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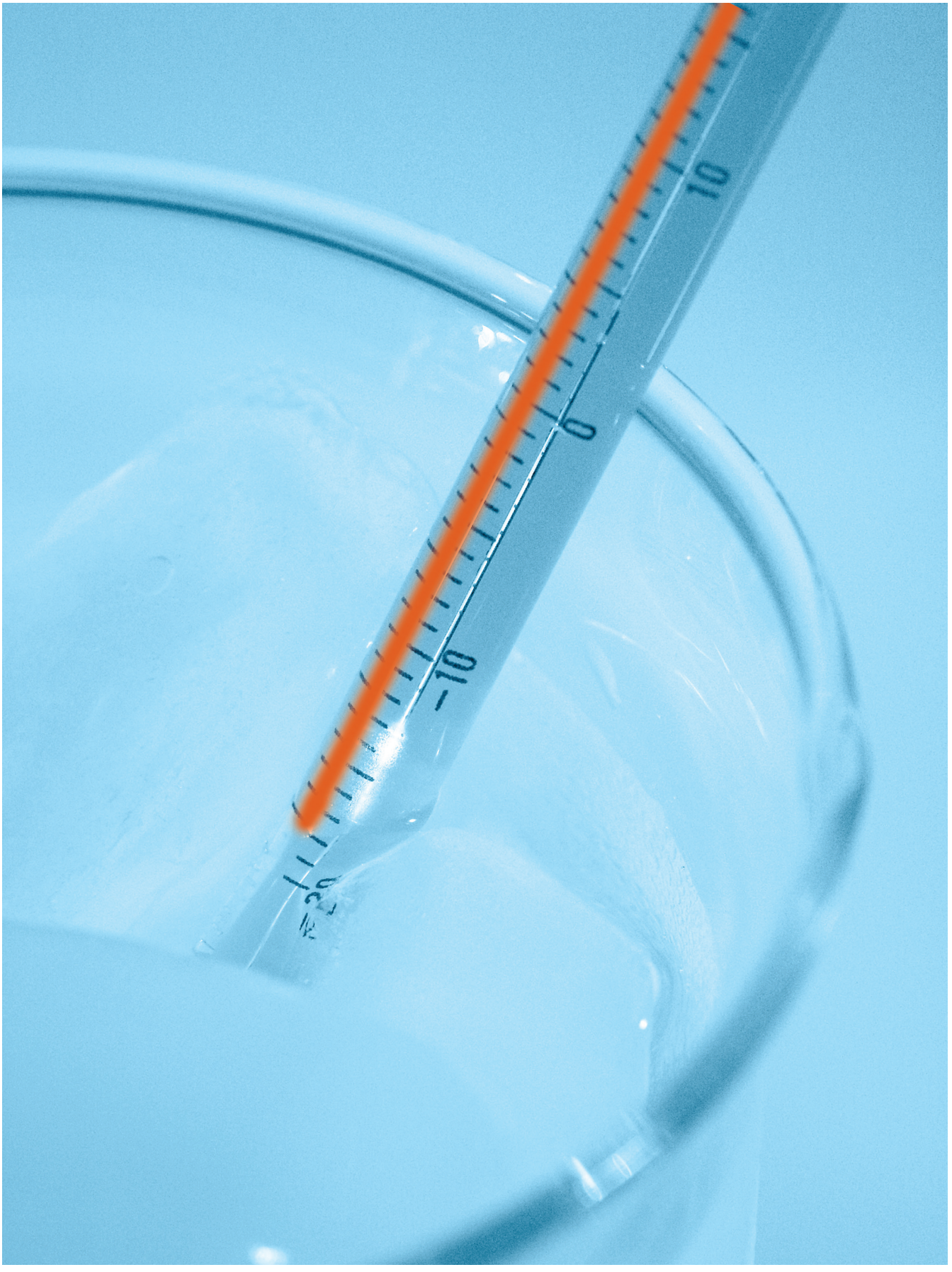
A practical guide for malignant hyperthermia management

