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LETTER TO THE EDITOR

Anesthetic management of a child with Ullrich myopathy

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To the Editor:

A 9-year-old boy was scheduled for surgery to correct bilateral fixed plantar flexion.

Because muscular weakness had been observed early in childhood, a muscle biopsy had been performed (result: myopathy), but a skin biopsy finally established the diagnosis of Ullrich's myopathy (UD), a congenital muscle dystrophy.

Preoperative physical examination showed a child with weight of 24 kg and height of 131 cm. Cervical spinal mobility was slightly limited; micrognathism and prominent incisive teeth were present. Creatinine phosphokinase (CK) level was at the upper limit of normal values for our laboratory. Cardiac and pulmonary function tests were normal.

The patient had already undergone one general anesthesia in the past (for the muscle biopsy) consisting of total intravenous anesthesia (TIVA) with alfentanil and propofol.

After careful positioning, standard monitoring was applied. Peripheral venous was obtained while the patient breathed O_2 and N_2O (50–50 %) by mask. Induction of anesthesia was performed with sufentanil (0.1 μg/kg) and propofol (3 mg/kg), followed with manual ventilation with sevoflurane. Orotracheal intubation was performed at first attempt (Cormack–Lehane stage 3).

Surgery lasted 2 h. A tourniquet was used for each site successively. Core body temperature and capnography remained normal; no significant increase in expired CO_2 was noticed when the tourniquets were released. In this surgery, as in the previous one, no muscle relaxant was used. Postoperative analgesia consisted of IV ketorolac, acetaminophen, and piritramide in the pediatric acute care unit (PACU). The postoperative evolution was satisfactory. No clinical signs of rhabdomyolysis (e.g., dark urine) were observed. No significant elevation in the CK levels was observed. The CK increased from 407 U/l before the surgery to 440 U/l after the surgery; the upper normal value of our laboratory is 400 U/l.

The clinical diagnosis of UD is based on general muscle weakness, contractures of multiple joints, hyperextensibility of distal joints, and other orthopedic and systemic signs [1, 2]. There is no evidence of cardiac dysfunction, but the diaphragm is often involved early. Tracheal intubation may be difficult because of the combination of micrognathism and contracture of the temporomandibular muscles.

When planning anesthesia for a child with a muscle disease, one of the anesthesiologist's concerns is to limit the risk of inducing a malignant hyperthermia (MH) crisis or rhabdomyolysis. In this case, the risk of MH was estimated to be not greater than in the general population because the familial history was negative and the gene mutations for UD are different and distant from the known mutations of the RYR1 and CACNL1A3 genes associated with MH [3–5].

In conclusion, this case, in addition to the other published cases, shows that halogenated agents can be used safely in children with UD as well as in other congenital muscle dystrophies.

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Conflict of interest None.

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