50% reduction of fistula. Post procedure, the infant developed Rt lower limb gangrene, cardiac arrest, ventricular fibrillation, renal dysfunction, lactic acidosis, consolidation and expired.

Another newborn who presented with increased head size from birth, delayed milestone, irritability, recurrent vomiting, neurological deficit and cardiac failure was diagnosed to have patent foramen ovale and pulmonary hypertension on echocardiography. He had hydrocephalus and gross cerebral atrophy. He underwent angiography and embolisation at an age of 9 month. He also had right aortic arch with mirror image branching diagnosed during angiography. He had earlier undergone ETV and VP shunt before embolisation. He was treated with NBCA by arterial route using glue in 98% concentration followed by embolisation with 2 coils placed in venous sac in another sitting after 2 weeks. Embolisation produced only partial reduction of shunt. He expired due to cardiac failure after aan episode of aspiration and subsequent pulmonary infection. Transient neurological deficits in the form of gaze paresis occurred in one patient immediately after embolization and spontaneously resolved within 3 days. After embolization one patient had transient hemiparesis, which resolved completely within 6 days. Another child had partial migration of glue to superior sagittal sinus and 1 episode of seizure after embolisation but improved subsequently with no further seizures in postprocedure period. One other child had a procedure related spasm of the iliac artery on the side that was used for arterial access. This complication was largely asymptomatic and resolved with conservative therapy.

In one infant with a choroidal malformation, there was migration of the first detachable platinum coil that was placed within the sac. Although the size of the coil was appropriate, after detachment of the coil, the high flow of the shunt resulted in migration of the coil into the falcine sinus and proximal part of occipital sinus. The procedure was continued and subsequent coils stayed within the venous sac at the site of detachment. The patient did not have any new neurological deficits after the procedure. In another procedure when attempting to negotiate the microcatheter over microguidewire, there was microcatheter perforation without any sequelae, the microcatheter was changed and the procedure continued subsequently. In another patient a Berenstein coil migrated while coiling through arterial route. It migrated via venous side to the pulmonary circulation and went to right lower lobe. There was only mild cough but no respiratory difficulty in immediate post procedure period and the patient is doing well at long term follow up of more than 7 years. In another patient there was migration of glue to the venous side without any sequelae. In another patient there was glue migration to venous side and left PCA branch. Thalamic infarct was identified in post procedure imaging but the patient did not have any new deficit and done well on long term follow up after embolisation.

All 20 surviving patients who underwent successful embolization and had improvement in clinical status beyond the post procedural period had variable clinical follow up. Duration of clinical follow up ranged from 15 days to 11 year 10 months (mean 4.3 years). All patients remained neurologically stable at follow up and did not develop any new symptom related to the malformation. One patient died after 6 months due to shunt related meningitis.

All patients who had normal cognitive functions before embolization continued to have normal functions and threee patients who presented with delayed milestones at the time of intervention had improvement after embolization. Follow-up angiograms demonstrated persistence of angiographic occlusion in all patients who had complete obliteration of the shunt.

Five patients had undergone embolisation in 2 sittings and 1 patient had undergone embolisation in 6 sittings. Rest of the 18 patients had single sitting for embolisation. The patients who underwent multiple sittings are mostly of choroidal type (4 choroidal types among 6). Three patients who showed near complete occlusion on immediate postprocedure angiography showed complete occlusion on subsequent imaging. One patient with partial occlusion on immediate post procedure angiogram showed near complete occlusion on subsequent imaging. These findings are highly important in the management of these lesions. Several authors have substantiated the efficacy of partial embolization to prevent congestive heart failure in neonates with high-flow lesions. This strategy may be extended to lesions presenting in other age groups. In centers with limited resources, partial embolization may be preferable to aggressive management of the whole lesion, especially in high-flow lesions such as choroidal malformations. Reduction of the arteriovenous shunt by partial embolization may promote further thrombosis and result in complete obliteration of the fistula without the need for a second procedure. Decisions regarding continuation of conservative management or timing of further embolization can be made on the basis of clinical follow up and periodic transcranial Doppler ultrasonography.

We routinely use vasodilator-induced hypotension during the injection of glue. This technique helps in the setting of glue at the site of the fistula. In the postprocedural period we maintain mean blood pressure 10 to 15% below baseline to avoid hemorrhagic complications. Maintenance of blood pressure below baseline also helps to reduce the flow within any residual shunts and aids in progressive thrombosis of the lesion, as we have seen in our cases. Postprocedural systemic hypotension also prevents normal perfusion pressure breakthrough, which can be associated with abrupt cessation of large arteriovenous shunts. However, induction of hypotension before closure of the fistula may be best avoided in children who have presented with congestive heart failure. The large shunt across the intracranial arteriovenous fistula significantly reduces the diastolic pressure within the aorta, causing reduced coronary artery flow. Induction of hypotension in these children may further reduce the subendocardial blood flow and result in myocardial infarction.

Only two patients in our series had major fatal complications which can be directly attributed to the procedure. We could not determine the exact source and mechanism of bleeding in these patients, as the relatives of the patients did not give consent for autopsy. Partial coiling of the venous sac was performed initially to achieve reduction of the arteriovenous shunt and trapping of the glue in both patients. In the first patient, extravasation of contrast medium was noted during catheterization of the arterial feeders after coil embolization of the venous sac. Venous hypertension induced by coiling of the venous sac could have been responsible for rupture of a subependymal vein, resulting in hemorrhage. The initial presentation of this patient with intraventricular hemorrhage may

have played an important role in the occurrence of this complication. In such patients, aggressive coiling of the venous sac to achieve reduction of the arteriovenous shunt may be avoided. Instead, reduction of arterial inflow with transarterial glue embolization may be performed. In the other patient with major complications, massive subarachnoid hemorrhage occurred about 3 hours after the procedure. Review of the angiographic images revealed a bleblike protrusion from the anterior aspect of the venous sac, at the point of entry of the feeders. This protrusion was likely the source of bleeding in this patient. Careful assessment of the angioarchitecture to identify such potentially weak spots may permit a less aggressive approach in the management of these lesions. Staged embolization may be carried out in such cases.

SUMMARY AND CONCLUSION

To summarise, our experience shows

1. A male and infantile preponderance, sizeable adult presentation and less of antenatally diagnosed cases.

2. Less than 2 years of age, there was more of presentations with macrocrania, thereafter headache was the dominant clinical feature.

3. Cardiac failure was usually associated with choroidal type of malformation.

4. Cardiac failure is independent of associated cardiac structural defect and is mainly a result of the extracardiac shunt.

5. Adult patients presented with headache.

6. Neurodevelopmental delay and cognitive decline was consistently associated with hydrocephalus.

7. Macrocrania is not invariably related to hydrocephalus.

8. Cardiac failure starts from neonatal age; it never started de novo later in life.

9. In contrast to CCF, hydrodynamic disorder of brain can manifest at any age.

10. Child with low neonatal score is destined to have a bad prognosis and this score needs to be calculated in all patients at the time of presentation.

11. Mural malformation is easier to treat.

12. Once completely occluded, VOGM never recanalises.

13. Near total or partial embolisation will progress to complete occlusion by subsequent thrombosis and can be helpful in limited resource condition and selected cases.

14. Aggressive coiling through venous route into the venous compartment is better avoided; instead transarterial approach for reduction of shunt is a favorable option.

