Continuous Infusion Propofol General Anesthesia for Dental Treatment in Patients With Progressive Muscular Dystrophy

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Progressive muscular dystrophy may produce abnormal reactions to several drugs. There is no consensus of opinion regarding the continuous infusion of propofol in patients with progressive muscular dystrophy. We successfully treated 2 patients with progressive muscular dystrophy who were anesthetized with a continuous infusion of propofol. In case 1, a 19-year-old, 59-kg man with Becker muscular dystrophy and mental retardation was scheduled for dental treatment under general anesthesia. General anesthesia was maintained by a continuous infusion of 6–10 mg/kg propofol per hour and an inhalational mixture of 67% nitrous oxide and 33% oxygen. No complications were observed during or after the operation. In case 2, a 5-year-old, 11-kg boy with Fukuyama type congenital muscular dystrophy and slight mental retardation was scheduled for dental treatment under general anesthesia. General anesthesia was maintained with a continuous infusion of 6–12 mg/kg propofol per hour and an inhalational mixture of 0.5–1.5% sevoflurane in 67% nitrous oxide and 33% oxygen. No complications were observed during or after the operation. It is speculated that a continuous infusion of propofol in progressive muscular dystrophy does not cause malignant hyperthermia because serum levels of creatine phosphokinase and myoglobin decreased after our anesthetic management. Furthermore, our observations suggest that sevoflurane may have some advantages in patients with progressive type muscular dystrophies other than Duchenne muscular dystrophy and Becker muscular dystrophy. In conclusion, our cases suggest that a continuous infusion of propofol for the patients with progressive muscular dystrophy is a safe component of our anesthetic strategy.

Key Words: Propofol; Progressive muscular dystrophy; General anesthesia; Sevoflurane.

There are multiple forms of muscular dystrophy. Becker muscular dystrophy (BMD) and Fukuyama type congenital muscular dystrophy (FCMD) fall into the category of progressive muscular dystrophies. Becker muscular dystrophy is a hereditary degenerative muscular disorder caused by mutations of the dystrophin gene.[1,2] Fukuyama type congenital muscular dystrophy is an unusual form of muscular dystrophy with autosomal recessive inheritance and is clinically characterized by an early age of onset, severe central nervous system involvement, facial muscle weakness, and multiple joint contractures.[3,4] When compared with Duchenne muscular dystrophy (DMD), BMD has milder muscle weakness and a better prognosis, and FCMD progresses at a slower rate and demonstrates lower serum levels of creatine phosphokinase (CPK).

Although few reports have addressed a continuous infusion of propofol in patients with progressive muscular dystrophy,[5–8] there is supportive consensus of opinion regarding its use.[5–14] However, an inhalational anesthetic might cause some complications in patients with

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DMD or BMD.[15–17] More specifically, there are no reports of patients with FCMD being anesthetized with an inhalational anesthetic. We had the opportunity to provide general anesthesia for patients with BMD and FCMD in our hospital. This report describes 2 patients with progressive muscular dystrophy who were successfully anesthetized with a continuous infusion of propofol and inhalation of sevoflurane for dental treatment.

CASE REPORTS

Case 1

A 19-year-old, 59-kg man with BMD and mental retardation was admitted for dental treatment under general anesthesia. He was diagnosed as having BMD when he was 3 years old. He was able to walk with assistance but could not run and was cared for at home. His preoperative serum level of CPK was measured at 5155 IU/L (18–186 IU/L).

Midazolam 10 mg and famotidine 10 mg were given orally for premedication. An intravenous catheter was inserted in the patient's forearm after he was admitted to the operation room. General anesthesia was induced with 50 mg propofol intravenously, and then a continuous infusion of propofol infusion was started with 67% nitrous oxide and 33% oxygen (Figure 1). An additional supplemental dose of 50 mg propofol was given to facilitate the topical anesthesia of the nasopharyngeal cavity with 4% lidocaine with 1 : 100,000 epinephrine and to spray the vocal cords with 4% lidocaine. Nasotracheal intubation was performed easily without muscle relaxants. Anesthesia was maintained with a continuous infusion of 6–10 mg/kg propofol per hour with 67% nitrous oxide and 33% oxygen. After the intraoral injection of 4.3 mL of 3% propitocaine with felypressin, 9 teeth were conservatively treated, 2 pulpectomies were performed, and 2 teeth were extracted. The recovery from anesthesia was smooth. The tidal volume and respiratory rate had returned sufficiently to baseline, and 15 minutes after the end of the operation the patient could be easily extubated. The operation lasted 2 hours 30 minutes, and the anesthesia lasted 3 hours 30 minutes. During this time the total infusion of propofol was 1169 mg. No complications were observed during or after the operation, and the patient was discharged from the hospital the following day. Four hours before discharge the serum level of CPK had decreased to 826 IU/L.

