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Malignant Hyperthermia

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Introduction

According to the Journal of Anesthesia and Intensive Care, Malignant Hyperthermia (MH) is an uncommon disorder of skeletal muscle, with autosomal dominant inheritance triggered almost exclusively by potent inhalational agents and suxamethonium (Chan, Bulger, Stowell, Gillies, Langton, Street, and Pollock, 2017). Although it is rare, it is a true medical emergency which can lead to death if left untreated. Mostly anesthesia medications trigger the response so it is imperative the anesthesia providers along with the postoperative nurses know the signs and symptoms that trigger malignant hyperthermia. It is a hypermetabolic state triggered by an abnormality of calcium release or reuptake by the sarcoplasmic reticulum with muscle contraction.

The incidence of MH in adults is one in every 50,000 to 100,000 and children is one in every 300,000 to 500,000 cases (Chan et al., 2017). Since MH occurs in genetically predisposed individuals, one must go through an invasive muscle biopsy procedure to get diagnosed. The test is very expensive and has a high false positive result. This topic was chosen due to my future interest as a practicing nurse anesthetist. Researching MH will prepare me and make me more aware as an anesthesia provider.

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Pathophysiological Process

Sign & Symptoms

Medical personnel should be aware and recognize the signs and symptoms of MH. The cardinal sign of MH is hyperthermia, which is relatively a late sign. MH can occur very suddenly with muscle rigidity or gradually and not show the signs till in the post-operative unit.

Signs and Symptoms include:
-Tachyarrhythmia's in 96% of cases
-Tachypnea in 85% due to rapidly rising carbon dioxide level
-Acidosis in 80% due to lactic acid and heat
-Oxygen saturation levels drop during normal ventilation
-Generalized muscle rigidity in 80% is one of the earliest signs (Hommertzheim & Steinke, 2006)

Therefore many complications can occur due to the profound impact on the body. Severe complications such as metabolic acidosis, bowel ischemia, compartment syndrome of the limbs, vital organ dysfunction, acute renal failure, and disseminated intravascular coagulation. Other less severe symptoms include muscle weakness, lightheadedness, and difficulty swallowing (Hommertzheim & Steinke, 2006).

Underlying Pathophysiology

MH is a hypermetabolic state triggered by an abnormality of calcium release or reuptake by the sarcoplasmic reticulum with severe muscle contraction. In genetically predisposed individuals the triggering event resides in the skeletal muscle at the level of calcium when it transfers into the muscle cell. Therefore the influx of calcium into the cell leads to hypermetabolism which increases carbon dioxide production, increases oxygen consumption, and increases sympathetic activity (Hommertzheim & Steinke, 2006).

-The most common mutation is found in the RYR1 gene on chromosome 19 that regulates the calcium pathway for the sarcoplasmic reticulum. The distribution of the mutation is varied among ethnic groups (McGoldrick, 2017).

-The increase or surge of intracellular hypercalcemia activates a cascade of metabolic pathways that deplete ATP, cause acidosis, membrane destruction, and cell death (McGoldrick, 2017).

-Testing can be performed to determine susceptibility of MH with a caffeine halothane contracture test (CHCT). The CHCT test requires invasive muscle biopsy from the thigh. Only six centers in the United States are approved for testing. A patient must travel to a center in order to get tested which is generally not covered by insurance at $6,000 (Hommertzheim & Steinke, 2006).

Significance of Pathophysiology

-Knowing the pathophysiology of MH allows individuals to recognize the signs and symptoms of MH faster. Also knowing the mechanism of action allows for treatment. Two known triggering agents are succinylcholine and halogenated volatile anesthetics (Hommertzheim & Steinke, 2006).

- Other triggers include butyrophenones, phenothiazines, various atypical anti-psychotics, anti-dopaminergic drugs, and abrupt withdrawal of anti-Parkinson's drugs (McGoldrick, 2017).

-Stop the trigger agent immediately and active the MH team to get help to quickly stop the crisis to prevent it from progressing to life-threatening conditions.

