A Neonate with a Vein of Galen Aneurysmal Malformation (VGAM) and Congestive Heart Failure Scheduled for Neuroembolization

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Learning Objectives:
Outline the diagnostic tools available to identify neonatal VGAM

Discuss the etiology of cardiac failure and pulmonary hypertension.

Formulate a rational evaluation of the patient's clinical status and understand the options for treating congestive heart failure and pulmonary hypertension.

Anticipate potential complications during intraoperative neuroembolism.

Stem Case and Key Questions:
Patient was a 37 week 3 kg product of a normal pregnancy and delivery. Two hours after birth he was noted to be tachypneic. Chest x-ray revealed cardiomegaly. Over the next 24 hours respiratory symptoms improved but radiographic evidence of cardiomegaly persisted. An echocardiogram demonstrated a dilated right ventricle with possible pulmonary hypertension. On day 3 of life, the patient was transferred to Children's Hospital.

What is the differential diagnosis?
How is the diagnosis confirmed?
What would be the indications for intubation, oxygenation and ventilation?

Uponn arrival at Children's Hospital, he exhibited signs and symptoms of respiratory distress and was intubated and appropriately oxygenated and ventilated. A chest x-ray revealed marked cardiomegaly with "hazy" lung fields. The diagnosis of cardiac failure was made and dopamine infusion was initiated at 5 mics/kg/minute. A cranial bruit was auscultated on the neonatal intensive care admission physical exam. An urgent echocardiogram, head ultrasound, and head CT scan were ordered.

The echocardiogram revealed normal segmental anatomy; marked right atrial and right ventricular enlargement with moderate to severe decreased right ventricular wall motion; moderate to severe tricuspid regurgitation with an estimated suprasystemic right ventricular pressure. Left ventricular function appeared normal, the ductus arteriosus was closed; the pulmonary veins to the left atrium were unobstructed; the foramen ovale was patent with moderate right-to-left shunt.

Head ultrasound demonstrated a Vein of Galen aneurysm with arteriovenous shunting.

CT scan of the brain revealed a marked enlargement of the vein of Galen measuring up to

2 cm in maximum thickness consistent with a vein of Galen malformation. There was marked enlargement of the straight, bilateral transverse and bilateral sigmoid sinuses due to the significant vascular shunting. The ventricles and sulci were normal in size for the patient·s age. There was no shift of the midline structures or extra-axial fluid collections or evidence of intracranial hemorrhage.

Neurosurgery consulted pediatric anesthesiology to determine the patient·s readiness for an intraoperative neuroembolism.

Describe the pathophysiology of this patient's congestive heart failure.

What inotropes could be used to treat this patient's high output cardiac failure? Are some inotropes a better choice than others?

Does pulmonary hypertension affect your decision to anesthetize this patient?

Would nitric oxide or prostaglandins E1 be useful for this patient's pulmonary hypertension?

The patient is stabilized and is brought to the OR for angiography and neuroembolization.

What are the approaches available to the neurosurgeon to achieve embolization?

The neurosurgeon decides to use the transtorcular approach?

What is this approach?

What are the advantages and disadvantages of this approach?

The baby underwent successful neuroembolism with a 140 cm 0.035 floppy coil. Three days later the infant returned to the operating room for further embolism because of continued signs and symptoms of high output cardiac failure. The patient underwent transarterial embolization with Onyx 34 embolic agent.

What precautions must be taken to avoid complications from embolism glue?
The patient's oxygen saturation and end-tidal CO_2 acutely decreases, what is your differential diagnosis?

The neuroembolism was successful with marked improvement of cardiac function. Echocardiogram performed at 5 weeks of age revealed a normal echocardiogram.

Model Discussion:
The vein of Galen aneurysmal malformation (VGAM) is a rare (1/25,000 deliveries) arteriovascular malformation that presents in the pediatric age group. VGAMs are congenital vascular malformations made from multiple arterial feeders establishing direct or indirect shunts with a large median venous collector. The latter is not the vein of Galen

but a persistent embryonic channel that normally participates in the formation of the vein of Galen. VGAMs classically present in the neonatal period with high-output cardiac failure and sometimes but not always with an audible cranial bruit. Severe pulmonary hypertension may also complicate the cardiac failure. In the past, neonates with VGAM presenting in cardiac failure had close to 100% mortality. Presently, endovascular treatment has emerged as the treatment of choice for patients with neonatal VGAM presenting in heart failure. Embolization, both of the feeding arteries and draining veins can result in reduction of blood flow and a greater survival rate in these infants. Neuroembolism will reduce and ultimately terminate excessive blood flow through the VGAM which is the key to improved cardiac function and brain injury prevention.

