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Malignant Hyperthermia

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Introduction

Malignant hyperthermia is an abnormal systemic response to volatile anesthetics and a certain depolarizing neuromuscular blocking drug that causes an uncontrollable release of intracellular calcium ions from the sarcoplasmic reticulum. This reaction subsequently causes sustained muscle contraction, over-utilization of the body's energy stores, and an increase in lactic acid production from anaerobic hypermetabolism, all of which can cause rapid death if not promptly treated. Due to the extreme rarity of malignant hyperthermia susceptibility and the even rarer occurrence of the manifested state, it is a pathophysiologic condition that many in the medical field are unfamiliar with. Unfamiliarity breeds discomfort. Discomfort in the healthcare setting can lead to ineffective and erroneous patient care. Therefore, the topic of this pathophysiology poster is malignant hyperthermia and its purpose is to further aid in spreading awareness of malignant hyperthermia and educate colleagues in the recognition and treatment of malignant hyperthermia. It is of extreme importance that perioperative nurses, especially nurse anesthetists, are comfortable with recognizing and treating malignant hyperthermia if it were ever to occur in the healthcare setting.

Pathophysiological Processes

Pathophysiology:

- Predisposition to MH is based on genetics, specifically an autosomal dominant disorder involving the mutation of the type 1: RyR1 (ryanodine receptor) or dihydropyridine receptor (Mullins, 2018)
- The mutation causes hypermetabolism and an unabated and abnormal release of calcium ions from the sarcoplasmic reticulum inside muscle cells when exposed to MH-triggering agents (Mullins, 2018)
- Atypical release of intracellular calcium causes an increased and sustained actin-myosin interaction (Riazi & Brandom, 2015)
- Mutated receptor prevents the sarcolemmal sodium-calcium pump from restoring homeostasis by reincorporating calcium via aerobic metabolism (Kim et al., 2019)
- MH-triggering agents include volatile anesthetic gases (desflurane, isoflurane, sevoflurane, and halothane) and depolarizing muscle relaxants (succinylcholine) (Mullins, 2018)
- MH can be triggered by heat and intense exercise (Mullins, 2018)

Signs & Symptoms:

- Muscle rigidity
- Increased core temperature
- Increased production of carbon dioxide
- Increased rate of oxygen consumption
- Unexplained tachycardia

(Kim et al., 2019)

Late Signs:

- Hyperkalemia
- Acidosis
- Myoglobinuria
- Increased creatine kinase
- Electrolyte imbalances
- Congestive heart failure
- Pulmonary edema
- Congestive heart failure
- Alterations in consciousness

(Kim et al., 2019)

Complications:

- Cardiac arrhythmias and dysfunctions
- Renal failure
- Disseminated intravascular coagulation (DIC)
- Central nervous system injury
- Death

(Riazi & Brandom, 2015)

Implications for Nursing Care

Preoperative:

- Perform comprehensive history
- MH-susceptibility can be phenotyped via caffeine-halothane muscle contracture tests (CHCT) (Mullins, 2018)
- All perioperative patients should be screened for MH history in self and family
- MH screening should help guide plan of care (Miller, 2017)
- All perioperative patients who have a family history of possible MH susceptibilities may benefit from being screened prior to receiving general anesthesia
- MH may develop after first exposure, but more commonly after three anesthetic exposures (Kim et al., 2019)

Intraoperative:

- MH episodes may resemble sepsis, pheochromocytoma, neuroleptic malignant syndrome, serotonin syndrome, and thyroid storm - differentiation is critical (Kim et al., 2019)
- Monitoring should always include, but not be limited to: continuous temperature, heart rate, oxygen saturation, blood pressure, and end-tidal carbon dioxide monitoring (Mullins, 2018)
- Know where designated MH supplies is stocked on the unit; consider dedicating a specific cart for the supplies if one does not already exist
- Know the facilities recommended treatment guidelines for MH
- Local anesthetics and/or spinal/epidural blocks are safe alternatives to general anesthesia in MH-susceptible patients (Riazi & Brandom, 2015)

Postoperative:

- Transfer patient to critical care
- Recrudescence of MH may occur after symptoms have resolved, close monitoring is recommended for multiple days (Kim et al., 2019)

Treatment

- Removal and cessation of all potential triggers (Riazi & Brandom, 2015)
- Abort surgery and close all incisions as soon as safely possible (Mullins, 2018)
- If surgery cannot be safely stopped, begin nontriggering anesthetics intravenously (Kim et al., 2019)
- Call Malignant Hyperthermia Association (MHAUS) hotline (Kim et al., 2019)
- Hyperventilate patient and utilize inline charcoal filters (if available) to remove trace gases from breathing circuit (Bilmen & Hopkins, 2019)
- Dantrolene (a muscle relaxant that antagonizes the ryanodine receptor, slowing the release of calcium) to be given 2.5mg/kg intravenously, with a maximum dose of 10mg/kg (Kim et al., 2019)
- Draw laboratories as soon as possible: arterial blood gas (ABG), complete metabolic panel, complete blood count, urine myoglobin
- Repeat creatine kinase (CK) levels and ABG analyses until acidosis and rhabdomyolysis resolves (Kim et al., 2019)
- Actively cool patient to less than 38 degrees Celsius with whatever appropriate methods are available (Kim et al., 2019)
- Treat hyperkalemia with calcium, glucose, insulin, and bicarbonate when necessary (Kim et al., 2019)
- Avoid calcium channel blockers when treating potential arrhythmias (can precipitate hyperkalemia) (Kim et al., 2019)
- Fluid resuscitation to achieve a urine output of greater than 1 ml/kg/h (Kim et al., 2019)

