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LETTER TO THE EDITOR

Anesthetic management of Prader–Willi syndrome: what if neuromuscular relaxants could not be avoided?

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To the Editor:

We read with interest the article by Meco et al. [1] concerning the anesthetic management of a 16-month-old child with the Prader–Willi syndrome avoiding the use of neuromuscular blockers. We would like to add our experience with anesthetic management involving the use of succinylcholine and non-depolarizing neuromuscular blockers in a child with the Prader–Willi syndrome because avoiding these drugs may not be possible in all situations.

A 5-year-old male child with Prader–Willi syndrome was scheduled for laparoscopy and repair of bilateral undescended testes. Preanesthetic examination revealed obesity (weight 30 kg, height 96 cm, body mass index 32 kg/m^2), short stature, small hands and feet, cryptorchid testes, and hypoplastic dental enamel. Following an inhalation induction with sevoflurane 8% in 100% oxygen, we succeed in obtaining venous access only after multiple failed attempts. However, the procedure was complicated by severe laryngospasm with an episode of desaturation (SpO_2 85%). Intravenous succinylcholine 50 mg was given immediately and effective bag-mask ventilation could then be delivered. The trachea was secured using a 4.5-mm cuffed endotracheal tube. The patient was then maintained on nitrous oxide–oxygen–sevoflurane and atracurium for relaxation. Analgesia was achieved with intravenous fentanyl 30 μg. His initial esophageal temperature was 36.5°C but this increased to 39.4°C after 40 min. This change was accompanied by an increase in heart rate and end-tidal carbon dioxide concentration. An arterial blood gas revealed respiratory acidosis. The patient was actively cooled with cold sponges, application of ice packs in the axilla, and gastric lavage using ice-cold saline. His temperature dropped to 37.8°C after 60 min. No dantrolene sodium was given. The total duration of surgery was 130 min. The neuromuscular blockade was then reversed using atropine and neostigmine, without difficulty. The patient was extubated awake with good respiratory efforts. There were no episodes of hyperthermia and obstructive sleep apnea in the postoperative period. The CPK obtained 8 h following anesthesia was 25 units per liter (normal, 8–132).

Certainly, use of anesthesia techniques [1, 2] avoiding the use of neuromuscular blockade in the Prader–Willi syndrome may reduce the risk of postoperative complications associated with this syndrome. However, avoiding neuromuscular blockade altogether may not be possible in all situations. We report this anesthetic experience because we believe that as experience is gained, our understanding increases concerning anesthetizing children with the Prader–Willi syndrome. (1) A narcotic-relaxant technique can be utilized safely in children with the Prader–Willi syndrome [3]. Both succinylcholine and non-depolarizing relaxants can be safely used in patients with Prader–Willi syndrome [4]. (2) Although thermoregulatory dysfunctions including intraoperative hypothermia are common in patients with Prader–Willi syndrome, a relationship to malignant hyperthermia has not been established [5]. Thus, the intraoperative increase in temperature with the use of succinylcholine does not always indicate malignant hyperthermia in these patients. (4) Despite the presence of hypotonia, neuromuscular blockade with good reversal is possible [3].

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