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Malignant Hyperthermia. Nursing Implications for Care.

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Introduction

Malignant Hyperthermia (MH) is a rare condition which can be developed during a surgical procedure or postoperatively. Unfortunately due to its nature, nurses are not fully aware of the signs and symptoms and moreover they do not possess the knowledge for its treatment.

The current article is a collection of the provided knowledge from the most related nursing and non-nursing articles of this particular condition.

Malignant Hyperthermia (MH) was first described by Denborough and Lovell back in 1960, when the two scientists first described an extreme reaction caused by anaesthetic drugs (Denborough and Lovell 1960, Pritchard 2003). The incidence of MH is between 1:50.000 to 1:100.000 in adults and 1:300.000 to 1:500.000 in children (Chipas 2005). Malignant hyperthermia occurs most commonly between the ages of 2 and 42 years. Seventy-five per cent of MH index cases have had previous anaesthesia prior to their MH crisis (Onofrei and Bromhead 2009). First-degree relatives of someone who has had MH or been diagnosed as MH susceptible are much more susceptible to MH than the general population. Second-degree relatives are less susceptible than first-degree relatives but have a greater than normal risk compared to the general population (Hommertzheim and Steinke 2006). It is obvious from the epidemiological data above that MH it is not a very common condition resulting to decreased exposure of nurses to it. But what is the mechanism for the development of the condition?

Calcium is an essential ion in normal cell energy production and muscle contraction. Normally, when a nerve impulse stimulates skeletal muscle, calcium is released from the sarcoplasmic reticulum, allowing muscle contraction to occur. In cases of MH, the triggering agent disrupts normal calcium ion concentrations, which causes an increase in calcium uptake in skeletal muscle fibres. This result in an increase in oxygen consumption and an increase in lactate and heat production leading to a hypermetabolic state (Martin 2009, Hommertzheim and Steinke 2006). The triggering agents are volatile anaesthetic drugs such as isoflurane, enflurane, desflurane, sevoflurane, halothane or intubation drugs such as suxamethonium.

These agents cause an abnormal calcium flux in the sarcoplasmic reticulum of the mitochondria, of the skeletal muscle cells and possibly cardiac muscle (Aithkenhead and Smith 1996, Neascu 2006). The inheritance gene for MH is identified on the long arm of chromosome 19 (Craft and Upton 1995, Morgan and Mikhail 1996) and as an inherited autosomal dominant trait, it requires only one parent to have the condition for a child to inherit it (Krivosic-Horber 1996, Saljoughian 2003).

Clinical Signs

Signs and symptoms of MH include the following (Mitchell-Brown 2012): Hypercapnia: Acidosis occurs in 80% of all patients experiencing MH in response to increased glycogenesis resulted to progressive, unexpected increase in ETCO2 lactic acid, and heat (DeLamar 2003, Mecca and Rosenberg 2004). This change is the earliest and most sensitive sign that a patient might be experiencing MH (Litman 2012). Normal ETCO2 is 35 to 45 mm Hg (Tautz et al 2010). In MH, the ETCO2 value may double or even triple on capnography. Usually this unexplained increase in ETCO2 doesn't respond to increased ventilation or additional administration of anaesthetic (Hsu 2007). The sudden increase in end-tidal CO2 is accompanied by oxygen destruction during normal ventilation. The sustained contracture of the skeletal muscle groups leads to increased use of available ATP and oxygen. Maintenance of normal oxygen saturation usually is dependent on increasing inspired oxygen content (De Lamar 2003, Mecca and Rosenberg 2004). Cyanotic or mottled skin is present in approximately 70% of patients experiencing MH and usually starts with a generalized erythematous flush (DeLamar 2003, Mecca and Rosenberg 2004). Tachycardia: Although an early sign of MH, tachycardia occurs in 96% of all patients experiencing MH but is nonspecific (DeLamar 2003, Mecca and Rosenberg 2004). Generalized muscle rigidity, especially of the jaw, trunk, and extremities. According to Noble (2007), muscle rigidity, especially masseter muscle rigidity, is seen in 75% of patients experiencing an MH episode. When masseter muscle rigidity is present, the patient's mouth can't be opened after exposure to the triggering drug.

However, transient masseter muscle rigidity is normally associated with the administration of succinylcholine and doesn't necessarily indicate MH unless it persists after potential triggering agents are discontinued (Litman 2012). ECG changes: Hyperkalaemia may cause premature ventricular contractions, which may progress to life threatening ventricular tachycardia or ventricular fibrillation (Litman 2012). Rhabdomyolysis: Destruction of skeletal muscle releases large amounts of the enzyme creatine kinase (CK) and myoglobin, which accumulates in the kidneys. Red or tea-coloured urine is a sign of myoglobinuria. Levels of serum CK and urine myoglobin, which peak about 14 hours after the acute MH episode, vary depending on the patient's muscle mass and the severity of the patient's condition (Litman 2012). Hyperthermia: An increase in body temperature is usually a later sign which confirms the suspicion of MH. The patient's temperature may rise 1° C every 5 minutes and might even exceed 40.6° C (Litman 2012). Electrolyte imbalances: Besides hyperkalaemia, it is important the continuous monitoring of the electrolytes balance since the MH crisis has an effect on the levels of phosphate and calcium also, leading to hyperphosphatemia and hypocalcaemia respectively (Mitchel Brown 2012).

