Rationale of using sugammadex in a patient with myotonic dystrophy

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Introduction

- Myotonic dystrophy (DM) is an autosomal dominant inherited disorder of the sarcolemma, resulting in abnormal responses to neuromuscular blocking agents (NMBAs).
- Acetylcholinesterase inhibitors such as neostigmine, are contraindicated in these patients due to their myotonic effect.
- Sugammadex is a novel pharmacologic agent, which encapsulates rocuronium or vecuronium to reverse their effect.
- We report our experience with using sugammadex to reverse NMBAs in a patient with DM.

Case Report

- 25-year-old male, initially admitted for cardiothoracic surgery, with progressive aortic valve insufficiency and DM type 1.
- Examination revealed symmetrical facial weakness, ptosis with temporal balding, and upper and lower limb muscle weakness.
- ECG showed a heart rate of 54 bpm, intraventricular conduction delay, LVH, and early repolarization.
- Echocardiogram showed an abnormal bicuspid aortic valve and aortic insufficiency.
- The patient was NPO for 6 hours and transported to the OR where ASA monitors were placed.
- A peripheral intravenous catheter was placed and a RSI with midazolam, fentanyl, etomidate, propofol, rocuronium, and lidocaine, intubation was uncomplicated.
- Maintenance anesthesia was provided with desflurane, dexmedetomidine, and fentanyl.
- Cardiopulmonary bypass time was 3:10 hours.
- Neuromuscular blockade was reversed with sugammadex (4 mg/kg).
- The patient's trachea extubated awake in the OR.
- The patient returned in 5 weeks for TEE and cardioversion due to atrial flutter with depressed myocardial function.
- He was NPO for 6 hours and ASA monitors were placed in the procedure area. RSI with endotracheal intubation using etomidate, fentanyl, and rocuronium. Maintenance anesthesia was provided with desflurane.
- Following electrical cardioversion, the patient returned to normal sinus rhythm.
- 45 minutes after the single dose of rocuronium, sugammadex (4 mg/kg) was administered.
- Extubation was successful without residual paralysis.

Discussion

- Perioperative morbidity in patients with DM may be due to respiratory failure, upper airway obstruction, cardiac failure, arrhythmias, or aspiration.
- Given the potential for exacerbating myotonia, choice of NMBA and reversal agents may impact perioperative course.
- Sugammadex allows reversal of NMBAs in patients with DM without exacerbating myotonia.
- In our patient, sugammadex (4 mg/kg) effectively reversed neuromuscular blockade and allowed early tracheal extubation.

Table 1: Previous reports of sugammadex use in patients with myotonic dystrophy

Author & reference	*Patient demographics*	*Outcome & anesthetic care*
Pickard A et al.[11]	14-month-old child with myotonic dystrophy for endoscopic gastrostomy tube placement, orchidopexy, and division of tongue tie.	General anesthesia with sevoflurane and fentanyl. Rocuronium (0.8 mg/kg) to facilitate endotracheal intubation. Fifty-seven minutes after rocuronium, no response of the TOF. Sugammadex (5 mg/kg) resulted in a TOF ratio of 96%. However, this fell to 60% and a second dose of sugammadex (5 mg/kg) was administered. The TOF ratio was 86% and remained stable. The patient's trachea was extubated.
Stourac P et al.[12]	37-year-old woman with myotonic dystrophy for Cesarean section.	Anesthetic induction and endotracheal intubation with propofol (2 mg/kg) and rocuronium (1 mg/kg). At the completion of the procedure (50 minutes), there were no twitches. Sugammadex (2 mg/kg) resulted in a TOF of 0.9 in 2 minutes and the patient's trachea was extubated.
Petrovski J et al.[13]	43-year-old woman with myotonic dystrophy for cystoscopy and colonoscopy.	Anesthesia was induced with sevoflurane in oxygen supplemented with propofol. Endotracheal intubation was facilitated by rocuronium (50 mg). Although there was no residual neuromuscular blockade on the TOF, sugammadex (200 mg) was administered to ensure complete reversal. Her trachea was extubated without problems.
Baumgartner P et al.[14]	59-year-old man with myotonic dystrophy for elective laparoscopy.	Anesthesia was induced with alfentanil (1 mg) and propofol (100 mg). Endotracheal intubation with rocuronium (30 mg). Surgery was completed 46 minutes after rocuronium. No twitches on the TOF. Sugammadex (2 mg/kg) was administered and four minutes later, TOF revealed four equal twitches. Within 10 minutes, the patient's trachea was extubated.
Matsuki Y et al.[15]	24-year-old woman with myotonic dystrophy for laparoscopic ovarian cystectomy.	Anesthesia was induced and maintained with propofol and remifentanil. Repetitive TOF stimulation was applied followed by rocuronium (0.3 mg/kg) for endotracheal intubation. At the completion of surgery, T2 of the TOF was present and sugammadex (2 mg/kg) was administered. Within two minutes, the TOF ratio was 0.9. The patient's trachea was extubated.
Gurunathan U et al.[16]	60-year-old, 70 kilogram man with myotonic dystrophy for elective laparoscopic cholecystectomy.	Anesthesia was induced with midazolam and propofol. Endotracheal intubation was facilitated by rocuronium (50 mg). After 45 minutes, there were no twitches on the TOF. Sugammadex (200 mg) was administered and within 30 seconds, there were 4 twitches without fade and the patient's trachea was.
Mavridou P et al.[17]	40-year-old, 74 kilgoram woman with myotonic dystrophy for laparoscopic cholecystectomy and ovarian cystectomy.	Anesthesia was induced with propofol (2 mg/kg) and rocuronium (30 mg). TOF was applied and endotracheal intubation was facilitated by rocuronium (30 mg). Anesthesia was maintained with propofol and remifentanil. After 90 minutes, there was a reappearance of T2 on TOF. Sugammadex (2 mg/kg) was administered and within 2 minutes, the TOF was 1. Opioid reversal was necessary, but the trachea was successfully extubated.