

Surgical Protocol

Label

Risk of hypoglycaemic coma, NEVER LEAVE THE PATIENT WITHOUT A CARBOHYDRATE

1 RISK OF HYPOGLYCAEMIA

No fasting without serum glucose infusion. Monitor capillary blood glucose +/- lactate every 3 hours.



NEVER clamp the glucose infusion: not in the emergency department, nor in the operating theatre and nor during patient transport (by stretcher-bearer or nurse): RISK OF DEATH

- Example of infusion:** serum glucose **G10%** with standard electrolyte supplementation* (not pure G10)
*e.g.: Polyionic, Bionolyte, B45, Glucidion, etc. if no solutes available, 10% glucose + 4g/L of NaCl (70 meq/L) and 2g/L of KCl (27 meq/L).

Age	0–24 months	2–4 years	4–14 years	> 14 years–adult	MAX FLOW RATE
Infusion 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)

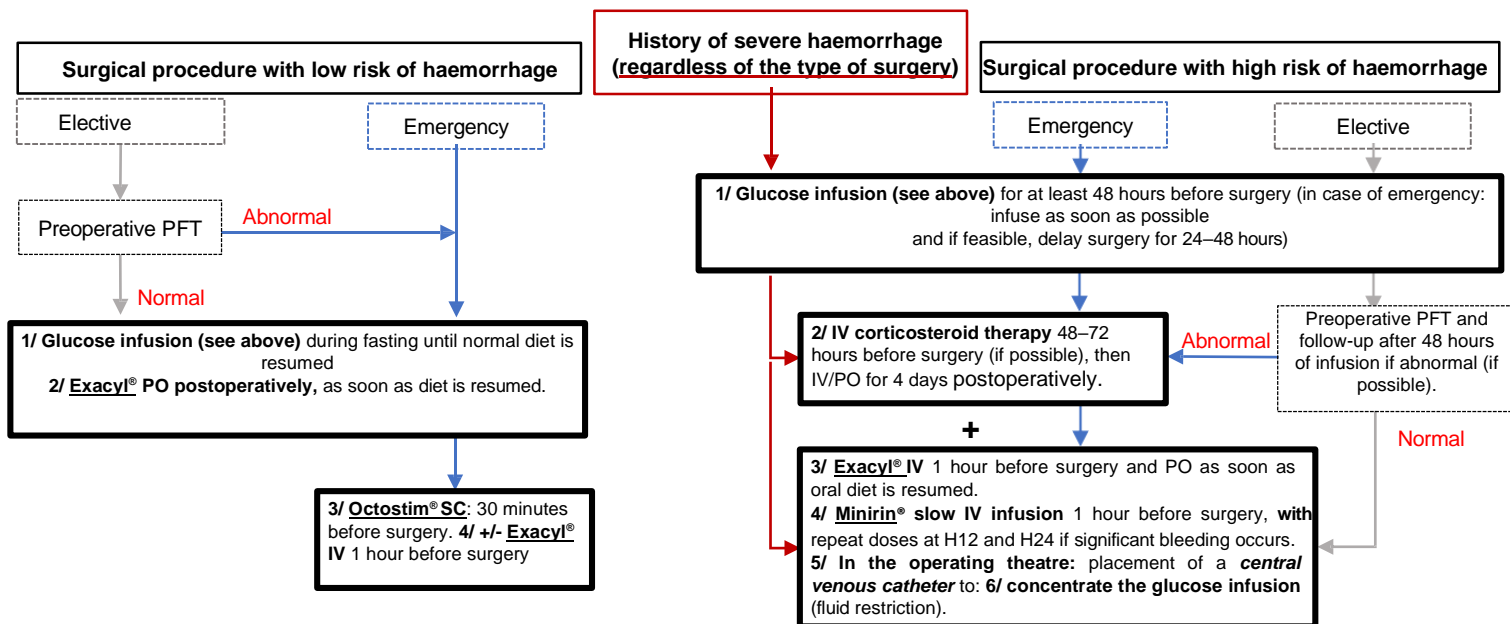
- In the event of hypoglycaemia (< 3.3 mmol/L or 60 mg/dL):**
 - Glucagon is CONTRAINDICATED**
 - Administer glucose as follows: **1 mL/kg of G30%** (max 30 mL) PO or enterally if patient is conscious or **3 mL/kg of G10%** IV if patient is unconscious (G30% IV also possible).
 - Check capillary blood glucose (dextro) 5 minutes later.
 - If hypoglycaemia persists, repeat glucose administration and recheck blood glucose (dextro) 5 minutes later.
 - Adjust the glucose infusion rate by 1–2 mg/kg/min (0.6–1.2 mL/kg/h)
- Target blood glucose: 3.3–6.6 mmol/L or 60–120 mg/dL.
- Hyperlactataemia may be habitual in some patients. If > 5 mmol/L: check blood gas lactate every 3–4 hours, add thiamine (B1) 100–200 mg/day orally or IV (500 mg in adults), and temporarily stop SGLT2 inhibitors for type 1B (for 1 week).

A central venous catheter is recommended to concentrate the glucose intake if the fluid intake is high over a prolonged period.

2 RISK OF HAEMORRHAGE

Haemostasis tests, including platelet function tests and von Willebrand factor testing, should be carried out regularly in specialised haemostasis consultations and prior to elective surgery.

