

Successful Embolization of a Large Vein of Galen Malformation in a Premature Infant Presenting with Congestive Heart Failure and Persistent Pulmonary Hypertension

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Abstract

Keywords

- ▶ vein of Galen malformation
- ▶ high-output heart failure
- ▶ endovascular embolization

We report a case of a premature infant with a vein of Galen malformation (VGM) who presented with high-output heart failure, pulmonary hypertension, and respiratory distress. An echocardiogram showed normal cardiac anatomy, patent ductus arteriosus, and retrograde flow in the descending aorta. Ultrasonography and computed tomography of head confirmed the diagnosis of a large VGM. The patient developed multi-organ failure. Endovascular embolization of the VGM was successfully performed with an excellent outcome.

Vein of Galen malformation (VGM) was first described in 1895 as an abnormal fistula between the cerebral arteries and the great vein of Galen.^{1,2} Neonates with VGM usually present with cardiac failure and cyanosis, and can be mistaken as having cyanotic congenital heart diseases. Without intervention the mortality rate among infants with VGM is close to 100%.³ Endovascular embolization revolutionized the management of VGM and significantly reduced its morbidity and mortality.

Case Report

The patient was a 3280-gram newborn female born at 35 weeks gestation. Apgar scores were 7 and 9 at 1 and 5 minutes, respectively. The patient developed early respiratory distress, mild cyanosis (O₂ saturation 88%), and required endotracheal intubation. Initial cardiac examination showed hyperdynamic precordium and a faint systolic murmur. Chest X-ray showed cardiomegaly and diffuse opacification of both lungs. Surfactant and nitric oxide were administered with the

assumption that the patient had respiratory distress syndrome and persistent pulmonary hypertension of the newborn.

The patient developed increased tachycardia (heart rate of 195/minutes), metabolic acidosis, and decreased urine output. An audible bruit was heard upon head auscultation. An echocardiogram showed normal cardiac anatomy, hyperdynamic function (systolic function [SF] 55%), and a large patent ductus arteriosus (PDA) with a bidirectional shunt flow. There was retrograde flow in the distal aortic arch to the dilated neck vessels (▶**Fig. 1**) and pulmonary hypertension (65 mm Hg). Ultrasonography and computed tomography of head confirmed the presence of a large VGM (2.81 × 1.65 cm) with high velocity turbulent flow.

The renal and cardiac function deteriorated (SF 25%), and a strain pattern was noted in electrocardiogram (EKG). On the third day of life, angiography revealed a large VGM (▶**Fig. 2A**). Transarterial embolization was performed using Guglielmi detachable coils, resulting in a substantial decrease in VGM flow.

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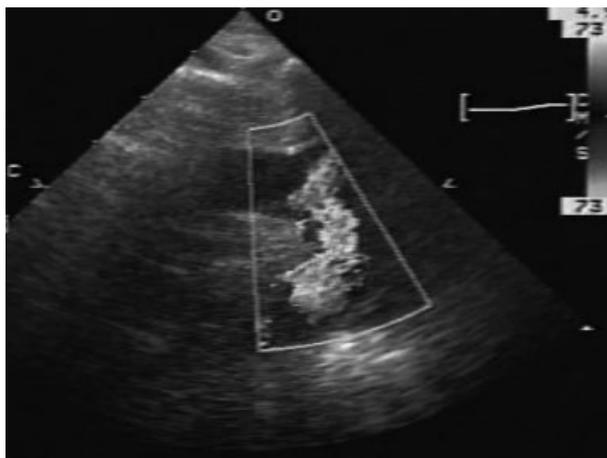


Figure 1 Doppler of the aortic arch showing retrograde flow in the descending aorta.

Initial clinical improvement was achieved, but the renal function continued to decline. On the ninth day of life, the VGM was reassessed and transvenous embolization was performed. Postembolization angiogram showed a significant decrease in shunting through the VGM and improved cortical artery flow (► **Fig. 2B**).

The hospital course which lasted 2.5 months was complicated by preprocedural clinical sepsis, renal failure, and persistent PDA requiring surgical ligation, and postprocedural seizures, chylothorax, pericardial effusion, and osteomyelitis of the right femur. Serial electroencephalograms (EEGs) showed improvement and the last EEG before discharge had no seizure activity. The chylothorax was treated with chest tube drainage, Portagen formula (Mead Johnson Nutrition, Glenview, IL), and octreotide.⁴

The patient did remarkably well despite of all these complications. At 26 months, she was seizure free, had a

normal physical examination and age appropriate milestones. An echocardiogram showed normal cardiac anatomy and function.

