Propofol infusion in an infant with glucose-6-phosphate dehydrogenase deficiency

We report the anesthetic management of 7-month-old infant with glucose-6-phosphate dehydrogenase (G6PD) deficiency undergoing tendo achilles tenotomy with safe use of propofol for induction and maintenance of anesthesia along with caudal block.

A 7-month-old male child weighting 7.6 kg had bilateral congenital talipes equinovarus and was planned for tendo achilles tenotomy and above-knee corrective casting under general anesthesia. The child was diagnosed to have G6PD deficiency along with Type 2 Arnold–Chiari malformation and developmental delay. He also had a history of lumbar meningomyelocele (MMC) repair at the age of 2 months and ventriculoperitoneal shunt surgery for hydrocephalus following MMC repair. His vitals and investigations were normal. On the day of surgery, standard monitoring was started and the infant was premedicated with glycopyrrolate (40 µg intramuscularly [IM]) and ketamine (30 mg IM) to secure the peripheral venous access. Dexamethasone (1 mg intravenously [IV]) and fentanyl (10 µg IV) were given. After preoxygenation, anesthesia was induced with propofol (2.5 mg/kg) and neuromuscular blockade was achieved with atracurium (4 mg IV) and LMA size no. 1.5 was inserted to secure the airway. Anesthesia was maintained with continuous manual infusion of propofol, 15 mg/kg/h for the first 15 min, 13 mg/kg/h for the next 15 min and 11 mg/kg/h till end of surgery, and O₂/air mixture. Before the start of surgery, caudal epidural block was given in lateral position by injecting 4 ml of 0.25% bupivacaine. Intraoperatively, he was hemodynamically stable and had good glycemic and temperature control using warm fluids and blanket. The surgery lasted for nearly 40 min, and at the end of the procedure, after reversing residual neuromuscular blockage, trachea was extubated. Postoperatively, daily hemogram was done for 3 days to detect possibility of hemolysis, which was not found and the patient was finally discharged without any complication.

G6PD deficiency is an inherited, X-linked recessive, metabolic disorder characterized by an enzyme defect in erythrocytes that predisposes to hemolysis in response to oxidative stress. The aim of the anesthetic management is to avoid the factors leading to oxidative stress and hemolysis including certain medicines (some anesthetic drugs), perioperative stress, metabolic conditions such as diabetic ketoacidosis and metabolic acidosis, hypothermia, hyperglycemia, and infection.

Among the anesthetic drugs, glycopyrrolate, halothane, fentanyl, sufentanil, tramadol, ketamine, propofol, thiopental, nitrous oxide, rocuronium, succinylcholine, neostigmine, heparin, and bupivacaine are considered as safe, whereas metoclopramide, diclofenac, lidocaine, methylene blue, prilocaine, for example, EMLA cream are considered as unsafe in G6PD deficiency. There are controversies regarding the safety of certain drugs in these patients such as alfentanil, isoflurane, midazolam, paracetamol, remifentanil, and sevoflurane. As per Altikat et al., also isoflurane, sevoflurane, and midazolam had an in vitro inhibitory effect on enzymatic activity of G6PD while no effect of halothane, ketamine, benzdiazepines, codeine/codeine derivatives, fentanyl, and propofol.
Beutler\[3\] reported that halothane has no effect on G6PD activity, but isoflurane and sevoflurane have, although there are few case reports where isoflurane and sevoflurane were safely used in G6PD deficiency patients.\[4\] Propofol is considered as safe in G6PD deficiency patient and we also used it for induction and maintenance of anesthesia in our case. The safety of use of propofol in neonates and infants is debatable,\[5\] but recent reports suggest that it can be safely used in infants.\[6\] With propofol infusion, care is to be taken as there are increased incidences of bradycardia, hypotension, and delayed awakening which are usually benign and self-limited, whereas the added advantages over conventional volatile anesthetic agents are quicker recovery, reduced nausea and vomiting, decreased postoperative delirium, and less environmental pollution. In our case in order to secure IV line, we preferred IM ketamine over EMLA cream as its constituent prilocaine which is unsafe and it takes long time. We used caudal block with bupivacaine 0.25% for intraoperative and postoperative analgesia which will not only decrease the overall anesthetic and analgesic requirement but also decrease the stress response of surgery. Lidocaine should be avoided in these patients.

To conclude, avoidance of oxidative stress along with the drugs causing it and better postoperative analgesia using regional techniques plays an important role in preventing the complications in G6PD deficiency patients.

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Conflicts of interest
There are no conflicts of interest.

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