

Vein of Galen Arteriovenous Malformation in a Neonate

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Abstract

The vein of Galen is the most common type of arteriovenous malformation in the fetus and neonate. Most vein of Galen arteriovenous malformations (VGAMs) are diagnosed in the neonatal period, with the remainder being identified in early childhood, typically via computed tomography scan. The VGAM is found in five different patterns where the vein of Galen and straight sinus extending to the torcula Herophili are dilated. This dilation can lead to significant compression of the posterior fossa structures. Clinically, the infant with this malformation can present with seizures or most commonly, high output cardiac failure. It is important, however, to keep a broad differential diagnosis as more prevalent neonatal conditions arise similarly. These conditions can include developmental delay, cerebral palsy, epilepsy, superior vena cava syndrome, hemangioendothelioma, and other arteriovenous fistulae. Treatment begins with early diagnosis and testing of initial sequelae. This is often accomplished in consultation with different pediatric subspecialists, particularly neurologists and cardiologists. The mainstay of therapy is with neurosurgical intervention. Although the mortality of a fetus or neonate with VGAM is very high, prognosis is dependent on the size of the malformation, age at diagnosis, and successful neurosurgical outcome. [*Pediatr Ann.* 2015;44(10):e243-e246.]

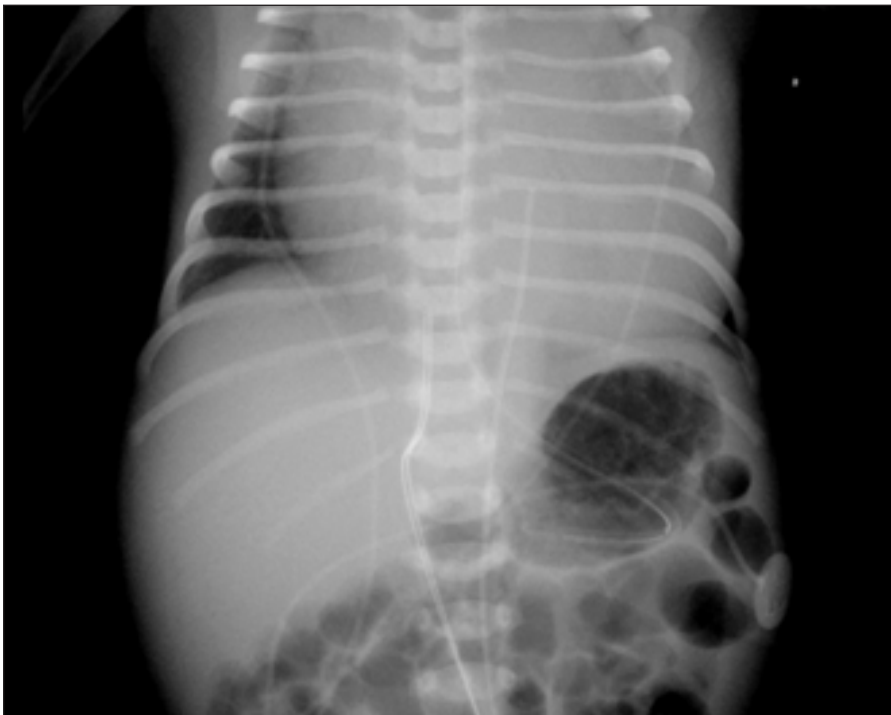


Figure 1. Cardiomegaly with cardiac apex left-sided. Note the minimal atelectasis of the left lower lobe and disorganized bowel gas pattern.

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A 33-year-old mother who had received adequate prenatal care with an uncomplicated pregnancy delivered an infant boy via emergency cesarean delivery weighing 3,240 g. Both the mother and father were carriers for spinal muscular atrophy, and chorionic villous sampling was conducted early in the pregnancy that demonstrated minimal risk to the infant. Maternal serologies were negative, but she was found to be group B *Streptococcus* positive for which she received one dose of intrapartum antibiotics.

Several hours after the mother was admitted for labor induction, there was a notable decrease in fetal movement, decreased variability in fetal heart rate, and late decelerations. As a result, an emergency Cesarean delivery was performed for fetal distress. Artificial rupture of membranes showed clear fluid and the procedure was vacuum assisted. Upon delivery, the infant was placed on a radiant warmer and was noted to be limp, apneic, and cyanotic. Heart rate at the time was <100 beats per minute, and positive pressure ventilation was then started; however, the infant remained apneic. A decision was then made to intubate and oxygen saturations improved from 40% to 85%. APGAR (Activity, Pulse, Grimace, Appearance, Respiration) scores were 1, 5, and 5 at 1, 5, and 10 minutes, respectively. Initial physical examination was notable for bilateral neck swelling with a varicosity of the left carotid area, a fontanel with a bruit, and an active precordium. He was admitted to the referring hospital's neonatal intensive care unit (NICU) for further evaluation and management.

