BIMDG

British Inherited Metabolic Disease Group

Contact Details Name:
Hospital
Telephone:

MANAGEMENT OF SURGERY IN CHILDREN WITH DISORDERS OF FATTY ACID OXIDATION

Patients with disorders of fatty acid oxidation are usually well controlled but can easily decompensate during surgery, particularly if catabolism is precipitated by fasting and surgery. Elective surgery in these patients is usually best done at the hospital with the regional metabolic unit. 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.

The following instructions apply to patients with:

- **Very long chain acyl CoA dehydrogenase deficiency (VLCADD**
- **Long chain 3-hydroxy-acyl CoA dehydrogenase deficiency (LCHADD)**
- **Multiple acyl CoA dehydrogenase deficiency (MADD)**
- **Carnitine palmitoyl transferase deficiency I & II**
- **And others**

A separate, simpler guideline is available for patients with MCAD deficiency – click here.

1. PRE-OPERATIVE MANAGEMENT - INTRAVENOUS THERAPY

If this is a routine procedure, check that the child is healthy. If the child 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 given below.

It is simplest if the operation is first on an afternoon list. The anaesthetist will probably then allow them to follow their usual overnight management, with an early breakfast and a drink containing glucose polymer 3-4 hours pre-op (concentration and volume as in their emergency regimen). Otherwise it may be safest to start the intravenous infusion at the beginning of the pre-operative fast.

If the surgery is first thing in the morning, it may be necessary to start the infusion the previous night: discuss with the metabolic consultant.

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. However, this depends on the child being able to fast for at least as long as the anaesthetist's minimum pre-operative fasting interval. This is likely to be true for most metabolic disorders but a few may not tolerate this. This management strategy is easiest if the operation is towards the end of the morning list as this allows the usual overnight/morning routine to be followed. If the child is scheduled to have his operation early on the list, the parents would have to persuade the child to take a drink containing glucose polymer in the early hours of the morning. Moreover, it will still be necessary 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 not just on the metabolic disorders but also 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 or via a nasogastric tube. If the child appears unwell, cannot be persuaded to take the glucose polymer or it is vomited or if the operation is delayed, such that the anaesthetic will start more than 4 hrs after the glucose polymer, an intravenous 10% glucose infusion must 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.

EXTRA INTRA-OPERATIVE INSTRUCTIONS

Does the child have regular nasogastric tube feeds (eg for overnight feeding)?

If so, make sure that the surgeons leave one in situ at the end of the operation, particularly if this was an ENT procedure.

POST-OPERATIVE PROCEDURE

The only medication that is essential is carnitine in carnitine transporter deficiency. This can be given orally or intravenously. For more details see BIMDG emergency protocol for carnitine transporter deficiency.

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 or enteral feeds. 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.

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