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# Bariatric surgery is not contraindicated in obese patients suffering from glycogen storage disease type IXa. A case report with follow-up at three years



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## ABSTRACT

**INTRODUCTION:** Glucose storage disease type IXa (GSD IXa) is an uncommon condition presenting with childhood onset hepatomegaly, growth retardation, and often, fasting ketosis and hypoglycemia. Despite its benign course, the lack of dietary counseling may favor uncontrolled weight gain. We investigated the efficacy of bariatric surgery in one 17 years old female suffering from GSD IXa and morbid obesity.

**PRESENTATION OF CASE:** The diagnosis was GSD type IXa in a patient with a body mass index (BMI) of 45.5 kg/m<sup>2</sup>. Onset of hypoglycemia was reported twice each month. She was treated her implanting an adjustable gastric banding through laparoscopy. Three years after surgery the patient presents a BMI of 30.1 kg/m<sup>2</sup> and an excess of weight loss (EWL) of 71.1%. Only once, following surgery, she had to deflate her band to allow a faster transit of food through her stomach, thus reaching a prompt euglycemic condition, due to an incoming hypoglycemic crisis.

**DISCUSSION:** Laparoscopic adjustable gastric banding (LAGB) is one of the most used approaches to treat morbid obesity. It is a restrictive procedure unable to affect the absorption of any nutrient, presenting a very low intra and perioperative complication rate. In our GSD IXa patient, it offered a prompt modification of food intake restriction whenever requested, thus avoiding hypoglycemia.

**CONCLUSION:** LAGB is effective in determining weight loss without inducing significant side effects or worsening hypoglycemia, in this morbid obese patient, suffering from GSD type IXa.

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## 1. Introduction

In last years, both clinical research and technical advance have led to an improvement of the efficacy in the diagnosis and treatment of several diseases of surgical interest.<sup>1,2</sup> Obesity is a chronic disease and bariatric surgical procedures allow the achievement of substantial weight loss<sup>3</sup> and major secondary health benefits<sup>4–6</sup> even in the long term.<sup>7</sup> Glycogen storage disease type IX (GSD type IX) results from a deficiency of phosphorylase kinase activity (PhK), which has a major regulatory role in the breakdown of glycogen. Two variants of PhK deficiency are known, liver PhK deficiency or GSD type IXa and muscle PhK deficiency or GSD type IXc, which is notably rarer.<sup>8</sup> Patients suffering from GSD type IXa, present early childhood onset hepatomegaly and growth retardation, and often, but not always, fasting ketosis and hypoglycemia. Although obesity

is uncommon in these patients, the suggested diet, high in complex carbohydrates and protein, to prevent hypoglycemia, may favor an uncontrolled weight gain. In this light, under the periodical evaluation of a metabolic nutritionist, routine monitoring of blood glucose and blood ketones concentration as well as reduced food intake, are prescribed.<sup>9</sup>

The case of a 17 years old female suffering from GSD type IXa and presenting with a body mass index (BMI) of 45.5 kg/m<sup>2</sup> treated with LAGB, is here reported.

## 2. Case report

In October 2010 a 17 years old female was referred from pediatrics department to our bariatric surgery unit. The diagnosis was GSD type IXa in a patient with a BMI of 45.5 kg/m<sup>2</sup> (weight 124 kg, height 165 cm). Her laboratory test showed a PhK of 11.0 μmol/min/gHb (normal hospital values 100–300) an amyloglucosidase level of 6 nmol/min/gHb (normal hospital values 0.6–3.5) a blood glucose level of 77 mg/dl (normal hospital values 60–110) an HbA1c of 6.3% (normal hospital values below 6.5%) and a glycogen level of 17 mg/dl (normal hospital values 0–10).

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**Table 1**

Pre- and postoperative clinical and biochemical status.

	Before surgery	At 1 year of follow up	At 3 years of follow up
Weight (kg)	124	100	82
BMI (kg/m <sup>2</sup> )	45.5	36.7	30.1
EWL %	–	40.6	71.1
Total bilirubin (mg/dl)	1.7 <sup>a</sup>	1.6 <sup>a</sup>	1.4 <sup>a</sup>
Conjugated bilirubin (mg/dl)	0.2	0.1	0.2
AST (U/l)	56 <sup>a</sup>	48 <sup>a</sup>	35
ALT (U/l)	116 <sup>a</sup>	75 <sup>a</sup>	70 <sup>a</sup>
Total cholesterol (mg/dl)	220 <sup>a</sup>	210 <sup>a</sup>	180
CPK (U/l)	156 <sup>a</sup>	144 <sup>a</sup>	95
AP (mmHg)	155/95 <sup>a</sup>	130/90	120/80
pH	7.30	7.40	7.41
pO <sub>2</sub> %	82.0	91	95
SatO <sub>2</sub> %	95	99	99

<sup>a</sup> Higher than normal hospital range.

