



British Inherited Metabolic Disease Group

**Contact Details Name:**

**Hospital**

**Telephone:**

- ..... has **FRUCTOSE 1,6 BISP HOSPHATASE 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 HYPOGLYCAEMIA and LACTIC ACIDOSIS. These are precipitated by fasting and intercurrent illness.
- **Start this treatment** if the patient is obviously unwell, drowsy or vomiting, hypoglycaemic (blood glucose <2.6 mmol/l), acidotic or hyperventilating (base deficit >10 mmol/l). Do not delay if you are uncertain.
- If unwell, **give glucose immediately** either intravenously (Glucose 200 mg/kg (= 2 ml/kg of 10% glucose or 1ml/kg of 20% glucose) over a few minutes. or into the buccal cavity using Glucogel ®. If an intravenous line cannot be established quickly, pass a naso-gastric tube to continue to give glucose.
- **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 the child deteriorates, check for rebound hypoglycaemia and call paediatrician urgently.
- 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 Fructose-1,6-bisphosphatase deficiency.