By Connie L. Martin, CNOR , BA, MSN

Malignant hyperthermia (MH) is a hypermetabolic disorder of skeletal muscle and is commonly triggered by the administration of general anesthetics in susceptible individuals.[1] MH is a genetic, autosomal dominant, life-threatening disorder that requires immediate intervention to prevent death. Today, continuing education promoting greater awareness of MH symptoms among OR personnel has led to better diagnoses and defined treatment protocols. Though MH once had a fatality rate of 70%, prompt intervention and treatment with dantrolene (Dantrium) have reduced the rate to less than 5%.[2]

Pathophysiology

Calcium is an essential ion in normal cell energy production and muscle contraction. Normally, when a nerve impulse stimulates skeletal muscle, calcium is released from the sarcoplasmic reticulum, allowing muscle contraction to occur.[1,3] In cases of MH, the triggering agent disrupts normal calcium ion concentrations, which causes an increase in calcium uptake in skeletal muscle fibers. This results in an increase in oxygen consumption and an increase in lactate and heat production leading to a hypermetabolic state.[1,3]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Anesthetic agents that trigger MH[5]

According to the Malignant Hyperthermia Association of the United States (MHAUS), the following are examples of anesthetic agents that are known to trigger MH, and aren't considered safe for use in the MH-susceptible patient.

Unsafe inhaled general anesthetic agents:
- desflurane (Suprane)
- enflurane (Ethrane)
- halothane (Fluothane)
- isoflurane (Forane)
- sevoflurane (Ultane)
- xenon (rarely used)

Unsafe depolarizing muscle relaxant:
- succinylcholine (Anectine)

Identifying patients at risk

MH occurs in one in 5,000 to 50,000 individuals who receive general anesthesia.[4]

Known triggers of MH include inhaled general anesthetic agents and the depolarizing muscle relaxant succinylcholine (see *Anesthetic agents that trigger MH)*. Non-MH triggering agents that are safe for use in MH-susceptible patients include nitrous oxide, opioids, barbiturates, nondepolarizing muscle relaxants, local anesthetics, ketamine, propofol, and anxiolytics.[5] MH may also be triggered by emotional stress, heatstroke, strenuous exercise exertion, and trauma.[1]

MH risk assessments should be performed by the perioperative nurse during the routine preoperative interview. Interview questions are developed to screen patients for a history of MH or those who may be susceptible to MH. Ask the patient if he has a history of caffeine intolerance, unexplained fever, dark cola-colored urine, or history of muscle weakness, cramps, or muscle group hypertrophy. Ask the patient if any family member ever had a problem or died after receiving anesthesia during surgery. Patients with Duchenne's or Becker's muscular dystrophy, King-Denborough syndrome, central core disease, and myotonia are considered at higher risk for MH crisis.[1,3] The obstetrical population should be carefully screened as well, since dantrolene crosses the placental barrier and may cause weakness in the child upon delivery.[1] Since MH is an inherited disorder, it's important to determine MH susceptibility of the pregnant patient and the father of the child[6] (see *Management of the non-MH susceptible pregnant patient whose partner is MH susceptible)*.

Management of the non-MH susceptible pregnant patient whose partner is MH susceptible[6]

MHAUS recommends the following for the non-MH susceptible pregnant patient whose partner is MH susceptible:
- Whenever possible, the father's MH susceptibility should be confirmed by review of the medical record to determine if genetic mutation is present.
- If the pregnant woman requires nonemergent surgery during the pregnancy, nontriggering anesthetic agents or epidural or spinal anesthesia should be used.
- Nitrous oxide is the only inhalation agent that may be used during surgery. However, during delivery, the mother may receive a volatile inhalation agent after delivery of a newborn.
- Dantrolene shouldn't be administered in preparation for surgery or labor and delivery.
- During delivery, if rapid sequence induction is necessary, succinylcholine (a known MH triggering agent) may be administered. However, according to the MHAUS, some MH experts don't recommend the use of succinylcholine, and an intubating dose of rocuronium should be used instead.
- Postdelivery, an umbilical cord sample should be obtained for genetic testing to determine MH susceptibility.

