Glycogen storage diseases (GSD) are a group of inherited disorders which result in deficiencies of enzymes involved in glycogen metabolism and lead to an accumulation of glycogen in the liver. Glycogen storage disease type 1a, a form of glycogen storage disturbance, is a rare metabolic disorder with important implications for anaesthesiologists. It is caused by the lack of the glucose-6-phosphatase, which is necessary for the liver to convert glycogen to glucose. The aim of this case report is to present an uncomplicated surgery with an uncomplicated general anaesthesia and an anaesthetic follow up in a patient with GSD type 1a in an elective day case surgery. In this case, there was a serious hypoglycemia problem due to the glycogen storage disturbance related to disease. To avoid severe hypoglycemia, it is crucial to keep oral feeding 2-3 hour for maintaining a normal blood sugar level.

More recently Huang detailed two cases in which the clinical course appeared satisfactory, but in both of them blood samples showed the presence of moderate and severe hypoglycemia during surgery. From the anaesthetist’s point of view: hypoglycemia and hepatic dysfunction are the major problems to deal with GSD type 1a patients at the perioperative period. Moreover, difficulties in intubation and ventilation and aspiration risks are the other encountered problems in GSD patients during surgery. 

The application of modern methods of management can correct or improve the clinical and biochemical features of GSD type 1a disease. If peri-
ods of hypoglycemia and metabolic acidosis can be avoided then these patients appear to tolerate anaesthesia and surgery satisfactorily.

The aim of this case report is to present an uncomplicated surgery with an uncomplicated general anaesthesia and an anaesthetic follow up in a patient with GSD type 1a in an elective day case surgery.

CASE REPORT

Tonsillectomy, adenoidectomy and circumcision were planned in a 2-yr-old boy with GSD-1a. He was diagnosed in the infancy period after a liver biopsy and an enzyme assay. His physical examination, the abdomen was distended and a remarkable hepatosplenomegaly was detected. His preoperative evaluation revealed normal platelet count and liver enzymes. His protrombin time (PT) and activated partial trombin time (PTT) levels were within normal limits (PT:11.6 sec [12-18 sec], aPTT: 28.8 sec [25-40 sec], INR:0.87% [0.80-1.22]). Blood count revealed leukopenia (4620 leukocytes per mm3). The patient was consulted to pediatrics and close follow up for hypoglycemia during and after surgery was advised.

The patient was fasted for 4 hr preoperatively. His preoperative blood sample was drawn for the assessment of blood glucose level after the insertion of an intravenous cannula and was found 59 gr.dL⁻¹ (70-100 gr.dL⁻¹). The infusion of 10% dextrose with 0.9% NaCl was started thereafter. His blood glucose was measured before premedication. He was premedicated with midazolam (Dormicum, RocheÒ) 0.5 mg (IV) after blood glucose reached 91 gr.dL⁻¹. Anaesthesia was induced with 3 mg.kg⁻¹ propofol (Propofol, Abbott) and muscle relaxation was achieved with rocuronium bromide (Esmeron, MSD) 8 mg. Orotracheal intubation was easily accomplished using a 4.5 mm ID endotracheal tube, and the lungs were mechanically ventilated. N2O/O2 combination and sevoflurane 2% was used for maintenance. Oxygenation, ventilation, and haemodynamic status were normal and stable. Venous blood was taken for gas analysis and for plasma glucose concentration measurements. Except for severe hypoglycemia (plasma glucose concentration of 65 mg/dL⁻¹) at the beginning of the procedure, which was corrected by intravenous administration of 10% dextrose, there were no episodes of metabolic acidosis. At the end of the surgery, volatile agents were discontinued and the patient was extubated. Recovery from anaesthesia was uneventful. At the postoperative period his venous blood sample was taken and his blood glucose and lactate levels were found 99 gr dL⁻¹, and 13 mg.dL⁻¹ (4.5-14.4 mg.dL⁻¹) respectively. The patient was infused with 10% dextrose and 0.9% NaCl for 4 hours and fed at the 2nd postoperative hour. He was discharged at the 12th postoperative hour without any other problems.

DISCUSSION

Glycogen storage diseases occur as a result of enzymatic abnormalities that lead to abnormal concentrations or structures of glycogen. In type 1a (GSD-1a), absence of glycogen-6-phosphatase is characterized with fasting hypoglycemia. As both glucose producing pathways are blocked due to this inborn error, glycogen accumulation in liver, kidney and intestine is unavoidable. Hypoglycemia and glycogen storage are the main reasons of the clinical findings and other biochemical abnormalities of the disease.¹ ³
The hypoglycemia is the major problem in the GSD-1a patients. This problem is caused by fasting before surgery. As fasting hypoglycemia is the most important problem of the disease, a short duration of preoperative fasting is recommended for such patients. Our patient experienced hypoglycemia, although we offered four hours fasting to our patient. The patient hospitalized and preoperative dextrose infusion started for the adequate control of blood glucose concentrations. Surgery is a major stress to the body and results in glucose accumulation into the blood due to the release of counter regulatory hormones in patients undergoing surgery. In regard to GSD-1a patients, the surgical stress results in glucose-6-phosphate accumulation in blood. The accumulation of this intermediate metabolite causes hypoglycemia in acute terms. The perioperative hypoglycemia can only be prevented by glucose infusion. We started infusion of dextrose solution 4 hours before induction of anesthesia. The overall stay time of the patient in the operating theatre was about 1 hour. We did not experience lactic acidosis during this period. At the postoperative period, as fasting lasted, we continued dextrose infusions for four hours. The patient was fed at the postoperative 2nd hour. We continued the dextrose infusion as soon as he tolerated his oral diet.

**References**