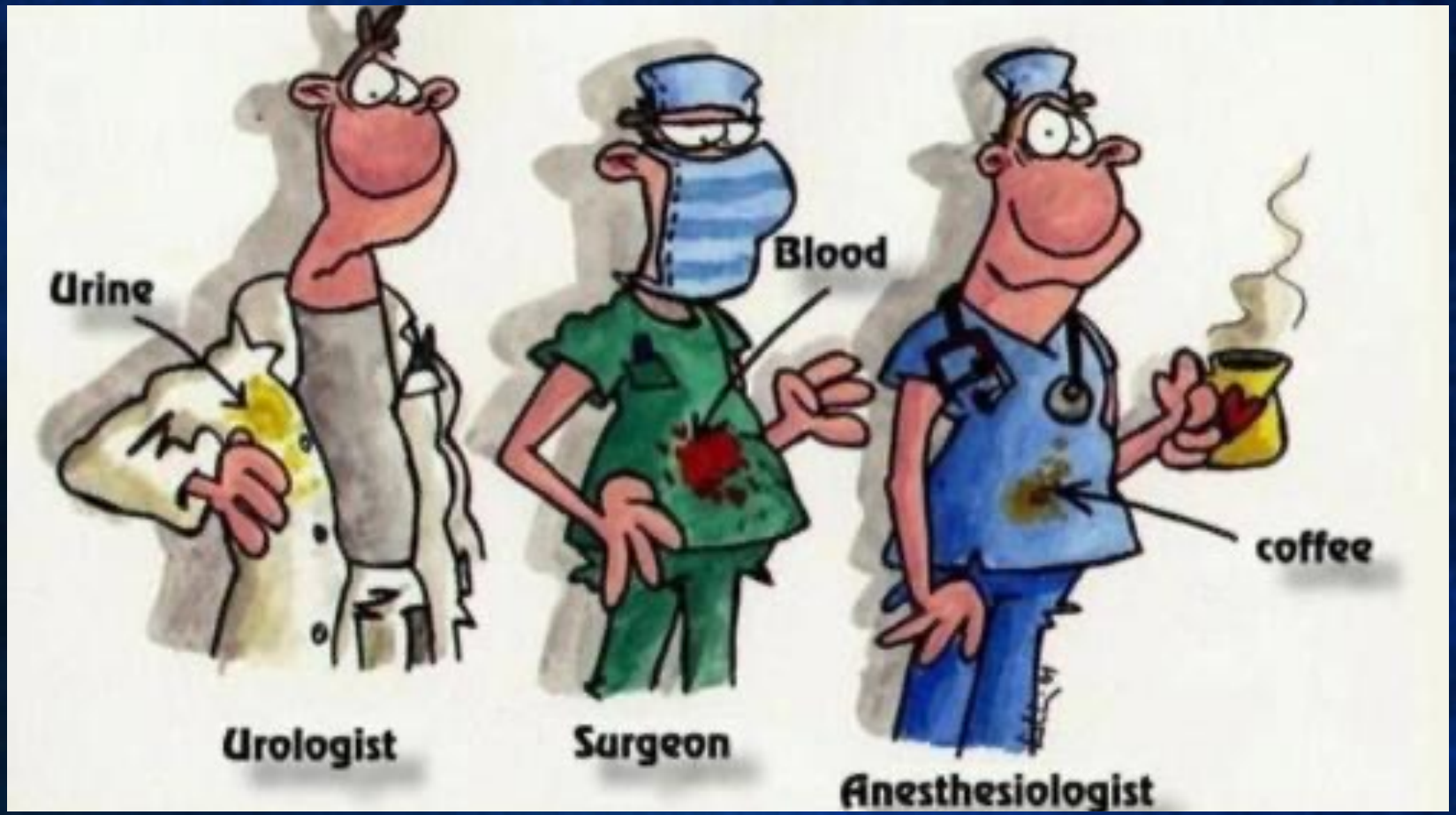
Malignant Hyperthermia

Nigam Sheth MD, FASA

Anesthesia Scheduling Services

Cardiothoracic Anesthesiology


Urine
Blood
coffee
Urologist
Surgeon
Anesthesiologist


"The anesthetist is running late. We'll have to start without him."
J. di Chiaro
CARTOONSTOCK .COM
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Introduction

- Incidence
- Causes
- Genetics
- Pathophysiology
- History
- Signs and Symptoms
- Differential Diagnosis
- Treatment
- Prevention
- Testing


ANESTHESIA
TECHNICIAN
BECAUSE FREAKIN'
MIRACLE WORKER
ISN'T AN OFFICIAL
JOB TITLE

Thank You Karen!

Definition:

Malignant Hyperthermia

- inherited disorder
- hypermetabolic state
- skeletal muscle
- triggered by
  - inhalational anesthetics
  - depolarizing muscle relaxants

Incidence

- Can be seen at either end of spectrum
- Most commonly seen in first 3 decades of life
- Exact incidence unknown
  - 1:3000 to 1:15,000 incidence in kids
    - Children under 15 account for 52% of cases
  - 1:50,000-1:100,000 incidence in adults
  - 1:30,000 incidence in general population
- Many cases are unreported due to various presentation and legal ramifications

Causes:

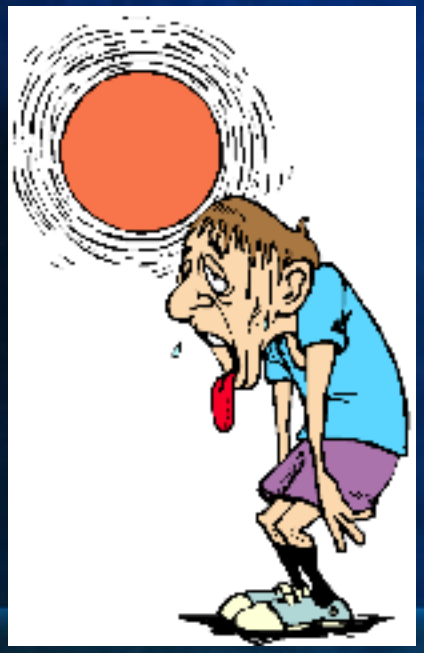
- Inhalational Agents
  - Halothane
  - Sevoflurane
  - Desflurane
  - Isoflurane

Causes:

- Depolarizing muscle relaxants

Causes:

