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LETTER TO THE EDITOR

Vecuronium was safely used in a patient with Engelmann's disease without muscle weakness

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To the Editor:

Engelmann's disease is a bizarre, autosomal dominantly inherited bone disorder characterized by progressive expansion and sclerosis mainly affecting the diaphyses of the long bones and cranium [1], and a possibility of restricted mouth opening and neck movements has been reported [2]. Although we had an interest in bone abnormalities, there are several reports that mention muscle abnormalities such as selective atrophy of type II fiber or thickening of the perivascular basement membrane [3, 4]. Therefore, a muscle relaxant may trigger an undesirable problem in these patients. However, to the best of our knowledge, there is no report regarding the use of a nondepolarizing muscle relaxant in a patient with Engelmann's disease.

The patient was a 37-year-old woman (height 171 cm; weight 46 kg). She was diagnosed with Engelmann's disease at the age of 23 by symptoms of the lower limbs, radiography of the long bones, and biopsy of the left tibia. She was scheduled for resection of bone tumor of her left little finger under general anesthesia. Her interincisor distance was about 4.5 cm, and neck movements were not restricted. Mallampati classification was class I. Although an electromyogram was not examined, her muscle strength was almost normal by manual muscle test. Her vital capacity was 3.73 l, $\%FEV_{1.0}$ was 90%, and all other laboratory data were within normal ranges. After administration of intravenous thiopental 225 mg, fentanyl 0.1 mg, and vecuronium 9 mg, her trachea was intubated without any difficulties at the first attempt. Anesthesia was maintained with 67% nitrous oxide in oxygen and sevoflurane 1–1.5%. Additional vecuronium was not administered. Spontaneous respiration appeared just before extubation because her lungs were ventilated mechanically during operation. The operation was uneventful, lasting 84 min. At the end of the operation, no palpable fade was present by train-of-four and tetanic stimulation of 50 Hz for 5 s at ulnar nerve. In addition, tidal volume was kept at more than 450 ml, and grip strength seemed sufficient. Reversal of vecuronium was not performed, and the trachea was extubated 141 min after administration of vecuronium.

Because we did not realize the potential harm by muscle relaxants in patients with Engelmann's disease at that time, we administered vecuronium inadvertently. Our patient might have microscopic muscular abnormalities even though no muscle weakness had been demonstrated. Therefore, we should have avoided administering vecuronium by using a supraglottic airway device. With regard to evaluation of neuromuscular blockade, no palpable fade by tetanic stimulation of 50 Hz for 5 s is considered to indicate adequate recovery from neuromuscular blockade [5], and we confirmed this. In addition, because tidal volume was 450 ml (about 10 ml/kg) and grip strength was sufficient at tracheal extubation, we deemed our patient recovered from neuromuscular blockade adequately.

In conclusion, vecuronium was used safely in a patient diagnosed with Engelmann's disease without muscle weakness or difficult airway. However, close attention should be paid to administering a muscle relaxant to the

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individual patient with Engelmann's disease because there is potential harm from muscle relaxants.

References

1. Vanhoenacker FM, Janssens K, Van Hul W, Gershoni-Baruch R, Brick R, De Schepper AM. Camurati–Engelmann disease. Review of radiological features. Acta Radiol. 2003;44:430–4.
2. Mason J, Slee I. Anaesthesia in Engelmann's disease. Anaesthesia. 1968;23:250–2.
3. Yoshioka H, Mino M, Kiyosawa N, Hirasawa Y, Morioka Y, Kasubuchi Y, Kusunoki T. Muscular changes in Engelmann's disease. Arch Dis Child. 1980;55:716–9.
4. Naveh Y, Ludatshcer R, Alon U, Sharf B. Muscle involvement in progressive diaphyseal dysplasia. Pediatrics. 1985;76:944–9.
5. Naguib M, Lien CA. Pharmacology of muscle relaxants and their antagonists. In: Miller RD, editor. Miller's anesthesia. Philadelphia: Elsevier Churchill Livingstone; 2005. p. 481–572.