Glycogen storage disease type Ia: Assessment and preanesthetic management - Case Report

Escala Braga

Celina Oliveira, Celina Gomes, Pedro Santos, Paulo Fragoso, Maria Crisóstomo, Henedina Antunes
Department of Anesthesiology, Braga Hospital, Portugal

BACKGROUND AND AIMS

Glycogen storage disease type Ia (G-Ia; von Gierke´s disease) is a rare inherited metabolic disorder resulting from glucose-6-phosphatase deficiency, a key enzyme in the glycogen metabolism (1). Since patients are unable to release glucose from the liver, hypoglycemia and lactic acidosis after short periods of fasting, and hepatomegaly are the most common signs. Prevention of complications by maintenance of normoglycemia, demands the ingestion of corn starch in a 3 hourly basis. Hyperuricemia, hyperlipidemia and platelet dysfunction are also common (1).

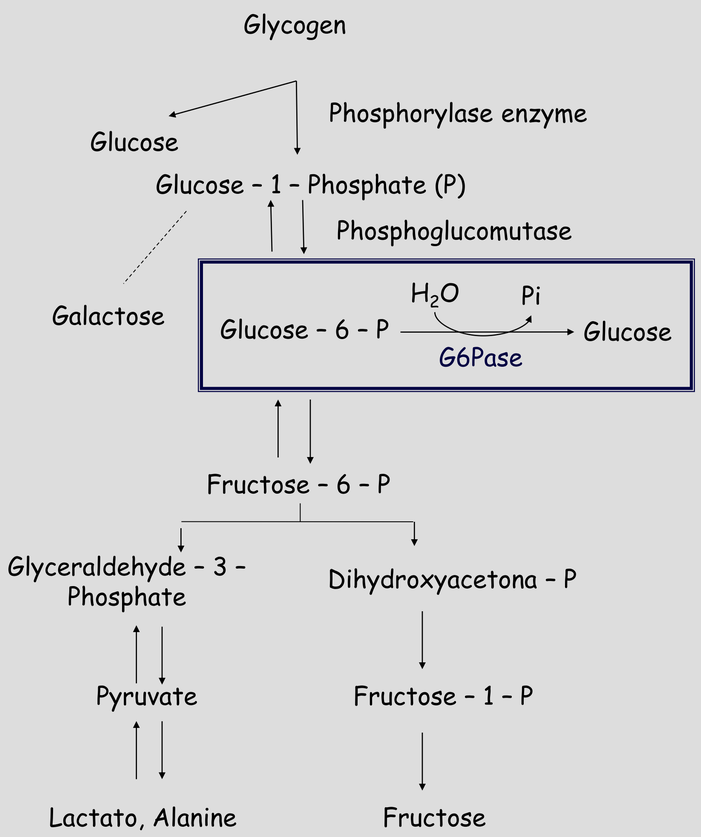

Fig. 1 – Schema of glycogen metabolism

CASE REPORT

Identification

Boy, 11 years old, 29,6 Kg , ASA III, with a diagnosis of G-Ia, since 12 months old.

Presented with hepatomegaly, growth retardation, elevated liver enzymes and hyperlipidemia.
No previous surgeries and no history of allergies or atopy.
Scheduled for elective circumcision.

Pre-anesthetic evaluation

- At 6am: last meal of corn starch;
- At 7am: admission and infusion of 5% glucose on half saline at a rate of 75cc/h;
- At 9am: hourly blood glucose determination was started ;
- At 12h the surgery took place.

Anesthesia

General Balanced Anesthesia
Induction: Midazolam 1 mg + Fentanyl 100 μg + Propofol 100 mg
Airway: laryngeal mask # 2,5
Mantenaince: Sevoflurane in nitrous oxide/oxygen and air mixture
Acetaminophen 500 mg iv
Length of anesthesia : 55 minutes
Per-operative period without accidents or complications.

Hourly blood glucose monitoring

Time	9am	10am	11am	12am	13pm
Glucose – venous blood (mg/dl)	128	110	84	95	83

Postoperative period

At 2pm oral feeding resumed without any complications.

DISCUSSION AND CONCLUSIONS

The pre-anesthetic evaluation and management of a patient with G-Ia is a challenge for the anesthesiologist, because these patients do not tolerate the fasting period required for the intervention (2). It is therefore necessary to adopt strategies to avoid hypoglycemia and lactic acidosis as well as its complications. According to the Pediatrics Department an adequate glycemic intake was selected, as well as its rate of administration; hourly blood glucose monitoring and infusion rate control were undertaken to maintain blood glucose levels above 60 mg/dl while avoiding acidosis. The use of Ringer´s lactate is not recommended by the risk of worsening lactic acidosis.

References: - Anesthesia and Co-Existing Disease 2008 5th ed: 319
- Anaesthesia 1980; 35:699-702