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### Malignant Hyperthermia

Brennon Pinion  
pinion1@otterbein.edu

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# Malignant Hyperthermia

Brennon Pinion, BSN, RN, CCRN  
Otterbein University, Westerville, Ohio

## Introduction

### Malignant Hyperthermia (MH)

- Malignant Hyperthermia is a rare hypermetabolic condition (Cornelius et al., 2019).
- MH is a disorder of the skeletal muscle (Lapisatepun & Arkarattanakul, 2020).
- It is caused by a genetic mutation of ryanodine receptor proteins in skeletal muscle cells (Cornelius et al., 2019).
- MH is caused by inhaled anesthetics and the paralytic medicated succinylcholine (Gallegos & Hennen, 2022).
- The first case in the United States was reported in 1922 (Cornelius et al., 2019).
- MH was fully described in 1951 (Cornelius et al., 2019).
- MH affects 1/100,000 patients undergoing surgery in an inpatient setting (Cornelius et al., 2019).
- It affects 1/500,000 patients undergoing surgery in outpatient settings (Cornelius et al., 2019).
- 1/15,000 pediatric surgical patients are diagnosed with MH (Lapisatepun & Arkarattanakul, 2020).

## Significance

- MH historically had a fatality rate of 70% (Gallegos & Hennen, 2022).
- The fatality rate is now 5% when appropriate treatment is given quickly (Cornelius et al., 2019).
- While there are treatments that can significantly improve outcomes, anesthesia providers must be able to quickly recognize the condition in order to begin treatment (Gallegos & Hennen, 2022).
- Without recognition, emergency medications likely cannot be given fast enough.
- The faster the medications are given, the higher the survival chance is (Gallegos & Hennen, 2022).

## Signs and Symptoms

- The initial symptoms are often unexplained (Hopkins et al., 2021).
- MH can cause:
- Jaw stiffness
  - Limb rigidity
  - Hyperthermia
  - Tachycardia
  - Hypercarbia
  - Flushed skin (Hopkins et al., 2021)

MH can lead to deadly conditions such as:

- Hyperkalemia
- Rhabdomyolysis
- Disseminated intravascular coagulopathy (DIC)
- Acidosis (Hopkins et al., 2021)

Due to the specific combination of symptoms that can mimic other problems, other conditions must be quickly ruled out. Other conditions that look similar to MH that should be ruled out include:

- Anaphylaxis
- Neuroleptic malignant syndrome (NMS)
- Sepsis
- Serotonin syndrome (Cornelius et al., 2019)

## Underlying Pathophysiology

- MH is caused by certain types of anesthesia medications, such as sevoflurane, isoflurane, desflurane, and succinylcholine (Cornelius et al., 2019)
- In normal muscle physiology, action potentials move down T-tubules and open channels by stimulating dihydropyridine receptors
- Dihydropyridine receptors then open ryanodine receptor proteins.
- When these proteins open, calcium that is stored in the sarcoplasmic reticulum is released.
- The calcium quickly binds to a troponin complex, which causes actin to bind to myosin, causing muscle contraction.
- The ryanodine receptor protein is inhibited by magnesium, stopping the outflow of calcium, stopping the muscle contraction (Cornelius et al., 2019).
- In Malignant Hyperthermia, there is a mutation in the ryanodine receptor proteins.
- This mutation causes the proteins to block the magnesium, preventing inhibition of the receptor.
- Because the receptor is not inhibited, too much calcium is released from the sarcoplasmic reticulum.
- With too much calcium flowing out and binding to troponin complexes, muscles are contracted for too long and cannot relax.
- This causes too much ATP to be used during the muscle contraction (Cornelius et al., 2019).

## Significance of Pathophysiology

- To keep muscle cells alive during the extended contractions, more ATP must be produced and used.
- This causes increased metabolism, creating the "hypermetabolic" condition.
- The increased utilization of ATP causes increased CO<sub>2</sub>, which causes acidosis and tachypnea (Cornelius et al., 2019).
- The increased metabolism also creates heat, causing hyperthermia and flushed skin (Cornelius et al., 2019).
- The hyperkalemia caused by hypermetabolism can cause deadly cardiac dysrhythmias (Hopkins et al., 2021).
- The pathophysiology of MH can be extremely dangerous and must be treated as fast as possible (Kleidon, 2020).
- MH can cause hemodynamic instability, dysrhythmias, and death (Gallegos & Hennen, 2022).
- It can also cause rhabdomyolysis (Otta et al., 2021).
- MH can lead to kidney injury (Tsutsumi et al., 2021).
- After treatment, patients go to the Intensive Care Unit for further monitoring and treatment (Kleidon, 2020).

