Malignant Hyperthermia: A Case Study in the Dental Ambulatory Surgery Setting

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Historically, patients who developed malignant hyperthermia had an extremely high rate of mortality. Today, if treated appropriately, patients who experience an episode of malignant hyperthermia will most likely survive. This dramatic decrease in mortality associated with malignant hyperthermia is due to several factors, including an increased understanding of the disease, improved diagnostic and monitoring equipment, and the development of lifesaving pharmacologic agents. This article presents the very likely case of acute malignant hyperthermia in a 24-year-old man with special needs, who presented for restorative dentistry under general anesthesia in the outpatient clinic of The Ohio State University's College of Dentistry.

Key Words: Malignant hyperthermia; Anesthesia complications; Dental anesthesia; Ambulatory surgery setting.

Prior to 1979, the mortality rate for fulminant malignant hyperthermia (MH) was 64%.^{1,2} The approval of dantrolene sodium by the US Food and Drug Administration in that year enabled health care providers to successfully treat the symptoms of MH and reverse the deadly effects of the disease. Today, the mortality rate of MH has dropped dramatically; a patient who develops an acute case of MH has a 95% chance of survival if treated appropriately.^{3,4}

MH is an extremely rare, hypermetabolic disease state associated with impaired calcium ion (Ca^{2+}) transport in the sarcoplasmic reticulum of skeletal muscles. The first known case of MH was recorded in 1922; however, it was not described in scientific literature until 1951.¹ Currently, the occurrence of MH in the United States cannot be stated with complete accuracy. Several studies have stated a wide discrepancy in the incidence of MH depending

upon location, gender, age, and genetic heritage. A widely accepted statistic claims that the prevalence of MH cases is approximately 1 per 100,000 surgical patients treated under anesthesia in the hospital setting and 1 per 500,000 surgical patients treated under anesthesia in the ambulatory surgery setting.⁵⁻⁷ These statistics imply that most anesthesiologists will never encounter a case of MH during their careers.

Notwithstanding the rarity of MH, anesthesiologists should understand the pathophysiology of this deadly disease and regularly review with their colleagues and support staff the proper manner in which to treat it perioperatively. Familiarity with and adherence to approved MH protocols is essential in assuring a positive outcome when MH presents.

CASE REPORT

Case Presentation

A 24-year-old white man presented with his mother to the College of Dentistry (COD) at The Ohio State University to complete his scheduled dental treatment

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plan, which had begun 2 months earlier. He was reported to be 188 cm (6 ft 2 in) tall and 135 kg (297 lb) with a body mass index of 38.13 kg/m². The patient's medical history was significant for autism spectrum disorder, epileptic seizure disorder, attention deficit/hyperactivity disorder, and obesity. The patient's autism spectrum disorder was classified as severe (level 3), as he was nonverbal and displayed continual repetitive behaviors. His last seizure episode had been 8 months prior to the visit and was classified as a generalized motor tonic-clonic seizure. The patient's home medications included olanzapine, valproate, and sertraline, all of which he had taken by mouth approximately 2 hours prior to the surgery. He had no known drug allergies. The patient had undergone general anesthesia 2 times prior, including a general anesthetic administered 2 months previously by the primary author of this paper. No complications occurred during either anesthetic. The anesthetic plan for the current procedure was roughly the same protocol used for the last anesthetic, with the only change being the substitution of a video laryngoscope in place of a traditional laryngoscope. The patient was deemed NPO appropriate and given an American Society of Anesthesiologists physical classification of II. An accurate airway assessment was impossible to obtain preoperatively because of the patient's severe autism and inability to cooperate. However, the previous anesthetic record stated the patient's airway was easily manageable, despite the notation of a grade IV Cormack-Lehane view of the glottis.

Because of the patient's inability to sit for intravenous (IV) cannulation, an intramuscular injection of ketamine (500 mg) was administered in the right deltoid. After 5 minutes, the patient displayed adequate signs of dissociative anesthesia and was transferred to the dental operatory via wheelchair. Once the patient was situated in the dental chair, standard American Society of Anesthesiologists monitors were applied, and a 22-gauge IV was placed in the dorsal surface of the right hand.

