

Duchenne Muscular Dystrophy: Management of Difficult Airway and Concurrent Bronchospasm

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Abstract

Using succinylcholine and inhalation agents for patients with Duchenne muscular dystrophy is extremely risky. Those risks include heart failure, cardiac dysrhythmias, rhabdomyolysis and malignant hyperthermia. Even in emergent situations, such as intraoperative bronchospasm, succinylcholine and inhalational agents are often considered contraindicated. Nevertheless, if intraoperative bronchospasm does occur in a patient with Duchenne muscular dystrophy, the benefits of using inhalational agents to treat bronchospasm may outweigh the risks in certain situations.

Keywords: Duchenne muscular dystrophy; Volatile anesthetics; Rhabdomyolysis; Difficult airway management; Bronchospasm

Introduction

Duchenne muscular dystrophy (DMD) patients often develop aspiration and inability to clear airway secretions effectively due to respiratory muscle weakness [1]. This quite often leads to pneumonia and further acute respiratory failure requiring emergent intubation. Due to commonly limited mouth opening, neck immobility and cervical spine deformation, DMD patient often present with difficult airway [2]. DMD patients are also known for compromised cardiopulmonary function and possible abnormal reactions to succinylcholine and volatile anesthetic agents.

Case Study

33 yr old male with PMH of DMD, chronic obstructive pulmonary disease, cardiomyopathy, and gastroesophageal reflux with spontaneously developed supraventricular tachycardia (SVT) and acute respiratory distress presented for emergent intubation. On airway exam, the patient's thyromental distance was less than two fingerbreadths; uvula was not visible on mouth opening. His neck appeared severely contracted and not capable of more than 30 degrees of extension or flexion. The patient was saturating at 84% on a facemask with 6 liters per minute oxygen flow, his heart rate was about 150 beats per minute and his blood pressure was approximately 90/60. Patient's chest auscultation revealed bilateral equal breath sounds with coarse rhonchi and crepitations. No recent chest X-ray was available at that time. The patient had a known difficult airway with a previous history of retrograde intubation done two months prior at an outside institution with no further details provided. We decided to perform an emergent awake fiberoptic intubation with ENT surgeons standing by for possible cricothyroidotomy. Because the patient appeared already severely lethargic from hypoxia, no sedatives were given prior to performing fiberoptic intubation. A superior laryngeal nerve block was done with bilateral injections of 2.5 cc of 1% Lidocaine at the level of the greater cornu of the hyoid bone. A transtracheal block was done by injecting 3 cc of 4% Lidocaine through the cricothyroid membrane. Fiberoptic camera was inserted orally until cricoids rings were visualized after passing the vocal cords. A 6.0 endotracheal tube placements was confirmed by visualization of the carina and by positive end-tidal carbon dioxide tracing. No tracheal or bronchial damage was noted during bronchoscopy. Soon after intubation, end-tidal carbon dioxide tracing on capnogram disappeared and bilateral breath sounds were completely absent. Rapid arterial oxygen desaturation with SpO₂ of 55%-60% followed. A dramatic increase in the

peak inspiratory pressures greater than 45 cm H₂O was noted and bag ventilation commenced. The endotracheal tube placement was confirmed once again via fiberoptic scope with carina in view. A presumed diagnosis of bronchospasm was made and ipratropium and albuterol were administered via the endotracheal tube. Ipratropium and albuterol were chosen for they were readily available in the operating room and easy to administer via nebulizer. Intravenous terbutaline could have been a reasonable alternative. Due to the preexisting SVT and terbutaline's significant protachyarrhythmic properties, terbutaline was avoided. No significant improvement was noted after nebulized ipratropium and albuterol was given. At that time, we proceeded to administer sevoflurane inhalation to break the intractable bronchospasm which resolved about two minutes after 1% sevoflurane inhalation was started. This was evidenced by the return of oxygen saturation to >90% and by adequate capnogram tracing. Ventilation became easier to perform along with the return of audible breath sounds over both lung fields. The use of sevoflurane raised significant concerns in regards to possible development of malignant hyperthermia and rhabdomyolysis. We followed post intubation CPK and potassium levels which appeared to be in normal range. Post intubation portable chest X-ray was of poor quality and consistent with blunting of the left costophrenic angle indicating pleural effusion or thickening (Figure 1). Breath sounds remained equal and bilateral and the endotracheal tube placement was reconfirmed again with fiberoptic carina visualization. No endobronchial tree damage was noted on follow up bronchoscopy.

