European Review for Medical and Pharmacological Sciences 2014; 18: 3347-3348

Letter to the Editor

# Anaesthesia management in a child with metabolic myopathy

*Dear Editor,*

Muscle contraction requires energy in the form of adenosine triphosphate (ATP), which is supplied through glycogen, glucose, and fatty acids metabolism. The metabolic pathways involving all these three energy sources converge into acetyl coenzyme A, which is oxidized in the mitochondria through the Krebs cycle and respiratory chain to ATP. In patients with metabolic myopathy, either defects of substrate use or disorders of lipid metabolism[1] lead to progressive dysfunction in tissues with a high energy requirement such as the brain or muscles[2]. In addition to muscular weakness and rhabdomyolysis, other manifestations including hypoglycemia, acidosis, cardiac conduction defects, cardiomyopathy and cardiac failure may also occur. The clinical picture can be complicated by hepatic and renal failure in severe cases. Despite the lack of definitive data regarding the incidence and prevalence of these disorders, 1 out of 4000 children is thought to be affected[3].

In an 8-year old male patient with a bodyweight of 22 kg adeno-tonsillectomy was planned due to recurrent occurrence of upper respiratory tract infections and presence of adeno-tonsillary hypertrophy. The patient was diagnosed with metabolic myopathy approximately 1 year ago after the development of gait disorder and difficulty in climbing stairs. He was currently receiving treatment with riboflavin (100 mg bid), and coenzyme Q10 (100 mg bid). Except for congenital ptosis of the right eye, the physical examination was unremarkable. Routine laboratory test results were within the normal ranges. The creatine kinase level measured 2 months before surgery was 326 U/L (normal range: 24-195 U/L), while the most recent value prior to surgery was 156 U/L. Due to possible risk of postoperative muscular weakness, an intensive care unit bed was allocated to him before the surgery, and he was instructed to take his medications in the morning of surgery as prescribed. His risk status according to ASA classification was ASA III. During 6 hours of pre-operative fasting, he was given a total of 500 ml of 5% dextrose solution by infusion. Also, pre-operative oral carbohydrate solution was given orally 2 and 4 hours before surgery. He was given the priority of being the first patient to be taken to the operation theatre on the day of surgery. Routine monitorization was performed in the surgery room (ECG, NIBP, $SpO_2$ and body temperature). After anaesthesia induction with sodium thiopental 150 mg and rocuronium 10 mg, endotracheal intubation was performed with a spiral endotracheal balloon tube of 5-mm internal diameter. Anaesthesia was maintained with propofol (10 mg/kg/h followed by 5 mg/kg/h and 3 mg/kg/h), remifentanyl (1 mcg/kg/h) and nitrous oxide (50%). During surgery, 5% dextrose solution admixed with a solution containing 1/3 of dextrose was infused. Following 20 minutes of surgery, sugammadex (50 mg) i.v. was administered to antagonize rocuronium. The patient regained consciousness uneventfully and was taken to recovery room, where his glucose was 185 mg/dl and lactate was 2.6 mmol/L (range: 0.5-2.0). Postoperatively, oral food intake was commenced at 2 hours and infusion with 5% dextrose solution was continued until adequate oral intake was achieved. At postoperative 16 hours, his creatinine kinase was 152 U/L. He was discharged without any complications at postoperative 24 hours.

Fasting, stress and hypothermia are among the principal factors responsible for the aggravation of metabolic myopathy[4,5]. Thus, this patient was given the priority to be operated as the first patient on the day of surgery and he was orally supplemented until 2 hours before the surgery and he received intravenous glucose support perioperatively. The use of i.v. solutions containing lactate was avoided. Also the body temperature was closely monitored during surgery to avoid hypothermia.

Since all anaesthesia agents have an impact on mitochondrial functions, no anaesthesia agent can be considered ideal for these patients. Similarly, due to accumulating evidence suggesting a link between the propofol infusion syndrome and mitochondrial function, care must be practiced during propofol infusion in these patients[2]. Therefore, in this patient sodium thiopental was preferred for anaesthesia induction, while remifentanyl and nitrogen protoxide were added to propofol for anaesthesia maintenance, due to the absence of effects of these two agents on oxidative phosphorylation, allowing the reduction of propofol dose.

*Corresponding Author:* Ayse B. Ozer, MD; e-mail: abelinozer@gmail.com

The association between metabolic myopathy and malignant hyperthermia is not well defined[4,5]. Therefore, succinylcholine and volatile anaesthetic agents were avoided. Since succinylcholine was not used, rocuronium was preferred for muscular blockade, which was antagonized by sugammadex.

Metabolic myopathy is a rare but manageable condition. A meticulous preoperative care will allow successful anaesthesia management in these patients.

## Conflict of Interest

The Authors declare that there are no conflict of interests.

## References

1) Urban MK, Lahlou S. Muscle disease. Fleisher LA. Anesthesia and uncommon disease. 5th edition. Philadelphia, Saunders Elseiver 2006; pp. 303-327.
2) Lammers C, Wyckaert C. Anesthesia for orthopedic surgery. Gregory GA, Andropoulos DB. Gregory's pediatric anesthesia. 5th edition. Oxford, Wiley- Blackwell 2012; pp. 757-776.
3) Dekker E. Anaesthesia and the paediatric patient with neuromuscular disease. S Afr J Anaesthesiol Analg 2010; 16: 18-21.
4) http://www.mitoaction.org/blog/anesthesia-risks-precautions-and-recommendations-mitochondrial-disease-patients
5) Zhou J, Allen PD, Pessah IN, Naguib M. Neuromuscular disorders and malignant hyperthermia. Miller RD (ed), Miller's Anesthesia. 7th edition. Philadelphia, Churchill Livingstone Elseiver 2010; pp. 1171-1195.

A.B. Ozer, I. Demirel, T. Gokdemir, O.L. Erhan
Department of Anesthesiology and Reanimation,
Firat University Faculty of Medicine, Elazig, Turkey