




Dental Treatment of a Child with Mitochondrial Myopathy Under General Anesthesia

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ABSTRACT The present case reports the dental treatment procedure under general anesthesia in a patient with mitochondrial myopathy. Mitochondrial diseases that cause prominent muscular problems are called mitochondrial myopathies whereas mitochondrial diseases that causes both prominent muscular and neurological problems are called mitochondrial encephalomyopathies. It is known that all general anesthetic agents directly inhibit the mitochondrial function and may cause perioperative complications. In the present case, general anesthesia was implied for dental rehabilitation of the severe early childhood caries which occurred due to inadequate dietary habits and ineffective oral hygiene practices. General anesthesia was maintained with intravenous propofol infusion (2 to 4 mg/kg/hour). This protocol is selected in order to avoid the mitochondrial suppression. The authors would like to underline important points regarding dental rehabilitation under general anesthesia in patients with mitochondrial myopathy.

Keywords: Anesthesia, general; dental care for children; mitochondrial myopathies

Mitochondrial myopathies (MM) are uncommon disorders caused by structural and functional abnormalities in mitochondria due to inactivation of the mitochondrial chain of respiration or oxidative phosphorylation.^{1,2} The condition occurs with an incidence of 1 per 4-5000 live births.³ Primary “target” organs are the central nervous system and the liver but the disorder may account for a wider range of neurologic, endocrine disorders as well as cardiac complications.

The major problems connect to mitochondrial disease are low energy, free radical production and lactic acidosis, which can result in a variety of symptoms in many different organs of the body.^{2,3} The precursor symptoms of the disorder include heart failure, muscle fatigue or rhythm disturbances, dementia, movement disorders, stroke-like episodes, deafness, droopy eyelids, blindness, limited mobility of the eyes, vomiting, and seizures. The severity of any of these symptoms varies greatly from person to person, even in the same family.

It is known that all general anesthetic agents directly inhibit the mitochondrial function and may induce perioperative complications.⁴ Thus, patients with mitochondrial disease are considered to be at special risk for general anesthesia with a main concern of malignant hyperthermia or rhab-

domyolysis that may result in perioperative morbidity or mortality.⁵ On the other hand, these patients often require general anaesthesia (GA) as part of their diagnostic work up, subsequent management or other invasive treatments such as dental care.

There are several reports in the literature of this type of disease tolerant to a wide range of anaesthetics including volatile anaesthetics, propofol and local anaesthetics.^{6,7} In 2013, Gentili et al. published a letter to the editor in which they described a severe case of mitochondrial myopathy (MELAS syndrome: mitochondrial myopathy, encephalopathy, lactic acidosis, and stroke-like episodes) that was operated for combined colectomy and ovariectomy under laparotomy with an intra-operative time of 5 hours.⁷ The operation was performed under general anaesthesia in combination with postoperative epidural anaesthesia. Yet, the evidence base for the use of GA is limited and inconclusive in patients with mitochondrial myopathies.⁵

CASE REPORT

The present case report of a male patient, aged 4.5 years, with mitochondrial myopathy and early childhood caries (ECC) who was referred for dental treatment to the Department of Paediatric Dentistry of the Baskent University. The patient was non-cooperative but requiring dental care. On account of the requirement for extensive dental treatment, along with his medical condition, treatment was planned to be accomplished under GA.

The patient had no systematically disorder except for myopathy. He had truncal deformities such as scoliosis, flexion contractures but no clinically important facial deformity. His mouth opening was normal and Mallampati score was class 1. His hemoglobin level was 11,8 g/dL, serum electrolytes, glucose level, organ function tests, coagulation profile, and platelet level were normal.

After 6 hours without food or drink, he was premedicated with oral 5 mg midazolam and taken to the operating room. Standard monitoring was performed with 5-lead electrocardiogram, pulse

oximeter and noninvasive blood pressure measurement. Anaesthesia induction was performed with inhalational sevoflurane 8% in 50/50 % air/oxygen mixture, then 22G intravenous line inserted from left arm. Antibiotic prophylaxis with cefazoline sodium (25 mg/kg) was administered before administration of intravenous anaesthetics of propofol (2 mg/kg), fentanyl (1 µg/kg), and midazolam (1 mg/kg). Then, laryngeal mask size of 2.5 was inserted to hypopharynx (Figure 1). Anaesthesia was maintained with intravenous propofol infusion (2 to 4 mg/kg/hour) and sevoflurane was discontinued. During the operation local anaesthetics applied to surgical site by the pediatric dentists. The patient was hemodynamically stable throughout and a warming system was applied with no changes in temperature or electrocardiogram.

A combination of restorative approaches and techniques was used, including the placement of composite resin (3M, Espe), pulpotomy with mineral trioxide aggregate (Proroot MTA, Dentsply) and several extractions in the primary dentition. Overall, surgery lasted 70 minutes. After a successful operation, the patient was extubated in the operating room and transferred to outpatient ward.

DISCUSSION

The inheritance of mitochondrial diseases is complicated, and often it is difficult to follow the condition through a family tree. In fact, many cases of mitochondrial disease are sporadic. The parents of



FIGURE 1: The view of the laryngeal mask without masking the operation site.

the presented case were in good health and the family history was non-contributory. A thorough literature search revealed only limited data regarding the dental treatment of patients with MP. Up to date, there is only one case presenting endodontic treatment in a 10 year old boy with MM under deep sedation via total intravenous anesthesia.⁸ The authors selected this protocol in order to avoid the mitochondrial suppression that may be induced with inhalation agents.

The patient described typical symptoms of mitochondrial myopathy, including gradual paralysis of eye movements. Mitochondrial myopathies can cause weakness in muscles of the face and neck, which can lead to difficulty with swallowing and, more rarely, slurred speech. In the present case, the patient had a minor difficulty in swallowing and he did not develop speech. The parents were feeding him mostly with pureed food rich in starch and sugar. This in turn, resulted in early development of carious lesions. Due to the lack of cooperation and the need for extensive dental treatment, dental rehabilitation under general anesthesia (DRGA) was planned after consulting the physician of the patient. The other main reason for deciding DRGA was the inability of the patient to maintain a patent airway due to muscle weakness. He could not sit alone, control his head, move his extremities, could not stand up or walk.

In the present case, none of the possible complications such as myocardial conduction disturbances, postoperative muscle hypotonia, malignant hyperthermia was observed. Postoperative care is also important in these patients. Individuals with mitochondrial disease generally should sunef valproic acid, statins, aminoglycoside antibiotics, and erythromycin.⁸ Thus, the surgeon should evaluate the cost and benefits of using postoperative antibiotics.

It should be noted that these patients must be monitored more closely than other patients when operating under GA. It is very important to titrate the induction drugs slowly. In addition, cautious documentation is needed to be sure that the effects of the anesthetics are largely washed-out before assuming that the patient can ventilate sufficiently. On the other hand, some patients with mitochondrial disorders are unable to metabolize lactate; and it is best to avoid the addition of lactate to their fluids.⁸

Informed Consent

Authors assure that the case presented herein was reported upon permission of the patient and his parents.

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Conflict of Interest

No conflicts of interest between the authors and / or family members of the scientific and medical committee members or members of the potential conflicts of interest, counseling, expertise, working conditions, share holding and similar situations in any firm.

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