To conclude, this is an Institutional experience with endovascular management of a considerable number of patients with Vein of Galen Malformation. The experience has taught us the natural history of the disease, has made us wiser in selecting embolisation protocol and decide on management options and we hope to continue this good work to help those unfortunate children who are born with this deadly malformation.

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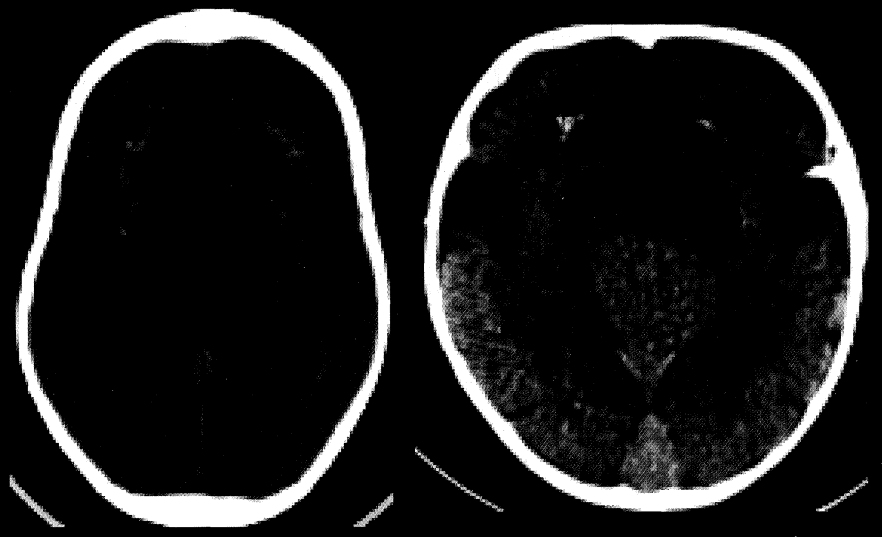
ABBREVIATIONS

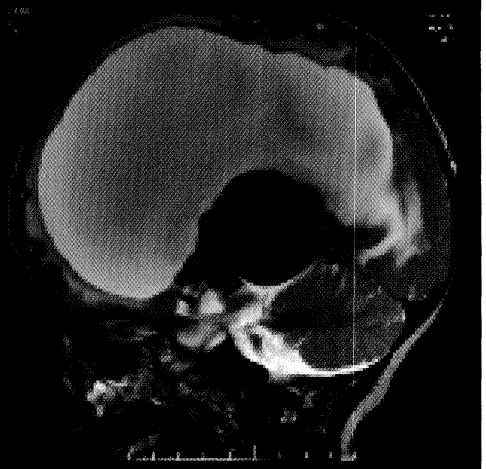
ASD- Atrial septal defect
AV-Arteriovenous
AVF-Arteriovenous fistula
AVM-Arteriovenous malformation
CAG-Cerebral angiography
CCA-Common carotid artery
CCF-Congestive cardiac failure
CHF-Congestive heart failure
CSF-Cerebrospinal Fluid
CT-Computed Tomography
DSA-Digital subtraction angiography
ECHO-Echocardiography
EEG-Electroencephalogram
ETV-Endoscopic third ventriculostomy
FiO2-Fractional inspired oxygen
IVH-Intraventricular hemorrhage
IJV-Internal Jugular vein
ICA-Internal cerebral artery
ICU-Intensive care unit
IVC-Inferior vena cava
LFT-Liver function test
MR-Mitral regurgitation
MRI-Magnetic Resonance Imaging
MR angiography-Magnetic resonance angiography
NBCA-N butyl cyanoacrylate
PCA-Posterior cerebral artery
PDA -Patent ductas arteriosus
PFO -Patent foramen ovale
PHT- Pulmonary hypertension
PICA-Posterior inferior cerebellar artery
PVH- Upper pulmonary vein hypoplasia
SAH-Subarachnoid hemorrhage
SCA-Superior cerebellar atery
TAH- Transverse arch hypoplasia
TB-Tuberculosis
TDS-Three times daily
USG-Ultrasonogram
VA-Vertebral artery
VGAD-Vein of Galen aneurysmal dilatation
VOGM-Vein of Galen Malformations
VP-Ventriculoperitoneal
VSD- Ventricular septal defect