Discussion

Our case deals with management of acute bronchospasm in a patient susceptible to inhalational agents and depolarizing muscle relaxant. Bronchospasm encountered during the perioperative period and especially after induction and intubation may involve an immediate hypersensitivity reaction. To prevent reflex bronchoconstriction caused

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Figure 1: DMD Case.

by endotracheal intubation induced vagal reflex, inhaled β_2 -agonists and anticholinergics are often administered as adjunct medications [3]. Preoperative treatment with combined corticosteroids and β_2 -agonists minimizes intubation-evoked bronchoconstriction [2] compared to β_2 -agonist alone. Nebulized racemic epinephrine can also effectively relax smooth muscle, but side effects of tachycardia and flushing can be seen. In our case we were reluctant to administer epinephrine since our patient was already in SVT with heart rate in the 150's. Ketamine also could be considered for its bronchodilator properties. However, due to significant sympathomimetic effects of ketamine in a presence of SVT, we decided to avoid it [4].

Use of inhalational agents in the treatment of severe bronchospasm is well advocated. With exception of desflurane, which at high alveolar concentrations has been shown to increase airway resistance, volatile anesthetic agents produce reliable bronchodilatation [5]. Cases of rhabdomyolysis and intraoperative cardiac arrest secondary to hyperkalemia during the use of inhaled anesthetics in patients with DMD are described [6,7]. Although, DMD patients are unlikely to have an increased risk of malignant hyperthermia, exposure to volatile anesthetics may be associated with life-threatening rhabdomyolysis and therefore should be used cautiously and when the benefits of their use outweigh the possible risks. Most retrospective reports on the anesthetic management of patients with DMD attest to the safe use of inhaled volatile anesthetics without succinylcholine and there is no clear evidence to pinpoint inhalational anesthetics as a culprit for malignant hyperthermia and rhabdomyolysis in DMD patients [8].

Using inhalational anesthetics in DMD patients to relieve bronchospasm is reasonable except, possibly, halothane which is associated with cardiac bradyarrhythmias. DMD patients have a propensity for cardiac arrhythmias due to dystrophic involvement of the myocardium [9]. This latter complication quiet often exacerbates postoperatively and volatile anesthetics should be used extremely cautiously; the benefits of their use should outweigh possible risks. Total intravenous anesthesia is obviously a preference to maintain anesthesia in a patient with DMD [2].

Succinylcholine administration is associated with life threatening hyperkalemia and should be avoided in patients with DMD. In one review study a literature search including Pub Med, Medline, OVID, yielded seven cases of rhabdomyolysis and intraoperative cardiac arrest secondary to hyperkalemia during the use of inhaled anesthetics in patients with DMD [8]. A recent large scale retrospective study did not find an increased risk of malignant hyperthermia susceptibility

in patients with DMD compared with the general population [10]. It is generally agreed that depolarizing muscle relaxants should not be used in DMD patients, but there is no strong evidence to confirm that volatile anesthetics are absolutely contraindicated. The risks related to anesthesia and sedation for patients with DMD include heart failure, cardiac dysrhythmias, and the consequences of rhabdomyolysis as leading causes of morbidity and mortality, but most probably not malignant hyperthermia. Preoperative administration of corticosteroid and β_2 agonist serves to minimize the risk of bronchospasm. If bronchospasm does occur in a patient with DMD, the benefit of inhalational agent outweighs the complication of hypoxia secondary to bronchospasm.

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