PAEDIATRIC ACUTE CARE GUIDELINE	
Fatty Acid Oxidation Disorders	
Scope (Staff):	All Emergency Department Clinicians
Scope (Area):	Emergency Department

Fatty Acid Oxidation Disorders

Contact the Metabolic Consultant and the Emergency Department Consultant urgently if the patient is unwell.

Background

- If a child with a fatty oxidation disorder: MCAD, LCHAD, VLCAD or CPT-1 deficiency presents to hospital, please ensure they are assessed immediately and emergency treatment commenced
- It is dangerous to base one's treatment on monitoring of blood glucose levels
- All patients must be discussed with the Metabolic Team Consultant at the earliest opportunity

General

Fatty Acid Oxidation Disorders:

- These are all inherited disorders of fat breakdown
  - MCAD = Medium chain acyl coenzyme A dehydrogenase deficiency
  - LCHAD = Long chain hydroxy acyl coenzyme A dehydrogenase deficiency
  - VLCAD = Very long-chain acyl coenzyme A dehydrogenase deficiency
  - CPT-1 = carnitine palmitoytransferase-1 deficiency
- Most of the time these children are healthy, but they can become unwell and unstable at times of illness

- Precipitants such as infection, fasting or vomiting episodes can lead to serious illness rapidly, with encephalopathy and rhabdomyolysis. This results from the accumulation of toxic fatty acids.
- Hypoglycaemia also may occur, but only at a relatively late stage
- The treatment of acute illness in these children aims to inhibit the mobilisation of fat stores by providing ample glucose substrate – either enterally or intravenously

Investigations

- All children with inherited metabolic disorders should have a capillary or venous blood gas at time of assessment (includes BGL, lactate) - if this is not possible, a bedside BGL must be done at a minimum
- Other investigations to consider depending on the clinical situation:
    - U&E
    - Ammonia (NH3) - remember this must be on ice
    - CK level and check urine for myoglobinuria in LCHAD and VLAD

Initial management

- Ascertain the reason why the child has been brought to hospital (vomiting, refusing drinks etc) and assess the patient rapidly
- If the problem is refusal of drinks/medicines or a single vomit, and there is no clinical suggestion of incipient encephalopathy (such as glazed look, lethargy or drowsiness), the child can be offered another high carbohydrate drink orally as per their emergency regime (e.g. lucozade or polyjoule) or via a nasogastric tube.
- The high carbohydrate fluid available at PMH is CarbPlus, which can be found in the Infant Formula Room
- If the child can be discharged home if they:
    - Tolerate the carbohydrate drinks
    - Has been observed for at least 2 hours
    - Looks well
    - Has been discussed with the Metabolic Team Consultant (Shanti Balasubramaniam, Adelaide Withers, Emily Boulter)

Further management

- If the child has further vomits or looks unwell:
    - Insert an IV cannula
    - Do a venous blood gas (if not already done)
    - Start 10% glucose infusion at 5mL/kg/hr immediately
    - Contact the Metabolic Team Consultant (as above)

Carnitine:

- For children with MCAD only, commence IV Carnitine at 30mg/kg 8 hourly
- For children with LCHAD and VLCAD the use of Carnitine during acute illness is controversial; please discuss with the Metabolic Team Consultant

Blood Glucose Level Monitoring:

- Despite its limitations, monitor the BGL 4 hourly initially if the child's condition is stable
- If unstable, monitor BGL hourly and U&Es and VBG 6 hourly

Admission criteria

- There is a low threshold for admission for children with inherited metabolic disorders.

Referrals and follow-up

- At the earliest opportunity please contact the Metabolic Team Consultant on call
- If they are not available, contact the General Paediatric Consultant on call

Nursing

- All metabolic patients are to be triaged as a Category 2
- Baseline observations: heart rate, respiratory rate, temperature, Sp02, capillary refill, BP and neurological observations
    - Monitor for signs of encephalophathy (glazed look, lethargy, drowsiness)
- Minimum of hourly observations should be recorded whilst in the emergency department
    - Any significant changes should be reported immediately to the medical team
- Hourly fluid input/output
- Investigations should include:
    - Bloods (without Emla® if unwell)
    - BGL (on arrival) and as directed by the medical team
    - Urine dipstick for ketones

Tags

breakdown, cpt-1, fat, fatty acid, fatty acid oxidation disorders, glucose, hypoglycaemia, lchad, mcad, metabolism, vlcad, vomiting

References
W:PharmacyPMHPharmacy PMH General|Clin Section_CLINICAL SERVICES HANDBOOKClinical Services Guidelines|Fatemulsion|PMH.Pharm.FatEmulsionUsageGuideline.docx Document Owner - L. Bint (Chief Pharmacist, last revised Jan 2014 http://cahs.hdwa.health.wa.go.au/_data/assets/pdf_file/0003/80859/Prep_IV_arg,_Carn,_Sod_Benz,_Sod_Phen_final_june2009.pdf PMH Pharm Hyperammonaemia Drugs Guideline doc -S Kassam

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