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Perioperative Considerations in Patients with Vein of Galen Malformations Undergoing Embolization–A Single-Institution Case Series

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Abstract

Vein of Galen malformation (VOGM) is a congenital, intracranial vascular malformation, with an extracardiac shunt. Neonates can present with high output cardiac failure, pulmonary hypertension, or multiorgan failure and are at high risk of perioperative complications, especially in remote locations. We conducted a retrospective single-center analysis of the perioperative management of patients with VOGM presenting for embolization. Patients were identified by querying both the hospital billing dataset using International Classification of Diseases-10 diagnosis or billing code and the Neuro-interventional Radiology Database, from January 2011 to March 2020. As many as 14 patients were identified, 12 of which underwent definitive treatment. Six patients who underwent embolization in the neonatal period had pulmonary hypertension. Those children required varying degrees of hemodynamic and respiratory support preoperatively and experienced significant intraoperative events, including one intraoperative cardiac arrest. Caring for these critically ill patients in a remote location requires proper planning to prevent adverse outcomes.

Keywords
- intraoperative
- pulmonary hypertension
- vein of Galen malformation

Introduction

The vein of Galen Malformation (VOGM) is a congenital, high flow, extracardiac shunt comprising 30% of all pediatric vascular and less than 1% of all cerebral arteriovenous malformations.[1,2] The clinical presentation varies depending on the age, with high-output heart failure (HF), pulmonary hypertension (PH), and/or multiorgan failure seen mainly in neonates.[2–4] Without treatment, mortality is over 90% by infancy, though with recent advances in endovascular technique this has been reduced to 11%.[2,4] These children are at high risk for perioperative complications, with only a few specialized centers encountering them on an annual basis. We describe our perioperative experience with these patients who present for neuroembolization.

Case Series

In this retrospective single-center analysis, patients with VOGM were identified by querying both the hospital billing dataset using International Classification of Diseases-10 diag-

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nosis or billing code and the Neuro-interventional Radiology database, from January 2011 to March 2020. We excluded patients who did not have a confirmed diagnosis of VOGM, who had their first embolization done at another facility or any subsequent embolization done after the initial admission.

Results

Out of 14 patients, 12 underwent a total of 24 embolizations.

Half of these patients were diagnosed prenatally, 21% immediately after birth, 21% during early infancy (increased head circumference, developmental delay, feeding difficulties and craniosynostosis), and 7% in the adolescence period (intracranial bleed).

All patients who underwent embolization in the neonatal period had echocardiographic evidence of extracardiac shunting and over circulation manifesting as PH (100%), dilatation of right ventricle (RV, 83%), diminished RV systolic function (33%), right to left shunt across patent ductus arteriosus (PDA, 66%), and retrograde flow during diastole in the descending aorta (50%). Considering the criticality of these patients, some of them required varying degrees of respiratory support; one or more pulmonary vasodilators such as phosphodiesterase inhibitor (sildenafil), endothelial receptor antagonist (bosentan), prostacyclin or prostacyclin analog (treprostinil or epoprostenol) or inhaled nitric oxide; vasopressors such as dopamine, milrinone or epinephrine; and multiple embolizations' (patients # 1, 3, 4 and 11 undergoing three, two, four, and seven embolization's respectively) to maintain hemodynamic and neurophysiological stability as highlighted in ►**Table 1**. The decision for intervention was based on a multidisciplinary team-based assessment of patient's clinical status rather than the use of Bicêtre Neonatal evaluation score. No intervention was done for two patients, due to poor prognosis.

The airway was secured in all patients prior to the intervention. Intraoperative anesthesia was maintained with combination of either inhaled anesthetic (isoflurane or sevoflurane), opioid administered either as bolus only, infusion, or both with agents such as morphine, fentanyl, or remifentanil), midazolam, dexmedetomidine, and muscle relaxant (vecuronium or rocuronium).

Eighty-three percent of the neonates developed significant intraoperative events (►**Table 2**), such as hypotension (defined as a documented event by the primary team, or based on intervention such as titration of existing vasopressors/ inotropes, addition of a new agent), desaturation with or without change in end-tidal CO_2 from the baseline (defined as a documented event by the primary team or based on an intervention such as hand bag ventilation with 100% fraction of inspired oxygen or titration of pulmonary vasodilators), or cardiac arrest. There was one death in our 12-patient cohort (8% mortality).

Lastly, patients embolized in the neonatal period had longer length of stay (median: 64.5 days, range: 15–187 days) compared to those presenting in infancy or later in life (median: 1.5 days, range: 1–23 days).

Discussion

In this retrospective review, we describe our perioperative experience with patients with VOGM presenting for their initial embolization at our institution. Patients who underwent embolization in the neonatal period were critically ill, requiring substantial cardiorespiratory support preoperatively, multiple embolization procedures, and had longer length of stay during their initial admission.

