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Malignant Hyperthermia

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Malignant Hyperthermia

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Overview

- ❑ The rare genetic condition of malignant hyperthermia (MH) is widely researched by anesthesia providers.
- ❑ MH is a condition in which the body goes into a hypermetabolic state affecting skeletal muscle in response to receiving inhaled anesthetics or the neuromuscular blocking agent, succinylcholine. Early recognition and treatment is necessary as this condition is fatal if left untreated.
- ❑ The exact incidence of MH is not well understood. MH presents in about 1 in 100,000 adult surgeries ; and 1 in about 30,000 pediatric surgeries.
- ❑ MH research allows for a more thorough understanding of the condition: the causes, clinical presentation, and most importantly how to treat the condition to prevent fatalities.

(Malignant Hyperthermia Association of the United States, 2020; Rosenberg et al., 2015).

Clinical Presentation

- ❑ MH results in a hypermetabolic state causing the following signs and symptoms:
- Hyperthermia
  - temperature rising 1-2 degrees every 5 min with temps greater than 44 degrees Celsius.
- Tachycardia
- Tachypnea
- Muscle rigidity
  - Occurs immediately after administration of anesthetic agent or succinylcholine
- Rhabdomyolysis
  - Dark color urine, marked increase in CPK levels
- Hypercapnia
  - Increase in Etc02 resistant to an increase in minute ventilation
- Hypoxia
  - Follows hypercapnia
- Acidosis
- Hyperkalemia
  - Can lead to dysrhythmias

(DeWel, B., & Claeys, K.G., 2018)

Table 2. Clinical Signs of MH[5,6,8,10]

Early Clinical Signs
• Abrupt increase in $ETCO_2$
• Cardiac arrhythmias
• Generalized muscle rigidity
• Hypoxia
• Profuse sweating
• Trismus / Masseter muscle rigidity (MMR)
• Metabolic-respiratory acidosis
• Mottling of the skin
• Tachycardia
• Tachypnea in spontaneously breathing patients
• Unstable arterial pressure
Late Clinical Signs
• Acute renal failure
• Circulatory failure
• Dark colored urine due to myoglobinuria
• Disseminated intravascular coagulation
• Elevated blood creatine phosphokinase levels
• Elevated blood myoglobin levels
• Hyperkalemia
• Hyperthermia (> 38.8° C)*
• Hypotension
• Rhabdomyolysis
• Severe cardiac arrhythmias and cardia arrest

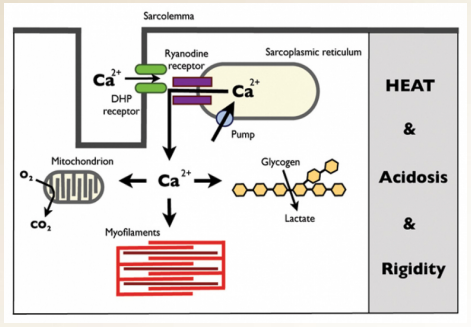
*A rapid temperature increase of >1° C in 15 minutes is more diagnostically relevant than peak temperature.[6]

(AANA, 2020)

Pathophysiology of Malignant Hyperthermia

- ❑ MH is an autosomal dominant disorder that is most closely related to a mutation in the ryanodine receptor type 1 (RyR1) gene.
  - Additional genetic mutations associated with MH include: CACNA1S
- ❑ Following the administration of triggering agents such as inhaled anesthetics or succinylcholine, the defective receptor (RyR1) causes an uncontrolled release of calcium from the sarcoplasmic reticulum.
- ❑ Increased intracellular calcium facilitates excessive excitation-coupling leading to inappropriate muscle contraction and a deadly hypermetabolic state.

(Rosenberg et al., 2015; Stephens et al., 2016)



(Malignant Hyperthermia, 2012)

Diagnosis

- ❑ Based on clinical presentation/ lab testing:
  - Elevated temperature: >38.8 increasing 1 degree every 5 minutes
  - Elevated EtC02 >55mmHg despite increased minute ventilation
  - Muscle rigidity
  - Rhabdomyolysis: elevated creatine kinase, cola-colored urine, elevated myoglobin
  - Blood/plasma/serum K+> 6 mEq/L
- ❑ The "gold standard" for diagnosis of MH is an in vitro contracture test.
  - Based on contracture of muscle fibers in the presence of halothane or caffeine

