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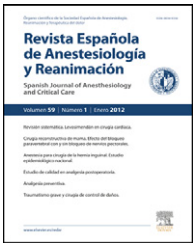

CASE REPORT

Sugammadex reversal of rocuronium-induced neuromuscular blockade in two types of neuromuscular disorders: Myotonic dystrophy and spinal muscular atrophy

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Rocuronium;
Sugammadex;
Residual neuromuscular blockade

Abstract Neuromuscular disorders like myotonic dystrophy (dystrophia myotonica or Steinert's disease) and spinal muscular atrophy are associated with perioperative complications related to muscle weakness. These patients have an increased sensitivity to non-depolarising neuromuscular blocking agents, which can lead to postoperative residual curarization (PORC) and its associated respiratory complications. Adequate reversal of neuromuscular blockade is essential to prevent this. Sugammadex is the first selective relaxant binding agent and it reverses rocuronium- and vecuronium-induced neuromuscular block. Two cases are reported in which the patients received sugammadex to reverse a rocuronium-induced neuromuscular block. Reversal of the rocuronium-induced neuromuscular block (NMB) in both cases was fast, effective and without recurarization, and no safety concerns were observed.

PALABRAS CLAVE
Trastornos neuromusculares;
Distrofia miotónica;
Atrofia muscular espinal;
Rocuronio;
Sugammadex;
Bloqueo neuromuscular residual

Sugammadex antagoniza el bloqueo neuromuscular provocado por rocuronio en dos tipos de trastornos neuromusculares: distrofia miotónica y atrofia muscular espinal

Resumen Las enfermedades neuromusculares como la distrofia miotónica (o enfermedad de Steinert) y la atrofia muscular espinal se asocian con las complicaciones perioperatorias relacionadas con la debilidad muscular. Estos pacientes presentan una hipersensibilidad a los bloqueantes neuromusculares no despolarizantes que podría derivar en curarización residual postoperatoria con complicaciones respiratorias. Para evitarlo conviene antagonizar satisfactoriamente el bloqueo neuromuscular (BNM). Sugammadex es el primer relajante selectivo y antagoniza los bloqueos neuromusculares por rocuronio y vecuronio. Se notifican dos casos donde los pacientes recibieron sugammadex para antagonizar un bloqueo neuromuscular

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provocado por rocuronio. El antagonismo de los BNM por rocuronio en ambos casos fue rápido, eficaz y sin recurarización, no se observaron preocupaciones de seguridad.

Introduction

Myotonic dystrophy, also referred to as dystrophia myotonica or Steinert's disease (SD) and spinal muscular atrophy (SMA) are progressively disabling neuromuscular disorders which are challenging to the anesthesiologist. SD is an autosomal dominant trinucleotide repeat disorder caused by a mutation on chromosome 19 and affects the gene which codes for myotonic dystrophy protein kinase, a protein expressed in skeletal, smooth and cardiac muscle.[1] This gene defect prevents cells in muscle and other tissues from functioning normally, and leads to muscle weakness, myotonia, cardiac abnormalities and cataracts.[1] SMA is an autosomal recessive deletion of the survival motor neuron (SMN1) gene located on chromosome 5.[2] This results in reduced levels of the SMN protein, leading to degeneration of alpha motor neurons of the spinal cord and resulting in muscle weakness, pulmonary insufficiency, autonomic and bulbar dysfunction and progressive paralysis.[2] Although these diseases have a different etiology, both are associated with an increased incidence of perioperative respiratory and cardiovascular complications, as occurs with neuromuscular disorders in general. Furthermore, postoperative residual curarization (PORC) is a major risk in these patients and therefore reversal of neuromuscular blockade (NMB) is important to prevent PORC.[3] However, reversal with cholinesterase inhibitors, especially in patients with neuromuscular disorders can also cause complications.[3] Sugammadex, a new reversal agent for rocuronium or vecuronium induced NMB, is able to encapsulate the aminosteroid relaxant molecule, resulting in rapid recovery from NMB. Reports of reversal of rocuronium- or vecuronium-induced NMB with sugammadex in patients with neuromuscular disorders are limited.[4–6] We report two cases in which two patients, one suffering from SD, and the other from SMA received sugammadex to reverse a rocuronium-induced NMB. Neuromuscular management and safety and efficacy of sugammadex in patients with neuromuscular disorders are discussed.

Case 1

A 38-year-old female patient, weight 76 kg, height 165 cm presented for elective laparoscopic cholecystectomy. Her medical history revealed SD diagnosed at the age of 36 years. Three years ago after delivery of her first child, she remained in hospital for two weeks due to a respiratory failure. Ten weeks after discharge she was readmitted to ICU with pneumonia and pericarditis. She was managed without mechanical ventilation and discharged on inhaled salbutamol and antibiotics. One year ago she underwent an uneventful thyroidectomy under general anesthesia without the use of neuromuscular blocking drugs. Her medication consisted of thyroxine 200 μg daily.

On examination she had muscle weakness of the lower limbs, slight slurring of her speech, mild ptosis, weakness of eye and mouth closure. She had weakness of neck flexors, the finger extensors, and the feet distally. She had bilateral percussion myotonia of the abductor pollicis brevis, and poor relaxation of grip on command. No reflexes could be elicited. Blood pressure, ECG and transthoracic cardiac echogram were normal. Respiratory function tests showed a Forced Vital Capacity (FVC) of 52% of predicted and Forced Expiratory Volume in One Second (FEV1) in 42% of predicted. DNA testing of the patient's leukocyte DNA confirmed SD, and nerve conduction tests showed profuse myotonic discharges and myopathic changes. Laboratory tests showed an elevated creatine kinase of 548 IU l^{-1} (range 0–192 IU l^{-1}). Full blood count, biochemistry and chest X-ray were normal.

After obtaining informed consent from the patient she was scheduled for laparoscopic cholecystectomy under general anesthesia. The neuromuscular function was monitored quantitatively with train-of-four (TOF) stimulation of the ulnar nerve using the neuromuscular transmission module (M-NMT, Datex-Ohmeda, Helsinki, Finland). The primary efficacy variable for reversal was defined as the time from the start of the administration of sugammadex to recovery of the TOF ratio to 0.9.

Premedication consisted of oral esomeprazole 40 mg before anesthesia. An intravenous line was inserted on arrival in the operating room. Standard intraoperative monitoring included pulse oximetry, ECG, non-invasive blood pressure (NIBP), radial arterial line, continuous capnography, temperature, inspired/expired oxygen concentration, State entropy (SE) and response entropy (RE). Intravenous fluids were warmed through a coil and the patient was kept normothermic by using warmed intravenous fluids and a warming blanket. The patient was preoxygenated for 5 min, cricoid pressure was applied and anesthesia was induced and maintained with propofol Target Controlled Infusion (TCI, range 3–6 μg ml^{-1}) and remifentanil TCI (range 2–5 ng ml^{-1}).

Procedures for the set-up, calibration, and stabilization of neuromuscular monitoring were performed. The patient then received rocuronium 35 mg IV (0.47 mg kg^{-1}). This was followed by endotracheal intubation, under excellent intubation conditions, within 80 s (at TOF 3) and the lungs were ventilated with a mixture of oxygen and air at a ratio of 2:3. The time that elapsed from the injection of rocuronium to a maximal NMB (TOF 0) was 220 s. Additional analgesia was provided with paracetamol 1000 mg IV, parecoxib 40 mg IV, and infiltration of the surgical incision with bupivacaine 0.5% and cefotaxime 2 g IV was also administered. Surgery was uneventful and the duration of anesthesia was 65 min. At the end of the procedure neuromuscular monitoring showed