



Clinical Guidelines

Vein of Galen Aneurysmal Malformation (VGAM)

Document Control Information

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1. Background

VGAM is a rare congenital midline arteriovenous vascular malformation causing shunting of arterial blood into the median prosencephalic vein of Markowski^{1,2}.

All neonates should be referred to the specialist service in Great Ormond Street Hospital NICU/PICU as clinical priority.

2. Presentation

- Antenatal diagnosis on foetal ultrasound
- Postnatal diagnosis most commonly high output heart failure³ (tachycardia, tachypnoea, cyanosis, pulmonary hypertension, hypoxaemia, multi-organ dysfunction)
- All unexpected heart failure signs in neonates require auscultation of fontanelle for bruit and cranial ultrasound to be performed

3. Management

Asymptomatic - antenatal diagnosis

- Consider high flow oxygen / Non-invasive CPAP
- IV access x2 (**Do not delay transfer, if access difficult consider IO**)
- Consider inotropic support
- NBM and fluid restrict to 60ml/kg/day 10% glucose/0.9% saline
- Consider diuresis 0.5mg-1mg/kg furosemide qds, urinary catheter insertion, monitor urine output
- Referral to CATS for urgent transfer to specialist centre

Symptomatic- postnatal diagnosis

- Medical management aim: to improve systemic, coronary and end organ perfusion by redirecting blood flow away from the VGAM
- Reassess after intervention – perform blood gas, paO₂, paCO₂, lactate, electrolytes, HR, rhythm, pulses and BP

First line therapy

- Elective intubation – consider cuffed ETT
- Consider starting inotropic support prior to induction - low dose adrenaline first choice
- Ventilate and oxygenate – target PEEP of 4-6cmH₂O, SaO₂ >95%
- Optimise BP and avoid tachycardia with cautious 5ml/kg crystalloid boluses – monitor for increasing hepatomegaly

- Neuroprotective strategies - sedate and muscle relax with morphine and vecuronium infusions, regular pupillary observations, 30 degrees head up tilt
- If signs of raised ICP, consider osmotherapy (3ml/kg of 2.7% saline aiming for Na 145-150mmol/L or 0.25g/kg mannitol)
- Aim for normothermia – to minimise further peripheral vasodilation and tachycardia
- If any clinical evidence/suggestion of seizure load with phenobarbitone 20mg/kg
- Fluid restrict as previous. Consider diuresis with furosemide 0.5-1mg/kg qds
- Insert urinary catheter and monitor urine output
- Consider central venous and arterial access - umbilical lines often used

Second line therapy

Refractory hypoxia

- Increase MAP – increase PEEP (8-10 cm H₂O)
- FiO₂ to 1.0
- **Discuss** with CATS Consultant – the following may be considered:
 - IV magnesium sulphate 50mg/kg IV/IO (over 20 mins)
 - Sodium bicarbonate 8.4% 1mmol/kg
 - Inhaled nitric oxide at 20ppm

Refractory cardiac failure

- Insert IO or central venous access
- Start adrenaline if not already started (0.1-1mcg/kg/min)
- Consider milrinone for diastolic dysfunction. Monitor diastolic BP, if low may require low dose noradrenaline
- Aim for normothermia, monitor with oesophageal temperature probe
- Ensure adequacy of sedation and muscle relaxation
- Discuss with CATS Consultant – consider prostaglandin E₂ infusion (using the duct as a pressure relief valve for failing right ventricle)

The only intervention that will resolve refractory high output heart failure is partial embolisation of VGAM to redirect flow.

A timely transport to a specialist centre via appropriate retrieval service is paramount.

References

1. Recinos PF et al (2012) *Neurosurg Clin N Am* 23:165-177
2. McSweeney N et al (2010) *ADC* 95:903-909.
3. Brinjikji et al (2017) *Am J Neuroradiology* 38 (12) 2308-2314