

## Crisis Management in Malignant Hyperthermia

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### ABSTRACT

The following case report describes malignant hyperthermia in an ambulatory surgical pediatric patient complicated by hypothermia and thrombophlebitis. It highlights the importance of teamwork and adhering to standardized checklists to maximize patient safety and efficiency while caring for a patient in a crisis situation.

**Keywords:** Protective devices; Equipment; Pandemics; Breathing filter system

### INTRODUCTION

Malignant Hyperthermia (MH) is a potentially life-threatening phenomenon occurring after the administration of succinylcholine or volatile anesthetic gases. The incidence of MH is between 1 in 10,000 to 1 in 250,000 anesthetics [1]. The pathophysiology of MH is the result of an autosomal dominant genetically mediated disorder due to a variant in the ryanodine receptor [2]. Release of calcium from the sarcoplasmic reticulum causes excessive muscle contraction which results in a hypermetabolic state, including a metabolic and respiratory acidosis, hyperthermia, and rhabdomyolysis. If untreated, hyperkalemia, myoglobinuria, elevated Creatine Kinase (CK), acute kidney injury, cardiac dysrhythmias, and in up to 10% of MH patients, even death may ensue [3].

### CASE STUDY

The patient was a 16 year old African American male 183 cm, 69.4 kg with no significant past medical history undergoing hip arthroscopy in a freestanding university hospital ambulatory surgical center. He had previously received one uneventful general anesthetic.

The patient was monitored with standard ASA monitoring, including a skin temperature probe. After anesthetic induction with fentanyl, lidocaine and propofol, a laryngeal mask airway was placed and anesthesia was maintained with sevoflurane.

Approximately 50 minutes after induction of anesthesia the end-tidal carbon dioxide (ETCO<sub>2</sub>) began to rise and peaked at 98 mmHg despite a tripling of the minute ventilation. The patient's temperature increased (peaked at 38.3°C) and he developed sinus tachycardia. A presumptive diagnosis of MH was made and

additional staff and the MH kit were called for emergently, which included all necessary drugs, equipment and a checklist. Sevoflurane was discontinued, the patient was started on a propofol infusion and the surgeon rapidly completed the surgery.

An anesthesiologist led the management of the crisis by reading aloud the MH checklist, and assigning tasks to specific personnel while receiving direct feedback and updates. The full range of skills involving the pharmacist, secretaries, nurses, surgical team (resident and attending), anesthesiology technician and anesthesiology staff (CRNAs and attending anesthesiologists) were appropriately utilized to ensure all of the tasks were completed timely and safely.

Dantrolene was prepared, the patient was intubated, the soda lime absorber and circuit were exchanged, a Foley catheter was placed, snow was obtained from outside and placed on the patient for active cooling, a rectal temperature probe was employed, a second large bore IV and arterial catheter were inserted. An arterial blood gas was drawn on 100% oxygen and driven directly to the hospital lab: pH 7.15, PaCO<sub>2</sub> 74, PO<sub>2</sub> 424, bicarbonate 25 mEq/L, base excess -5 mEq, potassium 5.1 mEq/L, lactate 2.4 mmol/L.

The patient received dantrolene 180 mg within 15 minutes of presumed MH diagnosis. He also received sodium bicarbonate and intravenous fluid. Shortly after dantrolene administration there was a reduction in ETCO<sub>2</sub> (98 to 66 mmHg), temperature (38.3°C to 37.4°C), and heart rate (initial 35-45 beats/min decrease).

The patient was transported with the anesthesiology team by ambulance with ETCO<sub>2</sub> and vital sign monitoring to the

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**Received:** March 01, 2021; **Accepted:** March 15, 2021; **Published:** March 22, 2021

**Citation:** Rubin MA, Lustik SJ, Spring CI, Pyne SG (2021) Crisis Management in Malignant Hyperthermia. J Anesth Clin Res. 12:995.

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hospital. Initial vital signs on arrival to the emergency department revealed a core temperature 33.8°C and the patient was actively warmed. The serum potassium was 7.6 mEq/L and the patient received calcium, insulin, and dextrose in addition to redosing of dantrolene.

The patient was admitted to the intensive care unit, continued to receive dantrolene therapy and was extubated on postoperative day 1. CK peaked 46 hours after induction at 940 units/L. His course was complicated by thrombophlebitis in the left upper extremity, the site of dantrolene administration. He was discharged home two days later in good health. He and his family were counseled regarding the diagnosis of malignant hyperthermia. They were referred to the Malignant Hyperthermia Association of the United States (MHAUS) and to follow up with their primary care physician.

## DISCUSSION

By using a checklist, the crisis team leader was able to ensure that all aspects of treating this event were considered and appropriately treated. This resulted in minimizing complications and successful recovery of the patient.

Known complications of MH include rhabdomyolysis, hyperkalemia, and death. The patient did not experience overt signs of rhabdomyolysis including no evidence of muscle necrosis, muscle pain, myoglobinuria, acute kidney injury, or markedly elevated CK levels. Surgery itself has been shown to cause elevated CK levels and often the CK peak after an MH event does not exceed the expected CK changes due to the surgery [4].

