An MH-Susceptible Shares Her Experience As A Potential Kidney Donor

by Barbara Matulionis

Four years ago, I became interested in donating a kidney to someone that I do not know. I contacted a local hospital in Cincinnati and proceeded with the work-up. In the end, I was declined as a donor due to having a strong family history of Malignant Hyperthermia. Unless I could prove that I was not MH susceptible, I would not be given the opportunity to donate. I was disappointed because I went through with nearly all of the testing. Additionally, I have had many surgeries in the past using non-triggering anesthesia. I believed I could safely have a kidney removed.

This was the impetus for my sister and I to get tested for MH. While being turned down as a potential kidney donor was the initiating event to start the process, we really wanted to know our status so we could, hopefully, clear our children of this condition. We knew we each had a 25% chance of being susceptible.

Using airline and hotel points, we went to Wake Forest Baptist in Winston Salem, NC, to have the muscle biopsies done. It turned out to be a fun and memorable trip. My sister had her testing done one day and mine was done the next. The medical care we received was outstanding. It was painful afterward, but since it was muscle pain, it was very tolerable. But we did use the wheelchair services at the airport to save walking!

Unfortunately, the results of the biopsies showed that we are both susceptible to MH. We were not able to clear our children, but we are very glad we had the testing done. I let the idea of kidney donation go, figuring it wasn’t meant to be.

Recently, though, I saw a news story about a woman in Cincinnati who was in need of a kidney. This re-inspired me to try again. My sister and I had some communication with (MH Hotline Consultant and MH expert) Dr. Joseph Tobin early in our testing process, so I reached out to him and asked if he was willing to write a letter of support.

After consulting the group of anesthesiologists at MHAUS, he wrote a brilliant letter that was educationally informative and indicated support by MHAUS that I should not be declined as a potential kidney donor due to being susceptible to MH.

I researched other transplant centers within driving distance from my home and made a decision. After doing the initial paperwork and labs, I drove to Columbus, OH, to meet with anesthesia, support letter in hand. I was approved to continue in the evaluation process. I left the center feeling confident that the medical professionals understood the condition and that I would be in good hands should I be approved to donate my kidney.

I am extremely grateful to Dr. Tobin for his support. I have nearly completed the donor evaluation and hope to donate my kidney in the near future.
Executive’s Corner ...

Better to have, and not need, than to need, and not have
– Franz Kafka

March was “MH Awareness Month,” and I was reminded of MHAUS’ internal focus on MH awareness and what that means to us – and to those who might unexpectedly need to react to an MH event and the lethal ramifications if healthcare providers are not prepared for it. Are you prepared for MH? This is the question we are here to help you address.

Just as many of us have “extra” items at home for those times we forget to get them at the store, healthcare facilities must have the tools on hand to be “ready” for an MH event. This includes an algorithm poster on the wall for all to use as well as the necessary drugs and equipment that can make all the difference in the final outcome. Where is your MH cart? Do you have the appropriate amount of the antidote, dantrolene sodium for injection, on hand? Are external cooling options/methods available? Is the MH Hotline number posted in every operating area? Most important of all: is your team trained to react?

So, you ask, what is MHAUS doing to help me in this regard? As a small non-profit, blessed with dedicated and selfless individuals, we focus on being there for you. We have an MH Hotline available 24/7/365, armed with doctors who are experts in the field of malignant hyperthermia. They are the needed cool head, when your team is frantically reacting to MH.

For general questions, we offer a contact form on our website at http://my.mhaus.org/?page=AskMHExperts where you can ask questions and usually receive a reply within 48 hours. Typing in words or questions in the “Search” feature at the top of the home page can also gain additional resources for basic research. This will lead you to a variety of available resources to help find the answers you seek.

More recently, we have listened to your comments about what you need to do a better job and, as a result, we now provide the 2018 updated version of the MH In-service available to you as a 2-year online subscription to be viewed, once you have completed the login process to activate the subscription, wherever you are and as many times as you like. The entire team can watch it all at once on an internal large screen (via the Internet) to address your MH preparedness plan. Providing it in this format allows us to make any necessary revisions directly on the website quickly. The purchaser can be confident the information is correct and up-to-date. If preferred, the subscription can also include a DVD of the presentation at a small additional fee. Also provided is a list of additional resources. A test to verify proper assimilation of the material by the team is available on the MHAUS website.

