

Malignant Hyperthermia

Care of the Patient

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Teenager Dies

Definition

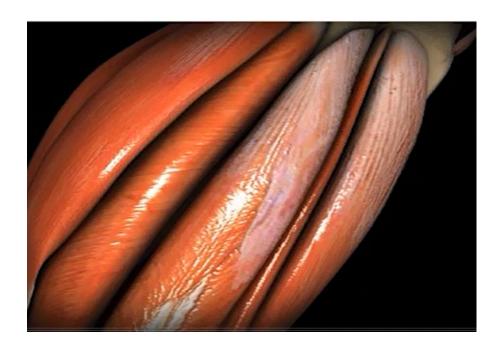
- MH is a rare genetic condition characterized by a severe hypermetabolic state and rigidity of the skeletal muscles.
- It occurs when affected individuals are exposed to a triggering agent such as inhalation anesthetics and succinylcholine, a depolarizing muscle relaxant.
- The triggering agent causes a series of chain reaction in the body related to an increase in intracellular calcium ion concentration.

Triggering Agents

Malignant hyperthermia (MH) is a hypermetabolic disorder of skeletal muscle that is triggered in susceptible individuals by several inhalation anesthetic agents (sevoflurane, desflurane, isoflurane, halothane, enflurane, and methoxyflurane) and succinylcholine.

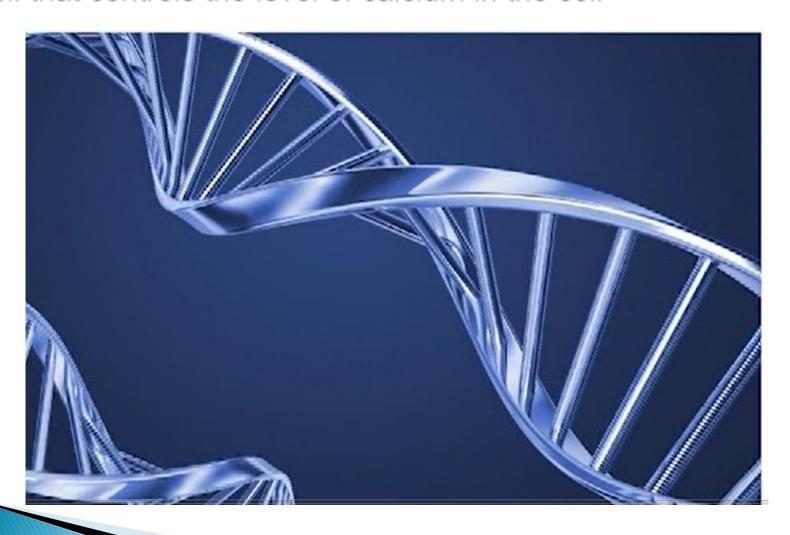


Inherited disorder of skeletal muscles



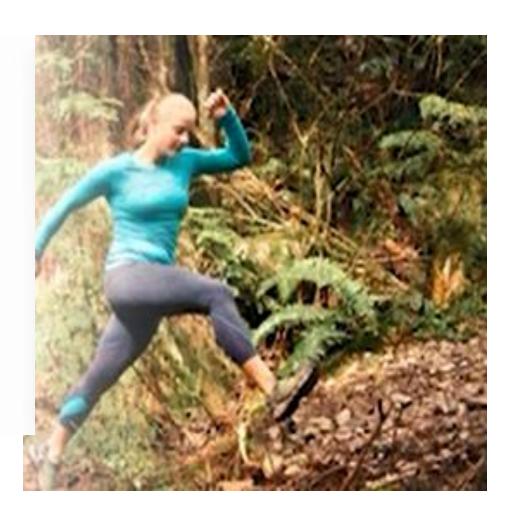
These anesthetic triggers cause intracellular hypercalcemia in skeletal muscle by decreasing the uptake of calcium by the sarcoplasmic reticulum. The intracellular hypercalcemia activates metabolic pathways that result in adenosine triphosphate (ATP) depletion, acidosis, membrane destruction, and ultimately cell death.

Genetic change that leads to an altered protein in the muscle cell that controls the level of calcium in the cell



Hypermetabolism

- ↑ Heart rate
- ↑ Respiratory rate and depth
- ↑ Body temperature



Pathophysiology of MH

- Primary defect resides in the skeletal muscle at the level of calcium released from the cell
- Results in intracellular hypercalcemia Lead to hyper metabolism
- Increased CO2 production
- Increased O2 consumption
- Leads to disruption of the cell membrane
- Muscle tissue unable to return to a resting state
- Creatinine kinase levels rise to 20,000 or more within 12 to 24 hours, putting the patient at risk for myoglobinuric renal failure
- Most susceptible individuals are completely asymptomatic until exposed to triggering agents.

