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Turning Up the Heat on Malignant Hyperthermia

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Introduction

Surgeries are common, everyday procedures within the walls of America's hospitals. According to Orser, Mazer, and Baker (2008), more than forty million patients in North America are given anesthetics annually. One of the major complications of anesthesia is malignant hyperthermia: a hyper-metabolic state that affects skeletal muscles. If left uncontrolled, malignant hyperthermia can cause multiple reactions within the body leading to metabolic and respiratory acidosis, cardiac dysrhythmias, kidney failure, coagulopathy, neurologic injury, and ultimately death (Seifert, Wahr, Pace, Cochrane, & Bagnola 2014). The incidence of this condition is estimated to be 1:15,000 in children and 1:20,000-50,000 in adults (Redmond, 2001). Although considered a rare event, the mortality rate of malignant hyperthermia was eighty percent in 1960 and has steadily declined to less than ten percent by 1980 (Saleh, 1992). The concern, however, revolves around the facts that malignant hyperthermia is a hereditary condition that is difficult to screen for prior to the administration of anesthesia, and it occurs in every country and race, male and female, child and adult (Donnelly, 1994). Prompt intervention is crucial and can adequately impede the progression of this heightened metabolic state, yet, delayed action will quickly increase risk of death (Redmond, 2001). Therefore, continued education in regards to the pathophysiology of malignant hyperthermia will aid the surgical team in identifying risk factors, recognizing early signs, and interceding appropriately to protect patients from a potentially life-threatening reaction.

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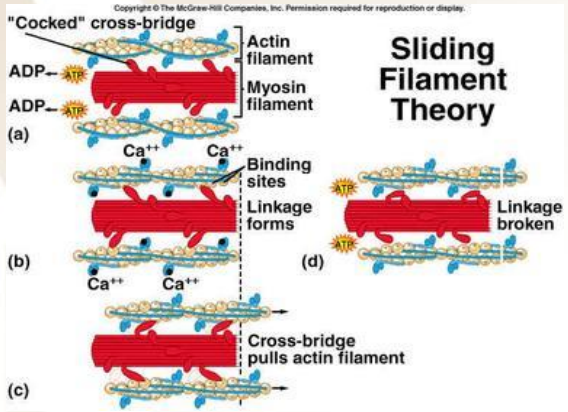
Pathophysiological Processes

Malignant Hyperthermia (MH) is considered a pharmacogenetic disorder as it is an autosomal dominant disorder activated by the introduction of certain anesthetic agents solely or in conjunction with a neuromuscular blocker known as succinylcholine (Stratman, Flynn, & Hatton, 2009). Those genetically susceptible have a defect of the ryanodine receptor (RYR 1), which encodes for the calcium channel on the sarcoplasmic reticulum of skeletal muscle cells. Once triggered by anesthesia agents, there is an exaggerated release of calcium resulting in a cascade of potentially lethal reactions. Uncontrolled increase in calcium stimulates actin and myosin to produce prolonged muscle contractions leading to accelerated adenosine triphosphate (ATP) depletion; oxygen consumption; glucose metabolism; lactate, carbon dioxide, and heat production. Consequently, acidosis, hyperthermia, and dangerously low ATP levels ensue thus causing the sarcolemma of the muscle cells to become damaged, cell death occurs, and intracellular contents such as magnesium, potassium, phosphate, and myoglobin are leaked systemically. The domino effect described places the patient in a hyper-metabolic state with potential for multiple organ damage and failure (Stratman et al., 2009).

Meanwhile, the hyper-metabolic state perpetuates the rise in body temperature potentiating the risk for coagulation disorders such as disseminated intravascular coagulopathy and neurologic damage (Dirksen, Van Wicklin, Mashman, Neiderer, Merritt, 2013). However, the pathophysiology of MH can be significant in a positive way as it identifies what interventions will be effective in hindering the devastating pathway from continuing. For example, understanding the genetic origin of MH has led to genetic testing to identify those susceptible, knowledge of the sustained muscle contraction that occurs and begins the lethal progression has led to the use of dantrolene, a muscle relaxant, as the first-line treatment (Stratman et al., 2009), and the awareness of the systemic effects have led to numerous interventions to prevent a secondary injury (Dirksen et al., 2013). Therefore, understanding the pathophysiology of MH can help direct the perioperative team to prompt recognition and appropriate treatment of such a destructive disorder.