Hyperkalemia is known to occur with MH due to the excessive muscle breakdown that can rapidly occur, particularly in muscular patients like this patient. The patient's initial serum potassium was 5.1 mEq/L and no treatment was administered, but following transfer to the hospital, serum potassium was 7.6 mEq/L. It seems likely that the patient continued to experience continued muscle breakdown and another dose of dantrolene should have been administered prior to transfer.

The patient's temperature peaked at only 38.3°C, possibly due to the early administration of dantrolene. Active cooling with ice unintentionally further cooled the patient to 33.8°C. Active cooling of the patient should have ceased when his temperature was 38°C to prevent hypothermia [5].

Dantrolene is the only known definitive treatment for MH as it directly reverses the hypermetabolic skeletal muscle process within minutes. Typical dosing consists of a 2.5 mg/kg IV loading dose followed by 1 mg/kg IV bolus doses as needed until signs and symptoms improve. Other treatment focuses on removing all potential triggers, ventilating with 100% oxygen, monitoring acid/base and electrolyte disturbances, coagulation parameters, as well as temperature management [6]. This should be followed by supportive care in a hospital floor or ICU depending on the severity of the reaction.

The patient experienced phlebitis and thrombosis in the left cephalic vein at the site of dantrolene infusion which required anticoagulation. Phlebitis occurs in up to 9% of patients

receiving dantrolene [7]. The mechanism is thought to be attributed to the insolubility of dantrolene and its alkaline pH. Another study suggested that dantrolene may cause skeletal weakness and result in a higher likelihood of thrombogenesis [8]; however, given the location of phlebitis in this patient, the latter mechanism seems less likely. Complications may be related to the total dantrolene dose administered [7]. Dantrolene should be administered through a large-bore IV or central access and sites should be assessed for evidence of phlebitis.

The importance and benefits of checklists in clinical practice is well established. Over the last decade healthcare has turned to the field of aviation industry and its ability to effectively utilize Crew Resource Management training and checklists to reduce complications [9]. Checklists were developed in aviation to help crews prevent the most common mode of failure: human error.

The Agency for Healthcare Research and Quality aims to create similar improvements in patient safety by advocating for the adoption of similar methods, such as checklists, that can increase patient survival and ultimately reduce harm. Although teams that follow checklists may be limited by the depth of their knowledge, judgment, and technical skills, it is the responsibility of the leaders of these teams to be familiar with these checklists so that when a crisis is identified, they are best equipped to handle the situation effectively and efficiently.

Checklists during a medical crisis improve teamwork, effective communication, and appropriate task delegation which will more likely reduce error and avoid harm to a patient. In a randomized study, anesthesiologists given a checklist to treat simulated patients for MH demonstrated improvements in adherence to guidelines, non-technical performance and speed to administer dantrolene [10]. In our patient, the crisis team leader was able to stand back and direct management without being distracted from hands-on care. The checklist ensured that all appropriate steps were taken and guided an effective handoff. The severity of this patient's symptoms was likely minimized by the appropriate and efficient action of the providers.

The clinical grading system established by Larach et. al. standardizes the qualitative likelihood of a patient having MH using clinical signs [11]. This patient had a total score of 51 points which made the diagnosis of MH almost certain (Table 1).

Indicator	Points
Rigidity	0
Family History of MH	0
Serum potassium >6 mEq/L	3
Inappropriate sinus tachycardia	3
Rapid reversal of MH signs with IV dantrolene	5
Arterial pH <7.25	10

Inappropriate rapid increase in 15 temperature	
ETCO <sub>2</sub> >55 mmHg with 15 appropriately ventilation controlled	
Total	51

<sup>a</sup>Calculated using the clinical grading scale by Larach, et al.

<sup>b</sup>Abbreviations: MH: Malignant Hyperthermia; ETCO<sub>2</sub>: End Tidal Carbon Dioxide

**Table 1:** Results of MH clinical grading scale in patient<sup>a</sup>.

A careful discussion with the patient's parents revealed an unexplained postoperative death in a family member, but few details were known. Given the autosomal dominant inheritance pattern of MH and a potential positive family history, it was recommended for the family to undergo further testing. Due to the high likelihood that our patient is MH susceptible, genetic testing has advantages over the Caffeine-Halothane Contracture Test (CHCT).

## CONCLUSION

The genetic test is more convenient, cheaper, and less invasive and if a causative mutation is identified it enables other family members to be tested. However, since not every MH susceptible patient has a known causative mutation, MH cannot be ruled out by a negative genetic test and the CHCT would be considered the gold standard because MH is potentially life threatening and rare, it is important to have a defined treatment strategy. Early detection and treatment is vital in minimizing morbidity and mortality in this condition. Dantrolene has side effects which should be considered during its administration. The use of standardized checklists allows for the greatest degree of patient safety while also improving efficiency, teamwork, and communication.

## CONFLICTS OF INTEREST

None

## CONTRIBUTION

**Michael Rubin:** This author helped with the literature search and composition and revision of the manuscript

**Stewart Lustik:** This author helped care for the patient and with the literature search, writing, and editing the manuscript.

**Camberly Spring:** This author helped with the literature search, editing and preparation of the manuscript.

**Sonia Pyne:** This author helped care for the patient and with writing, revision and editing of the manuscript.

**Attestation:** Sonia Pyne is the corresponding author, All authors approved the final manuscript.

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