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Preparedness of Nurses for Malignant Hyperthermia

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Introduction

Malignant hyperthermia (MH) is a potentially life threatening disorder that occurs following exposure to certain inhaled anesthetics such as halothane, isoflurane, sevoflurane, desflurane, enflurane, ether, and methoxyflurane alone or in combination with the depolarizing muscle relaxant, succinylcholine (Seifert,, Wahr, Pace, Cochrane, & Bagnola, 2014, p. 189). Patients experiencing malignant hyperthermia may progress to death if it is not recognized and treated early. Patient outcomes improve the earlier an intervention is given. Malignant hyperthermia is not a common condition and, therefore, nurses are frequently unfamiliar with the common signs, symptoms, and treatments. Malignant hyperthermia can occur in a variety of settings where these medications are used such as the operating room, post anesthesia recovery unit (PACU), emergency department, interventional radiology, labor and delivery, intensive care units, dental offices, and ambulatory surgery centers such as surgery centers and office based facilities, hence, nurses of all specialties may encounter MH (Hirshey Dirksen, Van Wicklin, Mashman, Neiderer, & Merritt, 2013, p. 332). Recurrent education and simulation with staff that work with patients receiving possible triggering agents would improve patient outcomes when they are faced with malignant hyperthermia emergencies. The purpose of this scholarly project is to provide education to nurses regarding malignant hyperthermia in hopes that it will increase their knowledge on the subject and prepare them for the medical emergency of malignant hyperthermia.

Pathophysiology

Malignant hyperthermia is a rare, autosomal dominant genetic disorder of the skeletal muscle that causes a life threatening hypermetabolic crisis. The people who are susceptible to MH have a gene mutation in the ryanodine receptor subtype 1 (RYR-1) gene which is found in all skeletal muscle. This gene encodes the calcium ion channel in skeletal muscle and is responsible for calcium release within myocytes. The RYR-1 is also the binding site for inhaled anesthetics and dantrolene (Cain, Riess, Gettrust, & Novalija, 2014, p. 302). When triggering agents such as volatile (inhaled) anesthetics and depolarizing muscle relaxants are administered to an individual with an abnormal RYR-1 gene, there is a prolonged opening of the altered RYR-1 receptor and an uncontrolled intracellular calcium release from the skeletal muscle sarcoplasmic reticulum into the cytosol. This causes a state of intense, sustained skeletal muscle contraction and rigidity which, in turn, causes increased sympathetic activity, oxygen consumption, and the production of carbon dioxide and heat from the rapid use of ATP (Hirshey Dirksen et al., 2013, p. 331). The contracted cells eventually deplete the oxygen and ATP and begin anaerobic metabolism which produces lactic acid and ultimately causes cellular death and destruction. Potassium, as well as other electrolytes (such as calcium, phosphate, and magnesium), creatine kinase(CK), and myoglobin then leak into the bloodstream as a result of the damaged cell membranes (Seifert et al., 2014, p. 195-196).

Significance of Pathophysiology

The significance of the pathophysiology in MH is due to the fatal complications that can occur as a result if it goes untreated. Hyperkalemia causes fatal cardiac dysrhythmias such as ventricular fibrillation and ventricular tachycardia. Myoglobinemia causes renal failure as the excessive CK and myoglobin released into the circulation blocks the renal tubules. Other complications, many of which are fatal, from MH include liver failure, compartment syndrome from profound muscle swelling, skeletal muscle damage, mixed respiratory acidosis or metabolic acidosis, abnormal blood coagulation and DIC, internal hemorrhage, neurologic injury, hyperthermia, rhabdomyolysis, bowel ischemia, CHF, pulmonary edema, end organ damage, cardiac collapse or arrest, and death (Hirshey Dirksen, et al., 2013, p. 333; Seifert et al., 2014, p. 189). These clinical signs are results of the inability of the body to maintain the increased metabolic demand during a malignant hyperthermia crisis.