The underlying pathophysiology in these patients typically manifests after birth with loss of low resistance placental circulation and rising systemic vascular resistance favoring the blood flow to the extracardiac shunt, leading to high output HF and systemic steal. Right HF occurs because of volume and pressure overload. This is exacerbated by circulatory steal impairing coronary blood flow. Elevated right-sided pressures cause shunting of blood right to left across the PDA and/or an atrial septal defect contributing to hypoxia. This leads to bowing of the intraventricular septum, which in turn compromises left ventricular volume, cardiac output and systemic perfusion, leading to lactic acidosis and potentially multiorgan system failure. Some of these physiological changes were evident on the preoperative echocardiographic findings in our neonates, including PH. They received varying degrees of respiratory and cardiac support and were at high risk of perioperative complications such as intraoperative pulmonary hypertensive crisis and cardiac arrest.

The key goals of anesthetic management for patients with PH include maintaining cardiac contractility and avoiding increase in pulmonary vascular resistance by maintaining adequate analgesia and avoiding factors that can cause increase in right ventricular strain such as increased afterload, decreased coronary blood flow, reduced preload, loss of sinus rhythm, and depressed right ventricular contractility.[5] PH crisis clinically manifests as an abrupt decline in end-tidal CO_2, desaturation, bradycardia, and cardiovascular collapse. Management of PH crisis includes increasing the fraction of inspired oxygen to 100%; optimization of sedation, analgesia, and ventilation (avoid overdistension of lungs or high positive end-expiratory pressures); and use of vasopressors/inotropes to increase systemic perfusion and initiating pulmonary vasodilator agents. Availability of arterial line to accurately monitor blood pressure and central venous access for administration of vasopressors is necessary.

Additionally, avoiding hypothermia, monitoring volume of flush used by interventional radiologist to avoid fluid overload, and worsening of HF and ensuring availability of trained personnel to assist during a crisis are essential.[6]

Limitations of our report include small sample size, data obtained from a retrospective chart review, missing data, difficulty on obtaining a composite picture regarding the clinical status of the patients (due to the use of the 'copy forward function'), variability of information in the electronic medical record, and lack of long-term outcomes due to loss of follow-up.

Table 1 Preoperative respiratory and hemodynamic support

Patient number	Inhaled prostacyclin	iNO	PH-specific medications	Supplemental oxygen	Inotropes/Vasopressors	Alive 30 days after first embolization
Patient 1 (embolization in neonatal period)						
Embolization 1	No	No	No	Yes, NC at 1L/min (21% O_2)	No	Unknown, transferred to outside hospital
Embolization 2	Yes	No (switched epoprostenol to iNO for OR)	Yes (sildenafil)	Yes, 54% FiO_2, intubated	Yes (dopamine)	
Embolization 3	Yes	No (started iNO for IR)	Yes (sildenafil)	Yes, 45% FiO_2, intubated	Yes (dopamine)	
Patient 2						
Embolization 1	No	No	No	No	No	Home
Patient 3 (embolization in neonatal period)						
Embolization 1	No	Yes	No	Yes, 100% FiO_2, intubated	Yes (dopamine, milrinone)	Yes, but passed away prior to discharge
Embolization 2	Yes	Yes	Yes (sildenafil, bosentan)	Yes, 80% FiO_2, intubated	Yes (milrinone)	Yes
Patient 4 (embolization in neonatal period)						
Embolization 1	No	No	No	Yes, 100% FiO_2, intubated	Yes (dopamine)	Yes
Embolization 2	No	Yes	No	Yes, 30% FiO_2, intubated	Yes (dopamine, epinephrine)	
Embolization 3	No	Yes, iNO at 1 ppm	No	Yes, 30% FiO_2, intubated	Yes (dopamine)	
Embolization 4	No	No	No	No, 21% FiO_2, intubated	Yes (dopamine)	
Patient 5 (embolization in neonatal period)						
Embolization 1	No	No (started in the OR)	No	Yes, 40% FiO_2, HFLNC	No	Yes
Patient 6						
	No	No	No	No	No	Yes
Patient 7						
	No	No	No	Yes, intubated	phenylephrine	Yes
Patient 8						
	No	No	No	No	No	Yes
Patient 9 (embolization in neonatal period)						
	No	No	No	No	PO digoxin	Yes
Patient 10						
	No	No	No	No	PO digoxin	Home

(*Continued*)

Table 1 (*Continued*)