- Heat
- Stress
- Vigorous Exercise

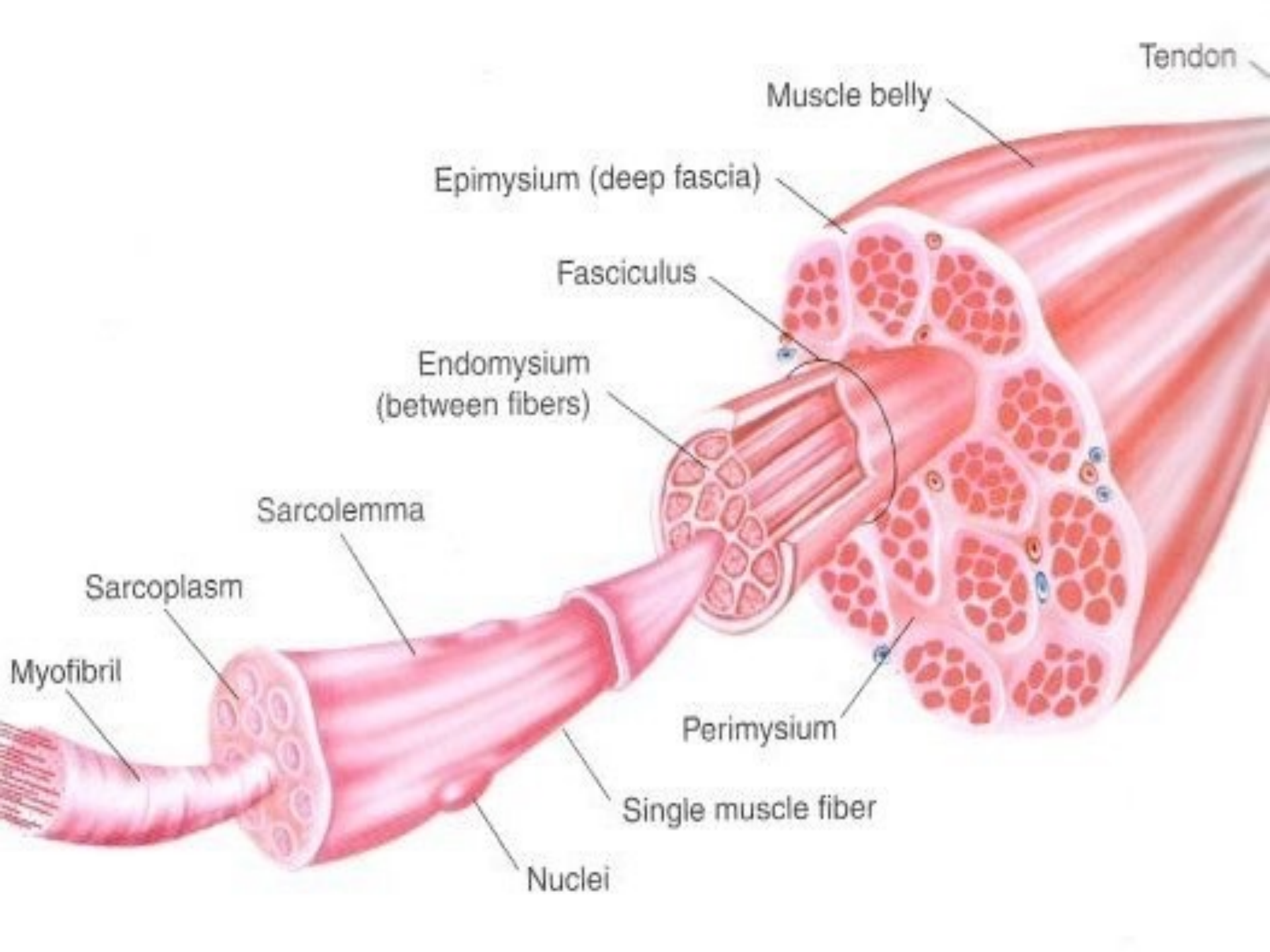


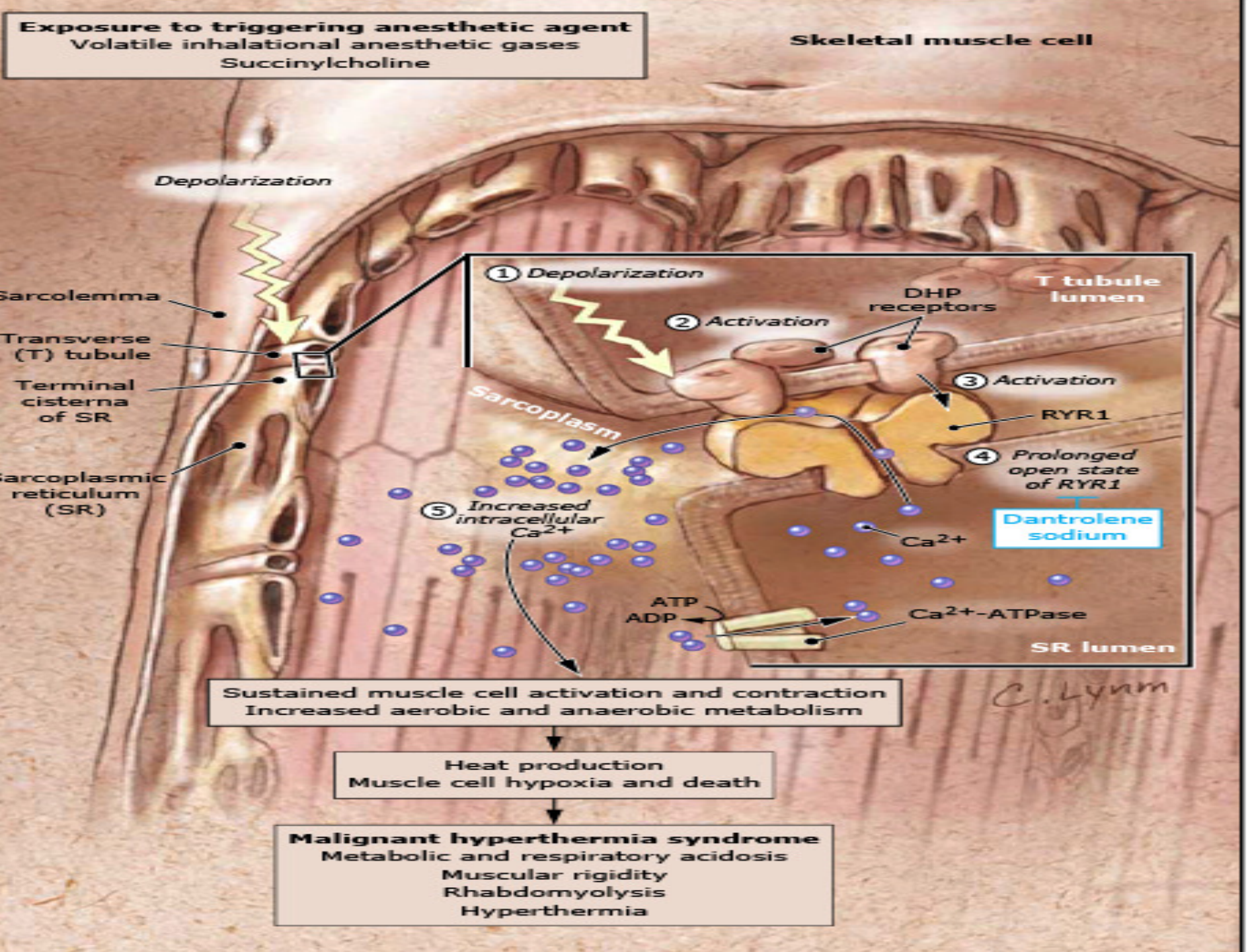
Causes:

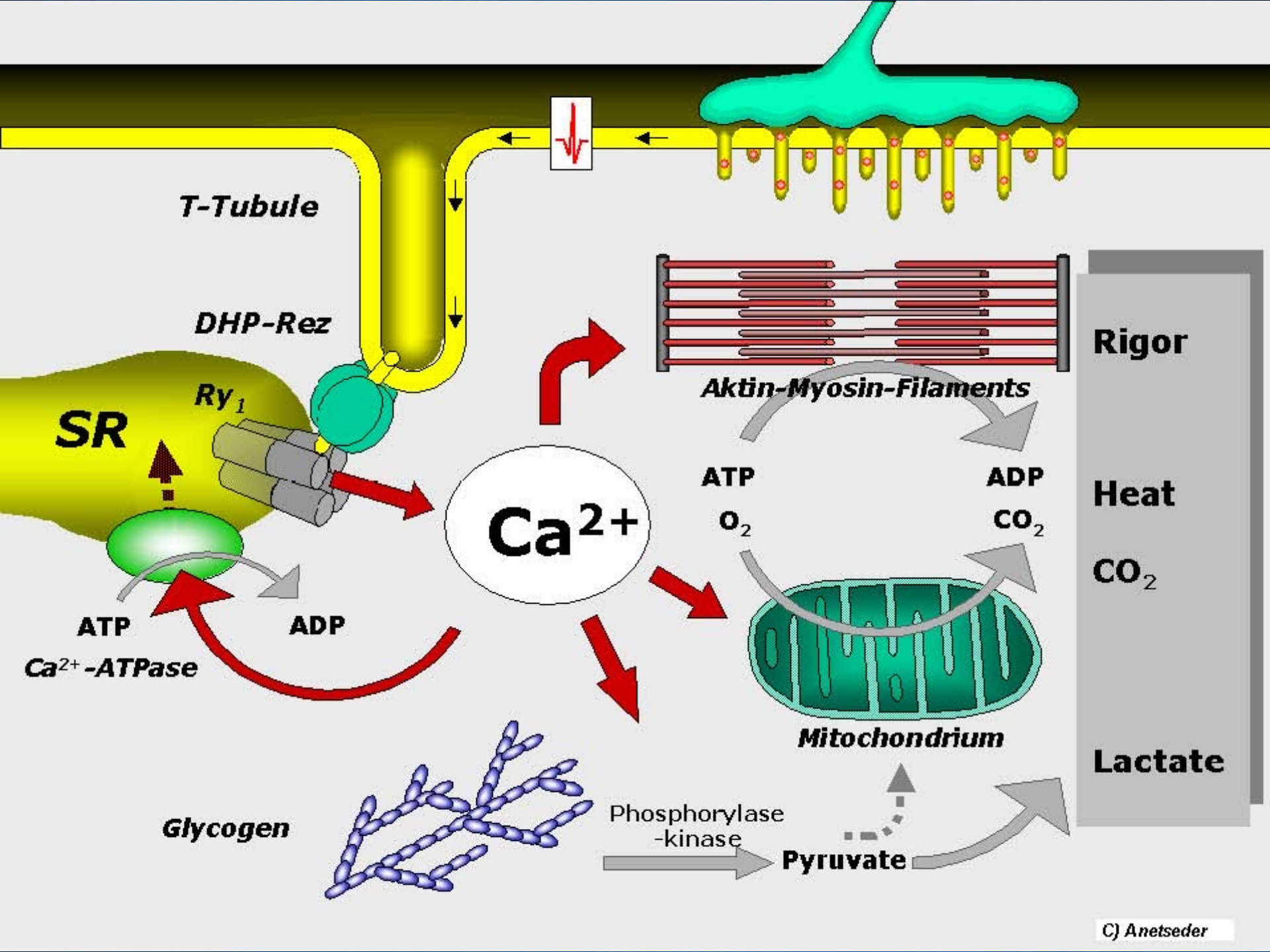
- 30% of cases need greater than 3 exposures to anesthetics to prompt reaction
- Variable penetration and presentation

Genetics:

- Chromosomal abnormality:
  - Chromosome 19
    - 50% of known MH cases (with multiple variations)
  - Chromosome 1
    - 1% due to mutations
- Abnormal receptor mutation of:
  - Dihydropyridine receptor (DHPR) or
  - Type 1 ryanodine receptor (RYR1) on skeletal muscles
- Inheritance:
  - Autosomal dominant