Discussion

VGM is a rare anomaly of the cerebral vasculature that results in significant aneurysmal dilation of the great vein of Galen. The clinical presentation of VGM varies with age. Neonates usually have multiple fistulae and present in high-output cardiac failure.⁵ Infants and younger children have a single fistula and present with seizures, hydrocephalus, distention of scalp veins, and failure to thrive. Older children and adults often present with neurological symptoms of headache and seizure.⁶ Due to collateral circulation, there is an increased blood flow in the pial veins leading to subarachnoid and intracranial hemorrhage.⁷

High-output cardiac failure in neonates with VGM is multifactorial in origin and responds poorly to medical management. During fetal life, the low resistance uteroplacental circulation diminishes the flow through the cerebral circulation. After birth, the placental circulation is lost and a significant portion of the cardiac output is directed toward the VGM. There is a compensatory increase in cardiac output and blood volume to maintain adequate systemic perfusion. This causes significant tachycardia, hyperdynamic cardiac function, and a wide pulse pressure. Since cardiac perfusion is dependent on the diastolic time and pressure, the cardiac function eventually deteriorates and ischemic changes can be noted in EKGs. The perfusion of all organs “including the brain,” is further compromised by stealing of blood from the systemic circulation by the low resistance VGM.

The management of VGM progressed through three historical eras. The first era involved the earliest descriptions

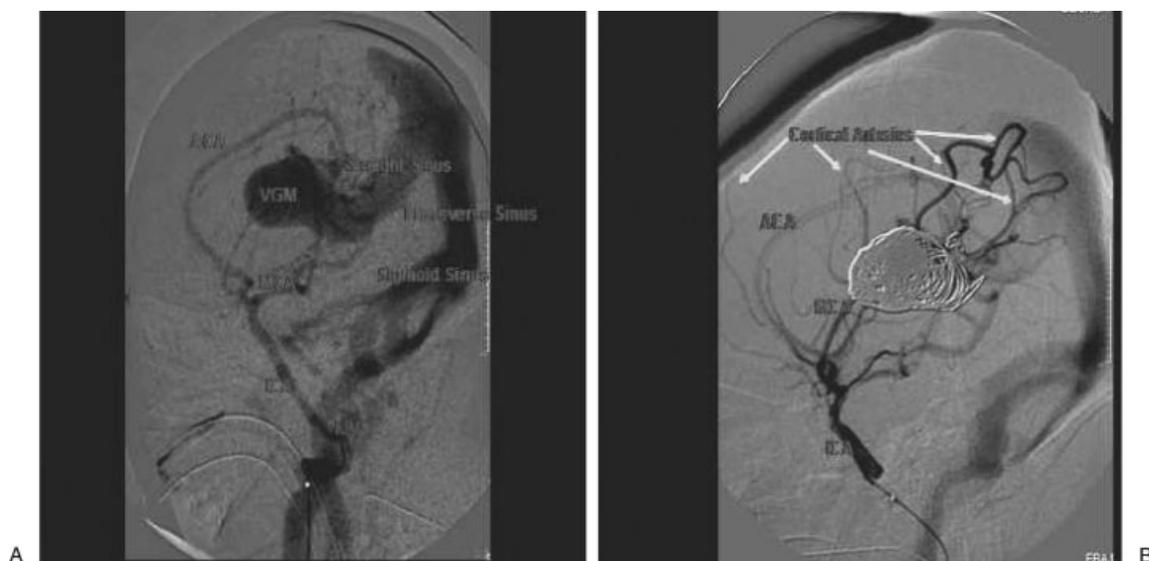


Figure 2 Angiograms of the vein of Galen malformation and the feeder arteries (A) pre-embolization and (B) postembolization, with improvement in filling of cortical arteries.

of pathology, anatomy, syndromes, and case reports. No interventions were offered to neonates with VGM and the mortality was close to 100%.³

During the second era, previously described syndromes were correlated with their outcomes. The main intervention was a combined open microsurgery through craniotomy and a direct transtrocular embolization, but mortality and morbidity remained as high as 91% regardless of surgery or medical management.⁸

The third and current phase is the neurointervention era. The development of intravascular occlusion devices has shown to be safer and more effective. A comparison study showed death of all six neonates with VGM who underwent direct microsurgery while six of the eight who were treated with endovascular embolization survived.⁹ In a case series of 36 VGMs treated with endovascular embolization, the mortality rate was 13%.¹⁰

Neonatal VGM remains a serious condition that requires early diagnosis and a multidisciplinary approach of management and intervention. Coordination among neonatologist, pediatric cardiologist, and interventional neuroradiologist is essential for better patient care and outcome. Prematurity and the presence of multiorgan failure should not be considered absolute contraindications for early intervention if the patient can be stabilized for the procedure.

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