ANNEXURE1-REPRESENTATIVE IMAGES

IMAGING FEATURES

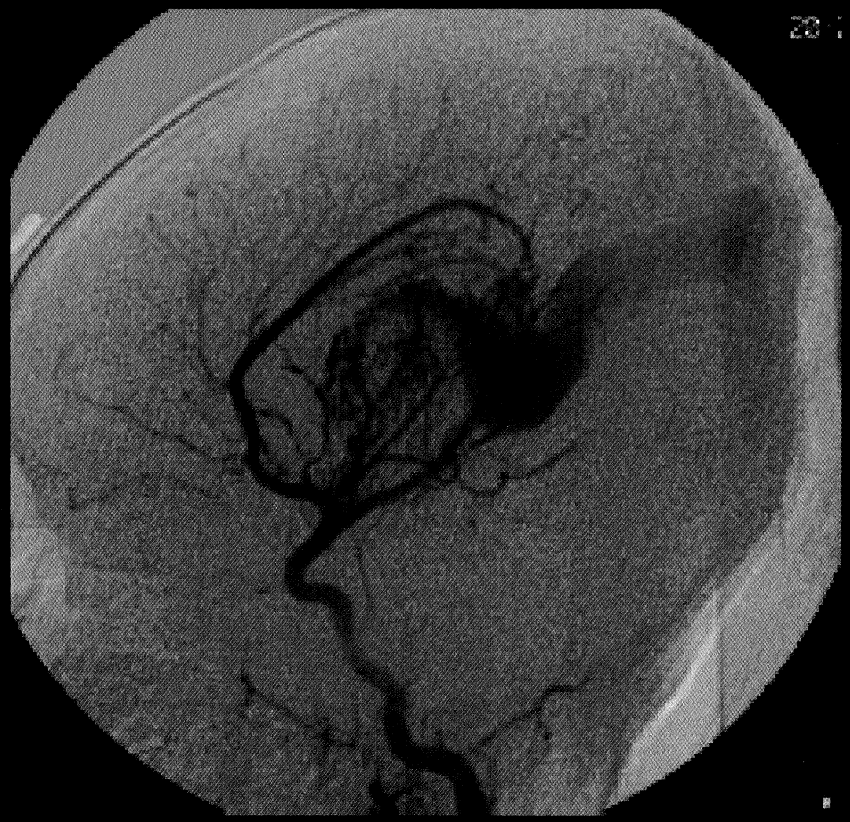


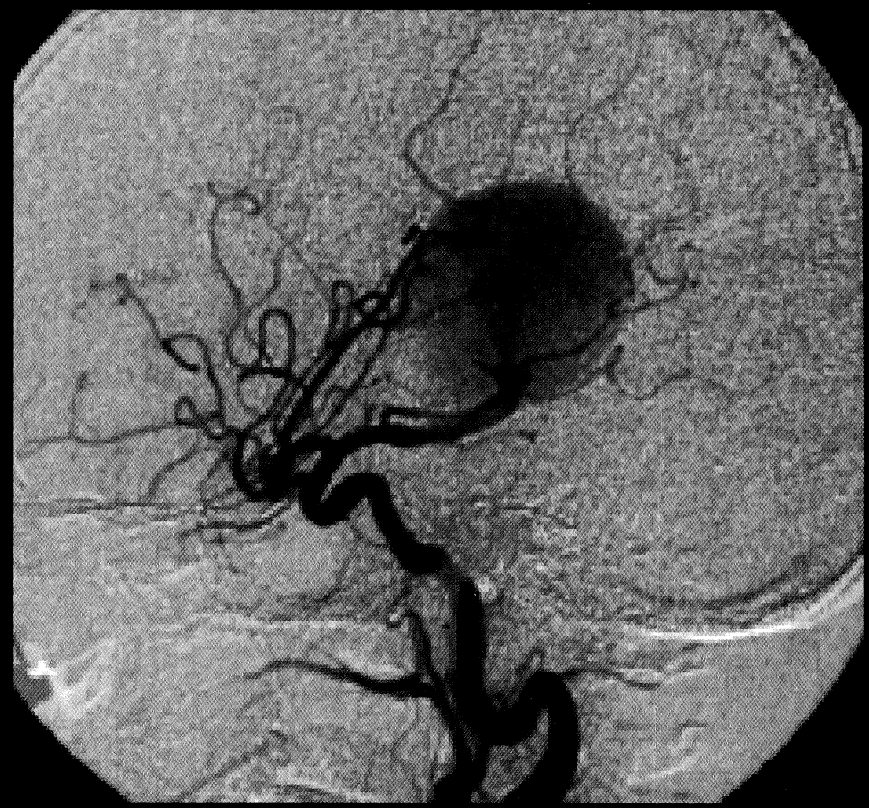
Cerebral Parenchymal Calcifications



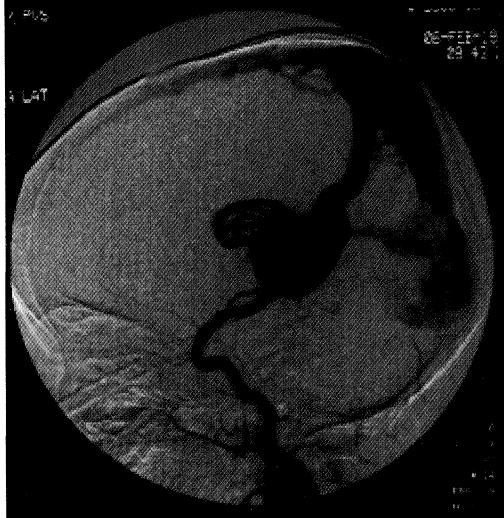
Hydrocephalus

CHOROIDAL AND MURAL

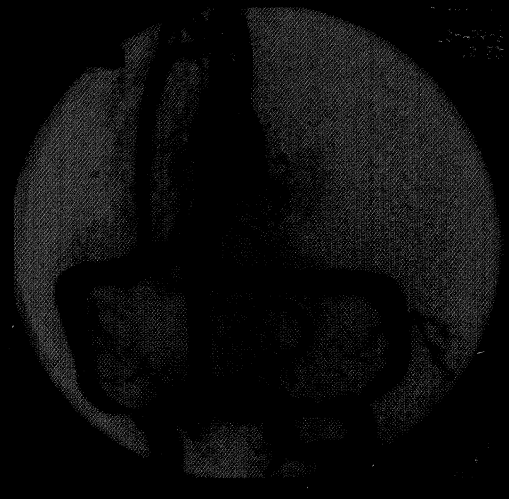




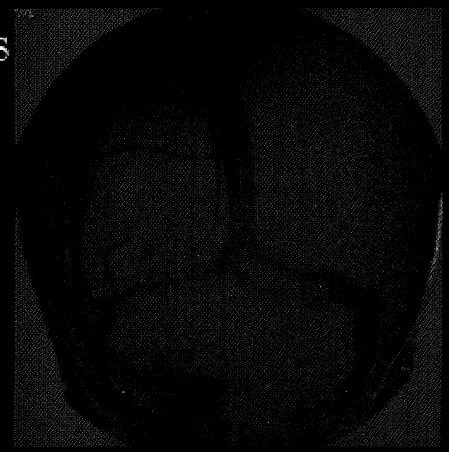
VENOUS ANOMALIES