What about your MH cart? Do you have the appropriate items available? Is there a checklist to use to assure it is ready? There is now! The MHAUS staff kept hearing about the need for a checklist that could be attached to the cart to assure there is nothing missing when an MH event strikes, and you wanted us to help. Reminded of the saying; REMEMBER; WHEN DISASTER STRIKES, THE TIME TO PREPARE HAS PASSED (Steven Cyros), we developed our newest product, the MH Cart Cards, to provide just what our customers needed. A more thorough description can be found on our website at https://mhaus.site-ym.com/store/ViewProduct.aspx?ID=10458471. The orders for this item are vigorous and the feedback has been positive. The cards are laminated, easy to read, easy to attach to the cart and fill this need very nicely.

We provide online webinars designed to answer questions relating to MH: what it is, how to be ready for it and how it affects patients. A webinar can address patients’ personal experiences and challenges evolving from their MH susceptibility and also involve all types of healthcare professionals who are chartered with their care. Goals have been set for the development of more webinars in coming months. One new idea is for a webinar to clarify questions surrounding the proper use of specific equipment needed in an MH event. This will be specifically designed for
healthcare professionals. We will be trying to put it into place with participation from the equipment manufacturers as well as our MH experts. We keep hearing from MH-susceptible (MHS) patients and their family members that the topic of muscle cramping and other “awake symptoms” are huge concerns. Is this true? If so, we can arrange a webinar that is set up as a panel of MH experts to listen to the questions/concerns posed by participants and then respond with their insight and perspective. Considering there is not a lot of clinical data for the questions being asked, this option would give MHAUS a forum to hear stories and personal experiences from MHS patients and caregivers directly. The top concerns can then be addressed and, if there is no specific answer during the webinar, we will try our best to find resources that might help after the webinar and will share them with you. To assure the audience is interested in this option and at least 100 strong, we must hear from you. If you want it badly enough, we will provide it. Go to this link at https://mhaus.site-ym.com/general/?type=emailpatientwebinar and give us your email address, first and last name. We will coordinate and arrange the webinar on our end once we have enough interest to move forward. Feel free to share the link with others you know will be interested via Facebook, Twitter or any other social media outlets you may use. Help us get you the answers/insight you need.

What about the MHAUS Recommendations? Do you know the various regulatory agencies have been known to ask whether your facility is following? Have you? Are you prepared to ask whether your facility is following regulatory agencies have been known to pose for over a year to develop additional recommendations designed to address the regularly asked questions and concerns they hear. You will find more recommendations are being added every few years for your team’s MH education and insight.

So, you ask, are we looking for new or improved products for the future? The answer is yes, we are! Many of you are using the MH Mock Drill Kit to set up your yearly or twice a year MH Mock Drill to assure you are ready for a possible MH disaster. Regulatory agencies are asking whether you have completed an MH Mock Drill from what we are hearing, so be ready for that question by putting the drill in your yearly team training plan. Some are even adding an MH Mock Drill Call to their practice run in order to get the most realistic experience they can design for their team. In your planning stages, we encourage you to give the MHAUS administrative office at least two weeks to coordinate an MH Mock Drill Call. There is a lot of internal management that needs to happen in order to assure the call goes off without a hitch, and please remember we need time to provide the best experience possible.

The MH Mock Drill Kit itself is beginning to go through updates and revisions for the next version. We will provide this product as an online 2-year subscription to improve the ease of access for the team. Just like the In-service, once the account is set up, the program can be viewed from anywhere at any time via the Internet and will remain up-to-date through the ease of use of this medium. We will also provide, for an additional fee, a DVD of the presentation. Look for it to be available by mid to late summer.

Watch the MHAUS website for updates and events as the year progresses. I hope you all are sharing pages you find helpful and retweeting as well! We rely on our members to help us spread the word about the need for MH preparedness and education across the world. If you are talking about it to someone, you are giving them an opportunity to learn something new that day and to gain a bit more insight into the depth of this particular uncommon disorder.