Conclusions

MH is estimated to occur in only 1 in 15,000 children and only 1 in 50,000 adults who are exposed to anesthesia, yet it is estimated that somewhere between 1 in 2000 and 1 in 3000 are susceptible to MH (Kim et al., 2019). MH poses an immediate threat to all healthcare facilities delivering anesthesia. It is the sole responsibility of the nursing team to ensure their facility and staff is well equipped and capable of promptly recognizing and treating MH episodes in the susceptible minority. Through vigilance and ongoing education, MH fatalities can be significantly reduced.

References

Figure 1: (Mullins, 2018, p. 583)

Figure 2: (Mullins, 2018, p. 584)

Additional Sources

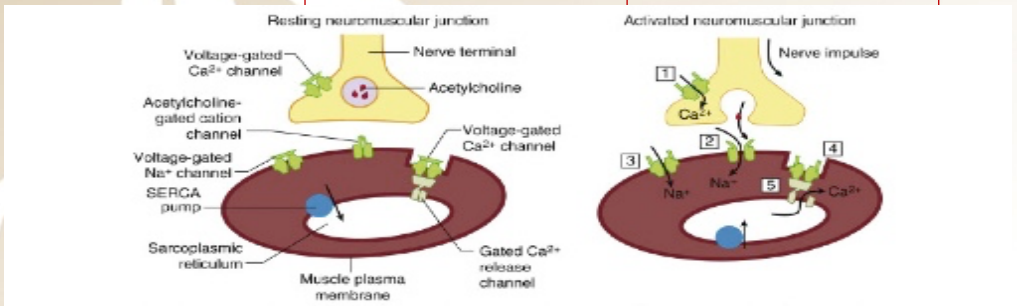

Figure 1. Key ion channels involved in neuromuscular transmission and excitation-contraction coupling. Nerve impulses arriving at the nerve terminal activate voltage-gated Ca^{2+} channels (1). The resulting increase in cytoplasmic Ca^{2+} concentration triggers the exocytosis process of acetylcholine. Binding of acetylcholine to postsynaptic nicotinic acetylcholine receptors (nAChRs) activates an integral nonselective cation channel that depolarizes the sarcolemma (2). Depolarizing the sarcolemma to threshold activates voltage-gated Na^+ channels (3), which initiates action potential impulses that propagate deep into the muscle through the transverse tubule system. Within the transverse tubule system, L-type voltage-gated Ca^{2+} channels sense membrane depolarization and undergo a conformational change (4). A physical link between the $\alpha 1$ subunit (Ca_V 1.1) of the dihydropyridine receptor (DHPR) and the ryanodine receptor (RyR1) is the means by which the electrical signal is transferred from the T tubule to Ca^{2+} release from the SR (5). Reprinted with permission from Elsevier. This figure is available in color online at www.jopan.org.

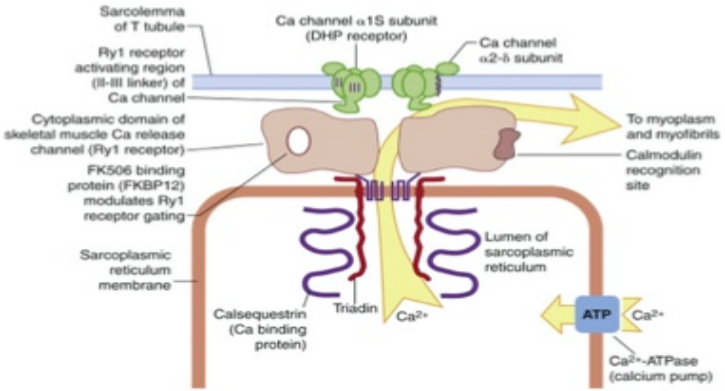

Figure 2. Schematic representation of the triadic junction of skeletal muscle shows the junctional foot protein (ryanodine [Ry1] receptor) and its associated proteins. In skeletal muscle, the α_{1S} subunit of the dihydropyridine receptor (DHPR) participate in the excitation-contraction coupling. These physical links transmit essential signals across the narrow gap of the triadic junction that activate the Ry1 receptor and release Ca^{2+} from the sarcoplasmic reticulum. Reprinted with permission from Elsevier. This figure is available in color online at www.jopan.org.

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