Clinical Guideline

WATCh – MANAGEMENT OF SUSPECTED METABOLIC EMERGENCIES

SETTING	Wales and West Acute Transport for Children (WATCh)
FOR STAFF	WATCh Team, South West and Wales District General Hospital medical and nursing teams.
PATIENTS	Children presenting with a suspected metabolic emergency

GUIDANCE

This guidance offers advice for the WATCh team and DGH staff treating children with a suspected metabolic emergency. A summary guideline is on page 2 and is available on the WATCh website (www.watch.nhs.uk)

Glossary:

IEM – Inborn Error of Metabolism
PVC – Peripheral Venous Cannula
NAGS – N-acetylglutamate synthase

RELATED DOCUMENTS	WATCh Clinical Guideline – Management of the Collapsed Neonate BRHC Clinical Guideline – Hyperammonaemia-Emergency Management of Undiagnosed Hyperammonaemia
AUTHORISING BODY	WATCh Governance Group
SAFETY	**Do not give L-carnitine if there is cardiomyopathy or cardiac arrhythmia or long chain fatty acid disorder suspected** Call the WATCh team for advice and support
QUERIES	0300 0300 789

MANAGEMENT OF SUSPECTED METABOLIC EMERGENCIES

METABOLIC EMERGENCIES	SIGNS AND SYMPTOMS
• Acute metabolic decompensation requires prompt recognition and intervention to prevent mortality and long-term morbidity. • Disorders of intermediary metabolism can present with acute, life-threatening illness, particularly organic acidurias, urea cycle disorders, maple syrup urine disease, and fatty acid oxidation disorders. • Neurotransmitter defects and related disorders can present with severe metabolic encephalopathy • If there is a suspicion of a metabolic disorder request an urgent glucose, lactate, blood gas and ammonia. **Contact WATCh and the duty Metabolic Consultant**	**Age:** Newborn to teenagers. Onset and severity may be influenced by changes in dietary intake, fasting, dehydration, intercurrent illness, medications, strenuous activity, childbirth, trauma, or surgery. **Neurological Signs:** Lethargy, coma, seizures, developmental delay or regression, peripheral neuropathy, abnormalities of tone, motor problems, ataxia. **Gastrointestinal:** Recurrent episodes of vomiting or dehydration, poor feeding, failure to thrive, decreased gastrointestinal motility, hepatomegaly or hepatosplenomegaly, and jaundice. **Cardiomyopathy:** Hypertrophic or dilated cardiomyopathy may occur in several IEMs and is typically related to impaired energy metabolism or storage material **Other:** Ophthalmologic, Dermatologic, Abnormal Odours and Dysmorphic Features

HISTORY AND EXAMINATION	DIFFERENTIAL DIAGNOSIS
• Neonatal history • Diet and last time of feed • Possibility of intoxication (e.g. alcohol) • Recent/current illness • Past history of unexplained seizures • Family history of unexpected or unexplained deaths • Parental consanguinity • Liver size • Association with lactic acidosis or hyper / hypo ketosis	• Sepsis • Encephalitis • Non-accidental injury • Urea cycle disorders • Amino acid disorders • Fatty acid oxidation disorders • Carbohydrate disorders • Mitochondrial disorders

IMMEDIATE MANAGEMENT

- **Respiratory Depression** - Respiratory acidosis should be managed with Intubation and Ventilation
- **Shock** - Fluid resuscitation with 10-20mL/kg, 0.9% Sodium Chloride or Plasmalyte 148, if shock persists after 40 mL/kg consider inotropic support. Antibiotic cover for sepsis.
- **Seizures** - Treat as per APLS guidelines, regular neurological observations and monitor serum sodium. Antibiotic cover for bacterial / viral meningitis. Consider CT Head

  Hyperammonaemia – **Seek urgent advice from WATCh and the duty Metabolic Consultant**. Start 10% Glucose at full maintenance rates. Aim for glucose 4-8mmol/l. Add insulin infusion 0.05units/kg/h if BM >10mmol/l in preference to cutting dextrose. Monitor BM hourly and adjust rate of insulin accordingly.
  - Ammonia <200 – Stop protein and repeat level.
  - Ammonia 200-350 – As above and treat with Sodium Benzoate, Sodium Phenylbutyrate, L Arginine and L Carnitine.
  - Ammonia >350 – As above plus CVVH
- **Metabolic Acidosis** - Bicarbonate should be administered with caution.
- **Liver Dysfunction** - Treat coagulopathy with Fresh Frozen Plasma
- **Lactic Acidosis** - Fluid resuscitation, inotropic support, reduce metabolic demand (aim anabolic state). Cover for sepsis.
- **Hypoglycaemia** - Treat with 2mL/kg 10% Glucose, blood samples for evaluation of metabolic causes of hypoglycaemia should be obtained before treatment. Normal glucose requirement is 6-8mg/kg/min. Intake can be increased to 12-15mg/kg/min (max 12.5% Glucose via PVC). Glucose Intake/Requirement (mg/kg/min) = $\frac{\%\text{glucose X rate (ml/hr)}}{\text{weight (kg) X 6}}$

MEDICATIONS

Drug	Loading dose over 90mins	Followed by maintenance dose over 24hrs	Max daily dose (every 24hrs there after)
Sodium Benzoate	250mg/kg	250mg/kg	500mg/kg
Sodium Phenylbutyrate	250mg/kg	250mg/kg	600mg/kg
L-Arginine	150mg/kg	300mg/kg	500mg/kg Higher doses can be given in discussion with the **Metabolic Team**
L-Carnitine (See safety note)	100mg/kg	100mg/kg or 100 - 200mg/kg in 4 divided doses may also be given	300mg/kg
Carglumic acid	Consider if suspect NAGS deficiency or organic acidaemia		250mg/kg stat dose enterally