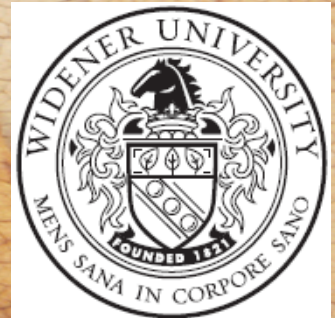


Malignant Hyperthermia: Its Hot, Hot, HOT!!!

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Objectives

Following participation in this educational offering, the learner will:

- Describe the incidence, genetic abnormality and diagnosis of MH.
- Describe normal skeletal muscle contraction and abnormality associated with MH.
- Discuss the triggers and management of MH.
- Apply knowledge of MH to perianesthesia case study.

Our Task for the Next 75 Minutes

- Close & personal with Malignant Hyperthermia.
- Complex topic!
- Sit back and enjoy!

The Problem

<http://www.youtube.com/watch?v=DDmlG63NuhA>

Incidence of MH

- First reported in 1960's.
- Mortality 80%; now 10%.
- Incidence:
 - 1:15,000 children (52%)
 - Mean age 15.2 years
 - 1:50,000 adults
- Range from minor to fulminant presentation.
- Symptom on induction to 1 hour after GA.

Genetics of MH

- >42 genetic mutations
- 4 associated with CCD
- Mutation in the skeletal muscle ryanodine receptor (RYR1)
- *Missense* mutation
- Geographically different

Screening is Key

- **Family history:**
 - Muscle disorders.
 - Unexplained intraoperative deaths.
 - Muscle rigidity; stiffness; fever under anesthesia.
- **Personal history:**
 - Muscle disorders.
 - Muscle rigidity; stiffness; fever under anesthesia.
 - Dark colored urine after surgery.
- **Elective surgery**

Diagnosis of MH

- History or suspicion.
- Muscle Contracture Test: Caffeine & Halothane Contracture Test (CHCT) (IVCT) Europe.
- Genetic Testing (Ryanodine Receptor [RYR1] gene sequencing).
- Testing Centers