Tendon
Muscle belly
Epimysium (deep fascia)
Fasciculus
Endomysium
(between fibers)
Sarcolemma
Sarcoplasm
Myofibril
Perimysium
Single muscle fiber
Nuclei


Exposure to triggering anesthetic agent
Volatile inhalational anesthetic gases
Succinylcholine
Skeletal muscle cell
Depolarization
Sarcolemma
Transverse (T) tubule
Terminal cisterna of SR
Sarcoplasmic reticulum (SR)
1 Depolarization
2 Activation
DHP receptors
T tubule lumen
3 Activation
RYR1
Sarcoplasm
4 Prolonged open state of RYR1
Dantrolene sodium
5 Increased intracellular Ca2+
Ca2+
ATP
ADP
Ca2+-ATPase
SR lumen
Sustained muscle cell activation and contraction
Increased aerobic and anaerobic metabolism
Heat production
Muscle cell hypoxia and death
Malignant hyperthermia syndrome
Metabolic and respiratory acidosis
Muscular rigidity
Rhabdomyolysis
Hyperthermia

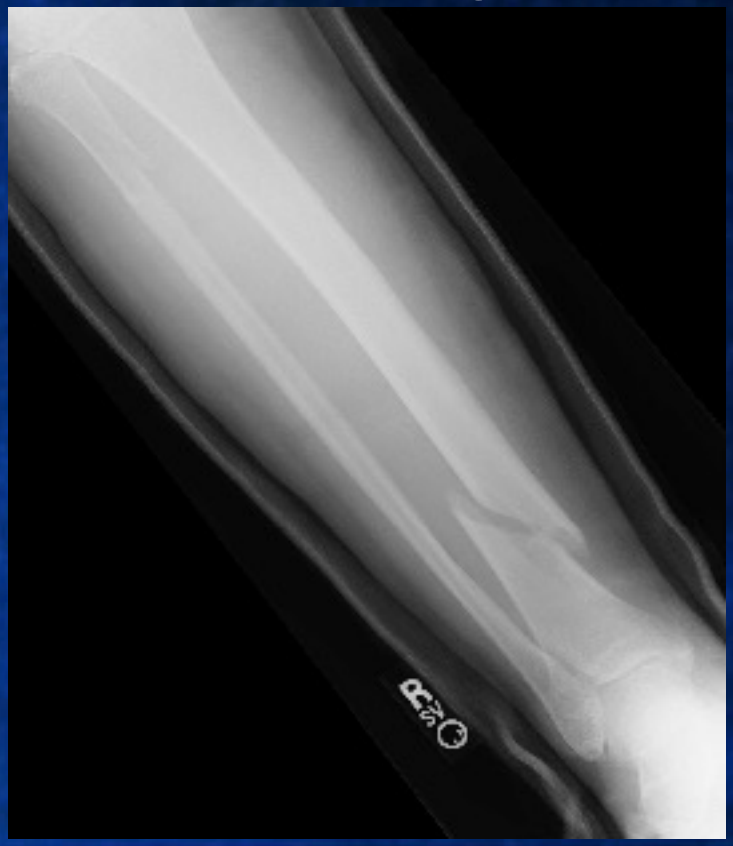

T-Tubule
DHP-Rez
Ry 1
SR
Ca2+
ATP
ADP
Ca2+ -ATPase
Aktin-Myosin-Filaments
ATP
ADP
O2
CO2
Mitochondrium
Glycogen
Phosphorylase
-kinase
Pyruvate
Rigor
Heat
CO2
Lactate
C) Anetseder

History:

- Between 1915 and 1925, one family suffered 3 deaths due to an anesthesia related event
- Rigidity and Hyperthermia in the patients
- Unclear picture
- Later confirmed MH susceptibility

History:

- 1960-21 year old male fractured his leg
- More concerned with the anesthesia than his leg.
- History of multiple deaths in his family due to anesthesia



History:

- Young man under the care of anesthesiologist Dr. Villiers
- During OR case, he suffered cyanosis, an increase in temperature, and tachycardia
- Case terminated, patient cooled
- First known survival of MH episode
- Case investigated by
  - Dr. Denborough-an internist and research fellow interested in genetics
  - Dr. Lovell-anesthesiologist
- Case report led to recognition of MH and further investigation

ANESTHESIA WAS INVENTED IN 1846

DOCTORS IN 1845:

History

- 1975-Dantrolene found to be effective in treating MH in pigs
- 1979-Dantrolene approved by FDA for MH treatment
- 1980's-awareness of MH increased. MH registry created in US (MHAUS)
- 1990's-molecular biologic techniques identified genes associated with MH
- 2003-genetic test for MH developed

Signs and Symptoms:

- MH can occur intraoperatively, in the PACU, or after discharge
- Most likely within 1 hour of initiation of inhalational agents or depolarizing muscle relaxant

Signs and Symptoms:

- Early Signs:
  - Rise in $ETCO_2$
    - Earliest sign of MH
    - Refractory to increased ventilation and higher tidal volumes
    - Due to an increase in metabolism and more CO_2 production
    - DDx: hypoventilation, machine malfunction, absorption of CO_2 during laproscopy

Signs and Symptoms:

- Early Signs:
  - Tachycardia
    - Inappropriate of patient condition
    - DDx: pain, surgical stimulus, sepsis, inadequate anesthesia depth, pheochromocytoma, etc

Signs and Symptoms:

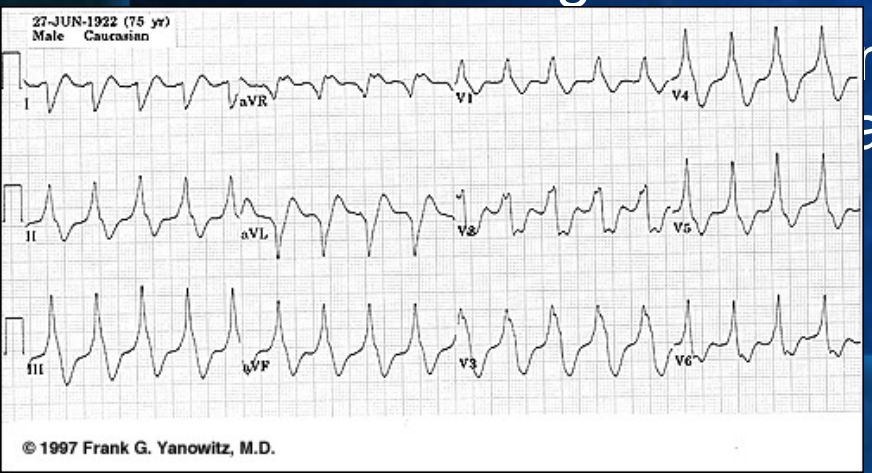
- Early Signs:
  - Masseter Muscle Rigidity
    - Inability to open patient's mouth after triggering event
    - If persistent, indicative of MH in 30% of all cases
    - Although **not** pathognomonic, if present, triggering agent should be discontinued and suspicions for MH high

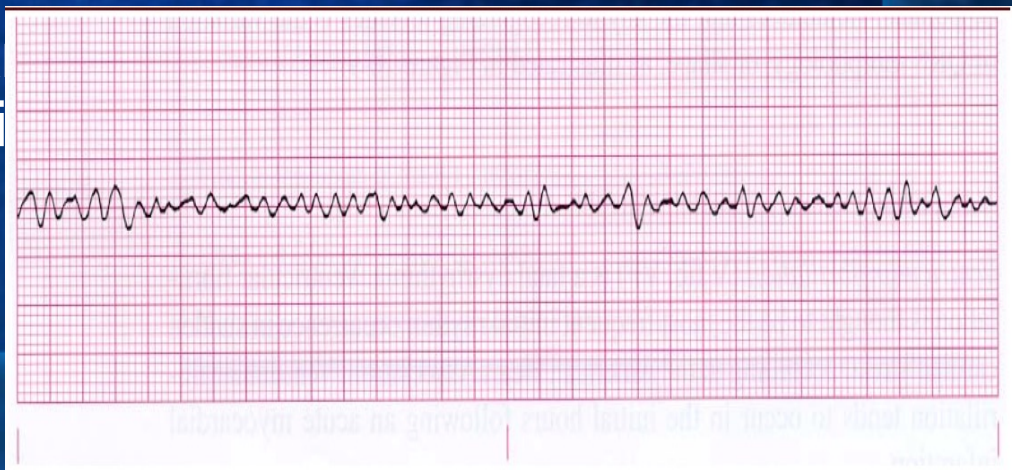
Signs and Symptoms:

- Early Signs:
  - Generalized muscle rigidity
    - Pathognomonic for MH if present with other signs

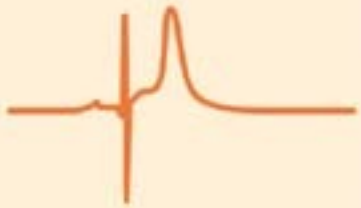
Signs and Symptoms:

- Late Signs:
  - ECG changes
    - Cell death from anaerobic metabolism and hypoxia can cause muscle breakdown
    - Muscle breakdown can lead to hyperkalemia, increasing potential for arrhythmias
    - Strengthens Dx of MH



