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Malignant Hyperthermia

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Introduction

Providing patient care in the operating room as a nurse anesthetist is a high-stress and constantly changing environment. The recognition of malignant hyperthermia (MH) is of paramount level of responsibility that must be addressed to ensure safe care. MH is an inherited autosomal dominant disease of skeletal muscle (Plaus, 2014). Recognition of MH is of extreme importance to ensure the patient and the future gen!erations are not affected. MH is currently diagnosed by muscle susceptibility testing (MST). These tests are performed on muscle biopsies obtained from individuals who are at risk of MH. A positive test result is defined in the absence of a triggering agent by hypermetabolism with muscular rigidity and rhabdomyolysis (Mitchell et al., 2012).

Pathophysiology

MH is a rare, autosomal dominant disease of skeletal muscle. MH arises from a genetic defect in the ryanodine receptor type 1 (RYR1) gene (Nagelhout, 2014; Mitchell et al., 2014). The RYR1 gene codes for the protein that controls calcium release from the sarcoplasmic reticulum (SRT) in muscle cells. Calcium plays a critical role in muscle contraction. In MH susceptible patients, volatile anesthetics (sevoflurane, isoflurane) or depolarizing muscle relaxant (succinylcholine) administration leads to increased intracellular calcium release, which results in muscle rigidity and rhabdomyolysis (Plaus, 2014).

Signs and Symptoms

Rigidity is the characteristic finding of MH. Other symptoms of MH crisis include tachycardia, tachypnea, fever, muscle rigidity, and seizures. In severe cases, lactic acidosis, metabolic acidosis, hyperkalemia, and hyperphosphatemia may develop (Plaus, 2014). The RYR1 gene codes for the protein that controls calcium release from the sarcoplasmic reticulum (SRT) in muscle cells. Calcium plays a critical role in muscle contraction. In MH susceptible patients, volatile anesthetics (sevoflurane, isoflurane) or depolarizing muscle relaxant (succinylcholine) administration leads to increased intracellular calcium release, which results in muscle rigidity and rhabdomyolysis (Plaus, 2014).

Incidence of MH varies between 1/50,000 to 1/250,000 and highest in young population with average age of 18-3 years upon exposure to volatile anesthetics or muscle relaxants and in association with 23 genetic mutations (Nagelhout, 2014). In contrast, according to Malignant Hyperthermia Association of the United States (MHAUS, 2015) MH arises in 1/100,000 in adults and 1/30,000 in young population with average age of 18-3 years upon exposure to volatile anesthetics or muscle relaxants and in association with 23 genetic mutations (Nagelhout, 2014).

MH is rare but potentially fatal skeletal muscle disorder which requires prompt intervention to decrease mortality rate. In the initial years of MH discovery, mortality rate due to MH was 50%. In recent years, this number has changed to approximately 5%, and it is contributed to increased education and awareness among healthcare providers, early recognition and management, and cultivated registry to stop MH crisis (Mitchell et al., 2012). Dantrolene inhibits calcium release by inhibiting RYR receptor at post-synaptic site (Gray & Good, 2002). Dantrolene can also be used to treat MH crisis complicated with other disorders, variable penetrance and absence of phenotypical characteristics in the absence of triggering agent. MH diagnosis is a clinical diagnosis. However, another study states that only 40% of MH suscep!tible patients will re!act to the presence of triggering agent (Mitchell et al., 2012).

Significance of Pathophysiology

Early recognition and prompt administration of dantrolene make it difficult to timely identify MH crisis and implement timely intervention. Understanding pathophysiology of MH could help to lead early recognition and intervention and better outcomes for patients experiencing MH crisis.

Implications for Nursing Care

At first suspicion of MH, discontinuation of anesthetic agent, call MH hotline immediately.

Conclusion

MH is a rare, autosomal dominant disorder and medical emergency that requires immediate recognition and treatment to avoid patient’s experience of death. MH has variable penetrance and is typically triggered by volatile anesthetics and/or depolarizing muscle relaxant (succinylcholine). CRNAs must be prompt and trained to provide “hands-on” care to minimize the risk of MH crisis. In MH susceptible patients, volatile anesthetics (sevoflurane, isoflurane) or depolarizing muscle relaxant (succinylcholine) administration makes it difficult to timely identify MH crisis and implement timely intervention. Understanding pathophysiology of MH could help to lead early recognition and intervention and better outcomes for patients experiencing MH crisis.