1066149 - CONGENITAL MYASTHENIA PATIENT FOR STRABISMUS REPAIR

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Purpose: To report a case and review the clinical features and anesthetic implications of a patient with congenital myasthenia syndrome (CMS) undergoing ambulatory strabismus repair.

Clinical Features: Consent and ethics approval were obtained. A two year-old nocturnally vented via tracheostomy CMS patient with 24-hour nursing care at a special need facility presented to an ambulatory center for serial elective strabismus repair secondary to sixth nerve palsy on treatment with diaminopyridine and pyridostigmine. CMS was diagnosed by multiple single fiber EMGs, though no genetic testing was done. The ophthalmologist involved in both cases routinely asks for muscle relaxation, presenting a challenge in a patient where muscle relaxation is contraindicated. The anesthetic technique involved maintenance with sevoflurane and remifentanil (bolus: 1mcg/kg and titrated infusion: 0.05 – 0.1 mcg/kg/min). Post-operatively the patient was transitioned to her home ventilator for transport and use in the PACU. There was no incident of oculocardiac reflex during either case. Upon arrival in the post-anesthesia unit the patient was weaned from ventilator-support and observed until discharge criteria were met. Had the patient failed to meet discharge criteria, post-operative pediatric ICU was available. The degree of muscle relaxation provided by remifentanil was deemed adequate by the surgeon.

Conclusion: CMS, like other forms of myasthenia, is a disease characterized by weakness and fatigue due to deficiency in neurotransmitter function. Myasthenia Gravis is an autoimmune disorder in which antibodies interfere with synaptic transmission of nerve impulses, whereas CMS has a genetic basis for failed neurotransmitter function. CMS is often grouped into three categories depending on site of defect: presynaptic, postsynaptic or synaptic. Presynaptic CMS is characterized by an inadequate release of acetylcholine for transmission of nerve impulse. Postsynaptic CMS can be caused by either too few acetylcholine receptors or an inadequate response (receptors stay open too short/long). Synaptic CMS is due to inadequate acetylcholinesterase activity in the synaptic cleft. Non-depolarizing muscle relaxants should be used with caution in patients with Myasthenia Gravis due to the potential of prolonged muscle relaxation.

Strabismus repair is the most common pediatric eye surgery performed, with 3-5% of the population affected. Muscle relaxation is frequently required in part to facilitate endotracheal intubation but also optimize surgical conditions, particularly during forced duction testing. The use of succinylcholine should be avoided during strabismus repair due to the prolonged contraction of the extra-ocular muscles. The use of volatile anesthetic and remifentanil has been used in patients with Myasthenia Gravis resulting in reversible muscle relaxation.

CMS presents substantial anesthetic challenges, particularly in procedures requiring muscle relaxation. To our knowledge there have been no reports of using remifentanil and volatile on patients with CMS. This technique provided adequate surgical conditions and facilitated same day discharge of a home ventilated patient from an ambulatory center.