BIMDG
British Inherited Metabolic Disease Group

This protocol has 3 pages

MEDIUM CHAIN ACYL COA DEHYDROGENASE DEFICIENCY (MCADD) – ACUTE ILLNESS / DECOMPENSATION

IMMEDIATE ACTION

- **Triage to high priority**
- **Hypoglycaemia occurs late – do not delay treatment because blood glucose is not low**
- **Management should be based upon clinical status as per Section 2.**
- **Run IV 10% Dextrose/0.45% Sodium Chloride at 5ml/kg/hr ONLY until fluids can be accurately calculated**
- **These guidelines cover the first 24hr of management only – ongoing management should be guided by the child's specialist metabolic team. Inform them EARLY about the child**

1. Background and Signs of Decompensation

- MCAD deficiency is the most common disorder of fat catabolism. The clinical manifestations occur due to energy deficit during fasting, mainly during acute illnesses with reduced intake.
- Infections, fasting, diarrhoea or vomiting can lead to serious illness, with encephalopathy and even sudden death.
- This results from the accumulation of toxic fatty acids.
- Hypoglycaemia only occurs at a late stage. The aim should always be to intervene whilst the blood glucose is normal.
- The early signs of decompensation may be subtle e.g. lethargy or 'floppiness'
- Treatment aims to prevent catabolism by providing energy in the form of glucose - enterally or intravenously (see section 2)

2.Management in hospital

- If the child is shocked or clearly very ill consider admission to ITU/High dependency.
- If admitted to a metabolic/general ward careful clinical assessment is essential including regular PEWS and neurological observations even if the patient does not appear encephalopathic.

The following blood tests should be considered:
- pH and gases
- Glucose (laboratory and bedside strip test)
- Urea and electrolytes
- Full blood count

Consider other tests as clinically indicated.

Management decisions should be based primarily on the **clinical** status. The first decision about therapy is whether the child can be treated orally or will need intravenous therapy. Intravenous fluids are indicated if:

1. the child is unable to tolerate oral fluids, or
2. there is moderate or severe clinical dehydration

- **If there is any doubt at all, put up an intravenous line.**
- Treat any infection

A. ORAL. If the child is relatively well and not vomiting, oral feeds may be given. For young children (typically under 2 years) or those who already have enteral feeding tubes, the emergency feed can be given via such tubes.

FULL ENTERAL EMERGENCY FEED – glucose polymer solution

Use patient's own ER recipe wherever possible.
Use age-based ER recipes below if not available.
If ER products not available use IV guidelines.

NB: MCT feeds and supplements contraindicated in MCADD
Oral rehydration solutions are low in CHO and not suitable

- Click Here for Emergency Regimen for Age ≤ 1 year (10%)
- Click Here for Emergency Regimen for Age 1- 2 years (15%)
- Click Here for Emergency Regimen for Age 2-9 years (20%)
- Click Here for Emergency Regimen for Age ≥ 10 years (25%)

EMERGENCY FEED ADMINISTRATION

- Give feeding volume for body weight (see recipe)
- Feed orally: 2 hourly day and night
- If not tolerated or fluid requirements not met, administer continuously by tube, without delay
- Administer bolus or continuously by tube feed, without delay for a maximum of 24-36hours
- Introduce usual diet/feeds as soon as clinically stable

Medications

- Antipyretics: as clinically indicated

Contact the child's specialist metabolic team and dietitian for further advice on the ER and introduction of usual diet/feeds

B. INTRAVENOUS. If the child is unwell and/or vomiting then IV treatment is needed:

IMMEDIATE FLUID RESUSCITATION:

- Give Glucose 200 mg/kg **at once if there is definite hypoglycaemia** (2ml/kg of 10% glucose or 1ml/kg of 20% glucose over a few minutes if blood glucose <3.0mM)

- Give 0.9% sodium chloride 20 ml/kg as a bolus **if the peripheral circulation is poor or the patient is frankly shocked**. Repeat the sodium chloride bolus if the poor circulation persists as for a shocked non-metabolic patient.

INITIAL FLUIDS AFTER RESUSCITATION:

- Run IV fluids of Glucose 10%/Sodium Chloride 0.45% at 5ml/kg/h ONLY until accurate fluid rates have been calculated **– do not leave on this high rate longer than necessary**. (for instructions to make this solution click here).

FURTHER FLUID MANAGEMENT IN FIRST 24 HOURS:

- Ongoing fluid management in based upon administering the fluid deficit plus maintenance over 24 hours as Glucose 10%/Sodium Chloride 0.45%.
- Deduct any fluid already given from the total for the first 24 hours.
- Potassium can be added once the plasma potassium concentration is known and the child is passing urine.
- Reassess hydration status and the need for ongoing IV fluids after 24 hours and if needed recheck the electrolytes every 24 hours.

3. Progress/Monitoring: Reassess after 4-6 hours or earlier if there is any deterioration or no improvement. Clinical assessment should include PEWS and neurological observations.

- If deteriorating, seek specialist help without delay.

4. Re-introduction of oral feeds:

- Intravenous fluids should not be stopped abruptly.
- Consider halving IV fluids for a few hours before stopping .
- IV fluids can be stopped once it becomes clear that oral feeds are being tolerated.
- For more information please refer to the MCADD dietary guidelines or consult your metabolic dietitian.
- Only allow the child home if you and the family are entirely happy. **It must be clearly demonstrated that the child can tolerate at least two successive feeds / meals before discharge.** The family must have a clear management plan and be prepared to return if the child deteriorates.

For further information please refer to:

Merritt JL 2nd, Chang IJ. Medium-Chain Acyl-Coenzyme A Dehydrogenase Deficiency. 2000 Apr 20 [Updated 2019 Jun 27]. In: Adam MP, Ardinger HH, Pagon RA, et al., editors. GeneReviews® [Internet]. Seattle (WA): University of Washington, Seattle; 1993-2021. Available from: https://www.ncbi.nlm.nih.gov/books/NBK1424/