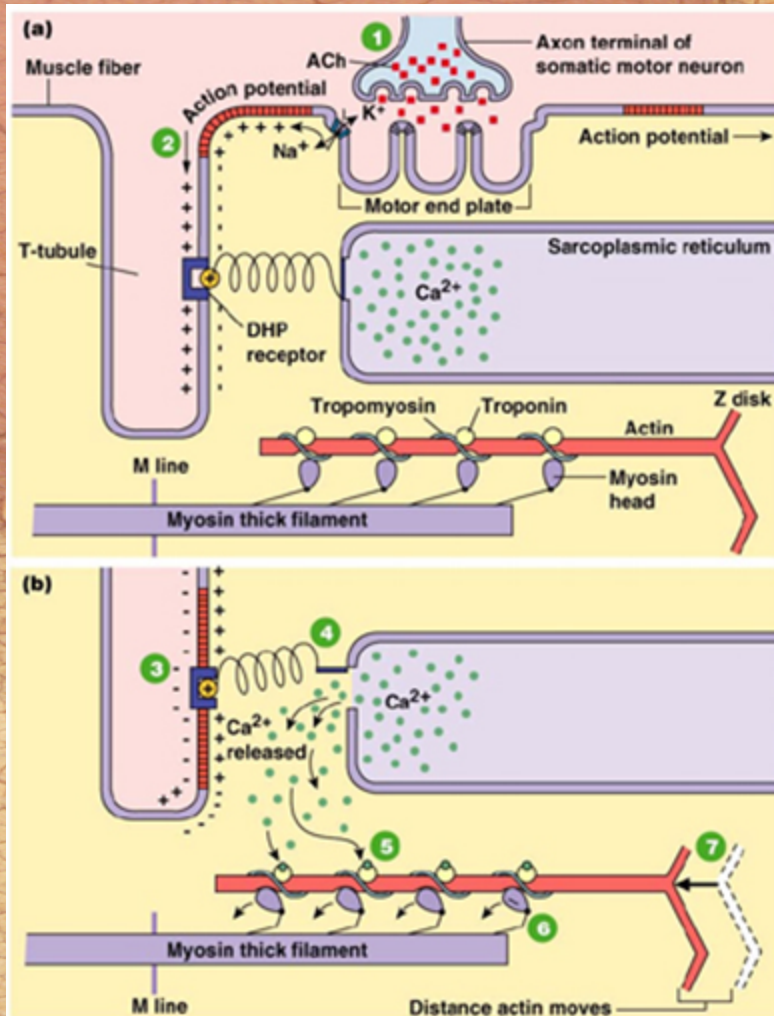
Skeletal Muscle Contraction

The background of the slide features a close-up photograph of several autumn leaves. The leaves are in various stages of decay, with colors ranging from bright yellow to deep orange and brown. The intricate vein structure of the leaves is clearly visible, creating a complex, organic pattern. A semi-transparent white rectangular box is positioned at the top of the image, containing the title text.

