



British Inherited Metabolic Disease Group

**Contact Details Name:**

**Hospital**

**Telephone:**

- ..... has **MAPLE SYRUP URINE DISEASE**
- **Please read carefully. ASSESSMENT AND TREATMENT ARE URGENT.** Treatment should be meticulous as there is a high risk of serious complications.
- The major complication is ENCEPHALOPATHY.
- **Start this treatment** if the patient is obviously unwell or vomiting, drowsy or ataxic. Do not delay if you are uncertain.
- **Give Glucose 200 mg/kg at once** (2 ml/kg of 10% glucose or 1ml/kg of 20% glucose) over a few minutes.
- **Give normal saline 10 ml/kg** unless the peripheral circulation is poor or the patient is frankly shocked, then give 20 ml/kg normal saline as a bolus immediately after the glucose. Repeat the saline bolus if the poor circulation persists as for a shocked non-metabolic patient.
- **Continue with glucose 10% / saline 0.45% at 5 ml/kg/h ONLY UNTIL THE NEXT SOLUTION IS READY AND AN ACCURATE INFUSION RATE HAS BEEN CALCULATED – DO NOT LEAVE ON HIGH INFUSION RATES FOR TOO LONG**
- If this is not immediately available, continue with glucose 10% until it is ready. (For instructions to make glucose 10% / saline 0.45% solution [click here](#))
- Call paediatrician for continuing management. **Do not delay. If paediatrician not immediately available please refer to additional instructions on the next page or the BIMDG website.**
- If there is any doubt at all, the child must be admitted, even if only necessary for a short period of observation.
- *This protocol is for the immediate management only.*

More information can be found in the [BIMDG standard emergency guideline for Maple Syrup Urine Disease \(Click Here\)](#)

*Continuing Intravenous management: Additional instructions*

- Quickly calculate the deficit and maintenance and prepare the intravenous fluids
  - Deficit: estimate from clinical signs if no recent weight available
  - Maintenance: Formula for calculating daily maintenance fluid volume (BNF for children) 100ml/kg for 1<sup>st</sup> 10kg then 50 ml/kg for next 10kg then 20ml/kg thereafter, using calculated rehydrated weight. Deduct the fluid already given from the total for the first 24 hours.
  - Give 0.45% saline/10% glucose ([for instructions to make this solution click here](#)).
- Having calculated the deficit and the maintenance, administer the appropriate rate of 0.45% saline/10% glucose to correct the deficit within 24 hours.
- Recheck the electrolytes every 24 hours if still on IV fluids.

If possible, a concentrated amino acid preparation should still be run whilst the child is receiving intravenous fluids.

[Click here for information about the enteral amino acid regimen for a child with maple syrup urine disease receiving intravenous fluids.](#)