Patient number	Inhaled prostacyclin	iNO	PH-specific medications	Supplemental oxygen	Inotropes/Vasopressors	Alive 30 days after first embolization
Patient 11 (embolization in neonatal period)						
Embolization 1	No	No	No	Yes, intubated (in NICU)	yes (dopamine)	Yes
Embolization 2	No	Yes	No	Yes, 95% FiO_2, intubated	Yes (dopamine, milrinone)	
Embolization 3	No	Yes	No	Yes, 90% FiO_2, intubated	Yes (dopamine, milrinone)	
Embolization 4	No	Yes	No	Yes, 80% FiO2, intubated	Yes (dopamine, milrinone)	
Embolization 5	No	No	Yes (sildenafil, bosentan, treprostinil)	Yes, 100% FiO2, intubated	Yes (milrinone)	
Embolization 6	No	No	Yes (sildenafil, bosentan, treprostinil)	Yes, 50% FiO2, intubated	No	
Embolization 7	No	No	Yes (sildenafil, bosentan, treprostinil)	Yes, NC at 4L/min (100% O_2)	No	
Patient 12 (no intervention)						
	N/A	N/A	N/A	N/A	N/A	Passed away
Patient 13 (no intervention)						
	N/A	N/A	N/A	N/A	N/A	Passed away
Patient 14						
	No	No	No	No	No	Home

Abbreviations: FiO2, fraction of inspired oxygen; HFLNC, High Flow Nasal Cannula; IR, intervention radiology; iNO, inhaled nitric oxide; N/A, not applicable; NC, nasal cannula; NICU, neonatal intensive care unit; OR, operating room; PH, pulmonary hypertension; ppm, parts per million.

Table 2 Intraoperative events

Patient number	Hemodynamic changes	Treatment	Outcome
1 (1st embolization)	Loss of end-tidal, desaturation, brady dysrhythmia, cardiac arrest	Chest compressions, epinephrine boluses, albuterol, iNO at 40ppm, epinephrine and dopamine infusions	Coiling halted, patient transported to the NICU intubated.
1 (2nd embolization)	Intermittent bigeminy, possible hypotension	Boluses of phenylephrine and titration of dopamine	Case completed
1 (3rd embolization)	Hypotension	Titration of dopamine	Case completed
2	None	N/A	Successful coiling
3 (1st embolization)	Desaturation, hypotension, metabolic acidosis	Epinephrine and vasopressin started, received sodium bicarbonate, titration of milrinone and dopamine	Successful coiling
3 (2nd embolization)	Desaturations and suboptimal ventilation Change in hemodynamics during groin pressure by IR	Improved with handbag ventilation and changed of endotracheal tube Improved hemodynamics with release of groin pressure	Successful coiling
4 (1st embolization)	Desaturation, increase in CVP, hypotension	iNO started, low-dose epinephrine infusion, hand ventilation with 100%	Successful coiling
4 (2nd embolization)	Hypotension	Phenylephrine, titration of pressors, iNO increased to 20ppm	Case completed
4 (3rd embolization)	None	N/A	
4 (4th embolization)	None	Titration of dopamine	Case completed
5	Desaturation, hypotension, decrease in end tidal CO_2 while coiling a large shunt	FiO_2 100%, iNO at 40 ppm, milrinone, dopamine infusions, one non-arrest dose of epinephrine	Successful coiling
6	None	N/A	Successful coiling
7	None	Rise in ICP during breath holding, CSF drained from intraventricular catheter, 3% saline infusion	Successful coiling
8	None	N/A	Successful coiling
9	None	N/A	Successful coiling
10	Hypotension	Fluid bolus	Successful coiling
11 (1st embolization)	Desaturation, no change in end tidal CO_2	Milrinone infusions initiated, iNO started, dopamine titrated	Successful coiling
11 (2nd embolization)	No data available	No data available	No data available
11 (3rd embolization)	None	N/A	Case completed
11 (4th embolization)	Desaturation, rising end tidal CO_2, acidosis	Increased iNO, started epoprostenol, titrated milrinone and dopamine	Case completed

(*Continued*)

Table 2 (*Continued*)

Patient number	Hemodynamic changes	Treatment	Outcome
11 (5^{th} embolization)	Desaturation, no change in end tidal CO_2	One non-arrest dose of epinephrine given	Case completed
11 (6^{th} embolization)	Desaturation, reduction in end tidal CO_2, decrease in heart rate, no hypotension	Bag ventilation with 100% FiO_2, increased iNO to 40, increased inhalational agent, albuterol given	Case completed
11 (7^{th} embolization)	Bronchospasm	Albuterol	Case completed
12	No intervention		
13	No intervention		
14	None	N/A	Successful coiling

Abbreviations: CSF, cerebrospinal fluid; FiO2, fraction of inspired oxygen; ETT, endotracheal tube; ICP, intracranial pressure; iNO, inhaled nitric oxide; N/A, not applicable; NICU, neonatal intensive care units.

In summary, care of these high risk critically ill patients in remote locations requires proper planning and arrangement of resources to manage crisis.

Conflict of Interest
None.

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