Brockville General Hospital

Policy Name:	Malignant Hyperthermia				**Number: CLIN-003**	
Manual:	General Clinical Policies	**Category:**	**C**linical			
Issued By:	VP Clinical Services/CNE					
Approved By:	C Cassidy-Gifford (Cathy Cassidy-Gifford)					
Applies to:	Dept Only	All Sites	✓CSS	GSS	FARC	McDougall
Original Date:	January 2005					
Last Reviewed or Revised Date:	October 2008. April 2016					
Retired Date:						

POLICY STATEMENT: to provide guidelines when caring for a patient with a known or suspected case of Malignant Hyperthermia(MH). Malignant Hyperthermia is a patient emergency. Recognition and immediate action are extremely important.

POLICY:

1. All patients who will receive general anesthesia must be screened for a history or family history of Malignant Hyperthermia (MH), or diseases related to MH such as muscular dystrophy.
2. Include family and support persons in preoperative teaching /instructions (e.g. Dantrolene, medical alert bracelet, potential for muscle testing, need for ongoing post-operative observation, familial/genetic testing)
3. Malignant hyperthermia is a potentially fatal, inherited disorder usually associated with the administration of certain general anesthetics and /or the drug succinylcholine.
4. MH can also be a response to extreme physical or emotional stress
5. The general signs of MH include:
    a) Muscle rigidity, masseter muscle rigidity (jaws of steel)
    b) Increase of end tidal C02
    c) Tachycardia/ tachypnea
    d) Arrhythmias
    e) Mixed respiratory and metabolic acidosis
    f) Temperature elevation (may exceed 40° C)
    g) Myoglobinuria from muscle breakdown
    h) Hyperkalemia
    i) hyper metabolism
    j) cyanosis or mottling of skin
6. Volatile triggering anesthetic inhalation agents include:
    a) Halothatne
    b) Isoflurane
    c) Sevoflurane
    d) Desflurane
7. When succinylcholine is combined with a volatile triggering anesthetic inhalation agent the chances of an MH crisis increases.

ALL STAFF IN OR/PAC AND ICU MUSTREVIEW THE CONTENTS OF THE MH CART ON A PRN BASIS, RECOGNIZE EARLY SIGNS AND SYMPTOMS OF MH, AND MAITAIN FAMILIARITY WITH THE TREATMENT PROTOCOL FOR MH. The **MH** education module is accessed on the BGH SharePoint site.

MH Cart locations:

ICU ground floor (between step-down and ICU beds)
Surgical suites in center core

PROCEDURE:

MANAGEMENT OF THE MH SUSCEPTIBLE PATIENT

In order to be prepared for the development of MH in a susceptible patient it is essential that:

1. The MH carts are checked and stocked on a regular basis for outdates and adequate vials of Dantrolene (36 vials), and IV solutions.
2. The anesthesiologist uses non triggering anesthetic agents. A spinal or epidural is the anesthetic of choice for the obstetrical patient.
3. The circulating nurse makes certain the MH cart and any additional supplies required are readily available.
4. The anesthesiologist ensures continuous monitoring of the patients temperature during the case. Standard monitoring procedures, including end tidal C02 are employed.
5. MH susceptible patients may be discharged home when the criteria for discharge for that area are met, and the minimum of 4 hours of observation and monitoring has been completed.
6. MH susceptible patients who undergo surgery in the OR and/or who are recovered in the PAR or ICU, REGARDLESS OF ANESTHETHETIC TECHNIQUE (INCLUDING LOCAL ANESTHETICS) MUST BE KEPT and monitored for a minimum of 4 hours post-surgery as follows:
    - Q 15 minutes x 1 hour then
    - Q30 minutes x 1 hour then
    - Q 1 hour x 2 hours
    - Blood glucose must be monitored q 2 h
    - Assess for hyperkalemia and electrolyte imbalances.