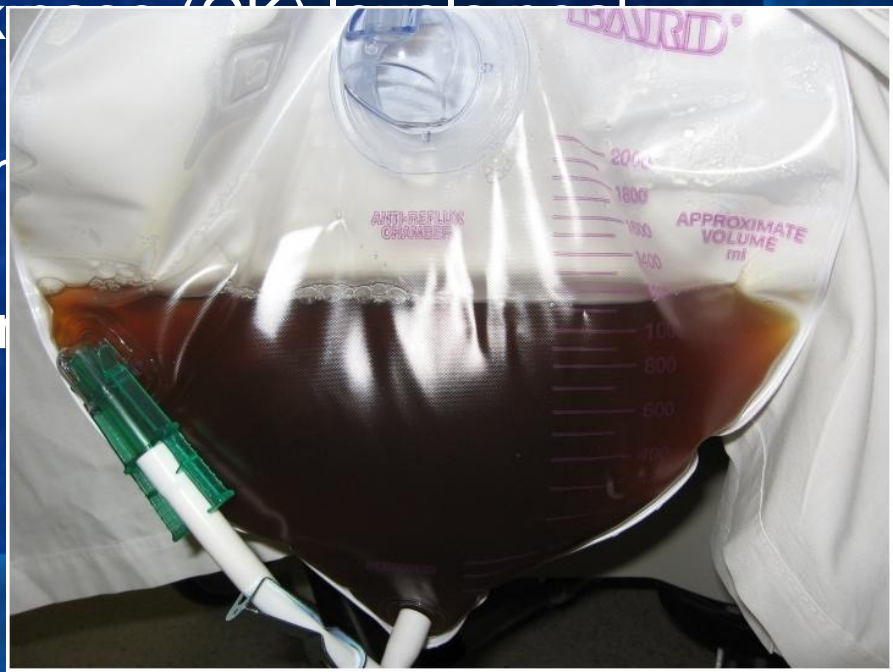


Signs and Symptoms:

Serum Potassium	Typical ECG Appearance	Possible ECG Abnormalities
Mild (5.5–6.5 mEq/L)		Peaked T Waves Prolonged PR Segment
Moderate (6.5–8.0 mEq/L)		Loss of P Wave Prolonged QRS Complex ST-Segment Elevation Ectopic Beats and Escape Rhythms
Severe (>8.0 mEq/L)		Progressive Widening of QRS Complex Sine Wave Ventricular Fibrillation Asystole Axis Deviations Bundle Branch Blocks Fascicular Blocks

Signs and Symptoms:

- Late Signs:
  - Rhabdomyolosis
    - Muscle fatigue will lead to muscle breakdown causing rhabdomyolosis
    - Elevation of creatine k
      around 14 hours after
    - Elevated CK levels ra
      100,000
    - Normal CK ranges fro
    - Can manifest as renal
      arrest



Signs and Symptoms:

- Late Signs:
  - Hyperthermia
    - Commonly misunderstood as an early presenting sign
    - From sustained muscle contraction from unregulated calcium release
    - Core temperature may rise 1ºC/5min
    - DDx: Intraoperative fever, faulty temperature probe, Neuroleptic Malignant syndrome
    - Extreme high temperatures (up to 45ºC) can lead to increased CO_2 production, increased O_2 use, and organ dysfunction

Differential Diagnosis:

- Neuroleptic Malignant Syndrome
  - Muscle rigidity
  - Temperature increase
- Meningitis
- Pheochromocytoma
- Sepsis
- Thyrotoxicosis
- Heatstroke
- Fever
- Cocaine/Amphetamine Toxicity

Treatment:

- TURN OFF AGENT
- Call for **HELP**
- Increase flows to 100% O_2
- Dantrolene
- Treat arrhythmias and hyperkalemia
- Cool patient
- ICU transfer

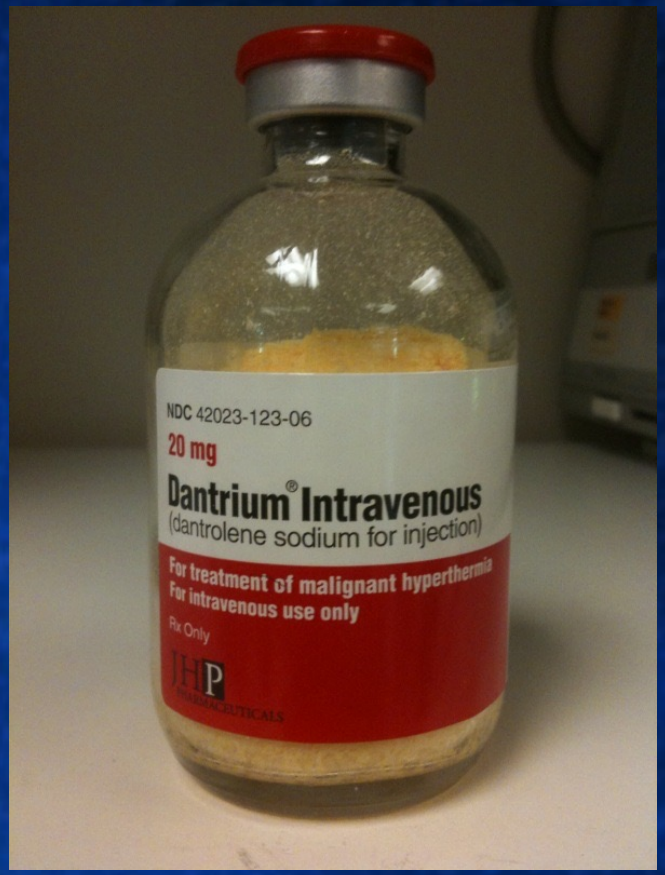
ANESTHESIOLOGIST

I DON'T BELIEVE IN MIRACLES...

I RELY ON THEM

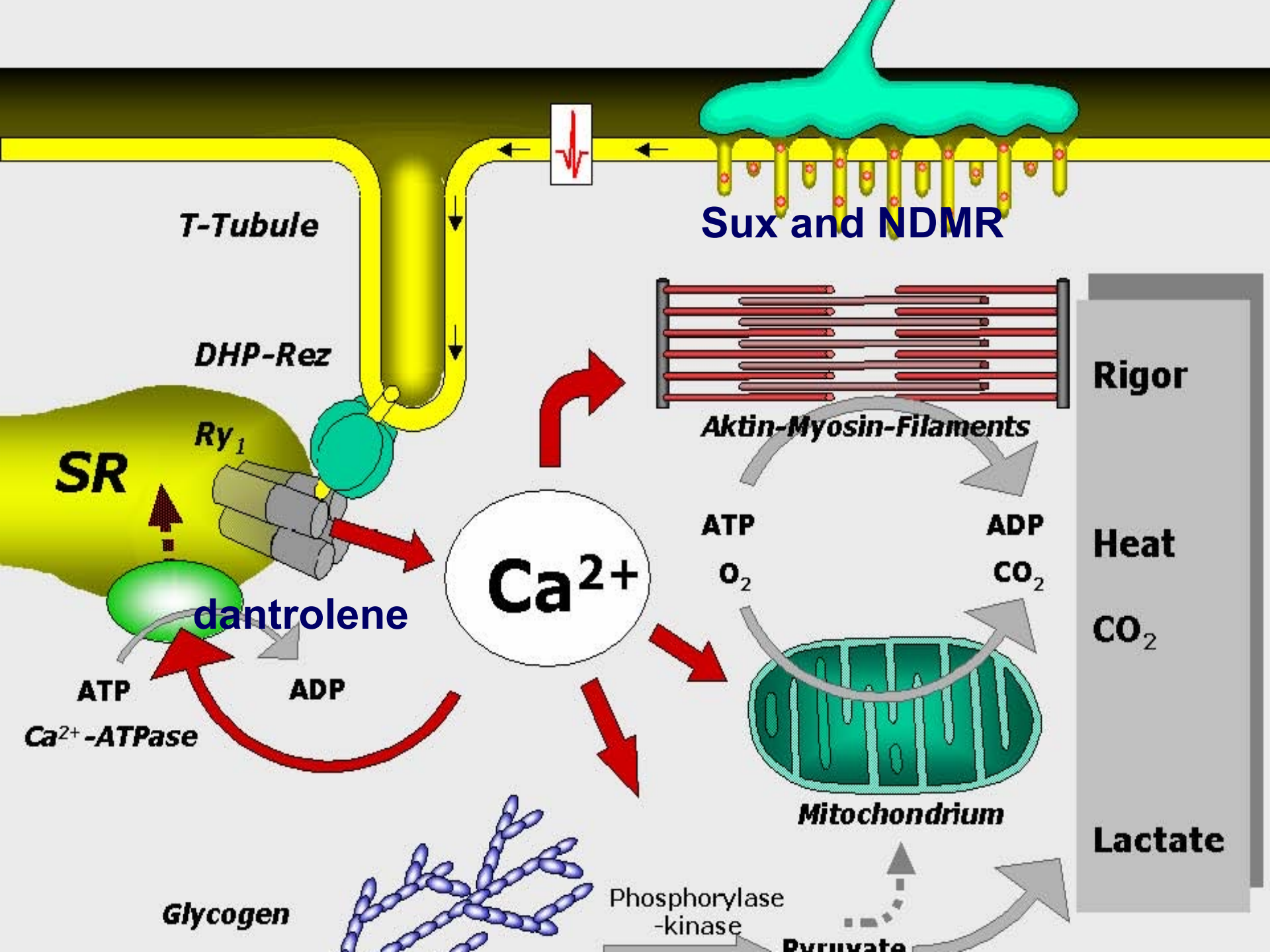
Treatment:

