British Inherited Metabolic Disease Group

Contact Details Name:
Hospital
Telephone:

…… has GLYCOGEN STORAGE DISEASE TYPE 1A/1B

- **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. Those with Glycogen Storage Disease Type1b have a neutrophil defect that predisposes to serious bacterial infections.

- **Start this treatment** if the patient is obviously unwell or vomiting, hypoglycaemic (blood glucose <2.6 mmol/l), acidotic or hyperventilating (base deficit >10 mmol/l), drowsy or a history of a convulsion. In GSD type 1b start treatment if obvious signs of infection. Do not delay if you are uncertain.

- **Give glucose immediately** either intravenously (Glucose 200 mg/kg at once (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 guidelines for glycogen storage disease type 1A and type 1B

Last Revised September 2016