Malignant Hyperthermia: Recognition and Treatment

NYSNA Continuing Education

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How to Take This Course

Please take a look at the steps below; these will help you to progress through the course material, complete the course examination and receive your certificate of completion.

1. **REVIEW THE OBJECTIVES**

   The objectives provide an overview of the entire course and identify what information will be focused on. Objectives are stated in terms of what you, the learner, will know or be able to do upon successful completion of the course. They let you know what you should expect to learn by taking a particular course and can help focus your study.

2. **STUDY EACH SECTION IN ORDER**

   Keep your learning "programmed" by reviewing the materials in order. This will help you understand the sections that follow.

3. **COMPLETE THE COURSE EXAM**

   After studying the course, click on the "Course Exam" option located on the course navigation toolbar. Answer each question by clicking on the button corresponding to the correct answer. All questions must be answered before the test can be graded; there is only one correct answer per question. You may refer back to the course material by minimizing the course exam window.

4. **GRADE THE TEST**

   Next, click on "Submit Test." You will know immediately whether you passed or failed. If you do not successfully complete the exam on the first attempt, you may take the exam again. If you do not pass the exam on your second attempt, you will need to purchase the course again.

5. **FILL OUT THE EVALUATION FORM**

   Upon passing the course exam you will be prompted to complete a course evaluation. You will have access to the certificate of completion **after you complete the evaluation**. At this point, you should print the certificate and keep it for your records.

Introduction

How could anything be more devastating than the death of a healthy individual undergoing minor surgery? Malignant hyperthermia (MH) is a rare, but dramatic, hypermetabolic reaction that individuals can experience if they are susceptible to certain anesthetic agents. When it does occur, it happens fast. Most times signs of MH will not appear until the triggering agent is given. Death can occur within 24 hours. Great strides have taken place in the identification and treatment of MH. Staff and patient education in the areas of detection and screening has made a significant impact on the reduction in mortality rate to approximately 5% (Rosenberg, 2010).

This course will provide an overview of malignant hyperthermia, triggering agents, preoperative considerations, signs and symptoms, and prompt intervention and treatment. The professional nurse has a responsibility to screen preoperative patients for the MH trait, to know the early signs of a MH crisis, and to be prepared to promptly and efficiently treat a malignant hyperthermia event. The goal of this course is to increase that knowledge to promote early recognition of the MH trait/MH susceptible patient and implementation of appropriate interventions.

Course Objectives

At the completion of this learning activity the learner will be able to:

- Describe the physiological mechanisms involved in the development of malignant hyperthermia (MH) syndrome.
- Discuss who is at risk for developing MH.
- Describe testing for potential MH.
- Identify the key symptoms of MH, particularly early symptoms of its development.
- Identify treatment interventions in MH crisis.
- Discuss operating room (OR) team roles.

About the Author

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Ms. Weyer received her bachelor's and master's degrees from the SUNY Institute of Technology in Utica/Rome. She has worked in the educational field for several years and is currently an educator with the Samaritan Medical Center in Watertown, NY. She has worked in the perioperative arena for ten years. During this time she has in-serviced staff on malignant hyperthermia on a periodic basis and has developed a self-learning module (SLM), updating information as needed.

Updates were completed in August 2010 by **Barbara Fane, RN, MS, APRN-BC.** Mrs. Fane received her bachelor's degree, Master of Science in Nursing, and post-graduate certificate for Adult Nurse Practitioner from the Sage Colleges in Troy, NY. With over 25 years in nursing, Mrs. Fane's experiences include faculty positions in baccalaureate nursing programs, staff orientation and development, as well as direct care in critical care and cardiology.

In March 2012, a new course was developed reflecting the most recent evidence in practice by **Halya Hebert, MS, RN**. Mrs. Hebert is employed as an Associate Director in the Education, Practice, and Research Program at the New York State Nurses Association, in Latham, NY.

This course originates from the e-leaRN™ course, *Malignant Hyperthermia: Is Your Surgical Patient at Risk?,* originally written by **Sandra J. Weyer, MS, RN, CS, ANP**.

The authors declare they have no vested interest.

Definitions

Malignant hyperthermia - a hypermetabolic syndrome (chain of reaction of symptoms) triggered in genetically susceptible individuals when certain potent inhalation anesthetics or succinylcholine are administered. The symptoms include a greatly increased body metabolism, muscle rigidity and an eventual high fever that may exceed 110 degrees F. Death or brain damage may result from cardiac arrest, internal hemorrhaging, or failure of other body systems.

