

GLYCOGEN STORAGE DISEASE TYPE 3

Priority patient: must not wait in A&E

Patient label

Risk of hypoglycaemic coma, cardiomyopathy and cardiac rhythm disorder
NEVER LEAVE THE PATIENT WITHOUT A SUPPLY OF SUGARS

Do not wait for signs of hypoglycaemia, in all cases initiate management as set out below

1 EMERGENCY WORKUP

Capillary and venous **blood glucose**, electrolytes, urea (BUN), creatinine, calcium, phosphate, magnesium, AST, ALT, CPK
+ Tests depending on the triggering intercurrent illness. Must not delay treatment.

If **cardiac signs** or abnormality on electrocardioscope: ECG, BNP, Troponin +/- echocardiogram; must not delay treatment.

2 IF HYPOGLYCAEMIA < 60 mg/dL (3.3 mmol/L)

- Administer **1mL/kg of 30% glucose** (max. 30 mL) **orally** or enterally if conscious or **3mL/kg of 10% glucose IV** if unconscious (30% glucose also possible via central line or intraosseous route, some teams allow 10mL of 30% glucose via a peripheral venous line for refractory hypoglycaemia).
- Check capillary blood glucose 5 minutes later.
- If still hypoglycaemic, administer a second dose of glucose and check capillary blood glucose 5 minutes later.
- URGENTLY** set up an infusion (2 quick attempts at a peripheral venous line, otherwise intraosseous) **without delaying** the glucose administration.
- Immediately start an infusion even if blood glucose levels have been corrected: Infusion using **10% glucose** with standard electrolyte additions* (not pure 10% glucose)

Age	0-24 months	2-4 years	4-14 years	>14 years - adult	MAX INITIAL RATE
Polyionic 10% glucose (glucose infusion rate)	5-6mL/kg/h (8-10mg/kg/min)	3.5mL/kg/h (6mg/kg/min)	2.5mL/kg/h (4mg/kg/min)	1.2mL/kg/h (2mg/kg/min)	120mL/h (3L/24h)

*e.g.: Bionolyte®, Glucidion®, etc. if no pre-made solution available, use 10% glucose in water + 4g/L NaCl (70 mEq/L) + 2g/L KCl (27 mEq/L)

If **IV line is impossible** => Nasogastric tube or gastrostomy: prepare the IV fluids listed above and pass them through the tube at the same rates.

- CONTRAINDICATION to glucagon.**
- If there are no gastrointestinal disorders and if the preparation is available: instead of infusion, **emergency diet by continuous enteral feeding** using nasogastric tube or gastrostomy (use dietician sheet from parents)

3 IN CIRCUMSTANCES WITH A RISK OF HYPOGLYCAEMIA

- Any circumstance in which the patient is deprived of a carbohydrate supply, e.g. in case of **vomiting, food refusal, diarrhoea** or fasting.
=> Infusion via peripheral line, or continuous enteral feeding of "emergency diet" to be started **IMMEDIATELY**.
- Failure to respect meal times (WARNING:** blood glucose levels can fall very rapidly within 5 minutes!). Hence, in the absence of hypoglycaemia or a situation creating a risk of hypoglycaemia: **Strictly respect** (within 5 minutes) the **meal times** of the patient's usual diet.

4 MONITORING after correction of blood glucose levels

- Check capillary blood glucose after 1h then every 3h.
- Adjust the rate of infusion of 10% glucose + electrolytes by +/- 5 mL/h. Target: capillary blood glucose between 60 and 120 mg/dL.
- If known cardiomyopathy or signs of heart failure, do an ECG, troponin, BNP +/- echocardiogram

PATHOPHYSIOLOGY:

Inherited metabolic disease due to deficiency in glycogen utilisation leading to accumulation of glycogen in the heart, liver and muscles. These patients are **at risk of hypoglycaemia after short periods of fasting, of cardiomyopathy, cardiac rhythm disorders, liver damage and myopathy.**

The usual treatment consists of:

- Meals at precise times of day containing precise quantities of carbohydrates (starch without quick acting sugars), with controlled lactose and fructose intake.
- Occasionally Maizena/Glycosade intake (uncooked corn starch, **not heated**) and/or night time enteral feeding with a precise rate of carbohydrate intake.
- High protein diet (in order to promote gluconeogenesis which is functional in these patients to maintain blood glucose levels), sometimes high lipid or even ketogenic diet and/or treatment with ketone bodies.

DRUG CONTRAINDICATIONS / GENERAL ADVICE:

Prohibited: glucagon (ineffective), oestrogens (hepatic adenoma), NSAIDs if there is a liver disease.

- All vaccinations are recommended (particularly influenza).
- **Never exceed the patient's usual fasting time: if admitted to hospital for a different reason, keep to the patient's usual diet and the precise meal times (which the patient or the parents will know)**
- Do not forget vitamins and trace elements when intake is exclusively parenteral.
- **In case of admission to hospital** (or attendance at A&E): patients must take with them their usual treatments and the special products that they have in order to prepare an emergency diet.
- The emergency treatment will be reassessed with the metabolic medicine specialist during the day.

SURGERY under General Anaesthesia:

WARNING: never leave the patient fasting without an infusion. Implement the emergency protocol with infusion as above, in preparation for surgery.

REFERENCE PHYSICIANS AND CONTACT DETAILS

On-call telephone numbers for metabolic emergencies of:

At night, only the medical teams can call in emergency situations and only if the emergency certificate has not been understood or if the clinical state or test results are worrying. As far as possible make calls before night-time.

Secretarial issues must be dealt with the outpatient office during the week or by email addressed to the patient's referring metabolic physician.

Certificate issued on :

Dr