After IV placement, the patient was preoxygenated via face mask with 100% O₂ in preparation for the further induction of anesthesia. During the preoxygenation period (approximately 2 minutes after IV placement), the patient demonstrated tonic-clonic seizure activity. The seizure was successfully broken following IV administration of midazolam (2 mg), fentanyl (100 mcg), and propofol (50 mg), plus 4% sevoflurane and O₂. Following cessation of the seizure activity, the patient's vital signs were stable, and the decision was made to continue with the procedure. Preoxygenation was performed until the end-tidal oxygen reached 90%, after which general anesthesia was induced with propofol (150 mg). Succinylcholine (200 mg) was

administered for skeletal muscle paralysis to help facilitate ease of intubation after confirming airway patency and ease of mask ventilation. Nasal intubation through the right naris was performed with a 6.5 Parker Flex-Tip cuffed endotracheal tube (ETT) using a McGrath MAC #4 video laryngoscope. A video laryngoscope was selected for this anesthetic because of the grade IV Cormack-Lehane view of the glottis that was reported in the previous anesthetic record. No complications or difficulties were reported negotiating the patient's airway while using the video laryngoscope. Proper placement of the ETT was confirmed by observation of chest rise, condensation noted within the ETT during manual ventilation, and auscultation of bilateral and equal breath sounds. The capnography waveform confirming endotracheal placement of the ETT was noted; however, the end-tidal carbon dioxide (EtCO₂) showed “+++” rather than the expected numerical value. The ETT was taped securely at a depth of 28 cm, the patient's head wrap was placed, the patient was covered with a light blanket, and a throat pack was inserted appropriately. The anesthesia machine was set to volume-control ventilation with a tidal volume of 600 mL and a rate of 14 breaths per minute administering 2% sevoflurane in 2 L/min of a 50/50% oxygen/nitrous oxide mix.

The patient was turned over to the dental team for the start of treatment approximately 6 minutes after arrival into the dental operatory. Upon reviewing the anesthesia monitors the EtCO₂ was noted to still be reading “+++.” It was postulated that the unfamiliar recording on the monitor meant 1 of 2 things: (a) there was a problem with the accuracy of the EtCO₂ monitoring, or (b) the patient's EtCO₂ was too high to be recorded.⁸

In order to elucidate the meaning of the “+++” display, several actions were immediately taken. The water trap, sample line, and CO₂ absorber were all exchanged with new parts. The breathing circuit and the ETT were checked for kinks or blockages and the patency of the 1-way valves on the machine was verified. Finally, the monitor was rebooted. When the display reappeared after the reboot, the EtCO₂ monitor continued to read “+++” and the pulse oximeter (SpO₂) dropped to 88%.

At this point, the fresh gas flows were increased to 100% oxygen at 10 L/min. The machine was switched from volume-control ventilation to spontaneous ventilation and the patient was manually hyperventilated at 30 breaths per minute. Manual ventilation was easy with no appreciable difficulties, increases in airway resistance, capnography waveform alterations, or elevated airway pressures noted. Auscultation of the lungs revealed slightly diminished breath sounds on the left side. The ETT was resecured at 27 cm and subsequent

auscultation revealed bilateral and equal breath sounds. The EtCO₂ sample line was switched to an alternate monitor and 10 puffs of albuterol were delivered directly into the ETT; the SpO₂ returned to 99%. After several minutes of manual hyperventilation, the alternate monitor showed an EtCO₂ of 80 mm Hg. The sample line was returned to the original monitor and the EtCO₂ of 80 mm Hg was confirmed. The sevoflurane was immediately discontinued.