A **malignant hyperthermia-susceptible (MHS) person** is someone identified has having risk factors for MH.

Triggering agents - all volatile inhalation anesthetics, including: sevoflurane, desflurane, isoflurane, halothane, enflurane, methoxyflurane and the muscle relaxant succinylcholine. Nitrous oxide is NOT a triggering agent.

MH trait - genetic link, probably the 19th gene, autosomal dominant, variable entrance (may skip a person).

Masseter muscle – muscle in the jaw; origin is on the zygomatic arch (cheekbone) and inserts to the mandible (jawbone).

History of Malignant Hyperthermia

In 1960, two Australian physicians, Denborough and Lovell, first described malignant hyperthermia. In their case study, a healthy 21-year-old male, injured in a motor vehicle accident, was to undergo a repair of compound fractures of the tibia and fibula. The young man gave a positive history of 10 close relatives who died during or shortly after anesthesia. The family attributed these deaths to the administration of ether. The patient agreed to general anesthesia, as long as ether was not used. The case was done with Halothane as the anesthetic of choice. Ten minutes after the surgery started, the patient developed signs and symptoms of what the two physicians later termed MH. He became tachycardic, hypotensive, febrile, diaphoretic and cyanotic. The anesthetic was stopped and the surgery concluded in 10 minutes. The patient was cooled with ice and given a blood transfusion. The patient remained deeply unconscious for 30 minutes after the anesthetic was stopped. He gradually recuperated over the next 1.5 hours (Stolworthy & Haas, 1998).

Once this case study was reported, other reports of this syndrome began to appear in the literature. The name “malignant hyperthermia” was coined. Anesthesia providers continued to develop an increased awareness. Yet, the mortality rate was then reported to be 80% (Stolworthy & Haas,1998).

In the late 1960s, it was noted that Porcine Stress Syndrome was related to MH. A syndrome that included very high body temperatures was occurring in swine, particularly in the Poland China Pig. This pig was known to drop dead during rooting and when autopsies were done, it was found the meat of the pig was cooked (from high body temperatures). This led to researchers isolating the 19th gene as the MH trait.

It was later identified that alterations in skeletal muscle were the primary defect in MH. Research led to the use of Dantrolene sodium to relax the muscle rigidity characteristic of MH. In 1979, the FDA approved the use of Dantrolene sodium in humans for the treatment of malignant hyperthermia. The use of this drug had decreased the mortality rate of MH to 10% (Stolworthy & Haas, 1998) and with most recent advancements and patient education this rate has dropped to 5-6% (Rosenberg, 2010).

In 1981, the Malignant Hyperthermia Association of the United States (MHAUS) was established. Its goal was to bring together both professionals and patients concerned with this condition. The North American Malignant Hyperthermia Registry, established in 1987, had as its goal to centralize, analyze and disseminate clinical and research information about MH to healthcare professionals and researchers. In 1995, these two organizations merged in order to consolidate the information and services.

For further information about this organization and their services go to: http://www.mhaus.org.

Incidence and Prevalence of Malignant Hyperthermia

The Malignant Hyperthermia Association of the United States (MHAUS) estimates MH to be as frequent as one in 3,000 or as rare as one in 65,000 administrations of general anesthesia with triggering agents. The incidence varies depending on the concentration of MH families in a given geographical area. High incidence areas in the United States have been identified as Wisconsin, West Virginia and Michigan. The incidence in New York State is estimated at 0.96 per 100,000 surgical discharges, with males slightly higher than females (Brady, Sun, Rosenberg, & Li, 2009).

The prevalence of MH is greater in children, although the reason for this is not fully understood. It is estimated to occur in one of 15,000 anesthetic administrations to children; and in one of 20,000 adults. Fifty-two percent of all MH cases are in children under age 15. Boys and girls are affected equally prior to puberty. There is a 50% chance from either parent carrying the gene for a child to inherit it (Brady et al., 2009).

Pathophysiology of Malignant Hyperthermia

Malignant hyperthermia is inherited as an autosomal dominant disorder that involves skeletal muscle calcium regulation, where exposure to specific triggering stimuli results in intracellular hypercalcemia. Disordered calcium regulation by the sarcoplasmic reticulum of the skeletal muscle occurs, causing a cascade of symptoms. The collective myofibrils in skeletal muscle are covered by the sarcoplasmic reticulum, a netlike sheath, which contains the majority of the calcium needed for muscular activity. During physical activity, muscular contraction is produced by the exchange of calcium between the sarcoplasmic reticulum and the cytoplasm and other vesicular structures of the muscle cell itself. Ryanodine receptors (RyR1) help to move calcium through its channels (Martin, 2009). Muscle contraction occurs and is sustained by the active transport and the calcium concentration of the intracellular compartment of the skeletal muscle is high. The muscle relaxes when calcium returns to the reticulum.