(Rosenberg et al., 2015)

Malignant Hyperthermia Diagnostic Grading Scale

Table 1 Criteria used in the Clinical Grading Scale for Malignant Hyperthermia

Process	Indicator
I: Rigidity	a. Generalized muscular rigidity (in absence of shivering due to hypothermia, or during or immediately following emergence from inhalational anesthesia)
	b. Masseter spasm shortly following succinylcholine administration
II: Muscle Breakdown	a. Elevated creatine kinase >20,000 IU after anesthetic that included succinylcholine
	b. Elevated creatine kinase >10,000 IU after anesthetic without succinylcholine
	c. Cola colored urine in perioperative period
	d. Myoglobin in urine >60 µg/L
	e. Myoglobin in serum >170 µg/L
	f. Blood/plasma/serum K^+ >6 mEq/L (in absence of renal failure)
III: Respiratory Acidosis	a. PET_{CO2} > 55 mmHg with appropriately controlled ventilation
	b. Arterial PA_{CO2} > 60 mmHg with appropriately controlled ventilation
	c. PET_{CO2} > 60 mmHg with spontaneous ventilation
	d. Arterial PA_{CO2} > 65 mmHg with spontaneous ventilation
	e. Inappropriate hypercarbia (in anesthesiologist's judgment)
	f. Inappropriate tachypnea
IV: Temperature Increase	a. Inappropriately rapid increase in temperature (in anesthesiologist's judgement)
	b. Inappropriately increased temperature > 38.8 °C (101.8 °F) in the perioperative period (in anesthesiologist's judgment)
V: Cardiac Involvement	a. Inappropriate sinus tachycardia
	b. Ventricular tachycardia or ventricular fibrillation

(Rosenberg et al., 2015)

Risk Factors

- ❑ Prior history of MH
- ❑ Musculoskeletal disorder
- ❑ History of rhabdomyolysis
- ❑ Greater incidence in males
- ❑ Highest incidence in young people, with average age of 18.3 years
- ❑ If parent has defect RyR1 gene, 50% chance of passing it to offspring (autosomal dominant)

(Rosenberg et al., 2015; Stephens et al., 2016)

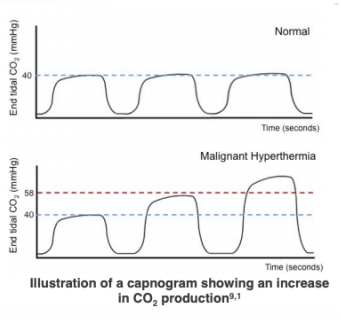


Illustration of a capnogram showing an increase in CO_2 production[9,1]

(Norgine, 2016)

Treatment

- ❑ Immediate discontinuation of trigger agents
- ❑ Dantrolene 2.5mg/kg q 10-15 min till resolution of symptoms
  - Works by directly decreasing calcium in the cell and interferes with muscle contraction
  - Only drug that specifically treats MH
- ❑ Supportive Care
  - Temperature cooling measures
  - Increase minute ventilation
  - Traditional hyperkalemia management

(Malignant Hyperthermia Association of the United States, 2020; Rosenberg et al., 2015)

(Malignant Hyperthermia Overview, 2020)

Nursing Implications

Medical interventions:
- ❑ Administration of dantrolene
- ❑ Management of hyperkalemia such as administration of insulin/ glucose
- ❑ Manage hemodynamics
- ❑ Maintain normothermia

Assessment:
- ❑ Understand and monitor for early signs and symptoms of MH
- ❑ Assess for susceptibilities of MH and past medical history
- ❑ Collect and monitor lab results

(AANA, 2020; Malignant Hyperthermia Association of the United States, 2020; Rosenberg et al., 2015)

Conclusion & Significance

- ❑ With the complexity of this disorder and its unique pathophysiological process it is crucial that all parts of the surgical team, especially anesthesia providers, understand how to treat a patient with MH.
- ❑ Certified registered nurse anesthetists (CRNA) are responsible for monitoring patients very closely. Clinical signs of MH must be recognized and treated early in order to decrease morbidity and mortality.
- ❑ Increased understanding of the clinical manifestation and pathophysiology of MH has led to a decrease in mortality from 80% thirty years ago to <5% in 2006.

(Rosenberg et al., 2015).

References and Additional Sources

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