- Dantrolene
  - Muscle relaxant
  - Mainline Treatment for MH



Treatment:

- Dantrolene
  - Unlike
    - neuromuscular blocking agents that act at the site of the neuromuscular junction or
    - nonspecific relaxants which act at the spinal cord (ex: flexeril)
  - Dantrolene acts within the muscle cell
    - decreases calcium release from the sarcoplasmic reticulum
    - reducing intracellular calcium levels
  - Exact mechanism of action is unknown


T-Tubule
Sux and NDMR
DHP-Rez
Ry1
SR
Aktin-Myosin-Filaments
Rigor
ATP
ADP
O2
CO2
Heat
CO2
Ca2+
dantrolene
ATP
ADP
Ca2+-ATPase
Mitochondrium
Lactate
Glycogen
Phosphorylase
-kinase
Pyruvate

Treatment:

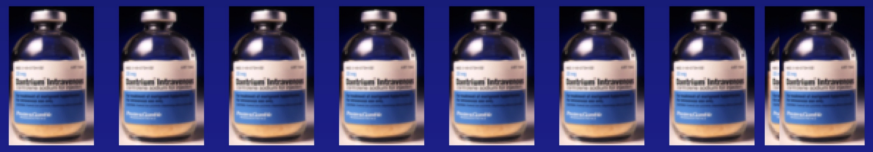
- Dantrolene:
  - 2.5mg/kg with repeat doses as needed.
  - Up to 10mg/kg
  - Most episodes controlled with 2-3mg/kg
  - Mixed with 60ml of sterile water
  - MUST be made with sterile water or it will precipitate

Dantrolene for MH Crisis

20 mg/60 ml = 1 mg/3ml

70 kg patient:

2.5 mg/kg = 175 mg or 525 ml (**9** vials)



~10 mg/kg = 700 mg or 2100 ml (35 vials)

Treatment:

- Ryanodex:
  - 250mg dantrolene per vial.
  - Mixed with 5ml of sterile water
    - Compare with 60ml for traditional formulations of Dantrolene
  - Significantly less sterile water mix

Treatment:

Table 1 Comparison of Dantrolene Formulations

Factor	Dantrium/Revonto	Ryanodex
Vial strength	20 mg	250 mg
Diluent volume per vial	60 mL	5 mL
Concentration after reconstitution	0.33 mg/mL	50 mg/mL
Mannitol content per vial	3,000 mg	125 mg
Number of vials needed	35	3
Average volume to be administered	2,100 mL	14 mL
Time to reconstitute	≥22 min for 13 vials	≤1 min for 1 vial
Shelf life	3 y	2 y
Approximate cost for suggested supply	$2,000-$3,000	$6,000

min: minute.
Source: References 16-18.

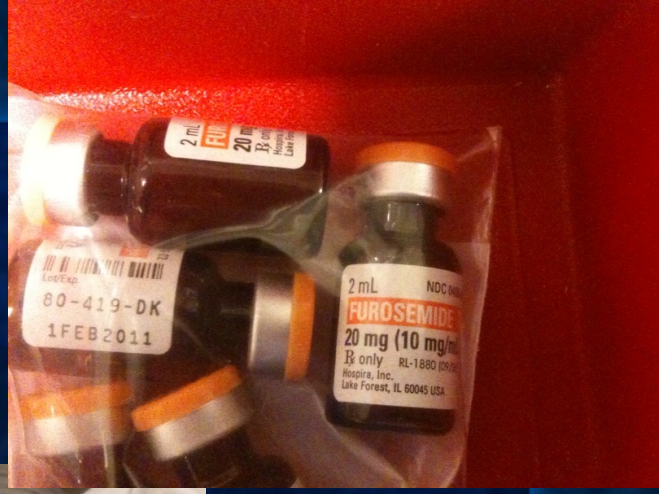
Treatment:

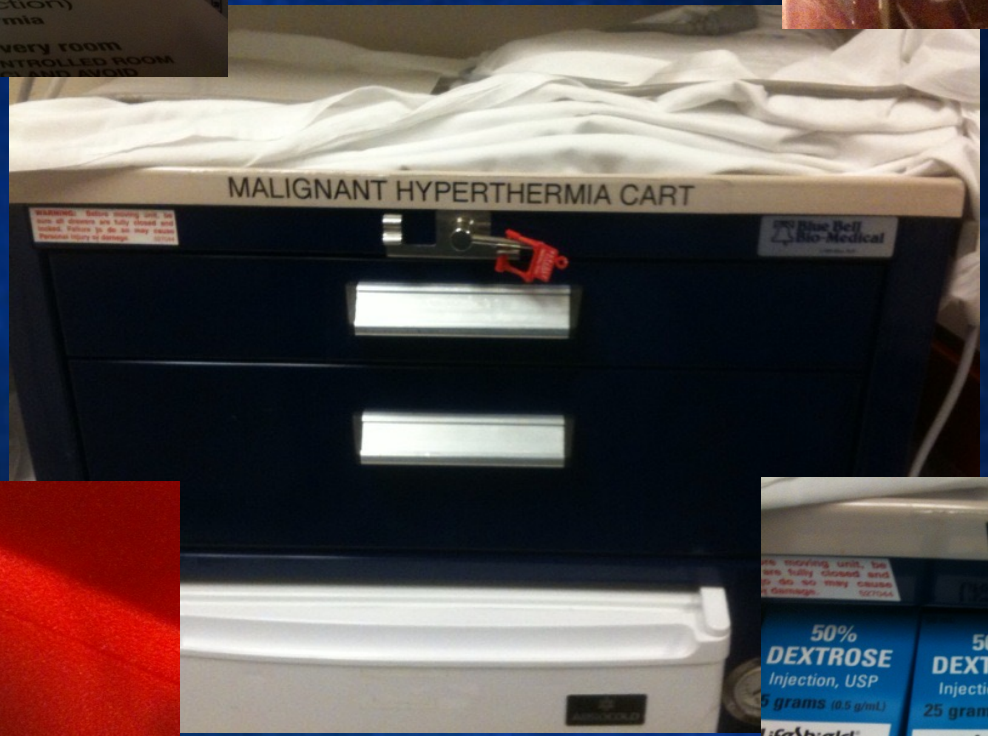
- Dantrolene:
  - Reduced Mortality from 80% 40 years ago, to 10% in current practice
  - Other studies have quoted reduced mortality 70% to between 1-17%
    - average of 1.4%
  - Delays in treatment lead to significant increases in complications
    - Dantrolene delayed 50 minutes can lead to 100% complication rate (renal dysfunction, DIC, coma, arrythmias, cardiac dysfunction, etc.)

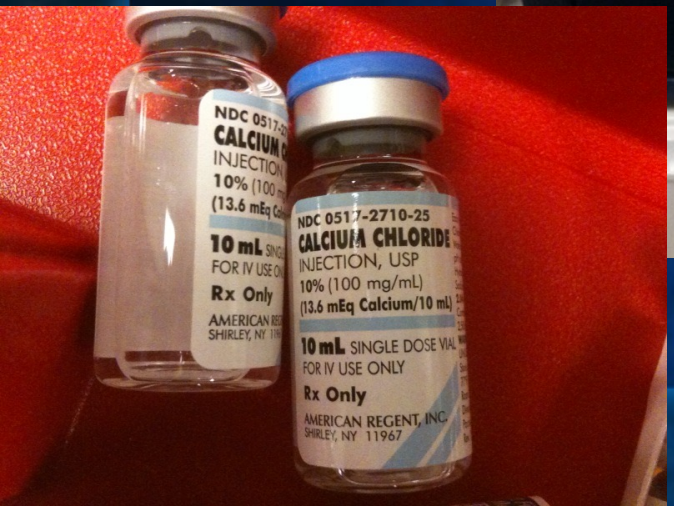
Treatment:

- Dantrolene: Cost
  - Cost of Dantrolene: 84 dollars/vial of basic formula.
    - $1008/year, assuming shelf life of 3 years
  - Does NOT include cost of other medications/supplies to treat MH
  - Does NOT include cost for maintaining MH Carts


LOT 142368
EXP 12/12
20 mg
Dantrium® Intravenou
(dantrolene sodium for injection
For treatment of malignant hyperthermia
FOR INTRAVENOUS USE ONLY
Store near operating room and recovery roc
STORE UNRECONSTITUTED PRODUCT AT CONTROLLED
TEMPERATURE (59°F TO 86°F OR 15°C TO 30°C) AND AVO
PROLONGED EXPOSURE TO LIGHT.
NDC 42023-123-06
20 mg
Dantrium® Intravenou
(dantrolene sodium for inject
For treatment of malignant hyper
For intravenous use only
Rx Only
20 mg
Dantrium® Intravenous
(dantrolene sodium for injection)
For treatment of malignant hyperthermia
FOR INTRAVENOUS USE ONLY
Store near operating room and recovery room


