

NATIONAL CENTRE FOR INHERITED METABOLIC DISORDERS

EMERGENCY LETTER FOR HOSPITAL – Glycogen Storage Disorder Type VI and IX

Addressograph

AT RISK OF HYPOGLYCAEMIA
Please attend to this patient without delay
Always contact the NCIMD for further advice

Glycogen storage disorders (GSDs) are a group of metabolic disorders affecting either the synthesis or breakdown of glycogen in the body, which is crucial for glucose homeostasis. There are many different types of GSD, a number of which affect the liver and therefore predispose to hypoglycaemia. This emergency letter is for the preliminary management of patients with GSD Type VI and Type IX, who may present with ketotic hypoglycaemia.

Maximum fasting time (well) - _____ hours (overnight)

Maximum fasting time (unwell) - _____ hours (day and night)

- Check glucose and ketones at point of care immediately on presentation
- If the patient is not tolerating oral intake, commence IV fluids at 100% maintenance rate, using 0.9% NaCl + 10% dextrose + 2mmol/kg/day of potassium chloride (KCl can be added once urine has been passed), adjusted depending on glucose and electrolyte levels
- If blood glucose is <3mmol/l and tolerating oral intake, give a rescue of 5g carbohydrate per 20kg body weight (e.g. with SOS or Lift drink) and recheck glucose in 10-15 minutes. Repeat if glucose still <3mmol/l. Follow with snack containing protein and carbohydrate
- If blood glucose is <3mmol/l and not tolerating oral intake or drowsy, give a bolus of 10% dextrose 2ml/kg IV and recheck glucose in 10-15 minutes. Repeat if glucose still <3mmol/l. Follow with IV maintenance fluids as described above
- Use of glucagon is contraindicated
- Do not stop IV fluids abruptly – the rate can be weaned gradually once the child has shown that they are tolerating oral intake in a satisfactory quantity
- Co-amoxiclav should be avoided as clavulanic acid is not recommended in GSDs – amoxicillin is safe to use if appropriate

The on call service for the National Centre for Inherited Metabolic Disorders is available to discuss this patient at +353 1 878 4200 via the CHI at Temple Street switchboard (available 24 hours a day). The metabolic ward nurse is also available to discuss at +353 1 878 4200, ask for bleep 836 (available 24 hours a day).