

Emergency Referrals to Quaternary Services (including ECMO and Vein of Galen)

Extracorporeal Membrane Oxygenation (ECMO)

ECMO for babies \geq 34+0 weeks gestation (and >1.8 kg) who have one of following, consider early discussion with the ECMO centre:

 Hypoxic respiratory failure with oxygenation index (OI) > 40 sustained for at least 4 hours despite optimising ventilation.

OI = (Mean airway pressure (MAP) (cm H_2O) x FiO₂(%)) / (Arterial PaO₂ (Kpa) x 7.5)

- Hypoxic respiratory failure with OI > 30 on inhaled nitric oxide and optimised ventilation
- Consider referral if hypoxic respiratory failure with OI >24 plus vasoactive inotrope score (VIS) > 18 (use of VIS to support decision-making is currently being evaluated).
 - VIS = (Dopamine + dobutamine (mcg/kg/min)) + $100 \times$ (adrenaline +noradrenaline (mcg/kg/min)) + $10 \times$ (milrinone (mcg/kg/min)) + $10 \times$ (vasopressin unit/kg/min)
- o If unable to measure OI (no arterial sample);
 - Inability to maintain pre ductal sats >85% or post ductal sats >70%
 - Ongoing need for high pressure ventilation (PIP > 28 cm H_2O on conventional ventilation, or MAP > 17 cm H_2O on HFOV)
- Hypercapnic respiratory failure with PaCO2 >10/pH < 7.15 despite optimal ventilation
- Inadequate tissue oxygen delivery (indicated by persistent pH< 7.15 or lactate > 5 mmo/L)
- Severe pulmonary hypertension resistant to pulmonary vasodilator therapy
- Severe cardiac compromise/dysfunction (discuss with cardiology team re appropriateness for ECMO)

Exclusion criteria:

- Significant coagulopathy or uncontrollable bleeding
- o Major intracranial haemorrhage
- Evidence of severe brain injury
- Irreversible lung injury
- Major life limiting congenital/chromosomal anomalies (including trisomy 13 and 18, NOT including trisomy 21).
- o Major cardiac malformation
- o Cardiac arrest other than immediately after birth

Conditions where ECMO may be considered;

Respiratory failure;

- \circ PPHN
- Air leak syndromes
- Sepsis (especially early-onset sepsis)
- Meconium aspiration syndrome
- Some congenital diaphragmatic hernias, especially where there is a correctable component to disease – infection or surfactant deficiency (see CDH guideline)



Cardiac failure;

- Post cardiac surgery
- Myocardial ischaemia following HIE, where there is relative preservation of cerebral function
- Other rare causes of myocardial ischaemia, eg myocardial infarction or amniotic fluid embolus
- Poor cardiac function (possible metabolic/genetic condition)

Information the ECMO centre will require (gather your information before dialling)

- Likely cause of the respiratory/cardiac severe disease (working diagnosis)
- Presence of any life-limiting congenital anomalies
- o Degree of cerebral injury (CFM pattern, fits, degree & duration of acidosis and hypoxia)
- All gas results (pH, pCO2, OI, BE and lactate)
- Lab results (FBC, clotting, renal and liver function tests)
- Progression of inotropes (doses and drugs used)
- o iNO duration and ppm
- Co-existing conditions (sepsis, PPROM and lung hypoplasia, clots in IVC/aorta, vascular catheter associated injury/ischaemia)
- o The condition or baby's broader condition needs to be potentially reversible
- Results of echo (if performed)
- o Result of a cranial US

Making a referral

- Southampton; Phone number; 023 8077 5502 (SORT- Southampton Oxford Retrieval Teamask for Southampton ECMO referral)
- Leicester (mobile ECMO centre). Phone number; 0300 300 3200 Leicester is the only service which offers mobile ECMO facility but this is not available 24/7.
- London centres. Phone number 0800 085 0003 (CATS -Children's Acute Transport Serviceask for ECMO referral)

Vein of Galen Malformation (VOGM)

- VOGM is a rare cerebral vascular malformation, causing a low resistance arterio-venous malformation of the choroidal arterial system.
- After birth it can present as progressive high output cardiac failure. Signs on echo including dilated SVC, dilated right side of heart, tricuspid regurgitation and right to left ductal flow. There may be pulmonary hypertension due to high volume load on right side of heart and persistent high pulmonary vascular resistance. Additionally, there may be signs of "steal" with diastolic flow reversal in descending aorta.
- o A bruit may be heard auscultating over the fontanelle.



- A cranial ultrasound with dopplers should be performed to confirm diagnosis if there is suspicion from the echo.
- Treatment is initially supportive (consideration for low dose inotropic support (adrenaline), peripheral vasodilatation (dobutamine), milrinone, diuretics and prostin, caution regarding use of of iNO as may increase overload to lungs further)
- Urgent referral for neurovascular intervention is required and should not be delayed to usual working hours.

Making a referral

- Great Ormond Street Hospital are the national centre for treatment of VOGM. Given the infrequency of this diagnosis and the number of teams involved, there are common problems with knowing who to call within GOSH for the referral. The pathway is;
 - Acute referrals should be made to the duty consultant paediatric neurologist via GOSH switchboard on 020 7405 9200.
 - Once the referral has been accepted, the transfer is to PICU (as opposed to CICU/NICU).

Liver Specialist

- Urgent referrals include;
 - Upper GI bleed secondary to liver pathology
 - Acute liver failure
 - Acute pancreatitis
 - Cholangitis
 - o Trauma to liver or pancreas
 - Neonatal cholestasis with acholic stools

Making a referral

- Birmingham Liver team;
 - o For urgent referrals call switch board on 121 333 9999 and bleep 200
- o Kings College Hospital Liver team
 - o Call 0203 299 9000 Ext 37812

Metabolic Specialist

Patients with following diagnoses require urgent referral to the metabolic team;

- o Hyperammonaemia
- Unexplained persistent lactic acidosis
- Unexplained persistent metabolic acidosis
- Unexplained hypoglycaemia



- o Neonate with suspected metabolic liver failure
- o New biochemical metabolic diagnosis for dietary management
- New diagnosis osteopetrosis with preserved vision
- o Newly diagnosed Pompe Disease prior to commencing enzyme replacement therapy
- Patient with known metabolic disorder on emergency regimen in local hospital and deteriorating, or not improving after 48hrs
- o Metabolic decompensation requiring total parenteral nutrition (TPN)
- o Undiagnosed possible metabolic disorder requiring urgent peri-mortem investigations

Making a referral

- Great Ormond Street Hospital; Call switchboard on 020 7405 9200 and ask for the on call metabolic medicine registrar.
- Evelina (Guy's and St Thomas Hospital); Call switchboard on 020 7188 7188 and ask for metabolic registrar or consultant.