



## 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<sup>1,2</sup>.

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 failure<sup>3</sup> (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 – consider blood gas, paO<sub>2</sub>, paCO<sub>2</sub>, 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 4-6cmH<sub>2</sub>O, SaO<sub>2</sub> >95%.
- Optimise BP and avoid tachycardia with cautious 5ml/kg crystalloid boluses – monitor for increasing hepatomegaly.
- Consider inotropic support – low dose adrenaline first choice.

- Neuroprotective strategies - sedate and paralyse 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 H<sub>2</sub>O)
- FiO<sub>2</sub> 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 paralysis.
- 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 transfer 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