VEIN OF GALEN EMBOLIZATION

Surgical Specialty:	Neurointerventional Radiology
Authors:	Marla Ferschl, University of California San Francisco, CA

Background:

- General Considerations
    - Vein of Galen aneurysmal malformations (VGAM) are rare congenital arteriovenous malformations that result from abnormal arteriovenous fistulas between the embryonic choroidal arteries and the median prosencephalic vein, which is the embryonic precursor of the vein of Galen. This abnormal shunting causes progressive dilation of the vein and prevents its involution.[1,2]
    - VGAM accounts for about 30% of all pediatric congenital vascular malformations and 1% of all intracranial vascular lesions.[1,2]
    - Estimated incidence is 1:10,000 to 1:25,000 births.
    - About 30% of cases are diagnosed prenatally with ultrasonography and fetal MRI.
    - Lesions are classified as mural or choroidal depending on the vascular architecture[1,2]
    - Lesions provide a low-resistance conduit for intracerebral blood flow, which can cause high-output cardiac failure and severe pulmonary hypertension.
    - Neurointerventional embolization of feeding arteries and draining veins with coils or glue is the treatment of choice.
- Patient Considerations
    - Neonates often present with high-output cardiac failure and pulmonary hypertension and have a poor prognosis; nearly 80% of cardiac output may be delivered to the head.
    - Patients presenting in infancy or later are more stable and have a better prognosis. They may present with hydrocephalus, seizures, or neurocognitive impairment.[1-3]
    - Associated Comorbidities/Syndromes
        - High output cardiac failure, pulmonary hypertension, stroke, seizures, hydrocephalus, neurocognitive impairment

Anesthetic Planning:

- Pre-Anesthetic Evaluation
    - Thorough preoperative history and physical examination with optimization of cardiac status
    - Additional labs/tests indicated during work-up
        - Echocardiogram
        - Chest X-ray

        - Labs: Complete blood cell count, coagulation profile, type and cross-match, and basic metabolic profile in the setting of furosemide use.
    - Discussions to have with surgeon/family/intensive care team
        - Code status/goals of care
        - Stopping point for intervention due to cardiac decompensation or volume overload.
        - Hemodynamic goals during the case
        - Plan in case of unanticipated vessel rupture during coiling
- Specific or Unique Room Set-Up Requirements
    - General endotracheal anesthesia with neuromuscular blockade required
    - Drugs/Infusions:
        - Vasopressor infusions, including epinephrine, norepinephrine, and/or dopamine.
        - Vasodilating medications, including milrinone, nitroglycerin, and /or nitroprusside. The use of vasodilators in combination with low-dose dopamine may improve systemic perfusion and reduce metabolic acidosis.[3]
        - Consider nitric oxide in the setting of pulmonary hypertension.
        - Diuretics to combat volume overload.
        - Heparin: patients are heparinized during the procedure.
    - Monitors:
        - Standard American Society of Anesthesiologists monitors.
        - Arterial line monitoring in the setting of cardiac failure.
        - Central venous pressure (CVP) monitoring in the setting of cardiac failure. Urine output monitoring will not reflect the volume status, as these patients are on periprocedural diuretics and osmotic agents for the management of the raised intracranial pressure (ICP). Cannulation of the internal jugular vein can be a challenge as the intracranial shunt makes even the venous column pulsatile, and venous blood appears bright red.
        - Adequate intravenous (IV) access with 2 “large bore” IVs is recommended.
        - Frequent monitoring of arterial blood gas and electrolytes.
        - Careful attention to temperature, especially in neonates and infants.
        - Foley catheter to monitor urine output.
    - Blood Availability
        - Washed, cross-matched blood should be immediately available for transfusion.
        - Fluid warmer and transfusion supplies should be prepared
    - ICU recovery is planned; neonates will often remain intubated postoperatively.