Post anesthetic monitoring for MH susceptible patients guidelines

DAY SURGERY: patients may be discharged to the community after 4 hours of observation if:

1. The patient meets discharge criteria
2. Vital signs stable including tympanic temperature, not >37.5° C.
3. Urine checked for myoglobinuria (cola or tea colored)
4. Patient assessed/ cleared by anesthesia.

SAME DAY ADMIT: patients may be discharged to ward after 4 hours in a monitored environment if:

1. The patient meets PACU discharge criteria
2. Vital signs are stable including tympanic temperature, not >37.5° C.
3. Urine checked for myoglobinuria (cola or tea colored)
4. Patient assessed/ cleared by anesthesia.

Obstetric patients and infants must be observed closely for at least 24 hours post-delivery for signs of MH.

Patients must remain in a monitored environment (ICU or step down) for at least 24-48 hours if:

1. Following a MH crisis
2. Myoglobinuria suspected or present
3. Temp >37.5° C tympanic and/or pulse, BP change >20% from pre-anesthetic baseline.
4. Anesthesiologist is to be notified if Temp >37.5° C tympanic and/or pulse, BP change >20% from pre-anesthetic baseline

Management of MH Crisis

Initial treatment

1. Call for help, obtain the MH cart and resuscitative equipment.
2. Notify surgeon to halt procedure and discontinue the triggering agents
3. If surgery must be continued, maintain general anesthesia with IV non triggering anesthetics and non-depolarizing neuromuscular blockers as needed.
4. Hyperventilate the patient with 100% oxygen at high gas flows, at least 10L/ minute to flush volatile anesthetics and lower ETC02.
5. Prepare and administer rapidly, Dantrolene (Dantrium) 2.5mg/kg bolus IV. In obstetrical patient, once the baby has been removed from the uterus, the Dantrolene may be given.
6. Obtain baseline blood work:
    - CBC
    - Electrolytes
    - Blood sugar
    - CK
    - Arterial blood gases
    - Urine and serum myoglobin
    - Coagulation profile
    - Calcium
    - Phosphorus

    Recognize and report any deviations.
7. Monitor core temperature and initiate cooling of patient in conjunction with
    - Cold IV saline lavage of the stomach, rectum, vagina, and open body cavities, as required
    - Surface cooling with hypothermia blanket and ice packs to the axilla and groin as ordered.
    - implement protective measures to prevent skin/tissue injury due to thermal sources
    - assess skin color, temperature and diaphoresis
    - stop cooling if temp <38° C

    N.B. Excessive and sudden cooling can cause severe shivering which precipitates ventricular tachycardia.

8. Treat acidosis with sodium bicarbonate (1-2 meq/kg) according to the ABG results.

9. a) Move the patient as little as possible. Moving the patient can precipitate ventricular tachycardia.
10. Treat hyperkalemia with IV glucose and insulin (10u Regular Insulin and 50mls of 50% Dextrose).
    Calcium Chloride (2-5mg/kg) may also be used to treat life threatening hyperkalemia. Note BS must be monitored q2hr.
11. Cardiac dysrhythmias may respond to treatment of acidosis and hyperkalemia, but may require treatment with anti-dysrhythmias. Note **AVOID** calcium channel blockers.
12. Ensure adequate urine output with administration of IV fluids and diuretics such as mannitol and/or Lasix. Monitor intake and output strictly.
13. Insert invasive monitoring equipment: CVP, arterial line, pulmonary catheter if necessary.
14. Continuously monitor end tidal CO2, HR with cardiac monitor, Resps with O2 sat, core temperature, BP and muscle tone.