## Treatment

- MH is difficult to diagnose, so the Larach Clinical Grading Scale is used to score patients based on symptoms to determine likelihood of MH (Cornelius et al., 2019).
- Once MH is recognized in the operating room, the medication that caused it must be stopped immediately.
- The treatment for MH is Dantrolene Sodium. Dantrolene blocks the effects of dihydropyridine receptors, decreasing the release of sodium.
- This stops the contractions that cause the hypermetabolic state (Lapisatepun & Arkarattanakul, 2020).
- The treatment starts with 2.5mg/kg of Dantrolene and then increased until symptoms stop.
- The maximum dose of Dantrolene is 10mg/kg (Cornelius et al., 2019).

After the initial MH treatment is given to prevent further damage, the effects of MH must be treated. These include:

- Sodium bicarbonate for acidosis
- IV calcium and glucose/insulin for hyperkalemia
- Amiodarone for dysrhythmias
- Sodium bicarbonate for myoglobinuria
- Coagulation transfusions for DIC (Hopkins et al., 2021)

## Implications for Nursing Care

- Because of the detrimental effects of MH, nurse anesthetists must always be aware of the possibility of MH, even though it is extremely rare (Cornelius et al., 2019).
- It is treatable when caught early, so nurse anesthetists must recognize the condition quickly (Cornelius et al., 2019).
- Understanding the pathophysiology is vital to recognize the condition and treat adequately.
- Susceptible patients can be screened with a surgical biopsy (Carlson et al., 2019).
- Cognitive aids should be posted anywhere that anesthesia is administered to shorten treatment time (Gallegos & Hennen, 2022).
- There should also be MH emergency kits anywhere that anesthesia is administered containing treatment medications and devices (Gallegos & Hennen, 2022).
- Dantrolene should be available to administer within 10 minutes (Lapisatepun & Arkarattanakul, 2020).

## Conclusion

- Malignant Hyperthermia is hypermetabolic condition that occurs after some patients receive certain anesthetic medications during surgery (Kleidon, 2020).
- MH is an extremely dangerous condition that everyone at a surgical facility needs to be aware of.
- Having a plan in place to treat MH is essential to preventing death caused by the condition.
- In order to prevent MH deaths anesthesia providers must understand the pathophysiology behind it and how to treat it.
- The death rate from MH has decreased dramatically but only if treatment is given quickly (Cornelius et al., 2019).
- All providers must remain vigilant of the disease even though it is very rare (Cornelius, et al., 2019).

## References



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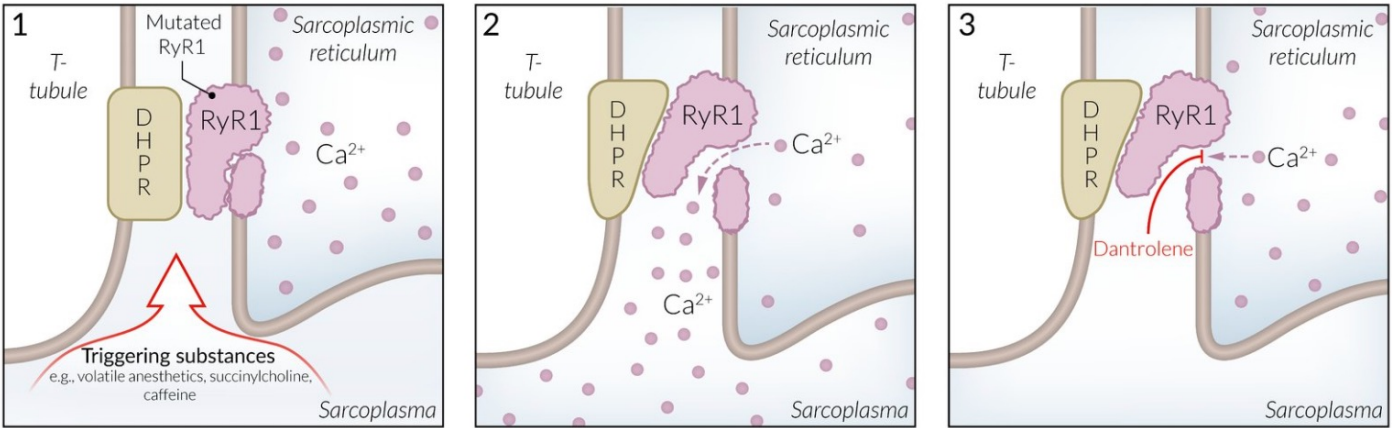


Figure 1: "Pathomechanisms and therapeutic approach in malignant hyperthermia" (Malignant Hyperthermia, 2021, p. 1)