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Emergencies at a Glance: Malignant Hyperthermia (MH)

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Definition of MH

A life-threatening genetic disorder triggered by certain general anesthetics and/or succinylcholine, causing an acceleration in skeletal muscle metabolism that leads to symptoms like muscle rigidity, tachycardia, hyperthermia, rhabdomyolysis, and acidosis. Immediate treatment with the drug dantrolene usually reverses the symptoms of MH.

Triggers

There are two categories of medications that can trigger an MH crisis: volatile inhaled anesthetics and depolarizing muscle relaxant. Volatile inhaled anesthetics include those ending with "ane," such as halothane, enflurane, isoflurane, sevoflurane, desflurane. Succinylcholine is currently the only depolarizing muscle relaxant available.

Clinical Signs and Symptoms of MH

There are several early signs of a malignant hyperthermia crisis and other signs that are seen later in the disease process.

Signs of MH include rising end-tidal CO_2, masseter muscle rigidity, generalized muscle rigidity, and hyperthermia.

Masseter muscle rigidity is defined as tightness of the jaw muscle after administration of a triggering agent. Mild masseter muscle spasm can be a normal assessment finding after the administration of succinylcholine because of the depolarizing nature of the medication; however, normal muscle fasciculations or spasms should resolve within approximately one minute and are not associated with generalized muscle rigidity. Masseter muscle rigidity is not a normal response to administration of volatile inhaled anesthetics.

Hypercarbia is indicated by an elevated exhaled CO_2 level (normal end-tidal CO_2 is 35 to 45 mm Hg). A sudden elevation of $EtCO_2$ after administration of a triggering agent is the most consistent presenting symptom of MH.

Hyperthermia is defined as a core temperature of >38° C (100.4° F). Core temperature should be monitored during any procedure involving general anesthesia that exceeds 30 minutes. Core temperature sites include the pulmonary artery, esophagus, nasopharynx, bladder, and tympanic membrane (if using thermocouple or thermistor). Hyperthermia can be a delayed symptom and is often seen after a sudden rise in $ETCO_2$ and tachycardia occur. Hyperthermia that develops at least an hour after discontinuation of triggering agents is unlikely to be MH and other causes or disorders of the febrile state should be ruled out. Late signs of MH include hyperkalemia, myoglobinuria, disseminated intravascular coagulation, severe cardiac dysrhythmias, and cardiac arrest.

Cardiac dysrhythmias typically include sinus tachycardia but can also devolve into worsening rhythms associated with the electrolyte abnormalities and acidotic state that occurs during an MH crisis.

Hyperkalemia (elevated potassium) occurs because of sustained muscular contractions during an MH

crisis. When this occurs, potassium is released from cells into the extracellular space. Electrocardiogram changes associated with hyperkalemia may be observed before laboratory test results are available and may include QRS widening and peaked T waves.

Myoglobinuria is characterized by dark-colored urine from myoglobin in the urine as a result from rhabdomyolysis.

Diagnosing MH

There is no definitive test to diagnose an MH crisis. Diagnosis is made based on evaluation of patient symptoms after known triggering agent is administered. Differential diagnosis can include sepsis or infection, insufficient ventilation or fresh gas flow (eg, anesthesia machine malfunction), anaphylaxis, pheochromocytoma, neuromuscular disorders, elevated $ETCO_2$ associated with CO_2 insufflation during laparoscopic procedures, hypoxemia, inadequate anesthesia, serotonin syndrome, thyroid storm, or the use of illicit drugs or stimulants.

Interventions and Management of MH – CALL MHAUS HOTLINE (1-800-644-9737)

1. Initial Steps include notifying the perioperative team that there is a concern the patient is having an MH crisis, calling for help and for the MH cart, stopping the triggering agent, and ventilating with 100% O_2 at high flow.

2. Dantrolene administration should be the prioritized intervention after the initial steps are completed. Reconstitution of the available formulations of dantrolene vary and clinicians should be familiar with the requirements associated with the brand stocked at their facility. Follow the Malignant Hyperthermia Association of the United States (MHAUS) recommendations for initial and repeat dosing.

3. If indicated, establish additional large-bore vascular access for IV fluid and medication management. The anesthesia professional may place an arterial line for hemodynamic monitoring. Laboratory test samples should be sent as soon as possible, and may include, an arterial blood gas (ABG), basic metabolic panel (BMP), creatinine kinase (CK), and lactate. Place an indwelling urinary catheter to monitor urine output and consider a temperature-sensing catheter (if available) for close monitoring of temperature.

4. Managing a patient in the acute phase of an MH crisis involves managing elevated $ETCO_2$, hyperthermia, and cardiac dysrhythmias;

correcting electrolyte imbalances; and maintaining urinary output.

To manage excess CO_2, expect the anesthesia professional to ventilate the patient with 100% O_2 at high flow. Ensure the anesthesia circuit has been changed out and that charcoal filters are added to prevent continued exposure to the inhaled anesthetic.

To manage hyperthermia, actively cool to a temperature of <38° C (100.4° F). Cooling measures include administering cooled IV fluid boluses, application of ice packs to the head, axillae, and groin, or cool irrigation in the abdomen if open from the surgical procedure. Stop cooling efforts when the patient reaches a core temperature of <38° C (100.4° F).

Cardiac dysrhythmias are common during an MH crisis because of the increased temperature and metabolic demand. Manage lethal rhythms using age-appropriate advanced life support algorithms. Avoid calcium-channel blockers and sodium-channel blockers.

Hyperkalemia is a common side effect of MH because of the sustained muscular contractions causing an increase in extracellular potassium in circulation. Manage hyperkalemia utilizing calcium chloride, insulin, and glucose.

Kidney damage can occur because of the vast amount of waste products generated during an MH crisis. Anticipate placement of a urinary catheter to assess urine output and target fluid therapy to a urine output of at least 1 to 2 mL/kg/hr.

Refractory MH

Approximately 80% of patients in one study experienced a recurrence of MH within 16 hours of the initial event prompting continued dantrolene therapy after initial stabilization.

See MHAUS recommendations for bolus doses and infusion rates of dantrolene. Dantrolene can be discontinued when the patient has been stable metabolically for >24 hours, has no muscle rigidity, a core temperature of <38° C (100.4° F), and improving or normalized laboratory test results. Critical care includes close monitoring of respiratory status, cardiac rhythm with treating of any dysrhythmias, acidosis, rhabdomyolysis, continued hyperkalemia, compartment syndrome, and DIC (disseminated intravascular coagulopathy). Additional treatment may include ECMO (extracorporeal membrane oxygenation) for patients in persistent cardiac arrest refractory to MH protocol treatments.