



## Information for Emergency Departments for infants

This child has medium-chain acyl-CoA dehydrogenase deficiency (MCADD). A child with this condition is at risk of serious illness, coma and death during prolonged fasting, particularly during infectious illnesses. He or she may have a deteriorating level of consciousness, may vomit and may have seizures.

**Hypoglycaemia may occur at a late stage but treatment must not be delayed even if the blood glucose is normal.**

Treatment aims to inhibit mobilisation of fat by providing ample glucose. During infections, parents will use an emergency regimen (ER) of frequent glucose polymer drinks but if this is not tolerated or there is clinical deterioration they have been instructed to attend hospital urgently for further management.

### Brief guide to MCADD hospital management

#### Assess

- Level of consciousness – record Glasgow coma score as modified for children
- Unresponsiveness, drowsiness, or irritability = encephalopathy
- U&E, Blood Gases, Glucose (near-patient testing + laboratory measurement), other tests as indicated

#### If not encephalopathic

- Oral or nasogastric Emergency Regimen until able to tolerate normal diet (formula feeds or breast milk and age-appropriate solids), blood glucose is within the normal range and clinically recovered and stable.

Note: Oral rehydration solutions do not contain sufficient glucose to avoid decompensation and therefore, if prescribed, must be fortified with glucose polymer – see the MCADD Dietary Guidelines, available at [www.imd.scot.nhs.uk](http://www.imd.scot.nhs.uk)

#### If encephalopathic or vomiting or diarrhoea or blood glucose $\leq 3.0$ mmol/l

1. IV 10% glucose bolus 2ml/kg (200 mg/kg)
2. If poor circulation / shock, follow with 20ml/kg 0.9% sodium chloride
3. Whilst the maintenance fluid is being made up, continue 10% glucose at 5ml/kg/hr
4. Maintenance fluid given as 10% glucose with 0.45% sodium chloride.

This solution can be made up as follows:

Remove and discard 50ml from a 500 ml bag of 0.45% sodium chloride & 5% glucose solution, then add 50ml of 50% glucose to the fluid remaining in the bag

OR

Add 7.5ml 30% (5 mmol/ml) NaCl solution to a 500ml bag of 10% glucose solution

5. Correct any fluid / electrolyte deficits; add potassium once U&E status is known
6. Admit & notify the on-call local consultant paediatrician and, if the child is very ill, the regional paediatric metabolic service (contact details below)
7. Monitor blood glucose 4 hourly during acute phase
8. Adjust IV infusion rate to maintain blood glucose 4-8 mmol/l
9. Continue infusion until blood glucose stable and tolerating usual oral feeds

**If this child is admitted, please notify the local paediatric consultant looking after this patient and, if very ill, the regional metabolic service (next page)**

## EMERGENCY CONTACT DETAILS

### DAYTIME:

*[Relevant Regional Metabolic Team]*

#### **Glasgow**

**Dr Peter Robinson, Consultant, Paediatric Inherited Metabolic Disease**

**Dr Bernd Schwahn, Consultant, Paediatric Inherited Metabolic Disease**

**07699 646865 (aircall for the on-call consultant)**

or

**0141 201 0000 (switchboard) and ask for page 8500 (Dr Robinson) or page 8341 (Dr Schwahn)**

#### **Edinburgh**

**Dr Alison Cozens, Consultant Paediatrician in Inherited Metabolic Disease**

**07895 713600**

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### EVENINGS and WEEKENDS and PUBLIC HOLIDAYS

There is a consultant in paediatric metabolic medicine on call at all times. Switchboards at RHSC Edinburgh and Glasgow will have the on-call rota and contact details



The aims of the National Managed Clinical Network are to improve diagnosis, treatment and support for all individuals and families affected by Inherited Metabolic Disorders, wherever in Scotland they live.