

GLYCOGEN STORAGE DISEASE TYPE 1A

Priority patient: must not wait in A&E / ED

Patient Label

Risk of hypoglycaemic coma
NEVER LEAVE THE PATIENT WITHOUT A SUPPLY OF CARBOHYDRATES

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

1 EMERGENCY WORKUP

Capillary **and** venous blood glucose, blood gases, lactate, electrolytes, urea (BUN), creatinine, triglycerides, AST, ALT + other tests in line with triggers and/or intercurrent illness depending on context. Must not delay treatment.

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

- Administration of **1mL/kg of 30% glucose** (max. 30 mL) **orally** or enterally if conscious or **3mL/kg of 10% glucose IV** if unconscious (IV 30% glucose is also possible via central line or intraosseous route; some teams allow 10 mL of 30% glucose via 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 (=dextrose) in water with standard electrolyte additions* (never 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)	6mL/kg/h (10mg/kg/min)	5mL/kg/h (8mg/kg/min)	3.5mL/kg/h (6mg/kg/min)	2.5mL/kg/h (4mg/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 available: instead of the infusion, set up the patient's **emergency diet** by **continuous enteral feeding** using nasogastric tube or gastrostomy (use dietician sheet from parents)



NEVER clamp off the glucose infusion: neither in A&E, in theatre, nor when moving the patient (porter / nurse): NEVER, risk of hypoglycaemic coma/seizure.

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, diarrhea** 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.

5 MONITORING after correction of blood glucose levels

- Check capillary blood glucose 1h after starting the infusion, 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 lactate > 5 mmol/L: check blood gases-lactate every 4h. If lactate > 10 mmol/L, add thiamine (B1), 100 to 200 mg/day orally or IV.

PATHOPHYSIOLOGY:

Error of inborn metabolism due to deficiency in the glycogen pathway, characterised by:

- Profound hypoglycaemia after a short period of fasting (2 to 4h depending on the patient). Usual treatment: Meals at precise times of day containing precise quantities of carbohydrates (starch without fast-acting sugars), with controlled lactose and fructose intake. Maizena/Glycosade intake (**uncooked** corn starch, not heated) and/or night time enteral feeding with a precise rate of glucose intake. If intercurrent disease: emergency diet by continuous 24h enteral feeding via NG tube / gastrostomy, with precise carbohydrate intake rate.
- Disorder of platelet aggregation: **risk of bleeding during surgery.**
- Possible **complications** during the course of the disease are renal involvement (tubular disease, renal failure), hepatic involvement (hepatomegaly, elevated liver enzymes, adenomas), hypertriglyceridaemia, hyperlactataemia and hyperuricaemia.

DRUG CONTRAINDICATIONS / GENERAL ADVICE:

Prohibited: antiplatelet drugs (acetylsalicylic acid, NSAIDs), **glucagon**, avoid Ringer's lactate

- All vaccinations are recommended (particularly influenza).
- **Never exceed the patient's usual fasting time: if admitted to hospital for a different reason, maintain the patient's usual diet (including quantities of carbohydrate), intake of uncooked cornstarch and the precise meal times (known by the parents).**
- **If the patient has to be fasted (e.g. for surgery), give the infusion described overleaf.**
- Do not forget vitamins and trace elements when intake is exclusively parenteral.
- **In case of admission to hospital** (or emergency consult): 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 specialist during the day.

SURGICAL PRECAUTIONS: THROMBOPATHY

Anaesthetic protocol: Contact the referring doctor in order to plan for precautionary measures.

- No risk of liver failure; no drug contraindicated apart from aspirin and NSAIDs;
- No additional risk with standard anaesthetic agents.
- **But: POTENTIAL RISK OF BLEEDING / THROMBOPATHY.**

BEFORE SURGERY:

- Investigation of haemostasis prior to planned surgery (and if any sign of bleeding: ecchymoses, haematomas, gingival bleeding, epistaxis => investigation of platelet function in addition)
- **Glucose infusion** (10% glucose + electrolytes), according to the table overleaf, starting ideally 24h before surgery.
- **The day before any surgical procedure: ORAL EXACYL** (tranexamic acid - antifibrinolytic) (1g/10mL or as 500 mg tablets) **20mg/kg/day** divided tid (max 1g x 3/day). Warning: will lower the seizure threshold: if patient has epilepsy, duly consider it's use.

DURING SURGERY:

- **If surgery involving bleeding:** on induction, give **EXACYL IV 10mg/kg** (max 0.5 to 1g by slow IV over 15min)
- **In addition to exacyl, if history of bleeding or known thrombopathy:**
 - **For minor surgery as outpatient: OCTIM® nasal spray** (desmopressin 150mcg/spray): **2 sprays** in one nostril 30 minutes before the operation. Contraindication: child under 2 years old.
 - **If risk of bleeding, or actual bleeding: MINIRIN® IV** (injectable desmopressin 1mL=4mcg) by slow IV over 30 minutes, starting 1 hour before the surgical procedure: **0.3mcg/kg** to be diluted in 50mL of normal saline, then after seeking haematologist's opinion, to be repeated after 12 and/or 24h depending on abundance. At the same time, restrict fluids for 24h, hence the need for a central line to enable a concentrated infusion of glucose (restrict to 20mL/kg/24h). If central line is impossible, closely monitor the blood sodium level.
- **If severe bleeding complication: consider platelet transfusion.**

AFTER SURGERY:

- Continue the glucose infusion post-operatively until the usual oral feeding is restored (normal quantities for 2 successive meals, respecting the patient's usual meal times and diet).
- **Monitor blood glucose and lactate every 3 h + blood gases if lactate >4mmol/L** during and immediately after surgery.
- Oral or slow IV of **EXACYL IN ALL CASES: 20 mg/kg/day** divided into 3 doses (max. 1g x 3/day) for 5 to 15 days as long as the risk of bleeding persists.

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