Malignant Hyperthermia

This lesson will take 15 minutes to complete.

Dignity Health

Objectives:

- Identify the causes of Malignant Hyperthermia (MH)
- Recognize the warning signs and symptoms of MH
- List common triggers and predisposing factors for MH
- Identify the steps to reverse an MH incident
- Discuss the resources available for reference

Malignant Hyperthermia

- Overview:
  - Malignant Hyperthermia (MH) is a potentially lethal syndrome caused by a hypermetabolic state that can be precipitated by the administration of volatile inhalation anesthetic agents and depolarizing agents, such as Succinylcholine (Anectine). The triggering agent causes an increase in intracellular calcium ion concentration. This elevated calcium level produces a chain of reactions.
  - During an MH crisis, calcium levels in the contractual part of the muscle become excessive, causing sustained muscle contractures. This causes increased oxygen consumption and increases the production of carbon dioxide and heat (a hypermetabolic state). If not quickly diagnosed and treated, the resulting ATP depletion, acidosis, and hyperthermia cause a large release of potassium, myoglobin and creatinine kinase (CK) into the extracellular fluid which results in dysrhythmias, DIC and renal failure.
  - Both a genetic predisposition and one or more triggering agents are necessary to evoke MH.

Malignant Hyperthermia

- Overview, cont.
  - The exact incidence of MH is unknown. The rate of occurrence has been estimated to be as frequent as one in 5,000 or as rare as one in 65,000 administrations of general anesthesia with triggering agents.
  - Malignant Hyperthermia occurs more frequently in children than in adults but occurs equally among boys and girls. In adults, it occurs more often in males. Susceptibility to MH is thought to be inherited as an autosomal dominant trait – therefore it requires only one affected parent for a child to inherit the condition.
  - It is believed that patients with pre-existing neuromuscular or skeletal diseases, including central core disease and muscular dystrophy, are at increased risk.
  - The mortality rate for malignant hyperthermia has decreased since 1960 from 80% to 6-7% today, primarily because of increased education, early diagnosis, and prompt treatment with dantrolene.

Malignant Hyperthermia

- Overview, cont.
  - Over 80 genetic defects have been associated with MH. MH susceptibility is inherited with an autosomal dominant inheritance pattern. This means that children and siblings of a patient with MH susceptibility usually have a 50% chance of inheriting a gene defect for MH and hence would also be MH susceptible. They, therefore, may develop an MH reaction upon exposure to triggers.
  - Those who are carriers for susceptibility may be completely unaware of this risk unless they or a family member developed a life-threatening crisis during anesthesia. It is important to know that not everyone who has a gene defect linked to MH develops the MH crisis upon each exposure to the triggering anesthetics.

Preventing Malignant Hyperthermia

The best treatment for malignant hyperthermia is prevention. A thorough assessment and evaluation of patients is necessary before anesthetics are given. This assessment should include:

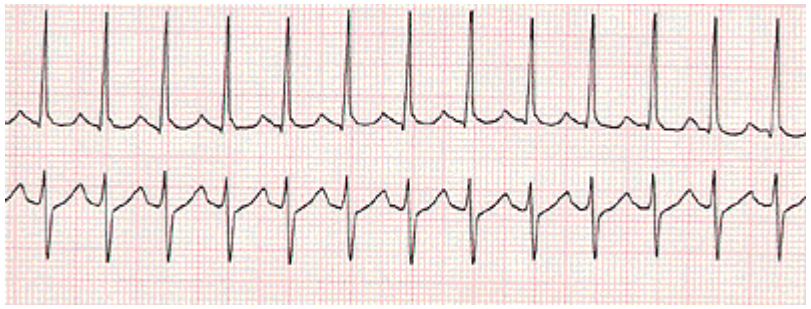
- History of past anesthetics and responses to them
- Family history of malignant hyperthermia
- Family history of anesthetic complications
- History of muscle disorders
- Specific screening questions for malignant hyperthermia are part of the pre-surgical assessment.
  - Typically, malignant hyperthermia is triggered by:
    - volatile anesthetics such as Halothane, Isoflurane or Desflurane
    - depolarizing muscle relaxants such as Succinylcholine.
    - Patients may have undergone many courses of anesthetics/surgery using these products with no complications, yet still have a malignant hyperthermia crisis.

Malignant Hyperthermia

- Cause:
  - MH-susceptible persons have a mutation that results in the presence of abnormal proteins in the muscle cells of their body. Although normal in everyday life, when these patients are exposed to certain anesthetic agents, it causes an abnormal release of calcium inside the muscle cell, which results in a sustained muscle contraction and the abnormal increase in energy utilization and heat production.
  - The muscle cells eventually run out of energy, and die, and release large amounts of potassium into the bloodstream, which can lead to heart rhythm abnormalities.
  - The muscle pigment myoglobin is also released and may be toxic to the kidney. Left untreated, these changes can cause cardiac arrest, kidney failure, blood coagulation problems, internal hemorrhage, brain injury, liver failure, and may be fatal.