Approximately 15 minutes after arrival to the operating room, 2 important events were noted: first, the patient's heart rate had increased from a baseline of 82 to 123 bpm, and second, the patient's axillary temperature had increased from 35.5 to 36.2°C. To confirm the occurrence of hyperthermia, a disposable temporal thermometer was applied; the reading was 38.9°C, so the blanket covering the patient was removed. The patient was examined for masseter and generalized skeletal muscle rigidity; however, all muscles were noted to be flaccid. An initial differential diagnosis of the patient's condition was highly suggestive of MH (Table 1).

As the suspicion of MH gained credibility, a cognitive aid was consulted in order to follow an approved algorithm.⁹ Additional help was recruited from the oral maxillofacial surgery faculty, and the Malignant Hyperthermia Association of the United States (MHAUS) hotline (1-800-MH-HYPER) was called.

Twenty-five minutes into the case, 7 people were enlisted to start the reconstitution of dantrolene. This was done by diluting 20-mg vials of dantrolene with 60 mL of preservative-free sterile water.⁹ Ice packs were retrieved from the freezer and packed around the patient's neck, chest, abdomen, axilla, and groin, rendering the axillary temperature probe defunct.

Approximately 5 minutes later the hypercapnia, hyperthermia, and tachycardia persisted, so the decision was made, in consultation with the MHAUS representative, to initiate dantrolene administration and activate emergency medical services by calling 911. Two additional large-bore (18-gauge) IVs were started in the patient's left and right forearms in preparation for the administration of dantrolene. Five minutes later, 18 vials of dantrolene (360 mg; ~2.66 mg/kg) were administered, all of the ice packs were replaced anew, and the patient's throat pack was removed.

Forty-five minutes after starting, the patient began to emerge from general anesthesia. A bolus of propofol (100 mg) was administered, followed by the initiation of a propofol infusion (300 mcg/kg/min) to keep the patient adequately sedated. Because the patient was in an ambulatory surgery setting, it was not possible to draw a blood sample for a blood gas analysis. Because of fear that a metabolic acidosis might be developing,

8.4% sodium bicarbonate (50 mEq) was administered via IV. Other than the sinus tachycardia noted at the onset of the case, the patient remained in sinus rhythm with no appreciable electrocardiographic abnormalities suggestive of acidosis or hyperkalemia, such as peaked T waves.

Fifty minutes after arrival to the operating room, chilled 0.9% normal saline was administered via IV and all ice packs were replaced again. At this point, emergency medical services arrived at the dental clinic. The patient was disconnected from the anesthesia circuit and hyperventilation was continued with a manual resuscitator and supplemental oxygen at 15 L/min. The patient was transferred to a gurney and removed from the COD to a waiting ambulance. The propofol infusion was discontinued prior to the patient leaving. At the time of discharge from the COD, the patient's EtCO₂ was 46 mm Hg, a sign that the dantrolene was having a positive effect.

The patient arrived at the Ohio State University Medical Center emergency department approximately 65 minutes after entry into the dental operating room. A handoff report as well as a copy of the MH cognitive aid was given to the emergency department team, who notably was unfamiliar with MH, by the primary dental anesthesia faculty member and emergency medical services personnel, and the patient was turned over to their care. At the time of turnover, the patient's EtCO₂ had risen to 51 mm Hg.

Postemergent Care

The emergency department team transferred the patient to the intensive care unit, where he remained for 4 days. He received 4 infusions of IV dantrolene via central line over the first 24 hours of his admission, during which time his creatine kinase (normal: <200 U/L) plateaued at 8979 U/L and began to trend downward. The patient's dantrolene infusions were subsequently discontinued and he was extubated. However, the patient's creatine kinase levels rebounded, so he was maintained on oral dantrolene until the levels began trending downward again. During his stay in the hospital, the patient's highest serum myoglobin level (normal: <90 mcg/L) was recorded at 2354 mcg/L, which indicated a significant amount of skeletal muscle breakdown, consistent with the anticipated rhabdomyolysis associated with MH. After 4 days in the intensive care unit, the patient was transferred to a floor bed, where he was monitored for 3 more days until being discharged, 7 days after general anesthesia was initially induced in the COD.

