New October 2018 Malignant Hyperthermia Guidelines: Is Your Emergency Department Prepared?

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CE Earn Up to 8.0 Hours. See page 226.

While working in the emergency department, you may be the nurse for a 6-year-old girl with status epilepticus who requires emergency intubation with weight-based etomidate or ketamine.[1] Later, the charge nurse assigns you a 9-year-old boy with status asthmaticus who requires intubation with weight-based succinylcholine.[1] Which intubation drugs will trigger malignant hyperthermia (MH)? Are you prepared to recognize, intervene, and provide follow-up care to children in the emergency department who are experiencing MH?

What is MH AND WHICH MEDICATIONS CAN TRIGGER IT?

MH is an inherited, pharmacogenetic skeletal muscle syndrome that appears after an exposure to a trigger presenting as a hypermetabolic reaction.[2-4] A patient with MH experiences fast-rising body temperature and extreme muscle contractions after receiving anesthesia.[1-4] Abnormal skeletal muscle receptors in patients susceptible to MH develop excessive amounts of calcium that accumulate as a result of the anesthetic agent.[1-4] Vigorous exercise and heat also can trigger MH.[2] Only one parent needs to carry the disease for the child to inherit MH.[2-4] Muscle disorders such as central core disease and myopathies have been linked to MH.[1-4] MH can be fatal if ED personnel do not immediately recognize the signs and symptoms and provide treatment. The Malignant Hyperthermia Association of the United States estimates that the prevalence of MH is 1:2000 in the general population.[3] Patients with MH usually are unaware of their predisposition to MH from anesthetics or other triggers.[1-4]

Patients may experience MH upon their first exposure to anesthetic agents, or they may receive anesthetics as many as three times before MH is triggered.[2] MH occurs in all ethnic groups and countries and in persons of all ages.[2] MH occurs 2 times more often in males than in than females; the mean age at presentation is 18.3 years.[2] Children younger than 15 years account for 52.1% of MH reactions.[2] The youngest and oldest age at which MH has been confirmed is 6 months and 78 years, respectively.[2]

MH is a fatal hypermetabolic response to volatile anesthetic agents (eg, desflurane, enflurane, halothane, isoflurane, methoxyflurane, and sevoflurane) and to succinylcholine, a depolarizing muscle relaxant.[1-4] ED providers can diagnose MH after administration of these agents on the basis of the following clinical findings: unexplained elevated expired carbon dioxide that continues to rise despite increasing ventilation support, rigid muscles, hyperthermia, tachycardia, tachypnea, respiratory/metabolic acidosis, rhabdomyolysis, and hyperkalemia.[2-4]

ED CASE SCENARIO: A 9-YEAR-OLD BOY WITH STATUS ASTHMATICUS REQUIRING INTUBATION

The physician orders 100% oxygen via nonrebreather mask with continuous end-tidal carbon dioxide ($ETCO_2$), cardiac and pulse oximetry, frequent blood pressure readings, and temperature monitoring per protocol. Beginning with arrival at the emergency department, comfort measures (including all care) are provided with age and development considerations; the family is present; respiratory treatments

