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Malignant Hyperthermia

Brandon Kinnamon
brandon.kinnamon@otterbein.edu

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Brandon Kinnamon, BSN, RN, CCRN
Otterbein University, Westerville, Ohio

Figure 1. Retrieved from https://www.thinglink.com/scene/705211532971081729 on July 17, 2017.

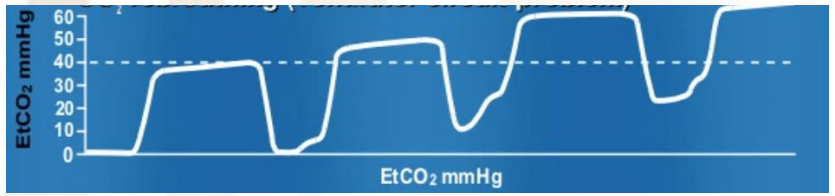
Introduction

- The pathophysiological concept selected for analysis is malignant hyperthermia (MH).
- Relevant in both the intensive care unit and the operating room while intubating patients or inducing anesthesia.
- Anesthetic inhalation agents like halothane and the neuromuscular blocking agent succinylcholine can trigger a pathological response involving intracellular calcium resulting in hyper-metabolism (Schneiderbanger, Johannsen, Roewer & Schuster, 2014, p. 356-357).
- It is vital for anesthesia professionals and critical care nurses to recognize MH and act quickly to limit rapid shifts of electrolytes (Donnelly, 1994, p. 395).

Underlying Pathophysiology

- Malignant hyperthermia is an autosomal dominant trait with a mutation of the ryanodine receptor (RYR1) (Hirshey Dirksen, et al., 2011, p. 1108).
- When exposed to an environmental (heat stroke) or pharmacological trigger, the mutation on RYR1 causes an uncontrolled release of calcium from the sarcoplasmic reticulum in skeletal muscles (Rosenberg, Pollock, Schiemann, Bulger & Stowell, 2015, p. 4).
- The only other gene showing potential MH-causing mutation is CACNA1S (Rosengerg, et al., 2015, p. 4).
- The depolarizing muscle relaxant succinylcholine and volatile anesthetics including halothane, isoflurane, enflurane, sevoflurane, methoxyflurane, and desflurane can all trigger MH (Schneiderbanger, et al., 2014, p. 356-357).
- The uncontrolled release of calcium in the muscle leads to sustained contraction causing a rise in exhaled carbon dioxide, the observable muscle rigidity, and generates heat which leads to a dramatic rise in body temperature (Seifert, Wahr, Pace, Cochrane & Bagnola, 2014, p. 189).
- When muscles become rigid they can no longer produce adenosine triphosphate (ATP) which leads to cellular damage and leakage of potassium, creatine kinase, and myoglobin from the muscle cells (Brandom & Callahan, 2015, p. 115).
- This leakage results in metabolic acidosis, cardiac arrhythmias, and myoglobinuria (Brandom & Callahan, 2015, p. 115).
- To compensate for the increased body temperature, hyperventilation, vasodilation, and catecholamine release all occur which lead to increased end-tidal carbon dioxide levels, flushing, and tachycardia followed by cyanosis of the periphery (Brandom & Callahan, 2015, p. 115).
- If not halted this hypermetabolic state can lead to cardiac arrest, hemorrhaging, and brain damage (Ali, Taguchi & Rosenberg, 2003).

Figure 2.
Retrieved from http://www.slideshare.net/larryide/capnography?next_slideshow=1 on July 17, 2017.



Signs and Symptoms of MH

- Hallmark and earliest sign of malignant hyperthermia is hypercapnia which can be detected as an increase in end-tidal carbon dioxide or EtCO2 (Lin et al., 2014, p. 209).
- Other major signs of MH include muscle rigidity, tachycardia, tachypnea, pyrexia, flushing followed by peripheral mottling, cardiac arrhythmias, hypotension, rhabdomyolysis, and even disseminated intravascular coagulation (Brandom & Callahan, 2015, p.119).
- Both metabolic and respiratory acidosis can develop simultaneously resulting in a profound acidotic state (Redmond, 2001, p. 261).
- Symptoms can result in what appears to be a comatose-state for the patient with absent reflexes, fixed pupils, apnea, and unconsciousness (Huether, McCance, Brashers & Rote, 2008, p. 314).

