Vein of Galen Aneurysms

A vein of Galen aneurysmal malformation is a dilation of the vein of Galen that results from the presence of an arteriovenous fistula between choroidal branches of the CA and/or branches of the basilar artery and the dilated veins of the mesencephalon. Lasjaunias and colleagues emphasize an important distinction: that these lesions exist within the subarachnoid space, unlike true pial AVMs, which develop from the subpial space. This feature allows for high rates of blood flow through the fistula without risk of rupture and hemorrhage.

The evolution of contemporary concepts that guide the approach to vein of Galen malformations can be broadly divided into three distinct eras. The first includes the time period before the 1960s, and could be characterized as the historical era. Each of these periods is detailed later, but the historical era could be fairly summarized as beginning with the earliest pathological descriptions and recognition of the lesion and concluding with a series of individual case reports that dominated the literature in the middle to late 1960s. Clinical syndromes were recognized and an early classification system was developed.

The second era, which we will define as the early era of treatment, was initiated with the emergence of a number of series from large pediatric neurosurgery centers. Clinical syndromes were correlated with outcome. The primary therapy during this period was surgery, and morbidity and mortality rates remained high (particularly for lesions in newborns with high-output cardiac failure). Broadly, this era occurred from the late 1960s until the early 1990s.

We will define the third as the era of neurointervention, and generally consider this to have lasted from the late 1980s and early 1990s until the current time. The hallmark of this era so far has been the critical observation that neurointerventional techniques in which occlusive materials are introduced into the malformations have demonstrated high rates of successful obliteration of the fistula with markedly reduced rates of mortality and morbidity. Advanced neuroangiographic capabilities have also allowed more detailed anatomical characterization of the lesions, which has prompted important developments in classification and insights into the pathophysiological features of these lesions. The latter part of this era and the most recent substantive papers on the treatment of vein of Galen aneurysmal malformations have reflected improved neurointerventional methodologies and techniques, or incorporation of multidisciplinary approaches in which surgical techniques were used in conjunction with neurointerventional ones.

Historical Era

As quoted by Hoffman, et al., and Johnston, et al., the first description of a vein of Galen aneurysm was probably published by Steinhilb in the German pathology liter-
ature in 1895. Steinhill reported the postmortem findings in a 49-year-old man with a vein of Galen aneurysmal malformation and a frontal AVM, and labeled the condition a varix aneurysm. The majority of the earliest papers were postmortem descriptions of the pathological anatomy associated with vein of Galen aneurysmal malformations. The first description of any treatment was that of Balance in 1905, as quoted by Pool and Potts in their 1965 text. Balance’s patient, an 11-year-old girl, like most others prior to the late twentieth century, underwent CA ligation as treatment for a vein of Galen aneurysmal malformation. A more complete description was provided in 1937 by Jaeger, et al., who also used CA ligation. The 4-year-old child in their report presented with hydrocephalus and was found to have a massively dilated vein of Galen.

Oschewitz and Davidoff were the first to perform an open operation for a vein of Galen aneurysmal malformation, and they reported their experience in 1947. The surgery consisted of intraoperative inspection of the aneurysm, needle puncture, and then control of the local hemorrhage from the needle site. Feeding vessels to a vein of Galen aneurysmal malformation were first clipped by Boldrey and Miller in 1949 after CA ligation had failed to resolve the preoperative hemorrhage in a patient with this lesion. French and Peyton described five patients with AVMs in the region of the great vein of Galen in 1954. In their discussion they comment that surgical treatment is unfavorable but appropriate if the arterial supply can be discerned angiographically and the lesion is symptomatic or increasing in size. By the late 1940s and early 1950s, advances in cerebral angiography had facilitated several important descriptive papers that served to increase awareness of the condition of vein of Galen aneurysmal malformation, which is reflected in a significant number of individual case reports in the 1950s and 1960s.

