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Malignant Hyperthermia

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Introduction

Working in the medical and neuro intensive care unit and my future nursing profession 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 fatal disease process and learn its clinical presentations and treatments.

- Malignant hyperthermia is an inherited autosomal-dominant disease trait which occurs in individuals shortly after the induction of anesthesia who encounter inhaled general anesthetics or the neuromuscular blocking agent, succinylcholine.
- Malignant hyperthermia is an emergent situation, and if it is not treated promptly and correctly, the outcome will be detrimental to the patient (Cain, Riess, Gettrust, & Novalija, 2014).

Pathophysiology

MH is an autosomal-dominant disease trait with incomplete penetrance and is associated with gene mutations of the ryanodine receptor 1 (RYR1) and calcium voltage-gated channel subunit alpha1 S (CACNA1S). The RYR1 functions to encode the skeletal muscle isoform of the calcium-release channel of the sarcoplasmic reticulum. CACNA1S functions to encode the alpha subunit of the L-type calcium channel isoform of the sarcolemma (Schneiderbanger et al., 2014). When functioning as intended, these genes are responsible to make proteins specifically for muscle movement. These proteins guide calcium in and out of the RYR1 ion channel in a coordinated technique with the end result being muscle contraction and/or relaxation. When there is a mutation 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 aberrant muscle contractions or rigidity. The increase in calcium concentration results in the processes which generate heat (leading to hyperthermia) and production of excess acid (leading to acidosis) and ultimately a hypermetabolic state (Riazi et al., 2014).

Signs & Symptoms

MH is an emergent condition which initially manifests itself as a severe increase in CO2 production (hypercapnia) with abrupt hyperventilation and increased end tidal CO2. Also occurring is tachycardia, ventricular arrhythmias, and masseter spams. More signs and symptoms of MH include skin that is flush, generalized body rigidity, hypoxia, 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 detrimental skeletal muscle breakdown which results in rhabdomyolysis. Myoglobinemia, hyperkalemia, and an increased creatine 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 (Schneiderbanger, Johannsen, Roewer, & Schuster, 2014).

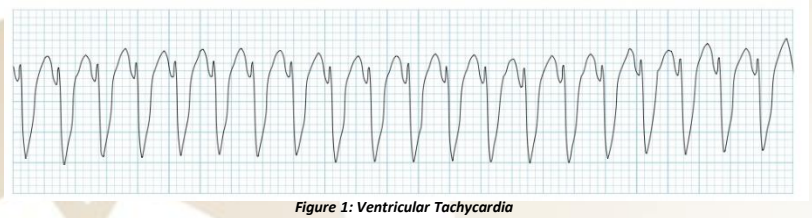

Figure 1: Ventricular Tachycardia
Retrieved July 12, 2016, from http://lifeinthefastlane.com/ecg-library/ventricular-tachycardia/

Significance of Pathophysiology

It is critical to understand the pathophysiological significance of malignant hyperthermia because of the following:

- If left untreated, the outcome is almost certain death (Cain et al., 2014).
- The cascade of consequences involved with MH are an increase of potassium (hyperkalemia), hyperthermia, and the release of myoglobin into the blood.
- Myoglobinemia is catastrophic in that it disrupts the renal tubules which results in renal failure.
- Hyperkalemia causes cardiac arrhythmias such as ventricular tachycardia or fibrillation which can lead to death.
- Hyperthermia is due to the hypermetabolic state and unsustaining APT expenditure (Schneiderbanger et al., 2014).
- Respiratory and metabolic acidosis
- Blood coagulopathies (DIC) and internal hemorrhage
- Pulmonary edema
- Bowel ischemia
- multi-organ failure (Dirksen, Van Wicklin, Mashman, Neiderer, & Merritt, 2013).

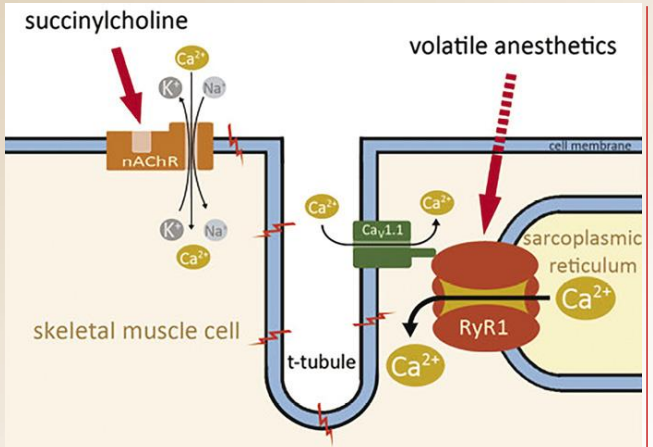

Figure 2: "Uncontrolled myoplasmic Ca2+ release is the key to malignant hyperthermia. The most prominent cytosolic Ca2+ elevation results from the freeing of stored sarcoplasmic Ca2+ mediated by ryanodine receptor type 1 (RyR1). While volatile anesthetics stimulate Ca2+ release via RyR1, succinylcholine acts indirectly by activating the nicotinergic acetylcholine receptor (nAChR), a nonspecific cation channel, resulting in continuous local depolarisation. The depolarization can trigger propagated action potentials and will further activate the dihydropyridine receptors (DHPR, CaV1.1) leading to the gating of both Ca2+ release from the SR via RyR1 and L-type Ca2+ current from the extracellular space." Graphic with text sourced from http://www.apsf.org/newsletters/html/2015/June/01_MHIQ.htm

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. Gathering a precise temperature of the patient before anesthesia induction could help the anesthesia provider recognize the abrupt increase in temperature commonly found in MH. This could lead to prompt treatment of the disease process (Cain et al., 2014). When a crisis is recognized, inhaled anesthetics must cease as well as any other MH triggering drugs. The surgery or procedure should be discontinued and the patient monitored very 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 dosing of this medication and time-consuming demand for reconstitution, accurate and swift administration can pose as difficult (Seifert, Wahr, Pace, Cochrane, & Bagnola, 2014). In addition to dantrolene administration, nurses must also prepare cooling blankets and refrigerated crystalloid IV solution to counter attack the lethalhyperthermia involved with the crisis. The anesthesia provider should deliver 100% FIO2 to avoid hypoxia and subsequent anaerobic metabolism, lactic acid accumulation, and ischemia (Dirksen et al., 2013). Sodium bicarbonate should be considered to treat acidosis. If cardiac arrest should occur, regular ACLS should commence. With education, experience, and familiarity of an MH crisis, nurses can effectively treat the disease which will not only save patient lives, but will also improve patient satisfaction and decrease patient length of stay in the hospital (Schneiderbanger et al., 2014).

Conclusion

Malignant hyperthermia is a rare disease trait and 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 other healthcare personnel be properly educated on MH crises. By detecting the signs and symptoms associated with the disease, providers can efficiently remedy the crisis and save patient lives (Seifert, 2014). Since the discovery of dantrolene in 1975 and the advancement of genetics regarding MH, death rates dropped from about 80% to about 5% (Schneiderbanger et al., 2014). Today there is a MH group called the Malignant Hyperthermia Association of 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).

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Figure 3: Dantrolene sodium medication label. "Administer dantrolene by intravenous push at a minimum dose of 1 mg/kg. If manifestations of MH 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." Graphic with text sourced from https://www.drugs.com/pro/ryanodex.html