HealthStream ASCA Regulatory Script

Malignant Hyperthermia

Release Date: November 2019
HLC non-PA Version: 1
HLC PA Version: 1
HLC CE Version: 1

Lesson 1: About Malignant Hyperthermia

Malignant hyperthermia (MH) is a rare pharmacogenetic disorder that can cause multi-organ failure and death if it is not recognized and quickly treated. Some individuals who have the genetic disorder may not know that they are at risk, so patient screening and team preparation for an MH crisis are critical in providing safe care and preventing mortality.

Upon completing this course, you will be able to:

- Identify individuals who may be at risk for malignant hyperthermia
- List triggering agents that can precipitate an MH reaction
- Describe changes that occur during an MH episode.
- Describe the necessary preparation for a possible MH episode
- Describe the response efforts necessary to resuscitate and care for someone during and after an MH event

As your partner, HealthStream strives to provide its customers with excellence in regulatory learning solutions. As new guidelines are continually issued by regulatory agencies, we work to update courses, as needed, in a timely manner. Since responsibility for complying with new guidelines remains with your organization, HealthStream encourages you to routinely check all relevant regulatory agencies directly for the latest updates for clinical/organizational guidelines.

If you have concerns about any aspect of the safety or quality of patient care in your organization, be aware that you may report these concerns directly to your organization's accrediting agency.

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Course Goals	
Lesson 1 will introduce malignant hyperthermia and provide background information on the disorder. Lesson 2 describes risk factors for MH. Lesson 3 reviews signs and symptoms of MH. Lesson 4 describes the necessary emergency treatment and transfer considerations. Finally, lesson 5 provides the necessary components of team preparation.	Lesson 1: About Malignant Hyperthermia • Genetic factors • Precipitating agents • Incidence • Mortality Lesson 2: Individuals at Risk • Screening considerations • Diagnostic testing • Testing sites Lesson 3: Signs and Symptoms of Malignant Hyperthermia • Early signs • Later signs Lesson 4: Treatment • Dantrolene • Team roles during an MH crisis • Post crisis monitoring • Patient transfer process • Post-event responsibilities • Providing for the patient who has malignant hyperthermia Lesson 5: Team Preparation and Personal Responsibility
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Competency Considerations

Have you ever worked with a patient who experienced malignant hyperthermia? It is such a rare occurrence that many clinicians have not.

What if you learned during the pre-surgical interview that a patient was at risk? Worse yet, what if a patient didn't know he or she was at risk, and symptoms suddenly appeared after anesthesia induction or later, during post-anesthesia care?

Would you be able to identify MH and intervene quickly and appropriately?

Are you prepared?

What is Malignant Hyperthermia?

Malignant hyperthermia is a rare, autosomal dominant inherited disorder of the skeletal muscles.

This muscle abnormality is caused by one of several genetic mutations in the muscle cells that cause the cells to release stored calcium ions when exposed to certain anesthetics. The resulting increase in calcium ion concentration inside the muscle cells stimulates a hypermetabolic reaction that causes muscle fibers to contract. Muscle rigidity is characteristic in individuals experiencing MH.

The reaction may be fatal if it is not recognized and treated immediately.

Precipitating Pharmacologic Agents

A malignant hyperthermia reaction can happen when volatile anesthetic gases or the depolarizing skeletal muscle relaxant succinylcholine are used.

The agents known to potentially cause an MH crisis include these inhaled general anesthetics:

- Desflurane
- Enflurane
- Isoflurane
- Halothane
- Sevoflurane
- Methoxyflurane
- Ether

Succinylcholine may trigger an MH event and risk increases when the drug is used in combination with volatile anesthetics.

Many individuals who carry the gene for MH may not experience symptoms after initial exposures to these anesthetics, making it difficult to predict whether a reaction will happen with a subsequent exposure.

Care Settings Where MH May Occur	

MH can occur anywhere where volatile anesthetics and succinylcholine are used. Care settings include:
• The hospital
  ○ Operating room suite
  ○ Emergency room
  ○ Interventional radiology
• An ambulatory surgical center (ASC)
• A surgeon's office
• An office where oral surgery is performed
 	
MH cases have also occurred in intensive care units in sedated patients.
 	
MH can occur during minor or major surgical procedures. It has occurred in individuals who have undergone:
• Tonsillectomy
• Appendectomy
• Plastic surgery
• Oral surgery
• Organ donation
• Other procedures or surgeries
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Review	
Select the answer that best fits the question. Which of the following statements is true? a. MH is an allergy. b. MH only occurs with major surgeries. c. Some individuals do not know that they have MH. d. A patient with flaccid muscles is experiencing MH.	Correct: C Rationale: Some individuals who have malignant hyperthermia may not know that they are at risk, so *screening* and *preparation* for an MH crisis are critical to providing safe care and preventing mortality. MH *is not* an allergy. It is a genetic disorder, brought on by certain volatile anesthetics and succinylcholine. Patients experience *muscle rigidity* among other symptoms related to a hypermetabolic state.
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Incidence of MH

The exact incidence of malignant hyperthermia is unknown because:

- It is not reported consistently.
- Many times, people with MH can undergo anesthesia without any crisis, before a subsequent surgery reveals it.

Studies reveal that MH occurs in about 1 in 100,000 surgeries in adults and about 1 in 30,000 in children. Individuals under 19 years of age account for about half of all reported MH cases.

The disorder is more common in males than in females. Incidence varies with geographical location, where the gene pool for this disorder is higher. These states include Wisconsin, Nebraska, West Virginia, and Michigan.