Skeletal Muscle Contraction Animation

<http://www.youtube.com/watch?v=CepeYFvqmk4>

Contraction Review

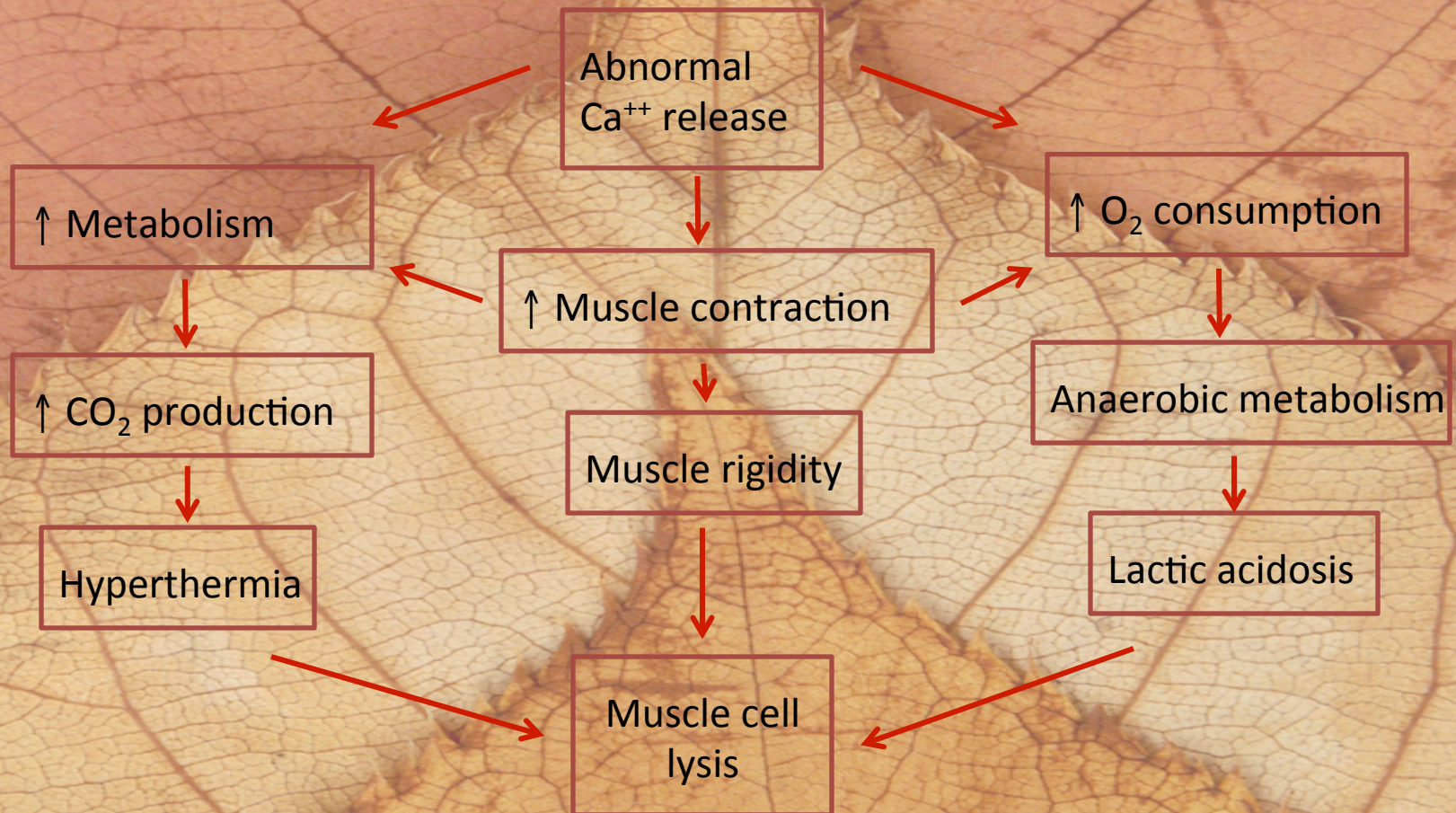


- ① NM stimulation
- ② Action potential
- ③ Voltage receptor “on”
- ④ Tugs ryanodine → Ca^{++}
- ⑥ Actin/myosin couple
- ⑦ Muscle shortens

Malignant Hyperthermia

- Genetic defect in ryanodine receptor → ↑ Ca^{++} release → ↑ intracellular Ca^{++} .
- Sustained muscular contraction → rigidity.
- Muscle metabolism ↑:
 - ↑ O_2 consumption; ↑ HR
 - ↑ heat & CO_2 production
 - Lactic acidosis
- Muscle cell lysis:
 - K^+ , CPK & myoglobin release

MH Downward Spiral



Triggers for MH

- Non-anesthetic triggers:
 - Overheating
 - Body exertion
 - Infection
 - Cocaine and ETOH
 - Myopathies
 - Evans myopathy
 - King Denborough syndrome
 - Central core disease

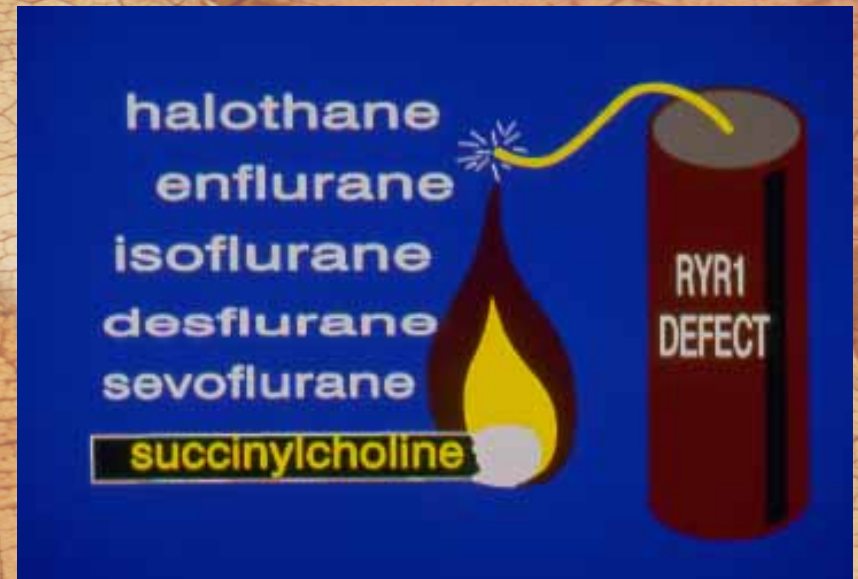
Anesthetic Trigger for MH

- **Volatile Inhalation Agents:**

- Chloroform
- Desflurane
- Enflurane
- Halothane
- Isoflurane
- Methoxyflurane
- Sevoflurane
- Trichloroethylene
- Xenon (rarely used)

