BIMDG

British Inherited Metabolic Disease Group

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MANAGEMENT OF SURGERY IN CHILDREN WITH AN ORGANIC ACIDAEMIA AND MSUD

Patients with organic acidaemias who are usually well controlled can easily decompensate during surgery, precipitated by a combination of stress and fasting. 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 that are also available on the BIMDG website.

The following instructions apply to patients with:
Methylmalonic acidaemia, Propionic acidaemia, Isovaleric acidaemia
Glutaric acidaemia type I, Maple syrup urine disease & other less common disorders.

IMPORTANT - ask if unsure

PRE ANAESTHETIC MANAGEMENT

If this is a routine procedure, check that the child is healthy. If he is not, postpone the operation. Emergency operations and major procedures (lasting longer than about 30 minutes) require special consideration: seek specialist advice.

a) For organic acidaemias other than Maple Syrup Urine Disease (MSUD):
On admission, check the plasma ammonia and pH and blood gases. If the plasma ammonia is >100 μmol/l, there is frank acidosis (pH<7.30 or base deficit > 10 mmol/l) or the child is unwell, cancel elective procedures & seek specialist advice.

b) For MSUD:
If the child is unwell, cancel elective procedures & contact the metabolic team. Otherwise check quantitative plasma amino acids (other tests are unhelpful in this condition). The results will not be available pre-operatively but serve as useful baseline.

Check the usual feeding regimen & drug dosages with the parents & the dietitian. Many of these patients are on overnight naso-gastric feeds.

INTRAVENOUS THERAPY

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 Pre and post-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 except patients with methylmalonic acidaemia (MMA). For these, only add potassium after plasma electrolytes have been checked.

If cannulation is difficult or the child is likely to pull out the cannula before getting to theatres, 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 will 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.

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.

Aminoacid supplements: Patients with MSUD/Glutaric Aciduria Type 1
A dose of amino acid supplement should be given preoperatively. Ideally, one third of the child's usual daily dose should be given with a light breakfast, 6 hours prior to surgery. If this is not possible, 0.25 g/kg should be added to the glucose polymer drink given 3 hours before surgery. For children aged < 1 year, the usual amino acid supplement comes in milk and so cannot be added to the glucose polymer drink: discuss with the dietician, who will supply a pure amino acid preparation, suitable for adding to glucose polymer.

Pre-operative glucose polymer

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 the drink in his emergency regimen: they may take it with flavouring or via a naso-gastric tube. If the child 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 drink:

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

4. 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.

5. POST-OPERATIVE PROCEDURE

Following the operation, feed the child (with their usual diet) at the time you would feed any other child following an equivalent procedure. Give whatever oral medicines he may be due at the same time. Patients with MSUD should receive their usual amino acid supplements (less whatever was given pre-operatively). 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.

If recovery is delayed or complicated by vomiting, seek specialist advice as intravenous medicines may be required. Check blood gases, electrolytes and ammonia as extra electrolytes or bicarbonate may be needed. Consult appropriate emergency regimen on BIMDG website.

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