2 mL
Exp.
80-419-DK
1FEB2011
2 mL
FUROSEMIDE
20 mg (10 mg/
℞ only
Hospira, Inc.
Lake Forest, IL 60045 USA


MALIGNANT HYPERTHERMIA CART
Blue Bell Bio-Medical


NDC 0517-2710-25
CALCIUM CHLORIDE
INJECTION, USP
10% (100 mg/mL)
(13.6 mEq Calcium/10 mL)
10 mL SINGLE DOSE VIAL
FOR IV USE ONLY
Rx Only
AMERICAN REGENT, INC.
SHIRLEY, NY 11967


MALIGNANT HYPE
50%
DEXTROSE
Injection, USP
25 grams (0.5 g/mL)
LifeShield
Glass
ABBOJECT®
Unit of Use Syringe
with male luer lock
adapter and
18-Gauge protected needle
℞ only
Hospira
50%
DEXTROSE
Injection, USP
25 grams (0.5 g/mL)
LifeShield
Glass
ABBOJECT®
Unit of Use Syringe
with male luer lock
adapter and
18-Gauge protected needle
℞ only
Hospira
8.4% SODIUM
BICARBONATE
Injection, USP
50 mEq
(1 mEq/mL)
LifeShield
Glass
ABBOJECT®
Unit of Use Syringe
with male luer lock
adapter and 18-Gauge
protected needle
℞ only
Hospira
8.4% SODIUM
BICARBONATE
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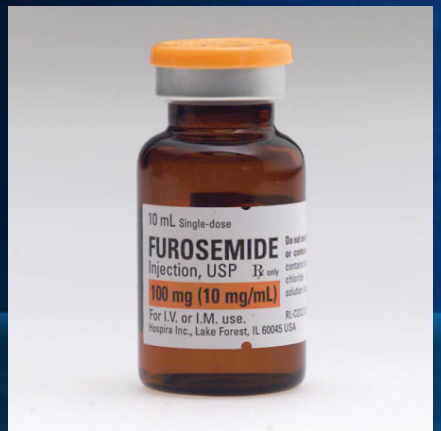
50 mEq
(1 mEq/mL)
LifeShield
Glass
ABBOJECT®
Unit of Use Syringe
with male luer lock
adapter and 18-Gauge
protected needle
℞ only

Treatment:

- Treat arrhythmias and hyperkalemia
  - Calcium gluconate
    - Stabilizes cardiac membrane potential
    - 2-3 ampules of calcium gluconate over 5 min or 1g $CaCl_2$ over 3 minutes
  - DO NOT GIVE CALCIUM CHANNEL BLOCKERS
  - Sodium Bicarbonate
    - Transfers K+ into cells
    - Maybe used in circumstances of severe metabolic acidosis
    - 50-100 meq over 5-10 min

Treatment:

- Treat arrhythmias and hyperkalemia
  - Glucose and Insulin
    - Insulin transfers K+ into cells
    - 10 units insulin IV with 50ml of 50% glucose solution
  - Other options: Diuretics (lasix), Beta-2 agonists, hemodialysis, etc.



Medications Used in Acute Treatment of Hyperkalemia

*Medication**	*Dosage*	*Onset*	*Length of effect*	*Mechanism of action*	*Cautions*
Calcium gluconate	10 to 20 mL of 10 percent solution IV over two to three minutes	Immediate	30 minutes	Protects myocardium from toxic effects of calcium; no effect on serum potassium level	Can worsen digoxin toxicity
Insulin	Regular insulin 10 units IV with 50 mL of 50 percent glucose	15 to 30 minutes	Two to six hours	Shifts potassium out of the vascular space and into the cells; no effect on total body potassium	Consider 5 percent dextrose solution infusion at 100 mL per hour to prevent hypoglycemia with repeated doses. Glucose unnecessary if blood sugar elevated above 250 mg per dL (13.9 mmol per L)
Albuterol (Ventolin)	10 to 20 mg by nebulizer over 10 minutes (use concentrated form, 5 mg per mL)	15 to 30 minutes	Two to three hours	Shifts potassium into the cells, additive to the effect of insulin; no effect on total body potassium	May cause a brief initial rise in serum potassium
Furosemide (Lasix)	20 to 40 mg IV, give with saline if volume depletion is a concern	15 minutes to one hour	Four hours	Increases renal excretion of potassium	Only effective if adequate renal response to loop diuretic
Sodium polystyrene sulfonate (Kayexalate)	Oral: 50 g in 30 mL of sorbitol solution Rectal: 50 g in a retention enema	One to two hours (rectal route is faster)	Four to six hours	Removes potassium from the gut in exchange for sodium	Sorbitol may be associated with bowel necrosis. May lead to sodium retention

When you gotta pretend you're awake at work but really ur just dead inside

Treatment:

- Cooling of patient
  - Severe increase in body temperature can be very dangerous
  - Cooling fans
  - Ice and ice water around head, groin area, axillae
  - Tell surgeon to flood field with cool solution
  - STOP at 38ºC to avoid hypothermia

Treatment:

- Transfer to ICU
  - Continue dantrolene therapy (1mg/kg q4-6hrs for 24-36hrs)
  - Monitor vital signs constantly
  - Lab tests: Basic metabolic panel, coagulation profile, CK levels, Liver function tests, urine output
  - Maintain urine output-diuretics, fluid
  - Watch for renal failure
  - Watch for muscle weakness
  - Watch for metabolic acidosis
  - Watch for DIC
  - As many as 25% of patients may experience a relapse within hours of episode
  - Mortality higher in patients with significant co-morbidities or larger muscle mass

MH Hotline
1-800-644-9737
Outside the US:
1-315-464-7079

Effective May 2008

EMERGENCY THERAPY FOR
MALIGNANT HYPERTHERMIA

DIAGNOSIS vs. ASSOCIATED PROBLEMS

Signs of MH:
- Increasing $ETCO_2$
- Trunk or total body rigidity
- Masseter spasm or trismus
- Tachycardia/tachypnea
- Mixed Respiratory and Metabolic Acidosis
- Increased temperature (may be late sign)
- Myoglobinuria

Sudden/Unexpected Cardiac Arrest in Young Patients:
- Presume hyperkalemia and initiate treatment (see #6)
- Measure CK, myoglobin, ABGs, until normalized
- Consider dantrolene
- Usually secondary to occult myopathy (e.g., muscular dystrophy)
- Resuscitation may be difficult and prolonged

Trismus or Masseter Spasm with Succinylcholine
- Early sign of MH in many patients
- If limb muscle rigidity, begin treatment with dantrolene
- For emergent procedures, continue with non-triggering agents, evaluate and monitor the patient, and consider dantrolene treatment
- Follow CK and urine myoglobin for 36 hours.
- Check CK immediately and at 6 hour intervals until returning to normal. Observe for dark or cola colored urine. If present, liberalize fluid intake and test for myoglobin
- Observe in PACU or ICU for at least 12 hours

ACUTE PHASE TREATMENT

1 GET HELP. GET DANTROLENE — Notify Surgeon
- Discontinue volatile agents and succinylcholine.
- Hyperventilate with 100% oxygen at flows of 10 L/min. or more.
- Halt the procedure as soon as possible; if emergent, continue with non-triggering anesthetic technique.
- Don't waste time changing the circle system and CO_2 absorbant.

2 Dantrolene 2.5 mg/kg rapidly IV through large-bore IV, if possible

To convert kg to lbs for amount of dantrolene, give patients 1 mg/lb (2.5 mg/kg approximates 1 mg/lb).
- Dissolve the 20 mg in each vial with at least 60 ml sterile, preservative-free water for injection. Prewarming (not to exceed 39° C.) the sterile water may expidite solublization of dantrolene. However, to date, there is no evidence that such warming improves clinical outcome.
- Repeat until signs of MH are reversed.
- Sometimes more than 10 mg/kg (up to 30 mg/kg) is necessary.
- Each 20 mg bottle has 3 gm mannitol for isotonicity. The pH of the solution is 9.

3 Bicarbonate for metabolic acidosis
- 1-2 mEq/kg if blood gas values are not yet available.

4 Cool the patient with core temperature >39°C, Lavage open body cavities, stomach, bladder, or rectum. Apply ice to surface. Infuse cold saline intravenously. Stop cooling if temp. <38°C and falling to prevent drift < 36°C.

5 Dysrhythmias usually respond to treatment of acidosis and hyperkalemia.
- Use standard drug therapy **except calcium channel blockers, which may cause hyperkalemia or cardiac arrest in the presence of dantrolene.**

6 Hyperkalemia — Treat with hyperventilation, bicarbonate, glucose/insulin, calcium.
- Bicarbonate 1-2 mEq/kg IV.
- For **pediatric,** 0.1 units insulin/kg and 1 ml/kg 50% glucose or for **adult,** 10 units regular insulin IV and 50 ml 50% glucose.
- Calcium chloride 10 mg/kg or calcium gluconate 10-50 mg/kg for life-threatening hyperkalemia.
- Check glucose levels hourly.

