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LETTER TO THE EDITOR

Reversal of neuromuscular blockade with sugammadex in a patient with spinal muscular atrophy type III (Kugelberg–Welander syndrome)

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To the Editor:

Spinal muscular atrophy (SMA) represents a group of neurodegenerative disorders, most commonly autosomal recessive, that are characterized by degeneration of motor neurons of the spinal cord, skeletal muscle atrophy, and generalized weakness [1]. SMA type III (Kugelberg–Welander syndrome) has an onset of symptoms after age 18 months and a protracted course with survival into adulthood [2].

A 61-year-old man (175 cm, 85 kg) with SMA type III was scheduled for elective percutaneous closure of an ostium secundum atrial septal defect (ASD). On physical examination he had a predominantly proximal tetraparesia with marked weakness and wasting of both upper and lower limbs, affecting more the proximal musculature and the lower limbs.

Anesthesia was induced with intravenous propofol (120 mg) and remifentanil (0.5 μg/kg/min) and maintained with continuous infusions of propofol (3–5 mg/kg/h) and remifentanil (0.1–0.3 μg/kg/min).

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Neuromuscular block (NMB) was monitored with repetitive train-of-four (TOF) stimulation (TOF-Watch SX) at the ulnar nerve. After induction of anesthesia but before the administration of rocuronium, setup and calibration of neuromuscular monitoring were performed. Baseline values of TOF ratio were above 0.90.

A single dose of rocuronium (40 mg), titrated in increments of 20 mg to a TOF count of 0, was administered intravenously to facilitate tracheal intubation.

Intraoperative transesophageal echocardiography confirmed the ASD with left–right shunt and showed an interauricular septum aneurysm, mild dilatation of right heart chambers, mild mitral regurgitation, and normal systolic function.

At the end of the 117-min procedure, sugammadex (170 mg) was administered intravenously for reversal of NMB (TOF ratio, 0.62). The time to complete reversal to a TOF ratio of 0.90 was 69 s, with no relevant cardiovascular changes. The trachea was extubated, and the patient was transferred to the recovery ward with the same preoperative pattern of muscle weakness. No signs of residual NMB or recurarization were observed during the postoperative period.

In patients with SMA there may be an increased sensitivity to nondepolarizing neuromuscular blocking agents, and suxamethonium may precipitate severe hyperkalemia [1, 2]. Our patient revealed an increased sensitivity to rocuronium as demonstrated by the presence of residual paralysis almost 2 h after a single intubating dose (approximately 20% lower than the usual standard dose of 0.6 mg/kg).

Anticholinesterase agents often do not guarantee an adequate recovery of neuromuscular function, including in cases of SMA [2, 3]. In addition, they may potentiate an existing NMB when administered during late recovery,

even in normal subjects [3]. Another drawback is the potential for cardiovascular side effects [4].

Sugammadex has been demonstrated to be safe and effective in patients with cardiovascular disease and other neuromuscular disorders [4, 5]. In this patient the reversal of a rocuronium-induced shallow NMB with sugammadex (2.0 mg/kg) was very efficient, with a time to complete reversal of 69 s and no adverse effects.

To our knowledge this is the first report of sugammadex use for NMB reversal in a patient with SMA. In conclusion, we suggest that the combination of rocuronium and sugammadex should be considered whenever there is indication for NMB in patients with SMA.

Conflict of interest The corresponding author has been involved in lecturing, education, and training, which was supported by Merck Sharp and Dohme (Portugal).

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