During MH crisis, a triggering agent causes the uncontrolled release of calcium into skeletal muscle cells. Because the calcium cannot be pumped back to the sarcoplasmic reticulum, it enters a state of sustained contraction and hypermetabolism. This consumes large amounts of energy and oxygen, resulting in heat production (hyperthermia). Body temperature can rise from 0.5 - 2.0 degrees C every 5 to 10 minutes. Once skeletal muscle has contracted, and remains contracted in response to a triggering agent, several things occur:

- Oxygen consumption is very high, causing carbon dioxide levels to increase, stimulating rapid, deep breathing (if the patient is not under the influence of a neuromuscular blockade);
- End tidal carbon dioxide ($ETCO_2$) increases, making it difficult for the anesthesiologist to give sufficient ventilation to the patient;
- Hypermetabolism causes a rise in body temperature and development of metabolic acidosis;
- Prolonged hypermetabolism causes rhabdomyolysis, the breakdown of muscle at the cellular level, which causes release of free myoglobin into the blood. This accumulates like sludge in major organs, particularly in the renal tubules, causing decreased urinary output and renal failure;
- Muscular damage leads to creatine kinase (CK) release into the blood, in proportion to the degree of muscular damage;
- An outpouring of potassium from the cell occurs, causing high levels of extracellular potassium affecting heart rhythm and requiring treatment;
- Serious end-organ damage may occur when the brain is deprived of oxygen;
- Untreated MH will lead to death.

Triggering Agents

The specific anesthetic agent used during a surgical procedure can make the difference in safety and outcomes.

Malignant Hyperthermia Trigger Agents
All volatile inhalation anesthetics: • Isoflurane • Desflurane • Halothane • Enflurane • Sevoflurane • Diethyl ether • Cyclopropane • Methoxyflurane
Depolarizing muscle relaxants: • Succinylcholine chloride • Decamethonium

Non-triggering anesthetic agents can be utilized while closing a surgical procedure during an emergency if MH occurs.

Non-Triggering Agents for Malignant Hyperthermia
Inhalation agents: • Nitrous oxide
Barbiturates: • Methohexital sodium • Thiopental sodium
Benzodiazepines: • Diazepam • Lorazepam • Midazolam hydrochloride
Intravenous induction agents: • Etomidate • Ketamine hydrochloride • Porpofol
Neuromuscular blocking agents non-depolarizing: • Atracurium besylate • Doxacurium chloride • Mivacurium chloride • Pancuronium bromide • Pipecuronium bromide • Rocuronium bromide • Vecuronium bromide
Narcotic and opioid analgesia: • Fentanyl citrate • Hydromorphone hydrochloride • Meperidine hydrochloride • Morphine sulfate • Sufentanil citrate • Alfentanil hydrochloride
Local anesthetic agents: • Lidocaine hydrochloride • Bupivacaine hydrochloride • Chloroprocaine hydrochloride • Tetracaine hydrochloride

Fortunato-Phillips (2000)

Nursing Assessment

Preoperative Assessment

All patients about to undergo general anesthesia should be asked specific questions regarding their personal and family medical history to determine any indication of the MH trait. These questions should include:

1. Is there a personal or family history of malignant hyperthermia or any anesthetic problems?
2. Is there a family or personal history of a muscle or neuromuscular disorder (e.g., muscle weakness)?
3. Have there been any unexpected deaths or complications arising from anesthesia (including the dental office) with any family members?
4. Is there a personal history of unexplained high fever following surgery?
5. Is there a personal history of muscle rigidity (especially jaw) during surgery?

(Fortunato-Phillips, 2000)

Testing for Malignant Hyperthermia

Early diagnosis and treatment of MH are absolute prerequisites for a satisfactory outcome. Identifying MH in affected families allows for the selection of safe anesthetics prior to administration.

The **caffeine/halothane contracture test**, or CHCT, is a test performed on biopsied muscle and is considered the "gold standard" for diagnosis of MH (Kwetny, 2010). This test can only be performed in approximately 40 treatment centers, eight of which are located in the United States, three in Canada. Use the following link to view a list of North American testing centers: http://www.mhaus.org/malignant-hyperthermia-testing/testing-centers/#.T8j5z1IbbK0.

