Clinical Guidelines

Vein of Galen Aneurysmal Malformation (VGAM)
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 failure\(^3\) (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, 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 4-6cmH\(_2\)O, SaO2 >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 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 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**