Patients with Huntington’s Chorea May Respond Normally to Succinylcholine

To the Editor:—A single report of prolonged paralysis in a patient with Huntington’s chorea is the basis for the recommendation that succinylcholine be avoided in such patients. We recently found it necessary to use succinylcholine to facilitate endotracheal intubation in a patient who had this hereditary disorder, and wish to report our experience.

A 46-yr-old, 55-kg man came to surgery for placement of a feeding gastrostomy. He had a 10-yr history of dementia and chorea, which had progressed to include swallowing dysfunction, with chronic drooling, and regurgitation. His father, brother, paternal aunt, and two paternal uncles had died from Huntington’s chorea; a daughter was also affected. Medications included haloperidol 2 mg daily and benztropine 1 mg bid. He appeared awake but did not respond appropriately to verbal commands, was unable to communicate, and made frequent, unpredictable, forceful athetoid movements. The remainder of the physical and laboratory examination was unremarkable. Because he was completely uncooperative, we felt that local or regional anesthesia would not be feasible. To minimize the risk of aspiration during induction of general anesthesia, we planned “rapid-sequence” endotracheal intubation with cricoid pressure and muscle paralysis.

We applied surface electrodes over the patient’s ulnar nerve at the wrist, applied supramaximal stimulation at 1 Hz, and measured the resulting muscle twitches using a Grass FT-10G strain gauge. After “preoxygenation,” we administered fentanyl (150 μg), thiopental (200 mg), and succinylcholine (55 mg, 0.6 mg/kg); with cricoid pressure applied, we accomplished tracheal intubation without difficulty. Nitrous oxide (70%) in oxygen maintained anesthesia, while we documented recovery from succinylcholine paralysis by continually recording muscle twitch strength. When twitch recovery was complete, we added isoflurane (up to 2% inspired) for the remainder of the 90-min procedure. Emergence from anesthesia was smooth; extubation and recovery were uneventful.

Analysis of the twitch-strength record revealed that muscle strength returned to 50% of control 555 s after succinylcholine and to 90% of control within 660 s. These recovery times are within the ranges (mean ± 2 SD) previously reported for normal patients receiving similar doses of succinylcholine.

Gualandi and Bonfanti’s report of a patient with Huntington’s chorea in whom succinylcholine 50 mg lasted 2 h has been excerpted in case reports as well as textbooks. Anesthesiologists subsequently avoided giving succinylcholine to patients with Huntington’s chorea. The unproven association has become accepted as fact!

Conversely, the diminished cholinesterase activity in Gualandi and Bonfanti’s patient may have been completely unrelated to his Huntington’s chorea. Even in the absence of a “linkage” between Huntington’s chorea and atypical cholinesterase, a patient with Huntington’s chorea has an approximately 1 in 2,500 chance of being homozygous for atypical cholinesterase. Our findings suggest that patients with Huntington’s chorea recover normally from succinylcholine and that this drug may be administered safely to these patients when clinically indicated.

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REFERENCES

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