# EMERGENCY CERTIFICATES - G2M NETWORK

V2 – Oct 2024

# GLYCOGEN STORAGE DISEASE TYPE IA

**PRIORITY PATIENT: Immediate capillary blood glucose test upon admission to the ED**
**Risk of severe hypoglycaemia**
**NEVER LEAVE PATIENT WITHOUT CARBOHYDRATE INTAKE even if there is no hypoglycaemia at admission**

Phone call only if the emergency certificate is not understood.

## 1 IF HYPOGLYCAEMIA < 60 mg/dL (= 3.3 mmol/L) → ADMINISTER GLUCOSE WITHOUT DELAY

- **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).
- Establish an **emergency IV line** (2 attempts at rapid peripheral venous puncture, otherwise IO access) without delaying glucose administration. → Do not wait for blood glucose correction to start the infusion (see table below)
- Check capillary blood glucose 5 minutes after glucose administration. If hypoglycaemia persists, administer a second glucose dose and check again 5 min later.

## 2 INFUSION or ENTERAL NUTRITION

- 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.
  ⇒ Infusion via peripheral IV or continuous enteral "emergency diet" must be started **IMMEDIATELY.**

Infuse 10% glucose with standard electrolyte supplementation* (not 10% glucose alone) →

| Age | 0 - 24 months | 2 - 4 years | 4 - 14 years | > 14 years/adult | MAX. FLOW RATE |
|---|---|---|---|---|---|
| Flow rate | 6 mL/kg/h (10 mg/kg/min) | 5 mL/kg/h (8 mg/kg/min) | 3.5 mL/kg/h (6 mg/kg/min) | 2.5 mL/kg/h (4 mg/kg/min) | 120 mL/h (3L/24h) |

***e.g.:*** Balanced electrolyte solution such as Bionolyte, B45, Glucidion. If unavailable: 10% 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 CONTRAINDICATED**

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

**NEVER clamp the glucose infusion — not in the ED, not in the operating theatre, not during transport (by stretcher bearer/RN): NEVER, LIFE-THREATENING**

**Assessment when starting the infusion:** Capillary and venous blood glucose, blood gases with lactate, serum electrolytes + additional tests as clinically indicated. Treatment must not be delayed.

## 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 correction

- Monitor capillary blood glucose 1 hour after starting the infusion, then every 3 hours.
- Adjust the infusion rate of 10% glucose + electrolytes by +/- 5 mL/h. Target: blood glucose between 60 and 120 mg/dL.
- If hyperlactataemia > 5 mmol/L: check blood gases with lactate every 4 hours. If hyperlactataemia > 10 mmol/L: add thiamine (B1) 100–200 mg/day PO or IV.

## PHYSIOPATHOLOGY:

Inborn error of metabolism by glycogen deficiency characterised by:

- **Severe hypoglycaemia after short fasting periods (2–4 hours depending on the patient).** Usual treatment: Daytime meals at set times with precise amounts of carbohydrate (starch without fast-acting sugars) and controlled lactose and fructose. **Uncooked, unheated** cornstarch (Maïzena, Glycosade) and/or nocturnal enteral nutrition with a precise carbohydrate intake. If intercurrent illness: 24-hour continuous "emergency diet" via NG tube/gastrostomy with a precise carbohydrate intake.
- **Platelet aggregation disorder** may occur, increasing **bleeding risk** during surgery.
- Possible **complications**: renal impairment (tubulopathy, renal failure), hepatic impairment (hepatomegaly, cytolysis, adenomas), hypertriglyceridaemia, hyperlactataemia, hyperuricaemia.

## DRUG CONTRAINDICATIONS/GENERAL ADVICE:

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

- All vaccinations are recommended (especially influenza).
- **Never exceed the patient's usual fasting time: if hospitalised for another reason, follow the usual diet (carbohydrate amounts), cornstarch intake and precise mealtimes (known to parents)**
- **If fasting is necessary (e.g. before surgery), give the infusion described on the front page.**
- 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.

## SURGICAL PRECAUTIONS: THROMBOPATHY

**Anaesthesia protocol: Contact the referring doctor to anticipate precautions.**

- No risk of hepatic failure; no contraindicated drugs other than aspirin and NSAIDs;
- No additional risk with standard anaesthetics.
- But: **POTENTIAL BLEEDING RISK/THROMBOPATHY**

→ See the specific protocol "surgery in glycogen storage disease Ia and Ib"

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