Table 1. Differential Diagnosis*

<i>Condition</i>	<i>Signs and Symptoms</i>	<i>Differences Compared With Malignant Hyperthermia</i>	<i>Actions to Consider</i>
Faulty equipment	If the expiratory valve is malfunctioning, hypercarbia may result	No muscle rigidity No hyperthermia	Clean or replace expiratory valve
	If the CO ₂ absorber is expired, hypercarbia may result and canister may be hot	No muscle rigidity No hyperthermia	Replace the CO ₂ absorber
	If fresh gas flows or ventilation is insufficient, hypercarbia, tachycardia, and hypertension may result	No muscle rigidity No hyperthermia	Increase fresh gas flows Increase FiO ₂
Insufficient anesthesia	Patient movement (if not paralyzed) Tachycardia Hypertension Tachypnea (if breathing spontaneously)	No muscle rigidity No hypercarbia No hyperthermia	Increase anesthetic depth Consider using a BIS monitor
Anaphylaxis	Increased airway pressures Hypotension Epidermal changes Hypercarbia	No muscle rigidity Hypercarbia responds to increased minute ventilation No hyperthermia	Administer epinephrine Activate EMS
Drug overdose (ie, cocaine, MDMA [“ecstasy”], methamphetamine)	Tachycardia/arrhythmias Hypertension Cardiac collapse Possible hyperthermia/diaphoresis	No muscle rigidity No hypercarbia Drug metabolites in urine	Supportive care and monitoring of vital signs Administer benzodiazepine Administer nitroglycerine for acute hypertension Activate EMS
Neuroleptic malignant syndrome	Use of neuroleptic medications or dopamine withdrawal Mental status changes Hyperthermia Muscle rigidity Autonomic instability	Takes place over 1–3 days (slow onset) Occurrence usually not concurrent with general anesthesia	Supportive care and monitoring of vital signs Administer benzodiazepine Administer dantrolene Administer dopamine if discontinued Activate EMS
Serotonin syndrome	Use/overdose of monoamine oxidase inhibitor or selective serotonin reuptake inhibitors Tachycardia Hyperthermia Muscle rigidity Labile blood pressure Tremor, clonus, hyperreflexia Dilated pupils	Dilated pupils No hypercarbia	Supportive care and monitoring of vital signs Administer benzodiazepine Administer serotonin antagonist (cyproheptadine) Activate EMS
Sepsis	Hyperthermia Metabolic acidosis Tachypnea Hypotension Altered mental status	Generally, no muscle rigidity No hypercarbia	Fluid resuscitation Administer antipyretic Antimicrobial therapy Administer vasopressors if needed Activate EMS
Thyroid storm	Untreated hyperthyroidism (ie, Graves disease, toxic multinodular goiter) Tachycardia/arrhythmias Hypercarbia Hyperthermia/diaphoresis Hypotension Cardiac collapse	No muscle rigidity No hyperkalemia Gastrointestinal symptoms (ie, diarrhea, vomiting)	Administer beta blocker Administer glucocorticoid Activate EMS

* From Litman¹¹ and Clinical Key Elsevier Point of Care.²⁹ BIS indicates bispectral index; EMS, emergency medical services.

Follow-up

The course of this patient's anesthetic care was discussed at length with the patient's mother. She was informed of the need for definitive testing and informed of the availability of both genetic and muscle biopsy testing options. His mother indicated that she would be pursuing the genetic testing alternative, primarily because of difficulties associated with traveling long distances with her autistic son. However, the genetic testing had not been completed prior to submission of this article.

