Anesthetic considerations for the patient with Huntington’s disease

Huntington’s disease (HD) is an autosomal, dominantly inherited disease with symptoms manifesting late in life. Patients suffer from motor dysfunction and progressive mental deterioration. HD patients present a number of anesthetic challenges as they are usually elderly, malnourished, and at risk for aspiration. Previous reports on anesthesia for patients with HD have warned that sodium pentothal and succinylcholine cause prolonged apnea in this patient population.

In this article, a 78-year-old female presented for cataract extraction. Although her movement disorder was under good control with haloperidol, a general anesthetic was chosen for the procedure in order to ensure a tranquil surgical field. This article presents the successful management of a patient with HD using a balanced anesthetic technique combining judicious amounts of narcotics and barbiturates with isoflurane.

Key words: Huntington’s disease, isoflurane, sodium pentothal, succinylcholine.

Huntington’s disease (HD) is an autosomal, dominantly inherited disease affecting the basal ganglia. The incidence is approximately 7 in 100,000. Expression of the disease is variable, with onset occurring most commonly between the third and fifth decade of life. HD is manifested by personality changes, choreathetoid movements, and progressive but variable dementia. Dysphagia is the most significant motor symptom as it hinders nutrition and increases the patient’s risk for aspiration. The involuntary movements, concomitant depression, and apathy lead to weight loss and cachexia. This, accompanied by the complications of aspiration, is the commonest cause of death, which usually occurs 10 to 30 years after onset of the disease.

Patients with HD present a number of anesthetic challenges. They are usually elderly and malnourished. Aspiration during induction and emergence is an added risk. Anesthetic management has been complicated by reports that barbiturates and/or succinylcholine may cause prolonged apnea in these patients. The authors report the successful management of a patient with Huntington’s disease.

Case report

A 78-year-old, 57-kg, white female presented for extracapsular cataract extraction with intraocular lens implant of the right eye. Physical examination revealed a frail elderly female with poor skin turgor. Laboratory values were all within normal ranges.

The patient’s medical history was significant
for a movement disorder diagnosed as HD at age 71, osteoporosis, and chronic dehydration. Family history was positive for a similar movement disorder present in three females among 10 siblings. The males were asymptomatic. Development of the symptoms in this family was consistently in the sixth or seventh decade of life. No family member had as yet exhibited any dementia. Current management consisted of the dopamine receptor blocking agent, haloperidol, at a dose of 1 mg at bedtime.

The patient was premedicated with ranitidine, 150 mg by mouth. Metoclopramide was omitted as its use has been associated with the manifestation of choreiform movements, possibly as a result of its action on a specific subpopulation of cerebral dopamine receptors. Pretreatment with cholinergic agents was also avoided as it is felt that these agents may further exacerbate the choreiform movements.

Although extracapsular cataract extractions are usually performed with local anesthesia and intravenous (IV) sedation in the authors' institution, a general anesthetic with endotracheal intubation was selected at the preoperative interview in order to ensure the patient's immobility during the surgical procedure.

In the operating room, IV access was established and Ringer's solution was infused. Baseline blood pressure and heart rate were 145/83 and 61 respectively. Oxygen saturation was 95% on room air. The patient received oxygen (6 L/min) by face mask, 50 μg fentanyl, 1.25 mg droperidol, and a priming dose of atracurium (0.06-0.08 mg/kg).

Repeat blood pressure was 155/71 with a heart rate of 63. Oxygen saturation was now 99%. Three minutes after the priming dose of atracurium was injected, the patient received 150 mg sodium thiopental (2.5 mg/kg), 100 μg fentanyl, and 25 mg atracurium. Intubating conditions were realized within 90 seconds. Cricoid pressure was applied during the induction sequence to minimize the risk of aspiration.

Isoflurane (0.5%-1.0%) in 50% nitrous oxide/oxygen mixture was administered during surgery. Additional muscle relaxant was given as indicated by the peripheral nerve stimulator (2 mg/kg).

At the conclusion of surgery, neuromuscular blockade was reversed with 1 mg glycopyrrolate and 5 mg neostigmine. Adequate neuromuscular reversal was demonstrated by tetanus stimulation without fade. The patient began spontaneous ventilation, was extubated, and was transferred to the postanesthesia care unit (PACU) in stable condition. She demonstrated typical choreiform movements in the operating room after emergence. These movements remained throughout her stay in the PACU but did not necessitate the use of restraints.

Discussion

Huntington's disease is a genetic disorder characterized by the premature destruction of neurons in the caudate nucleus and putamen from an unknown mechanism. Although choreoathetoid movements are classically associated with excessive dopaminergic transmission within the basal ganglia, studies have demonstrated that the dopamine concentration in the brains of persons with HD is no different from that of normal individuals. Other compounds have been implicated as responsible for the development of the disease including quinolinic acid and somatostatin.