Differential Diagnosis

Although all these signs and symptoms are unexplained and life threatening for the patient, they are not exclusive to the MH crisis. There are other conditions that have similar symptoms, which the health care providers must be aware of, in order to perform a rapid differential diagnosis. These conditions could be: inadequate anaesthesia or analgesia, inappropriate breathing circuit / fresh gas flow / ventilation leading to inadequate ventilation, cocaine toxicity, endocrine disorders such as phaeochromocytoma and thyrotoxicosis, sepsis, hypoxic encephalopathy, and intracranial trauma (Onofrei and Bromhead 2009, Hommertzheim and Steinke 2006).

Management

One of the important factors for the successful treatment of the condition is time. The collaboration and the rapid response of the multidisciplinary team are very crucial (Hernandez et al 2009). DeJohn (2008) believes there is a strong correlation between the timing of interventions and positive patient outcomes. During surgery, the priorities are to discontinue the use of any inhalational anaesthetics and succinylcholine, optimization of oxygenation and ventilation, and administration of dantrolene immediately. The team must make sure that the patient should have arterial access line and central venous access. Administer dantrolene. Considered the gold standard, dantrolene sodium is the only known effective treatment for MH. Dantrolene is classified as a direct-acting skeletal muscle relaxant. It may stop skeletal muscle contraction by interfering with calcium release from the sarcoplasmic reticulum, reversing skeletal muscle hypermetabolism. (Litman 2012, British National Formulary 2012). Because MH signs and symptoms recur in up to 25% of patients after initial treatment, maintenance doses of dantrolene should continue for 48 hours after the last observed sign of acute MH. If signs continue despite on-going treatment, additional dantrolene doses or a dantrolene infusion may be required. Monitor the venous access site closely for signs of dantrolene extravasation, including pain, erythema, and oedema, which can lead to tissue necrosis due to its high pH. Provide adequate oxygenation. Administer oxygen at 100% to meet the patient's high metabolic demands. If the patient is not intubated consider preparing for endotracheal intubation. Close monitoring ETCO2 by continuous capnography. Assess the patient for signs and symptoms of fluid volume overload, such as pulmonary crackles, because the patient will be receiving large amounts of I.V. fluids. Closely monitor continuous pulse oximetry and ABG values. Maintain haemodynamic stability. Patient should be under continuous cardiac monitor and frequently assess BP, heart rate, core body temperature, and central venous pressure. An arterial line is beneficial not only for continuous BP monitoring but also for frequent arterial blood sampling (Stratman RC, Flynn JD, Hatton KW 2009, Mitchel Brown 2012).

Furthermore the management of MH is a continuous process which it doesn't stop after the end of the crisis itself. Following initial stabilisation, the patient should be transferred to an intensive care or high dependency unit for at least 24 hours or until vital signs have returned to normal. Dantrolene should be continued during this period depending on the patient's response to treatment. The patient's close monitoring continuous including monitoring of electrolytes (potassium) and creatine phosphokinase levels, renal function in case of the onset of myoglobinuria, clotting status and temperature (Pritchard M. 2003).

In the previous paragraphs, there was an introduction of the signs and symptoms of the condition plus the management once it is developed. Nurse should be aware though, of the pre surgery tests that can be done in order to assess if a patient has increased chance of developing MH. The most known test is the Caffeine–Halothane Contracture Test.

The caffeine– halothane contracture test (CHCT) requires approximately 2gr of muscle excised from the vastus lateralis or vastus medialis muscle. The tissue sample is tested for its viability with electrical stimulation in order to assess the level of muscle contracture. The sample is divided in pieces which then are being exposed to either halothane or caffeine under continuous evaluation of the muscle contracture.

If a contracture of 0.7 g or greater develops in any halothane-exposed muscle strip or if a contracture of 0.3 g or greater develops in any strip exposed to caffeine at 0.5, 1, or 2 mM, then the test is considered to be positive, and the patient has MHS (Rosenberg, Antognini, Muldoon 2002, Baur et al 2000).

But what are the implications for nursing practice further to the knowledge of the condition's signs, symptoms and management?

Nursing training. Given the complexity of treating MH the literature suggests the development of training days with simulated situations of MH crisis and incidents, at least once a year. These kinds of exercises, not only keep the staff updated and alerted but also improve the staff collaboration. It's important that all staff members who may be involved in caring for the patient during an MH crisis attend an MH education session and participate in the MH simulations. Furthermore the staff should be aware of their Trust's protocols and guidelines, how to access their emergency trolley and what it should include The MH cart should be checked frequently to ensure that expired supplies are removed and replaced, access to ice, cooled I.V. fluids and cooling blanket must be ensured (Mitchel Brown 2012, Martin 2009).