7 Follow $ETCO_2$, electrolytes, blood gases, CK, core temperature, urine output and color, coagulation studies. If CK and/or K+ rise more than transiently or urine output falls to less than 0.5 ml/kg/hr, induce diuresis to >1 ml/kg/hr and give bicarbonate to alkalanize urine to prevent myoglobinuria-induced renal failure. (See D below)
- Venous blood gas (e.g., femoral vein) values may document hypermetabolism better than arterial values.
- Central venous or PA monitoring as needed and record minute ventilation.
- Place Foley catheter and monitor urine output.

POST ACUTE PHASE

A Observe the patient in an ICU for at least 24 hours, due to the risk of recrudescence.

B Dantrolene 1 mg/kg q 4-6 hours or 0.25 mg/kg/hr by infusion for at least 24 hours. Further doses may be indicated.

C Follow vitals and labs as above (see #7)
- Frequent ABG as per clinical signs
- CK every 8-12 hours; less often as the values trend downward

D Follow urine myoglobin and institute therapy to prevent myoglobin precipitation in renal tubules and the subsequent development of Acute Renal Failure. CK levels above 10,000 IU/L is a presumptive sign of rhabdomyolysis and myoglobinuria. Follow standard intensive care therapy for acute rhabdomyolysis and myoglobinuria (urine output >2 ml/kg/hr by hydration and diuretics along with alkalinization of urine with Na-bicarbonate infusion with careful attention to both urine and serum pH values).

E Counsel the patient and family regarding MH and further precautions; refer them to MHAUS. Fill out and send in the Adverse Metabolic Reaction to Anesthesia (AMRA) form (www.mhreg.org) and send a letter to the patient and her/his physician. Refer patient to the nearest Biopsy Center for follow-up.

Non-Emergency Information
MHAUS
PO Box 1069 (11 East State Street)
Sherburne, NY 13460-1069
Phone
1-800-986-4287
(607-674-7901)
Fax
607-674-7910
Email
info@mhaus.org
Website
www.mhaus.org

Since 1981 MHAUS Malignant Hyperthermia Association of the United States
Dedicated to Patient Safety

CAUTION: This protocol may not apply to all patients; alter for specific needs.

ORPO 5/08/SK

Produced by the Malignant Hyperthermia Association of the United States (MHAUS). MHAUS is a non-profit organization under IRS-Code 501(c)3. It operates solely on contributed funds. All contributions are tax deductible. For more information, go to www.mhaus.org.

Treatment:

- Malignant Hyperthermia Hotline
  1-800-MH-HYPER
  (1-800-644-9737)

Prevention:

- History and family history
- Family awareness and testing
- Avoid triggering agents
- Use other anesthesia techniques
  - Nondepolarizing neuromuscular blocking drugs (rocuronim, vecuronium, etc), remifentanil, propofol, local anesthetics, TIVA

SAFE DRUGS	UNSAFE DRUGS
Antibiotics Antihistamines Barbiturates Benzodiazepines Droperidol Ketamine Local anesthetics Nitrous oxide Nondepolarizing neuromuscular blockers Opioids Propofol Propranolol Vasoactive drugs	All inhalation agents (except nitrous oxide) Succinylcholine

Prevention:

- Machine checkout
  - Tape vaporizers in OFF position
  - Flow 10L O_2 through machine for 20 minutes
  - New unused breathing circuit
  - Check with machine's manufacturer individually for specifications!
    - Ex: Drager Fabius requires 60 min of preparation

What YOU can DO:

- Be Present!
  - If you hear about an MH emergency, GO HELP
- Call the MH Hotline
- Get the MH Cart
- Prepare Ice to Cool Patient
- Get Sterile Water
- Help Mix and Prepare Dantrolene
- Preventive Tactics
  - Tape vaporizers
  - New Breathing Circuits
  - Call Anesthesia Machine company for recommendations

Post-Op:

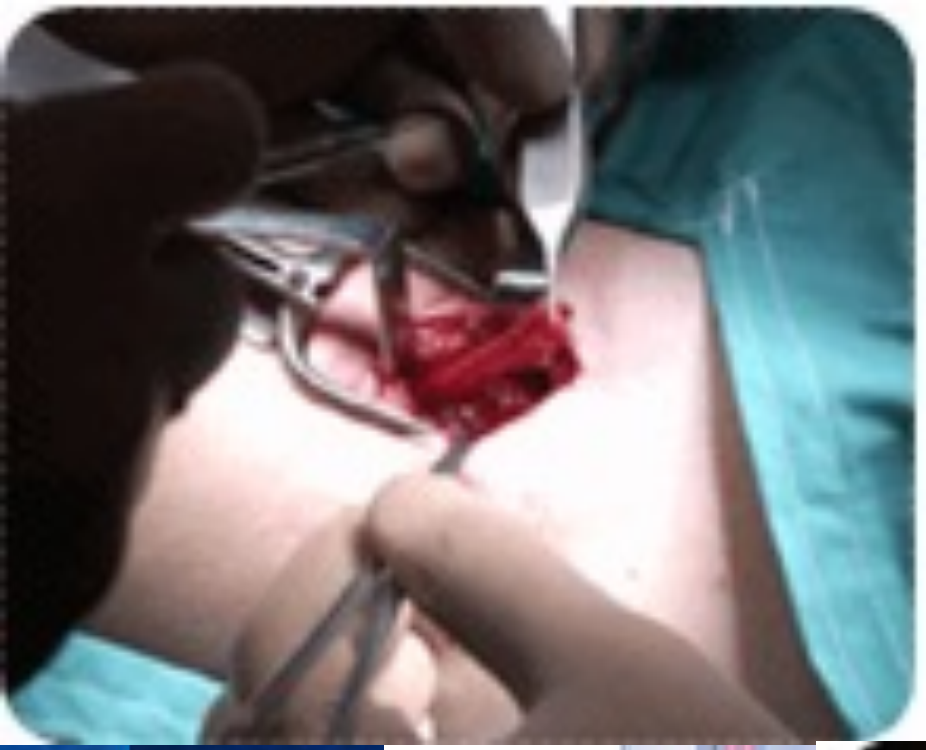
- Must be watched closely in PACU for 2.5 hrs after surgery
- If no complications, can be discharged home

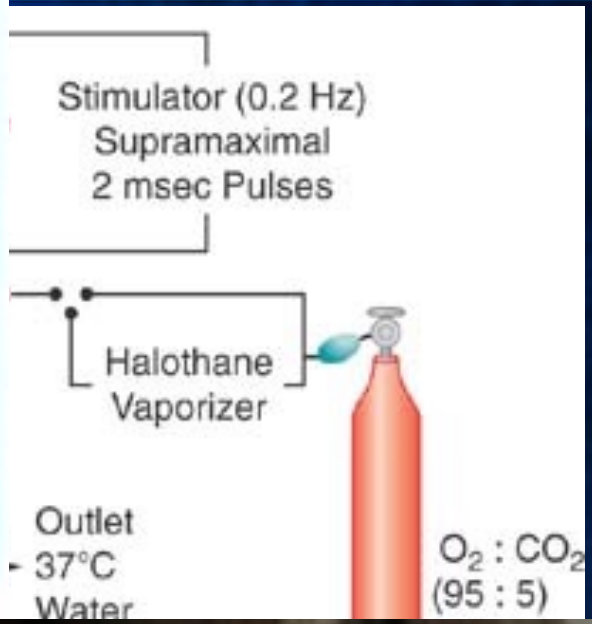
Testing:

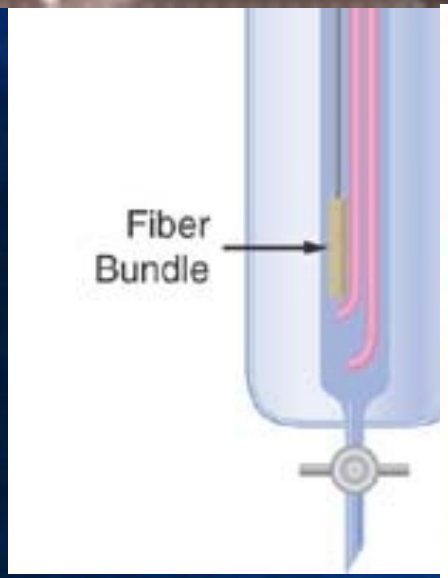
- Contracture testing:
  - Gold standard
  - Can establish a definitive diagnosis
  - Sensitivity 100%
  - Specificity 80%