NICU COURSE AND DIAGNOSIS

Upon admission to the referring NICU, passive cooling was initiated. Umbilical lines were placed and ini-

tial tests were conducted. Glucose was noted to be 11 mg/dL, and 10 mL of 10% dextrose in water (D10W) was administered. Repeat glucose was 30 mg/dL and another 10 mL of D10W was given. Complete blood count was notable for a platelet count

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of 16,000 for which 20 mL/kg of platelets were given along with fresh frozen plasma. A stat head ultrasound (HUS) showed a large cerebral arterial venous malformation with ventriculomegaly and cerebral atrophy affecting the left hemisphere more than the right. A chest X-ray (CXR) was also performed, which showed an enlarged heart. The decision was made to transfer the infant to our NICU for further management and neurosurgery consultation. Just prior to the transfer, the infant was noted to have no activity, no spontaneous respirations, was hypotonic, and had an intermittent suck on the endotracheal tube.

On arrival, the infant was placed on a radiant warmer. Vital signs at this time showed a heart rate of 80 to 90 beats per minute, and oxygen saturations of 70% to 80%. Cooling was discontinued at this time and dopamine was started, as mean arterial pressures were noted to be <30 mm Hg. The patient was in cardiac failure, maintained by vasopressors and a CXR confirmed the findings of cardiomegaly (**Figure 1**). Computed tomography (CT) scan of the head showed arteriovenous malformations with evident

engorgement and dilation of both superficial and deep veins. The vein of Galen and straight sinus extending to the torcula Herophili were extremely dilated leading to significant compression of the posterior fossa structures (**Figure 2**). In addition to the vein of Galen findings, there were multiple calcifications throughout the supratentorial parenchyma bilaterally and diffuse edema across the bilateral cerebral hemispheres.

Using the Bicêtre Neonatal Evaluation Score for embolization,¹ he was noted to have a score of 2. Scores from 8 to 12 are typically needed for embolization to have potential success in vein of Galen malformations. With the attending neonatologist, the patient's prognosis was discussed with the family and the decision was made to transport the infant back to the referring hospital to be with his mother, at which time life support would be withdrawn.

DISCUSSION

Development and Classification

Vein of Galen arteriovenous malformations (VGAM) are the most common arteriovenous malformation in fetuses and infants.² Functionally, the vein of Galen drains the blood from the anterior and central regions of the brain into the sinuses of the posterior cerebral fossa. The vein of Galen drains into the confluence of the inferior sagittal sinus and the anterior extremity of the straight sinus. This is the area located just under the cerebral hemispheres. The aneurysmal dilatations begin during embryologic formation from weeks 6 to 11 of gestation. At this time, fistulae form between the primitive pia arachnoidal arteries and veins that cross one another before draining into the vein of Galen. The most likely persistent embryonic vascular channel contributing to the VGAM is the median prosence-

phalic vein. Formation of these fistulae persist in various patterns and can include one or many vessels in the aneurysmal malformation.¹ With age, the malformation will continue to increase in size. There are five different patterns of malformation that have been observed (Table 1).

Physical Examination and Differential Diagnosis

VGAM is most commonly associated with high-output cardiac failure. This failure is the result of decreased resistance and high blood flow into the VGAM from the arachnoidal vessels. These changes in flow and shunting of blood can cause sequelae, such as a steal phenomenon. At times, the arteriovenous shunt can steal up to 80% or more of the total cardiac output, causing heart failure.³ As blood is backed up in the venous system, it can lead to right heart and pulmonary artery dilation. Furthermore, with shunting, certain areas of the brain have decreased flow. Patients can present with progressive hemiparesis or less commonly ischemic strokes or hemorrhage from eventual rupture of vessels. In a few patients, it has been noted that the VGAM can be so large, it causes a mass effect and focal neurologic results, as was the case with the patient illustrated in this article.

Depending on the primary presentation, patients with VGAM can have features similar to other, often more common conditions. It is important to maintain a broad differential diagnosis. In the neonatal period these include: seizures, cranial bruits, cavernous sinus syndromes, central vein thrombosis, hydrocephalus, or intracranial hemorrhage. In older children with a later presentation, it is important to add: developmental delay, cerebral palsy, epilepsy, superior vena cava syndrome, and pseudotumor cerebri.⁴

DIAGNOSIS

There are many diagnostic modalities to help with the identification and characterization of this condition. Initial imaging to consider is a cranial ultrasound (US). The cranial US will help to identify and localize a lesion. Hemodynamics of the VGAM can be further understood with the Doppler functionality.

The gold standard for diagnostic imaging of a VGAM is with a cranial magnetic resonance imaging or CT—with and without contrast. These studies will help to confirm the diagnosis as well as establish the degree of involvement with the surrounding parenchyma. In particular, these studies help to determine if the patient is a surgical candidate. Other imaging studies to consider are chest radiograph followed by cardiac echocardiography to analyze the grade of involvement.

