EMERGENCY CERTIFICATES - G2M NETWORK

V2 – Oct 2024

HYPERINSULINISM

PRIORITY PATIENT: MUST NOT BE KEPT WAITING IN THE EMERGENCY DEPARTMENT
In the event of vomiting, diarrhoea, fever or fasting
= risk of hypoglycaemia/seizures or hypoglycaemic coma

Label

Do not wait for signs of hypoglycaemia, always begin treatment according to the protocol below

1 EMERGENCY ASSESSMENT

Phone call only if the emergency certificate is not understood.

Capillary blood glucose, venous blood glucose if possible, without delaying treatment

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

- **If coma or seizures or difficulty obtaining venous access quickly**:
  - **GLUCAGON** 1 mg/mL: inject **1 mg SC**, regardless of age /BAQSIMI if available: 3 mg single-dose intranasal (adults and children > 4 years)
  - **Then always begin: Infusion of** 10% glucose with standard electrolyte supplementation* (not 10% glucose alone)
    Initial infusion rate table, to be adjusted later (see paragraph below)

Age	0 - 24 months	2-14 years	> 14 years/adult	MAX. FLOW RATE
Balanced electrolyte solution 10% glucose	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, etc. If unavailable: 10% glucose + 4–6 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.

  - If hypoglycaemia persists or if no venous access with digestive intolerance: Continuous glucagon (SC or IV) 1 mg/24h. Dilute 1 mg = 1 mL in 11 mL of 0.9% NaCl, administer continuously at 0.5 mL/h

- **If the patient is conscious:**
  - **Rapid PO or enteral glucose administration**: 30% glucose solution 1 mL/kg, max 30 mL, or 1 sugar cube/20 kg body weight
  - Check capillary blood glucose 5–10 minutes later. If hypoglycaemia persists, administer a second glucose dose and check again 5 to 10 mins later.
  - **If hypoglycaemia still persists**: see above (infusion and/or glucagon)
  - **If blood glucose normalises:**
    1/ Immediate snack, then resume normal oral or enteral feeding
    2/ In the event of food refusal or intolerance (vomiting, diarrhoea): infusion (see above)
- **Titration of infusion:**
  - Check capillary blood glucose every 30 minutes and adjust the infusion rate in increments of +/-10% of the initial rate until 2 consecutive values are between 3.3 mmol/L (0.6 g/L) and 5.5 mmol/L (1 g/L)
  - BG < 3.3 mmol/L (0.60 g/L): Administer 30% glucose (see above) and increase infusion rate by 10%. Check again 10 minutes later
  - If hypoglycaemia persists despite increasing the infusion rate: Continuous IV or SC glucagon, see above.

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

3 IN THE ABSENCE OF HYPOGLYCAEMIA

- Assess food intake and treatment compliance
- If food intolerance, even without hypoglycaemia: start the infusion above

4 CONTINUE USUAL TREATMENTS (depending on the patient)

- Diazoxide (Proglycem) PO or NG tube
- Somatostatin analogues by continuous SC infusion (pump) or regular IM injections
  - ➢ NEVER STOP THESE TREATMENTS! Continue exactly as prescribed to the parents.
  - ➢ If the subcutaneous pump has to be removed (e.g. for a brain MRI): inject 1/3 of the daily dose SC.

HYPERINSULINISM

PHYSIOPATHOLOGY:

Patients with congenital hyperinsulinism have dysregulated **insulin secretion**, leading to excessive insulin release and **hypoglycaemia**.
Usual treatment, depending on the patient:

- Somatostatin analogue, SC or IM
- Diazoxide
- Sometimes continuous daytime and/or night-time enteral tube feeding.

CAUTION: never stop this continuous feeding without an infusion!

CIRCUMSTANCES WITH RISK OF HYPOGLYCAEMIA:

- **Unexpected discontinuation of treatment**/situations where food or treatments are no longer absorbed (vomiting, diarrhoea).
- Intercurrent infection, fever, anorexia, vomiting, surgery

In all these situations, the patient must be monitored, starting an infusion if feeding is impossible, to avoid hypoglycaemia.

GUIDANCE ON PRACTICAL ADMINISTRATION OF TREATMENTS /ADVERSE REACTIONS:

- GLUCAGON if severe hypoglycaemia/unconscious child: 1 mg = 1 mL SC injection
- Continuous IV or SC GLUCAGON infusion: If hypoglycaemia persists despite infusion: 1 mg/24 h. Dilute 1 mg = 1 mL in 11 mL of 0.9% NaCl, administer continuously at 0.5 mL/h
- Somatostatin analogues (SANDOSTATIN/SOMATULINE/OCTREOTIDE/PASIREOTIDE): continuous SC infusion (pump) or regular IM injections.
  - ➢ Personalised prescriptions: continue exactly as prescribed to the parents.
  - ➢ If the subcutaneous pump has to be removed (e.g. for a brain MRI): inject 1/3 of the daily dose SC

  Side effects: Before 1 month of age: enterocolitis and poor GI tolerance After 1 month of age: gallstones, drug-induced hepatitis.
- DIAZOXIDE/PROGLYCEM: 25 mg or 100 mg capsules or 50 mg/mL oral suspension (under temporary authorisation [ATU]) Side effects; before 6 months of age: monitor for pulmonary arterial hypertension (PAH) and oedema. After 6 months: rare cases of pericarditis.

DRUG CONTRAINDICATIONS/GENERAL ADVICE:

- Prohibited: none
- All vaccinations are recommended (especially influenza).
- Prolonged fasting contraindicated: Adhere to the fasting times specific to each patient.

SURGERY

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