Silverman and colleagues communicated important observations about the pathophysiological features of the fistula in a vein of Galen aneurysmal malformation and its causative role in prompting high-output cardiac failure in neonates and infants. Before birth, the low-resistance placental circulation absorbs as much as 40% of cardiac output, so that the right ventricle supplies the placenta and lower body while the left ventricle supplies the head. In the case of a vein of Galen aneurysmal malformation, the fistula is supplied by the left ventricle while fetal circulation persists. Once the child is born and the low-resistance bed of the placenta is eliminated, the flow through the fistula abruptly increases. Each ventricle must pump blood individually and in series, and therefore the demand on each ventricle is increased. This is compounded by the opening of the lungs and transmission of blood through the pulmonary capillary beds. Excessive blood flow in the pulmonary circuit contributes to pulmonary hypertension. In turn, the increased venous return to the right side of the heart can increase right-to-left shunting of blood if there is a patent foramen ovale. Cardiac perfusion is dependent on diastolic time and pressure. The combined factors of reduced diastolic time (due to the increased rate needed to support cardiac output) and increased intracardiac pressure from the right-to-left shunting can significantly impair coronary artery perfusion and contribute to myocardial ischemia. The combination of myocardial ischemia and cerebral ischemia, which arises due to shunting of blood through the fistula at the expense of perfusion of normal cerebral capillary beds, is thought to represent the central mechanism by which vein of Galen aneurysmal malformations work their devastation in infants and neonates.

Johnston, et al., observed that there were 30 cases of vein of Galen aneurysmal malformations reported in 22 papers between 1940 and 1960. Many of these were individual case reports that documented associated pathological conditions or unusual presentations or findings in adults with these lesions. Several central observations emerged. In a seminal paper in 1964, Gold, et al., established the clinical features of a vein of Galen malformation and defined three characteristic clinical syndromes associated with these lesions. Neonates were found to present in extremis in severe congestive heart failure. None survived at that time and all were found to have signs of severe cardiovascular decompensation at postmortem analysis. Infants characteristically presented with seizures or hydrocephalus. Distention of the scalp veins, particularly over the glabella, was another common finding in infants. An important diagnostic sign in this age group was the cranial bruit that was present in 10 of the 19 children in this cohort. A loud, harsh, pansystolic bruit is described. Older children and young adults were found to suffer from headaches or signs and symptoms of subarachnoid hemorrhage.

In 1973 Amacher and Shillito reviewed five cases from the Children’s Hospital Boston pediatric neurosurgery service, in addition to 37 cases that they found in the literature. They defined a fourth group as well, of patients who presented later in life with headache, exercise syncope, or subarachnoid hemorrhage. Syndromes are essentially the same as those defined by Gold, et al., in their earlier manuscript, except that Amacher and Shillito noted early mild heart failure that was medically controlled and presaged hydrocephalus as a presenting symptom. These authors advocated a right parietooccipital craniotomy with a parasagittal corridor of approach to isolate and divide feeding vessels. They advised against attempts to remove the sac and advocated instead that its brisk arterial inflow be divided.

Long, et al., outlined insights gleaned from 12 patients who underwent surgery for vein of Galen malformations at the University of Minnesota in 1974. These authors emphasized the critical importance of management of the cardiac status of these small infants. Babies with vein of Galen malformations have an enlarged blood volume, and their compromised heart is incapable of managing the greater volume against a suddenly (surgically) increased resistance. Therefore, the authors embraced measured reductions in blood volume and gradual obliteration of the malformation to reduce cardiac complications. The gradual occlusion was accomplished by ligating feeding vessels individually and slowly, and allowing time for the heart to equilibrate between ligations. Temporary clips were liberally used if there was uncertainty whether an individual branch should be immediately sectioned.

Another important contribution during this era was the paper from Norman and Becker, who evaluated seven newborns who had died of congestive heart failure arising from a fistula associated with a vein of Galen malformation. Postmortem analysis demonstrated severe cerebral gliotic infarction “which clearly antedated birth by sever-
al weeks.” The authors concluded that neonates presenting with a high-flow lesion had probably already sustained significant cerebral damage because of steal and blood shunting from dilated leptomeningeal vessels.

