
CASE REPORTS

MONITORED ANESTHESIA CARE FOR A PATIENT WITH ADVANCED HUNTINGTON'S CHOREA

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Introduction

Huntington's disease (HD), a rare, autosomal dominant disorder of the central nervous system, has been associated at times with unusual responses to anesthetic agents such as thiopental, midazolam, succinylcholine, and nondepolarizing neuromuscular blocking drugs. We describe the anesthetic management of a 50 year-old female with advanced HD, complicated by chorea, dementia, dysphagia, and dysarthria, undergoing percutaneous endoscopic gastrostomy (PEG) placement. To the best of our knowledge, there have not been any prior reports describing the use of propofol for sedation in a patient with Huntington's disease.

Case

A 50 year-old, 50 kilogram female with Huntington's disease (HD) complicated by chorea, dementia, dysphagia, and dysarthria, with no history of cardiopulmonary disease, presented for percutaneous endoscopic gastrostomy (PEG) placement. Despite redirection by the patient's sister at bedside, the patient was agitated in the endoscopy holding area and suffering from violent choreoathetoid movements. Although she was unable to follow commands, she spontaneously demonstrated a Mallampati Class 1 airway, with adequate mouth opening, thyromental distance, and neck range of motion.

Given her agitation, intramuscular sedation with a benzodiazepine and/or an antipsychotic was considered; however, with the aid of the nursing team and the patient's sister, a 20-gauge intravenous catheter was placed in the left forearm and well secured. Midazolam was administered in one-milligram increments over the span of approximately 20 minutes, to a total dose of four milligrams, with little decrease in the patient's movements or level of agitation. The decision was made to proceed to the endoscopy suite to continue with further sedation.

Standard ASA monitors and a nasal cannula were placed with difficulty, and an initial 30-milligram bolus of propofol was administered. Within 60 seconds, the patient's choreoathetoid movements ceased, and she closed her eyes. With intermittent propofol boluses to a total dose of 200 milligrams, and local anesthesia administered by the proceduralist, PEG placement

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proceeded without incident, with adequate respiratory and cardiovascular parameters throughout the fifteen-minute procedure. The patient's recovery room course was uneventful, and she was able to return to her skilled nursing facility without any undue delay.

Discussion

Huntington's disease was first fully described by 22-year old New York family physician George Huntington in 1872¹. It is a devastating disease of the human central nervous system, now known to be caused by a toxic gain-of-function mutation of the Huntingtin gene². This mutation, an increase in the number of cytosine-adenine-guanine trinucleotide repeats present in the gene's coding portion, creates a polyglutamine region in the Huntingtin protein which alters its function and leads to neuronal degeneration, particularly affecting the basal ganglia. Greater than 40 CAG repeats produces fully penetrant disease, which is inherited in an anticipatory autosomal dominant pattern².

Through the latter portion of the 20th century, case reports have at times associated HD with prolonged recovery from benzodiazepines and barbiturates, as well as with increased duration of paralysis after administration of both depolarizing and nondepolarizing neuromuscular blocking drugs (NMBDs)^{3,4}. In addition, at least one genetic study has found an increased incidence of atypical pseudocholinesterase in patients with HD⁵.

A recent review of eleven patients with HD who underwent seventeen general anesthetics, by Kivela *et al* of the Mayo Clinic, did not find any atypical reactions to midazolam, sodium thiopental, succinylcholine, nor nondepolarizing NMBDs⁶. Prolonged sedation

observed after benzodiazepines and barbiturates in prior reports was attributed by these authors to relative overdosing of these drugs, not abnormal patient response. While no abnormal response to NMBDs was observed, the authors still recommended caution with succinylcholine given the association with atypical pseudocholinesterase. Succinylcholine may still be needed, however, due to the possible increased risk of aspiration in these patients.

The administration of propofol to individuals with HD has been previously described^{7,8}. However, in those two case reports, the tracheas of both patients were intubated, and the anesthetics were general anesthetics with mechanical ventilation. There was no report on any effect that propofol had on the chorea. The use of sedation has been reported previously in a patient with Huntington's disease, but the sedation was achieved solely with midazolam, which in that case did have a beneficial effect on the choreiform movements⁹.

With regards to our case, midazolam did not control the patient's motor symptoms or agitation. In addition, we note that the recovery course was not prolonged despite this 50-kilogram patient's receiving four milligrams of midazolam prior to her brief procedure. A small dose of propofol quickly and effectively ceased her choreoathetoid movements and established a plane of anesthesia appropriate for beginning the PEG placement. Propofol has been associated with involuntary movements including athetosis, seizures, and dystonia, likely due to inhibition of inhibitory pathways in the basal ganglia, leading to a net increase in excitatory cholinergic outflow¹⁰. In that case report, the movements were lessened by benztropin. The use of propofol in our case was effective in quieting the debilitating motor symptoms of Huntington's disease.

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