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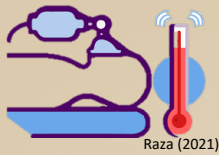
Malignant Hyperthermia

Shannen Steinbrunner
steinbrunner1@otterbein.edu


Raza (2021)

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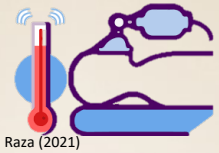
Shannen S. Steinbrunner, BSN, RN, CCRN, SCRN
Otterbein University, Westerville, Ohio


Raza (2021)

Introduction

Malignant Hyperthermia

- Malignant hyperthermia (MH) is a rare, life-threatening series of hypermetabolic reactions (Hopkins et al., 2021)
- Exposure to any trigger can elicit a response: anesthetic gases (halothane, sevoflurane, desflurane, isoflurane), and depolarizing muscle relaxant (succinylcholine) (Larach et al., 2019)
- Genetic predisposition (autosomal dominant) and specific characteristics (vigorous exercise and high temperatures, and several neuromuscular disorders) can increase the risk (van den Bersselaar et al., 2022).

Importance

- The first reported case of MH was in 1960, and the mortality rate was 80-90% (Rosenbaum & Rosenburg, 2022).
- The mortality rate is less than 10%; in cases involving anesthesia, 1:100,000 people may be affected (Rosenbaum & Rosenburg, 2022)
- Those with a genetic predisposition may have a risk of 1:3,000.
- While MH may be inherited, it is possible to carry the mutation without knowing (Cleveland Clinic, 2022).
- A healthy patient who underwent 30 anesthesia procedures can develop MH during the 31st time; it can happen to anyone, anytime (Hopkins et al., 2021).
- All ethnics groups worldwide are affected; the highest incidence occurs in children <19 (50% cases) (Litman et al., 2019).
- Caffeine halothane contracture test (CHCT) is only available in four US locations (Cleveland Clinic, 2022).
- Prompt recognition and treatment can treat MH but multiple organ dysfunction syndrome and death can still occur.

Signs and Symptoms

(Early Signs)

- Tachypnea
- Unexplained increased CO2 or ETCO2
- Acidosis
- Unexplained Tachycardia
- Hyperthermia
- Rigid muscles/spasms ("jaws of steel" – masseter muscle rigidity)

(Cleveland Clinic, 2022)

Signs and Symptoms

(Late Signs)

- Arrhythmias
- Hyperthermia (1-2C q5 mins)
- Mottled skin
- Hyperkalemia
- Rhabdomyolysis
- Dark/amber urine
- Bleeding (DIC)
- Seizures

(Cleveland Clinic, 2022)

Laboratory Values

Increased

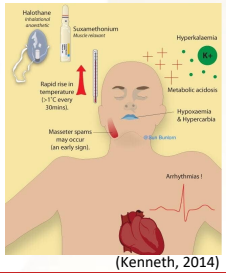
- Calcium and lactate
- Potassium >6 mEq/L
- PaCO2 >60 in controlled ventilation or >65 in spontaneous ventilation
- CK >20,000 units/L after administering succinylcholine or >10,000 units/L without succinylcholine administration
- Urine myoglobin >60 mcg/L
- Serum myoglobin >170mcg/L
- Base deficit >8 mEq/L

(Rosenbaum & Hosenberg, 2022)

Decreased

- Mixed venous O2
- Arterial pH <7.25

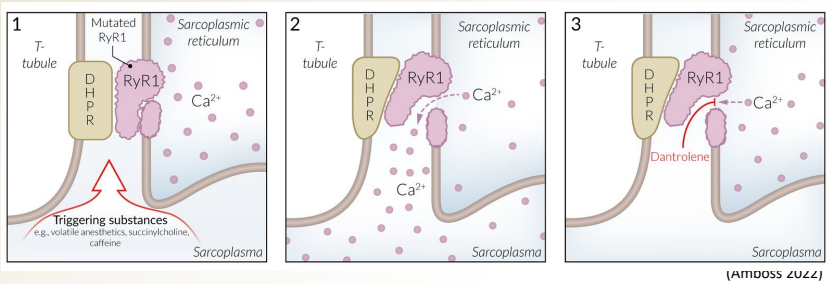
(Rosenbaum & Hosenberg, 2022)



(Kenneth, 2014)

Underlying Pathophysiology

- Those with genetic abnormalities on the skeletal muscle receptor allow a large amount of calcium to enter the cell and cause a muscle contraction. This is because the receptor is defective and unable to close (Cully et al., 2018).
- Normally, a muscle cell experiences depolarization which activates dihydropyridine (DHP) receptors in the t-tubule membrane. The DHP receptors are coupled to ryanodine receptors (RYR1), which control the efflux and influx of calcium within the sarcoplasmic reticulum. Calcium binding to troponin causes the actin and myosin to crosslink, thus causing a muscle contraction (Lawal et al., 2021)
- In MH, the RYR1 receptor is defective and does not close, allowing calcium to rush into the cell, leading to sustained muscle contraction. This consumes oxygen consumption and depletes ATP, thus producing large amounts of carbon dioxide and heat (Cully et al., 2018).
- With little/no ATP, the cell membrane is damaged, which leads to potassium, creatinine kinase, and myoglobin leaking into circulation (Rosenbaum & Rosenburg, 2022).
- Dantrolene is a skeletal muscle relaxer that inhibits calcium to be released from the sarcoplasmic reticulum. Because this acts as an antagonist of the RYR1 receptor, it lessens the depolarization of the muscle cell and, therefore, the contraction (Larach et al., 2019).
- Coding genes such as CACNA1S and STAC3 have also been linked to the cause of MH (Lawal et al., 2020).
- Even if a patient has never had MH prior, several neuromuscular diseases also increase the susceptibility: central core disease, multiminicore disease, and ing-Denborough syndrome (Cleveland Clinic 2022).