### Early Treatment Period

By the late 1970s to early 1980s, there was widespread recognition of the clinical entity of vein of Galen malformations. Using cerebral angiography, physicians could readily identify the lesions, and the newly emerging computerized tomography scanning technology enabled noninvasive visualization of the malformation. The primary treatment was open microsurgery, and a number of reports addressed technical points in the open surgical treatment of these lesions. As these lesions became better recognized, referral centers began to accumulate experience and single-institution series began to appear.7

Yaşargil, et al.,39 reported experience with nine cases from Zürich and emphasized the utility of a posterior interhemispheric approach performed with the patient in the sitting position along with comprehensive study of the preoperative angiogram and meticulous microsurgical technique. These authors further proposed that the vessels feeding the lesion be taken in order, starting with the distal pericallosal arteries, then proceeding sequentially to the supply from the cisternal vessels of the superior cerebellar and posterior cerebral arteries. Finally, the perforating vessels from the posterior thalamus should be taken. Again, the aneurysmal dilation is not resected.

Menezès, et al.,23 emphasized the importance of interrupting the fistula and leaving the aneurysm mass or sac in place as an important modification in reducing the considerable morbidity and risk of death that aneurysm excision entailed.

The largest and most influential single-center report during this era was published by Hoffman, et al.,11 in 1982. In addition to summarizing experience recorded in the international literature (128 cases to date), these authors reported on 29 patients with vein of Galen malformations treated at the Hospital for Sick Children in Toronto between 1950 and 1980. Important conclusions that the authors drew from published international experience included the following points: 1) patients who harbor vein of Galen malformations and who remain untreated do poorly; 2) treated patients have higher survival rates than untreated ones (despite some very primitive means of treatment reported in the literature); and 3) the best surgical results occur in older children and adults. Like other series, there was a preponderance of male patients (2:1 ratio) in the report from Toronto. Sixteen neonates were evaluated and all did poorly. Fourteen of the 16 presented with congestive heart failure. Seven underwent surgical treatment and only one survived (with dense hemiparesis).

The malformations were typically fed by numerous vessels that most often entered the aneurysm at its anterolateral fistula than open surgery. The deep central fistulous communications that define vein of Galen aneurysms make them less amenable to excision than endovascular obliteration.

### New Treatment Paradigms

Advances in neurorinterventional techniques that occurred in the late twentieth century revolutionized the diagnosis, treatment, and understanding of vein of Galen malformations. Progressive intravascular occlusion of the lesion with particles or balloons was rapidly shown to be a far safer and more effective method of eliminating the fistula than open surgery. The deep central fistulous communications that define vein of Galen aneurysms make them less amenable to excision than endovascular obliteration.
ation. This finding facilitated aggressive, early treatment, which has become recognized as the clinical cornerstone to good outcomes in these challenging lesions. This evolution in treatment was succinctly summarized in the report by Ciricillo and colleagues. They contrast their initial surgical experience, in which five infants with a vein of Galen aneurysmal malformation treated before 1983 underwent direct surgical treatment, with subsequent cases in which eight infants were treated using endovascular techniques. All patients treated with direct surgery died, and six of the eight treated with endovascular occlusion survived.

Selective angiography facilitated detailed examination of the anatomy of vein of Galen aneurysms and promoted the development of angiographic classification systems. Just as the clinical classification systems reflected and explained clinical presentation, the angiographic classification systems are useful in therapeutic decision making. The two most widely referenced angiographic classifications are those of Lasjaunias and colleagues and Yaşargil. Lasjaunias defined two major angiographic forms. The first is the true vein of Galen malformation. These lesions occur because of a dysembryogenetic event involving the median vein of the prosencephalon. As a result, a fistulous connection develops between arteries and veins in the wall of the dilated vein of Galen. A fistula is in the wall of the vein (median vein of the prosencephalon) or the choroidal type, in which the drainage is into tributary veins of the medial vein of the prosencephalon. Typically, mural-type true vein of Galen aneurysmal malformations have smaller numbers of fistulas than choroidal types, and they present in infancy. Lasjaunias and colleagues advocate transarterial obliteration with coils and acrylic glues for mural malformations of the vein of Galen. These lesions can be treated later in life than choroidal types because the number of fistulas is smaller and the resulting flow is less. Choroidal artery–vein of Galen malformations typically present in neonates with high-flow fistulas and cardiac compromise from congestive heart failure. Staged endovascular obliteration is advocated for these lesions. The first intervention obviously attempts to decrease the massive flow across the multiple fistulas as much as possible. Later stages involve serial obliteration of additional fistulas to achieve complete closure of the malformation.