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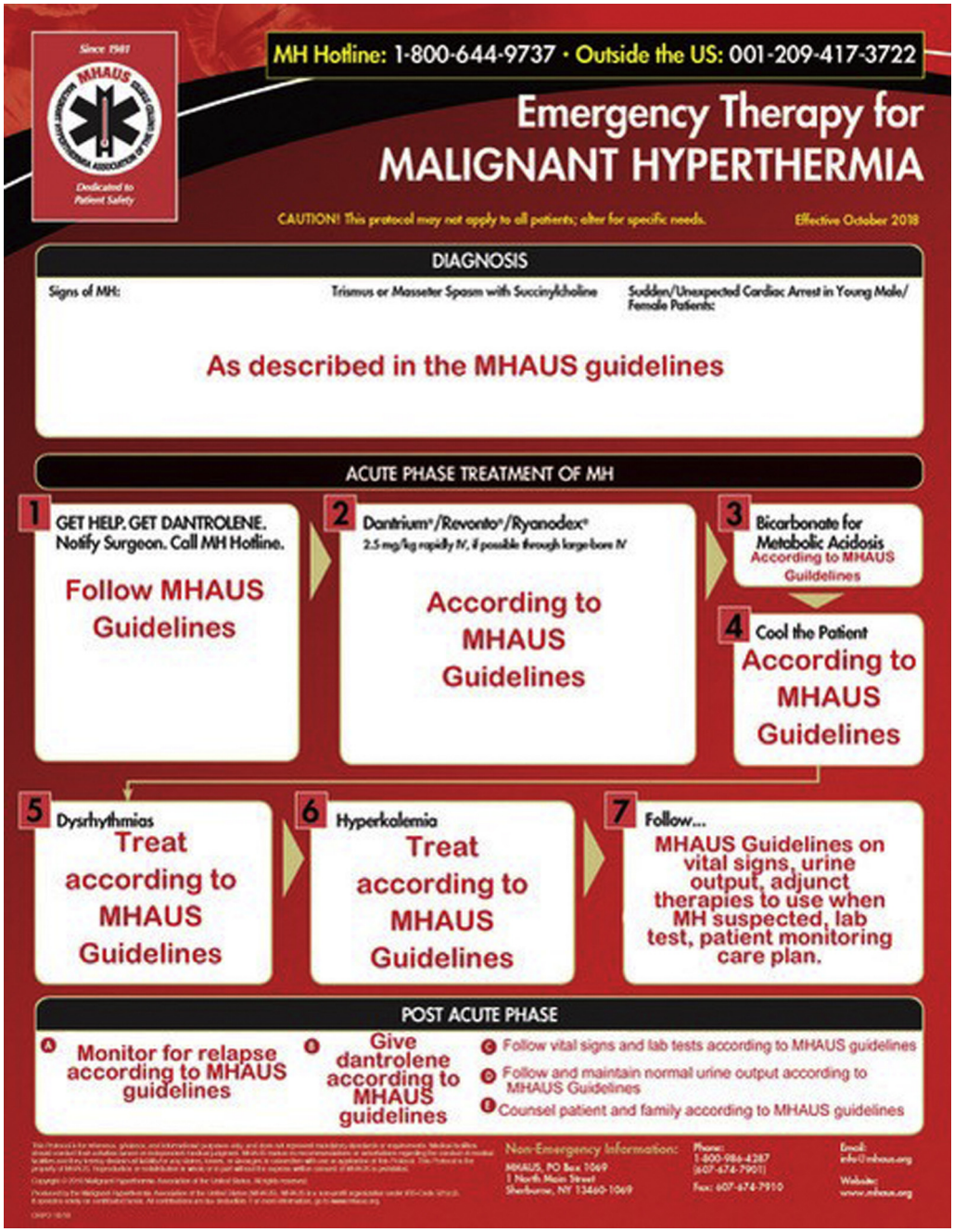

FIGURE

Emergency therapy for malignant hyperthermia. Reprinted with permission from the Malignant Hyperthermia Association of the United States. The malignant hyperthermia poster and cards may be purchased at https://my.mhaus.org/store/ViewProduct.aspx?ID=1512933.

are administered; and IV access is secured. Medical history, food or drug allergies, current medications, and family history of difficulties with anesthesia are reviewed. Consent forms are signed. During the administration of succinylcholine, the child's $ETCO_2$ level continues to rise higher than 55 mm Hg, and tachycardia, tachypnea, severe muscle

rigidity in all extremities, and a temperature of 40° C develop. What is your emergency care plan?