The patient is required to travel to one of these sites because the test must be completed rapidly after muscle is removed from the patient. This test is usually reserved for patients with a positive family history and/or has had a previous suspicious reaction to anesthesia. The test costs could range from $2,500 up to $6,000 and are partly covered by some insurance companies in the U.S.

The following are indications for muscle biopsy testing:

Definite indications:

- Suspicious clinical history for MH.
- Family history of MH.
- Severe masseter muscle rigidity.

Possible indications:

- Unexplained rhabdomyolysis during or after surgery (may present as sudden cardiac arrest due to hyperkalemia).
- Moderate to mild masseter muscle rigidity with evidence of rhabdomyolysis.
- Exercise-induced rhabdomyolysis.

Probably not indicated:

- Sudden, unexpected cardiac arrest during anesthesia or early post-operative period not associated with rhabdomyolysis.
- Most centers do not perform biopsy on patients less than approximately 40 lbs. or five years of age.
- Neuroleptic malignant syndrome.

Approximately 2 grams of skeletal muscle (less than 1/10 of an ounce) is removed through a 2-3 inch incision, usually from the vastus lateralis or vastus medialis muscles of the thigh. This procedure is usually done on an outpatient basis under local, spinal, epidural or non-triggering general anesthesia; it must be performed within 5 hours after muscle harvest. The rate and tension at which the muscle contracts after exposure to halothane and/or caffeine in the lab is then recorded electronically. The results of this are then compared to the strength of contractions with previously established standards. Muscle from MH susceptible patients is more sensitive and contracts with greater force than normal muscle.

Roughly 10% will return a false positive. Although the CHCT is still considered the gold standard, other less invasive blood tests are being studied (Kwetny, 2010).

Postoperatively, patients should avoid strenuous activities, including work, for at least three days. There is a minimal chance of postoperative bleeding, infection, or numbness.

Intraoperative Assessment

Early signs of MH include:

- Generalized erythematous flush, skin feels warm, core temperature normal.
- Unanticipated increase end tidal CO_2 ($ETCO_2$).
- Doubling or tripling of exhaled carbon dioxide.
- Tachypnea and tachycardia may be initial signs; secondary to hypermetabolism and hypercarbia.
- May occur over a brief period of time or may develop over 10-20 minutes.

Most sensitive signs of MH:

- Masseter muscle rigidity (MMR) on intubation following administration of succinylcholine.
- Intubation is difficult (should raise a red flag - 20% will develop clinical MH).
- Children experience MMR to greater degree than adults.
- If anesthesia is continued with use of triggering agents, signs and symptoms of MH occur in about 20 minutes in approximately 50% of cases.

Late signs of MH:

- Skin becomes mottled, followed by cyanosis.
- Elevated temperature (may increase 1 degree C or 1.8 degrees F every 5 minutes; may reach as high as 110 degrees F).
- Decreased platelet count.
- Unstable hypotension.
- Left ventricular failure with signs of pulmonary edema.
- Hyperkalemia.

- Hypoxia.
- Elevated creatine phosphokinase (CPK) liver enzymes, lactic acid, and magnesium.
- Nausea, vomiting, fever without rigidity.
- Diaphoresis.
- Dark blood, hot skin and tissues at wound site.
- Generalized muscle rigidity - most specific sign.
- Red or brown urine, followed by oliguria or anuria.
- Metabolic and respiratory acidosis.
- Falling serum calcium levels.

Treatment/Interventions

Prior to the introduction of **Dantrolene** (dantrium sodium) in 1979, the mortality rate for MH was 80%. Patients were treated for their symptoms by cooling instead of identifying the underlying cause. With Dantrolene and extensive education, the mortality rate has decreased to approximately 5%.

Dantrolene is a muscle relaxant that works by reducing the release of calcium from skeletal muscle sarcoplasmic reticulum, counteracting the abnormal intracellular calcium levels associated with MH. It does not work at the neuromuscular junction, as do standard neuromuscular blocking drugs, such as pancuronium.

***Should not be used with calcium channel blockers (may produce life threatening hyperkalemia and myocardial depression).**

***May cause significant muscle weakness in patients with pre-existing muscle disease and should be used with extreme caution in those patients.**

Dantrolene is supplied in vials containing 20 mg of Dantrolene powder with 3 g of Mannitol and sodium hydroxide.

