Glucose-6-phosphate dehydrogenase (G6PD) deficiency affects approximately 400 million people over the world. The enzyme protects cells against oxidative damage by creating NADPH and reducing the glutathione level via catalyzing first step of pentose phosphate pathway. Any exposure to oxidative stress such as infections, drugs and fava bean intake as well as some metabolic conditions results in acute hemolysis in those patients. The lack of the enzyme becomes lethal for erythrocytes which do not have other metabolic pathway and oxygenation injury rapidly occurs in red cell membrane in case of contact to a triggering factor.1

Although not confirmed clinically, certain anesthetics, especially volatile agents have an in-vitro inhibitory effect on G6PD activity and may carry risk of hemolysis in G6PD-deficient patients.2 The suitable approach to prevent hemolysis during perioperative period is to suppress the stress induced by surgery with proper anesthetic regimens based on drugs that are safe and have not been shown to cause hemolytic crises. Additionally, it is crucial to observe the patients closely in postoperative period up to three days to identify any sign of drug-induced hemolysis. Because the incident may be seen in a period of one to three day after the contact of triggering factors.1

We describe a 6-year-old male child, weighing 24 kg, presented to our hospital for circumcision. He was diagnosed with G6PD-deficiency after a prolonged jaundice and anemia in his neonatal period. He has no other known diseases and found normal in physical examination. He had no previous story of exposure to any anesthetic agent. His preoperative laboratory profile was normal with a hemoglobin level of 13.1 g/dL and hematocrit 37.4%, as well as serum biochemistry within physiologic limits included serum bilirubin level. After a six hours of fasting, the child was accepted to the operating room following an oral premedication of 10 mg midazolam. The patient was monitored with ECG, pulse oximetry, non-invasive blood pressure during the procedure. Following insertion of a 22
G intravenous cannula a balanced electrolyte solution was started. The anesthetic care was achieved by a combination of intravenous ketamine 0.5 mg/kg and midazolam 0.05 mg/kg. The patient’s spontaneous breathing was maintained and supported by 2 L/min oxygen supplementation via a face-mask attached to the auxiliary fresh gas outlet of the anesthesia machine. The circumcision was performed by diathermy method after a dorsal penile nerve block with 3 ml of 0.025% bupivacaine infiltration. The intervention lasted 15 minutes and no additional anesthetic was applied till to the end of surgery. After a trouble-free recovery period the patient was transferred to the ward. The hemogram ranged in normal limits during subsequent 3 days and the patient was discharged at 72\textsuperscript{th} hours.

Anesthesiologists have to be cautious to manage patients with G6PD deficiency. First, a safe anesthetic regimen based on agents which have not been shown to cause hemolytic crises has to be chosen. Then the whole perioperative period should be normalized particularly by effective analgesia and temperature management as well as strict protection against infections. Finally the patient should be monitored up to three days after surgery to detect any hemolysis. A minor surgery in our case, a children with G6PD deficiency have been managed during circumcision by a monitored anesthesia care plan with ketamine to be known safe, and midazolam. Although previously mentioned that midazolam has an in-vitro inhibitory effect on the enzymatic activity, no documented cases were found to demonstrate a midazolam triggered hemolytic crisis in G6PD-deficient patient.\textsuperscript{2} Additionally, bupivacaine, a local anesthetic without methemoglobinemia risk was chosen for penile block. It’s already stated that lidocaine and prilocain should be avoided in those group of patients because methylene blue is ineffective in case of the methemoglobinemia in G6PD-deficient patients.\textsuperscript{3}

In conclusion, a “monitored anesthesia care” plan supported by peripheral nerve block with safe agents and close observation till the end of risky period was the keynotes of our experience with the G6PD-deficient children undergoing circumcision.

REFERENCES