BE SURE YOU ARE READY FOR MH; AND YOU WON’T BE SORRY!
Available from Par Sterile Products, LLC

For more information on Dantrium® IV and for Full Prescribing Information please visit parsterileproducts.com
If you swim, ride bike, run, walk, do-yoga, or climb mountains – literally any kind of physical activity – we’re asking you to help raise awareness and funds for MHAUS this summer by participating in our “Exercise for MH” summer event.

You can choose to take donations or simply participate to help raise awareness of MH. You choose how much or how little you want to participate. For example, you can dedicate just one day in June to “Exercise for MH” or you can do it every day of the month to spread the word about MH. Maybe you want to take things a step further and run that first marathon of your life, or hit the century mark on your bicycle.

A specific summer month is not necessary. The important thing is to get involved! Tell your family, friends, coworkers, customers, everybody, what you are doing and get them to register for the “Exercise for MH” summer event. You can ask for donations from your community, family, and friends, or just make a donation yourself.

Together, we can help save lives by raising awareness for MH. All proceeds will go towards funding the important work of MHAUS. When you donate to MHAUS, you are providing the resources needed to promote optimum care and scientific understanding of Malignant Hyperthermia and related disorders. Donations have helped to reduce morbidity attributed to MH from greater than 85 percent in 1981 to less than five percent today. But there’s still more to do.

Donations support calls to the 24-hour MH Hotline, continuation of research by medical professionals using the North American MH Registry of MHAUS’ database, papers written by medical professionals affiliated with MHAUS, MH information shared with attendees at medical meetings, and MHAUS-sponsored scientific conferences. These are just some of the ways we put your donation dollars to work.

So what do you say? Gather some friends, family, and co-workers and get out there.

MHAUS Will Help Promote Your MH Activity

You can help build awareness about Malignant Hyperthermia by sharing information about the activities related to MH that you are participating in. MHAUS will share news of your activity with the MH community through bulk email, newsletters, social media, and press releases. (Did you know MHAUS has a nearly 6,000 person database?) To promote your activity, contact us as soon as you know all the details. The more information you include, the more interesting your submission will be to people reading about your activity. And the more likely they will sign up to either join you or sponsor you. Contact MHAUS at info@mhaus.org or call 607-674-7901.

Do Your Shopping on AmazonSmile To Support MHAUS

AmazonSmile is a simple and automatic way for you to support MHAUS every time you shop, at no cost to you!

When you shop at smile.amazon.com, you’ll find the exact same low prices, vast selection and convenient shopping experience as Amazon.com, with the added bonus that Amazon will donate 0.5% of the purchase price to MHAUS.

To shop at AmazonSmile simply go to smile.amazon.com from the web browser on your computer or mobile device and set up your Amazon-Smile account. You may also want to add a bookmark to AmazonSmile to make it even easier to return and start your shopping at AmazonSmile.

Tens of millions of products on AmazonSmile are eligible for donations. You will see eligible products marked “Eligible for AmazonSmile donation” on their product detail pages. And yes, you can use the same account on Amazon.com and AmazonSmile. Your shopping cart, Wish List, wedding or baby registry, and other account settings are also the same.
Choose RYANODEX®:
formulated for rapid reconstitution and administration with fewer vials and less fluid volume.¹

RYANODEX® is formulated for speed and efficiency during the critical challenges presented by malignant hyperthermia (MH)²–⁴:

- Simple and rapid reconstitution within 10 seconds²
- One-minute administration of a loading dose by 1 provider¹,³
- Significantly fewer vials and less IV fluid volume required¹,³,⁵,⁶
  - One vial of RYANODEX® provides the same amount of dantrolene sodium as 12.5 vials (13 vials reconstituted) of other formulations

RYANODEX®: Because every minute counts³,⁴

Learn more at Ryanodex.com or call 855.318.2170

Indications
RYANODEX® (dantrolene sodium) for injectable suspension is indicated for the treatment of malignant hyperthermia in conjunction with appropriate supportive measures, and for the prevention of malignant hyperthermia in patients at high risk.