Nurses are also responsible for the education of their patients. When the MH patient is able to be discharged, it's important to educate him or her about MH. The patient should also understand the signs and symptoms of non-acute MH expression such as muscle cramps, low-grade fever, and fatigue. The patients should be instructed to inform all healthcare providers about MH susceptibility before undergoing surgery or dental work (Martin 2009).

Conclusion

Malignant hyperthermia is a rare but emergency hyper metabolic syndrome and Dantrolene is the only medication to treat it. Perioperative nurses play a critical role in helping maintain a safe environment for the patient in surgery. It is essential then for the nurses, to be able to identify the risk factors for MH and be prepared to respond fast and effectively after the development of an MH crisis.

References

Aitkenhead A, Smith G (1996) Textbook of Anaesthesia. Third edition. Churchill Livingstone, London.

Baur CP, Bellon L, Felleiter P, Fiege M, Fricker R, Glahn K, Heffron JJA, Herrmann-Frank A, Jurkat-Rott K, Klingler W, Lehane M, Ording H, Tegazzin V, Wappler F, Georgieff M, Lehmann-Horn F (2000) A multicenter study of 4-chloro-mcresol for diagnosing malignant hyperthermia susceptibility. Anesth Analg. 90, 00–5.

British National Formulary (2012) British National Formulary No. 63. British Medical Association and Royal Pharmaceutical Society of Great Britain, London.

Chipas T "Malignant hyperthermia,"seminar presented at Via Christi Regional Medical Center, Wichita, Kan, 8 Feb 2005.

Craft TM, Upton PM (1995) Key Topics in Anaesthesia. Second edition. Bios Scientific Publishers, Oxford.

DeJohn P (2008) Be prepared: malignant hyperthermia. OR Manager. 24,6,26-27.

DeLamar L (2003) "Malignant hyperthermia," in Alexander's Care of the Patient in Surgery, 12th ed, J Rothrock, ed. Mosby, St Louis 247-249, 1219.

Denborough MA, Lovell RRH (1960) Anaesthetic deaths in a family (letter). Lancet 2,45.

Hernandez JF, Secrest JA, Hill L, McClarty SJ (2009) Scientific advances in the genetic understanding and diagnosis of malignant hyperthermia. J Perianesth Nurs. 24,1,19-31.

Hommertzheim R., Steinke E (2006) Malignant hyperthermia-The perioperative nurse's role. AORN J. 83, 1, 149-164.

Hsu SC, Huang WT, Yeh HM, Hseieh AY(2007) Suspected malignant hyperthermia during sevoflurane anesthesia. J Chin Med Assoc. 70,11,507-510.

Krivosic-Horber R (1996) The Molecular Biology of Malignant Hyperpyrexia. www.jlar.com/Congres_anterieurs/SENFA%2097/resume%202.htm (Last accessed: June 2013.)

Litman RS (2012) Malignant hyperthermia: clinical diagnosis and management of acute crisis. UpTo Date. http://www.uptodate.com (Last accessed: June 2012).

Martin C. L (2009) A practical guide for malignant hyperthermia management. OR Nurse. 3, 3, 20-26.

Mecca R, Rosenberg H (2004) Malignant Hyperthermia: Knowing Your Role. Philadelphia: Malignant Hyperthermia Association of the United States.

Mitchell-Brown F (2012) Malignant hyperthermia: turn down the heat. Nursing. 42, 5, 38-44.

Morgan EG, Mikhail MS (1996) Clinical Anaesthesiology. Second edition. Appleton and Lange, Stamford CT.

Neascu (2006) Malignant hyperthermia. Nursing Standard. 20,28,51-57.

Noble KA (2007) Malignant hyperthermia: hot stuff! J Perianesth Nurs. 22,5,341-345.

Onofrei M, Bromhead H (2009) Malignant Hyperthermia. ATOTW 131, 1-6.

Pritchard, M. J. (2003) Malignant hyperthermia: clinical features and management. Nursing Times; 99: 23, 32–34.

Rosenberg H, Antognini J F, Muldoon S (2002) Testing for Malignant Hyperthermia. Anaesthesiology. 96, 232–7.

Saljoughian M (2003) Malignant hyperthermia: prevention and therapy. US Pharmacist. 28,3,25-30.

Stratman RC, Flynn JD, Hatton KW (2009) Malignant hyperthermia: a pharmacogenetic disorder. Orthopaedics. 32,11,835.

Tautz TJ, Urwyler A, Antognini JF, Riou B(2010) Case scenario: increased end-tidal carbon dioxide: a diagnostic dilemma. Anaesthesiology. 112,2,440-446.