Transfontellar Doppler sonography is a noninvasive technique enabling bedside evaluation of the cerebrovascular system in neonates. Once the diagnosis is made, further studies such as computed tomography (CT), computed tomography angiography and magnetic resonance imaging and angiography may also be performed if the neonate is stable. A CT scan is usually performed to identify ventricular enlargement, brain atrophy and parenchymal calcifications associated with severe intracranial hypertension and brain injury. Babies with large VGAM shunts can have early ischemic brain injury. These children are not candidates for repair.

The etiology of cardiac failure may begin in utero. Several reports of antenatal echocardiograms reveal increased flow through the pulmonary artery and retrograde diastolic flow through the isthmus of the aorta. In one case report, infants with concommitant pulmonary hypertension showed signs of pulmonary hypertension on in utero echocardiography. Excessive pulmonary blood flow occurs because pulmonary flow far exceeds the dilatory capacity of the ductus arteriosus. Fetal imaging by ultrasound is now able to identify fetuses with VGAM allowing expectant delivery in a tertiary care center able to treat neonatal AVMs.

Cardiac high output failure is associated with VGAM because 60 to 80% of aortic blood flow is directed through the VGAM'S low resistant shunt. This increases blood flow via the superior vena cava to the right atrium and right ventricle. The neonate will respond to the increased flow by increasing pulmonary artery pressure. If the infant experiences hypoxia, hypercarbia and acidosis the pulmonary hypertension can lead to persistent pulmonary artery hypertension of the neonate. This scenario will cause right to left shunting both through the patent ductus arteriosus and foramen ovale. The right ventricle enlarges and loses compliance, worsening right heart failure. Cardiac interdependence will lead to left heart failure. Blood flow travels through the VGAM during diastole creating a “steal phenomena”. The reduction of diastolic pressure can lead to myocardial ischemia secondary to reduced coronary blood flow.

Improving cardiac function is imperative in order to avoid multiple organ failure. Cardiac failure is difficult to treat and most beta agonists along with diuretics have been used with varying success. Milrinone shows some promise because it is an inotrope and vasodilator. Although total systemic vascular resistance is reduced through the VGAM, the child in cardiac failure has an increased extracranial vascular resistance. Any intervention that

reduces extracranial systemic resistance is likely to improve systemic perfusion. Nitric oxide has been used with varying success to treat pulmonary hypertension in these patients. It does not appear to worsen cardiac failure but has rarely been documented to improve hemodynamics.

Cranial arteriovenous malformations are often initially confused clinically with congenital heart disease. Congestive heart failure with as much as 80% of cardiac output perfusing the brain results in increased precordial activity and diminished lower extremity pulses. Thus, aortic stenosis is a common misdiagnosis. Obviously an echocardiogram will rule out this congenital cardiac lesion.

Prostaglandins E1 was serendipitously found to lower right ventricular pressure and pulmonary artery pressure in babies with VGAM. Since patent ductus arteriosus dependent congenital heart diseases are included in the differential diagnosis for acute neonatal heart failure, VGAM patients are placed on prophylactic prostaglandins E1 before definitive diagnosis. This decision is made via protocols for treatment of acute neonatal cardiac failure regardless of etiology. In at least three VGAM patients described in the literature, neonates with a VGAM placed on protocol prostaglandins E1 infusion for acute neonatal heart failure demonstrated improvement.

Historically, babies with suprasystemic pulmonary hypertension have a poor prognosis. The mortality rate for all neonates in heart failure undergoing transcatheter embolization can be up to 50%, mortality is much higher when pulmonary hypertension is present. Babies with suprasystemic pulmonary artery pressure at birth may represent a severe form of VGAM with a higher degree of shunt that overloads the pulmonary vascular system in utero. Autopsy findings on two young babies with VGAM and pulmonary hypertension who expired despite aggressive treatment of heart failure and early neuroembolism of the VGAM are described by Dahdah et al. They found that both neonates had severe pulmonary hypertensive disease with medial muscle comprising 50% of the external diameter of small muscular pulmonary arteries, far in excess of the 5 to 15% that is expected in the normal newborn infants. In both these infants the ductus arteriosus was patent.

