# **EM** CASE OF THE WEEK.

BROWARD HEALTH MEDICAL CENTER DEPARTMENT OF EMERGENCY MEDICINE



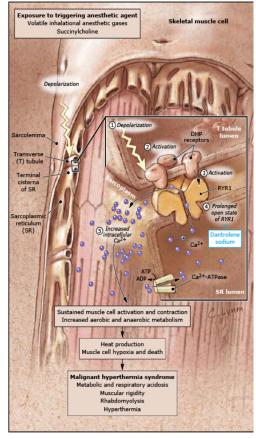
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## Malignant Hyperthermia

A 16-year-old male with a past medical history of asthma presents to the ED with shortness of breath for the last 6 hours. He has been frequently hospitalized in the last year for similar episodes. He denies any fever, chest pain, palpitations, cough, or sputum production. Patient is tachypneic and tachycardic, saturating at 75% on room air. He is afebrile and normotensive. On physical exam the patient is in severe respiratory distress with accessory muscle use and intercostal retractions. Lung exam reveals diffuse inspiratory and expiratory wheezing bilaterally. After several rounds of bronchodilators and positive pressure ventilation failed to improve his saturation, the decision is made to intubate. Several minutes after intubation the patient becomes cyanotic, diaphoretic, with trismus and generalized muscle rigidity. Which of the following is the most appropriate initial treatment for this patient's condition?

- A. IV Dantrolene Sodium at 2.5mg/kg
- B. Start cooling measures including cooling blanket, ice packets, and cold IV solutions.
- C. Administer bicarbonate to treat acidemia
- D. IV insulin and dextrose to treat hyperkalemia
- E. Discontinue any potential triggering agent, call for dantrolene cart, and call for multiple assistants



©2018 UpToDate® Pathophysiology of acute malignant hyperthermia

Malignant hyperthermia results from the uncontrolled release of calcium from the sarcoplasmic reticulum by a triggering agent, producing sustained muscle contractions.

### EM Case of the Week is a weekly "pop quiz" for ED staff.

The goal is to educate all ED personnel by sharing common pearls and pitfalls involving the care of ED patients. We intend on providing better patient care through better education for our nurses and staff.

#### **BROWARD HEALTH MEDICAL CENTER**

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The correct answer is E. Though episodes of malignant hyperthermia (MH) are rare in the ER setting, succinylcholine, a paralytic administered during intubation, can trigger such an event. Suspicion should be high if the patient displays the characteristic signs. Administration of dantrolene is critical when treating MH. However, the very first step in treating MH is ensuring that no additional triggering agent is administered and that multiple assistants, including nursing and paramedics, are rounded up to assist in the labor-intensive mixing of dantrolene sodium, cooling of the patient, and frequent lab draws.

#### **Discussion**

Malignant hyperthermia (MH) has a complex inheritance pattern, with mutations can be present on multiple genes and on multiple chromosomes. It occurs in approximately one in 40,000 adults and one in 15,000 pediatric patients. These mutations result in massive amounts of calcium being released into the intracellular space within muscle cells after exposure to a triggering agent. This causes sustained muscle contraction and severe hyperthermia. The effects are frequently fatal if untreated. Triggering agents include the depolarizing agent succinylcholine and the inhaled anesthetics desflurane, sevoflurane, isoflurane, methoxyflurane, halothane, and ether. The earliest signs of malignant hyperthermia include muscle rigidity, tachycardia, and hypercarbia. Having two or more of these signs increase the suspicion for MH. The most sensitive sign of MH is an observed doubling or tripling of end tidal CO2. Hyperthermia is a late sign with potential temperature increases of 1.8 degree Fahrenheit every 5 minutes.

Complications of MH include acute kidney failure and disseminated intravascular coagulopathy (DIC). The mortality rate of malignant hyperthermia even with treatment is approximately 5-30%. The most common cause of death is from organ failure secondary to DIC.