Case Study

A 22 year old male presented to a military hospital after being injured by an improvised explosive device. He presented with a penetrating injury to his left thigh; radiological findings confirmed a left femur fracture. The patient was conscious and able to answer questions; he denied any allergies or past medical history. The man was prepared for surgery and given ketamine, 50 mg; etomidate, 30 mg; and succhinylcholine, 120mg for rapid sequence intubation. Post-intubation vitals are as follows: blood pressure (BP) 115/60 mmHg; heart rate (HR) 90/min; arterial oxygen saturation (Sao2) 100%; end tidal carbon dioxide (ETCO2) 42 mmHg; temperature 98.6 degrees F. General anesthesia was maintained with sevoflurane. Total operative time was two hours with no complications.

However, upon completion, the BP was 118/42; HR 106; Sao2 100%, ETCO2 65; and temp 100.4F. Pain medicine was given for increased heart rate yet fifteen minutes later the ETCO2 was 90, BP 140/78; HR 110, temp 100.6. Hyperventilation was initiated to reduce ETCO2, and the anesthesia machine was inspected but no malfunction found. Concurrently, severe bilateral masseter muscle contractions and increased abdominal muscle rigidity were noted. The temperature climbed to 100.9F, labs were sent revealing hyperkalemia and a diagnosis of MH was made. The patient was then given dantrolene, 2.5mg/kg. Once the dantrolene bolus was given, muscular rigidity disappeared. Twenty minutes following bolus, ETCO2 was 35 and temperature was 100.5F, all other vital signs were stable. Seventy minutes after treatment, the patient was completely stable and was transported to another facility (Banet, Weatherwax, Spence, Perry, Muldoon & Capacchione, 2013).

Signs and Symptoms

Unfortunately, other diseases such as thyroid storm, heat stroke, pheochromocytoma, cocaine overdose, etc. can present in phenotypically similar ways as MH making timely diagnosis a challenge. Moreover, many genetically susceptible patients undergoing general anesthesia are unaware of their genetic defect; it is likely there will be no forewarning of this rapidly debilitating disorder. Therefore, detection of the signs and symptoms of malignant hyperthermia is imperative for the healthcare team in order to improve patient survival. The signs and symptoms for MH can be classified as either early or late. Typically, the earliest signs can occur within minutes of exposure and include: increased end tidal carbon dioxide, tachycardia, and contraction of the masseter muscle of the jaw. Other early signs are tachypnea, generalized muscle rigidity, cardiac arrhythmias, and electrolyte imbalances.

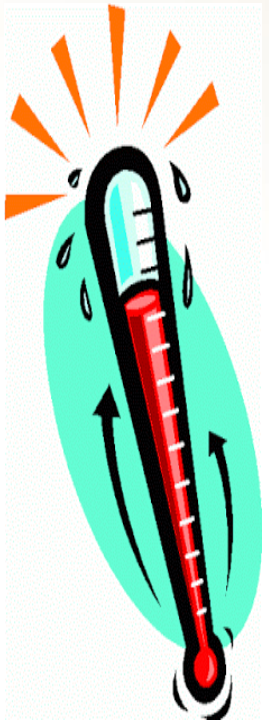
Late signs can occur hours after exposure and are associated with mottled skin, disseminated intravascular coagulation, rhabdomyolysis, myoglobinuria, renal failure, respiratory and metabolic acidosis, and hyperthermia. Once hyperthermia sets in, shock and lethal arrhythmias can occur as quickly as twenty minutes due to oxygen consumption that is two to three times the normal rate of consumption (Stratman et al., 2009). All in all, the longer the disorder has to disturb the body's homeostasis, the harder it is to combat. Early detection is paramount when dealing with malignant hyperthermia.

