

Anesthesia Management Of A Patient With Glucose-6-Phosphate Dehydrogenase Deficiency

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Abstract

The most common red blood cell enzymatic defect is Glucose-6-phosphate dehydrogenase deficiency (G6PD). Hemolysis can be caused by oxidant agents during peroperative medications in patients with Glucose-6-phosphate dehydrogenase deficiency. This report presents a case of general anaesthesia management in a patient with glucose-6-phosphate dehydrogenase deficiency.

A 4 year-old male with G6PD deficiency was scheduled for inguinal hernia repair under general anesthesia. G6PD deficiency diagnosed when he was 1,5 years-old. Intraoperative and postoperative course was uneventful such as stress, hemolytic problems, malignant hyperthermia, or methemoglobinemia.

We think that general anaesthesia can be performed successfully with special attention in patients with G6PD deficiency.

INTRODUCTION

Glucose-6-phosphate dehydrogenase (G6PD) deficiency is the most common disease-causing enzyme defect in humans affecting an estimated 400 million people, but there are few publications relating patients with G6PD deficiency under going to general anesthesia (1). This X-linked inherited disorder most commonly affects persons of African, Asian, Mediterranean, or Middle-Eastern descent (2,3). Persons with this condition also may be asymptomatic. Homozygotes and heterozygotes can be symptomatic, although the disease typically is more severe in persons who are homozygous for the deficiency. The conversion of nicotinamide adenine dinucleotide phosphate to its reduced form in erythrocytes is the basis of diagnostic testing for the deficiency. This usually is done by fluorescent spot test. Different gene mutations cause different levels of enzyme deficiency, with classes assigned to various degrees of deficiency and disease manifestation. Because acute hemolysis is caused by exposure to an oxidative stressor in the form of an infection, oxidative drug, or fava beans, treatment is geared toward avoidance of these and other stressors. Acute hemolysis is self-limited, but in rare instances it can be severe enough to warrant a blood transfusion. The variant that causes chronic hemolysis is uncommon because it is related to sporadic gene mutation rather than the more common inherited gene mutation(4).

This is a report on a pediatric G6PD-deficient patient submitted to inguinal hernia repair, under general anesthesia

CASE REPORT

A 4 years old 14 kg, male patient with G6PD deficiency was admitted for left inguinal hernia repair. He had developed serious haemolytic anemia with a fall in haemoglobin to 6.5 g/dL three days after ingestion of fava beans when he was 1,5 year-old. After treatment with intravenous fluids and blood transfusions he recovered and was discharged from hospital after ten days. At 3.5 years old a left inguinal hernia repair was performed under local anaesthesia with 3 ml prilocaine 2% without any complication. Since then, after all care had been taken to prevent hemolytic crises triggering factors, the patient had a good evolution without new crises.

Before operation at the preanesthesia policlinic, no cardiovascular, respiratory, digestive and renal systems abnormalities were detected. Preoperative blood tests were as follows: red cells 4.700/mm³; Hemoglobin 12.5 g /dL , hematocrit 36.7 %, time of activated thromboplastin 89.7% and INR 1.14, platelet 341.000 mm³. His electrolytes were normal.

At the operation day after 6 hours of fast, the patient was accepted to preoperative care unit and a 24 Gauge cannula was inserted into a vein on the dorsum of left hand and crystalloid infusion was started. After 15 minutes the patient

was taken to the operating room. He was monitored with peripheral oxygen saturation (SpO₂), electrocardiogram, cutaneous temperature (T) and noninvasive blood pressure (NIBP). Anesthesia was induced by 1 µg/kg fentanyl, 2.5 mg/kg propofol, and 0.1 mg/kg vecuronium intravenously. Tracheal intubation was performed. Maintenance was achieved using sevoflurane 1 MAC in a 2: 1 nitrous oxide : oxygen mixture and intravenous vecuronium. Hemodynamic and other vital parameters were stable during intraoperative period. The duration of surgery was 45 minutes. After recovery of muscle tone and spontaneous breathing was adequate, trachea was extubated. No respiratory or hemodynamic problems occurred. The patient was directed to the Post Anesthetic Recovery Unit without pain or other complaints and he remained there for 60 minutes without interferences or medications. With stable vital signs (mean blood pressure: 90/40mmHg, heart rate 100 beats.min⁻¹, peripheral oxygen saturation 99%), he was send to the pediatric surgery clinic and had a good recovery without interferences and was discharged 24 hours after surgery.

DISCUSSION

Glucose-6-phosphate dehydrogenase deficiency, the most common enzyme deficiency worldwide, causes a spectrum of disease including neonatal hyperbilirubinemia, acute hemolysis, and chronic hemolysis. Some drugs evoke production and accumulation of toxic peroxides, cause oxidation of hemoglobin and red blood cell membrane, and the use of these kinds of drugs results in excessive hemolysis in the patients with G6PD deficiency (Table 1).

G6PD catalyzes first step of pentose phosphate metabolic pathway which is an exclusive source of NADPH in red blood cells (5). The most important role of NADPH in erythrocyte consists in regeneration of reduced glutathione, which prevents hemoglobin denaturation, preserves the integrity of the red blood cell membrane sulfhydryl groups, and detoxifies peroxides and oxygen free radicals in the red blood cells (6, 7). NADPH production is decreased in G6PD deficiency. Our patient had developed serious haemolytic anemia with a fall in haemoglobin to 6.5 g/100 ml three days after ingestion of fava beans

Figure 1

Table 1: Durugs Able to Induce Hemolysis in G6PD

Analgesics	asetylsalicylic Acid	Nitrofurantoin	Methylene blue
Acetaminophen	Vitamin K	Nalidixic acid	Acetanilid
Probenecid	Antimalaria	Primaquine	Ascorbic acid
Chloroquine	Nitrates	Pentaquine	Quinidine
Sulfonamides	Naphthalene	Sulfones	Chloramphenicol
Quinine	Quinidine		

Effects of many drugs on G6PD enzymatic activity have already been investigated (8). The anesthetics widely used in clinical practice, effect on G6PD activity lacking. Halothane was the first introduced halogenated agent and it was widely used. But its use is now declining in favor of isoflurane and other drugs. Isoflurane and sevoflurane are the most favorable volatile anesthetic drugs due to their low toxicity (9). However, the results of this study have shown that halothane has no effect on G6PD activity, but isoflurane and sevoflurane have. Because of this, the use of halothane may be appropriate in a patient with G6PD deficiency.

Additionally, the use of diazepam or midazolam, which have inhibitory effects on in vitro G6PD enzymatic activity together with isoflurane or sevoflurane may increase severity of hemolysis(10). Therefore we did not administer midazolam during preoperative and peroperative medication. But Emilio et al. (11) administered midazolam 5 mg orally and peroperative sevoflurane to a patient 2 years old with G6PD deficiency and detected no changes on vital parameters during postoperative period. Alticat et al (10) conclude that ketamine may be chosen instead of midazolam or diazepam for balanced anesthesia, as intravenous anesthetic drug, and furthermore, prilocaine should be chosen for local anesthesia if it is required.

The use of a G6PD inhibitor as general anesthetic drugs may worsen health of the patients with G6PD deficiency and may lead to fatal outcome. In conclusion, we think that the results of this case report may be useful in choosing of anesthetic drugs for use in a patient with G6PD deficiency.

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