DISCUSSION

Pathophysiology

In humans, MH occurs primarily only after a patient is exposed to specific triggering agents. Current literature supports the theory that volatile anesthetic gases (sevoflurane, isoflurane, desflurane, etc) and the depolarizing neuromuscular blocker succinylcholine trigger MH in susceptible people, as do excessive exercise and heat stress in very rare cases.^{3,10,11} MH is not triggered by a multitude of drugs, including statins, phosphodiesterase III inhibitors, 5-HT_{2A} agonists, 5-HT₃ antagonists, local anesthetics, nitrous oxide, or methylene blue, despite previous speculation.¹⁰

Normal skeletal muscle contraction occurs by a mechanism called excitation-contraction coupling (ECC) and is modulated by the movement of Ca^{2+} through the sarcoplasm of stimulated cells. When a skeletal muscle cell is depolarized by a motoneuron, an action potential is generated and propagates along sarcolemma and down into the invaginations of the cell membrane called T tubules.¹² T tubules allow the action potential to spread from the cell membrane deep into the interior of the muscle cell where the contraction units of the cell's myofibrils lie. The surfaces of T tubules have specialized calcium channel receptors embedded in them called dihydropyridine receptors (DHPRs). When the action potential wave contacts DHPRs, voltage sensors in the receptors move and cause a plug in the ryanodine receptor (RYR) protein to open. RYR proteins are calcium channels located in the terminal cisternae of the sarcoplasmic reticulum. When they are “unplugged” by the DHPRs in the nearby T tubules, large stores of Ca^{2+} are allowed to flow out of the sarcoplasmic reticulum into the cytosol of the muscle cell¹² (see Figure 1). Within milliseconds, the influx of Ca^{2+} binds with the troponin complex of the thin filament of the actin-myosin complex. The calcium-bound troponin complex induces a conformational change in tropomyosin,

(a) Calcium release from the sarcoplasmic reticulum

(b) Calcium uptake into the sarcoplasmic reticulum

Figure 1. (a) Calcium release from the sarcoplasmic reticulum. (b) Calcium uptake into the sarcoplasmic reticulum.

uncovering the active binding site of the actin, which binds to the myosin heads from the thick filament.¹³ The binding of myosin to actin causes a ratcheting motion of the thick and thin filaments as they slide past one another, ultimately resulting in muscle contraction.

Magnesium ions (Mg^{2+}) play an important role in ECC by causing an inhibitory effect on the RYR/DHPR complex.¹⁴ Under normal conditions, Mg^{2+} prevents the spontaneous opening of the RYR protein and the subsequent Ca^{2+} release from the sarcoplasmic reticulum. This magnesium inhibition is effectively overcome only when the T tubules are depolarized; depolarization of the T-tubule membrane provides the stimulus necessary to cause the DHPRs to override the magnesium inhibition and “unplug” the RYR protein from the terminal cisternae of the sarcoplasmic reticulum.

In MH, a genetic mutation of the RYR protein prevents proper inhibition of the protein by magnesium.¹⁴ As a result, triggering agents cause the RYR protein to open more easily and remain open longer, resulting in prolonged ECC and subsequent skeletal muscle contraction.^{10,15} During normal ECC, adenosine triphosphate (ATP) is metabolized every time Ca^{2+} is utilized. For example, ATP is used when Ca^{2+} unbinds from troponin during muscle relaxation and during

Table 2. Clinical Signs of Malignant Hyperthermia*

<i>Clinical Sign</i>	<i>% of Patients Demonstrating Sign</i>
Hypercarbia	92
Sinus tachycardia	73
Rapidly increasing temperature	65
Elevated temperature	52
Generalized muscle rigidity	41
Masseter spasm	27
Tachypnea	27
Sweating	18
Other	17
Cola-colored urine	14
Cyanosis	9
Skin mottling	6
Ventricular tachycardia	4
Excessive bleeding	3
Ventricular fibrillation	2

* From Larach et al.¹⁷

removal of Ca^{2+} from the sarcoplasm back into the sarcoplasmic reticulum.^{15,16} The hypermetabolic state associated with MH occurs as a result of the abnormal amounts of Ca^{2+} being released by defective RYR proteins and the exaggerated amount of ATP that is hydrolyzed as a direct result of the increased sarcoplasmic Ca^{2+} .¹⁵ Hypercarbia, hyperthermia, and lactic acidosis are the results of the MH-induced hypermetabolism.