Transcatheter embolism is the procedure of choice to occlude the VGAM's shunt. It is less invasive and has a higher survival rate than open neurosurgical procedures. The advantages of transcatheter embolization include hemodynamic stability with minimal pain. Heart failure is an indication for urgent transcatheter embolization. The goal is to achieve partial embolization with the endpoint of improving heart failure. Thus, the embolization may be staged. Each session is limited by the volume of contrast media delivered and the patient's tolerance of the procedure. The ultimate goal of these sessions is to completely occlude the VGAM while avoiding neurological and cardiac injury. Since 2000, the mortality and morbidity rates of neonates in cardiac failure without suprasystemic pulmonary hypertension have been reduced to between10 and 15%.

The goals of anesthesia are to maintain cardiac stability and provide a quiet field for successful VGAM embolization. Opioid based anesthesia preserves cardiac function, and

is a good technique for this procedure. In addition, the liberal use of oxygen, hyperventilation, and nitric oxide have been advocated to control pulmonary hypertension. Patients are usually maintained on the ventilator postoperatively so that the patient can remain sedated, and cardiac and respiratory stability maintained. In addition, hypertension needs to be avoided in the first 48 hours after the procedure to prevent neurological hyperperfusion injury.

There are many approaches to occluding the VGAM. Because our neuroradiologist is also a neurosurgeon, the transtorcular approach was selected. This technique decreases flow through the malformation and usually improves heart failure quicker and with less contrast media than the transarterial approach. The neurosurgeon must create a small burr hole to reach the torcular. Multiple detachable coils are then placed into the venous pouch of the vein of Galen. This provides for partial occlusion of the shunt and improvement of the infant's cardiac status. In the next several days a second transarterial embolization takes place using embolic glue. This glue will usually completely occlude the VGAM. The previous placed coils help prevent unwanted vascular spread of the glue through the pulmonary artery which can cause pulmonary vasoconstriction and symptoms consistent with pulmonary embolism.

Intracerebral hemorrhage due to venous hypertension is a potentially fatal complication of endovascular management. Some authors feel this complication can be avoided by staging the embolization procedure. Perforation of the venous sac has been reported to occur during positioning of the microcatheter during coil embolization, and can usually be managed by reversal of anticoagulation and continuation of coil embolization. Ischemic neurological deficits can occasionally be encountered after embolization. Pulmonary embolization with embolic agents is a concern due to the high flow across the intracranial shunt that drains immediately into the central venous system.

References:

Ashida Y, Miyahara H, Sawada H, Mitani Y, Maruyama K. Anesthetic management of a neonate with vein of Galen aneurysmal malformations and severe pulmonary hypertension. Paediatr Anaesth. 2005 Jun; 15(6):525-8.

Brunelle F. Arteriovenous malformation of the vein of Galen in children. Pediatr Radiol 1997 Jun ;27(6):501-13

Burry M, Reig A, Beierle E, Chen M, Mericle R. Extracorporeal membrane oxygenation combined with endovascular embolization for management of neonatal high-output cardiac failure secondary to intracranial arteriovenous fistula. Case report. J Neurosurg 2004 Feb; 100:197-200.

Chevret L, Durand P, Alvarez H, Lambert V, Caeymax L, Rodesch G, Devictor D, Lasjaunias P. Severe cardiac failure in newborns with VGAM. Prognosis significance of hemodynamic parameters in neonates presenting with severe heart failure owing to vein of Galen arteriovenous malformation. Intensive Care Med 2002 Aug; 28(8):1126-30.

Covert R. Response to prostaglandin E1 in neonates with intracranial arteriovenous malformation treated for suspected congenital heart disease. Pediatr Cardiol 1994 Mar-Apr; 15(2):81-4.

Cumming G. Circulation in neonates with intracranial arteriovenous fistula and cardiac failure. Am J Cardiol 1980 May; 45(5):1019-24.

