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Malignant Hyperthermia

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Introduction

Malignant Hyperthermia (MH)
- Pathophysiological processes
- Pathophysiology
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- Significance
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In peri-operative anesthetic practice, complications occur frequently but are rapidly identified and abated by astute clinicians. Rarely, unexpected complications such as MH develop and require emergency intervention to diminish the probability of mortality. Malignant hyperthermia is a potentially lethal hypermetabolic complication of a rare inherited muscle disorder which is caused by inhaled volatile anesthetics and neuromuscular blocking agents. (McCance & Huether, 2010, p. 500).

Understanding the susceptibility, risk factors, pathophysiology, and treatment are foundational for the advanced practice nurse developing his practice, but all clinicians must be knowledgeable about MH so that evidenced-based treatment may be swiftly implemented. Quantifying MH events is challenging, but that the prevalence for MH is 1 in 100,000 which differs from MH susceptibility (MHS) of *between* 1 in 1,200 and 1 in 1,300 which may be explained by sub-clinical manifestations of MH crises. (Belani et al., 2020, p. 553)

Pathophysiological Processes

Pathophysiology

- Inherited autosomal dominant trait, more than 80 genetic defects associated with MH (Spruce, 2020, p. 282)
- Genetic mutation RYR1 encodes ryanodine receptors, CACNA1S encodes dihydropyridine calcium channels in skeletal muscle (Nagelhout & Elisha, 2017, p. 774)
- Reduced magnesium ion affinity responsible for closing calcium channels and triggering agents greatly open RYRI channels leading to uncontrolled, unopposed calcium release from the sarcoplasmic reticulum (Belani et al., 2020, p. 553)
- Sustained muscle contraction consumes adenosine triphosphate depleting stores leading to "protracted muscle rigidity, rhabdomyolysis and cellular membrane breakdown releasing intracellular contents (potassium, CPK, and myoglobin into circulation" (Schneiderbanger, 2014, n.p.)
- Triggering agents include volatile anesthetics (halothane, enflurane, isoflurane, desflurane, sevoflurane) and depolarizing neuromuscular blocking agents, namely succinylcholine (Schneiderbanger, 2014, n.p.)

Signs & Symptoms

Key Diagnostic Features
(Hopkins et al., 2021, p. 658)

- Unexplained, unexpected rise in *end-tidal CO2 (ETCO2)*
- Unexplained, unexpected increase in *heart rate*
- Unexplained, unexpected increase in *temperature*
- Unexplained, unexpected *skeletal muscle rigidity*

Other Diagnostic Features
(Hopkins et al., 2021, p. 660)

- Cardiac arrythmia
- Cardiac arrest
- Acidosis
- Hyperkalemia
- Rhabdomyolysis
- Disseminated intravascular coagulation (DIC)
- Compartment syndrome
- Masseter muscle rigidity

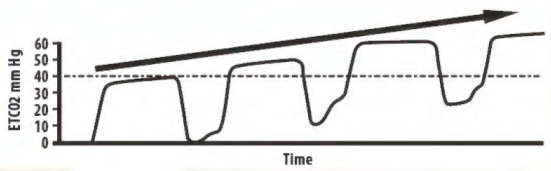

Figure 1: Retrieved from https://www.capnoacademy.com/

Significance of Pathophysiology

Although MH has a relatively low incidence, the MH crisis patient is at a heightened risk for death and all clinicians – especially anesthesia providers – in the perioperative environment must be familiar with the signs, symptoms, and protocols for treatment to reduce mortality.

Pathogenesis: Excitation-Contraction Coupling

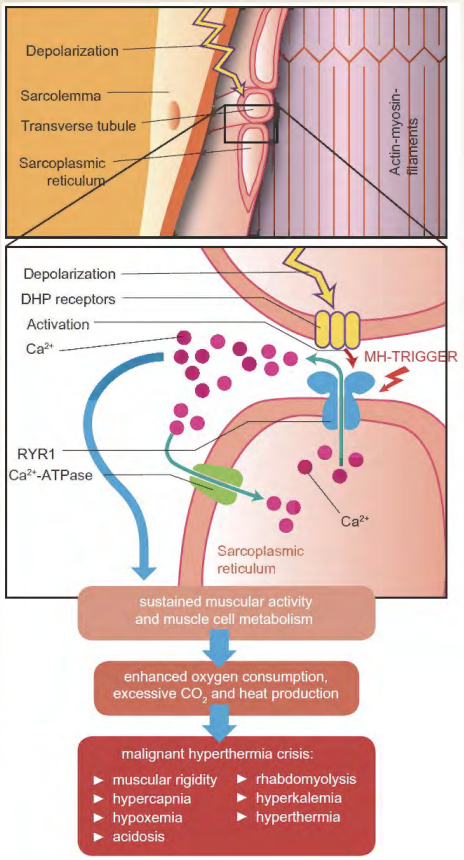

Figure 2: Retrieved from https://www.dovepress.com/cr_data/article_fulltext/s47000/47632/img/fig1.jpg

Risk Factors and Genetics

Risk Factors
(Nagelhout & Elisha, 2017, p. 775)
- Known MH susceptibility
- Providers must use plain language and consider healthcare literacy when obtaining pre-operative
- Family history of unexpected intraoperative complications or death
- Family history of MH or MH-like response to anesthesia

Genetics
- Caffeine Halothane Contracture Test is a muscle biopsy test for establishing MHS (MHS) and is the gold standard currently (Belani et al., 2020)
- Other genetic testing methods such as next-generation sequencing are being investigated to rule out MHS but genetic counseling and CHCT remain best-practice (Belani, et al., 2020)

(MHAUS, 2021)

Nursing Implications

Reversing the MH Process
(Hopkins et al., 2021, p. 659)

- Eliminate the agent (turn off/remove vaporizer
- Administer 100% Oxygen, maximal flow
- Increase minute ventilation 2-3x normal
- Insert activated charcoal filters into inspiratory and expiratory limbs of the circuit
- Administer dantrolene sodium, 2.5 mg/kg as frequently as needed until a decrease in ETCO2, decreased muscle rigidity, and lowering heart rate are appreciated (MHAUS, 2021)
- Initiate active body cooling
- Supportive therapy for other symptoms
- When body temperature rises above 41° C, DIC is the most common cause of death (Gong, 2021, p. 7)
- When dantrolene is unavailable, early warning signs must be identified with prompt, effective therapies (Gong, 2021, p. 7)
- Providers should advocate for dantrolene availability, even when succinylcholine is used without volatile anesthetics as MH crisis may occur as demonstrated by Larach et al. (2019)

MH Resources

Malignant Hyperthermia Association of the United States

24/7 Emergency MH Hotline
1(800)MH-HYPER (644-9737)

https://www.mhaus.org

Conclusion

MH is relatively rare, though potentially fatal genetic condition that requires prompt, effective treatment to reduce mortality. This complexity requires pre-operative identification, thorough history and physical, a team-based treatment approach, and effective communication. Anesthesia providers are integral team members who must be prepared with knowledge about MH and need to be comfortable with the mixing of dantrolene in a crisis.

References and Resources

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