BIMDG

British Inherited Metabolic Disease Group

Contact Details Name:
Hospital
Telephone:

- …… ……has **a DISORDER OF THE UREA CYCLE** *(complete disorder)*
- **Please read carefully.** ASSESSMENT AND TREATMENT ARE URGENT. Treatment should be meticulous as there is a high risk of serious complications.
- The major complications are hyperammonaemia and encephalopathy.
- **Start this treatment** if the patient is obviously unwell, vomiting, drowsy, uncooperative or is behaving oddly. Do not delay if you are uncertain.
- Insert intravenous cannula and send blood (heparinised tube) for plasma ammonia urgently
- **Give Glucose 200 mg/kg intravenously 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%** at 5 ml/kg/h **ONLY UNTIL THE NEXT SOLUTION IS READY – DO NOT LEAVE ON HIGH INFUSION RATES FOR TOO LONG**
- **Call Paediatrician on duty** as patients need specialised medication URGENTLY. DO NOT DELAY
- Call pharmacist and request intravenous preparations of sodium benzoate, sodium phenylbutyrate and arginine (Note: some parents carry supplies of these)
- 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 guidelines for hyperammonaemia and urea cycle disorders; UCD 1 for OTC and CPS deficiencies, UCD 2 for citrullinaemia and argininosuccinic aciduria, and a separate one for NAGS deficiency.

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