- **Depolarizing Muscle Relaxants:**

- Succinylcholine (Anectine)



<http://metrohealthanesthesia.com/edu/mh/mh6.htm>

Similar MH Manifestations

- **Tachycardia:**
 - Hypoxia
 - Hypercarbia
 - Hypovolemia
 - Insufficient anes. Depth
 - Pharmacology
 - Pheochromocytoma
- **Hyperpyrexia:**
 - Heatstroke
 - Transfusion reaction
 - Infection
 - Drug reaction
 - Neuroleptic malignant syn.
 - Serotonin syndrome
 - Hypermetabolism
- **Tachypnea; Hypercapnia:**
 - CHF; pulmonary edema
 - Hypermetabolic states
 - Intraperitoneal CO₂ insufflation
 - Airway obstruction; PTX
 - Excess dead space; ↓ minute vol.
- **Masseter muscle rigidity:**
 - ↓ neuromuscular blockade
 - Temporomandibular joint syn.
 - Neuroleptic malignant syn.
 - Myotonia

Early Symptoms

- Sudden rise in end-tidal CO₂.
- VS: tachycardia, tachypnea, labile BP or arrhythmias.
- Masseter rigidity.
- Acidosis.
- Hyperthermia.
- Cola-colored urine.
- Mottled, cyanotic skin.
- Decreased SaO₂.




A MH Case Study: Here We Go!

You are called in for an emergency appendectomy on a 21 year old male TT. TT is in his junior year of nursing school at a large urban university in the eastern United States. You complete the admission nursing data base with TT and his parents in preparation for his surgery. TT has a negative past medical history, takes no medications and has one hospitalization for severe heat stroke following his first participation in a triathlon 18 months earlier. TT has never has anesthesia or surgery. He is 6'2", weighs 164 pounds (75 kg) and is very muscular in appearance. TT's mother reports TT has had intermittent abdominal pain for the past week, but she became concerned when he was unable to complete his normal morning run. He has also had nausea and vomiting today and has not had solid food since dinner the prior evening. His lab work is all within normal limits, with the notable exception of a white blood count of 18,000.

MH Case Study: The Story Continues...

TT is taken to the operating room and moved to a warmed operating table. Monitoring equipment is applied and he is given 100 μg of *fentanyl* and 2 mg *midazolam* followed by a rapid sequence induction with *propofol*, *succinylcholine* *oxygen*, *nitrous oxide* and *sevoflurane*. TT's baseline temperature with a skin probe was recorded as 37.4° C (99° F) on induction and his heart rate was 114, increasing to 128 and noted to have frequent multifocal premature ventricular contractions (PVC's) with induction. He was also noted to have masseter rigidity with intubation. Surgical incision was made but then surgery was suspended related to an increase in muscular rigidity and a rapid rise in end-titile carbon dioxide (CO₂). He was noted to be cyanotic with skin that was very warm to touch and his temperature had increased to 39° C (102° F).

Rapid Assessment

- Vigilance is  .
- Rapid symptom identification.
- Error on the side of caution.
- Stop all triggers.
- Notify surgeon.
- 100% high-flow O2
- Call for **HELP!**

Sx of Possible MH: **Tell Me Why?**

- Masseter rigidity.
- Color change in CO₂ absorber; ↑ end-tidal CO₂ (> 55 mmHg).
- Labile VS
- Acidosis
- Significant ↑ temperature
- Ruddy urine
- Mottling

MH Party Invitations

- Surgeon
- Anestheologist
- Nurse anesthetist
- Role of Circulating RN
- RN #2: The MH cart
- RN #3: The mixer
- RN #4: The crash cart
- RN #5: The coolest
- MHAUS?