It is estimated that about 1 in 2,000 individuals harbor a genetic change that makes them susceptible.

The annual number of MH cases in the U.S. is about 700.

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Mortality

In recent decades, the mortality of malignant hyperthermia has dramatically decreased from 70-80% to less than 5%.

This is largely due to these important factors: the introduction in 1979 of dantrolene sodium for the treatment of MH; the use of capnography for early detection of an MH episode; and the introduction of diagnostic testing for malignant hyperthermia susceptibility (MHS).

Emergency preparedness and consistent, thorough screening contribute to decreased mortality as well.

- **Dantrolene**
- **Capnography for early detection of MH**
- **Diagnostic testing**
- **MH preparedness**
- **Screening**

Lesson 2: Individuals at Risk

Many individuals who carry the gene for malignant hyperthermia may not experience symptoms after initial exposures to the list of volatile anesthetics and succinylcholine presented earlier. This makes it difficult to predict whether a reaction will happen with a subsequent exposure.

Certain groups have been identified as being at high risk for an MH crisis. The Malignant Hyperthermia Association of the United States (MHAUS) [glossary] recommends asking specific questions about a patient's medical history to prompt recognition of this potential problem.

Individuals at risk are discussed on the next few slides.

Individuals at Risk: A History of Complications

A History of Anesthetic Complications

A history of anesthetic complications, especially an unexplained personal or family history of perioperative fever may be suggestive of MH. People may not realize that a fever or other complications during a surgical procedure in a close relative might suggest MH in themselves.

Consider, for a moment, the true story of a man who shared that his sister died in 1947 from complications of an appendectomy. He also had a brother who died from complications after an auto accident. Prior to a needed surgical procedure for himself, his physicians suggested he be tested for MH and he was found to be positive. Because of this, the surgical team was prepared to safely manage the man, who had surgery without complications, unlike his siblings. Source: (MHAUS, 2018)

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Individuals at Risk: Family History of Malignant Hyperthermia

A Family History of Malignant Hyperthermia

MH is an autosomal dominant trait. If it is present in one parent, it may be passed down to their offspring.

- People who have a parent, sibling, or child with MH have a 50% chance of having MH.
- If aunts, uncles, or grandchildren have the condition, a person's chance of having MH is 25%.

Family members of people with MH must be informed and be sure to tell healthcare providers which of their relatives have MH, understanding that they themselves may be at risk.

In 1984, a 7-year-old boy needed ear tube surgery and adenoid removal. Shortly after surgery began, the child's temperature rose and his muscles became rigid. He was experiencing an MH crisis.

The boy had undergone two prior uneventful surgeries, at six months and at three years of age. He had been fortunate not to experience a crisis during either of those surgeries, because dantrolene had not yet been identified as a drug to treat MH.

During the boy's third surgery in 1984, the doctor and anesthesiologist recognized the symptoms of MH and quickly administered dantrolene, which saved the child's life.

His parents promptly informed family members of their risk, and they diligently inform their healthcare providers. The 7-year-old, who is now in his mid thirties and a father himself, will have his own son tested. (Olsen & Olsen, 2017)

Individuals at Risk: Skeletal Muscle Disorders

Skeletal Muscle Disorders

Certain muscle disorders are known to be associated with malignant hyperthermia. These skeletal muscle abnormalities include:

- King-Denborough syndrome [glossary]
- Central core disease [glossary], also known as central core myopathy
- Multiminicore disease [glossary], also known as multiminicore myopathy

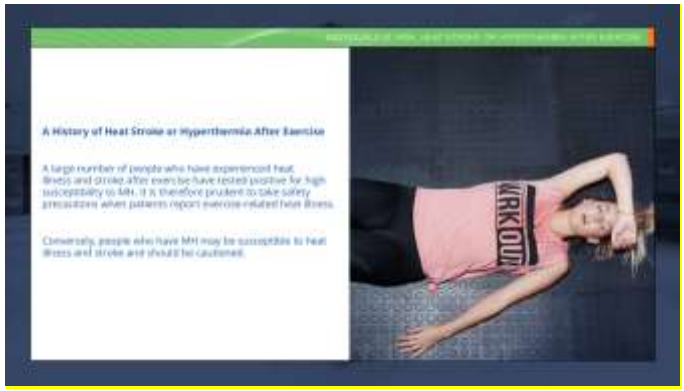
Duchenne muscular dystrophy [glossary] is probably not associated with MH, but it is noteworthy because succinylcholine may precipitate hyperkalemic cardiac arrest in these individuals. They also have anesthesia-induced rhabdomyolysis, but do not experience the hypermetabolism associated with MH. (Chapin, 2018) Individuals with any form of myotonia (muscle weakness) *should not* receive succinylcholine. (Rosenberg, 2010)

Individuals at Risk: Heat Stroke or Hyperthermia After Exercise

A History of Heat Stroke or Hyperthermia After Exercise

A large number of people who have experienced heat illness and stroke after exercise have tested positive for high susceptibility to MH. It is therefore prudent to take safety precautions when patients report exercise-related heat illness.

Conversely, people who have MH may be susceptible to heat illness and stroke and should be cautioned.



Individuals at Risk: Diagnostic Testing	
Patients identified as at risk for malignant hyperthermia should be tested if there is a strong suspicion that an individual is MH susceptible. This includes a review of family history for possible MH. The caffeine halothane contracture test (CHCT) is the standard test used. Although costly, it is approved by most insurance companies. In this test, a muscle sample is obtained, usually from the thigh. The muscle is tested for its contractile response to caffeine or halothane. If the results are positive, the patient has MH.	NO IMAGE
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Individuals at Risk: Testing Sites

Because the muscle sample must be tested immediately upon extraction, the patient must travel to one of the available testing locations.