Dantrolene must be reconstituted with 60 ml sterile water for injection USP (without a bacteriostatic agent) before it is administered. Mixing may take several minutes and may require extra personnel.

36 vials of Dantrolene should be kept in the OR at all times.

Dantrolene is administered via IV as quickly as possible. Dosing begins at 2.5 mg/kg. Sometimes more than 10 mg/kg (up to 30 mg/kg) is necessary. Administration is continued until signs of MH abate.

Tissue necrosis may result if extravasation of IV Dantrolene occurs. Side effects include skeletal muscle weakness, drowsiness, dizziness, blurred vision, and generalized weakness.

<u>Emergency Therapy for Malignant Hyperthermia</u>

The Malignant Hyperthermia Association of the United States offers a poster, "Emergency Therapy for Malignant Hyperthermia" (MHAUS, 2008), which should be posted in every OR/surgical suite. In addition, there are now new transfer of care guidelines for patients at risk for MH that the Association of periOperative Registered Nurses (AORN) has published.

Late signs of MH:

- Increased $ETCO_2$.
- Trunk or limb rigidity.
- Masseter spasm or trismus.
- Tachycardia/tachypnea.
- Acidosis.
- Increased temperature (late sign).

Sudden/unexpected cardiac arrest in young patients:

- Presume hyperkalemia and initiate treatment (see #6 below, Acute Phase Treatment).
- Measure CK, myoglobin,arterial blood gases (ABG) until normalized.
- Consider Dantrolene.
- Usually secondary to occult myopathy (e.g., muscular dystrophy).
- Resuscitation may be difficult and prolonged.

Trismus or masseter muscle spasm with succinylcholine:

- Early sign of MH in many patients.
- If limb muscle rigidity, begin treatment with Dantrolene.
- For emergent procedures, continue with non-triggering agents; consider Dantrolene.
- Follow CK and urine myoglobin for at least 36 hours.
- Observe in intensive care unit (ICU) for at least 12 hours.

Acute Phase Treatment

1. **GET HELP. GET DANTROLENE.**

   Notify Surgeon

   - Discontinue volatile agents and succinylcholine.
   - Hyperventilate with 100% oxygen at flows of 10 L/min or more.
   - Halt the procedure as soon as possible; if emergent, use non-triggers.

2. **Dantrolene** 2.5 mg/kg rapidly IV

   - Repeat until there is control of the signs of MH.
   - Sometimes more than 10 mg/kg (up to 30 mg/kg) is necessary.
   - Dissolve the 20 mg in each vial with at least 60 ml sterile preservative-free water for injection.
   - The crystals also contain NaOH for a pH of 9, Mannitol 3 g.

3. **Bicarbonate** for metabolic acidosis

   - 1-2 mEq/kg if blood gas values are not yet available.

4. **Cool** the patient with core temperature 39 degrees C via cold saline IV. Lavage open body cavities, stomach, bladder, or rectum. Apply ice to surface. Stop cooling if temperature less than 38 degrees C and falling to prevent drift less than 36 degrees C.

5. **Dysrhythmias** usually respond to treatment of acidosis and hyperkalemia.

   - Use standard drug therapy **except calcium channel blockers which may cause hyperkalemia or cardiac arrest in the presence of Dantrolene sodium.**

6. **Hyperkalemia** - Treat with hyperventilation, bicarbonate, glucose/insulin, calcium.

   - 10 units regular insulin and 50 ml 50% glucose **(adult)**

   or

   - 0.15 units insulin/kg and 1 ml/kg 50% glucose **(pediatric)**.
   - Calcium chloride 10 mg/kg or calcium gluconate 10-50 mg/kg for life-threatening hyperkalemia.

7. **Follow** $ETCO_2$, electrolytes, blood gases, CK, core temperature, urine output and color, coagulation studies.

   - Venous blood gas (e.g., femoral vein) values may document hypermetabolism better than arterial values.
   - Central venous or pulmonary artery (PA) monitoring as needed.

Post-Acute Phase

1. Observe the patient in an ICU for at least 24 hours, due to the risk of reoccurrence. Pain management should begin before the patient is admitted to ICU.

2. Dantrolene 1 mg/kg every 4-6 hours for at least 36 hours. Further doses may be indicated.

3. Follow labs as above (see #7 above, Acute Phase Treatment) with:

    - Frequent ABGs.
    - CK every 6-8 hours.

