NHS Children's Acute Transport Service

Clinical Guidelines

Vein of Galen Aneurysmal Malformation (VGAM)

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Applicable to	**All CATS employees**

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 specialist centre in Great Ormond Street Hospital or Glasgow NICU/PICU as clinical priority.

2. Presentation

- Antenatal diagnosis on fetal ultrasound.
- Postnatal diagnosis as heart failure (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 antenatally diagnosed:

- 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/k/d 10% dextrose/0.9% saline
- Consider diuresis 0.5mg-1mg/kg furosemide qds, urinary catheter insertion, monitor urine output
- URGENT transfer to specialist centre, call CATS.

Symptomatic postnatally diagnosed:

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

First line therapy:

- Elective intubation – consider cuffed ETT, ketamine and atracurium induction, cautious doses, consider starting inotropic support first.
- Ventilate and oxygenate – PEEP of 4-6cmH2O, RR 30-35, Itime 0.5-0.75sec, aim for PaCO2 4.5-5kPa, PaO2 12-14kPa, 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.
- Sedate and paralyse with IV morphine and vecuronium infusions, regular pupillary observations, 30 degrees head up tilt.
- If bradycardia or pupillary changes suggestive of raised ICP, consider osmotherapy (3ml/kg of 2.7% or 3% saline aiming for Na 145-150mmol/L or 0.25g/kg mannitol).
- Avoid pyrexia – to avoid further peripheral vasodilation and tachycardia.
- If any clinical evidence/suggestion of seizure activity load with phenobarbitone 20mg/kg.
- Fluid restrict as previous. Consider diuresis furosemide 0.5-1mg/kg qds.
- Urinary catheterisation and monitor urine output.
- Central venous access and arterial access 'desirable', umbilical lines often used.

Second line therapy:

Refractory hypoxia:

- Increase MAP – increase PEEP (8-10 cm H2O), increase PIP, increase i-time
- 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 (this may not be beneficial for the patient, discuss first).

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, if started monitor diastolic BP, if low will 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 blow off valve for failing right ventricle).

Note: the only thing that will resolve refractory high output heart failure is partial embolization of VGAM to redirect flow; a timely transfer to a specialist centre via appropriate retrieval service is paramount. Telephone CATS 0800 085 0003.

References

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