POST –ACUTE PHASE MANAGEMENT IN ICU

The plan of therapy for the patient includes the following:

1. Transfer the patient to ICU for 24-48 hrs.
2. Administer Dantrolene sodium 1mg/kg IV q4-6 h for 24-48 hours following the acute stage.
3. Monitor ABG's, electrolytes, CK, Ca++, urine and serum myoglobin, PTT, PT, INR, q6h for 24-48 hours.
4. Monitor core temperature continuously until the patient is stable.
5. Watch for MH relapse by evaluating the patient at least every 4 hours for the first 36 hours after an MH event.
6. Advise possible post-dantrolene therapy symptoms (i.e., nausea, diarrhea, muscle weakness, double vision, dizziness or light headedness).
7. Educate the family and the patient

There is a **Canadian MH Association 1-416-340-3238**
There is a **MH Association in the USA 1-800-644-9737**

DANTROLENE SODIUM (DANTRIUM)

Notify the pharmacy services (extension 1136) that Dantrium was required.

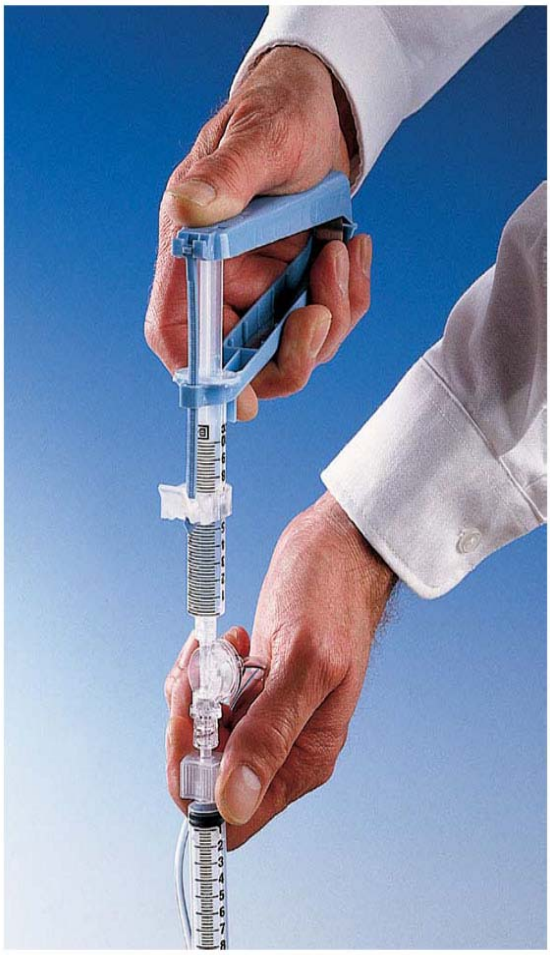
The Dantrium blocks the ongoing release of Calcium from the storage sites (sarcoplasmic) in skeletal muscles cells. The commercial preparation consists of:

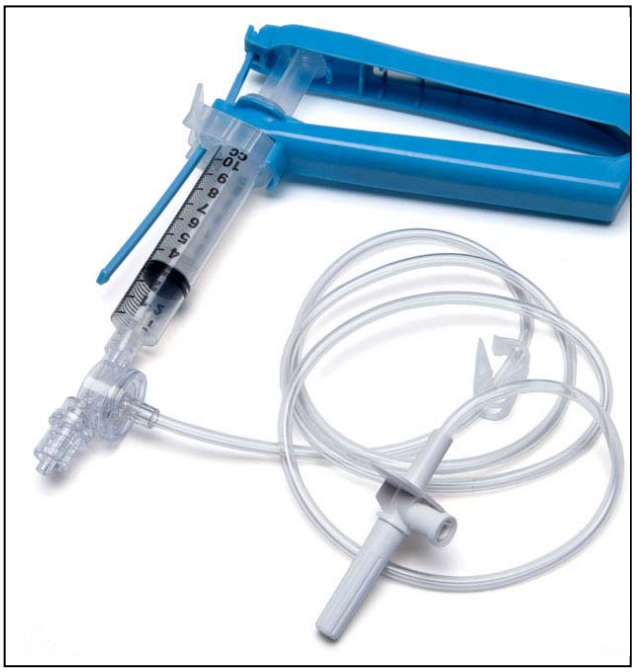
- Dantrolene sodium 20mg
- Mannitol 3000mg
- Sodium hydroxide (to yield a Ph greater than 9.5 that if interstitial can cause local tissue damage.