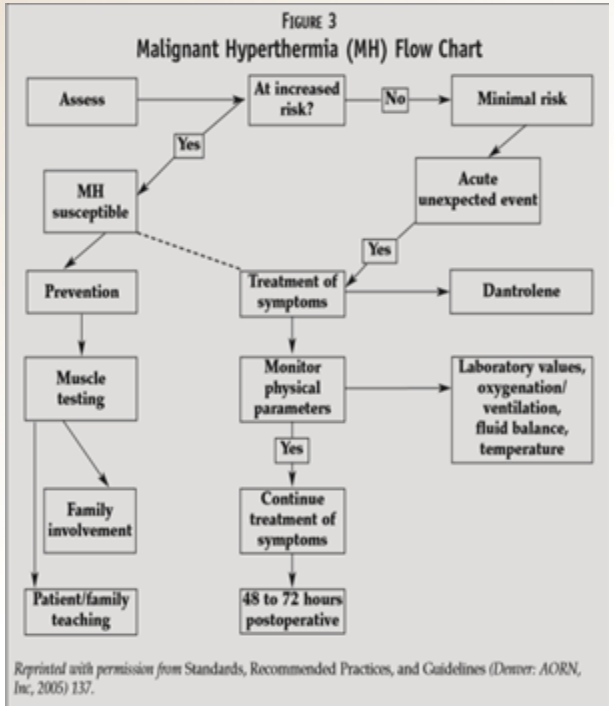

FIGURE 3
Malignant Hyperthermia (MH) Flow Chart

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Implications for Nursing Care

The anesthesia staff along with the registered nurses working in the postoperative unit must know the response plan for an MH event. Chances for a successful outcome will increase with a rapid, accurate diagnosis and a coordinated, swift, multidisciplinary team to deliver the appropriate treatment (Dirksen, Van Wicklin, Mashman, Neiderer, Merritt, 2013). Therefore, proper education and training is imperative to all the necessary team members based on the organizations environment. Practice through simulation and mock drills are essential to ensure the staffs roles are defined during a MH crisis. Many facilities have a designated emergency cart solely responsible for a MH crisis. Staff must know where the carts are located and know how to use the equipment within the carts. Perioperative nurses also screen patients prior to anesthesia and maybe able to uncover red flags such as a prior occurrence or known family member who has experienced MH.

-If and MH event occurs the procedure should be stopped immediately unless emergent. If the procedure must continue the anesthesia team is responsible for discontinuing all triggering anesthetics and begin the treatment for MH. Treatment begins by hyperventilating the patient with 100% oxygen and administering a dose of dantrolene sodium. Dantrolene is a skeletal muscle relaxant that depresses muscle contraction by decreasing intracellular calcium concentration (National Institute s of Health, 2018). Electrolyte replacement and monitoring will have to be corrected. The patient would most likely be transferred to the intensive care unit for monitoring for the next day or two for observation.

Conclusion

-MH is rare genetic ailment that is a life-threating condition when provoked by the administration of anesthetic agents.

-The earliest signs of MH are generally non-specific, which can make MH very difficult to diagnose by even experienced medical professionals (Issak & Stiegler, 2016).

-Simulation and mock drills are necessary to all anesthesia providers and nurses know how to monitor and treat a patient if MH develops.

-Testing is expensive and not convenient for a majority of the population.

-Before the introduction of dantrolene, the mortality of MH was 84%. Now with proper monitoring and education the mortality has been reduced to 9% (Chan et. al, 2017).

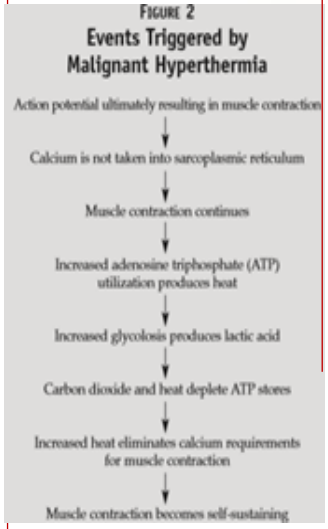

FIGURE 2
Events Triggered by Malignant Hyperthermia

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