Primary signs of MH

- Muscle rigidity (Stiffness)
- Unexplained Tachycardia (often 1st sign) 90%
- Hypercarbia due to increased CO2 (elevated end tidal CO2) 1st sign in surgery due to ventilator resistance
- Respiratory and Metabolic Acidosis
- Increased Core Temperature 1 to 2 degrees Celsius every 5 minutes
- Skin changes/Cyanosis or Mottling
- Myoglobinura. Renal function altered due to leakage of muscle contents. Dark color urine.
- Fever

Signs and Symtoms

- In most cases, the first signs and symptoms of the disorder are evident in the operating room.
- Malignant hyperthermia can also occur in the recovery room or even after the patient has been transferred to the patient floor.
- It is important to remember that none of the signs and symptoms occur in all cases.

Affected Population

- Ethnic groups all parts of the world affected.
- ➤ In the US higher incidences of MH reported in the following states:
 - Wisconsin
 - Nebraska
 - West Virginia
 - Michigan
- > Episodes of Malignant hyperthermia occur most often, but not exclusively, in children.
- The mean age for Malignant hyperthermia is 15 years, although cases have been reported in infants as well as the elderly.
- The Malignant hyperthermia incidence ranges from approximately 1 in 10,000 to 1 in 50,000 individuals who are exposed to the triggering agents.

Mortality Rate

MH mortality rate has been reduced from as high 70% to less than 5%.



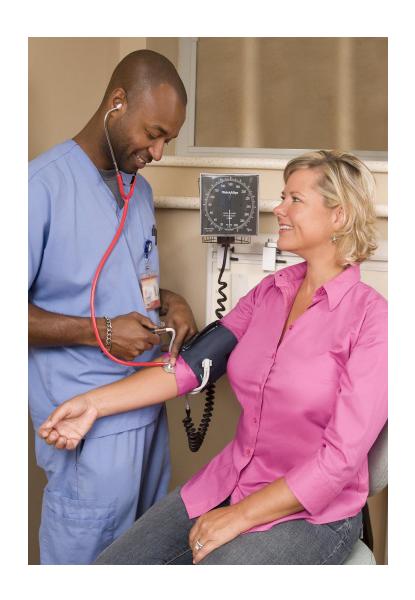
Factors Responsible

- Better screening through careful preoperative interviews and caffeine-halothane contracture testing.
- Pharmacologic basis for MH has been established so triggering agents can be avoided
- Sophisticated monitoring give an early warning so diagnosis and treatment can be started
- The use of Dantrolene

Pre-op Questions:

Question patients about:

- Unexpected death
- Waking up "packed in ice"
- Re-admission to hospital with signs of muscle breakdown after anesthesia
- Pain
- Brown or cola-colored urine without exposure to anesthesia
- History of heat stroke



Preparing Patient cont.

- Peri-op RN assessment
- Review of lab studies i.e. electrolytes, CBC, coagulation studies
- MH can develop over a period of several hours.
- Locate MH cart in pharmacy and bring to OR suite. Blue bin from refrigerator taken during a MH crisis.
- Include training in orientation for OR/PACU/Birth Center staff
- Annual on going review of MH and care of the patient
- May take two-four licensed staff to reconstitute the required amount to accomplish rapid administration.

Preparing Patient with Personal/Family History of MH

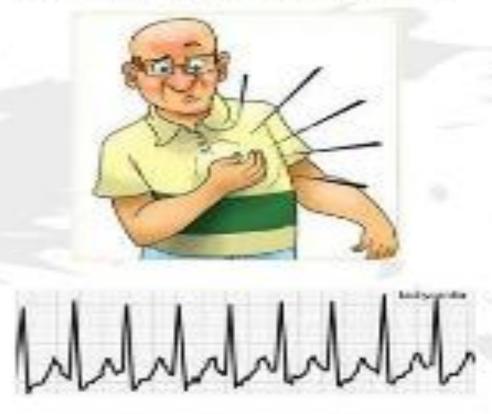
- Preoperative assessment to determine if patient has susceptibility
- Evaluation by anesthesia/health screening
- Consider large bore IV access and arterial line
- Preparation of anesthesia machine.
- Change soda lime canister Flush with 5L/min. of vapor-free oxygen for at least 20 min.
- Consider regional anesthesia and/or use of nontriggering agents

