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### Malignant Hyperthermia

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# Malignant Hyperthermia

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## Introduction

Malignant hyperthermia (MH) is a serious, life-threatening emergency that any diligent certified registered nurse anesthetist (CRNA) needs to be aware of. Along with a myriad of conditions and potential complications that can occur during anesthesia, MH stands out as one that requires prevention, prompt recognition, and treatment to avoid rapid deterioration of the patient's condition.

Malignant hyperthermia is known as a pharmacogenetic disorder which manifests itself in the skeletal muscle (Heytens, Forget, Scholtès, & Veyckemans, 2015). When a susceptible patient, who carries the autosomal dominant trait, is exposed to volatile anesthetics and/or the neuromuscular blocker succinylcholine, a detrimental response can occur. This response is a hypermetabolic state with hypercapnia, hemodynamic instability, rigidity, hyperthermia, and signs of rhabdomyolysis (Heytens et al., 2015). This response is due to the calcium release from the sarcoplasmic reticulum into the cytosol (MacKay, Wilkerson, Kraeva, Rosenberg, & Kennedy, 2016). MH is a rare condition with incidences between 1/5,000 and 1/50,000 (Nagelhout and Plaus, 2014, p. 829), and often occurs during the induction of anesthesia but can also occur intraoperatively or one hour post operatively (Cain, Riess, Gettrust, & Novallija, 2014). Because MH can manifest itself at several different periods during the perioperative phase, CRNA's must be conscientious health care providers. Incidences occur in patients who carry the mutated autosomal dominant type-1 ryanodine receptor (RYR1) gene (Riaz et al., 2014). If these patients who carry the mutated gene are exposed to triggering agents, and MH is not properly recognized and treated, the result could be irreversible damage and potential death due to the sustained hypermetabolic state the body endures. It is worth exploring the pathophysiology, signs, symptoms, and treatment to prevent this condition from happening.

## Pathophysiology

Malignant hyperthermia occurs in patients who have a genetic defect in their ryanodine receptor subtype 1 (RYR1), which is a large ion channel that facilitates the release of calcium from the sarcoplasmic reticulum (SR) in skeletal muscle (Schneiderbanger et al., 2014). Calcium is released from the SR during muscle contraction and normally undergoes reuptake after muscle contraction has been completed. During an MH crisis, when susceptible patients are exposed to triggering agents (volatile anesthetics and succinylcholine), there is a prolonged opening of the RYR1 receptor channels, which results in an uncontrolled release of calcium and unrelenting muscle activation which presents as rigidity (Schneiderbanger et al., 2014). This sustained muscle contraction causes increased sympathetic activity such as tachycardia and hypertension, increased oxygen consumption, production of carbon dioxide, and high body temperature (Schneiderbanger et al., 2014). Cellular adenosine triphosphate stores are depleted, and aerobic metabolism is eventually useless and anaerobic metabolism begins. This produces lactic acid and ultimately causes cell death and destruction (Riaz et al., 2014). This breakdown of cellular membrane integrity leads to the release of potassium, myoglobin, and creatine phosphokinase into the bloodstream (Nagelhout and Plaus, 2014). The release of these contents into circulation can further cause rhabdomyolysis, cardiac arrhythmias, pulmonary edema, and disseminated intravascular coagulation (DIC) (Riaz et al., 2014).

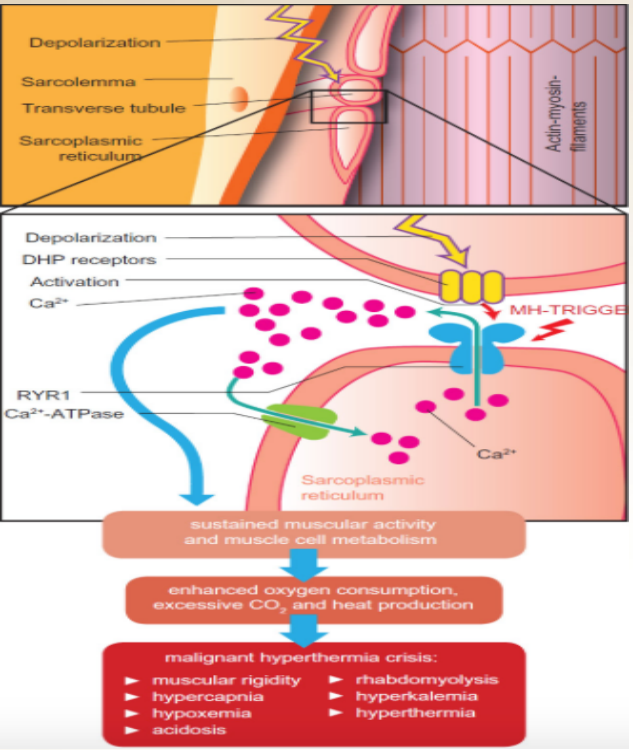
Table 1

Early Signs and Symptoms	Late Signs and Symptoms
Increased ETCO2	Cyanosis
Muscle Rigidity	Pyrexia
Cardiac abnormalities	Disseminated intravascular coagulation
Generalized erythematous flush	Left ventricular failure
Electrolyte imbalance	Metabolic acidosis