Case 2

A 5-year-old, 11-kg boy with FCMD and slight mental retardation was admitted for dental treatment under general anesthesia. He was diagnosed as having FCMD when he was 15 months old. He had such marked loss of muscular mass that he could not walk.

Diazepam 6 mg and famotidine 2.5 mg were given orally for premedication. General anesthesia was induced with 5% sevoflurane in 67% nitrous oxide and 33% oxygen (Figure 2). An intravenous catheter was inserted in the patient's arm after induction, and general anesthesia was maintained by an inhalational mixture of 67% nitrous oxide and 33% oxygen supplemented by a continuous infusion of propofol. A dose of 1.0–2.0% sevoflurane was added when the patient moved slightly in response to the administration of topical anesthetic (4% lidocaine with 1 : 100,000 epinephrine) to the nasopharyngeal cavities. Nasotracheal intubation was easily performed without muscle relaxants after the vocal cords were sprayed with 4% lidocaine. General anesthesia was then maintained with a continuous infusion of 6–12 mg/kg propofol per hour and 0.5–1.5% sevoflurane in 67% nitrous oxide and 33% oxygen. After the intraoral injection of 1.5 mL of 2% lidocaine with 1 : 80,000 epinephrine, 2 pulpectomies were performed, 4 teeth were restored, and 3 teeth were extracted. Fifteen minutes after the end of the operation, the patient was extubated, and recovery from anesthesia was smooth and uneventful. The operation lasted 2 hours 20 minutes and the anesthesia lasted 3 hours 45 minutes. The total infusion of propofol was 312 mg. No complications were observed during or after the operation, and the patient was discharged from the hospital the following day. The serum level of myoglobin had decreased from 583.5 ng/mL at the induction of anesthesia to 391.7 ng/mL (60 ng/mL) at the end of the operation.

DISCUSSION

In case 1, inhalational anesthetics, muscle relaxants, and opioids were not administered for 3 reasons. First, it has been suggested that DMD and BMD predispose patients to developing significant complications when an inhalational anesthetic is administered.[15–17] Second, because muscle weakness and wasting associated with progressive muscular dystrophy makes it difficult to monitor the effects of muscle relaxants, prolonged apnea might occur postoperatively even when lower-than-recommended dosing regimens are used.[12,13] In case 1, as an alternative to using muscle relaxants, we performed nasotracheal intubation with topical anesthesia of the nasopharyngeal cavity by using 4% lidocaine with 1 : 100,000 epinephrine after spraying the vocal cords with 4% lidocaine. Third, opioids have been reported to produce severe respiratory depression and prolonged apnea