Crisis Priorities

- Stop surgery.
- 100% FIO₂.
- Lines everywhere.
- Stop the Ca⁺⁺ shuttle:
 - **Dantrolene.**
- Rapid cooling.
- Lab assessment.
- Stabilization.
- Call report.
- Learning opportunity.

A Team Approach

<http://www.youtube.com/watch?v=kSOvl1lzSNY&feature=related>

<http://www.youtube.com/watch?v=WfmvyrkWqeE&feature=related>

Dantrolene

- 2.5 mg/kg IV bolus.
 - [70kg: 175 mg **8.75 vials**]
- Repeat prn
- Dose of 10mg/kg may be needed.
 - [70kg: 700 mg **35 vials**]
- If dose 20 mg/kg without improvement reassess.
- Stable: 1mg/kg q 4-6 hrs or 0.25mg/kg drip for 24 hours.
- Remember: mannitol
- Administration.
- 36 vials → **\$2400**

Laboratory Analysis

- ETCO₂
- ABG (arterial & venous)
- Blood sugar & electrolytes
- CK
- Urine output & myoglobin
- Coagulation studies

Tubes-R-Us

- Ice packs.
- Irrigation per temperature.
 - Surgical incision.
 - NGT.
- Multiple large bore IV's.
- Foley.
- Central line or PA monitoring.
- Arterial line.

Additional Medications

- Bicarbonate 1-2 meq/kg initial then ABG-driven.
- \uparrow K^+ : Hyperventilation
 - Bicarb; glucose/insulin; Ca^{++}
 - Peds:
 - insulin 0.1u/kg
 - 1ml/kg 50% glucose
 - Adult:
 - 10 u regular IV
 - 50ml 50% glucose
- Calcium channel blockers contraindicated.

Additional Medications

- Follow lab values.
- Significant \uparrow K^+ or CK or u/o $< 0.5\text{ml/kg/hr}$: diuresis
- Maintain u/o $> 1\text{mk/kg/hr}$.
- Give bicarbonate to alkalinize urine to prevent myoglobin-induced renal failure.

Phase I Care

- Late complications.
- Frequent VS/labs.
- Respiratory control.
- Critical care bed.
- Recommend follow-up.

Recommendations

- Notify MHAUS.
- Council family & patient:
 - MH:
 - Precautions
- Complete AMRA:
 - www.mhreg.org
- Written notification to patient & attending MD.
- Refer to nearest biopsy center.

Emergency Preparedness

- MH Drills:
 - A competency test.
 - Quarterly recommended.
 - Play different roles.
 - Plan different settings.
 - Many hands make light work.
 - Coordination is central.

MH Case Study: The Story Continues...

All anesthetics were discontinued, a second large bore intravenous was inserted for rapid cool fluid administration and TT was hyperventilated with 100% oxygen. The MH protocol was put into place and the PACU nurse assisted with the aggressive cooling and the insertion of a foley catheter, noted to immediately drain cola-colored urine. *Dantrolene* was mixed and administered and arterial blood gas analysis and serum electrolytes were obtained. *Bicarbonate* was administered for a severe metabolic acidosis (pH 6.55) and *glucose, insulin and calcium carbonate* were given for a serum potassium of 7.1 mEq/L. Arrangements were made for an emergent intensive care bed (ICU) bed and the patient was transported to ICU and placed on mechanical ventilation.

MH Case Study: The Story Continues...

TT's condition stabilized in 24 hours and he returned to the operating room for appendectomy for ruptured appendix 48 hours later with *propofol* and epidural anesthesia without problem. MH testing was strongly recommended for TT and his four siblings.

**Taken from: Noble, K. A. (2007). Malignant hyperthermia: hot stuff!
Journal of PeriAnesthesia Nursing, 22(5), 341-345.**

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Questions?

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