Clinical Guidelines

Vein of Galen Aneurysmal Malformation (VGAM)

Document Control Information

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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. 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 (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