MH screening

Diagnosing MH susceptibility is a team effort in the OR. Perioperative nurses can help avert an MH crisis by properly screening patients for MH susceptibility.

For many years, there were no simple tests for MH screening. In fact, until recently, the only definitive test to indicate the possibility of MH response in a patient was the caffeine–halothane contracture test (CHCT). This test exposes a biopsy of skeletal muscle from the thigh to caffeine and halothane, both of which cause muscle contracture. In a susceptible person, the contracture

response is much stronger than normal. CHCT isn't a definitive test; it gives false-positive results in up to 22% of cases. Additionally, the procedure is very expensive, which limits its use as a screening tool. Only a few testing centers are equipped to perform a CHCT. There are currently only six testing centers available, four in the United States and two in Canada.[7] The test is made more difficult because the patient must travel to the testing center since the CHCT has to be performed on freshly biopsied muscle to be accurate. The CHCT test is currently recommended for individuals with a history of a hypermetabolic reaction to anesthetic agents, a family history that suggests MH, or a known genetic susceptibility in the family.[1]

Today, MH testing possibilities have been expanded to include genetic testing, which can point to a genetic basis for MH. Genetic testing isn't definitive either; it only identifies approximately 25% of susceptible patients. The MH genetic test is recommended for patients who have a positive CHCT, have relatives who have had a positive CHCT, have a known genetic mutation for MH, or a highly suspicious clinical episode that occured in the patient or a blood relation.[2] The gene associated with MH is the ryanodine receptor form 1 (RYR1) gene; currently there are 29 listed MH causative RYR1 mutations.[7]

Clinical manifestations

MH develops after a susceptible patient is exposed to a triggering agent. Perioperative nurses need to be able to identify the clinical manifestations of an impending MH crisis (*see Signs and symptoms of MH crisis*).

A rapid differential diagnosis is performed by the anesthesia provider to determine if MH is the cause of the symptoms.[1] Usually, when other causes are ruled out (such as thyrotoxicosis, sepsis, pheochromocytoma, or cocaine toxicity) the patient with clinical symptoms is treated for MH crisis.[1,3,8] Additionally, neuroleptic malignant syndrome is another condition that may mimic MH. Neuroleptic malignant syndrome is a potentially fatal drug-induced hypermetabolic reaction to antipsychotic agents.[3] It isn't an inherited disorder, and is thought to result from dopamine receptor blockade. Dantrolene may also be used to successfully treat this syndrome.[3]

Signs and symptoms of MH crisis[1,3]

Although the anesthesia provider is likely to identify the potential MH episode, the perioperative nurse must also be able to recognize the symptoms of MH and be ready to help organize and coordinate a team response. Signs and symptoms of MH include:

Early signs

- Unexplained tachycardia and dysrhythmias (usually ventricular tachycardia and premature ventricular contractions)
- unexplained tachypnea in an effort to compensate for a rising carbon dioxide level (end-tidal CO_2 levels increase)
- Masseter muscle rigidity after administration of succinylcholine (more common in children)
- generalized erythematous flush and skin warm to the touch
- hyperkalemia and acidosis
- generalized muscle rigidity

Late signs

- Temperature elevation (marked temperature elevation is the hallmark of MH, but it's a later sign)
- hypoxemia
- cyanosis and mottled skin
- coagulopathy (disseminated intravascular coagulation)
- rhabdomyolysis
- myoglobinuria
- kidney failure (resulting from myoglobinuria)