4. Counsel the patient and family regarding MH and further precautions. Refer them to MHAUS, fill out and send in the Adverse Metabolic Reaction to Anesthesia (AMRA) form and send a letter to the patient and her/his physician.

5. Refer patient to the nearest biopsy center for follow-up.

    (MHAUS, 2008)

CAUTION: This protocol may not apply to all patients; alter for specific needs. MH MAY OCCUR WITHIN 24 HOURS AND CAN RECUR WITHIN 24 HOURS!

The Malignant Hyperthermia Emergency Cart/Equipment

Being prepared to intervene immediately when a MH crisis occurs is essential. It is part of the nurse's responsibility to ensure that needed supplies are on hand. The OR and post-anesthesia care unit (PACU) must have an emergency cart readily available that accommodates all necessary supplies, medications and forms.

The Malignant Hyperthermia Association of the United States recommends the following:

Drugs

- Dantrolene – 36 vials should be available in each institution where MH can occur, each to be diluted at the time of use with 60 ml sterile water for injection USP (without a bacteriostatic agent).
- Sterile water for injection USP (without a bacteriostatic agent): each vial of Dantrolene should be reconstituted by adding 60 ml of sterile water for injection USP (without a bacteriostatic agent) and the vial shaken until the solution is clear. If the MH episode is proceeding rapidly, simply mix and inject. It is mandatory to get Dantrolene to its effective site, the skeletal muscle. The sterile water for reconstitution must be stored in 100 ml vials, not bags, to avoid accidental IV administration of this hypotonic solution.
- Sodium bicarbonate (8.4%) – 50 ml x 5;
- Furosemide 40 mg/amp x 4 ampules;
- Dextrose 50% – 50 ml vials x 2;
- Calcium chloride (10%) 10 ml vial x 2;
- Regular insulin 100 units/ml x 1 (refrigerated);
- Lidocaine* for injection, 100 mg/5 ml or 100 mg/10 ml in preloaded syringes (3). Amiodarone is also acceptable. ACLS protocols, as prescribed by the AHA, would be followed when treating all cardiac derangements caused by MH.

* Lidocaine or procainamide should not be given if a wide-QRS complex arrhythmia is likely due to hyperkalemia; this may result in asystole.

General Equipment

- Syringes (60 ml x 5) to dilute Dantrolene;
- Mini-spike IV additive dispensing pins x 2 and multi-administration fluid transfer sets x 2 (to reconstitute Dantrolene);
- Intravenous catheters 16G, 18G, 20G, 2-inch; 22G, 1-inch; 24G, 3/4-inch (4 each) (for IV access and arterial line);
- Nasogastric (NG) tubes;
- Toomy irrigation syringes (60 ml x 2) with adapter for NG irrigation;
- Micro drip IV set (x 1).

Monitoring Equipment

- Esophageal or other core (e.g., nasopharyngeal, tympanic membrane, rectal, bladder, pulmonary artery catheter) temperature probes;
- Central venous pressure (CVP) kits (sizes appropriate to your patient population);
- Transducer kits for arterial and central venous cannulation.

Nursing Supplies

- A minimum of 3,000 ml of refrigerated cold saline solution for IV cooling;
- Large sterile Steri-Drape (for rapid drape of wound);
- Urine meter x 1;
- Irrigation tray with piston (60cc irrigation) syringe;
- Large clear plastic bags for ice x 4;
- Small plastic bags for ice x 4;
- Bucket for ice;
- Test strips for urine analysis.

Laboratory Testing Supplies

- Syringes (3 ml) for blood gas analysis or ABG kits x6;
- Blood specimen tubes (each test should have 2 pediatric & 2 large tubes): (A) for CK, myoglobin, SMA 19 (LDH, electrolytes, thyroid studies); (B) for PT/PTT, fibrinogen, fibrin split products; and lactate; (C) CBC, platelets; (D) blood gas syringe (lactic acid level). If no immediate laboratory analysis is available, samples should be kept on ice for later analysis. This may well prove useful on retrospective review and diagnosis. Blood cultures are very useful and should be included to rule out bacteremia.
- Urine collection container for myoglobin level. Pigmenturia (e.g , brown or red urine and heme positive dipstick) indicates that renal protection is mandated, when the urine is centrifuged or allowed to settled, and the sample shows clear supernatant, i.e., the coloration is due to red cells in the sample.

