British Inherited Metabolic Disease Group

Contact Details Name:
Hospital
Telephone:

MANAGEMENT OF SURGERY IN CHILDREN WITH MCAD DEFICIENCY

Patients with MCAD deficiency are usually well controlled but there is a risk of decompensation during surgery, particularly if catabolism is precipitated by fasting and surgery. It is important to follow an appropriate protocol, minimising catabolism by providing adequate amounts of carbohydrate. This protocol should be used in conjunction with the emergency regimens on the BIMDG website.

PRE-OPERATIVE MANAGEMENT - INTRAVENOUS THERAPY

If this is a routine procedure, check that the child is healthy. If the is unwell, postpone the operation. Emergency operations and major procedures (lasting longer than about 30 minutes) require special consideration: seek specialist advice.

By the time the operation starts the child will need to be receiving intravenous 10% glucose/ 0.45% saline (for instructions to make this solution click here). at the rate given by the formula below.

For patients with MCAD deficiency, it is generally satisfactory to start the infusion on the morning of the procedure if on a morning list (or from lunchtime if on an afternoon list).

Formula for calculating for peri-operative intravenous therapy :

Suitable rates for 10% glucose 0.45% saline (for instructions to make this solution click here).
Fluid/24 hours = 100ml/kg for 1st 10kg then 50 ml/kg for next 10kg then 20ml/kg thereafter.
Potassium should be added to this solution 10 mmol in 500 ml.

If cannulation is difficult or the child is likely to pull out the cannula before getting to theatre, it may be possible to postpone insertion of the cannula until after induction of anaesthesia. It may still be necessary, however, to start the infusion before anaesthesia if the surgery is delayed.

The intravenous infusion must continue throughout the operation.

PRE-OPERATIVE ORAL MANAGEMENT

The exact arrangements will depend on the timing of the surgery/anaesthesia and the views of the anaesthetist.

(i) Is the child is late enough on the list to allow breakfast?
Generally a light breakfast is given to children >6 hrs before their minor operations. Thus, children whose operations are scheduled for 12.00 or later will generally be given breakfast, but a parent may tell you that their child is very unlikely to take breakfast before a certain hour, which should be taken into consideration.

(ii) Pre-operative glucose polymer
Provided the anaesthetist agrees, a drink of glucose polymer should be given to patients 3 hrs pre-operatively unless an infusion of 10% glucose has already been started. Suitable volumes and concentrations are given in the table below. Contact your local dietitian for these solutions – details can also be found here.

Ask the child's carer how they normally take glucose polymer in the emergency regimen: they may take it with flavouring for example. If the child appears unwell, cannot be persuaded to take the glucose polymer or it is vomited or if the operation is delayed an intravenous 10% glucose infusion should be started before the anaesthetic.

Table : Pre-operative drinks: Suitable doses & concentrations of glucose polymer[§]

Age (yrs)	Concentration (%)	Volume
0-1	10	14 ml/kg
1-2	15	8 ml/kg
2-6	20	100 ml
6-10	20	150 ml
>10	25	180 ml

DELAYED OPERATION

If the operation is delayed, the glucose infusion should be started at the time the operation was due to begin. **Important Note:** hypoglycaemia is a late event in these disorders and blood glucose should not be used to monitor them.

POST-OPERATIVE PROCEDURE

Feed the child at the time you would feed any other child following an equivalent procedure. Discontinue the intravenous infusion ONLY after the child has been seen to tolerate food. Remove the cannula ONLY when there is no chance of the child vomiting. Seek specialist help if there are any problems.

Discharge the child ONLY when absolutely sure they have fully recovered and they have been discussed with the metabolic team. This will often be the following day.

Date last reviewed April 2017