Treatment

Treatment of an MH crisis is complex, and performing all of the immediate necessary actions is beyond the capabilities of any one nurse. In the case of a suspected MH crisis, the very first action is to call for help. Ideally, it takes about four nurses along with the anesthesia provider and surgeon to treat the MH patient competently because so many actions must be done simultaneously.[1] The circulating nurse initiates the MH protocol and calls for additional nursing staff to assist. Nursing staff from the postanesthesia care unit (PACU) and preoperative areas may be needed to assist, depending on the number of OR

MH supplies[12]

Another consideration for MH preparedness is the MH cart itself. Does your OR have the recommended supplies in one container that's mobile enough to be moved into any OR as needed? Small surgical departments usually have one MH supply source available. But if a very large surgical department with many OR suites also only has one MH supply source readily available, there may be delays in getting the cart to the OR in a timely manner.

The following supplies should be available on the MH cart:

- dantrolene (Dantrium) 20 mg vial (36 vials)
- sterile water (preservative-free) for drug dilution
- sodium bicarbonate
- dextrose 50%
- regular insulin
- calcium chloride
- furosemide
- lidocaine
- I.V. cannulas (assorted sizes)
- I.V. administration sets
- syringes (assorted sizes, including a 60 ml syringe to dilute dantrolene) and needleless cannulas to inject the drug quickly into the I.V. site
- central venous pressure kit
- transducing pressure system for arterial and central venous pressure monitoring
- blood collection tubes
- arterial blood gas kit
- nasogastric tubes
- irrigation tray with piston syringe
- indwelling urinary catheter tray
- I.V. fluids (0.9% sodium chloride) and 0.9% sodium chloride irrigation solution (refrigerated)
- bucket and plastic bags for ice

staff available and especially if the crisis occurs during the evening or night shifts. The surgeon should stop the surgical procedure as soon as possible. The anesthesia provider is responsible for discontinuing the inhaled anesthetic agents and succinylcholine, administering 100% oxygen to the patient, and changing the anesthesia machine circuitry. The anesthesia provider will also start any necessary I.V. infusions, insert an arterial line, and draw arterial blood gases (ABGs) and blood work (such as electrolytes, serum glucose, creatine kinase, complete blood cell (CBC) count, and coagulation studies) as needed. The anesthesia provider monitors the patient's vital signs, core temperature, oxygen saturation, end-tidal CO_2, and heart rate and rhythm.

The circulating nurse should document the event, insert an indwelling urinary catheter, and assist the anesthesia provider as needed.

A second nurse will bring the MH cart or supply box into the OR and begin mixing and administering the dantrolene as ordered. The initial dose of dantrolene is 2.5 mg/kg rapid I.V. bolus through a large-bore I.V. site. The dose is repeated until signs of MH are reversed.[9] Each 20 mg vial of dantrolene (up to 36 vials may be needed) is mixed with 60 mL of preservative-free sterile water for injection.[9] Mixing the dantrolene is time consuming and difficult, but using warmed diluent has been shown to decrease the time needed for mixing[10] (see *MH supplies*).

A third nurse should be assigned to bring the emergency resuscitation cart, draw up, and administer medications as per the anesthesia provider's orders. Acidosis is managed with sodium bicarbonate, hyperkalemia is treated with I.V. glucose, insulin and calcium chloride, and cardiac dysrhythmias are treated with antiarrhythmic agents.[1,9] Calcium channel blockers aren't used to treat dysrhythmias because concurrent administration of a calcium channel blocker with dantrolene may cause hyperkalemia or cardiac arrest.[9] Diuretics are administered to prevent or manage myoglobinuria, which causes renal breakdown.[11]

A fourth nurse should bring ice bags and iced saline for lavage. Cooling a patient's rising temperature is instituted with iced saline lavage, iced irrigation of the open abdomen in an abdominal case, ice bags placed under the arms and in the groin areas, iced lavage of any open body cavities such as the stomach, bladder, or rectum, as well as administration of dantrolene as soon as possible. Cold I.V. fluids are administered using refrigerated 0.9% sodium chloride, but lactated Ringer's is avoided so that acidosis is not worsened.[1] Treatment is directed to cooling the patient to 100.4° F (38° C) and managing symptom cascade.[9]

