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Malignant Hyperthermia
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Introduction
Working in the medical and nursing intensive care units and the future practicing nurse as an anesthesia provider has sparked a spirit of inquiry in me regarding malignant hyperthermia. By choosing this research topic, I will begin to develop an understanding of this potentially deadly disease and learn from the clinical presentations and treatments.

Malignant hyperthermia is an inherited autosomal-dominant disease that occurs in individuals shortly after the induction of anesthesia who encounter inhalation anesthetics or the neuromuscular blocking agent, succinylcholine. Malignant hyperthermia is an emergency situation, and if it is not treated promptly and correctly, the outcome will be detrimental to the patient (Cain, Gettrup, & Novella, 2014).

Pathophysiology
MH is an autosomal-dominant disease that results when a disturbance occurs in the sarcoplasmic reticulum’s calcium release channels, which is characterized by the denervation of acetylcholine (nAChR) (Cain et al., 2014). MH is a complex inherited disease with a genetic predisposition (Dirksen et al., 2014). When functioning as intended, these genes are responsible for making proteins specifically for muscle movement. These proteins guide the sarcoplasmic reticulum’s Ca2+ release channel in a coordinated technique with the end result being muscle contraction and/or relaxation. When there are abnormalities in these genes, the channels are dysfunctional and lead to calcium channels that open too easily and close too slowly. This defect allows for a high influx of calcium into muscle cells which causes abnormal muscle contractions or rigidity. The increased calcium concentration results in the processes which generate heat (leading to hyperthermia) and production of excess acid (leading to acidosis) and ultimately a hypertensive state (Riess et al., 2014).

MH is an emergent condition which initially manifests itself as a severe increase in CO2 production (hyperpyrexia) with abrupt hyperventilation and increased end tidal CO2 (Kraeva et al., 2013). Also occurring is tachycardia, ventricular arrhythmias, and myoglobinuria. More signs and symptoms of MH include skin that is flush, generalized muscle rigidity, hypotension, respiratory acidosis, and coagulopathy. Despite the name’s implication, a quick increase in temperature (> 38.8°C) is typically a late sign. As the MH crisis continues, there is diminished skeletal muscle function which results in rhabdomyolysis. Myoglobinuria, hyperkalemia, and an increased creatinine phosphokinase all lead to an acute renal failure. If left untreated, the end stage of MH is displayed by circulatory collapse, multi-organ failure, and death (Schneiderbauer, Johnson, Rosee, & Schuster, 2014).

Figure 2: "Untouched myoplasmic Ca2+ release is the key to malignant hyperthermia. The most prominent cytosolic Ca2+ elevation results from the freeing of stored sarcoplasmic Ca2+ mediated by ryanodine receptor type 1 (RyR1). While volatile anesthetics stimulate Ca2+ release via RyR1, succinylcholine acts indirectly by activating the nicotinic acetylcholine receptor (nAChR), a non-sarcoplasmic cation channel, resulting in continuous local depolarization. The depolarization can trigger propagated action potentials which further activate the ryanodine receptor (RyR1) leading to the gating of both Ca2+ release from the SR via RyR1 and Ca2+ current from the extracellular space." Graph with text sourced from http://www.apsf.org/newsletters/html/2015/June/01_MHIQ.htm

Signs & Symptoms
MH is an emergent condition which initially manifests itself as a severe increase in CO2 production (hyperpyrexia) with abrupt hyperventilation and increased end tidal CO2 (Kraeva et al., 2013). Also occurring is tachycardia, ventricular arrhythmias, and myoglobinuria. More signs and symptoms of MH include skin that is flush, generalized muscle rigidity, hypotension, respiratory acidosis, and coagulopathy. Despite the name’s implication, a quick increase in temperature (> 38.8°C) is typically a late sign. As the MH crisis continues, there is diminished skeletal muscle function which results in rhabdomyolysis. Myoglobinuria, hyperkalemia, and an increased creatinine phosphokinase all lead to an acute renal failure. If left untreated, the end stage of MH is displayed by circulatory collapse, multi-organ failure, and death (Schneiderbauer, Johnson, Rosee, & Schuster, 2014).

Conclusion
Malignant hyperthermia is a rare disease but can take place in a variety of settings. If not treated in a timely manner, the consequences will be dire. It is recommended that nurses and anesthesiologists be prepared personally be employed on MH crisis. By detecting the signs and symptoms associated with the disease, providers can efficiently remedy the crisis and save lives (Sefert, 2014). Since the discovery of dantrolene in 1975 and the advancement of genetics regarding MH, death has dropped from about 80% to about 5% (Schneiderbauer et al., 2014). Today there is a list group calling for the MH Association for the United States which offers education and support for victims of MH. There is also a 24-hour hotline for people to call during an MH emergency (Dirksen et al., 2013).

References Cited

Implications for Nursing Care
It is of paramount importance for nurses to understand and be familiar with the treatment for MH. Anesthesia providers must maintain an accurate preoperative assessment, ensuring a precise temperature of the patient before anesthesia induction so that the anesthesia provider recognize the abrupt increase in temperature commonly found in MH. This could lead to prompt treatment of the disease prior to the onset of the crisis (Cain et al., 2014). When a crisis is recognized, inhospital anesthetics must be maintained as well as any other MH triggering drugs. The surgery or procedure should be canceled and the patient monitored every closely. The primary treatment for MH is dantrolene sodium – a medication that is reconstituted with sterile water and pushed quickly through a large bore IV catheter. Because of the unique dose of this medication and time-consuming demand for reconstitution, accurate and swift administration can prove as difficult (Sefert, Wahr, Pace, Cochran, & Bagnola, 2014). In addition to dantrolene administration, providers must also prepare cooling blankets and rehydrated crystalloid IV solution to counter the attack on the fatal hyperthermia involved with the crisis. The anesthesia provider should deliver 100% FIO2 to avoid hypoxia and ventilate patients on lactic acid accumulation, and ischemia (Dirksen et al., 2013). Sodium bicarbonate should be considered to treat acidosis. If cardiac arrest occurs, regular ALS should occur. With confidence, education, and familiarity of an MH crisis, nurses can effectively treat the disease which will not only save patients lives, but will also improve patient satisfaction and decrease patient length of stay in the hospital (Schneiderbauer et al., 2014).

Figure 3: "Dantrolene sodium medication label. "Dantrolene dextrorotatory is a medication labeled for intravenous use at a minimum dose of 1 mg/kg. In case of malignant hyperthermia, dantrolene sodium will continue, administer additional intravenous boluses up to the maximum cumulative dosage of 10 mg/kg. If the symptoms reappear, repeat dantrolene dosing by intravenous push starting with 1 mg/kg." Graph with text sourced from https://www.drgas.com/pr/ryanodine.html

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