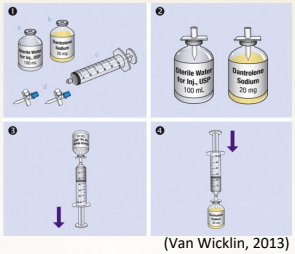


(AMBOSS 2022)

Significance of Pathophysiology

- Nurse anesthetists must rapidly be able to recognize a patient experiencing MH. Severe contracture progresses quickly into respiratory acidosis (Hopkins et al., 2021).
- These patients increase their metabolic function by increasing their O2 consumption, therefore, increasing their CO2 release, eliciting a sympathetic response as HR increases and body temperature rises (Rosenbaum & Rosenburg, 2022).
- The syndrome progresses to metabolic acidosis and, later, rhabdomyolysis as more cells continue to die (Hopkins et al., 2021).
- Rhabdomyolysis results in hyperkalemia and myoglobinuria, leading to fatal arrhythmias (Hopkins et al., 2021).
- Increasing the FiO2 and minute ventilation is also beneficial as the patient is using up their ATP. Oxygen is needed to make ATP; therefore, supplemental O2 or artificial ventilation is vital (JSA, 2017).
- Aggressively cooling the patient with ice packs or cool IV fluids will slow the ATP from being used up, ultimately preventing multiple organ dysfunction syndrome (MODS) (Rosenbaum & Rosenburg, 2022).
- Health care providers should educate the patient and blood relatives about the increased susceptibility and likelihood of MH reoccurring (Cleveland Clinic, 2022).

Implications for Nursing Care


(Van Wicklin, 2013)

- Dantrolene (2.5mg/kg then titrate to tachycardia and hypercarbia). Continue at 1mg/kg every 5 minutes up to 10mg/kg (Larch et al., 2019).
- Once dantrolene has been used, the patient needs to stay in the ICU for a minimum of one day to monitor signs and symptoms (Cleveland Clinic, 2022).
- Stop inhalation agent, succinylcholine or reverse trigger agent (Larach et al., 2019)
- Treat arrhythmias. Avoid Ca blockers as it can interact with dantrolene and cause hyperkalemia (Hopkins et al., 2021)
- Increase minute ventilation to lower ETCO2 (Rosenbaum & Rosenburg, 2022).
- Ice packs/cooling blanket (take aggressive measures) (Hopkins et al., 2021).
- Urine output goal: 2 ml/kg/hour with mannitol, furosemide, and fluids as needed (Rosenbaum & Rosenburg, 2022).
- Obtain coagulation labs every 6-12 hours as DIC can occur with temps above 41C (Rosenbaum & Rosenburg, 2022).
- MH hotline should be contacted for further guidance (1-800-MH-HYPER) (Rosenbaum & Rosenburg, 2022).
- Notify the surgeon and stop the procedure as soon as possible.
- There is no bedside test available; testing available at birth: in vitro contracture test (IVCT) (Litman et al., 2019).
- Genetic testing can also be performed for RYR1, STAC3, and CACNA1S DNA mutations (Litman et al., 2019).

Conclusions

- Malignant hyperthermia is a rare but fatal disorder if not treated promptly.
- MH be easily preventable if clinicians know the factors that exacerbate the reaction.
- Triggers include volatile anesthetics (desflurane, enflurane, isoflurane, etc.) or succinylcholine, as well as high fevers and intense exercise (Carlson et al., 2022).
- Those with a genetic defect of the RYR1 gene are highly susceptible, as well as CACNA1S gene (Lawal et al., 2020).
- If MH does occur, nurse anesthetists must quickly be able to recognize the symptoms and administer dantrolene to minimize the patient's adverse effects.
- Common signs and symptoms include increased heart rate, CO2, rate of breathing, temperature, and muscle spasms (Rosenbaum & Rosenburg, 2022).
- The first and most important step in treating MH once a reaction has begun is to remove the stimuli.
- Dantrolene is the drug of choice if MH is suspected.
- The patient must stay in the ICU for monitoring overnight once dantrolene has been given (Cleveland Clinic, 2022).
- Primary family members of the patient with the genetic variation should be tested in order to prevent its occurrence.
- Children contribute to 50% of the cases reported for MH (Litman et al., 2019).
- A patient may have surgery with the same anesthetic multiple times prior and not have a reaction, then develop MH during the last surgery (Hopkins et al., 2021).

References

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