As soon as the immediate crisis has been successfully managed, the patient is prepared for

transfer to the ICU and will stay for about 24 hours after the symptoms have subsided. An I.V. infusion of dantrolene may be administered for at least 24 hours in the ICU to ensure the MH symptoms have been adequately treated.[9] Adverse reactions to dantrolene include muscle weakness, drowsiness, nausea, and fatigue. A recurrent episode of MH can occur within hours of the initial episode, so the patient must be closely monitored and complications treated in the ICU.[1]

When the crisis is over and the patient has been safely transferred to the ICU, debriefing the OR personnel who handled the crisis can help staff understand what was done correctly and what other steps could have been taken to avoid problems that came up during the event. A properly conducted debriefing may provide all personnel the chance to turn the crisis into a learning experience, so that next time a crisis occurs, they will be able to handle things more smoothly. The debriefing may affirm that the tools and procedures are in place to handle an MH crisis successfully, or that some remediation steps need to be taken to provide a proper response given the resources of the OR.

Treatment of an MH crisis is complex, and performing all of the immediate necessary actions is beyond the capabilities of one nurse.

Ensuring competency

Given the complexity of treating MH, once-a-year competency testing may not provide adequate MH education and practice for perioperative nurses. One suggestion is to hold an MH drill at least quarterly, which instructs staff on quick and proper handling of MH. This drill would help perioperative staff practice recognizing an MH crisis, and to smoothly move into the roles best suited to handle the crisis efficiently. A quarterly drill would also improve the OR team coordination and give nurses the opportunity to serve in each of the four roles mentioned previously. Some surgical departments may not have additional nurses available to fill the recommended roles. Possible solutions to this situation may include training the scrub person to assist the circulating nurse, training orderlies or aides to bring the supplies to the OR room, and recruiting managerial personnel and staff from the PACU to assist. It's important that all staff members who may be involved in caring for the patient during an MH crisis attend an MH education session and participate in the MH drill and the annual competency validation.

All staff members should know how to access the facility's protocol for MH, where the emergency supplies are kept, and what additional resources are available. Resources include the MH hotline (1–800–644–9737) from the Malignant Hyperthermia Association of the United States (MHAUS), which is immediately available with expert help to guide healthcare providers in managing an MH crisis.

There is also a Web site maintained by the MHAUS, www.mhaus.org, which contains tutorials on mixing dantrolene, and an online brochure for handling MH. An online presentation for managing MH syndrome is also available for medical professionals.[3]

Patient education

When the MH patient is able to be discharged, it's important to educate him or her about MH. The patient should be taught the definition of MH, and should understand that most people with this susceptibility never have MH-related health problems unless exposed to a triggering agent or circumstance. The patient should also understand the signs and symptoms of nonacute MH expression such as muscle cramps, low-grade fever, and fatigue. Also, advise patients to wear a medical

alert ID. If possible, provide referral for genetic counseling. Instruct the patient to inform all healthcare providers that he's MH susceptible and needs a nontriggering anesthesic agent or a local or regional anesthetic before undergoing surgery or dental work.

Prevention

The best treatment for an MH crisis is prevention. It's important to screen patients preoperatively to identify those who may be MH susceptible. Provide counseling for the patient and family members about MH, necessary precautions, and follow-up testing.

As new information becomes available and is incorporated into the perioperative nurse's knowledge, safer practices for patients have resulted. Perioperative nurses continue to be one of the best safeguards patients have while under anesthesia and are key responders to any anesthetic crisis. **OR**

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Connie L. Martin is a nursing instructor at the University of Nebraska Medical Center, Omaha.

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