The secondary types of vein of Galen aneurysms were termed vein of Galen dilations by Lasjaunias and colleagues. This reflected and extended the earlier observations (in 1960) of Litvak et al., who discerned the difference between primary fistulas involving the great vein of Galen and adjacent AVMs that drain through the vein of Galen. These lesions are characterized by a deep midline true AVM that drains selectively into the great vein of Galen and thereby causes distention and enlargement of the vein of Galen. These lesions are characteristically filled by branches of the middle cerebral artery (thalamoperforating, lenticulostriate, or transsylvian branches), whereas the choroidal and mural true vein of Galen malformations are typically filled by choroidal and pericallosal vessels.

 Yaşargil defined four types of lesions: Type 1 is a simple small fistula involving branches from the pericallosal or posterior cerebral arteries; Type 2 involves more feeding vessels from middle cerebral artery branches (thalamoperforating vessels); Type 3 involves high-flow lesions with large numbers of fistulous connections from a wide range of feeding vessels; and Type 4 is a midline AVM with drainage into the vein of Galen (analogous to Lasjaunias’ vein of Galen dilations). Mickle and others have also proposed an angiographic classification system, but this one differs in that it centers on the patterns of venous drainage rather than the source, number, and site of fistulous connections as in the other grading systems.

Mickle and Quisling described a transfemoral approach for embolization of vein of Galen malformations in 1986. The procedure involves a small craniotomy made directly over the torcular herophili and subsequent puncture of this structure with an angiocatheter. A guidewire and angiography catheter are introduced, through which embolizing coils are placed into the malformation. A reduction of flow by one half is the authors’ recommended end point in depositing embolizing coils. An associated plunger device was simultaneously developed to assist in securing any coil material that became dislodged in an undesired location. In the original manuscript, three patients were initially described in whom successful outcomes were achieved using these methods, but other groups have reported similarly good outcomes after using these techniques.

Due to the rarity of these lesions, there are few large published series of patients treated with endovascular approaches for vein of Galen aneurysmal malformation. In 1993 Lylyk et al., reported on a group of 28 children who were treated for vein of Galen aneurysms at the University of California Los Angeles between 1986 and 1990. Fifteen of these patients had severe congestive heart failure. Eleven children underwent a combined transarterial and transvenous procedure, whereas eight underwent a pure transfemoral approach. Immediate postprocedure improvement was noted in 23 patients and complete occlusion of the fistula was noted in 13. Overall, 60% of patients demonstrated a good or excellent clinical outcome.

King and colleagues reported on two infants and a 10-year-old child treated with transfemoral embolization. All did well and were cured of their preoperative symptoms.

In 1989, Lasjaunias and colleagues reported a large series of 36 cases of vein of Galen malformations treated using endovascular procedures. Twenty-eight children were included, and all obliteration was completed via a transfemoral route. In the short follow-up period, these investigators demonstrated a mortality rate of 13% with no technical morbidity, a 27% rate of complete occlusion, and a 53% rate of significant clinical improvement. Further follow-up data were included in a large cumulative report of 179 cerebral AVMs published by this group in 1995. The combining of vein of Galen malformations with pial AVMs reflects this group’s contention that angiographic classification is most important in determining treatment and overall bias toward neurointerventional techniques for all AVMs (see Part III of this review for further discussion). Seventy-seven patients with vein of Galen malformations underwent evaluation and treatment, and more than half attained complete obliteration without neurological insult. Thirteen of 18 untreated patients died.
Conclusions

Rarely has a lesion with such an ominous prognosis as the untreated vein of Galen malformation undergone such a radical metamorphosis in prognosis in such a short time. Most children harboring these lesions now have a good to excellent prognosis, provided that experienced neurointerventional care can be promptly obtained. Neonates with severe congestive heart failure still have elevated mortality rates that may approach 40 to 50%. Gradual embolization with particulate materials via transarterial or transchoroidal approaches is the preferred treatment, and although direct surgical attack is occasionally necessary to supplement endovascular occlusion, treatment has evolved so that this earlier approach is of mostly historical significance.

References


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