Important Safety Information
RYANODEX® is not a substitute for appropriate supportive measures in the treatment of malignant hyperthermia (MH), including:

- Discontinuing triggering anesthetic agents
- Increasing oxygen
- Managing the metabolic acidosis
- Instituting cooling when necessary
- Administering diuretics to prevent late kidney injury due to myoglobinuria (the amount of mannitol in RYANODEX® is insufficient to maintain diuresis)


Please see Brief Summary of full Prescribing Information on the adjacent page.
RYANODEX® (dantrolene sodium) for injectable suspension, for intravenous use.

Brief Summary of Prescribing Information. See Package Insert For Full Prescribing Information

INDICATIONS AND USAGE
RYANODEX® is indicated for the:
- Treatment of malignant hyperthermia in conjunction with appropriate supportive measures (see Warnings and Precautions)
- Prevention of malignant hyperthermia in patients at high risk.

DOSAGE AND ADMINISTRATION (Selected Information)
In addition to RYANODEX treatment, institute the following supportive measures:
- Discontinue use of malignant hyperthermia (MH)-triggering anesthetic agents (i.e., volatile anesthetic gases and succinylcholine).
- Manage the metabolic acidosis.
- Institute cooling when necessary.
- Administer diuretics to prevent late kidney injury due to myoglobinuria (the amount of myoglobin in RYANODEX is insufficient to maintain diuresis).
- Administer RYANODEX by intravenous push at a maximum of 1 mg/kg/h.
- If the physiologic and metabolic abnormalities of MH continue, administer intravenous boluses up to the maximum cumulative dosage of 10 mg/kg.
- If the physiologic and metabolic abnormalities remain, repeat RYANODEX dosing by intravenous push starting with 1 mg/kg.

Dosage for Prevention of Malignant Hyperthermia
- The RYANODEX prophylactic dose of 2.5 mg/kg administered intravenously every 4 to 6 hours for a period of at least 1 minute, starting approximately 75 minutes prior to surgery. Avoid agents that trigger MH. If surgery is prolonged, administer additional individualized RYANODEX doses during anesthesia and surgery.

Dosage for Pediatric Patients
- The recommended weight-based dose of RYANODEX for pediatric patients in the treatment and prevention of MH is the same as for adults for these indications (see Dosage and Administration).

Reconstitution and Administration Instructions
The suspension is packaged in vials that are to be reconstituted prior to administration.
- Reconstitute each vial of RYANODEX lyophilized powder by adding 5 mL of sterile water for injection as indicated.
- (Do not reconstitute with any other solution, e.g., 5% dextrose injection, 0.9% sodium chloride injection.)
- Shake the vial to ensure an orange-colored uniform suspension. Visually inspect the vial for particulate matter and discoloration prior to administration.
- Must use the contents of the vial within 6 hours after reconstitution. Store reconstituted suspensions at controlled room temperature (68°F to 77°F or 20°C to 25°C).

(For complete Dosage and Administration Section, see Full Prescribing Information)

CONTRAINdications
- None.

WARNINGS AND PRECAUTIONS
Muscle Weakness
ephyrocolagogue and an antidiuretic agent may potentiate their effects on the central nervous system (see Warnings and Precautions).

.Bean et al. 1983)

equal at delivery; neonatal levels then fell approximately 50% per day for 2 days before declining sharply. No fatal respiratory and neuromuscular side effects were observed in this study.

Nursing Mothers
- Dantrolene is present in human milk. In one case report, low dantrolene concentrations (less than 2 micrograms per milliliter) were measured in the breast milk of a lactating woman during intravenous dantrolene administration over 3 days. Because of the potential for serious adverse reactions of respiratory depression and muscle weakness in nursing infants from dantrolene, a decision should be made whether to discontinue nursing or to discontinue the drug, taking into account the importance of the drug to the mother.

Pediatric Use
- The safety and efficacy of RYANODEX in the treatment and prevention of malignant hyperthermia in pediatric patients is based on clinical experience with other intravenous dantrolene sodium products, which suggests adults-weight-based doses are appropriate for pediatric patients.