Testing is available at only five centers in North America:

- Toronto General Hospital in Toronto, Ontario
- Uniformed Services University of the Health Sciences in Bethesda, Maryland
- University of California, Davis, California
- University of Minnesota, Minneapolis, Minnesota
- Wake Forest University in Winston-Salem, North Carolina

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Individuals at Risk: Diagnostic Testing

Genetic testing is also an option. The DNA is isolated from a patient sample and mutations of the RYR1 gene are identified. There are over 30 causative mutations and the presence of any one of them is diagnostic for malignant hyperthermia susceptibility.

Once a causative mutation is found, family members can be tested for that specific causative mutation and the individual can avoid the pain and discomfort of a muscle biopsy.

Absence of a causative mutation does not rule out MH, however, and a CHCT would be needed to confirm that an individual is susceptible.

Health insurance may not cover genetic testing.

Detailed information on testing sites for both the CHCT and genetic testing is available on the MHAUS website and includes contact individuals and phone numbers.

Lesson 3: Signs and Symptoms of Malignant Hyperthermia

Reactions to volatile, inhaled anesthetics or succinylcholine may occur during surgery or shortly afterward, often occurring within an hour of induction.

Severe reactions can cause multi-organ failure and death if not identified and treated immediately.

Preparedness, attention, and excellent assessment skills can prevent complications and possible mortality.

MHAUS recommends continuous core temperature monitoring for all patients receiving general anesthesia for longer than 30 minutes, regardless of exposure to a triggering agent. Capnography is also recommended because an elevated CO_2 is one of the first signs of MH.

Signs and symptoms occur in order of the body's response to the agent and include both early and later signs.

Early Signs of Malignant Hyperthermia	

Early signs of malignant hyperthermia include:
• A rapid heart rate; ventricular arrythmia
• Tachypnea resulting from a rapid rise in CO_2—an attempt to reduce the rising CO_2
• Oxygen consumption is increased threefold due to oxygen demands of this hypermetabolic state, putting the patient at risk for myocardial distress
• **Increased end tidal CO_2 (>60 mmHg) *** causing respiratory and metabolic acidosis with a pH below 7
• Decreased oxygen saturation
• Blood lactate increases 15-20 times
• Masseter muscle rigidity (MMR) lasting longer than 1 minute
  ○ More common in children
  ○ Occurs after succinylcholine
• **Generalized muscle rigidity**
• Flushed skin, cyanosis, and mottling
 	
*Bolded symptoms are specific to MH
	Symptoms Specific to MH
	
	• Increased end tidal CO_2 (>60 mmHg)
	• Generalized muscle rigidity
	• A dramatic and rapid rise in body temperature
	• Rhabdomyolysis (elevated creatine kinase)

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Later Signs of Malignant Hyperthermia	

Later signs of malignant hyperthermia include:
• **A dramatic and *rapid* rise in body temperature** greater than 38.8° C (101.8° F) and may exceed 110° F (43.3° C), resulting from a hypermetabolic state and an attempt to reprocess the calcium and return it to the storage areas in the cell. Contracting muscles generate excessive heat.
• Sweating
• Brown or cola-colored urine (a sign of myoglobinuria)
• Confusion
• Muscle weakness and **rhabdomyolysis** (muscle breakdown) as evidenced by an elevated creatine kinase (CK) level and leading to:
  ○ Hyperkalemia
  ○ Increased myoglobin and resultant kidney damage
  ○ Edema
• Disseminated intravascular coagulation (DIC) because of thromboplastin release
• Multiple organ failure
• Death
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Rapid Treatment Prevents Complications

If MH is suspected, it must be treated rapidly to prevent complications. When treated, symptoms usually resolve within 12-24 hours and the patient can recover completely.

After the event, the patient should be monitored in the intensive care unit. If the MH crisis occurs in a non-hospital setting, such as an ambulatory surgery center, the patient will have to be transferred, once stable.

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Review	
Select the answer that best fits the question. To be prepared for any MH crisis, MHAUS recommends which of the following interventions? a. Continuous core temperature monitoring for all patients receiving general anesthesia for longer than 30 minutes b. Capnography c. Asking specific questions about one's medical history that might identify susceptibility to MH d. That all patients get a CHCT prior to any surgery e. A, B, and C f. All of the above	Correct: E Rationale: The Malignant Hyperthermia Association of the United States (MHAUS) recommends the following: continuous core temperature monitoring for anesthesia lasting longer than 30 minutes; capnography, since a high CO_2 is one of the specific signs of MH; and taking a thorough history asking specific questions related to MH susceptibility. CHCT is recommended only if there is a strong suspicion of MH. This test confirms susceptibility to MH.
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Dantrolene

Dantrolene is the only FDA-approved treatment for malignant hyperthermia crisis. The drug inhibits the release of calcium within the cells and reverses the process. Initially approved in 1979, dantrolene is now available in two formulations (20 mg/mL or 250 mg/mL). Check your state regulations and your organization's policies and procedures to learn which formulation is used.

The likelihood of significant MH complications doubles for every 30-minute delay in dantrolene administration.

MHAUS recommends that *any facility* that stocks triggering agents or succinylcholine should have dantrolene on hand, even if those agents are not routinely used. A stock supply should be available within 10 minutes when MH is identified. The sooner it is used, the better the outcome.

