



British Inherited Metabolic Disease Group

**Contact Details Name:**

**Hospital**

**Telephone:**

- ..... has **GLUT1 DEFICIENCY**
- Please read carefully. **ASSESSMENT IS URGENT**. Meticulous treatment is important as there is a high risk of serious complications.
- **Important note: The management of illness in GLUT1 deficiency is quite different from other metabolic disorders.**
- The major acute complications are fits and the patients are treated with a ketogenic diet.
- **If fitting**, check blood glucose (laboratory and bedside strip test) and urine ketones.
  - If blood glucose < 2 mmol/l and urine ketones > 2 mmol/l consider giving anticonvulsants (carbamazepine or phenytoin but not phenobarbitone or diazepam).
  - If blood glucose < 2 mmol/l and urine ketones < 2 mmol/l consider giving glucose orally or intravenously 200mg/kg.
- **Give normal saline 10 ml/kg** unless the peripheral circulation is poor or the patient is frankly shocked, and 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 normal saline at 5 ml/kg/h UNTIL AN ACCURATE INFUSION RATE HAS BEEN CALCULATED – DO NOT LEAVE ON HIGH INFUSION RATES FOR TOO LONG**
- 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 BIMDVG standard emergency guideline for GLUT1 deficiency.