Geriatric Use
Clinical studies of RYANODEX did not include sufficient numbers of subjects aged 65 or over to determine whether they respond differently from younger subjects. Other reported clinical experiences have not identified differences in responses in the elderly and younger patients. In general, dose selection for an elderly patient should be cautious, reflecting the greater frequency of decreased hepatic, renal, or cardiac function, and of concomitant disease or other drug therapy.

OVERDOSAGE
Overdosage Symptoms
Overdosage symptoms include, but are not limited to, muscular weakness and alterations in the state of consciousness (e.g., lethargy, coma), somnolence, dizziness, and crystals.

Management of Overdosage
- Employs general supportive measures for acute overdosage of RYANODEX.

PaTIent Counseling Information
Inform patients, their families, or their caregivers of the following:

- Muscle Weakness
Muscle weakness (i.e., decrease in grip strength and weakness of leg muscles, especially walking down stairs) is likely to occur with the use of RYANODEX. Patients should be provided assistance with standing and walking until their strength is restored to normal (see Warnings and Precautions).

- Difficulty Swallowing
Cautiously indicated at meals on the day of administration because difficulty swallowing and choking have occurred with the use of dantrolene sodium products in general; dysphagia has been reported with the use of RYANODEX (see Warnings and Precautions).

- Dizziness and Somnolence
The use of RYANODEX has been associated with dizziness and somnolence. See Warnings and Precautions.

- Driving or Operating Machinery
Symptoms such as “dizziness” may occur. Since some of these symptoms may persist for up to 40 hours, patients must not operate an automobile or engage in other hazardous activity during this time (see Warnings and Precautions).

- Revised: 7/2014
Marketed by: Eagle Pharmaceuticals, Inc.
Woodcliff Lake, NJ 07677

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Woodcliff Lake, NJ 07677 7/2016

Table 1: Adverse Events in Healthy Volunteers

<table>
<thead>
<tr>
<th>Number of Subjects</th>
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<th>Number of Subjects</th>
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<tbody>
<tr>
<td>RYANODEX</td>
<td>DANTROLENE</td>
<td>SODIUM CONCENTRATOR</td>
</tr>
<tr>
<td>3</td>
<td>1</td>
<td>3</td>
</tr>
<tr>
<td>Headache</td>
<td>1 (3)</td>
<td>4 (3)</td>
</tr>
<tr>
<td>Nausea</td>
<td>1 (3)</td>
<td>2 (3)</td>
</tr>
<tr>
<td>Nausea</td>
<td>1 (4)</td>
<td>4 (3)</td>
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<tr>
<td>Tachycardia</td>
<td>1 (3)</td>
<td>0 (0)</td>
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<tr>
<td>Tachycardia</td>
<td>1 (3)</td>
<td>0 (0)</td>
</tr>
<tr>
<td>Infusion site pain</td>
<td>1 (3)</td>
<td>0 (0)</td>
</tr>
<tr>
<td>Dizziness</td>
<td>1 (3)</td>
<td>0 (0)</td>
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</tbody>
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Postmarketing Experience
- The following adverse reactions have been identified during postapproval use of another formulation of dantrolene sodium for injection. Because these reactions are reported voluntarily from a population of uncertain size, it is not always possible to reliably estimate their frequency or establish a causal relationship to drug exposure.

Pulmonary Edema
There have been reports of pulmonary edema developing during the randomization to receive treatment with RYANODEX or an active comparator at doses ranging from 1 mg/kg to 2.5 mg/kg.

- The RYANODEX dose was infused over the course of 1 minute for each of the doses evaluated.
- The active comparator was an ineptic formulation of dantrolene sodium that differed from RYANODEX in that it contained dantrolene sodium and mannitol at concentrations of 0.33 mg/mL and 50 mg/mL, respectively, whereas reconstituted product contains its prescribing information. The active comparator was infused at a rate that administered 20 mg of dantrolene sodium per minute for each of the doses evaluated.