Practice Standards

An ASC should follow federal and state regulations as well as the standards of their accrediting organization related to the identification of signs and symptoms of MH and preparation of appropriate drugs and equipment available to treat it.

The American Association of Nurse Anesthetists recommends that an MH cart or kit be available in any facility where triggering agents or succinylcholine are used.

Recommended MH cart supplies and drugs are listed on the MHAUS website, along with very useful training tools, guidance documents, and posters to help staff review and remember.

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Stock Dantrolene Recommendations

The Centers for Medicare and Medicaid Services (CMS) regulations specify that an ASC is expected to maintain a comprehensive, current, and appropriate set of emergency equipment, supplies, and medications that meet current standards of practice and that are needed to respond to a patient emergency in the ASC.

States also have specific regulations regarding the emergency equipment, supplies, and medications an ASC must have readily available.

The amount of dantrolene that is recommended to be kept readily available includes:

- 36 vials of Dantrium® and Revonto®, 20 mg/vial, each to be mixed with 60 mL of preservative-free sterile water

OR

- 3 vials of Ryanodex®, 250 mg/vial, each to be mixed with 5 mL of preservative-free sterile water.

Once reconstituted, dantrolene must be used within 6 hours since preservative-free diluents are used.

Agency Protocols Direct the Team

Your organization's policies and procedures should allow for multiple staff to perform interventions in synchrony to ensure reversal of malignant hyperthermia symptoms and to avoid possible complications. Depending on your organizational policies and procedures, this may require:

- Calling for additional nursing help
- Calling for a code or rapid response team
- Getting the MH cart and code cart
- Calling the MHAUS hotline at 1-800-644-9737

The MHAUS toll-free number provides guidance from experienced staff who are on call around the clock, every day of the year.

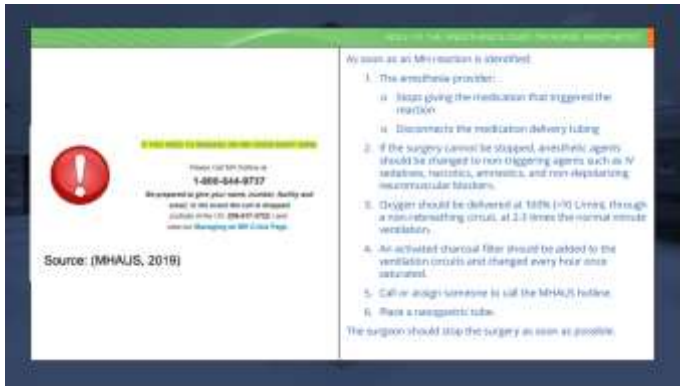
Critical interventions and staff roles are reviewed on the next few slides.

Role of the Anesthesiologist or Nurse Anesthetist

As soon as an MH reaction is identified:

1. The anesthesia provider:
   - Stops giving the medication that triggered the reaction
   - Disconnects the medication delivery tubing
2. If the surgery cannot be stopped, anesthetic agents should be changed to non-triggering agents such as IV sedatives, narcotics, amnestics, and non-depolarizing neuromuscular blockers.
3. Oxygen should be delivered at 100% (>10 L/min), through a non-rebreathing circuit, at 2-3 times the normal minute ventilation.
4. An activated charcoal filter should be added to the ventilation circuits and changed every hour once saturated.
5. Call or assign someone to call the MHAUS hotline.
6. Place a nasogastric tube.

The surgeon should stop the surgery as soon as possible.



The Role of the Nurse

Nurses assume various roles during an MH crisis. Their responsibilities vary with each facility and should be detailed in your facility's policies and procedures.

1. One nurse, usually the circulating nurse, records interventions, prepares needed lab supplies, and draws labs as ordered. These include:
    - Arterial blood gases
    - Electrolyte panel
    - Complete blood count
    - Coagulation studies
    - Serum for creatine phosphokinase (CPK), myoglobin, BUN/creatinine, glucose and lactate, liver enzymes

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The Role of the Nurse	
2. Another nurse *or two*, prepares and labels dantrolene (Dantrium®, Revonto® or Ryanodex®) as ordered until the patient is stabilized: a. **Dantrium intravenous** should be administered by continuous rapid intravenous push beginning at a *minimum dose of 1 mg/kg* and continuing until symptoms subside or the maximum cumulative dose of 10 mg/kg has been reached. (Drugs.com, 2017) **MHAUS recommends an initial dose of 2.5 mg/kg IV push up to 10 mg/kg. Check your organization's policies and procedures to know the initial dosing.** **For the dantrolene product you are using, know where the dosing chart is located and consult it for guidance during reconstitution.** b. The effective dose to reverse the crisis is directly dependent on the individual's degree of susceptibility to malignant hyperthermia, the amount and time of exposure to the triggering agent, and the time elapsed between onset of the crisis and initiation of treatment. (Drugs.com, 2017)	The amount of dantrolene needed depends on: • The degree of MH susceptibility • The time of exposure to the triggering agent • The time between the onset of symptoms to initiation of treatment

The Role of the Nurse

3. A third nurse prepares and administers medications as ordered to control the heart rate and rhythm and to correct the acidosis and hyperkalemia. These medications include but may not be limited to:
    a. Sodium bicarbonate (1-2 mEq/kg) to correct acidosis
    b. Regular insulin (10 units) in 50 mL of 50% glucose, calcium chloride (10 mg/kg) to correct hyperkalemia
    c. Hydration and furosemide to induce diuresis and avoid myoglobin-induced renal failure
    d. Amiodarone or lidocaine to correct arrythmias (**do not use calcium channel blockers**)

The Role of the Nurse

4. A fourth nurse provides additional cooling measures and monitors the core temperature. Procedures may include the use of:
   - Ice packs to the neck, axillae, and groin
   - Cooling blankets
   - Cooled normal saline administered IV
   - Cool mist and fans
   - Cooled normal saline administered via peritoneal lavage, particularly if the abdomen has been surgically opened
   - Cooled normal saline also can be inserted into a Foley catheter, although the surface area of the bladder is small.