Signs and Symptoms

The signs and symptoms of malignant hyperthermia usually occur within minutes or up to an hour after the administration of the triggering agent, however, if succinylcholine is used in conjunction with an inhaled anesthetic, the onset of symptoms is generally quicker. It is possible that MH may develop up to 36 hours after exposure to the triggering agent. The signs and symptoms of MH can present in any sequence, but there are typically three signs that manifest the earliest (Cain et al., 2014. p. 302-303). The first sign of MH is hypercarbia or an unexplained, progressive increase in carbon dioxide production. The ETCO2 can double or triple a normal capnography value in MH. The next signs that typically manifest are masseter (jaw) muscle spasm and sinus tachycardia. Other early signs include flushed skin, dysrhythmias, such as PVC's and bigeminy, and generalized muscle rigidity. Hyperthermia is actually a late sign of MH despite the name of this disorder. The rapid increase in body temperature can exceed 43.3° C [109° F], and it can increase by 1° C to 2° C [1.8° F to 3.6° F] every five minutes (Seifert et al., 2014, p. 189). Other late signs are metabolic acidosis, hypoxia, mottled skin, cyanosis, coagulopathy, left ventricular failure evidenced by pulmonary edema, rales, and frothy sputum, and brown, cola-colored urine from rhabdomyolysis. Other possible signs and symptoms of MH include mixed respiratory acidosis, tachypnea, sweating, myoglobinuria, hyperkalemia, ventricular tachycardia, and ventricular fibrillation (Stewart, 2014, p. 253-254). MH can recur within 24 to 48 hours after the initial event, so the patient with MH should be monitored for recrudescence, which is the recurrence of symptoms after a period of remission (Hirshey Dirksen, 2013, p. 333).

Signs and Symptoms

- Hypercarbia
- Tachycardia
- Masseter spasm
- Flushed skin
- Cardiac dysrhythmias
- Generalized muscle rigidity
- Tachypnea
- Hyperthermia
- Hypoxia
- Acidosis
- Coagulopathy

Implications for Nursing Care

The goal of treatment of malignant hyperthermia is to reverse the hypermetabolic state and prevent the potentially lethal consequences of MH. The likelihood of a positive outcome after diagnosis of MH improves with expeditious treatment. If MH is not treated rapidly, the mortality rate is high. The most effective treatment for MH is the administration of dantrolene sodium. The mortality rate from MH used to be as high as 80%, but since the discovery of dantrolene in 1975, it is now down to 5% (Hirshey Dirksen et al., 2013, p. 330). Dantrolene is a specific ryanodine receptor antagonist and reverses the MH related muscle contractions by decreasing the calcium in muscle cells (Seifert et al., 2014, p. 192). The longer time that elapses between the onset of MH and the first dose of dantrolene results in an increase in complications associated with MH. It is well known that the preparation of dantrolene is difficult. Some of the difficulties with dantrolene preparation are that it requires large quantities of diluents to be mixed with large quantities of the medication and it requires complicated medication calculations. The required dose of dantrolene is 2.5 mg/kg rapidly through a large bore IV every 5 minutes and repeat until symptoms subside or a maximum dose of 30 mg/kg has been reached. Dantrolene comes in 20 mg vials and must be mixed with 60 ml sterile water (Seifert et al., 2014, p. 192-194). For a patient who weighs 100 kg, the initial dose of dantrolene would be 250 mg. That would require 12.5 vials of dantrolene to be mixed and a total of 750 ml sterile water. If this medication preparation must be done every five minutes, it can become time consuming and difficult to calculate. It may take several nurses mixing dantrolene at the same time to get the first critical dose in a timely manner. In a rare situation like MH, this skill is not second nature and must be practiced to be effective in an emergency. Another obvious treatment of MH is to stop the volatile anesthetic and either abort the procedure or switch to a non-triggering anesthetic. An increase in oxygenation to 100% FIO2 will help prevent anaerobic metabolism, and hyperventilation will decrease the excess CO2 build up resulting from MH (Seifert et al., 2014, p. 195). Also, sodium bicarbonate can be given to treat metabolic acidosis, and insulin and calcium can be given to treat hyperkalemia (see Figure 1). Nurses should be prepared to start cooling measures immediately by using cooling blankets, gastric lavage, body cavity irrigation such as the surgical site, and ice packs. Shivering should be avoided, though, because it will increase the metabolic state. The goal is to keep the temperature of the patient with MH lower than 39° C [102.2° F] (Cain et al., 2014. p. 302). Cardiac dysrhythmias should be treated with normal ACLS medications, but calcium channel blockers should not be used because they interact with dantrolene at the receptor site (Seifert et al., 2014, p. 196). After swift and appropriate treatment, the patient with MH would hopefully revert back to a normal metabolic state with no complications.

Conclusion

Malignant hyperthermia is a very rare occurrence and only occurs approximately once in every 3,000 to 50,000 procedures where volatile anesthetics are administered (Dirksen et al., 2013, p. 330). It is of importance, though, because it can progress quickly to a life threatening situation and is a medical emergency. Because it is rare, many nurses are unprepared to care for a patient experiencing MH. The specifics about the signs and symptoms, diagnosis, treatment, and location of MH emergency supplies should be reviewed frequently by personnel that may encounter it (Hirshey Dirksen, 2013, p. 333). The areas where MH may occur are vast which means nurses of all specialties need to review this information at a minimum of each year to give the patient experiencing MH a chance for the best outcome. Education and simulation are integral parts of MH preparedness.

