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Letter to Editor

Short-Time Anesthesia Management for Duchenne Muscular Dystrophy

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Duchenne muscular dystrophy (DMD) is a rare X-linked inheritance disorder that due to the deficient of dystrophin. Dystrophin gene is located on chromosome Xp21and plays an important role in sarcolemmal stability and muscle membrane integrity. The hallmark of the disorder is progressive muscles dysfunction that commonly leads to respiratory failure [1-3].

Occasionally, one of the challenges for anesthesiologists is when a short, simple operation is to be performed on a patient with a rare and complex underlying disease. In these cases, on the one hand, the surgeon expects the patient to be anesthetized quickly and easily due to the simplicity of the surgery, and on the other hand, despite the simplicity of the surgery, the anesthesiologist faces many challenges in managing these patients. Additionally, case reports or reference texts usually mention to the management of these patients in difficult and complex surgeries, and we rarely find anesthesia management of these patients for simple and short surgeries.

Therefore, we report a 15- year-old boy who diagnosed with DMD in the early childhood time was planned for femoral shaft fracture repair. He was in a wheelchair. The physical examination was normal except macroglossia (Figure 1). All routine laboratory investigations were normal and potassium level was 4 mEq/liter. The electrocardiogram revealed sinus tachycardia and right bundle branch block (Figure 2). In the operating room, standard monitoring was attached and vital signs were: HR 127 bpm, BP 101/61 mmHg, and SpO2 98%. Initially, anesthetic gas vaporizer demounted from the anesthesia machine and the soda lime was changed. The anesthesia machine was flushed with oxygen about 20 minutes. Following pre-oxygenation, IV fentanyl 2 µg/kg and propofol 1.5 mg/kg were administered. During procedure, he received 100% oxygen via the oxygen facemask throughout spontaneous breathing. The operative course (less than 10 minutes) was uneventful and he was completely awakened (less than 10 minutes) later without any problems. DMD is commonly the most severe type of muscular dystrophy that is characterized by fast progressive muscle weakness. Symptoms usually appear the ages of 2 to5. Some of clinical symptoms and signs are macroglossia (cause of difficult tracheal intubation), waddling and toe walking, gowers sign (using hands to climb up thighs to stand), pulmonary dysfunction (due to progressive truncal and respiratory muscle weakness), highly susceptible to infection, and cardiac dysfunction due to degeneration of cardiomyocyte and fibrosis. The characteristic electrocardiographic abnormality usually is sinus tachycardia. Chronic respiratory insufficiency generally indicates increasing weakness of respiratory muscles and loss of diaphragmatic strength. They are also at risk for life-threatening hyperkalemia, and malignant

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hyperthermia-like reaction due to inhalational anesthetics and succinylcholine. Therefore, these medications are better to be avoided. Furthermore, it is recommended to demount gas vaporizers from the anaesthesia machine and washout the machine by flushing with oxygen 100% to decontamination of halogenated compounds before induction of anesthesia. Propofol, opiates, ketamine, dexmedetomidine, and midazolam have been used successfully for them. Benzodiazepines and opiates (due to respiratory insufficiency), and nitrous oxide in patients with cardiomyopathy should be used with caution. If the surgery does not require muscle relaxation, it is better to avoid muscle relaxants. Although most of anesthesiologists recommend total intravenous anesthesia, there is no evidence of specific anesthesia management for them [1-4]. In our case, since the procedure was supposed to take less than a few minutes (< 10 minutes) and propofol has been used in DMD without major adverse effects (such as unexplained fever or rhabdomyolysis), it was decided to use it. Propofol in combination with fentanyl was injected titrated to avoid hemodynamic and myocardial impairment.

Finally, this case showed that short time anesthesia with combination of propofol and fentanyl could be feasible and safe in disabled DMD patients.

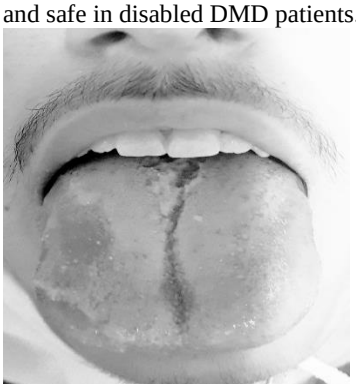

Figure 1- Macroglossia with mallampati grade 4 score in the boy with DMD

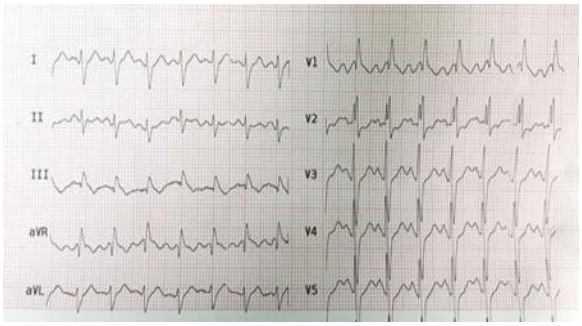

Figure 2- ECG showing RBBB (QRS duration greater than 120 milliseconds, rsR "bunny ear" pattern in the anterior precordial V2 lead, Slurred S waves in leads I, aVL and V5), sinus tachycardia and right axis deviation.

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