Recognize that the temperature can continue to decline after cooling measures stop, so stop cooling once the patient's temperature reaches 100.4º F (38º C).

According to MHAUS recommendations, cooling should never distract from dantrolene administration and hyperventilation interventions.

Post MH Crisis Care

After an MH crisis, call 911 for transport to an inpatient intensive care unit. The anesthesia provider will confirm stability prior to transfer. Some key indicators of stability include:

- $ETCO_2$ is declining or normal.
- Heart rate is stable or decreasing without dysrhythmia.
- IV dantrolene was started.
- Temperature is declining.
- Muscle rigidity is resolving.

About 25% of the time, symptoms will reoccur and can be deadly if not recognized.

IV dantrolene may be required for up to 36 hours at a dose of 1 mg/kg every 4-6 hours. Intermittent bolus is ideal if there is no central line because dantrolene can cause necrosis if IV infiltrates.

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Post MH Monitoring for Relapse and Complications	
Post event monitoring includes: • Vital signs including core temperature • ECG monitoring for arrythmia • $ETCO_2$ • Urine output and color • Arterial and central venous monitoring depending on severity • Oxygen saturation • Muscle tone • Labs for electrolytes, blood gases, CPK until decreasing, urine myoglobin • Liver function studies (SGOT, SGPT, alkaline phosphatase, total bilirubin) MH complications include: • Change in level of consciousness • Cardiac dysrhythmias • Pulmonary edema • Renal dysfunction and failure • DIC • Muscle weakness • Compartment syndrome (secondary to rhabdomyolysis)	NO IMAGE
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Post MH Crisis Transfer – Receiving Facility Capabilities

Per MHAUS recommendations, non-hospital centers such as ASCs should have agreements in place with an inpatient facility that can manage MH.

The receiving facility should have:

- An adult and pediatric intensive care unit
- Continuous core temperature monitoring ability
- Cardiopulmonary monitoring
- Invasive and non-invasive cooling measures
- Continuous sedation
- Dantrolene for maintenance and for a crisis*
  - 36 vials of Dantrium® or Revonto®
  - 3 vials of Ryanodex®
- Dysrhythmia treatment
- Hemodialysis

*State regulations will guide staff in the required product needed.

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Post MH Crisis Transfer – Transport Team Capabilities

Per MHAUS recommendations, the transport team must have what they need to manage the patient until arrival at the inpatient facility.

The transport team should have:

- Ventilation support
- Cardiopulmonary and temperature monitoring
- Fluid resuscitation
- Appropriate medications:
  - Dantrolene
  - Non-depolarizing muscle relaxants
  - Sedatives and hypnotics
  - Analgesics and opioids
  - Insulin, glucose, and calcium chloride
  - Life-support medications

The transport team must be able to communicate by phone with the MHAUS hotline and the identified receiving staff at the hospital.

Transitioning Care

Any time there is a transition of care, there is a risk of patients receiving fragmented care.

Miscommunication can occur as responsibilities for care are transferred from the ASC to the hospital staff. This often results in adverse events, emergency department (ED) visits, and often, rehospitalization.

Effective communication with members of the healthcare team is essential in ensuring a successful and safe transfer of care.

If a caregiver is present (and deemed appropriate), he or she should be apprised of the event in order to support communication with the receiving team.

Hand-off Communication to the Receiving Facility	

The **sender** *transfers* responsibility by:
• Providing prompt, complete, and accurate information to the receiver (911 personnel and hospital staff), including:
  ○ Baseline labs and vital signs
  ○ Details of the event including monitoring results
  ○ The response to treatment with dantrolene and other drugs
  ○ Current status including:
  ▪ The presence or absence of muscle rigidity
  ▪ The color of the urine
  ▪ The integrity of the IV
  ▪ When the next dose of dantrolene is due
• Verifying receipt by the receiver
• Clarifying information sent
• Verifying that medications, equipment, and services will be available in the new setting as soon as they are needed
• Documenting the transaction
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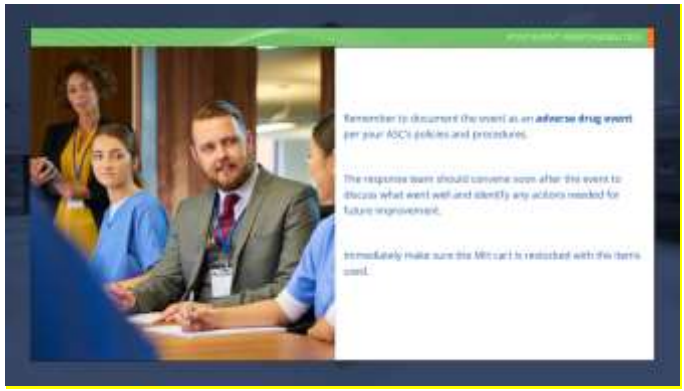
Post-Event Responsibilities

Remember to document the event as an **adverse drug event** per your ASC's policies and procedures.

The response team should convene soon after the event to discuss what went well and identify any actions needed for future improvement.

Immediately make sure the MH cart is restocked with the items used.



