

Dexmedetomidine and Duchenne Muscular Dystrophy

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Introduction: Duchenne muscular dystrophy (DMD) is an x-linked myopathy resulting in progressive muscle weakness, cardiac dysfunction and ventilatory failure. Nocturnal hypoventilation, sometimes associated with obstructive sleep apnea (OSA), is a predictor of impending respiratory failure (1). Death occurs by the second decade of life from respiratory failure in more than 70% of cases (2). We report the use of dexmedetomidine for sedation on a patient with DMD, severe OSA, hypercapnic respiratory failure and history of difficult intubation undergoing percutaneous gastrostomy (PEG).

Case report: A 22 year old male with Duchenne Muscular Dystrophy (DMD) was admitted to the pediatric intensive care unit for severe hypercapnic respiratory failure. Venous blood gases on admission showed a pCO₂ of 123 and a pH of 7.1. The application of nasal CPAP improved his respiratory status and avoided mechanical ventilation. Past medical history was significant for mild left ventricular dysfunction, necrotizing pneumonia with residual bronchiectasis, severe OSA requiring nocturnal auto-CPAP and progressive dysphagia with malnutrition (weight, 36 Kg). In addition, the patient had type I Von Willebrand disease and history of difficult intubation during a prior general anesthetic that required multiple attempts of fiberoptic intubation. Five days after admission, the patient was scheduled for percutaneous gastrostomy (PEG) to improve his nutritional status. Considering the history of difficult intubation and the potential for difficult weaning from mechanical ventilation in patients with DMD, it was decided to do the procedure under conscious sedation. Dexmedetomidine was chosen to provide patient sedation and analgesia with minimal respiratory depression. In the OR, nasal CPAP was continued, and electrocardiogram, oxygen saturation (SaO₂), respiratory rate (RR), blood pressure (BP) and heart rate (HR) were monitored. Equipment for difficult intubation was available. To provide fast anxiolysis and amnesia 1mg of midazolam was administered. A loading dose of 1µg/kg of dexmedetomidine was given over 15 minutes and a maintenance infusion of 0.6 µg/kg/hr was started. After patient sedation, the hypopharynx was topically anesthetized with 4% lidocaine and the endoscope was advanced without difficulty. Local infiltration of the abdominal incision was done with 1% lidocaine. The gastric tube advancement was difficult requiring additional sedation with 1mg of midazolam and an increase of the dexmedetomidine infusion to 0.8 µg/Kg/ hr. To improve analgesia 50 µg of fentanyl were given. The patient tolerated the procedure well, was arousable and cooperative and tended to sleep during periods of minimal stimulation. No airway obstruction or changes in RR or SaO₂ were noted. BP and HR remained at baseline levels and only a brief increase in HR was noted during the enlargement of the abdominal incision. At the end of the procedure the dexmedetomidine infusion was discontinued and the patient remained awake and in stable condition. There were no complications or side effects during the postoperative period.

Discussion: There are multiple concerns for the perioperative care of patients with DMD. Succinylcholine is associated with rhabdomyolysis and hyperkalemic cardiac arrest in patients with diagnosed or undiagnosed DMD. There are also numerous case reports of rhabdomyolysis when inhalational anesthetics alone were used in patients with DMD. Some authors recommend that volatile agents be avoided in these patients, especially now that intravenous anesthetics are readily

available (3). Conscious sedation can be provided with opioids, benzodiazepines, barbiturates or propofol, but all of these pharmacologic agents have the potential for respiratory depression and airway obstruction. These side effects can be life threatening in a patient with limited respiratory reserve and a difficult airway. Dexmedetomidine is a highly selective α_2 adrenergic agonist with sedative, analgesic and opioid sparing effects and is not associated with respiratory depression at therapeutic doses (4, 5). At higher doses, dexmedetomidine decreases the respiratory response to CO_2 and produces a rightward shift of the CO_2 response curve (6). However, a recent report showed preservation of respiratory drive when it was used in high doses as a total intravenous anesthetic (7). In this report, the use of dexmedetomidine as the primary sedative agent allowed us to minimize the use of opioids and benzodiazepines and provided effective sedation on a DMD patient with limited respiratory reserve and a difficult airway during a painful procedure. This is to our knowledge the first report of the safe use of dexmedetomidine in DMD.

- References:**
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