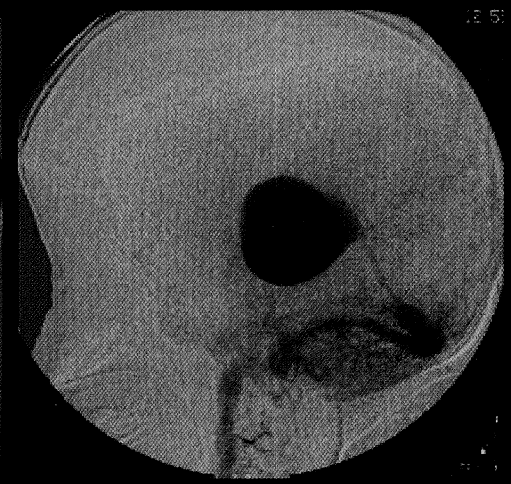
FALCINE SINUS



OCCIPITAL SINUS

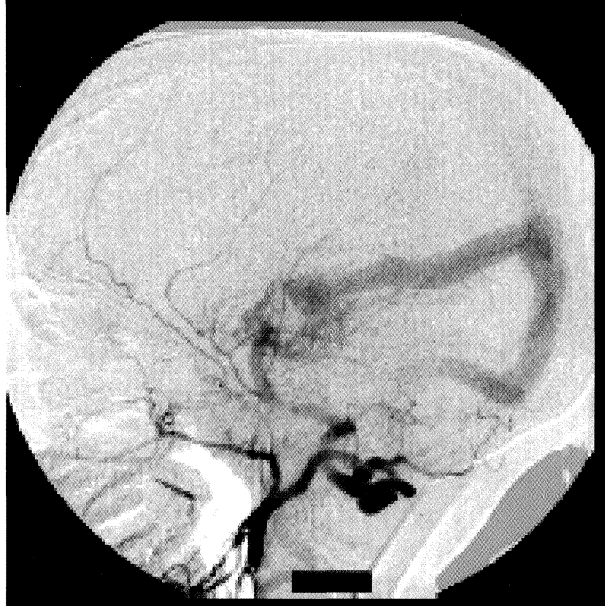


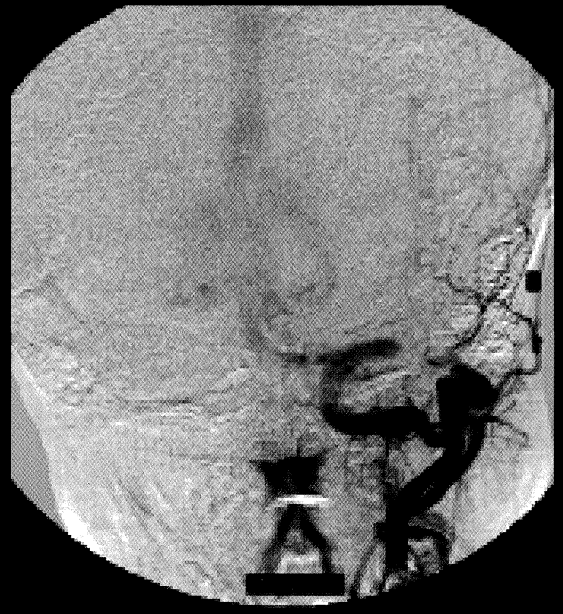
DUPLICATION OF SINUS



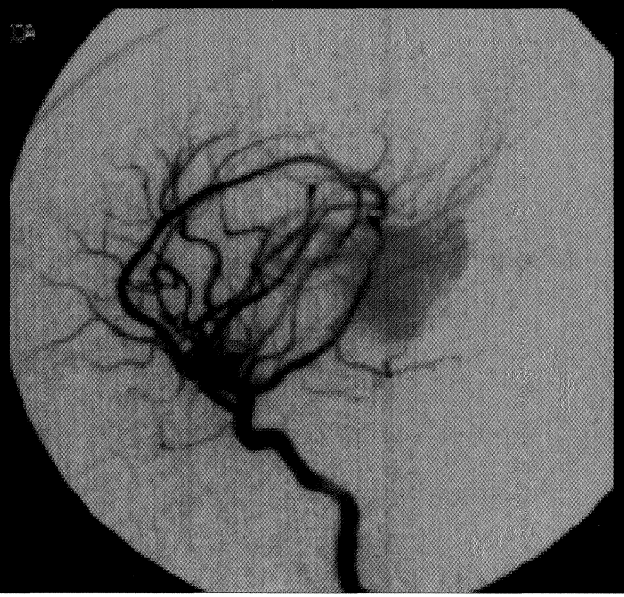
ATRETIC STRAIGHT SINUS

ARTERIAL ANOMALIES



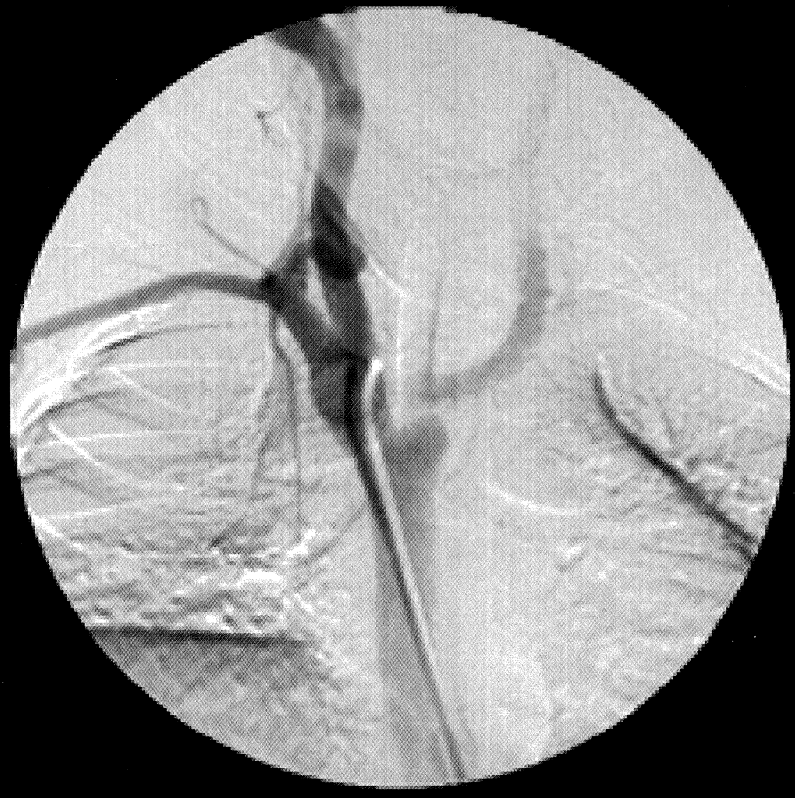


PERSISTENT PROATLANTAL INTERSEGMENTAL ARTERIES