See Table 1 for other values. A liver biopsy confirmed the diagnosis. Liver ultrasound showed hepatomegaly. A clinical presentation of hypoglycemia was reported twice each month. It was usually treated with the assumption of sugar or chocolate. Due to the benign course of the illness, dietary counseling had been abandoned in favor of an uncontrolled diet. As a consequence of frequent consumption of junk food, both her mother and her sister suffered from morbid obesity. Besides this clinical presentation, cardiopulmonary, hormonal and psychological parameters were normal. After obtaining the authorization from our university ethic committee, following a thorough staff discussion, it was decided to implant in this patient an adjustable gastric banding by laparoscopy. In order to get a signed informed consent to surgery, the patient and her family were educated about the risk benefit ratio of a bariatric procedure. Surgery was then performed as usual in about 45 min. The “pars flaccida technique” was used.<sup>10</sup> As a technical variation, the regulation port was placed palpable into subcutaneous tissue of left groin, and not fixed to the deep muscular fascia. The postoperative period was uneventful and the patient was discharged on second postoperative day. During the first control, at one week from surgery, besides from all planned visits, she was taught to deflate her band by aspirating water saline solution from the regulation port.

At three years from surgery the patient is now 82 kg (BMI 30.1 kg/m<sup>2</sup>) with an excess of weight loss (EWL) of 71.1%, in good conditions, with a glycemic status mostly unmodified and an acceptable biochemical panel, see Table 1. She is now following dietary counseling again although she admits the occasional consumption of sweets. Following hospital discharge, during the postoperative port regulation, the young patient and her family were taught about the management of the device. A staff nurse on duty was however available 24 h a day. In only one circumstance, due to an incoming hypoglycemic crisis, she had to deflate her band to allow a faster transit of food through the stomach, thus reaching a prompt euglycemic condition.

### 3. Discussion

The association between obesity and GSD type Ia has been reported.<sup>11</sup> This more aggressive GSD variant is characterized by an unfavorable prognosis. GSD type IXa has on the other hand a more benign course, although hypoglycemia and hepatomegaly are however present. This is caused by the deficiency of PhK. It makes the physiological amount of blood glucose from the breakdown of glycogen, unavailable, whenever requested.<sup>8,9</sup> Fasting hypoglycemia is routinely controlled by a monitored nutritional support, which includes frequent daytime feedings that are high in complex carbohydrates and protein; conversely an uncontrolled or excessive food intake may turn into weight gain probably sustained by insulin resistance.<sup>11</sup>

The management of morbid obese patients suffering from GSD type IXa, in which dietary counseling and a change in life habits have failed to induce weight loss, is complex. On one hand, bariatric surgery may be indicated to achieve substantial weight loss and improve or heal comorbidities; on the other, a surgical procedure leading to restriction or malabsorption of food intake may worsen hypoglycemia if present.

According to this, while our patient fulfilled in general the operative criteria suggested by the Italian Society for bariatric and Metabolic Surgery (SICOB) guidelines<sup>12</sup> and by many other scientific societies,<sup>13</sup> on the other hand, our goal was to offer her a restrictive bariatric procedure presenting a very low perioperative complication rate<sup>14</sup> and which was, at the same time, unable to affect the absorption of any nutrient. Furthermore, a prompt modification of food intake restriction whenever needed by the patient, thus avoiding hypoglycemia, was requested. Laparoscopic adjustable gastric banding (LAGB) met these criteria. Other known restrictive bariatric procedures, such as intragastric balloon or sleeve gastrectomy are in fact not adjustable in response to the nutritional needs of the patient, and may determine a high risk to worsen hypoglycemia. Furthermore, LAGB efficacy in the long term does not only depend on the mechanical restriction to stomach filling, but also on the neurological regulation of hunger and satiety possibly mediated by mechanoreceptors present in the gastric wall.<sup>7,13</sup>

### 4. Conclusion

To our knowledge, this is the first report of bariatric surgery in a morbid obese patient suffering from GSD. Although type IXa is a very mild expression of GSD and has a benign course, severe obesity may represent a serious complication; in the present case a morbid obese patient suffering from GSD type IXa was effectively treated with LAGB. This surgical procedure determined weight loss, without inducing significant side effects or worsening hypoglycemia.

### Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

### Conflict of interest

No conflict of interest to disclose.

### Funding

No funding sources to declare.

**Ethical approval**

Approved by “Federico II” University Ethic Committee.

**Authors' contributions**

Mario Musella, Marco Milone, Paola Maietta and Paolo Bianco have made substantial contribution to conception and design

of the study and acquisition of data by performing the surgery reported.

Mario Musella, Marco Milone, Anna Pisapia, Dario Gaudio and Rubina Palumbo have been involved in drafting the manuscript and revising it critically.

All authors have given final approval of the version to be published.

**Key learning points**

- Efficacy of Bariatric Surgery in obese patients suffering from Glycogen storage disease type IXa.

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