Helping with Data Collection	
The North American Malignant Hyperthermia Registry of MHAUS is a database that records detailed events surrounding MH episodes and correlates the clinical history along with genetic and CHCT results. Information can come from physicians or patients. The registry acts as a service to patients, families, and healthcare professionals to communicate and store important information relating to the risk of MH. Information about the registry, including a contact number, is available on the MHAUS website.	NO IMAGE
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Patient and Family Education and Counseling

Once the patient recovers, the hospital staff will have to teach the patient and family about malignant hyperthermia. ASC staff may have the opportunity to begin to educate the person who accompanies the patient on key information before the patient is transferred.

Important teaching points include the following:

- Emphasize the importance of informing providers about the MH episode prior to any future surgical procedure, including oral surgery or other outpatient procedures where volatile anesthetics or succinylcholine may be used.
- Explain that MH is related to an autosomal dominant trait, meaning that only one parent has to have the gene in order for it to be passed on.
- Provide them with the link to the MHAUS website and their phone number.
- Make sure that they are tested using the CHCT test at one of the four testing facilities in the United States.
- Encourage genetic testing to help identify the genetic mutation that caused the event.
- Stress the importance of wearing a medical alert bracelet if they are found to be susceptible to MH.
- Notify family members to help them identify their potential risk for MH.

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Managing the Patient Who Has Malignant Hyperthermia

By now you may be thinking, can a patient who has malignant hyperthermia ever have surgery and avoid an MH crisis?

Yes, they can, however certain preparation is required by various members of the team to ensure patient safety.

Some patients with a known or suspected personal or family history of MH have been denied surgery with general anesthesia prior to diagnostic testing for MH. Some are told that they can't have surgery in an ASC and that a hospital setting is their only choice.

Provided that non-triggering agents are used, surgery can be safely done in ASCs.

ASCs should review and follow their policies regarding patients with MH.

Safe Drugs for the Malignant Hyperthermia Patient	
There is a broad range of medications that are safe to use for a person with malignant hyperthermia. Some examples of anesthetics include: • Local anesthetics • Ketamine • Barbiturates • Nitrous oxide • Propofol (Diprivan) • Thiopental (Pentothal) • Etomidate • Narcotics • Benzodiazepines Examples of non-depolarizing muscle relaxants include: • Pancuronium • Cisatracurium • Atracurium • Mivacurium • Vecuronium • Rocuronium A full list of safe medications appears on the MHAUS website.	NO IMAGE
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Managing the Patient Who Has Malignant Hyperthermia

The anesthesia provider must make sure that residual anesthetic does not remain in the anesthesia machine that will be used for a patient who is MH-susceptible. There are three ways to do this, but always follow the manufacturer instructions.

1. Use a clean machine:
   - Use a dedicated vapor-free machine for MH patients.
   - Use an ICU ventilator that has never been exposed to volatile anesthetic agents.
2. Flush an existing machine, which may take 10 to 104 minutes:
   - Physically disconnect the vaporizer.
   - Use a new, disposable breathing circuit and reservoir bag.
   - Replace the carbon dioxide absorbent.
   - Keep the fresh gas flow at 10 L/min during the procedure.
3. Use in-line activated charcoal filters such as Vapor-Clean™:
   - These filters remove trace levels of volatile anesthetic agents following a 90-second flush.
   - Place them at both inspiratory and expiratory ports.

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Managing the Patient Who Has Malignant Hyperthermia

If a procedure is to be done in an ASC for a patient with MH susceptibility, the ASC must ensure the following:

- The staff is prepared to recognize and treat an MH crisis.
- Dantrolene is accessible within 10 minutes of the first signs of MH.
- The facility has the capacity to administer at least 10 mg/kg of dantrolene in the event of an acute MH episode requiring multiple dantrolene doses to abort the crisis.
- The anesthesia machine is flushed according to its specific manufacturer's recommendations and/or charcoal filters are placed on both inspiratory and expiratory limbs to minimize residual volatile agent in the circuit.
- There is a formal agreement in place between the ASC and hospitals for transfer of patients to a higher level of care after a suspected MH episode.

Source: (MHAUS, 2012)

Review	
Select the answer that best fits the question. The Centers for Disease Control and Prevention keeps track of patients who have MH. a. True b. False	Correct: B Rationale: The North American Malignant Hyperthermia Registry of MHAUS is a database that records detailed events surrounding MH episodes and correlates the clinical history along with genetic and CHCT results. Information about the registry is available on the MHAUS website.
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Lesson 5: Team Preparation and Personal Responsibility	

In order for an ASC to provide a surgical procedure to an MH patient, or to manage an unexpected MH crisis, the ASC staff must be prepared.
 	
To become competent in the management and care of a patient who is MH susceptible, be sure to do the following:
• Review your facility policies and procedures related to MH.
• Visit the MHAUS website to view their recommendations and articles of interest. Subscribe to their newsletter.
• Seek professional guidance by contacting your national professional organization and participating in their educational resources. Some reputable organizations include:
  ○ The American Association of periOperative Registered Nurses (AORN)
  ○ The American Association of Nurse Anesthetists (AANA)
 	
Ask your supervisor for assistance if you can't find the information that you need.
	NO IMAGE

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Staff Training, Drills and Competency Requirements

Ongoing training, drills, and competency assessment is critical for staff confidence and successful patient outcomes. Drills should include:

- Requirements for pre-operative screening
- Anesthetic selection including triggering and non-triggering agents
- MH cart review including dosing and how to prepare commonly used medications, including dantrolene. (Use expired drugs labeled “For Training Use Only.")
- Anesthesia work station preparation
- Signs and symptoms of MH
- Other treatments and management
- Documentation forms and tools
- Staff roles and responsibilities including handling multiple roles
- Transfer protocols and communication
- Components of patient and family education and counseling