Signs and Symptoms

MH can manifest in a variety of ways. It is critical to understand that all the signs and symptoms associated with MH may not be present in any given episode. After reviewing 255 cases of MH from the North American Registry database between 1987 and 2006, Larach and colleagues¹⁷ found that of the patients who experienced MH, 92% had significant hypercarbia, 73% demonstrated sinus tachycardia, and 65% had rapidly increasing body temperature. Larach et al¹⁷ compiled a list of the clinical signs of MH encountered during this extensive review. These signs and the relative frequencies of their presentation during MH are documented in Table 2. It is helpful to be aware that the earliest signs of MH are usually masseter spasm, tachycardia, hypercarbia, muscle rigidity, and tachypnea.^{17,18}

Diagnosis and Treatment

MH is an autosomal dominant disease that affects the RYR protein on the terminal cisternae of the sarcoplasmic reticulum.¹⁹ Mutations to the RYR protein have

been identified in 3 genes: RYR1, CACNA1S, and STAC3. However, the majority of relevant genetic variants associated with MH are found on the RYR1 gene.²⁰ Genetic testing is available for patients who have family members known to have MH susceptibility. Conclusive diagnosis of MH susceptibility by current genetic testing, however, is limited by the specificity of those tests.²⁰ Therefore, definitive diagnosis of MH susceptibility is reliant solely upon caffeine-halothane contracture testing. Unfortunately, there are only 5 locations in North America that currently provide caffeine-halothane contracture testing, making diagnosis difficult for patients in some populations.

Dantrolene sodium is currently the only Food and Drug Administration–approved drug for the treatment of MH.¹⁹ The parenteral form of dantrolene sodium comes in 2 formulations: 20 mg per vial (Dantrium, Revonto) and a concentrated 250 mg per vial (Ryanodex). In order to administer Dantrium and Revonto, 60 mL of sterile water must be used to reconstitute each 20-mg vial, whereas only 5 mL of sterile water must be used to reconstitute a 250-mg vial of Ryanodex. Obviously, this implies that the preparation of Ryanodex is much easier and quicker than the preparation of Dantrium or Revonto. According to the Food and Drug Administration as well as each manufacturer, the proper dosage of dantrolene sodium begins at 1 mg/kg and should continue until symptoms abate or until a maximum dosage of 10 mg/kg has been reached.^{21–23} However, MHAUS recommends that dantrolene sodium be initially administered at 2.5 mg/kg; MHAUS also recommends dosing dantrolene sodium according to actual weight, not ideal weight.^{2,24}

Dantrolene sodium works by increasing the RYR protein's affinity for Mg^{2+} .¹⁴ Because Mg^{2+} inhibits the RYR protein's ability to release Ca^{2+} from the terminal cisternae of the sarcoplasmic reticulum, dantrolene sodium effectively stops the uncontrolled cascade of Ca^{2+} that causes the hypermetabolic state of MH. There are no contraindications to the administration of dantrolene sodium if a patient is suspected of having an episode of MH.²¹ Patients who receive dantrolene sodium may experience generalized skeletal muscle weakness, respiratory depression, pulmonary edema, thrombophlebitis, and urticaria.²¹ It is important that dantrolene sodium is not allowed to extravasate into peripheral tissues, as it has a basic pH of 9.5, which can cause tissue necrosis.

Recognizing MH

Because access to definitive diagnostic testing is limited, and because the initial presentation can vary, a clinical

Table 3. Signs and Symptoms in Determining MH According to the Larach Clinical Grading Scale*

