Malignant Hyperthermia during Sevoflurane Anesthesia in a Child with Central Core Disease

HIROSHI OTSUKA, M.D.,* YOSHIHIRO KOMURA, M.D.,† TAKAHISA MAYUMI, M.D., PH.D.,‡ TAKEYASU YAMAMURA, M.D., PH.D.,§ OSAU KEMMOTSU, M.D., PH.D., F.C.C.M.,¶ KEIKO MUKaida, M.D., PH.D.**

Malignant hyperthermia (MH) is a catastrophic, hypermetabolic syndrome that arises in susceptible individuals when they are exposed to certain inhalational anes-

* Instructor of Anesthesiology, Hokkaido University School of Medicine.
† Instructor of Anesthesiology, Hokkaido University School of Medicine.
‡ Assistant Professor of Anesthesiology, Hokkaido University School of Medicine.
§ Associate Professor of Anesthesiology, Hokkaido University School of Medicine.
¶ Professor and Chairman of Anesthesiology, Hokkaido University School of Medicine.
** Instructor of Anesthesiology, Hiroshima University School of Medicine.

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Address reprint requests to Dr. Otsuka: Department of Anesthesiology, Hokkaido University School of Medicine, N15 W7, Sapporo, 060 Japan.

Case Report

A 4-yr-old, 14.2-kg girl with congenital ptosis was scheduled to undergo a Berke operation (contraction of bilateral levator palpabrae superior muscles). Preoperative physical examination and laboratory data were unremarkable except for the presence of ptosis and scoliosis. Although there were no associated neuromuscular problems among family members, her mother also had congenital ptosis and had undergone uneventful regional anesthesia for the same operation. The patient had no known allergies and was receiving no medication.

She received a bromazepam suppository 3 mg and came to surgery well sedated. A precordial stethoscope, a blood pressure cuff, electrocardiogram leads, and a pulse oximeter sensor were applied before induction of anesthesia. Her heart rate was 142 beats per min. Anesthesia was induced with nitrous oxide (67%) and oxygen, and the inspired sevoflurane (MAC 1.71%) concentration was increased gradually to 3% over 5 min. An intravenous catheter was inserted, and vecuronium bromide 2 mg was administered to facilitate tracheal intubation using a 5.5-mm oral endotracheal tube. Ventilation was controlled, and anesthesia was maintained with 2–3.5% sevoflurane and 50% nitrous oxide in oxygen using a semiclosed anesthesia circuit. Because the patient was scheduled for surgery of 4 h duration, an intratracheal catheter, a rectal thermometer, and a bladder catheter were placed. Blood gas analysis before surgery revealed a pH of 7.45, arterial carbon dioxide tension of 32 mmHg, oxygen tension of 278 mmHg, and base excess of 0.1 mm. Her body temperature was 37.3°C.

Surgery proceeded uneventfully for 20 min, at which time the patient’s heart rate increased to 180 beats per min and her blood pressure increased to 142/74 mmHg. Peak inspiratory pressure also increased to 50 mmHg, but hemoglobin saturation by pulse oximetry was maintained at 98%. During the next 10 min rectal temperature increased to 38.5°C, and her legs became rigid. The changes in body temperature, heart rate, blood pressure along with blood gas analysis data are summarized in figure 1. Arterial blood gas analysis at this point showed a severe, mixed acidosis. Because of the hyperthermia, rigidity, and severe acidosis, MH was strongly suspected, and body surface cooling was immediately initiated with ice packs and a cooling mattress. Surgery was stopped immediately. Sevoflurane and nitrous oxide were discontinued, and 100% oxygen was administered through a non-rebreathing circuit.

The patient then received fentanyl 0.2 mg, dantrolene sodium 20 mg, sodium bicarbonate 1400 mg (16.7 meq), and furosemide 10 mg. A bundle branch block and slurring of the R waves were noted on the electrocardiogram. Her rectal temperature continued to increase to 40.5°C during the next 15–20 min despite aggressive cooling and improved arterial blood gas values. An additional dose of dantrolene sodium 10 mg was administered, followed by a continuous infusion at 14 mg h⁻¹. Muscle rigidity subsided slightly and rectal temperature began to decrease during the next 10 min. The patient became alert 30 min after discontinuation of anesthesia. At this time, arterial blood gas values were pH 7.35, arterial carbon dioxide tension 40 mmHg, arterial oxygen tension 575 mmHg, and base excess -1.9 mm. Urine output was 150 ml over 30 min, and no gross myoglobinuria was noted.