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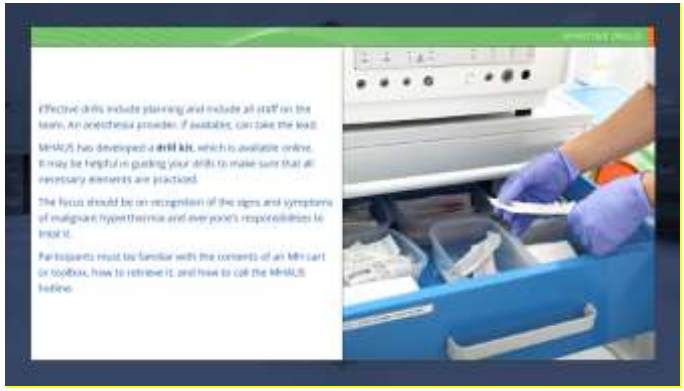
Effective Drills

Effective drills include planning and include all staff on the team. An anesthesia provider, if available, can take the lead.

MHAUS has developed a **drill kit**, which is available online. It may be helpful in guiding your drills to make sure that all necessary elements are practiced.

The focus should be on recognition of the signs and symptoms of malignant hyperthermia and everyone's responsibilities to treat it.

Participants must be familiar with the contents of an MH cart or toolbox, how to retrieve it, and how to call the MHAUS hotline.



Role Assignment

Roles should be assigned to prevent overlap and/or distraction from priority functions. In other words, not *everyone* needs to be focused on cooling when many other interventions must occur concurrently, such as:

- Stopping the trigger anesthesia
- Oxygenation
- Dantrolene preparation and administration
- Correction of the acidosis
- Lab draws

Staff must understand the priorities of treatment and work in synchrony to respond appropriately to an MH crisis.

Be sure to debrief after the drill to discuss what was done well and what needs improvement. Discuss how well the individuals functioned as a team and identify any gaps in knowledge or skill set. A competent team is confident and can skillfully manage a patient in crisis.

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Review	
Select the answer that best fits the question. A competent team is one that: a. Identifies and addresses gaps in knowledge and skills b. Seeks information from credible sources c. Practices needed skills d. Applies best practices when caring for patients e. All of the above	Correct: E Rationale: Competency requires identification of knowledge and skill gaps and taking appropriate action to fill those gaps. This includes seeking information from credible sources, practicing needed skills, and applying best practices.
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Summary	
You have completed the course on malignant hyperthermia (MH). During this course you have reviewed: • Who may be at risk for MH • Triggering agents that can precipitate an MH reaction • Changes that occur during an MH episode • The necessary preparation for a possible MH episode • The response efforts necessary to resuscitate and care for someone having an MH crisis	NO IMAGE
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Please remember that compliance is the responsibility of each organization. Provision of this list does not imply that the content of this course wholly or partially addresses the guidelines and references provided here.

Glossary

Term	Definition
Central core disease	A disorder in skeletal muscle cells due to a mutation in the RYR1 gene (the same gene associated with MH susceptibility) that changes the structure of the ryanodine receptor, allowing calcium ions to leak through an abnormal channel. This disruption in calcium ion transport leads to muscle weakness, which can range from slight weakness to very severe. Central core disease gets its name from disorganized areas called cores that are found in the center of muscle fibers in many affected individuals. (U.S. National Library of Medicin, 2019)
Duchenne muscular dystrophy	A group of genetic conditions characterized by progressive muscle weakness and wasting. Affecting the skeletal and heart muscles, it is found mostly in males. Individuals with this rapidly progressive disease may only live into their 20s.
King-Denborough syndrome	A congenital myopathy associated with susceptibility to malignant hyperthermia, skeletal muscle abnormalities, and dysmorphic features with characteristic facial features. Facial and proximal limb weakness may become more pronounced with increasing age. Thoracic kyphosis, lumbar lordosis, and scoliosis often develop as the condition progresses. Source: (GARD, 2013)
Malignant Hyperthermia Association of the United States (MHAUS)	MHAUS is a non-profit organization founded in 1981 to promote optimum care and scientific understanding of MH and related disorders
Multiminicore disease	A skeletal muscle disorder caused by a mutation of the RYR1 and SELENON genes, which leads to muscle weakness beginning in infancy and early childhood affecting the trunk and neck muscles. Infants appear floppy, which can delay the development of motor skills. Muscles used for breathing are affected and may cause severe and life-threatening respiratory problems and scoliosis, which worsens over time. (U.S. National Library of Medicine, 2019)

EXAM

1. Which of these medications does/do NOT trigger a malignant hyperthermia reaction? (Select all that apply.)

a. Methoxyflurane
b. Ether
c. Enflurane
d. Etomidate
e. Nitrous oxide

Correct: D and E
Rationale: Etomidate and nitrous oxide are both safe to use for patients who are susceptible to malignant hyperthermia.
Reference: Slide 6

2. For a patient weighing 156 lbs. (~70 kg), how many **vials** of Dantrium® (20 mg/vial) should be mixed for an **initial** dose of 2.5 mg/kg?
    a. 3 vials
    b. 7 vials
    c. 9 vials
    d. 35 vials

Correct: C
Rationale: Using the patient's weight in kilograms (~70), figure 2.5 mg x 70 = 175 mg / 20 = 8.75 (round to 9). Nine vials of Dantrium® should be mixed for the initial dose.
Reference: Slide 26