DOSAGES OVERLEAF



BEFORE SURGERY

For elective surgeries:

- Schedule as the first procedure in the operating theatre
- plan for at least four full days of hospitalisation before surgery if the procedure carries a high risk of haemorrhage.
- Stop cornstarch (Maizena, Glycosade) 12 to 24 hours before surgery (risk of aspiration), while ensuring glucose infusion is maintained

If **Mimirin® IV** is used → **Fluid restriction:** 20 mL/kg/24 hours or 750 mL/24 hours for patients > 40 kg, requiring placement of a central venous catheter with concentrated glucose solution (G30, G50%, etc.) to maintain the same carbohydrate supply. Monitor electrolytes twice daily for 48 hours post-injection.

INTRA- AND POSTOPERATIVE PERIOD

In the event of intraoperative haemorrhage: platelet transfusion (in addition to standard management)

Discontinue the glucose infusion postoperatively: Continue the glucose infusion postoperatively until the patient has resumed a normal oral diet (normal quantities for two consecutive meals, according to the patient's usual meal times and diet). Progressive tapering of the glucose infusion is essential once the patient is fully fed, as abrupt cessation can rapidly lead to rebound hypoglycaemia.

Blood glucose and lactate monitoring: Every 3 hours, with GDS if lactate > 5 mmol/L during the perioperative and immediate postoperative period.

Drug	INN	Dosage	Administration
Exacyl IV: 500 mg/5mL	Tranexamic acid	IV: 10 mg/kg (max 0.5 to 1 g by slow IV infusion over 15 min) CI: History of thromboembolic events Caution: Lowers the seizure threshold	Slow IV infusion over 15 min 1 hour before surgery
Exacyl® PO (oral suspension or 500 mg tablets) Exacyl IV: 500 mg/5mL	Tranexamic acid	PO: Adults: 3–5 tablets/day in 2–3 intakes/day; children: 20 mg/kg/intake, 2–3 times/day CI: History of thromboembolic events Caution: Lowers the seizure threshold	PO, as soon as oral diet is resumed. Duration: 3 to 15 days, as long as the risk of haemorrhage persists
Octostim solution for SC injection, 1 mL ampoule (15 µg/mL)	Desmopressin acetate	0.2–0.3 µg/kg (max 30 µg)	SC, 30 minutes before surgery
Minirin® solution for injection IV, 1 mL ampoule (4 µg/mL)	Desmopressin	0.3 µg/kg diluted in 50 mL saline solution (max)	Slow IV infusion over 20–30 min 1 hour before surgery. Repeat at H12 and H24 if significant bleeding occurs
Corticosteroid therapy IV	Methylprednisolone IV Prednisolone PO	2 mg/kg/day (max 60 mg/day)	IV, start 48–72 hours before surgery. Continue for 4 days after surgery if there is a risk of haemorrhage. Switch to PO at H24 if there is no bleeding.

PATHOPHYSIOLOGY:

Inherited metabolic disease due to impaired glycogen utilisation, characterised by:

- acute complications:

- **Profound hypoglycaemia (with hyperlactataemia) after a short fast (2 to 4 hours depending on the patient).**
- **Impaired platelet aggregation**, leading to a **risk of haemorrhage** during surgery.
- Acute hypertriglyceridaemia (with (rare) risk of pancreatitis).

- chronic complications:

- Possible **complications** in the course of the disease include: renal involvement (tubulopathy, lithiasis, renal failure), hepatic involvement (hepatomegaly, cytolysis, adenomas), hypertriglyceridaemia, hyperlactataemia and hyperuricaemia (risk of gout attacks).
- **Standard treatment:** diet with precisely timed meals during the day and precise quantities of carbohydrates (starch, no simple sugars), including doses of Maizena/Glycosade (raw, unheated cornstarch) and/or nocturnal enteral feeding with a precise carbohydrate infusion rate.

Type 1B patients have neutropenia and require specific treatment with GCSF or SGLT2 inhibitors.

View the Emergency section of the G2M website

**DRUG CONTRAINDICATIONS / GENERAL ADVICE:**

Prohibited: platelet aggregation inhibitors (acetylsalicylic acid, NSAIDs), **glucagon**. Avoid Ringer's lactate.

SURGICAL PRECAUTIONS: THROMBOPATHY

Anaesthesia protocol: Contact the referring doctor in advance to discuss precautions.

- No risk of liver failure; no contraindicated medications apart from aspirin and NSAIDs;
- No additional risk with standard anaesthetics.
- **But: POTENTIAL RISK OF HAEMORRHAGE/THROMBOPATHY Patients may present with a specific thrombopathy with characteristics similar to those of von Willebrand disease. Therefore, the risk of haemorrhage may be increased, which must be anticipated and managed. This risk is assessed by platelet aggregation testing during a dedicated consultation or in partnership with a haemostasis specialist.**
- **BEFORE SURGERY: See specific instructions overleaf** (diagram)
- Plan for assessment of haemostasis and platelet function tests (PFT) before elective surgery (and check for signs of bleeding: ecchymosis, haematoma, gingival bleeding, epistaxis)
- The GSD I diet must be maintained on a regular schedule in the preoperative period until fasting begins. **Cornstarch should be discontinued at least 12 hours and up to 24 hours before surgery due to the risk of aspiration.**

REFERRING DOCTORS 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 patient's clinical condition or test results are concerning. Where possible, calls should be made before night time.

Secretarial issues must be dealt with via the medical secretariat during the week or by email addressed to the patient's referring metabolic doctor.

Certificate issued on

Dr