Identifying a Malignant Hyperthermia Crisis

- Early diagnosis of malignant hyperthermia is essential for treatment to be effective. Untreated, malignant hyperthermia has an 80% mortality rate. Unexplained tachycardia and an increase in end-tidal CO2 may be the first indicators of an impending crisis. Tachycardia is seen in 96% of patients experiencing an MH crisis and generally occurs within 30 minutes of induction of anesthesia. Increased end-tidal CO2 is often the earliest indicator.



Malignant Hyperthermia

- The general signs of the MH crisis that occur shortly after the induction of anesthesia include:
  - Tachycardia
  - A greatly increased body metabolism (↑ end tidal CO2)
  - Masseter (jaw) muscle rigidity
  - Total body rigidity
  - Dark blood in the surgical field
  - Arterial desaturation
  - Fever that may exceed 108 degrees F (late sign).
  - Severe complications include: cardiac arrest, brain damage, internal bleeding or failure of other body systems. Thus, death, primarily due to a secondary cardiovascular collapse, can result.

Identifying a Malignant Hyperthermia Crisis - Continued

The following lab results could be expected in an MH crisis:

- Metabolic/Respiratory acidosis
- Hyperkalemia
- Hypercalcemia
- Hyperphosphatemia
- Creatine kinase levels > 1000IU
- Myoglobinurea
- MHAUS has a 24/7 “hotline” to assist anesthesiologists in managing a Malignant Hyperthermia crisis.
- 1 (800) 644-9739 or 1 800-MH HYPER
- It is recommended that a staff member contact the hotline in a crisis.

Malignant Hyperthermia

- What Drugs Trigger MH?
  - The volatile gaseous inhalation anesthetics are MH triggers:
    - Sevoflurane
    - Desflurane
    - Isoflurane
    - Halothane
    - Enflurane
    - Methoxyflurane
    - Succinylcholine (Anectine), the depolarizing muscle relaxant

Treatment of a Malignant Hyperthermia Crisis

- Once malignant hyperthermia is suspected, the focus of the entire surgical team must transfer from the surgery to managing the crisis.
  - Staff should immediately request assistance
  - Stop the anesthetic**.** All triggering agents must be discontinued and the patient should be hyperventilated with 100% oxygen at high flow
  - Rapidly administer an initial bolus of IV Dantrolene sodium (2-3mg/kg at CRMC & MGMC) (2.5mg/kg at STHMC & SJWMC). Dantrolene is difficult to mix and each vial needs to be mixed with 60cc sterile preservative free water for injection. **5% Dextrose, 0.9% Sodium Chloride injection, and other acid solutions are not compatible with Dantrium Sodium**
  - Call the Pharmacy
  - Supplemental increments up to 10mg/kg total should be given until the signs of MH are controlled but clinical reassessment is suggested
  - Onset of action is 2-3 minutes.
  - Each vial contains 20mg Dantrolene and 3g Mannitol
  - The surgery/procedure should be terminated as quickly as possible
  - A central line and arterial line should be initiated if possible but with no delay in administering specific interventions.

Treatment of a Malignant Hyperthermia Crisis

- Give Bicarbonate - Patients produce large amounts of lactic acid in a MH crisis. Treat acidosis with Bicarbonate, if not promptly reversed by Dantrolene.
- Cool the patient - Give cold IV fluids, externally cool patient with ice and cold saline. Irrigate the wound, stomach, and bladder with cold saline (insert 3-way catheter for bladder irrigation). Monitor core temperature of patient.
- Treat dysrhythmias - Patients in an acidotic state produce dysrhythmias
- Monitor end-tidal CO2 - One of the best indicators of MH. This will help determine when the patient is improving.
- Ensure urine output - Myoglobin from blood cell breakdown is toxic to the kidneys, urine output should be watched carefully.
- Measure - Creatinine Phosphokinase, myoglobin, and ABG's until normalized.