3. Which interventions or drugs are used during a malignant hyperthermia (MH) crisis to help correct arrythmias? (Select all that apply.)
    a. Calcium channel blockers such as amlodipine, diltiazem, and verapamil
    b. Insulin, glucose, and calcium chloride to correct the hyperkalemia
    c. Amiodarone and lidocaine
    d. Etomidate and propofol

Correct: B and C
Rationale: Calcium channel blockers are **not** used during a malignant hyperthermia (MH) crisis because they can cause more hyperkalemia and cardiac arrest when used in combination with dantrolene. Insulin, glucose, and calcium chloride correct the high potassium that can cause arrest. Etomidate and propofol are anesthetic agents and not antiarrhythmics.
Reference: Slide 31

4. Which symptoms listed may NOT be specific to MH?
    a. A rapid heart rate and tachypnea
    b. Generalized muscle rigidity
    c. Decreased oxygen saturation
    d. Increased end tidal CO_2 (>60 mmHg)

Correct: A and C
Rationale: Increased end tidal CO_2, generalized muscle rigidity, a rapid rise in body temperature, and rhabdomyolysis are signs and symptoms that indicate and are specific to MH. A rapid heart rate, tachypnea, and a decreased oxygen saturation may indicate MH, or they may indicate complications other than MH, so it is important to know characteristic signs among others.
Reference: Slide 20

5. During a preoperative interview, Mr. T. reports that he had an appendectomy as a teenager and a knee arthroplasty 2 years ago. He tells you that he never experienced any problems during or after either surgery. He mentions he was glad that he didn't because both of his siblings had problems with the anesthesia and ended up in intensive care. He arrived at the Ambulatory Surgery Center for repair of a ruptured plantar plate and bunionectomy. How should you proceed? (Select all that apply.)
    a. Ask more questions about the problems his two siblings experienced to find out more about their events.
    b. Mr. T. may be predisposed to an MH crisis, so notify the anesthesiologist and the surgeon.
    c. Proceed as usual since he stated that he had no complications during his prior surgeries.
    d. Review the signs and symptoms of MH and discuss the plan of care with your team.

Correct: A, B, and D
Rationale: Mr. T.'s siblings having had problems with anesthesia could indicate that they experienced MH. This is a cue to ask more questions. Since inheritance of MH susceptibility is related to a dominant gene, if Mr. T.'s *siblings* had an MH reaction, then Mr. T. is at risk for MH despite his two uneventful prior surgeries. More questioning about the experiences of Mr. T.'s siblings may help you identify whether those events could have been due to MH. Preparing for an MH event requires clinicians to review signs and symptoms and procedures to follow if it occurs. All team members should be aware.
Reference: Slide 12

6. The effective dose of dantrolene during an MH crisis depends upon which of the following factors? (Select all that apply.)
    a. The number of people in the patient's family who have MH
    b. The patient's degree of susceptibility to MH
    c. The amount and time of exposure to the triggering agent
    d. The time that elapsed between the onset of the crisis and the initiation of treatment

Correct: B, C, and D
Rationale: The effective dose to reverse the crisis is directly dependent upon the individual's degree of susceptibility to malignant hyperthermia, the amount and time of exposure to the triggering agent, and the time elapsed between onset of the crisis and initiation of treatment. (Drugs.com, 2017)
Reference: Slide 30

7. Which individual is more likely to have a malignant hyperthermia (MH) reaction during surgery?
    a. A teenager with Down syndrome
    b. A 50-year-old woman undergoing a hysterectomy
    c. A teenager with proximal limb weakness and kyphosis
    d. An infant undergoing surgery on her ears

Correct: C
Rationale: The teenager's proximal limb weakness and kyphosis is indicative of a chronic muscle disorder and may be a result of King-Denborough syndrome.
Reference: Slide14

8. Which medical history information may be indicative of malignant hyperthermia susceptibility? (Select all that apply.)
    a. The patient tells you of an aunt who has malignant hyperthermia.
    b. The patient tells you that he almost had heat stroke during a recent workout and the outside temperature was in the 50s.
    c. The patient tells you that his temperature dropped during his last surgery and that the OR team had to use warming blankets to bring it back up.
    d. The patient reports muscle weakness in his thigh after he broke his femur in a motorcycle accident.

Correct: A and B
Rationale: When a patient has an aunt or uncle who has malignant hyperthermia (MH), there is a 25% chance the patient has it, too. Heat intolerance after exercise is a sign that the patient might be susceptible to MH. A drop in temperature after surgery is not a sign of MH but an unexplainable rise in temperature would be. Muscle weakness after an injury is not associated with MH.
Reference: Slides 12-15

9. A triggering agent that causes masseter muscle rigidity (MMR) is:
    a. Isoflurane
    b. Halothane
    c. Sevoflurane
    d. Succinylcholine

Correct: D
Rationale: Succinylcholine is known to cause masseter muscle rigidity in patients. This is more common in children. If MMR lasts for longer than a minute, it is likely due to malignant hyperthermia and is followed by generalized muscle rigidity.
Reference: Slide 20

10. One of the first signs of malignant hyperthermia is:
    a. Increased end tidal CO_2 (>60 mmHg)
    b. A rapid rise in core temperature to 38.8° C (101.8° F)
    c. Brown-colored urine

d. An increase in serum potassium, creatine kinase, and myoglobin

Correct: A
Rationale: An increase in end tidal CO_2 (>60 mmHg) as monitored by capnography, is one of the very first signs that malignant hyperthermia may be occurring.
Reference: Slide 20