Nursing Implications

Malignant hyperthermia is a rare and fatal disorder that can be challenging to detect and treat. Due to the low incidence of MH events, medical personnel are often not familiar and/or confident in the management of such an event. Therefore, it is the responsibility of the healthcare team to familiarize themselves with the disorder, the signs and symptoms, expected treatments, and evaluation of successful intervention. A study was done in the Midwest with all surgical personnel being placed in various simulations that were set to resemble a malignant hyperthermia crisis. The staff was educated on the disorder, simulation was completed, and all members were debriefed. The findings produced protocols and exercises that are done on a monthly basis to ensure all parties of the OR team are prepared for MH if it should arise. One recommendation that was vital to the teams was assigning each person with a specific task in order to accomplish all the interventions needed to safely care for a MH patient. For example, multiple members needed to dilute dantrolene as this can be a tedious step, another cooled the patient with ice, another documented the event, etc. (Cain, Reiss, Gettrust, & Novalija, 2014). PACU and ICU nurses also need to be educated as many signs and symptoms of MH may not present in the OR but in the PACU or critical care units (Barnes, Stowell, Bulger, Langton, & Pollock, 2015). Nurses need to provide education for the patients and families of those whom have suffered with MH, assuring they understand how critical this disorder can be and how pertinent it is that they allow future anesthesia teams know of their susceptibility to the deadly disorder.

Conclusion

Malignant hyperthermia is a complex, life-threatening crisis that presents multiple challenges yet can be successfully impeded with rapid and proper treatment. Continued education is essential for early detection, educated planning, swift implementation, and proficient evaluation of an MH event. Thorough pre-operative assessments, strong intra-operative knowledge, and efficient interdisciplinary roles will only bolster the fight against MH and ensure significant reduction in morbidity and mortality (Stratman et al., 2009).



References

Banek, L. R., Weatherwax, L. J., Spence, U. C. D., Perry, L. S., Muldoon, S., & Capacchione, J. (2013). Delayed Onset of Suspected Malignant Hyperthermia During Sevoflurane Anesthesia in an Afghan Trauma Patient: A Case Report. AANA journal, 81(6), 441.

Barnes, C., Stowell, K. M., Bulger, T., Langton, E., & Pollock, N. (2015). Safe duration of postoperative monitoring for malignant hyperthermia patients administered non-triggering anaesthesia: an update. Anaesthesia and intensive care, 43(1), 98-104.

Brownstown Biomes (2015). [Thermometer]. Retrieved from brownstownbiomes.wikispaces.com.

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.

Dirksen, S. J. H., 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), 329-353.

Donnelly, A. J. (1994). Malignant hyperthermia: epidemiology, pathophysiology, treatment. AORN journal, 59(2), 393-405.

Katielyn & Kelsey (2015). [sliding filament theory]. Retrieved from www.katelynanderson.wikispaces.com

Orser, B. A., Mazer, C. D., & Baker, A. J. (2008). Awareness during anesthesia. Canadian Medical Association Journal, 178(2), 185-188.

Redmond, M. C. (2001). Malignant hyperthermia: perianesthesia recognition, treatment, and care. Journal of PeriAnesthesia Nursing, 16(4), 259-270.

Saleh, K. L. (1992). Practical points in the management of malignant hyperthermia. Journal of post anesthesia nursing, 7(5), 327.

Seifert, P. C., Wahr, J. A., Pace, M., Cochrane, A. B., & Bagnola, A. J. (2014). Crisis Management of Malignant Hyperthermia in the OR. AORN journal, 100(2), 189-202.

Stratman, R. C., Flynn, J. D., & Hatton, K. W. (2009). Malignant hyperthermia: a pharmacogenetic disorder. Orthopedics, 32(11), 835.