ganglion using CT imaging. This may prove to be more reliable and selective than previously described methods for sympathetic blockade of the head and upper extremities.

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Muscle Rigidity Following Halothane Anesthesia in Two Patients with Freeman–Sheldon Syndrome

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The musculoskeletal and soft-tissue manifestations of Freeman-Sheldon Syndrome (FSS) often require orthopedic and plastic reconstructive surgery. Patients may require correction of the small oral commissure with pursed lips that results in the characteristic appearance of a "whistling face"; joint contractures with or without ulnar deviation of the fingers; equinovarus; and scoliosis. This constellation of features appears to be the result of an underlying myopathy.[1,2] Malignant hyperthermia (MH) is clearly associated with certain myopathies such as central core disease and is possibly associated with other myopathies such as Duchenne muscular dystrophy.[3] The few reports describing anesthesia in patients with FSS focus on the difficulty of tracheal intubation but also report that two potent MH triggers (halothane and succinylcholine) have been used uneventfully for anesthesia.[4–6] We report two instances of muscle rigidity that occurred in unrelated children without a family history of MH and that occurred only after exposure to these anesthetic agents.

Case Reports

Case 1. A 3-yr-old, 9.5-kg boy with FSS required craniofacial surgery. Two previous anesthetic experiences at age 5.5 and 7.5 months using non–MH-triggering agents had been uneventful. For this procedure, anesthesia was induced *via* mask with oxygen, nitrous oxide, and halothane. Within 10 min, generalized rigidity of skeletal muscles developed and rapidly became more severe. Intubation was attempted and deferred because of masseter rigidity. Hemoglobin oxygen saturation measured by pulse oximeter (Sp_{O_2}) was 93%, and an arterial blood gas analysis (FI_{O_2} 35%) showed *p*H 7.03, P_{CO_2} 64 mmHg, and P_{O_2} 101 mmHg. Halothane and nitrous oxide were discontinued, and a second anesthetic machine, unused for at least 18 h, was used to provide 100% oxygen. Sodium bicarbonate 2.5 mEq/kg was then given intravenously. Two doses of pancuronium 0.1 mg/kg each caused neither masseter muscle relaxation nor relaxation of peripheral muscles. Dantrolene 2 mg/kg was given 35 min after starting the induction. Skeletal muscle relaxation occurred within 5 min. His Sp_{O_2} increased to 95%, and arterial P_{O_2} was 84 mmHg. His heart rate rose from less than 130

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Key words: Anesthetics, volatile: halothane. Complications: Freeman-Sheldon syndrome; malignant hyperthermia. Enzymes: creatine phosphokinase. Neuromuscular relaxants: dantrolene; succinylcholine.

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beats/min in the first 15 min to 150 beats/min by 30 min, and ventilation was assisted with positive pressure delivered by bag and mask until he had sufficient respiratory drive and the ability to maintain patency of his upper airway.

Serum creatine phosphokinase at the time of the rigidity was 285 IU/l; it increased to 9,215 IU/l the next day and was 7,300 IU/l the third day. The patient never developed hyperthermia, hyperkalemia, cardiac arrhythmias, or myoglobinuria. Muscular weakness was still evident at the time of discharge 2 days after the event, but after 4 more days, his muscle strength had improved, enabling him to stand by holding on to objects. He subsequently has undergone three complex craniofacial surgical procedures using agents not associated with precipitating MH episodes.

Case 2. A 4-yr-old, 15-kg girl with FSS was scheduled for bilateral oral commissurotomy. Anesthesia was induced with 10% rectal methohexital (30 mg/kg) followed by halothane and nitrous oxide *via* mask. After several attempts at tracheal intubation, the glottis was visualized, but the patient developed laryngospasm and received atropine 0.02 mg/kg and succinylcholine 1.5 mg/kg. With the laryngoscope blade in the mouth, she developed masseter muscle spasm (train-of-four was zero). The trachea was intubated, and halothane was replaced by fentanyl and pancuronium. Because there was no increase of exhaled P_{CO_2}, cardiac arrhythmias, tachycardia, hyperkalemia, temperature increase, or myoglobinuria, surgery was allowed to proceed. Creatine phosphokinase increased from 208 IU/l during the operation to 1,193 IU/l 6 h later; it was 933 IU/l the next day.

Discussion

Masseter muscle spasm or generalized muscle rigidity developed in two patients with FSS after induction of anesthesia using known MH-triggering agents. Baseline creatine phosphokinase levels were increased in both cases and then increased after the event. The reaction to induction was more severe in the first case, requiring dantrolene, bicarbonate, and supplemental oxygen to modify the severe muscle rigidity and to treat the metabolic component of acidosis. In the second case, discontinuing halothane was sufficient to treat masseter rigidity.

Only a limited number of the classic features of MH[7] were manifest in these two cases. This may be because of the prompt administration of dantrolene in case 1 and the more limited spectrum of isolated masseter muscle spasm in case 2.

Despite previous reports of successful administration of anesthesia using halothane and/or succinylcholine to patients with FSS, these two patients reacted unfavorably to these MH-triggering agents. This seeming inconsistency with reports in the literature may actually reflect the genetic heterogeneity of FSS. Although an autosomal dominant inheritance pattern is the most common in FSS,[8] a recessive inheritance pattern also has been recognized.[9–11] Brownell[3] has speculated that the association of MH with certain types of myopathies suggests that its gene is located in close proximity to the genes for various myopathies. The predictability of clinical MH from the *in vitro* muscle contracture test in myopathic patients and their families remains uncertain.[12]

We conclude that halothane and succinylcholine should not automatically be assumed to be safe in patients with FSS. In the absence of results from a caffeine–halothane muscle contracture test, nontriggering anesthetic agents should be considered for all patients with FSS.

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