-Hypercapnia -Acidosis
-Rigidity -Hyperthermia

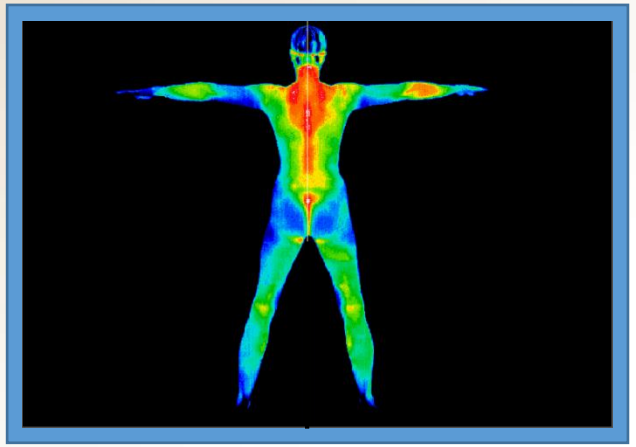


Figure 3. Retrieved from http://www.applewoodacupuncture.com/infrared-thermal-imaging/ on July 17, 2017.

Significance of Pathophysiology

- Malignant hyperthermia is life-threatening and is associated with administration of anesthetic agents and neuromuscular blocking agents.
- It can occur rapidly, or slightly delayed, in any facility that administers the potential triggering agents. This means that reactions can occur in the operating room (OR), post anesthesia care unit (PACU), or the intensive care unit (ICU) (Seifert, et al., 2014, p. 190).
- Without proper recognition, the affected patient can suffer dire consequences. Cardiac arrest, disseminated intravascular coagulation, kidney failure, liver failure, brain damage, and even death are potential outcomes if treatment is not initiated and supportive care provided (Seifert, et al.,2014, p. 189).
- This makes the pathophysiology of MH extremely significant and it is vital that the anesthesia provider, PACU nurse or ICU nurse recognize the condition so it can be treated.

Implications

- Careful monitoring of end-tidal carbon dioxide and core body temperature should be conducted during surgical procedures involving possible triggering agents (Lewis, Dirksen, Heitkemper, Bucher & Camera, 2011, p. 363).
- The failure to monitor capnography and core temperature puts patients experiencing MH at an increased risk of death (Brandom & Callahan, 2015, p. 119).
- Once a patient develops MH the stoppage of all triggering agents and immediate administration of the skeletal muscle relaxant dantrolene must begin (Lilley, Harrington & Snyder, 2007, p. 190).
- Dantrolene blocks the release of calcium from the skeletal muscle cells and halts the sustained contraction causing the hypermetabolic state (Brandom & Callahan, 2015, p. 120).
- Beta blockers and oxygen are also needed to treat the tachycardia and hypoxia associated with MH (Lilley et al., 2007, p. 171).
- Cooling the patient and correcting hyperkalemia and acidosis are also frequently necessary (Myers, 2009, p. 1135-1136).
- Supportive care and dantrolene administration will continue from the operating room (OR) to the intensive care unit (ICU) for at least forty-eight hours of monitoring and continued care (Ali, et al., 2003, p. 526).
- Screening patients preoperatively can prevent MH episodes before they happen.
- A thorough individual (including muscular pathologies) and family health history can identify patients at risk for MH, since it can be an inherited condition (Schneiderbanger, et al., 2014, p. 360).
- Muscular disorders including Central Core Disease, Myotonic Muscular Dystrophy, and King-Denborough syndrome are all conditions predisposed to developing MH (Rosenberg, et al., 2015, p. 3-4).
- Testing can be performed to determine susceptibility of MH in at-risk individuals with the caffeine halothane contracture test (CHCT) (Hirshey Dirksen, et al., 2011, p. 1112). During the test caffeine and halothane are applied directly to muscle bundles and will cause contracture at lower concentrations in MH susceptible patients (Hopkins, 2017, p. 323).
- Families with the RYR1 mutation are referred to the Malignant Hyperthermia Registry also known as MHAUS for entry into the national database (Myers, 2009, p. 1136).

Conclusions

- Malignant hyperthermia is a rare and life-threatening condition which is elicited by anesthetic agents used during intubation and general anesthesia (Cain, Riess, Gettrust & Novalija, 2014, p. 301).
- It is vital that nurses and anesthesia providers know what to monitor and what actions to take if a patient develops MH.
- Simulations can assist in preparing personnel for a potential MH crisis in a safe environment, and this practice has been used regularly by anesthesia professionals since 1994 (Cain et al., 2014, p. 302).
- MHAUS has organized mock drills with the funding help of professional anesthesia associations and is a valuable data bank for families and anesthesia providers on malignant hyperthermia (Brandom & Callahan, 2015, p. 124).
- Future research on MH will likely focus on genetics and the phenotypic variability of RYR1-related conditions (Hirshey Dirksen, 2011, p. 1110).
- Accurately predicting which patients are susceptible to MH will allow providers to avoid possible triggering agents while still providing excellent anesthetic care to those patients.

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