PREPARATION OF DANTROLENE SODIUM

Dantrolene sodium is provided in a 20mg vial that is reconstituted with **STERILE WATER FOR INJECTION, RED LABELED BAG (NO BACTERIOSTATIC AGENT)** as follows:

1. Remove the aluminum cap from the dantrolene sodium vials.
2. Place vented 18 G needle on a **cornwall syringe** (see below) and draw up 60 ml of sterile water from the RED LABELED 500 ml sterile water bag for injection. The cornwall syringe has a 10 cc syringe on it, so you must pull up 10 cc's at a time, 6 times per vial.
3. Add the 60ml of sterile water to the vial. ENSURE VENTED NEEDLE SLEEVE ENTERS THE VIAL DAM. At least 2 Nurses need to mix up dantrolene as it is timely process.
4. Remove the syringe and recap using single-handed technique.
5. Shake the dantrolene sodium vial vigorously until the solution appears clear. (yellow /orange)
6. Draw up the reconstituted dantrolene sodium into a 60ml syringe ensuring that the vented needle sleeve enters the vial dam.
7. Change to a non-vented needle before handing the syringe to the physician for administration.





Administration of Dantrolene Sodium

The physician administers an initial dose of 1-3mg/kg by rapid IV bolus. This is repeated until signs of malignant hyperthermia subside or to a **maximum cumulative dose of 10mg/kg.** The anesthesiologist is provided with sterile water for this purpose.

Example: For a 60 kg patient at an initial dose of 3mg/kg

Amount of Drug required
Weight 60 kg **x dose per kg** 3mg/kg =180 mg (total 9 vials)

Note:

- Protect the reconstituted solution from direct light (drape the vials)
- The reconstituted solution is stable for 6 hours at room temperature
- The solution is not compatible with 5DW or NS for reconstitution. It can be administered through an IV with NS but not LR.

REFERENCES:

AORN Standard, Recommended Practices and Guidelines 2015, Malignant Hyperthermia Association of the United States (MHAUS), http://www.mhaus.org/

Dantrium Intravenous product Monograph , Jan. 22, 2009

Effective February 2015

Emergency Therapy for MALIGNANT HYPERTHERMIA

MH Hotline: 1-800-644-9737 • Outside of the US: 001-209-417-3722

DIAGNOSIS

Signs of MH:

- Increasing $ETCO_2$ *(despite hyperventilation)*
- Trunk or total body rigidity
- Masseter spasm or trismus
- Tachycardia/tachypnea
- Mixed respiratory and metabolic acidosis *(MH can occur without significant metabolic acidosis)*
- Increased temperature (may be an early or a late sign)
- Myoglobinuria

Sudden/Unexpected Cardiac Arrest in Young Male Patients:

- Presume hyperkalemia and initiate treatment *(see #6)*
- Measure blood gases and electrolytes
- Measure CK, myoglobin, ABGs, until normalized
- Usually secondary to occult myopathy *(e.g., muscular dystrophy)*
- Resuscitation may be difficult and prolonged
- Myoglobinuria is common

Trismus or Masseter Spasm with Succinylcholine

- Early sign of MH in many patients
- If limb muscle rigidity, begin treatment with dantrolene.
- For emergency procedures, continue with non-triggering agents, evaluate and monitor the patient, and consider dantrolene treatment.
- Check CK immediately and at 6-8 hr intervals until returning to normal. Observe for dark or cola-colored urine. If present, liberalize fluid intake and test for serum and urine myoglobin. *(see D below)*
- Observe in PACU or ICU for at least 24 hours if metabolic signs of MH were present.