Treatment of a Malignant Hyperthermia Crisis

- Treat hyperkalemia - with glucose, insulin, and calcium
- Cease cooling efforts - when temperature has fallen to 38°C.
- Watch for reappearance of symptoms by appropriate monitoring in an ICU for 24 hours. Recrudescence occurs in about 25% of MH cases. Core temperatures should be monitored throughout.
- Ensure urine output - of at least 2 ml/kg/hr by hydration and diuretics.
- Avoid IV potassium - during ongoing rhabdomyolysis (breakdown of muscle tissue).
- Follow coagulation profile - DIC may occur. Measure CK's every 6 hours until decreased.
- Continue monitoring - Observe the patient in ICU for at least 24 hours

Treatment of a Malignant Hyperthermia Crisis

- Post-Crisis Care
  - Administer Dantrolene 1 mg/kg every 4-6 hours for 24 hours after the episode. Dantorlene may then be given orally if ordered by physician
  - Follow vitals and labs (ETCO2, blood gases, CK)
  - Continue monitoring urine output and color
  - Monitor temperature
  - Support patient as there may be muscle weakness following Dantrolene therapy
- Counsel the patient and family about malignant hyperthermia and further precautions. Refer the patient to MHAUS. www.mhaus.org. 1-800-986-4287
- Fill out an Adverse Metabolic Reaction to Anesthesia (AMRA) report, available through the North America Malignant Hyperthermia Registry (1 717-531-6936

Malignant Hyperthermia

- Preoperative planning for the known or suspected MH susceptible patient:
  - Do not use MH-triggering agents on patients susceptible to MH or their undiagnosed relatives
  - Ensure that anesthetic vaporizers are disabled by removing or taping in the “off” position.
  - Obtain an accurate & thorough pre-operative history and assessment
  - Use a clean, vapor-free anesthetic machine.
  - Flow 10L/min. O2 through circuit via the ventilator for at least 20 minutes (if fresh gas hose is replaced, 10 minutes is adequate).
  - Use a new or disposable unused breathing circuit and bag.
  - Monitor body temperature for all patients undergoing general anesthesia
  - End-tidal CO2 monitoring for all patients undergoing general anesthesia.
  - Dantrolene available where trigger anesthesia agents are used.

Malignant Hyperthermia

- Safe anesthetic drugs include:
  - Local anesthetics
  - Nitrous Oxide
  - Barbiturates
  - Narcotics
  - Propofol
  - Benzodiazopines
  - Ketamine
  - Etomidate
- Safe non-depolarizing muscle relaxants include:
  - Pancuronium
  - Cisatracurium
  - Atracurium
  - Mivacurium
  - Vecuronium
  - Rocuronium

Malignant Hyperthermia

- Preoperative Planning:
  - Obtain an accurate & thorough pre-operative history and assessment
  - Use a clean, vapor-free anesthetic machine.
  - Monitor body temperature for all patients undergoing general anesthesia
  - End-tidal CO2 monitoring for all patients undergoing general anesthesia.
  - Dantrolene available where trigger anesthesia agents are used.
- In the past, Dantrolene sodium was given to pretreat MH susceptible patients. Currently, prophylactic treatment with Dantrolene is not recommended because patients have experienced muscle weakness and gastrointestinal upset. It also does not influence the likelihood of MH if safe techniques are used.
- Communication should occur between the entire surgical team regarding the patient's possible MH susceptibility.

Malignant Hyperthermia

- The MH Cart is checked monthly by the unit staff for outdated drugs and supplies. Pharmacy replaces the drug tray. The MH monthly drug and supply forms are attached to the policy. The cart should not be changed/removed from the unit while cases are in progress.
- Report patients who have had acute MH episodes to MH Registry of MHAUS
- For consultation to help with patient management, call the MH Hotline
- For a complete list of drugs and supplies, refer to the Malignant Hyperthermia policy.

Malignant Hyperthermia

- Is There A Test For MH?
  - There is currently no simple diagnostic test available for screening the general population (e.g., a blood test). Genetic testing is available, which is useful in identifying some patients with MH. Since not all of the genes responsible for MH have been identified, the genetic tests are only useful in families with a suspicious history of MH. Even if the genetic tests do not show that you have one of the genes for MH, you may still have other unidentified genes and therefore may be at risk for MH.
  - The Caffeine Halothane Contracture Test (CHCT), a test performed on freshly biopsied muscle, is the "gold standard" for diagnosis of MH. It can be performed only in roughly 30 centers worldwide, eight of which are located in the United States and Canada. The patient must travel to one of these sites for the test because the test must be completed within hours after muscle is removed.

References

- Dignity Health. Malignant Hyperthermia, Guidelines for the Treatment of. Policy #6100-M-01
- Dignity Health. Management of Malignant Hyperthermia. Policy # 600.039
- Malignant Hyperthermia Death Holds Many Lessons , Henry Rosenberg, MD and Al Rothstein
- Malignant Hyperthermia Association
  of the United States (MHAUS)
  11 E State St, PO Box 1069
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  607-674-7901 or 800-98MHAUS
  Web site: www.mhaus.org

Thank You

You have completed the information portion of the lesson.

You must pass the post-test to successfully complete this activity.

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