#### **Treatment**

Initial step to treat malignant hyperthermia focus on terminating the episode and preventing complications from acidosis and hyperthermia. Removing exposure to the triggering agent is the first step in treating the patient. Ventilator lines should be flushed with oxygen or carbon filters placed in line with the respirator. The physician should call for help as the treatment of malignant hyperthermia involves many interventions that require multiple people. Respiratory support is necessary with 100% oxygen and the patient should be hyperventilated to reduce hypercarbia. Dantrolene Sodium (a ryanodine receptor binder that inhibits calcium release into the intracellular space) should be administered at a dose of 2.5mg/kg. Higher dose may be required if the patient does not respond. Doses up to 10mg/kg may be needed, particularly in patients with significant muscle mass. Dantrolene is commonly stored in 20mg vials as a dry powder that must be individually reconstituted with 60ml sterile water. Multiple assistants will be needed to expedite the mixing and hasten administration.

After symptoms are controlled, dantrolene should be administered at 1 mg/kg every 6h for the next 24-48hrs. Monitoring for acidosis is critical. Acidosis is treated with intravenous bicarbonate and with hyperventilation. Hyperthermia should be treated with cooling measures including ice packs over major arteries, cold air convection, cooling blankets, chilled IV fluids, and iced saline lavage if the patient is having abdominal surgery. Temperature should be frequently monitored. Monitoring for hyperkalemia is also critical as it can lead to arrhythmias. The patient should be administered IV dextrose with regular insulin to promote the intracellular shift of potassium into cells. Furosemide can also be administered to promote kaliuresis. Close monitoring of both glucose and potassium is required. Because of the risk of multiorgan failure, coagulation studies should also be performed to monitor for DIC.

For a list of educational lectures, grand rounds, workshops, and didactics please visit **BrowardER.com** and **click** on the **"Conference" link**.

All are welcome to attend!



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## Protocol for immediate treatment of malignant hyperthermia

- **1.** Discontinue volatile anesthetic and succinylcholine. Notify the surgeon. Call for help.
- 2. Mix dantrolene sodium with sterile distilled water and administer 2.5 mg/kg intravenously as a soon as possible.
- 3. Administer bicarbonate for metabolic acidosis
- **4.** Institute cooling measures (lavage, cooling blanket, cold intravenous solutions).
- **5.** Treat severe hyperkalemia with dextrose, 25-50 g intravenously, and regular insulin, 10-20 units intravenously (adult dose).
- **6.** Administer antiarrhythmic agents if needed despite correction of hyperkalemia and acidosis
- 7. Monitor end-tidal CO2 tension, electrolytes, blood gases, creatinine kinase, serum myoglobin, core temperature, urinary output and color, and coagulation status.
- 8. If necessary, consult on call physicians at the 24-hour MHAUS hotline, **1-800-644-9737**

The Malignant Hyperthermia Association of the United States outlines the treatment of MH in the table above. There is a 24-hour a day hotline that healthcare providers can call in the event of a MH episode that will connect them to a physician trained in the treatment of MH.

### **Take Home Points**

- Malignant hyperthermia is a life-threatening event that requires quick removal of the offending agent (succinylcholine, volatile anesthetic) and administration of dantrolene sodium.
- One of the earliest and most sensitive signs of malignant hyperthermia is the doubling or tripling of end tidal CO2. Other findings include tachycardia and muscle rigidity.
- Hyperthermia is a late finding of malignant hyperthermia.
- Close monitoring for acidosis, hyperkalemia, hyperthermia, and DIC are critical in preventing complications of malignant hyperthermia
- Multiple people are needed to carry out the labor-intensive task of treating malignant hyperthermia.



ABOUT THE AUTHOR

This month's case was written by Benjamin Eike. Benjamin is a 4th year medical student from NSUCOM. He did his emergency medicine rotation at BHMC in March 2018. Benjamin plans on pursuing a career in anesthesiology after graduation.

#### REFERENCES

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