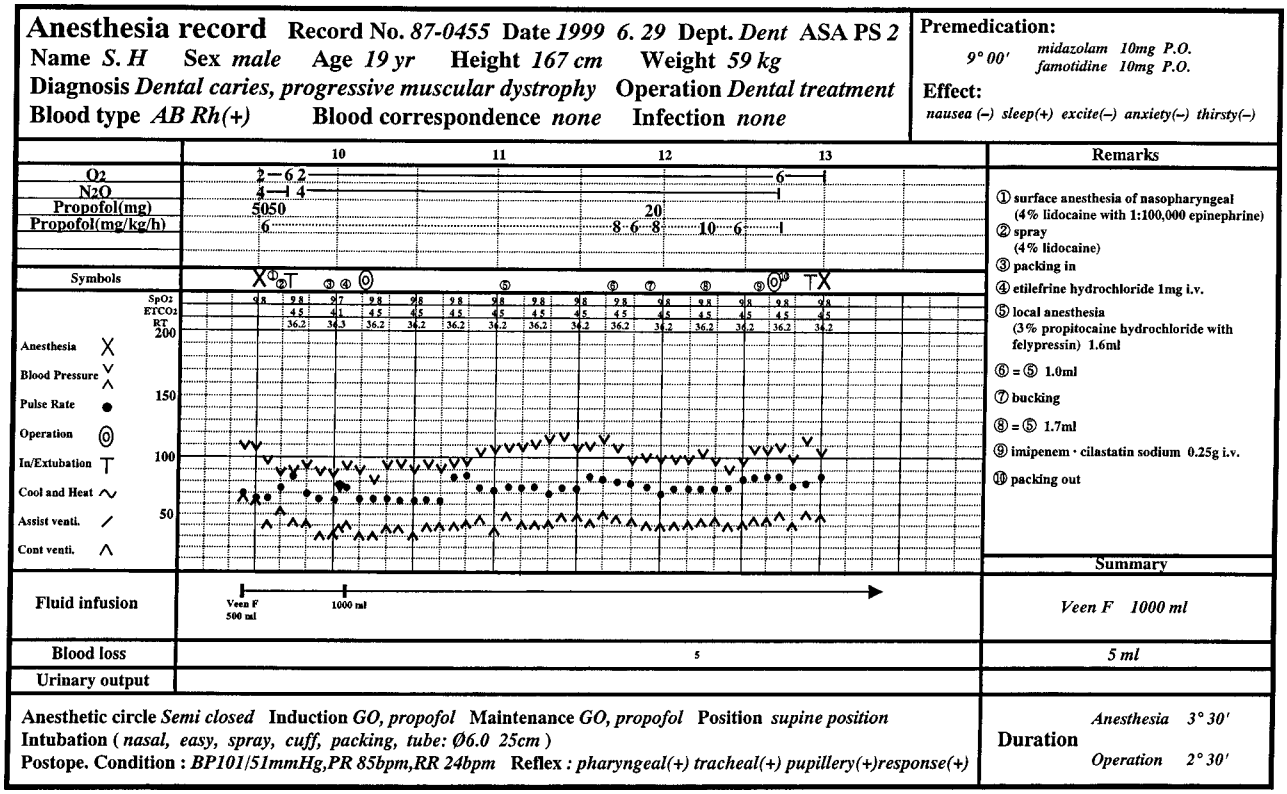

Figure 1. General anesthesia was induced with 50 mg propofol intravenously, and then maintained with a continuous infusion of 6–10 mg/kg propofol per hour with 67% nitrous oxide and 33% oxygen.

postoperatively even in patients with myotonic dystrophy that is not as severe as progressive muscular dystrophy.[18] As an alternative to postoperative pain control with opioids, we injected the local anesthetic 3% propitocaine. As a result of the combined use of local anesthetics (4% lidocaine and 3% propitocaine) and the continuous infusion of propofol plus nitrous oxide and oxygen, general anesthesia was successfully accomplished in this first case.

In case 2, general anesthesia could not be performed with the same method as used in case 1. The patient was unable to cooperate to the degree required to make placing an intravenous catheter in the arm a safe procedure. Therefore, general anesthesia was induced with 5% sevoflurane in 67% nitrous oxide and 33% oxygen. After the patient was sedated deeply, we tried to maintain the general anesthesia with a continuous infusion of 8–12 mg/kg propofol per hour in 67% nitrous oxide and 33% oxygen after the insertion of an intravenous catheter. However, the patient moved in response to the administration of topical anesthetic to the nasopharyngeal cavities for nasotracheal intubation. This indicated that the patient's level of general anesthesia was insufficient and needed either sevoflurane or opioids to supplement the continuous infusion of propofol and inhalation of 67% nitrous oxide and 33% oxygen. Anecdotal reports have suggested that inhalational anesthetics are not safe for patients with DMD or BMD.[15–17] On the other hand, some reports have claimed that sevoflurane is safe for patients with myotonic dystrophy,[19–21] and 2 reports claim that sevoflurane is used safely for the patient with progressive muscular dystrophy.[9,10] In light of these reports, and because the patient in case 2 needed a deeper level of anesthesia and because FCMD is different from DMD and BMD genetically, we decided to supplement the anesthetic with sevoflurane. We considered using opioids; however, we selected sevoflurane to prevent postoperative respiratory depression and any risk of prolonged apnea. As a result, there was no delay in recovery from the anesthetic, and the patient was extubated 15 minutes after the end of the operation. This case supports the idea that sevoflurane can be used for patients with FCMD.

The development of malignant hyperthermia and rhabdomyolysis in general anesthesia for patients with progressive muscular dystrophy is a concern. We are concerned that propofol and sevoflurane might trigger malignant hyperthermia and rhabdomyolysis in our anesthetic management. Reports have shown that a propofol infusion for a patient susceptible to malignant hyperpyrexia is safe[22] and that propofol does not trigger malignant hyperthermia because it does not activate the