Early Symptoms



Very Fast Increase in Body Temperature (Hyperthermia)

Early Symptoms



Tachycardia - increased heart rate

Late Symptoms

Within minutes, the patient can die from cardiac arrest



Summary of Symptoms

Early Signs	Late Signs
Skeletal Muscle Rigidity	Within minutes Cardiac Arrest
Very fast increase in body temperature	Within hours pulmonary edema or coagulopathy
Tachycardia	Within days neurological damage or obstruction renal failure
Metabolic Respiratory and acidosis	
Cellular Ion Imbalance	
Muscle Spasm	
Sweating	

Conditions Resembling MH

Sepsis:

- Hyperthermia, acidosis, hypermetabolism and muscle breakdown
- Difficult to differentiate from MH
- Rigors resembling muscle rigidity
- Hyperthermia
- Tachycardia
- Tachypnea
- Acidosis



Conditions Resembling MH

Rapid absorption of carbon dioxide during laparascopic procedures:

- Tachycardia
- Acidosis
- Hyperthermia in patients being actively warmed



Resemble MH-Failed Equipment:

- Stuck or defective anesthesia flow valve allowing rebreathing to occur.
- Misplaced LMA

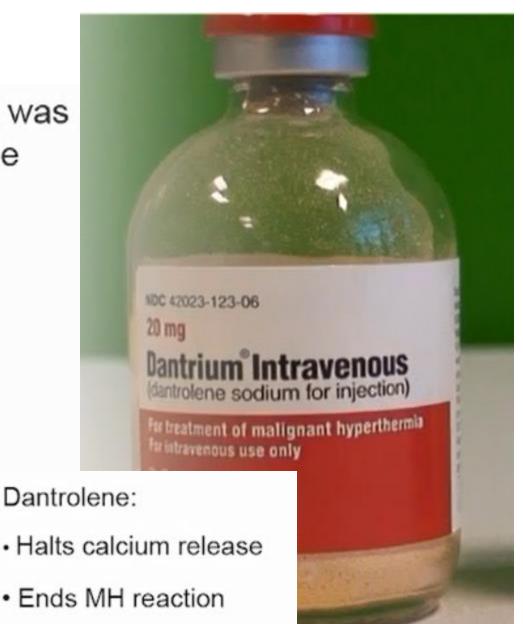


Dantrium

In the late 1970s, a drug was found that will reverse the challenges of malignant hyperthermia.

Dantrolene sodium for injection (Dantrium ®)

Must be administered intravenously.



What is in Dantrium/Revonto?



Each vial contains a sterile lyophilized mixture of 20 mg dantrolene sodium, 3,000 mg mannitol, and sufficient sodium hydroxide to yield a pH of approximately 9.5 when reconstituted with 60 mL sterile water for injection USP (without a bacteriostatic agent).

Use within 6 hours after reconstitution.

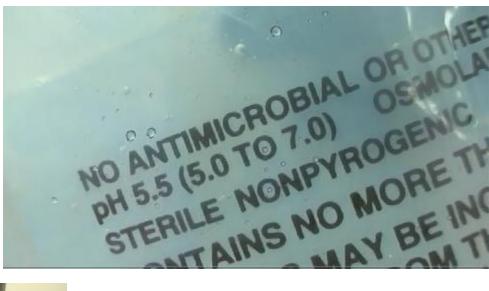
Initial dosing 2.5mg/Kg

Mixing Dantrolene

- Reconstitute each vial by adding 60ml of preservative-free sterile water
- Mix minimum 36 vial.
- Shake vigorously until solution is clear
- Protect from light
- Use within 6 hours after reconstitution

Mix with 60ml Sterile Water



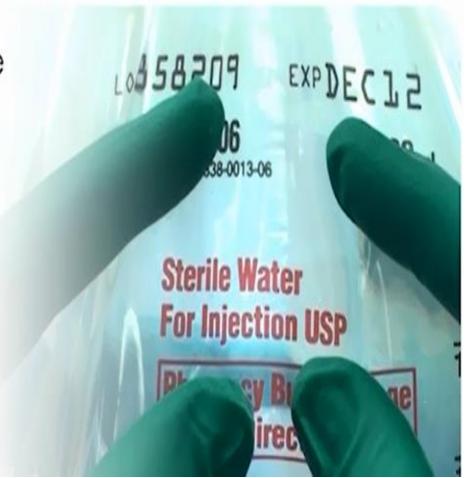




Mixing Dantrolene/MH Cart

Sterile water for injection must be used.

Do not use cold water.



Fairley rapid onset < 10 minutes



