

NHS CATS
Children's Acute Transport Service



Clinical Guidelines

Vein of Galen Aneurysmal Malformation (VGAM)

Document Control

Author	A Jones	Author Position	CATS Consultant
Document Owner	M Clement	Document Owner Position	CATS Operational and Nursing Lead
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Background

VGAM is a rare congenital midline arteriovenous vascular malformation causing shunting of arterial blood into the median prosencephalic vein of Markowski, resulting in high output cardiac failure and poor systemic blood flow. *Definitive management is neurovascular embolisation.* Neonates with an antenatal or postnatal diagnosis of VGAM should be referred to GOSH PICU, regardless of clinical condition. The lead team at GOSH for VGAM patients is interventional radiology/neurology. This is a nationally commissioned service.

Presentation

- Antenatal diagnosis on foetal ultrasound.
- Postnatal presentation is most commonly with signs and symptoms of high output heart failure (tachycardia, tachypnoea, cyanosis, pulmonary hypertension, hypoxaemia, multi-organ dysfunction).
- All neonates with heart failure and/or PPHN require auscultation of the fontanelle for a bruit, and a cranial ultrasound.

Management

Antenatally diagnosed VGAMs can be asymptomatic. Consider:

- High flow oxygen or CPAP if developing respiratory symptoms or signs of heart failure.
- NBM and fluid restrict to 60ml/kg/day with 10% glucose/0.9% sodium chloride.
- Diuresis with 0.5mg-1mg/kg furosemide IV qds; urinary catheter insertion, monitoring of urine output.
- Potential for deterioration; including need for intubation and inotropic support.

Postnatal diagnosis is typically symptomatic.

The goal of medical management is the optimisation of cardiac function to maintain organ perfusion until the endovascular procedure can be carried out. This procedure reduces the effects of the AV shunt on cardiac function by occlusion of the pathological arterial feeders of the vascular malformation. This is the only definitive therapy. *A timely transport to the specialist centre is essential.*

Transport considerations

First line therapy for the symptomatic neonate:

- Non-invasive respiratory support may be sufficient for transport.
- If not, intubation with a cuffed ET tube.

- Consider starting inotropic support prior to induction of anaesthesia - low dose adrenaline is first line.
- Ventilate and oxygenate - PEEP of 5 cm H₂O, SaO₂ >95%.
- Optimise BP and avoid tachycardia with cautious 5ml/kg crystalloid boluses – monitor for increasing hepatomegaly and worsening pulmonary oedema.
- Close monitoring of urine output, lactate and SvO₂ is desirable, with regular reassessment.
- Neuroprotective strategy if haemodynamic status allows - sedate and muscle relax; perform regular pupillary observations, 30 degrees head up tilt, normothermia.
- If signs of raised ICP, consider osmotherapy (3ml/kg of 2.7% saline aiming for Na 145-150mmol/L).
- If any clinical evidence/suggestion of seizures load with anticonvulsant.
- Consider central venous and arterial access - umbilical lines are often used initially.
- Avoid inserting lines in the femoral vessels if possible.

Second line therapy for the symptomatic neonate with refractory hypoxia:

- Discuss with the CATS consultant.
- Increase mean airway pressure – increase PEEP (8-10 cm H₂O) and assess response; high PEEP may adversely affect cardiac function.
- Manage as per severe PPHN – normal pH, consider sodium bicarbonate, avoid hypercarbia, ensure adequacy of sedation and paralysis.
- Inhaled nitric oxide at 20ppm can be trialled but the very limited evidence available suggests it may not be beneficial.

Second line therapy for the symptomatic neonate with refractory cardiac failure:

- Discuss with the CATS consultant.
- Start adrenaline if not already started, avoiding high doses if possible.
- Consider milrinone for diastolic dysfunction; monitor diastolic BP and if low consider noradrenaline.
- Aim for normothermia, monitor with oesophageal temperature probe.
- Ensure adequacy of sedation and muscle relaxation.
- Consider prostaglandin E2 infusion, using the duct as a 'pressure relief valve' for the failing right ventricle.