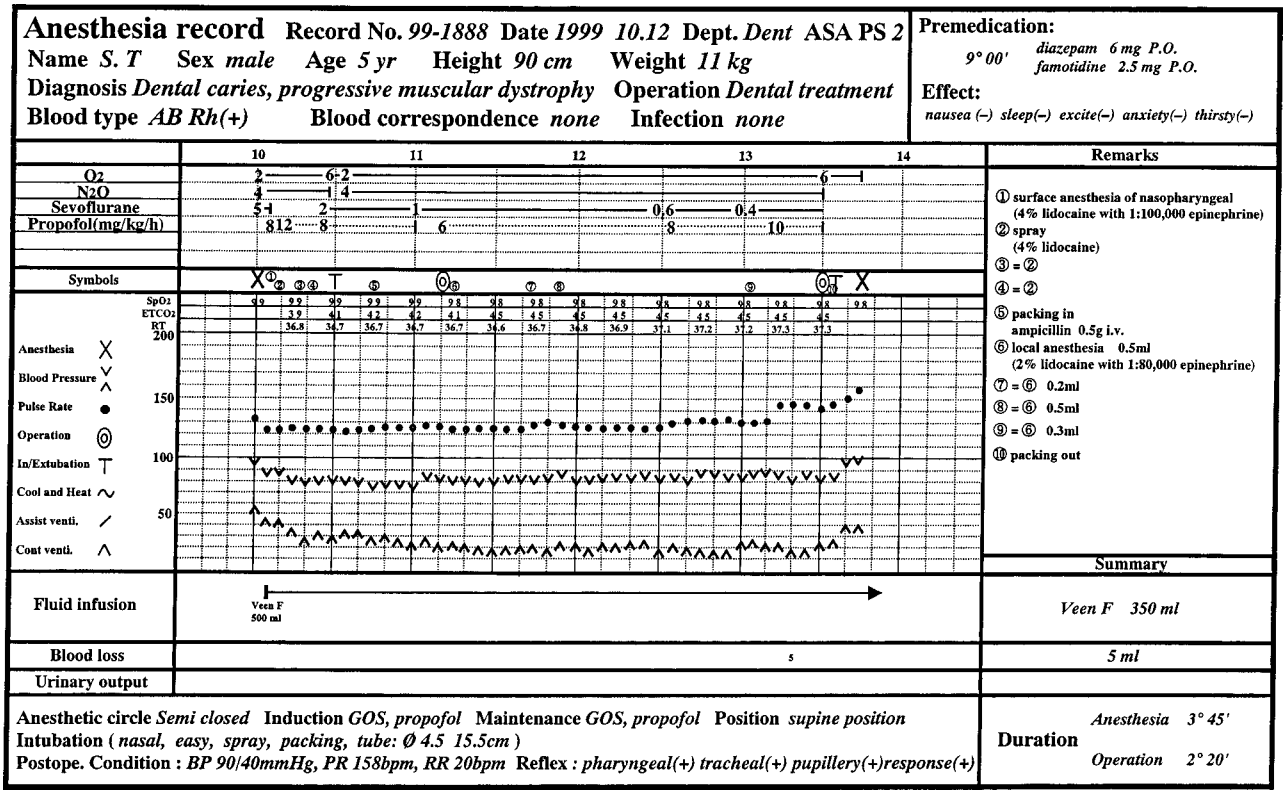

Figure 2. General anesthesia was induced with 5% sevoflurane in 67% nitrous oxide and 33% oxygen, and then maintained with a continuous infusion of 6–12 mg/kg propofol per hour and 0.5–1.5% sevoflurane in 67% nitrous oxide and 33% oxygen.

ryanodine Ca^{2+} receptor in contrast to inhalational anesthetics.[23] However, a case suspected to be malignant hyperthermia caused by propofol infusion was reported in 1996 in a letter to the editor.[24] One year later, 2 rebuttal letters were published that concluded it was unlikely the child had developed malignant hyperthermia because (a) the onset was days, not hours, after the use of a triggering agent; (b) hypercapnea was not observed as evidence of a hypermetabolic state; and (c) hyperkalemia was not present.[24,25] Also, the cause of fatalities in children sedated with prolonged infusions of propofol had not yet been clearly identified.[26] In our cases, serum levels of CPK and myoglobin decreased after operation. Generally, serum levels of CPK and myoglobin increase in malignant hyperthermia, muscular disease, and major surgery. The nature of our procedure was such that it would not be expected to influence the serum level of CPK or myoglobin. This result suggests that propofol does not cause malignant hyperthermia in progressive muscular dystrophy. Regarding sevoflurane, malignant hyperthermia and rhabdomyolysis have been reported to occur after the administration of sevoflurane in a patient with DMD.[12] However, it has also been reported that no complications were observed in general anesthesia with sevoflurane for the patients with progressive muscular dystrophy.[9,10] To prevent malignant hyperthermia and rhabdomyolysis, we assume that propofol is more appropriate than sevoflurane for patients with progressive muscular dystrophy. However, in patients with both mental retardation and progressive muscular dystrophy (except for DMD and BMD), controversy exists as to which induction method is safer, the intravenous anesthesia induction after restraint to start the intravenous infusion or the smoother induction of inhalational anesthesia with minimal restraint.

Our cases suggest that propofol infusion for the patients with progressive muscular dystrophy is safe and appropriate as an anesthetic strategy.

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