Preanesthesia Screening: Review Exclusion Criteria and Contraindications

Prior to administering anesthesia, each ED nurse must review exclusion criteria if succinylcholine is the planned anesthetic agent. Preanesthesia safety checks should include MH contraindications. Patients who screen positive for any potential complications of MH or who have a family history that indicates complications may occur should not receive succinylcholine in the emergency department because the mechanism of action is unknown.[4] Contraindications are any skeletal muscle disorders, muscle diseases, myopathies, or muscle weakness, as well as King-Denborough syndrome and Native American myopathy.[3,4] Use of MH-triggering anesthesia agents should be avoided in any patient with a history of exertional rhabdomyolysis or heat intolerance.[4]

Diagnosis: Signs and Symptoms of MH

MH signs and symptoms include severe muscle rigidity of the masseter muscle (trismus) or generalized rigidity, a rapidly increasing or late increasing core temperature of higher than 39° C (102.2° F), tachycardia, and tachypnea.[3] The ED nurse may see increasing $ETCO_2$ levels despite increasing or doubling minute ventilation.[3] MH may present with respiratory acidosis with the $ETCO_2$ level greater than 55 mm Hg or greater than 60 mm HG partial pressure of carbon dioxide with controlled ventilation.[3,4] Metabolic acidosis can develop with a base deficit greater than 8 mEq and pH less than 7.25.[3,4] Dysrhythmias can develop if acidosis and hyperkalemia are not treated.[3] Hyperkalemia should be treated. Muscle breakdown causes serum creatine kinase to be greater than 20,000 IU/L after succinylcholine administration.[4] Brown-colored urine indicates myoglobinuria.[3,4]

Acute-Phase and Postacute-Phase Treatment of MH

One must get help right away to manage MH (see Figure). The triggering agent must be discontinued, and dantrolene (Revonto or Ryanodex) should be adminisered as directed.[3] The new October 2018 guidelines should be used; their US hotline is 1-800-644-9737, and outside the US, practitioners should call 001-209-417-3722.[3] Sodium bicarbonate should be administered for metabolic acidosis, and the patient should be cooled according to your agency's cooling policy until his or her temperature decreases to less than 38° C (100.4° F).[3] All vital signs must be continuously monitored, including cardiac monitoring, to identify dysrhythmias and temperature, and a Foley catheter should be placed to monitor urine output.[3] Treatment of acidosis and hyperkalemia usually corrects dysrhythmias.[3] Administer standard drug therapy if dysrhythmias occur. Emergency practitioners need to be aware to avoid calcium channel blocker medications during treatment of dysrhythmias if dantrolene is being given due to the risk of hyperkalemia and cardiac arrest.[3] Avoid use of lidocaine or procainamide if the patient is hyperkalemic.[3] Hyperkalemia should be treated with hyperventilation, bicarbonate, glucose, insulin, and calcium.[3] Diuretics should be administered to prevent myoglobinuria-induced renal failure.[3]

Patients who have experienced MH should be continuously monitored for 24 hours after MH signs and symptoms have resolved to ensure that a relapse does not occur.[3] Signs of recurring MH may be subtle and difficult to diagnose. Patients with MH recurrence may demonstrate increasing rigidity of muscles without shivering, inappropriate rise in temperature, respiratory acidosis, and metabolic acidosis. MH patients receiving respiratory support should be closely monitored for subtle signs of MH recurrence, such as rigidity of muscles that does not lessen with adequate medication for sedation and paralysis.[3] Relapse events occur in 25% of patients who have experienced MH and can be fatal. An MH recurrence should be treated immediately using the exact treatment used for acute-phase MH.[3,4]

Prevention and Conclusion

ED nurses must be aware that succinylcholine can trigger a fatal case of MH if the condition is not rapidly recognized and treated. Every emergency department should practice their response to MH events and have MH code supplies, and the MH hotline number should be easily accessible.[3,4] ED providers should avoid administration of succinylcholine if there is a family history of problems with anesthesia, exercise- or heat-induced rhabdomyolysis, or musculoskeletal disorders or if the history is unknown. Discharge teachings should include the need to wear a medical alert bracelet, how to follow the postacute phase Malignant Hyperthermia Association of the United States guidelines, and the need to avoid stimulant drugs (eg, ecstasy, amphetamines, or cocaine) and other MH triggers.[3,4] ED educators and administrators must provide training regarding recognition of MH, training, and practice drills to prevent fatal MH events and comply with regulatory agency requirements.

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