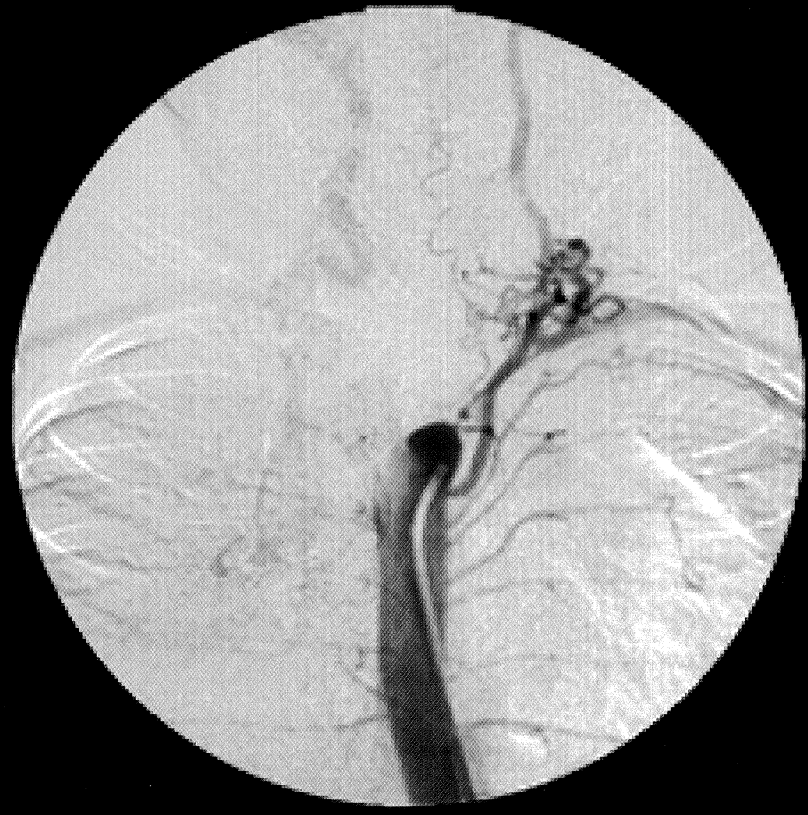


LIMBIC ARCH

ARTERIAL ANOMALIES

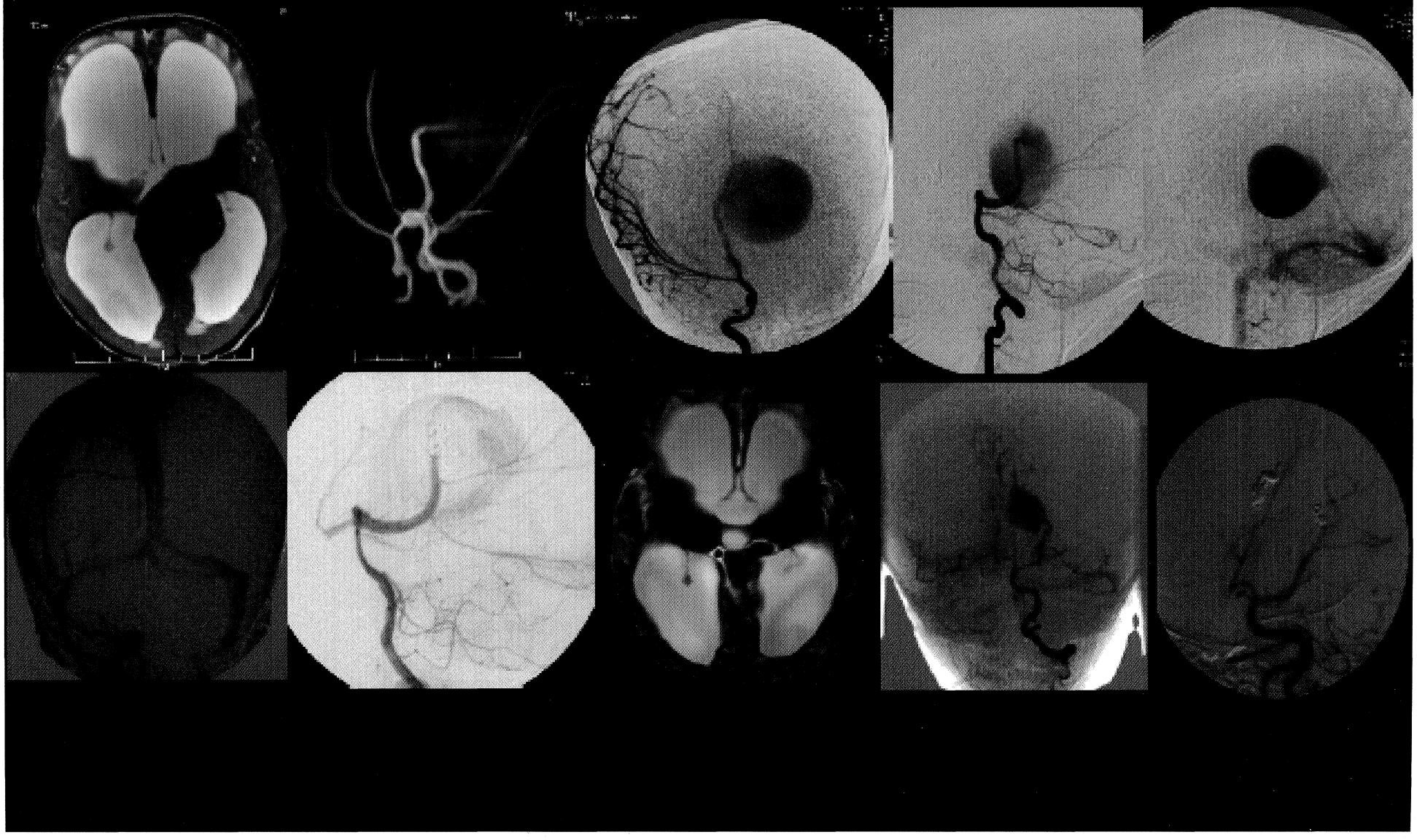


RIGHT AORTIC ARCH

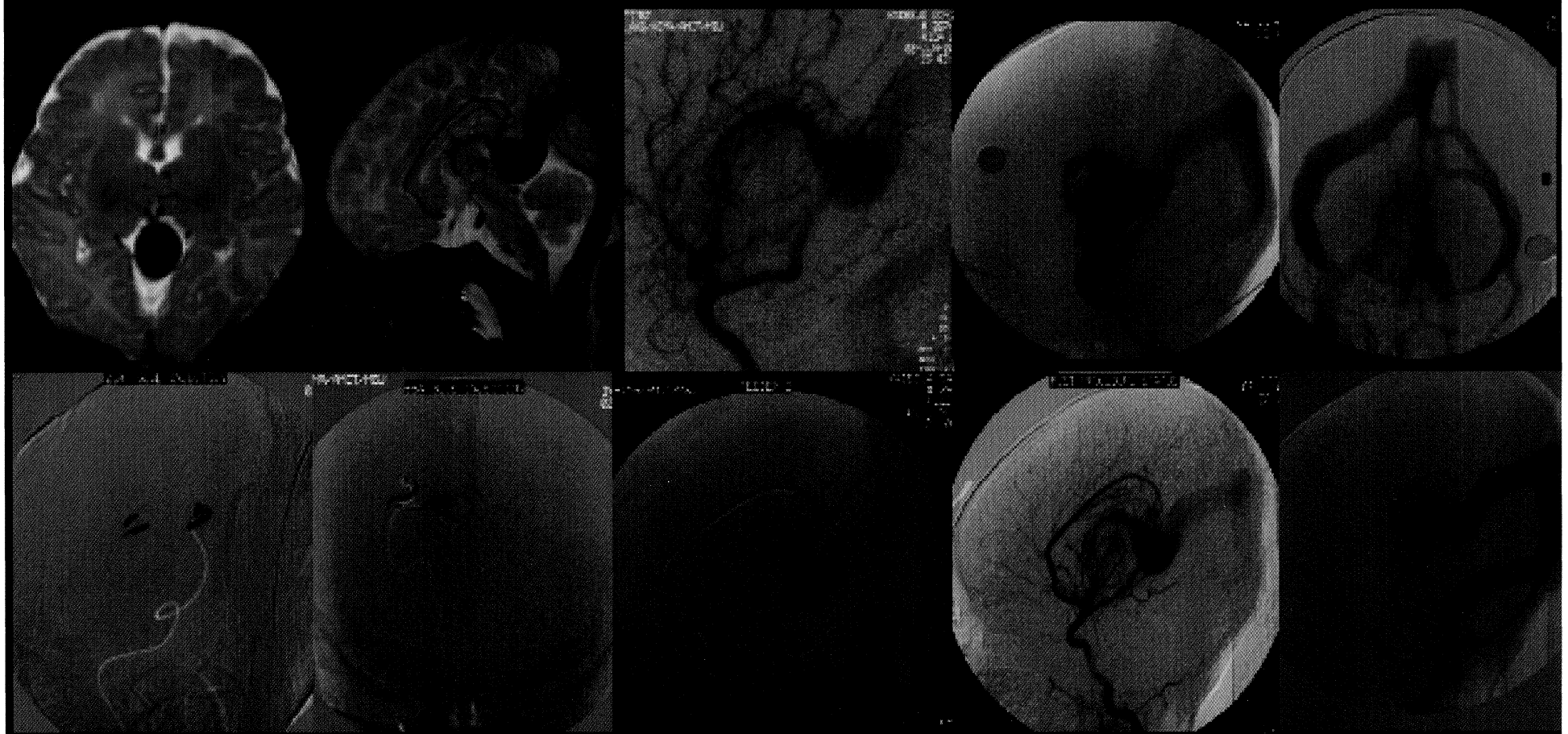


KOMMEREL'S DIVERTICULUM

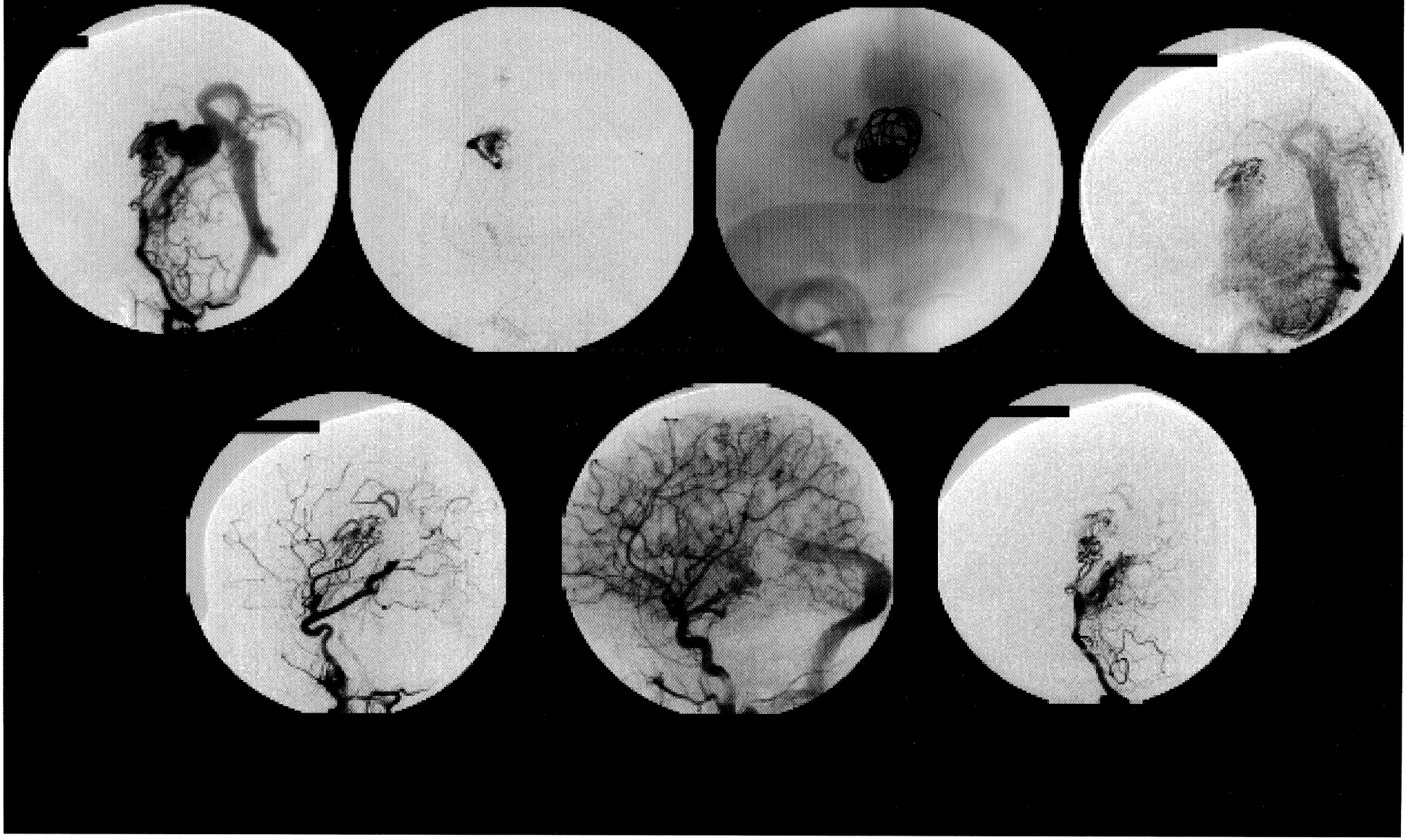
9mth old male child, presented with progressive increase in head size



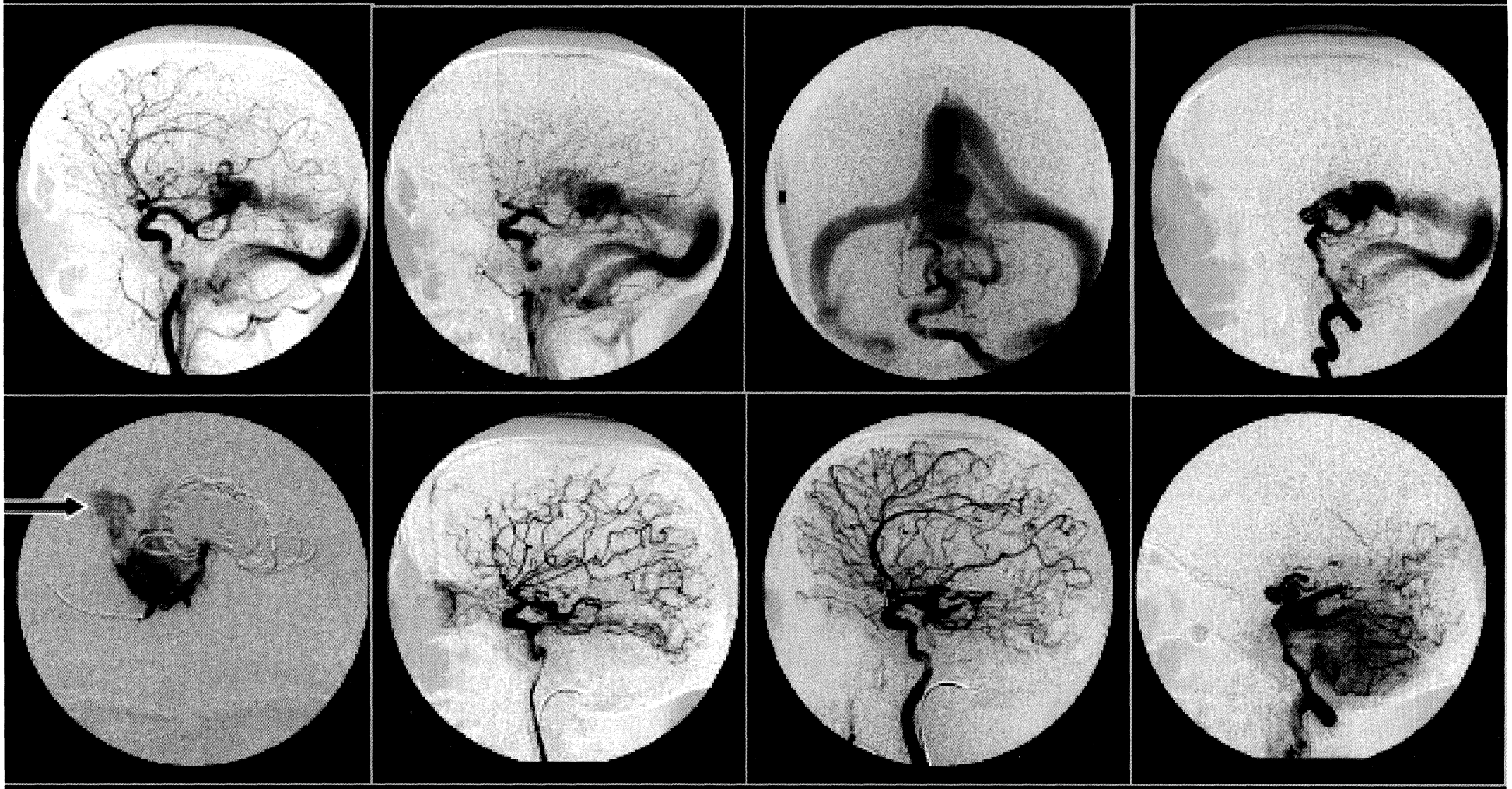
2mth old female child presented with feeding difficulty. Associated cardiac anomaly



8 year old male child with chronic headache



20 year old male patient, presented with IVH



hemorrhage after transvenous coiling. ? Due to rupture of small vein. Managed by immediate reversal of anticoagulation, glue injection and external ventricular drainage. Patient expired 3 days later