(MHAUS, 2010)

Prevention and Patient Education for MH Susceptible Patients

- Screen patients preoperatively;
- Avoid the use of MH-triggering anesthetics with local or regional anesthesia;
- Be familiar with the signs and symptoms, as well as treatment of MH;
- MH cart or kit in OR with Dantrolene for future OR procedures;
- Observe patient for 4 hours following ambulatory surgery prior to discharge;
- Patient to wear band and inform healthcare provider of susceptibility.

(Martin, 2009)

Surgical Team Roles in MH: Who Does What?

The entire perioperative team plays a key role in the pre-screening for risk, early identification of symptoms and signs, as well as prompt and appropriate treatment for the patient at risk for MH.

*Preoperative/Preanesthesia Nurse***:**

Careful screening, as previously indicated, is critical. Preanesthesia assessment should include the patient and the patient's family. Information about how the patient responded to previous surgery or anesthesia must be obtained; the same information must be obtained regarding the patient's family. Has the patient or anyone in the patient's family had a history of an MH episode, a positive muscle biopsy, history of muscle disorders, history of dark-colored urine after anesthesia, history of unexplained fever or muscle rigidity during surgery, history of any family member who had an unexpected death during surgery and anesthesia. Any positive response raises a red flag and must be carefully investigated.

A physical examination is indicated after the history is taken. It is important to note:

- Extraocular muscle abnormalitites (ex. - ptosis or strabismus);
- Spinal deformities (ex. – scoliosis or pigeon breast);
- Congenital hernias;
- Clubfoot;
- Frequent joint dislocations.

Circulating Nurse:

The circulating nurse must prepare the OR for any suspected or known MH susceptible patient. Supplies and equipment must be readily available, such as fully stocked MH cart, ice, cold intravenous fluids, hypothermia blanket and other emergency supplies.

Anesthesia Provider:

Information obtained from the preanesthesia assessment is used to provide a safe course of anesthesia for the MH susceptible patient. Non-triggering MH agents must be utilized. It is standard of practice for anesthesia to flush anesthesia machinery to remove residual gases that are triggering agents (Wappler, 2010).

During the surgical procedure, the anesthesia provider must scrupulously monitor the patient for any early signs of MH. This includes:

- End-tidal carbon dioxide concentration;
- Arterial oxyhemoglobin saturation;
- Cardiac status;
- Core temperature.

Because an MH crisis is a true emergency, careful monitoring and assessment of all patients, whether or not they are MH susceptible, is necessary.

PACU Nurse:

Although MH is more likely to occur in the OR, it can occur within the first hour and up to 24 hours postoperatively. The PACU nurse must be familiar with the early as well as late signs of MH. It may be the PACU nurse who first alerts the surgical team of the onset of MH.

If the MH crisis occurred in the OR, or if it occurred but is now resolving in the PACU, the PACU nurse will continue to manage the MH patient. This will include the nursing interventions of:

- Any alterations in cardiac output;
- Altered thermoregulation;
- Altered ventilation;
- Altered fluid and electrolytes.

Any patient that has had a MH episode in the OR or in the PACU will be admitted to the intensive care unit for 24 to 48 hours for further observation, assessment and treatment (Kaplow, 2010).

<u>If the MH Crisis Occurs Intraoperatively</u>

- Anesthesiologist generally identifies and announces the impending MH crisis, and takes charge of the OR.
- All inhalation anesthetics and depolarizing muscle relaxants are immediately stopped and the patient is hyperventilated with 100% oxygen.
- Timekeeper is designated.
- Surgeon is instructed to close as expeditiously as possible and remains in the room as part of the team.
- The staff assigned to the room remains; the sterile team should remain sterile.
- The OR desk is notified that there is an emergency and asks for stand-by help that should remain outside the room until help is requested.
- MH protocol by MHAUS is followed (MHAUS, 2008).

Evaluation

Documentation: After the crisis, it is critical that the nurse fully document the situation. Important elements to include are:

- Patient responses;
- Interventions;
- Times;
- Personnel involved;
- Patient outcomes.

Issues in Litigation: When adverse outcomes have led to lawsuits, the following are cited:

- Failure to monitor temperature intraoperatively;
- Failure to elicit family history of MH;
- Failure to stop anesthetic and begin prompt treatment;
- Failure to defer surgical procedure.

Key Elements to Nursing Liability: Nurses, as key members of the operative team, are responsible for providing the care that meets the standards applicable to this situation. When lawsuits have occurred, nurses have been liable for the following omissions:

- Assessment of crisis situation;
- Protection of patient - nursing interventions;
- Provision of needed supplies and equipment;
- Planning, coordination and communication with receiving nursing staff.