(Stratman, Flynn, & Hatton, 2009)

## Signs and Symptoms

- Early clinical signs of MH include increasing heart rate and end tidal carbon dioxide (Stratman, Flynn, & Hatton, 2009)
- Hyperthermia, with temperature rising as quickly as 1 degree Celsius every 5 minutes (Stratman, Flynn, & Hatton, 2009)
- Muscle rigidity, especially in the jaw, chest, and extremities (Stratman, Flynn, & Hatton, 2009)
- Later signs include acidosis, cyanosis, and various electrolyte imbalances that can also manifest as cardiac arrhythmias (Stratman, Flynn, & Hatton, 2009)
- Rhabdomyolysis with myoglobinemia, myoglobinuria, hyperkalemia and acute renal failure (Stratman, Flynn, & Hatton, 2009)
- Any of the above listed signs and symptoms can quickly turn into cardiac arrest (Stratman, Flynn, & Hatton, 2009)
- Refer to Table 1 for detailed early and late signs and symptoms



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## Significance of Pathophysiology

Malignant hyperthermia causes significant and potentially lethal distress to the body if not properly recognized and treated. But it's important to realize that since this condition is genetically linked, it can be avoided. Because of its significance, malignant hyperthermia should be a priority during an anesthesia provider's preoperative assessment. Patient's should be asked if they or their family members have ever experienced any complications related to anesthesia, specifically regarding a high fever or muscle rigidity. This simple assessment can avoid the risks associated with volatile anesthetics and depolarizing neuromuscular blockers, and prevent the patient from experiencing deadly symptoms such as ventricular fibrillation, rhabdomyolysis and renal failure, pulmonary edema, DIC, etc. This condition shares many of the same signs and symptoms of other pathological disorders. It becomes even more important to be diligent with assessment skills and understand the pathology to comprehend how this might happen and cause a lethal condition. Because it is such a pathologically serious disorder, increased education for patients, their families, and providers are necessary and important to provide safe anesthetic care.

## Implications for Nursing Care

As mentioned previously, the goal should be to avoid triggering agents in the presence of someone who has a history of MH or has a family member with a history of MH. There are often special OR's with specific equipment that can be used in these circumstances. However, the RYR1 defect may not be known and a MH crisis may occur in the middle of surgery. MH should be promptly recognized and many things needs to happen at the same time.

- All triggering agents should be discontinued, an increase in oxygenation to 100% FiO<sub>2</sub> should be completed, and the patient should be given non-triggering relaxants and anesthetics such as propofol and/or midazolam (MHAUS, 2015)
- Dantrolene should be administered. Dantrolene is a specific ryanodine receptor antagonist that prevents calcium from being released and prevent the muscle cells from contracting (Seifert, Wahr, Pace, Cochrane, & Bagnola, 2014). The dose of dantrolene is 2.5 mg/kg every 5 minutes repeatedly until a maximum dose of 30 mg/kg has been reached. This medication should be rapidly infused through a large bore IV (Seifert et al., 2014)



\* Retrieved from <http://embed.wistia.com/deliveries/224a3f7c8025681839b7662b42a9ccc6c17f9a0a.jpg>

- The patient should also be cooled with ice packs, cold wraps, and/or chilled IV normal saline with an attempt to keep their temperature less than 39 degrees Celsius (Dirksen, Van Wicklin, Mashman, Neiderer, & Merritt, 2013)
- Metabolic acidosis and hyperkalemia may be treated with sodium bicarbonate, hyperventilation, glucose and insulin and arterial blood gases, electrolytes, and blood levels should be checked every 15 minutes (Dirksen et al., 2013)
- It should also be a goal to maintain the patient's urine output greater than 2 ml/kg/hr with adequate hydration, furosemide, and mannitol as needed (Nagelhout and Plaus, 2014, p. 829).
- While everything else is going on, an OR team member should also call the MH hotline at 1-800-644-9737 and be prepared to give name, number, facility and email (MHAUS, 2015)

## Conclusion

Although malignant hyperthermia is a rare medical emergency, it is vital that clinicians and patients are prepared and educated for the surgical procedure and the risk for complications with a detailed plan in place (Johns, Stoudt, Scholtis, & Gavel, 2012). Malignant hyperthermia requires immediate recognition and treatment to increase the patient's chance of survival. Providers need to know its triggers (volatile anesthetics and succinylcholine), and a thorough pre-operative assessment must be completed to identify any history or risk factors for MH. It is important that all OR staff regularly review the standards and procedure for dealing with an MH crisis, as it is a rare occurrence. The whole operating room must work as a team and be familiar with signs and symptoms of MH and understand the protocol in order to properly react and treat the patient for the best possible outcome.

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