Management of the disorder is directed at the relief of the choreiform movements with dopamine receptor blocking agents. Haloperidol, a butyrophenone, is the most commonly used agent. This drug has implications for the anesthetist in that butyrophenones may interact with commonly used anesthetic agents. In combination with enflurane or halothane, butyrophenones may cause a decrease in arterial blood pressure. They also may augment the respiratory depression of narcotics and increase the hypnotic effect of barbiturates.

Prolonged apnea after induction with sodium thiopental (STP) has been previously reported. However, in a case reported by Davies, a male "of average build" with HD received 600 mg STP and 60 mg gallamine for induction. The patient was not intubated. He did not breathe spontaneously for 60 minutes. Fifty minutes after induction, the patient received atropine (0.6 mg) and neostigmine (2.5 mg) without effect. He then received 2 mL nikethamide (a central respiratory stimulant). The patient resumed spontaneous respirations soon after receiving the nikethamide. Recovery of consciousness was delayed. During the recovery period, the patient suffered generalized tonic spasms.

As the patient's weight was not given, the mg/kg dose of STP used could not be determined; however, it is probable that 600 mg was too great an amount for this patient. Excessive doses of STP may saturate the capacity for redistribution of this drug, causing a prolonged recovery. Nikethamide may provoke tonic-clonic convulsions, and it is possible that this caused the patient's tonic spasms.

The same patient underwent another operation 15 days later and received 400 mg STP for induction. He began to breath spontaneously within 12 minutes, but the recovery period was again prolonged and complicated by generalized tonic spasms, although nikethamide was not administered this time.

Blanloeil and associates induced a 41-year-old, 40-kg male with a 20-year history of HD with 300
mg STP and 40 mg succinylcholine. The patient received a further 100-mg dosage of STP prior to surgery. The operation lasted 15 minutes with an apneic period of 1 hour. It is also likely in this instance that the total dose of STP was excessive, contributing to a prolonged recovery period.

In subsequent case reports, STP has been used for induction without adverse effects. In these cases STP was administered in doses of less than 4 mg/kg.

The postoperative shivering seen with inhalational agents has also been noted to precipitate tonic spasms. No postoperative shivering was seen in the case presented.

Rodrigo has reported the occurrence of a bradycardia (heart rate of 34) in a patient with HD who received 50 µg fentanyl prior to induction; however, in this article, the patient had a baseline heart rate of 46. Fentanyl has been successfully used elsewhere without untoward cardiovascular effects. The patient described in this article received 150 µg fentanyl without adverse effect.

Atracurium was chosen as the muscle relaxant for intubation as it has been successfully used in this patient population, and succinylcholine has been implicated as contributing to prolonged apnea postoperatively. Gualandi and Bonfanti have suggested that patients with HD might be sensitive to depolarizing muscle relaxants and reported one case in which a female who received 50 mg succinylcholine for intubation remained apneic for 2 hours. No mention was made in their report of studies for abnormal plasma pseudocholinesterase. Although Whittaker has suggested that there is the presence of an abnormal gene controlling biosynthesis of the enzyme pseudocholinesterase in this patient population, Browne and Cross administered succinylcholine to brothers with HD without adverse effects. Dibucaine numbers in these two patients were normal. Costantino and Gross administered succinylcholine at a dose of 0.6 mg/kg to an uncooperative HD patient for a rapid sequence induction. They reported that recovery time from this drug was within the range previously reported for normal patients receiving similar doses of succinylcholine. With atracurium (0.6 mg/kg), good intubating conditions can be produced within 90 seconds, a factor desirable in a patient at risk for aspiration. Pancuronium has also been successfully used in these patients.

Reversal of the muscle relaxant was with glycopyrrolate and neostigmine. Although other authors have used atropine on this patient population, Stewart feels that anticholinergic agents may further exacerbate the choreiform movements in patients with HD and advised against their use. Glycopyrrolate, being a quaternary ammonium salt, does not cross the blood brain barrier and may be preferred over atropine because of its lack of central effects.

Propofol is an agent that may also be safely used in this patient population. Kaufman and Erb induced anesthesia in a 108-kg, 42-year-old female with 120 µg of propofol. Anesthesia was maintained with decreasing amounts of the drug in infusion and supplemented with fentanyl. At the end of the procedure, the patient opened her eyes on command 7 minutes after termination of the propofol infusion. Regaining airway reflexes as early as possible after termination of anesthesia minimizes the risk of postoperative aspiration in this patient population. Propofol may be considered as another drug in the armamentarium for patients with HD.

Summary

This article presents the successful management of a patient with Huntington's disease using a balanced anesthetic technique combining judicious amounts of narcotic and barbiturates with isoflurane. HD affects the inhibitory central effects on skeletal muscle. Any anesthetic care plan should take this into consideration, as well as the effect of the patient's nutritional status, age, and concomitant medications.

Patients with HD may be severely depressed, apathetic, irritable, or aggressive. Their families may also be highly stressed by living in the shadow of such a severely debilitating disease. They will require the support of a therapeutic caregiver who is able to understand both the physiological and psychological ramifications of this disease.

REFERENCES

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