Medical Management

The care of an infant with a VGAM can be divided into both medical and surgical aspects. From a medical standpoint, management is based largely on control of sequelae. The cardiac management of the high-output heart failure is essential and often accomplished with consultation with a pediatric cardiologist.

If the patient is having seizures, they should be managed with antiepileptic medications, most commonly phenobarbital or phenytoin. Regular visits with a pediatric neurologist are also recommended. Lastly, it is important to assess for hydrocephalus by regularly measuring head circumference and noting delays in development during well-child examinations.⁵

Surgical Management

When surgery is an option, it is the mainstay of therapy for a VGAM. Initially, neurosurgical procedures are

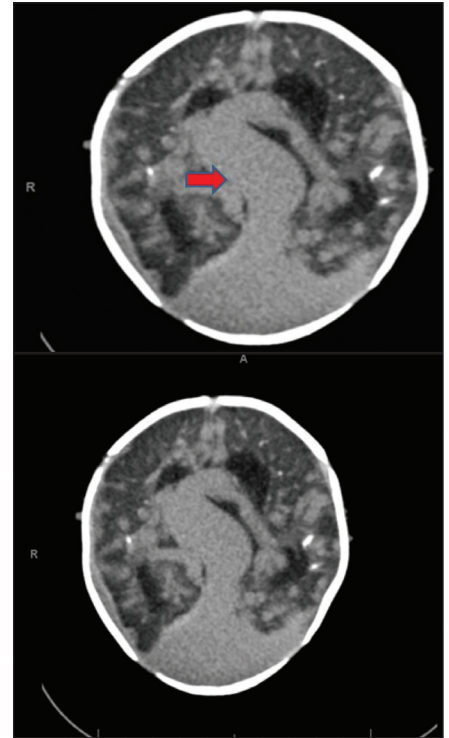


Figure 2. Arteriovenous malformation with engorgement and dilation of both the superficial and deep veins (red arrow). The vein of Galen and straight sinus extending to the torcula Herophilli are extremely dilated. Multiple calcifications throughout the bilateral supratentorial parenchyma and diffusely edematous bilateral hemispheres are also noted.

most valuable in relieving the pressure of hydrocephalus. This is typically accomplished using a ventriculoperitoneal shunt. In most cases of VGAM, the contributing fistulae must be ablated, thus reducing flow into the already dilated vein of Galen. Open surgery carries a high mortality rate and, until the past decade or so, was the only means of definitive treatment.

Emerging therapies include radiotherapy, which uses focused beams aimed to damage the contributing vessels. Unfortunately, not only does this modality take several months to years to complete, the beams can also damage the surrounding brain tissue.

Lastly, vaso-occlusive therapy including catheterization and embolization is currently the standard of care. This embolization is accomplished through the use of coils placed in the

TABLE 1.

Vein of Galen Arteriovenous Malformation Patterns

Pattern 1	Many vessels, including anterior cerebral arteries, thalamic perforating arteries, and superior cerebellar arteries, discharge into the vein of Galen
Pattern 2	A single posterior choroidal artery drains into the vein of Galen
Pattern 3	One or both posterior choroidal and one or both anterior cerebral arteries drain directly into the Galenic system
Pattern 4	An angiomatous network of posterior choroidal and thalamic perforating arteries enter the vein of Galen directly
Pattern 5	A high-flow arteriovenous malformation in the right inferior frontal lobe drains via the inferior sagittal sinus and pericallosal vein into the vein of Galen

Adapted from Nicholson et al.³

contributing vessels during the intravascular catheterization. As with any interventional procedure, there is significant risk to be considered—the most frequent adverse event is hemorrhage from coil misplacement or rupture of a vessel. Optimal age for intervention by any modality has been noted to be between ages 4 and 5 months.⁶

EPIDEMIOLOGY AND PROGNOSIS

Most VGAMs are diagnosed in the neonatal period, with the remainder being identified in early childhood. This condition does not have a racial

predilection and cases have been distributed equally among the male and female genders. Neonates with poor outcomes were those who have neurologic manifestations at birth, focal cerebral parenchymal changes, brainstem or tonsillar herniation, calcification of cerebral tissue, or more than one affected vessel leading into the malformation.

Mortality is very high. Over 50% of patients with a VGAM have a malformation that cannot be corrected and in fact 77% of untreated cases result in death. Despite surgical treatment, the mortality rate still approaches 40%.⁷

CONCLUSION

VGAM, although not a common occurrence, can have many different clinical presentations that vary with severity of the malformation. Early diagnosis and treatment with neurosurgical intervention are the mainstays of therapy. The prognosis of this condition is greatly dependent on the size of the malformation, age at diagnosis, and success of neurosurgical outcome.

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