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References

Cain, C. L., Riess, M. L., Gettrust, L., & Novalija, J. (2014). Malignant hyperthermia crisis: Optimizing patient outcomes through simulation and interdisciplinary collaboration. *AORN Journal*, *99*(2), 300-311. doi: http://dx.doi.org/10.1016/j.aorn.2013.06.012.

Hirshey Dirksen, S. J., Van Wicklin, S. A., Mashman, D. L., Neiderer, P., & Merritt, D. R. (2013). Developing effective drills in preparation for a malignant hyperthermia crisis. *AORN Journal*, *97*(3), 330-352. doi: http://dx.doi.org/10.1016/j.aorn.2012.12.009

Hutton, D. (2011). Emergency preparedness in the OR. Malignant hyperthermia---Part 1. *Plastic Surgical Nursing*, *31*(1), 23-28. doi: http://dx.doi.org/10.1097/PSN.0b013e31820a6363

Johns, C. D., Stoudt, R. S., Scholtis, M. P., & Gavel, T. (2012). Malignant hyperthermia: a crisis response plan. *OR Manager*, *28*(6), 18-21. Retrieved from http://ezproxy.otterbein.edu/login?url=http://search.ebscohost.com/login.aspx?direct=true&db=rzh&AN=2011564093&site=eds-live

Mitchell-Brown, F. (2012). Malignant hyperthermia: Turn down the heat. *Nursing*, *42*(5), 38-45. doi: http://dx.doi.org/10.1097/01.NURSE.0000413609.98565.96

Mullen, L., & Byrd, D. (2013). Using simulation training to improve perioperative patient safety. *AORN Journal*, *97*(4), 419-427. doi: http://dx.doi.org/10.1016/j.aorn.2013.02.001

Musselman, M. E., & Saely, S. (2013). Diagnosis and treatment of drug-induced hyperthermia. *American Journal of Health-System Pharmacy*, *70*(1), 34-42. doi: http://dx.doi.org/10.2146/ajhp110543

Schneiderbanger, D., Johannsen, S., Roewer, N., & Schuster, F. (2014). Management of malignant hyperthermia: Diagnosis and treatment. *Therapeutics & Clinical Risk Management*, *10*, 355-362. doi: 10.2147/TCRM.S47632

Seifert, P. C., Wahr, J. A., Pace, M., Cochrane, A. B., & Bagnola, A. J. (2014). CRISIS CONSIDERATIONS. Crisis management of malignant hyperthermia in the OR. *AORN Journal*, *100*(2), 189-202. doi: http://dx.doi.org/10.1016/j.aorn.2014.06.014

Stewart, M. W. (2014). Anesthetic drugs and malignant hyperthermia. *Journal of PeriAnesthesia Nursing, 29*(3), 253-255. doi: http://dx.doi.org/10.1016/j.jopan.2014.03.002

[Untitled photograph of tachometer on fire]. (n.d.). Retrieved from http://aa2day.org/2014/11/december2014/

Medication Doses and Treatments for Malignant Hyperthermia (MH)

AORN

DANTROLENE TREATMENT

- Mix each ampule with 60 mL sterile (ie, preservative-free) water
- Administer 2.5 mg/kg IV every 5 minutes until symptoms subside; may require up to 30 mg/kg
- Ensure that a sufficient supply of dantrolene is available because MH symptoms can "wax and wane" (eg, symptoms may reoccur after treatment seems successful even during the postoperative period)
- Use warm, sterile water (113° F [45° C]), if available, for reconstitution, which may help dissolve the dantrolene into the solution more quickly, thereby enabling faster administration of dantrolene to the patient; however, the health care provider who mixes the dantrolene should not allow warming of the diluent to cause a delay in administration

SODIUM BICARBONATE TREATMENT (for suspected metabolic acidosis)

- Administer 1 mEq/kg to 2 mEq/kg slow IV push (may be administered if arterial blood gas values are not available)

HYPERKALEMIA TREATMENT

- Administer calcium gluconate 30 mg/kg or calcium chloride 10 mg/kg IV
- Adult patients: administer regular insulin 10 units IV with 1 to 2 amps (ie, 50 mL to 100 mL) $D_{50}W$
- Pediatric patients: administer regular insulin 0.1 units/kg and 1 mL/kg $D_{50}W$

Adapted from the Malignant Hyperthermia Crisis Checklist [2013] *with permission from Ariadne Labs, Brigham and Women's Hospital and Harvard School of Public Health; Boston, MA.*

Figure 1. Medication doses and treatments for malignant hyperthermia. Reprinted from "CRISIS CONSIDERATIONS. Crisis Management of Malignant Hyperthermia in the OR", by P. C. Seifert, J. A. Wahr, A. B. Cochrane, and A. J. Bagnola, 2014, AORN Journal, 100(2), p. 198. Copyright 2014 by Elsevier Inc. Reprinted with permission.