

REVERSION OF NEUROMUSCULAR BLOCK WITH SUGAMMADEX IN CHARCOT-MARIE-TOOTH DISEASE: CLINICAL CASE

1. BACKGROUND

- Charcot-Marie-Tooth disease (CMTd) is an hereditary demyelinating peripheral neuropathy;
- Is characterized by progressive muscular atrophy and sensory-motor abnormalities;
- The major anesthetic consideration is increased sensitivity to non-depolarizing muscle relaxants (NDMR)¹.

2. CASE REPORT

Male patient, 17-year old, ASA III, with CMTd. Sensory-motor deficits more evident in the lower limbs (Fig.1).



Fig. 1 – Cavus foot of the Patient.

Diagnosis: acute appendicitis.

Proposed surgery: lap. appendectomy.

Anesthetic approach: Balanced General Anesthesia (BGA).

ANESTHETIC INDUCTION: fentanyl (2µg/kg) and propofol (2mg/kg).

The supramaximal neuromuscular response was measured and recorded using the Organon TOF-Watch SX monitor and rocuronium (0.6mg/kg) was administered. Orotracheal intubation was performed 3'30" after NDMR administration, when the 4 responses to TOF disappeared.

ANESTHESIA MAINTENANCE: Desflurane.

The TOF stimulation was repeated every 5' with a frequency of 2Hz and an intensity of 70mA. During the procedure, neuromuscular blockade monitoring revealed 2 to 3 responses on the TOF-Watch SX monitor.

ANESTHETIC EMERGENCY: For reversion of the residual neuromuscular block, sugammadex (2mg/kg) was used and after 2' a TOF-ratio of 100% was obtained (TOF-ratio of 70% at 1'20", and 90% after 1'45").

The surgery lasted 90 minutes without complications.

The postoperative period was uneventful.

The neurological and musculoskeletal examination did not present any new deficits.

Preoperative Period

Intraoperative Period

Posop. Period

Monitorization

Standard ASA monitorization

TOF watch

Bispectral Index™

Neuromuscular relaxation was assessed through nerve stimulation of the ulnar nerve with a train-of-four (TOF) stimulus sequence and subsequent quantification of the thumb adductor response.

3. DISCUSSION

- The response to NDMR is variable and its effects may be prolonged.
- Due to the chronic demyelination present, the use of succinylcholine can trigger a hyperkalaemic response².
- There are no references to the use of sugammadex in CMTd.

4. LEARNING POINTS

- We report the absence of adverse events in the intraoperative and postoperative periods of a laparoscopic appendectomy under BGA using a NDMR in a patient with CMTd.
- We describe, for the first time, the use of sugammadex for the reversal of neuromuscular blockade in a patient with CMTd which occurred successfully and without adverse events.