NATIONAL CENTRE FOR INHERITED METABOLIC DISORDERS

EMERGENCY LETTER FOR HOSPITAL – Glycogen Storage Disorder Type III

Addressograph

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

DIAGNOSIS

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 management of GSD Type III, which may present with ketotic hypoglycaemia.

Complications of GSD III include hepatomegaly, hyperlipidaemia, short stature, myopathy and osteopenia. Children with GSD III may also have cardiomyopathy. Patients are managed with reduced fasting times and a diet with regular intake of complex carbohydrate and protein, and low in simple sugars. The use of uncooked cornstarch or a modified cornstarch (Glycosade®) may be indicated. Some children will need enteral tube feeding to meet their requirements.

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

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

NOTES FOR PARENTS/CARERS

Contact the metabolic team at the first signs of illness to discuss your child's care. Metabolic Clinical Nurse Specialist contact: (01) 878 4409 (during working hours).

If you are advised to attend the emergency department, remember to bring your medication, feeding supplies, a copy of your diet plan and a copy of this letter with you.

EMERGENCY MANAGEMENT

Always take illness in a patient with GSD III seriously. Always discuss this patient with the metabolic consultant on call without delay.

- Assess and resuscitate as necessary as per APLS protocols
- Check glucose and ketones at point of care immediately on presentation
- Obtain IV access and send the following bloods to the lab:
    - glucose, ketones, VBG, renal/liver/bone profile, CK
    - additional tests as dictated by presentation (e.g. CRP, urine culture, NPA, etc)
- If blood glucose <3mmol/l and tolerating oral intake, follow the guidance in the patient's diet plan (if they have one) - or 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 <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. Always follow a bolus of dextrose with IV fluids as described below
- If unable to obtain IV access, Glucogel or equivalent can be given to treat the hypoglycaemia
- Use of glucagon is contraindicated
- 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 the patient is tolerating oral intake, follow their recommended diet plan (if they have one) orally/via NG/via PEG with particular attention to fasting intervals (may need continuous feed)
- Monitor glucose and ketones at point of care before each feed (or 4 hourly if on continuous feed/IV fluids) and adjust glucose infusion rate (GIR) to maintain blood glucose between 4mmol/l and 6mmol/l
- Give ondansetron for management of nausea and vomiting
- If restarting feeds, do not stop IV fluids abruptly – the rate can be weaned gradually once the child has demonstrated that they are tolerating oral intake in a satisfactory quantity
- If infection suspected, manage in line with the hospital sepsis pathway. Consider discussion with Micro/ID especially in cases of sepsis or if the patient has multi-drug resistant organisms. Co-amoxiclav should be avoided as clavulanic acid is not recommended in GSDs – amoxicillin is safe to use if appropriate
- **This plan is for the immediate management only and continued management must be discussed with the metabolic team**

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).