NHS Children's Acute Transport Service

Clinical Guideline

CATS Metabolic Referrals

Document Control Information

Author	E. Randle	**Author Position**	CATS Consultant
Document Owner	Metabolic/CATS Team	**Document Owner Position**	CATS/metabolic services
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Children's Acute Transport Service provides paediatric intensive care retrieval for Great Ormond Street, The Royal Brompton and St Mary's NHS Trusts. Funded and accountable to the North Thames Paediatric Intensive Care Commissioning Group through Great Ormond Street NHS Trust

Establish if a metabolic condition is known and to which centre.

Known metabolic conditions should be discussed with the duty GOSH (or base centre) metabolic consultant.

If a new metabolic condition is suspected, early discussion with the metabolic on-call team is advisable, especially for hyperammonaemia.

Assessment

Presentation

- encephalopathy, seizures and apnoea
- metabolic acidosis but **NB** most metabolic acidosis is due to poor tissue perfusion (sepsis/low cardiac output) rather than metabolic disease.
- hypoglycaemia
- hypocapnia with tachypnoea
- cardiac failure/cardiomyopathy
- liver dysfunction
- dysmorphism
- hydrops
- hyperammonaemia

History and clinical examination

Ask about

- involvement of a metabolic team
- family history of metabolic disorder
- previous pregnancy losses
- consanguinity
- any problems during the pregnancy
- poor feeding, growth or development

Examination should focus on cardiorespiratory and neurological status.

Initial screening tests for metabolic disease

- odour of baby and urine
- blood glucose
- venous sample for serum ammonia (trend data is important)
- acid-base status
- anion gap ([Na+ + K+] - [Cl- + HCO3-] (normal <12 mmol)
- lactate (trend data is important)
- liver function tests
- creatine kinase
- urinary/plasma ketones

Initial management

If a metabolic disease is suspected, the case should be discussed with the duty GOSH consultant in metabolic medicine.

Supportive therapy.

- Profound encephalopathy, intractable seizures or apnoea – intubate and provide mechanical ventilation
- Circulatory failure - intravascular volume expansion ± inotrope therapy
- Stop oral feeds and give 10% dextrose ± electrolytes
- Give dextrose at an infusion rate of at least 9 mg/kg/min to promote anabolic state.
- If hyperglycaemic, may require insulin
- If hypoglycaemic, give dextrose infusion to maintain
- Euglycaemia (may need to use a dextrose concentration >10%, ideally through a central venous catheter)
- Seizures – treat hypoglycaemia, commence APLS protocol anticonvulsants if necessary
- Metabolic acidosis - correct pH using sodium bicarbonate (discuss with metabolic team).
- Hyperammonaemia – consider sodium benzoate, sodium phenylbutyrate for known organic acidaemia. Additionally, consider L-arginine and L-carnitine for a suspected new diagnosis (loading and maintenance doses below). Consider carbaglu. Discuss this management with the metabolic team.

Indications for intubation

- Depressed level of consciousness
- Intractable seizures
- Apnoea
- Severe circulatory failure

Management following intubation

- Sedate and paralyse
- Ensure ETT well secured.
- Attain adequate venous access, consider arterial access, in neonates consider UAC and UVC.
- Give sodium bicarbonate if acidosis intractable and CO2 allows.

Samples to bring with patient (if possible)

- Lithium Heparin (2ml)
- Urine (pre-treatment sample very helpful for diagnosis, will be sent for organic acids)
- Blood spot on Guthrie card if possible

Transport considerations

- Prepare fluid boluses
- Consider preparing infusions of dopamine and adrenaline ready to commence immediately if necessary.
- Be prepared to treat seizures
- Be prepared to administer a glucose bolus for hypoglycaemia

Metabolic Drugs:

- Sodium Benzoate & Sodium Phenylbutyrate **can be** made up together in same syringe if both are the same dose.
- Monitor Na levels.
- Discuss with metabolic team regarding the need for loading dose or infusion.

Sodium Benzoate:
(2g in 10mls), Load: 250mg/kg over 90 mins then 10-20 mg/kg/hr infusion. Dilute 2g in 40mls with 10% glucose (max concentration).

Sodium Phenylbutyrate:
(2g in 10mls), Load: 250mg/kg over 90 mins then 10-20 mg/kg/hr infusion. Dilute 2g in 40mls with 10% glucose (max concentration).

Carnitine:
(1g in 5mls), Load: 100mg/kg over 30 mins, then 4mg/kg/hr infusion. Dilute 400mg in 50mls with 10% glucose.

Arginine:
(5g in 10ml), Load: 300mg/kg over 90 mins, then 12.5mg/kg/hr infusion. Dilute with 10% glucose, max concentration 50mg/ml.

British Inherited Metabolic Disease Group(BIMDG) website and guidelines:

www.bimdg.org.uk
Use 'Emergency Guides' on the red tab