Intraoperative Considerations:

- General
    - The procedure is often staged; patients will be least stable during the first procedure.
    - ***Neonatal mortality approaches 50% in the setting of heart failure***[1]
- Induction
    - Slow controlled induction to avoid significant changes in blood pressure; opioids may help smooth hemodynamic variability. Avoid hypo or hypertension, hypoxia, and hypo or hypercarbia. Use caution with ketamine if the ICP is elevated.
- Intubation
    - Increased risk of difficult airway if the head size is large. The head should be stabilized and can be positioned in a head ring using a small pillow/roll under the

back or shoulders to improve the view. Backup airway devices (e.g., video laryngoscope) should be available and ready.

- Positioning
    - Supine position in the neuro-interventional radiology suite.
    - Patient access may be limited by equipment/spacing. The patient is usually rotated 90-180 degrees from the anesthesia machine.
    - Avoid kinking of neck veins with head positioning, which may occur with macrocephaly.
- Maintenance
    - Anesthetics agents that reduce the cerebral metabolic rate and reduce oxygen consumption are preferred (total intravenous anesthesia (TIVA) +/- low dose volatile anesthetic)
    - Maintain neuromuscular blockade and monitor the depth of neuromuscular blockade.
    - Avoid nitrous oxide in the setting of pulmonary hypertension and possible air embolism.
    - Consider dextrose-containing maintenance IV fluids in neonates to avoid hypoglycemia.
    - Patients on anti-epileptic medications may have increased metabolism of muscle relaxants and/or narcotics.
- Hemodynamic/Physiologic goals
    - Goal is to maintain cardiovascular stability; hypotension will worsen already compromised cardiac perfusion.
    - Hypotension may occur due to intra-arterial use of vasoactive agents by the neuroradiologists. It can be treated with phenylephrine
    - If the AVM is completely coiled, ventricular afterload may suddenly rise, leading to decompensation.
- Surgical Considerations
    - A significant volume of contrast may be administered by the radiologist, which may exacerbate heart failure. Close communication about volume status and hemodynamics is essential.
    - The use of a continuous flush system by the radiologist may also result in the administration of a large volume of crystalloid over the course of the procedure.
    - Intraoperative rupture is possible and requires rapid protamine administration and blood transfusion.
- Emergence/Disposition
    - Patients usually return to the intensive care unit (ICU) intubated.
    - If extubated is planned, a smooth emergence that avoids hypertension is preferred.
- Postoperative Care
    - Hypertension should be avoided in the first 48 h after the procedure to prevent neurological hyperperfusion injury, as blood previously transmitted through the AVM is redirected to smaller, normal vasculature after the AVM has been removed, with the subsequent inability of the vessels to handle the increased flow.

    - Among all patients who survive embolization, 66% are neurologically normal, 14% have transient neurological symptoms, 11.5% have mild neurological symptoms, and 8.5% have a severe irreversible deficit.[5]

Case-Specific Complications/Pitfalls

- High-output cardiac failure: Many neonates present with cardiac failure; those who do not are prone to developing failure associated with large volume infusions intraoperatively. Close cardiovascular monitoring and tight blood pressure control are imperative.
- Coronary perfusion: Increased diastolic flow to the VGAM reduces coronary blood flow; this is exacerbated by increased ventricular pressure, which reduces subendocardial perfusion, leading to myocardial ischemia and potentially exacerbating right heart failure.
- Hypothermia: Patients are prone to heat loss due to age; this is exacerbated by high fluid volumes given by the radiologist. Care should be taken to maintain body temperature with the use of forced-air warming devices, fluid warmers, and a humidified breathing circuit.
- Pulmonary hypertension: common in neonates with VGAM and severe heart failure, due to arteriolar changes in utero associated with high fetal pulmonary blood flow. Steps to avoid increasing pulmonary vascular resistance should be taken. Inhaled nitrous oxide may be beneficial.[5]

References

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Reviewed by:

Reviewer #1: Alina Lazar, M.D., Lurie Children's Hospital

Reviewer #2: Debnath Chatterjee, MD, University of Colorado

Reviewer #3: Gina Whitney, MD, University of Colorado

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