<i>Process</i>	<i>Sign or Symptom Present</i>	<i>Points</i>	<i>Present in This Case</i>
I. Rigidity	Generalized muscular rigidity	15	
	Masseter spasm after succinylcholine administration	15	
II. Muscle breakdown	Elevated creatine kinase >20,000 if succinylcholine used	15	
	Elevated creatine kinase >10,000 if succinylcholine not used	15	
	Cola-colored urine in perioperative period	10	
	Myoglobin in urine >60 mcg/L	5	
	Myoglobin in serum >170 mcg/L	5	✓
	Blood/plasma/serum K ⁺ >6 mEq/L	3	
III. Respiratory acidosis	PETCO ₂ >55 mm Hg with controlled ventilation	15	✓
	Arterial PaCO ₂ >60 mm Hg with controlled ventilation	15	
	PETCO ₂ >60 mm Hg with spontaneous ventilation	15	
	Arterial PaCO ₂ >65 with spontaneous ventilation	15	
	Inappropriate hypercarbia (anesthesiologist's judgement)	15	
	Inappropriate tachypnea	10	
IV. Temperature increase	Inappropriate rapid rise in temperature (anesthesiologist's judgement)	15	✓
	Increased temperature >38.8°C (101.8°F)	10	
V. Cardiac involvement	Inappropriate sinus tachycardia	3	✓
	Ventricular tachycardia or ventricular fibrillation	3	
VI. Family history	Positive MH family history in relative of first degree	15	
	Positive MH family history in relative not of first degree	5	
VII. Other indicators	Arterial base excess more negative than −8 mEq/L	10	
	Arterial pH <7.25	10	
	Rapid reversal of MH signs of metabolic/respiratory acidosis with IV dantrolene	5	✓
Total score in this case			43

* From Larach et al.²⁵ MH indicates malignant hyperthermia; K⁺, ??; PETCO₂, ??; PaCO₂, ??; and IV, intravenous.

grading scale was developed by Larach and colleagues²⁵ to predict the likelihood of MH when signs and symptoms are observed in suspect patients. This scale was designed using the Delphi method to be an objective aid in identifying the disease and has been used and referenced in many peer-reviewed journals.^{3,10,26–28} The Larach clinical grading scale assigns weighted points to each of the signs and symptoms observed during a suspected MH episode.³ Those points are then summed to give a total score, which is translated into an MH risk score between 1 and 6. A risk score of 1 means that the cumulative observed signs and symptoms “almost never” indicate a case of MH. A risk score of 6 means that the cumulative observed signs and symptoms

indicate that a case of MH is “almost certain.”²⁵ It is important to understand that the score is not a percentage of the chance of MH, but rather is only to be seen as a qualitative indicator of the disease.²⁵ It is also important to understand that this grading scale has limitations because all of the observations and tests may not be available during an episode of MH.³ Despite the deficiencies of the Larach clinical grading scale, it provides an interesting and helpful insight into the likelihood of a true MH episode. It is worth noting that the suspected MH case outlined in this report received 43 points according to the Larach clinical grading scale. This translates into a risk score of 5, indicating that a definitive diagnosis of MH in this case was “very likely.” A summary of the Larach clinical grading scale and the signs and symptoms observed and scored during this case are presented in Tables 3 and 4.²⁵

Table 4. Interpretation of the Larach Clinical Grading Scale*

<i>Points From Observed Signs and Symptoms</i>	<i>Risk Score</i>	<i>Likelihood of MH</i>
0	1	Almost never
3–9	2	Unlikely
10–19	3	Somewhat less than likely
20–34	4	Somewhat greater than likely
35–49	5	Very likely
50 +	6	Almost certain

* From Larach et al.²⁵ MH indicates malignant hyperthermia.

CONCLUSION

MH is a rare yet potentially fatal disease that must be recognized and treated appropriately by the anesthesia team for the patient to have a good chance at survival. The hypermetabolism of MH is related to the unchecked influx of Ca²⁺ into the sarcoplasm of skeletal muscles that results in abnormal and prolonged ECC. MH is

Figure 2. (a) Relaxation. (b) Contraction.

typically triggered by exposure to volatile anesthetic agents and/or succinylcholine. Common signs and symptoms of the disease includes masseter spasm, tachycardia, persistent hypercarbia despite hyperventilation, skeletal muscle rigidity, and increased body temperature. The definitive treatment for MH is IV dantrolene as soon as a valid suspicion of the disease is confirmed. This article presents the very likely case of acute MH in a 24-year-old special needs male patient in a dental ambulatory setting and the corrective actions taken to satisfactorily manage this condition.

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