Dahdah N.S., Alesseh A., Dahms B., Saker F. Severe pulmonary hypertensive vascular disease in two newborns with aneurysmal vein of galen. Pediatr Cardiol 2001 Nov-Dec;22(6):538-41.

Frawley, G, Dargaville P, Mitchell P, Tress B, Loughnan P. Clinical course and medical management of neonates with severe cardiac failure related to vein of Galen malformation. Archives of Disease in Childhood Fetal and Neonatal Edition 2002; 87:F144-F149.

Gailloud P, O'Riordan D, Burger I, Levrier O, Jallo G, Tamargo R, Murphy K, Lehmann C. Diagnosis and management of vein of galen aneurysmal malformations.
J Perinatol 2005 Aug; 25(8):542-51.

Hamasaki T, Kai Y, Hamada J, Miura M, Kondo Y, Ushio Y. Successful treatment of a neonate with vein of Galen aneurysmal malformation.
Pediatr Neurosurg 2000 Apr; 32(4):200-4.

Hendson L, Emery D, Phillipos E, Bhargava R, Olley P, Lemke R. Persistent pulmonary hypertension of the newborn presenting as the primary manifestation of intracranial arteriovenous malformation of the Vein of Galen. Am J Perinatol 2000; 17(8):405-10.

Holden A, Fyler D, Shillito J, Nadas A. Congestive heart failure from intracranial arteriovenous fistula in infancy. Clinical and physiologic considerations in eight patients. Pediatrics 1972 Jan; 49(1):30-9.

Jee S, Grow J, Faix R. Intraparenchymal brain hemorrhage and remote soft tissue arteriovenous malformation in a newborn infant. J Perinatol 2002 Dec; 22(8):664-6.

Levrier O, Gailloud P, Souei M, Manera L, Brunel H, Raybaud C. Normal galenic drainage of the deep cerebral venous system in two cases of vein of Galen aneurysmal malformation. Childs Nerv Syst 2004 Feb; 20(2):91-7.

McConnell, Aronin P, Vitek J. Congestive heart failure in neonates due to intracranial arteriovenous malformation: endovascular treatment. Pediatr Cardiol 1993 Mar; 14(2):102-6.

Menovsky T., van Overbeeke J.J. Cerebral arteriovenous malformations in childhood: state of the art with special reference to treatment. Eur J Pediatr 1997 Oct ;156(10):741-6.

Mericle R, Mickle J, Quisling R. Vein of galen malformation. Pediatr Neurosurg 1999 Nov; 31(5):279-80.

Mitchell P, Rosenfeld J, Dargaville P, Loughnan P, Ditchfield M, Frawley G, Tress B. Endovascular management of vein of Galen aneurysmal malformations presenting in the neonatal period. AJNR Am J Neuroradiol 2001 Aug; 22(7):1403-9.

Patton D.J., Fouron J-C Cerebral arteriovenous malformation: prenatal and postnatal central blood flow dynamics. Pediatr Cardiol 1995 May-Jun ;16(3):141-4.

Swanstrom S, Flodmark O, and Lasjaunias P. Conditions for treatment of cerebral arteriovenous malformation associated with ectasia of the vein of Galen in the newborn. Acta Paediatr 1994 Mar; 83(3):255-7.

Selective References:

Ashida Y, Miyahara H, Sawada H, Mitani Y, Maruyama K. Anesthetic management of a neonate with vein of Galen aneurysmal malformations and severe pulmonary hypertension. Paediatr Anaesth. 2005 Jun; 15(6):525-8.

Chevret L, Durand P, Alvarez H, Lambert V, Caeymax L, Rodesch G, Devictor D, Lasjaunias P. Severe cardiac failure in newborns with VGAM. Prognosis significance of hemodynamic parameters in neonates presenting with severe heart failure owing to vein of Galen arteriovenous malformation. Intensive Care Med 2002 Aug; 28(8):1126-30.

Frawley, G, Dargaville P, Mitchell P, Tress B, Loughnan P. Clinical course and medical management of neonates with severe cardiac failure related to vein of Galen malformation. Archives of Disease in Childhood Fetal and Neonatal Edition 2002; 87:F144-F149.