Conclusion

The steps to successful management of malignant hyperthermia are:

- Be knowledgeable about MH;
- Be prepared for it;
- Know what signs to look for;
- Know your role if and when it occurs.

OR and PACU nurses should participate in educational programs related to MH on a periodic basis to gain familiarity with the treatment of MH, including supplies needed, knowledge of how to reconstitute Dantrolene and an understanding of the sequence of events during an MH crisis.

While MH is a rare entity, it can kill in minutes if not properly recognized and managed promptly. Bringing an episode to a successful conclusion depends on each and every member of the operating team (Martin, 2009).

Resources

American Society of PeriAnesthesia Nurses (ASPAN)
90 Frontage Road
Cherry Hill, NJ 08034-1424
Phone: 877.737.9696
Fax: 856.616.9601
Email: aspan@aspan.org
website: www.aspan.org

Association of periOperative Registered Nurses (AORN)
2170 South Parker Road
Suite 400
Denver, CO 80231
Phone: 1-800-755-2676 (303-755-6300)
Fax: 800-847-0045
email: custsvc@aorn.org
website: www.aorn.org

Malignant Hyperthermia Association of the United States (MHAUS)
39 East State Street
PO Box 1069
Sherburne, NY 13460-1069
Phone: 800-986-4287 (607-674-7901)
Fax: 607-674-7910
email: mhaus@norwich.net
website: www.mhaus.org

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Malignant Hyperthermia: Recognition and Treatment

Course Exam

After studying the downloaded course and completing the exam, you need to enter your exam answers ONLINE; answers cannot be answered and graded on this downloadable version of the course. To enter your answers, return to e-leaRN's website, **www.elearnonline.net** and click on the Login/My Account button. Next, login using your username and password, follow the prompts to access the course material, and proceed to the course exam.

Note: Contact hours will be awarded for this online course until **May 31, 2015**.

1. Malignant hyperthermia is defined as hypermetabolic syndrome in genetically susceptible individuals triggered by:

    A. Barbiturates, succinylcholine
    B. Lidocaine, propofol
    C. Inhalation agents, succinylcholine
    D. Neuromuscular blocking agents, nitrous oxide

2. With the introduction of Dantrolene in 1979, and rigorous preoperative screening, the mortality rate of malignant hyperthermia has decreased to:

    A. 5%
    B. 15%
    C. 40%
    D. 80%

3. A preoperative history indicative of MH susceptibility may include all EXCEPT:

    A. Unexplained high fever following anesthesia
    B. Brother-in-law with MH trait
    C. Cola-colored urine following anesthesia
    D. Family member who had an MH crisis

4. The gold standard for testing for MH susceptibility is:

    A. Hyperthermia/sweat test
    B. Positive liver biopsy
    C. Caffeine/halothane contractor test
    D. Elevated CPK

5. Tommy Lane, 17, has come to your surgery center for his preoperative testing prior to his septoplasty surgery scheduled for next week. His previous surgical history was a tonsillectomy four years ago. When questioned about any complications during the surgery, mom remembers the anesthesiologist telling her something about difficulty putting in the “breathing tube” because Tommy’s jaw was stiff, but he went home and did fine. What early sign of MH is mom referring to?

    A. Lockjaw
    B. Masseter muscle rigidity
    C. Laryngospasm
    D. Increased end tidal CO_2

6. Dantrolene must be dissolved in:

    A. 60 cc sterile water for injection, with a bacteriostatic agent
    B. 60 cc saline with a bacteriostatic agent
    C. 60 cc saline without a bacteriostatic agent
    D. 60 cc sterile water for injection, without a bacteriostatic agent

7. A late sign of MH is an elevated temperature reaching as high as 110 degrees F. At what rate would you expect it to rise?

    A. 1.8 degrees F every 5 minutes
    B. 1 degree F every 15 minutes
    C. 2.6 degrees F every hour
    D. 2.8 degrees F every 30 minutes

8. Who is in charge of OR during an MH crisis?

    A. Surgeon
    B. Circulating nurse
    C. Anesthesiologist
    D. Charge nurse

9. MH may reoccur in 24 hours.

    A. True
    B. False

10. As the manager of an OR/surgical center, how can you best prepare your staff for an MH crisis?

    A. In-service presentations with a mock MH crisis
    B. Post policies in the OR manual, as MH episodes are rare
    C. Send staff to Sherburne to tour MHAUS facilities
    D. Nothing since no mention of JCAHO is the good decision