BIMDG

British Inherited Metabolic Disease Group

Contact Details Name:
Hospital
Telephone:

…… has **CPT1 DEFICIENCY OR HMG COA SYNTHASE DEFICIENCY**

- **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 encephalopathy and hypoglycaemia but this is a late complication.
- **Start this treatment** if the patient is obviously unwell or vomiting, hypoglycaemic (blood glucose <2.6 mmol/l) or drowsy. 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, even if not hypoglycaemic.
- **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)
- If there is any doubt at all, the child must be admitted, even if only necessary for a short period of observation.
- Inform the child's metabolic team.
- *This protocol is for the immediate management only.*

More information can be found in the BIMDG standard emergency guideline for CPT1 or HMG CoA Synthase deficiency

Last Revised January 2017