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Sevoflurane and mivacurium in a patient with Huntington's chorea

P. Nagele* and A. F. Hammerle

Department of Anaesthesiology and General Intensive Care, General Hospital, University of Vienna, Währinger Gürtel 18–20, A-1090 Vienna, Austria

**Corresponding author*

There is little experience of anaesthesia for patients with Huntington's chorea. These patients have an increased risk of intraoperative complications such as pulmonary aspiration. We present the successful anaesthetic management of a 17-yr-old patient suffering from Huntington's chorea requiring urgent appendectomy. After rapid-sequence induction with thiopental 400 mg and succinylcholine 100 mg, anaesthesia was maintained with sevoflurane. For maintenance of neuromuscular blockade mivacurium 10 mg was administered and repeated 15 min later. Except for a short episode of postoperative shivering, the perioperative course was uneventful. Sevoflurane and mivacurium were used safely and effectively in this patient.

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Only a few case reports exist concerning the anaesthetic management of patients suffering from Huntington's chorea.^{1–9} Huntington's chorea is a rare hereditary disorder of the nervous system. Symptoms usually appear between the ages of 30 and 45 yr and include choreatic movements, progressive mental deterioration and ataxia. Early onset of symptoms is associated with more rapid and severe progression of the disease. Patients suffering from Huntington's chorea are at higher risk of intraoperative complications, including pulmonary aspiration,¹ a prolonged response to succinylcholine³ and thiopental,⁵ and increased sensitivity to midazolam.⁶ The primary goal in general anaesthesia

for these patients is to provide airway protection and a rapid and safe recovery. The development of new short-acting drugs, such as propofol, has led to recommendation of a total intravenous anaesthesia (TIVA) technique^{6–8} in patients with Huntington's chorea, thus avoiding compromised postoperative recovery and the increased risk of postoperative shivering from potent inhalational agents. However, the recently introduced inhalational agent sevoflurane could eliminate the problem of prolonged recovery because of its favourable pharmacokinetic profile. Secondly, the use of the short-acting, non-depolarizing neuromuscular blocking drug mivacurium could provide adequate relaxation without

the danger of prolonged paralysis at the end of the operation.

Case report

A 17-yr-old, 85 kg girl with a 4-yr history of Huntington's chorea was admitted for urgent appendectomy. Her sister had died from Huntington's chorea at the age of 25 yr. She suffered from progressive dementia and swallowing dysfunction with regurgitation, but was not taking any medication. She appeared anxious and uncooperative and did not respond appropriately to verbal commands. Laboratory investigations were normal.

To minimize the risk of aspiration, we performed rapid-sequence induction consisting of thiopental 400 mg and succinylcholine 100 mg with cricoid pressure applied. To determine the degree of muscle relaxation we used train-of-four (TOF) monitoring (Innervator; Fisher & Paykel Health Care, Auckland, New Zealand). After induction the TOF was no longer detectable and the trachea was intubated. Within 10 min, all four twitches of the TOF returned to control levels and mivacurium 10 mg was administered. Anaesthesia was maintained with fentanyl 0.25 mg and sevoflurane 1.5–2% inspired. After 15 min, a second increment of mivacurium 10 mg was given, as four twitches were detectable and the TOF ratio was >0.75. At the end of surgery, which lasted 70 min, the TOF had recovered to control values and sevoflurane was discontinued. After 5 min the patient regained consciousness and was extubated uneventfully without any reversal of residual neuromuscular block. To prevent postoperative nausea and vomiting, and thus reduce the risk of aspiration, we administered ondansetron 8 mg and ranitidine 50 mg i.v. The postoperative course was uneventful except for a short episode of shivering, which was successfully treated with pethidine 50 mg. The patient was discharged from hospital on the third postoperative day.

Discussion

Different anaesthetic techniques have been recommended for use in patients suffering from Huntington's chorea. Spinal anaesthesia has successfully been performed in one patient.⁹ Some authors recommend using a TIVA technique and avoiding potent inhalational agents to reduce the risk of postoperative shivering and the precipitation of generalized

tonic spasms.^{6–8} Others have used inhalational agents, such as halothane² and isoflurane,⁴ without problems. In spite of recommendations to avoid potent inhalational agents,^{6–8} we decided to use sevoflurane to maintain anaesthesia because of its favourable pharmacological properties. It is easy to titrate and provides rapid recovery. It proved to be effective and safe in our patient; the short episode of postoperative shivering did not evoke generalized spasms and was easily treated by the administration of pethidine.

We did not find any prolonged effects from thiopental or succinylcholine, which have been found previously.^{3,5} The cause of the prolonged response to succinylcholine, as found in one patient in a previous report,³ may have been abnormal plasma cholinesterase and completely unrelated to Huntington's chorea.⁴ The use of rocuronium for rapid-sequence induction in this patient might have been a feasible alternative, but the authors' experience concerning rocuronium was too limited at that time. The recovery time for mivacurium was within the range reported in normal patients who have received similar doses. We could not find any evidence of altered mivacurium pharmacodynamics in our patient despite prior administration of succinylcholine.

The use of sevoflurane and mivacurium for general anaesthesia in patients suffering from Huntington's chorea seems to be effective and safe.

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