ACUTE PHASE TREATMENT

1. GET HELP. GET DANTROLENE.
Notify Surgeon. Call MH Hotline.

- Discontinue volatile agents and succinylcholine.
- Hyperventilate with 100% oxygen at flows of 10 L/min. to flush volatile anesthetics and lower $ETCO_2$. If available insert activated charcoal filters into the inspiratory and expiratory limbs of the breathing circuit. The Vapor-Clean™ filter may become saturated after one hour; therefore, a replacement set of filters should be substituted after each hour of use.
- Halt the procedure as soon as possible; if it is not possible to stop surgery, continue with non-triggering anesthetic technique.
- Don't waste time changing the circle system and CO_2 absorbent.

2. Dantrium®/Revonto®/Ryanodex®
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).

- Dantrium/Revonto – Each 20 mg vial should be reconstituted with at least 60 mL sterile water for injection, USP (without a bacteriostatic agent). There are 3 grams of mannitol in each 20 mg vial of Dantrium and Revonto.
- Ryanodex – Each 250 mg vial should be reconstituted with 5 mL sterile water for injection, USP (without a bacteriostatic agent) and shaken to ensure an orange-colored uniform, opaque suspension. There are 125 mg of mannitol in each 250 mg vial of Ryanodex.
- Repeat until signs of MH are reversed.
- Sometimes more than 10 mg/kg *(up to 30 mg/kg)* of dantrolene is necessary.

3. Bicarbonate for metabolic acidosis

- 1-2 mEq/kg if blood gas values are not yet available

4. Cool the patient

- If core temperature > 39°C Apply ice to surface.
- Infuse cold saline intravenously.
- Lavage open body cavities.
- Other cooling techniques may be applied at clinician's discretion.
- Stop cooling if temperature < 38°C and falling to prevent hypothermia.

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

Continued on other side...

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ACUTE PHASE TREATMENT Continued

5. Dysrhythmias

Usually respond to treatment of acidosis and hyperkalemia.

- Use standard drug therapy **EXCEPT avoid calcium channel blockers — (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 regular insulin/kg and 2 mL/kg 25% dextrose or for adult,10 units regular insulin IV and 50 mL 50% dextrose
- Calcium chloride 10 mg/kg IV or calcium gluconate 10-50 mg/kg IV for life-threatening hyperkalemia
- Check glucose levels hourly.

7. Follow...

$ETCO_2$, minute ventilation 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 alkalinize urine and prevent myoglobinuria-induced renal failure *(see D below).*

- Venous blood gas *(e.g., femoral vein)* values may document hypermetabolism earlier than arterial values.
- Central venous or PA monitoring as needed.
- Place Foley catheter and monitor urine output.

POST ACUTE PHASE

A. Watch for MH relapse by continuously evaluating the patient for at least 24 hours following cessation of signs of MH. 25% of MH events relapse, which can be fatal. Treat immediately if relapse occurs. Signs of MH relapse include:

- Increasing muscular rigidity in the absence of shivering
- Inappropriate hypercarbia with respiratory acidosis
- Metabolic acidosis without other cause
- Inappropriate temperature rise

B. Give dantrolene, 1mg/kg IV q 4-6h or 0.25mg/kg/hr by infusion and continue for at least 24 hr and sometimes longer as clinically indicated.

Dantrolene can be stopped, or the interval between doses increased to q8h or q12h if all of the following criteria are met:

- Metabolic stability for 24 hours
- Core temp is less that 38°C
- CK is decreasing
- No evidence of myoglobinuria
- Muscle is no longer rigid

C. Follow vital signs and labs as above (see #7)

- Frequent blood gases as per clinical signs
- CK every 6 hours; less often as the values trend downward

D. Follow urine myoglobin and institute therapy to prevent myoglobin 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 by hydration and diuretics (urine output >2 mL/kg/hr along with alkalinization of urine with No-bicarbonate infusion and 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/registry) and send a letter to the patient and her/his physician. Refer patient to the North American MH Registry and the nearest Biopsy Center for follow-up.

NON-EMERGENCY INFORMATION

MHAUS
1 North Main Street
PO Box 1069
Sherburne, NY 13460-1069

Phone: 1-800-986-4287
(607) 674-7901
Fax: (607) 674-7910
Email: info@mhaus.org **Website:** www.mhaus.org

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