The patient received 245 ml of 2.5% dextrose/0.45% saline solution over 2 h peripherally. When her rectal temperature decreased to 36.0°C, she was transferred to the intensive care unit. Creatine phosphokinase values were 526 IU/l (normal = 32–180 IU/l) 1 h after surgery and 5512 IU/l the next morning. The urine myoglobin concentration remained less than 10 mg/ml. Dantrolene sodium was discontinued after 2.5 h (total dose 50 mg), and the trachea was extubated 8 h later. The patient experienced no further episodes of muscle rigidity or hyperthermia and returned to the ward on the second morning.

Five days later, a quadriiceps muscle biopsy was performed under local anesthesia with procaine and intravenous sedatives for a Ca²⁺ release test and histologic evaluation. Skinned fibers were examined for the rate of Ca²⁺-induced Ca²⁺ release from the sarcoplasmic reticulum with various concentrations of Ca²⁺ ion. The results revealed a significant increase in the rate of Ca²⁺-induced Ca²⁺ release (fig. 2), compatible with a diagnosis of MH susceptibility. Histologic examination disclosed changes typical of central core disease.

The patient was uneventfully discharged from the hospital on the 8th postoperative day.
DISCUSSION

Although Shulman et al. have reported that sevoflurane triggers MH in MH-susceptible swine, there has been no report of MH during sevoflurane anesthesia in humans. It has been shown that the release of myoglobin and creatine phosphokinase from the muscle cells during sevoflurane anesthesia is less than that during halothane anesthesia, and in MH-susceptible pigs halothane provokes more severe MH than does sevoflurane. Vecuronium, which was used in this patient, is not a triggering agent for MH in susceptible pigs, and thiopental and pancuronium have been shown to delay the onset of MH. We used no anesthetics considered to be triggering agents for MH except sevoflurane.

During the preoperative evaluation, our attention was drawn to the patient’s congenital palsy—a clinical sign, like scoliosis, that has been observed in MH susceptible patients. However, neither the family history nor the preoperative laboratory studies suggested MH susceptibility. Therefore, a diagnostic contracture test was not performed, and dantrolene was not administered prior to anesthesia and surgery.

This case report demonstrates both that sevoflurane can trigger MH in susceptible patients and also that MH triggered by sevoflurane can be successfully treated with intravenous dantrolene.

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REFERENCES


Intraoperative Use of Transesophageal Echocardiography with Pulsed-wave Doppler Evaluation of Ventricular Filling Dynamics during Pericardiectomy

CHARLES W. HOGUE, JR., M.D.,* MITCHELL PLATIN, M.D.,† BENICO BARZILAI, M.D.,‡ LARRY R. KAISER, M.D.§

Transthoracic echocardiography is the most frequently used method for the diagnosis of pericardial effusions. In addition to detecting the presence of pericardial fluid, M-mode and two-dimensional echocardiography can establish the presence of tamponade. Previous investigators have examined transthoracic M-mode, two-dimensional, and pulsed-wave Doppler echocardiography in nonanesthetized patients undergoing pericardiocentesis. We report a case in which transesophageal echocardiography proved clinically useful during pericardiectomy in a patient receiving general anesthesia.

CASE REPORT

A 59-yr-old man with a history of controlled essential hypertension presented with a recent history of a flulike illness and a pericardial effusion. Two weeks before admission the patient had had an episode of paroxysmal atrial fibrillation associated with severe shortness of breath and fatigue. The patient was treated with metoprolol, digoxin, and quinidine with successful conversion of the atrial fibrillation to sinus rhythm. Diagnostic evaluation included a transthoracic echocardiogram that showed a large pericardial effusion without tamponade. Because of increasing shortness of breath at rest, a pericardiectomy (pericardial window) was scheduled. Preoperative examination revealed an arterial blood pressure of 118/60 mm Hg and no pulse paradox or jugular venous distention, and the electrocardiogram revealed sinus rhythm at a rate of 90 beats per min.