NHS Children's Acute Transport Service

Clinical Guideline

CATS Metabolic Referrals

Document Control Information

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Document Version	1.3	**Replaces Version**	1.2

First Introduced	2001	**Review Schedule**	Annually
Active Date	January 2016	**Next Review**	January 2018

CATS Document Number	22072012-2
Applicable to	**All CATS employees**

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

Known metabolic conditions should be discussed with the duty GOSH metabolic consultant.

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
- 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
- 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
- urinary reducing substances
- 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.
- Hyperammonaemia – consider sodium benzoate, sodium phenylbutyrate or for known organic acidaemia consider carbaglu with the metabolic consultant

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.

Samples to bring with patient (if possible)

- Plasma (2ml)
- Lithium Heparin (2ml)
- Urine

Transport considerations

- Prepare fluid boluses
- Consider preparing infusions of dopamine and adrenaline ready to commence immediately if necessary.

Metabolic Drugs:

Metabolic Drugs: Sodium Benzoate & Sodium Phenylbutyrate **can be** made up together in same syringe if both are the same dose. Montior 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 conc).

Sodium Phenylbutyrate:
(1g in 5mls), Load: 250mg/kg over 90 mins then 10-20 mg/kg/hr infusion. Dilute 2g in 40mls with 10% glucose (max conc).

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

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

http://www.bimdg.org.uk/protocols/topics.asp