Sugammadex in Pediatric Congenital Myasthenic Syndrome

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Introduction

• Congenital myasthenic syndrome (CMS) is an inherited neuromuscular disorder caused by a defect in the acetylcholine receptor in the neuromuscular junction.[5]

• While CMS differs from myasthenia gravis (MG) both in age of onset (pediatric vs. adult) and etiology (congenital vs. autoimmune), both are characterized by 1) reduced acetylcholine activity at the NMJ, and 2) muscle weakness that gets worse with exercise. [2,5]

• Due to their baseline restrictive lung pathology and muscle weakness, myasthenic patients are predisposed to respiratory complications following anesthesia.

•The use of rocuronium is controversial in myasthenic patients.[4]

• Sugammadex is routinely used in adults and case reports show safety and efficacy in adults with myasthenia gravis.[4]

• While there is a lack of evidence as to its effectiveness in children, we believe sugammadex offers an attractive alternative to reversal of neuromuscular blockade without the use of anticholinesterases and antimuscarinics.[1,3]

Background

Fig. 1

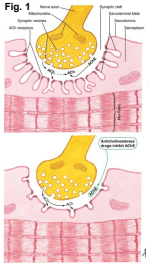

Fig. 2

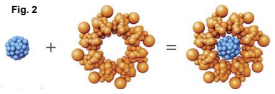

Fig. 3

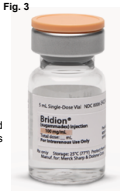

Fig. 1: Myasthenic patients have a decreased number and depth of folds in the neuromuscular junction. CMS patients have a defect in the post-synaptic acetylcholine receptor.[2]

Fig. 2: Sugammadex, a γ-cyclodextrin, forms a very tight complex around steroidal neuromuscular blocking drugs (rocuronium, vecuronium) at a 1:1 ratio in the plasma. The encapsulated NMBA is thus inactivated and then renally cleared.[3]

Fig. 3: A vial of sugammadex

Case History

Patient History

• A 6-year-old boy with congenital myasthenic syndrome, DOK7 gene mutation, and bilateral congenital hip dysplasia presents for bilateral varus derotation osteotomies, acetabular osteotomies, and soft tissue lengthening.

• No prior history of anesthesia.

• Requires BiPAP on room air at night for sleep. Home medications included 3,4-Diaminopyridine/amifampridine phosphate, pyridostigmine, and albuterol.

Case Details

• Anesthesia was induced via standard inhalational induction.

• After induction, a peripheral IV and arterial line were placed with subsequent administration of fentanyl, propofol, and dexamethasone.

• Acute pain team placed epidural catheter for postoperative pain control.

• Ulnar twitch monitor placed to assess muscle relaxation. Giving rocuronium (0.3 mg/kg) achieved complete relaxation. Patient was redosed twice to maintain 0/4 to 1/4 twitches on train of four.

• At the end of the case, he had return of twitches – 4/4 with fade.

• Patient was given Sugammadex 2mg/kg and subsequently had 4/4 twitches with sustained tetany.

• Patient was extubated to home BiPAP and taken directly to the ICU.

Follow up

• Patient remained on supplemental oxygen via BiPAP for a few hours but was quickly weaned to room air and transferred out of the ICU that day

• Stable, uneventful postoperative hospital course with discharge on postoperative day 3.

Discussion

• CMS patients are on chronic anticholinesterases that interact with traditional reversal agents and make it difficult to differentiate residual neuromuscular blockade from cholinergic crisis.[2]

• Patients are often more sensitive to anesthetics and require a stay in the ICU for careful monitoring of their respiratory status. These stays can lead to an accumulation of charges for both the patient and hospital.[2]

• Sugammadex is a reversal agent that sequesters relaxants, thus eliminating the uncertainty in dosing both relaxants and reversal agents in myasthenic patients.[1]

• Following surgery, our patient did not require extra time in the operating room or recovery room, or even an overnight stay in the ICU.

• Although the current first line therapy for pediatric neuromuscular blocker reversal is glycopyrrolate/neostigmine, this case describes the use of sugammadex in a pediatric congenital myasthenic syndrome patient. Administration of sugammadex resulted in complete reversal of neuromuscular blockade, positive effects on respiratory status, and reduced length of stay in the ICU, thereby justifying its initial higher cost and greatly benefitting patient care.

References

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