ANNEXURE2-PROFORMA

Sl. No-

Name- **Hospital No-** **Age at presentation-**

Sex- **Address-** **Contact no-**

Referring Hospital/Physician-

Admission dates- **Discharge dates-**

Presentaion

Neurological- **Increased head circumference**

Headache

Delayed milestone

Prominent scalp vein

Neurodeficit

Poor scholastic performance

Seizure

SAH

Others

Cardiac-

Antenatal-

Others-

Examination

Neurological-

Cardiac-

Developmental-

Echocardiography-

USG/CT/MRI Head- **Ventriculomegaly**

Cerebral atrophy

Parenchymal infarct/edema

Parenchymal calcification

Wall calcification

VOGM thrombus

Others

DSA- **Type- choroidal/mural**

Feeder

Associated venous anomaly- persistent falcine sinus

occipital sinus

atretic straight sinus

other atretic sinus

duplication of sinus

venous sac at torcula

Associated arterial anomaly-

persistent limbic arch

Rt aortic arch

kommorels diverticula

persistent proatlantal artery

others

LFT- **RFT-**

Treatment - Embolisation /VP shunt/ ETV

embolisation route- arterial/venous/combined

material- muscle/balloon/glue/coil/combined

Complication-

IVH

Spasm

transient neurodeficit

pulmonary embolism

shunt infection/obstruction

other

death

Follow up- Duration-

Imaging/Angiographic- complete occlusion/near total occlusion

Clinical- improved clinical status-milestone/static/deteriorate/death

ANNEXURE3-QUESTIONNAIRE

Dear.....,

Being an embolized (old patient) of this institute we would like to know your current health status. I request you to come to our OPD (NROPD) with this letter on any working day before 15th August for medical check up in the morning at 10AM.

In case you cannot come please send back this following proforma after filling it up and putting it in the self-addressed stamped envelope that accompanies this letter. Please post the letter at the earliest.

1. Do you have severe headache	-	Yes	No
2. Do you have visual problem other than spectacle related	-	Yes	No
3. Do you have any paralysis or weakness of hand or leg	-	Yes	No
4. Do you have any abnormal sensation in body	-	Yes	No
5. Do you suffer from recurrent cough and cold	-	Yes	No
6. Do your legs swell up when you walk	-	Yes	No
7. Do you need to visit cardiologist or neurologist regularly	-	Yes	No
8. Do you suffer frequently from breathing difficulty	-	Yes	No
9. Are you better after the embolisation at our hospital	-	Yes	No

Thanks for your cooperation.

Sincerely,

(Dr. A. K. Gupta)
Prof. & Head
Dept. of IS & IR

ANNEXURE4-MASTER CHART

Serial no	Age presenting	Sex	head size increase	headache	milestone d	scalp vein	neurodeficit	poor scholastic performance	seizure
1	0	m	0	0	0	0	0	0	0
2	6yr	f	0	0	0	0	0	0	0
3	28yr	m	0	1	0	0	1	0	0
4	1yr 2 months	f	1	0	0	1	0	0	0
5	1yr	m	0	0	0	0	1	0	0
6	1yr6mth	m	1	0	0	0	0	0	0
7	1yr6mth	m	0	0	0	0	0	0	0
8	7yr	m	0	1	0	0	1	0	0
9	6yrs	m	0	1	0	0	1	0	0
10	0	m	1	0	1	0	1	0	0
11	0	m	0	0	0	0	0	0	0
12	7mth	m	0	0	1	0	1	0	0
13	4yr	f	0	1	0	0	0	0	0
14	14days	f	0	0	0	0	0	0	0
15	6mth	m	1	0	0	0	0	0	0
16	6mth	m	1	0	1	0	0	0	0
17	27yr	f	0	1	0	0	0	0	0
18	1yr 9mth	m	1	0	0	0	1	0	0
19	10yr 9mth	f	0	1	0	0	1, due to paralytic polio le	0	0
20	3mth	m	1	0	0	0	0	0	0
21	10mth	m	1	0	0	0	0	0	0
22	9yr	m	0	1	0	0	0	1	0
23	5yr 5mth	f	0	1	0	0	0	0	0
24	5month	m	1	0	0	0	0	0	0

SAH	vomitting	Irritability	antenatal	cardiac failure	other	neurodeficit	head size l
0	0	0	1	1	facial dysmorphism	0	0
0	0	0	0	0	incidental, common cold-CT done	0	0
0	1	0	0	0	0	1	0
0	0	0	0	0	bruit	0	1
0	1	0	0	0	hernia,hydrocele	0	0
0	1	0	0	0	hyperactive	0	0
0	0	0	0	0	incidental,carotid bruit	0	0
0	0	0	0	0	0	0	0
0	0	0	0	0	0	1	0
0	1	1	0	1	hyperpigmented macule on abd	1	1
0	0	0	0	1	**deranged LFT**	0	0
0	0	0	0	0	h/o meningitis,left mastoid bruit	1	0
0	0	1	0	0	h/o tbm, cough,fever	0	0
0	0	0	0	1	**transient neonatal hyperbilirubinemia**	0	0
0	1	1	0	0	0	1	1
0	0	0	0	1	deranged LFT, alkaline ptase 2623u/l, cupping of epiphyses	0	1
1	0	0	0	0	0	0	0
0	0	0	0	1	0	1	1
0	0	0	0	0	0	1, left leg polic	0
0	0	0	0	0	0	0	1
0	0	0	0	0	0	0	1
0	0	0	0	0	0	1	0
0	1	0	0	0	0	0	0
0	0	0	0	0	0	0	1

prominent scalp vein	cardiac signs	cardiac structural defect	hydrocephalus	cerebral atrophy
0	1	PDA, PHT	0	0
0	0	0	1	0
0	0	0	0	0
1	0	0	1	0
0	0	asd	0	0
0	0	concentric LVH	1	0
0	0	trivial mr	0	0
0	0	0	1	0
0	0	0	1	0
0	1	PFO,PHT	1	1
0	1	asd,pht,transverse arch hypoplasia, rt upper pulmonary vein hypolasia	0	0
0	0	cardiomegaly	1	0
0	0	0	0	0
0	1	tiny muscular vsd,pfo	1	0
0	0	small os asd	1	1
0	1	cardiomegaly	1	0
0	0	0	0	1
1	0	asd,cardiomegaly	0	0
0	0	0	0	0
0	0	0	1	0
0	0	0	0	0
0	0	0	1	0
0	0	0	1	0
0	0	0	1	0

infarct	calcification	hemorrhage	thrombosis	type	anteriorchoroidal	posterior choroidal	dural	P1perforators	pericallosal
0	0	0	0	c	1	1	0	1	0
0	0	0	1,partial	m	0	1	0	0	1
0	0	1	0	c	1	1	0	1	0
0	0	0	0	m	1	1	1, occipital	0	0
0	0	0	0	m	0	1	0	1	0
0	1	0	0	m	0	0	0	1	0
0	0	0	0	m	0	1	0	1	0
1	0	0	0	m	1	1	0	1	0
0	0	0	0	c	1	1	0	1	0
0	0	0	0	c	1	1	0	1	0
0	0	0	0	c	0	1	0	0	1
0	0	0	0	c	1	1	0	1	0
0	0	0	0	m	0	1	0	0	1
0	0	0	0	c	0	1	0	0	0
0	0	0	0	m	1	1	0	0	1
0	0	0	0	c	0	1	0	1	0
0	1	0	0	m	0	0	0	1	0
0	0	0	0	m	0	0	0	1	0
0	0	0	0	c	0	1	0	1	0
0	0	0	0	c	0	1	0	0	0
0	0	0	0	m	0	1	0	0	0
0	0	0	0	m	0	1	0	0	0
0	0	0	0	c	0	1	1, occipital	0	0
0	1,wall	0	0	m	0	1	0	1	0