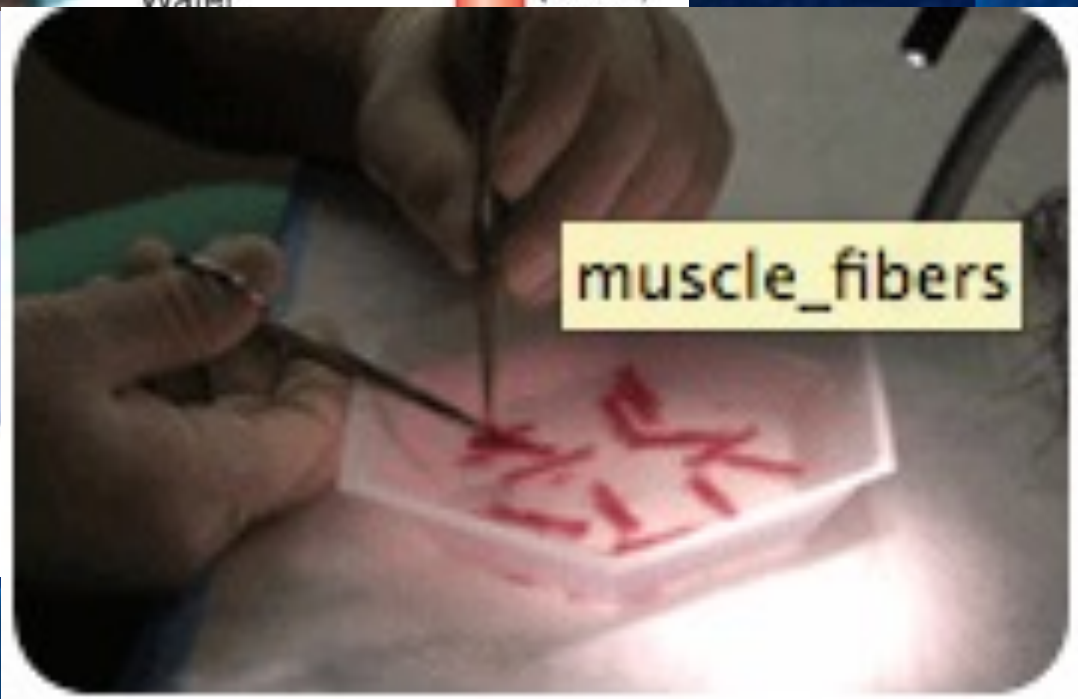
Testing:

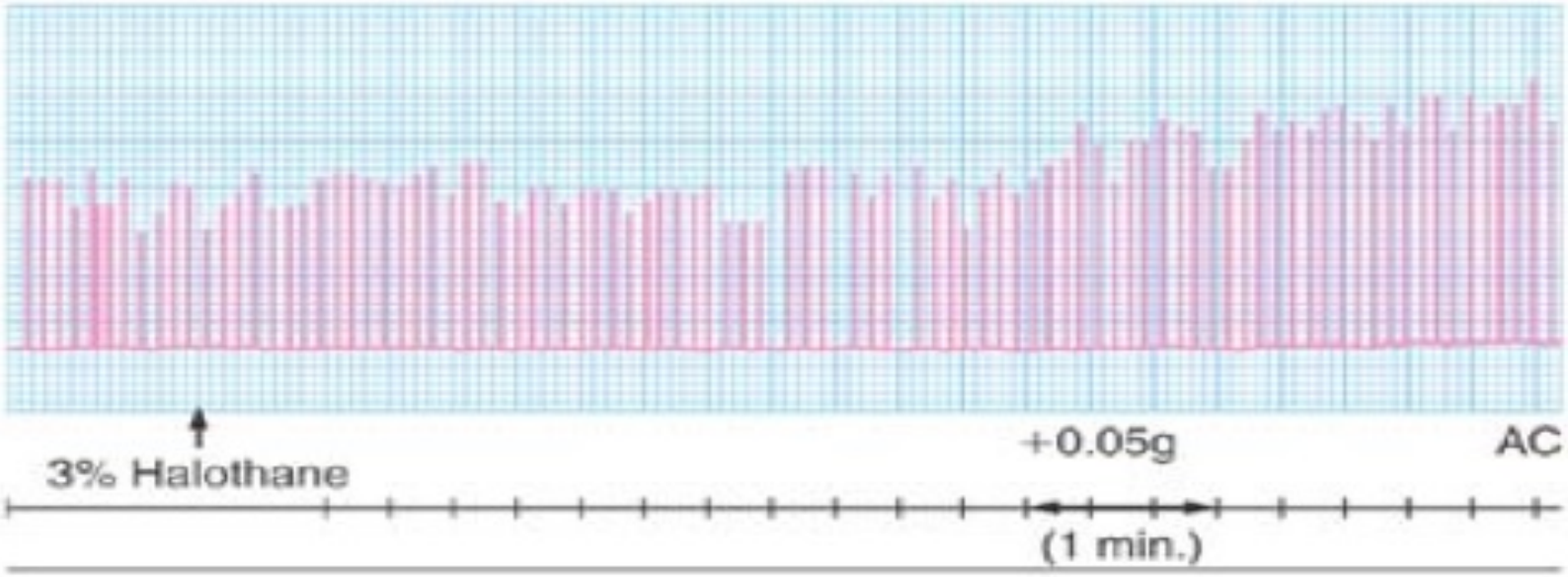
- Caffeine Halothane Contracture Test:
  - Exposure of a FRESH muscle biopsy specimen
    - caffeine
    - halothane
  - Produces contractures at lower thresholds than non-MH individuals



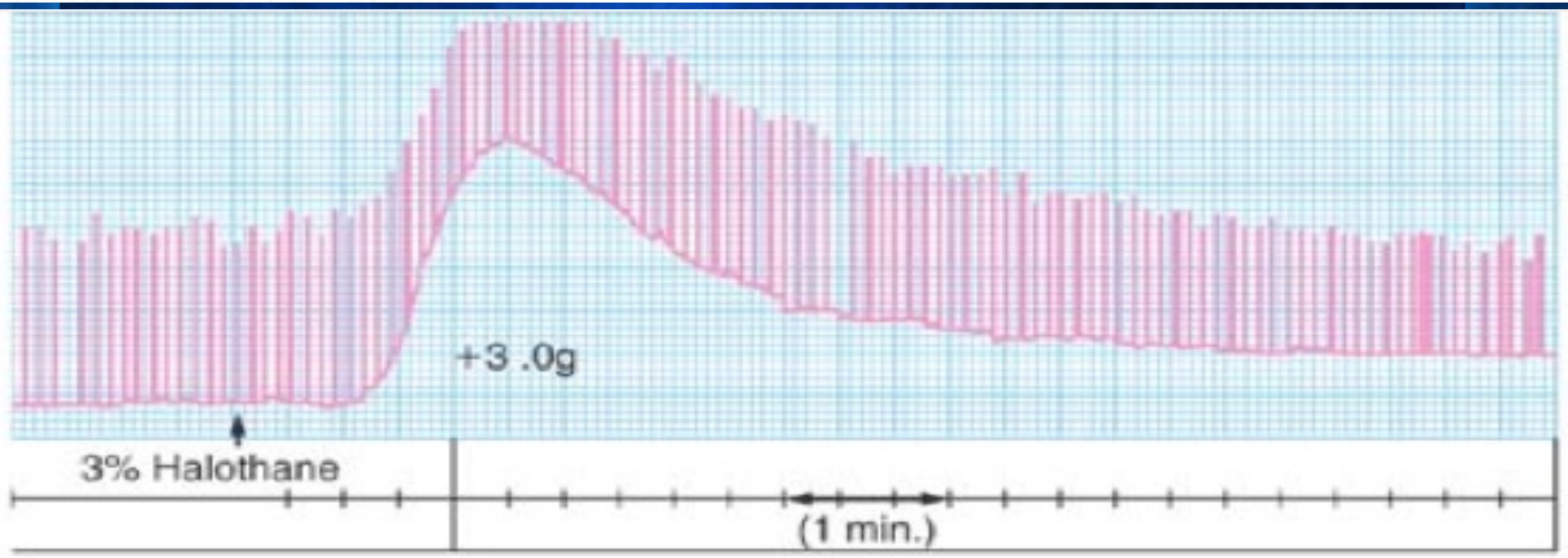

Stimulator (0.2 Hz)
Supramaximal
2 msec Pulses
Halothane
Vaporizer
Outlet
37°C
Water
O_2 : CO_2
(95 : 5)


Fiber
Bundle


muscle_fibers



A Normal Halothane Contracture



Abnormal Halothane Contracture

Testing:

- Caffeine Halothane Contracture Test
  - Costly
    - Insurance will sometimes pay
    - Sometimes will not
    - (6-10,000 U.S.dollars)
  - Requires 2-7 days from work
  - Even if test is negative, caution must be used
  - Requires a FRESH sample
  - Can be done at few centers around the U.S. and Canada

Testing:

- Test Centers:
  - University of Minnesota- Minneapolis, MN
  - University of California- Davis, CA
  - Wake Forest University- Winston-Salem, NC
  - Uniformed Services University of the Health Sciences- Bethesda, MD
  - The Ottawa Hospital - Civic Campus- Ottawa Ontario
  - Thomas Jefferson University- Philadelphia, PA
  - MHAUS- Sherburne, NY

Testing:

- Genetic Testing:
  - Presence of causative mutation in RYR1 gene is diagnostic for MH
  - Less invasive
  - Less costly—but STILL 4-8 thousand U.S. dollars
  - Less traveling
  - Contracture test might still be needed due to heterogeneous nature of MH gene
  - Limited due to incomplete genetic panel of all causative mutations for MH

Summary:

- Malignant Hyperthermia is a rare but potentially fatal disease in the OR
- Early signs and symptoms (hypercapnia, tachycardia, rigidity) may go unnoticed initially, but suspicions should be high if seen
- Treatment should be quick and thorough to prevent severe damage to patient
- If a MH patient presents, be aware of other possibilities for anesthetic choices
- Family counseling and testing are options to identify and prevent future complications with MH
- Overall, VIGILENCE!!!

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Surgeons be like

Questions?


I will not ask dumb questions
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