EMERGENCY CERTIFICATES - G2M NETWORK

V2 – Oct 2024

GLYCOGEN STORAGE DISEASE TYPE 3

PRIORITY PATIENT: MUST NOT BE KEPT WAITING IN THE EMERGENCY DEPARTMENT
Risk of hypoglycaemic coma, cardiomyopathy and cardiac arrhythmia
NEVER LEAVE PATIENT WITHOUT CARBOHYDRATE INTAKE

Label

Do not wait for the onset of hypoglycaemia; always initiate the following management protocol:

Phone call only if the emergency certificate is not understood.

1 EMERGENCY ASSESSMENT

Capillary and venous **blood glucose**, serum electrolytes, urea, creatinine, calcium, phosphorus, magnesium, AST, ALT, CPK + tests as clinically indicated.
If cardiac signs or abnormalities on vital signs monitor: ECG, BNP, troponin +/- echocardiography
Do not delay treatment.

2 INFUSION OR ENTERAL NUTRITION

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

- **Administer glucose** 1 mL/kg of 30% glucose solution (max 30 mL) orally or via enteral tube. If enteral route is not possible: 10% glucose solution 3 mL/kg IV bolus (30% glucose solution also possible via central venous catheter [CVC] or intraosseous [IO] access; some teams administer 30% glucose solution via peripheral IV in refractory hypoglycaemia).
- Check capillary blood glucose 5 minutes later. If hypoglycaemia persists, administer a second glucose dose and check again 5 min later.
- Establish an **EMERGENCY** IV line without delaying glucose administration.

B/ INFUSION

Any circumstance in which the patient is deprived of carbohydrate intake, i.e. **vomiting, refusal to eat, diarrhoea, fasting. even in the absence of hypoglycaemia.**

⇒ Start infusion via a peripheral IV or start continuous enteral "emergency diet" **IMMEDIATELY**

- Infuse 5% glucose with standard electrolyte supplementation* (not glucose alone)

The carbohydrate intakes below are for guidance. Switch to 10% glucose in a balanced electrolyte solution if hypoglycaemia persists despite infusion.
Adjust infusion volume and intake to hydration status.

Age	0 - 24 months	2 - 4 years	4 - 14 years	> 14 years/adult	MAX. FLOW RATE
Flow rate	5–6 mL/kg/h (4–5 mg/kg/min)	3.5 mL/kg/h (3 mg/kg/min)	2.5 mL/kg/h (2 mg/kg/min)	1.2 mL/kg/h (1 mg/kg/min)	120 mL/h (3L/24h)

e.g.: Balanced electrolyte solutions such as Bionolyte, B26, etc. If unavailable: 5% glucose + 4 g/L NaCl (70 mEq/L) and 2 g/L KCl (27 mEq/L)
If IV infusion is not possible **=>** Nasogastric tube or gastrostomy: prepare the above IV solutions and administer via the tube at the same rates.

- **Glucagon is CONTRAINDICATED.**
- In the absence of gastrointestinal issues, instead of infusion, **emergency diet** possible via **continuous** enteral feeding through a nasogastric tube or gastrostomy (preparation known to parents according to the dietary sheet)

3 If presenting to the ED for another reason

CAUTION: Always adhere to the patient's specific mealtimes (blood glucose can drop very quickly, within 5 minutes). Therefore, in the absence of hypoglycaemia or a situation at risk of hypoglycaemia: Adhere strictly to "maintenance diet" mealtimes (no more than 5 minutes difference).
Check capillary blood glucose before each meal and if there is any clinical uncertainty

4 MONITORING after blood glucose correctio..

- Check capillary blood glucose 1 hour after starting the infusion, then every 3 hours.
- Adjust infusion rate by +/-5 mL/h. Target: blood glucose between 60 and 120 mg/dL.
- If known cardiomyopathy or signs of heart failure, obtain ECG, troponin, BNP +/- echocardiography

PHYSIOPATHOLOGY:

Inborn error of metabolism by glycogen deficiency with glycogen accumulation in the heart, liver and muscle. Patients are **at risk of short-fasting hypoglycaemia, cardiomyopathy, arrhythmias, liver involvement and myopathy.**

Usual treatment:

- Daytime meals at set times with precise amounts of carbohydrate (starch without fast-acting sugars) and controlled lactose and fructose.
- Sometimes uncooked, **unheated** cornstarch (Maïzena/Glycosade) and/or nocturnal enteral nutrition with a precise carbohydrate intake.
- High-protein diet (to promote gluconeogenesis, which is functional for maintaining blood glucose), sometimes high-fat or even ketogenic, and/or treatment with ketone bodies

DRUG CONTRAINDICATIONS/GENERAL ADVICE:

Prohibited: **glucagon** (ineffective), oestrogens (risk of hepatic adenoma), NSAIDs if liver disease is present

- All vaccinations are recommended (especially influenza).
- **Never exceed the patient's usual fasting time: if hospitalised for another reason, follow the usual diet and precise mealtimes (known to parents)**
- Remember vitamin and trace element supplementation if the patient is on exclusive parenteral nutrition.
- **In the event of hospitalisation** (or emergency consultation): patients must bring their usual treatments and any special products they have with them to prepare an emergency diet.
- Emergency treatment must be reviewed with the metabolic specialist during the day.

SURGERY with general anaesthesia

CAUTION: never leave the patient fasting without an infusion. Apply the emergency protocol with the above infusion in preparation for surgery.

Refer to the **Emergency section** on the G2M website.

REFERRING PHYSICIANS AND NUMBERS:

At night, only medical teams may call in emergencies and only if the emergency certificate is not understood or if the clinical condition or test results are concerning. Whenever possible, calls should be made before nightfall.
Secretarial issues must be dealt with via the medical secretariat during the week, or by email addressed to the patient's metabolic medicine specialist.

Certificate issued on Dr