other perforator	persistent falcine sinus	atretic straight sinus	atretic transverse-sigmoid	occipital sinus
0	0	0	0	1
0	1	1	0	0
1	0	0	0	0
0	0	0	0	0
0	1	0	0	0
0	1	0	0	0
0	1	1	0	0
1	0	0	0	0
0	0	0	0	0
sca	0	1	1	1
0	1	1	0	1
0	1	1	0	1
0	1	1	0	1
0	0	1	0	1
0	1	1	0	0
0	0	1	0	1
0	0	1	1	1
0	0	0	0	0
0	0	0	0	0
0	0	0	0	0
0	0	0	0	0
0	0	0	0	0
sca,pica	0	0	0	0
0	0	1	1	1

other	associated anomaly	embolisation	age	route	material
marginal sinus	PAISA	1	4mth	c	b
limbic arch	0	1	6yr	a	g
0	0	1	28yr	c	c
epsilon vein	Rt arch, aberrant lt sca with kommorels	1	1yr 2mth	c	b
0	0	1	1yr	c	b
limbic arch	0	1	2 yr 6mont	a	b
0	0	1	2yr 6mth	a	g
0	0	1	12yr 2mth	a	g
duplicated straight sinus	0	1	8yr	c	b
sss atrophy	rt aortic arch,mirror image	1	9mth	c	b
interrupted IVC, marginal,duplicated left transverse	hypoplastic aortic arch, bovine lt CCA, direct VA origin	1	2mth	a	g
0	0	1	3yr	c	b
marginal sinus	0	1	4yr	a	g
epsilon vein	PAISA	1	6mnth	c	b
0	0	1	9mth	a	c
0	PAISA	1	6mnth	c	b
0	0	1	27yr	a	m-1983
0	0	1	1yr 10mth	a	g
0	0	1	11yr	a	g
0	0	1	4mth	a	g
0	0	1	10mth	a	g
0	0	1	9yr	a	g
0	0	1	5yr 8mth	a	g
0	0	1	11mnth	a	b

coil no/type	glue concentration	microcath	no of sitting
4/vog,gdc	95	spinneker	1
0	85	spinneker	1
14/gdc,vog		echelon	1
4/ vog,fibrecomplex	33,75	tracker18 perforation,spinneker	1
18/fibre,cook,bernstein	15	tracker18,magic	1
1/bernstein	90	tracker18	1
0	50,60,90	spinneker,magic	2
0	50,85	spinneker	1
5/	66,80,75,33,50,20,17,25	spinneker,magic,excelsior	6
2/gdc,berensteinliquid	98	echelon,spinneker	2
0	25,66	spinneker	1
2/gdc,fibre	na	tracker18	1
0	85	spinneker	1
12/	20,40,60	spinneker,magic	1
9/gdc,matrix	0	echelon	2
16/gdc	80	echelon	1
0	0	0	1
0	na	spinneker	1
0	na	spinneker	2
0	na	spinneker	2
0	na	spinneker	1
0	85	tracker18	1
0	95	spinneker	1
6/gdc3d, helix	20	echelon	1

complication	technical success
partial coil migration to straight sinus	nt
seizure, glue to SSS	t
sah(17% glueing of rent, drain)	nt
microcatheter perforation	t
IVH,SAH, EVD	t
pulmonary coil migration	nt
0	t
0	t
glue migration to venous side	nt
0	p
rt lower limb gangrene, cardiac arrest, ventricular fibrillation, renal dysfunction, lactic acidosis, consolidation	p
0	p
0	t
glue migration to venous side and left PCA branch, thalamic infarct, hydrocephalus	nt
0	nt
0	nt
ocular gaze paresis,improved	t
rt iliac artery spasm,conservative management successful	nt
0	t
infarct, left hemiparesis, resolved completely in 6days	nt
0	nt
0	t
0	i
0	nt

clinical follow up duration	imaging follow up duration	imaging followup result	clinical follow up result	surgery	shunt	ventriculostomy
6 yr 7 month	3 yr 8mnth	t	i	0	0	0
0	0	0	0	0	0	0
6days	3days	c	d	0	0	0
9yr 6month	4yr 10mth	t	i	0	0	0
2days	0	0	d	0	0	0
7yr4mth	7yr4mth	t	i	0	0	0
0	0	0	0	1	1	0
11mths	0	0	i	0	0	0
2yr9mth	2yr	nt	i	0	0	0
0	0	0	d	1	1	1
0	0	0	d	0	0	0
10yr8mth	1yr	nt	i	0	0	0
4mths	0	0	i	0	0	0
3yr3mth	0	0	i	0	0	0
2yr2mth	1yr	nt	i	0	0	0
10yr4mth	9.5yr	t	i	0	0	0
9yr 1mth	1yr 2mth	t(calcified)	i	0	0	0
8yr	0	0	i	0	0	0
6mth	6mth	t	i	0	0	0
6mth	0	0	I,shuntmeningitis,expired	0	1	0
4yr	0	0	i	0	0	0
11 yr 10months	2.5yr	t	i	0	0	0
3mth	0	0	i	0	0	0
15d	4d	t	i	0	0	0

surgical complication	surgical follow up	DSA	CT	MRI	TCD	LOST
0	0	1	1	0	1	0
0	0	1	1	0	0	0
0	0	1	1	1	0	0
0	0	1	1	0	1	0
0	0	1	1	0	0	0
0	0	1	1	1	0	0
0	0	1	0	0	0	0
0	0	1	1	1	0	0
0	0	1	1	0	1	0
0	0	1	1	1	0	0
0	0	1	0	1	0	0
0	0	1	1	0	0	0
0	0	1	0	1	0	0
0	0	1	0	1	0	0
0	0	1	1	1	1	0
0	0	1	1	0	0	0
0	0	1-conventional	1, preop only xray	0	0	0
0	0	1	1	0	0	0
0	0	1	1	0	0	0
meningitis	0	1	1	0	0	1
0	0	1	0	1	0	0
0	0	1	1	0	0	0
0	0	1	1	1	0	0
0	0	1	1	1	1	0

ANNEXURE5-KEY TO MASTER CHART

KEY TO MASTER CHART

SAH-subarachnoid hemorrhage
DSA-digital subtraction angiography
CT-Computed Tomography
MRI-Magnetic Resonance Imaging
yr- year
mth-month
sex column m-male
f-female
0-no
1-yes
h/o-history of
ASD- atrial septal defect
VSD- ventricular septal defect
PDA -patent ductas arteriosus
PFO -patent foramen ovale
PVH- upper pulmonary vein hypoplasia
PHT- pulmonary hypertension
MR-Mitral regurgitation
LVH-Left ventricular hypertrophy
Type column- m-mural
c-choroidal
sca-superior cerebellar artery
pica-posterior inferior cerebellar artery
SSS- superior sagittal sinus
IVC-inferior vena cava
Rt-right
CCA- common carotis artery
SCA-subclavian artery
VA-vertebral artery
PAISA-proatlantal intersegmental artery
LT-left
Route column- c-combined arteriovenous
a- arterial
Material column- c-coil
g-glue
b- both coil and glue
m- muscle
gdc- guglielmi detachable coil
vog-vein of galen coil
LFT-liver function test
IVH-intraventricular hemorrhage
EVD-endoscopic ventricular drainage
PCA-posterior cerebral artery