Table 1 displays the most common adverse events in this study. These data are not an adequate basis for comparison of the types or frequencies of adverse event types between RYANODEX and the dantrolene sodium comparator.

- Adverse events increased in frequency with increasing doses in the trials, but did not differ in frequency between the two treatment groups. RYANODEX-treated subjects were more likely to report immediate adverse events of flushing, dyspnea, and dizziness than those receiving the active comparator.

- In all dose groups, hand grip strength declined after dosing. In general, the decline in hand grip strength was more pronounced and occurred more rapidly in the RYANODEX-treated subjects in the 1.0, 1.75, 2.0, and 2.25 mg/kg treatment groups. In the 2.5 mg/kg treatment group, the decline in hand grip strength both in amount and between the two treatment groups.

...
**New MHAUS Recommendation:**

Can Patients with a Suspected Personal or Family History of MH be Safely Anesthetized Prior to Diagnostic Testing for MH Susceptibility?

**Developed by Dr. Lena Mayes**

**Background:**

Patients with a known or suspected personal or family history of MH are often denied access to general anesthesia prior to diagnostic testing for MH susceptibility, resulting in cancellation and postponement of necessary surgical procedures. Also, MH susceptible patients may be told they cannot have surgery in ambulatory surgery centers but must have surgery at inpatient hospitals.

**Discussion:**

A suspected personal or family history of possible MH is not uncommon in patients requiring general anesthesia for medical or surgical procedures. The details of the presumed episode may be unclear, and in many instances, it is impossible to determine these details because medical records cannot be accessed in a timely manner. However, some patients suspected of being MH susceptible may require surgical management before formal MH susceptibility testing has been performed. Additionally, for many patients, diagnostic testing for MH susceptibility is not feasible because of the geographical distance to an MH biopsy-testing center, or their lack of insurance coverage for muscle contracture or genetic testing.

**Conclusions:**

Care of MH susceptible patients need not be restricted by the lack of formal MH susceptibility testing, nor should care be limited to inpatient hospitals facilities. MH susceptible patients can be safely cared for in most anesthetizing locations, including appropriately staffed and resourced ambulatory surgery centers, provided non-MH triggering agents are used. However, the chosen anesthetizing location should meet the following criteria:

1. The facilities should be prepared to recognize and treat an MH crisis according to the established guidelines by MHAUS and accrediting organizations. 2. Dantrolene should be accessible within ten minutes of the first signs of MH, and the facility should have the capacity to administer at least 10mg/kg of dantrolene in the event of an acute MH episode requiring multiple dantrolene doses to abort the crisis. The anesthesia machine should be flushed according to its specific manufacture’s recommendations and/or charcoal filters placed on both inspiratory and expiratory limbs to minimize residual volatile agent in the circuit (http://www.mhaus.org/healthcare-professionals/be-prepared/preparing-the-anesthesia-machine/).

**References**


New MHAUS Recommendation: What evidence-based interventions are recommended to alleviate hyperthermia associated with Malignant Hyperthermia?

Developed by Mohanad Shukry, MD

Background: The most important treatment of Malignant Hyperthermia (MH) is discontinuing MH triggering agents, hyperventilation, and timely administration of dantrolene. However, prolonged hyperthermia worsens patients’ outcomes and should also be treated when occurs.

Many cooling strategies are available, but in practice it is impossible to implement all of them simultaneously without distracting from the key tasks of administering dantrolene and treating the patient’s metabolic and respiratory abnormalities.

Many experts believe that hyperthermia is a sign of inadequate physiological treatment and clinicians’ priority should be to stop MH with dantrolene and adequately treat hypercarbia and acidosis before focusing time and efforts on thermal management (personal communication, Dr. Daniel Sessler). It is therefore important to prioritize cooling approaches based on efficiency, ease of use, and safety.

Discussion: Thermal management can be divided into three categories: pharmacologic, noninvasive, and invasive. Pharmacologic treatment of hyperthermia includes dantrolene, acetaminophen, and nonsteroidal anti-inflammatory drugs. Dantrolene is the only clinically available specific treatment for MH and, after discontinuation of triggering agents, should always be the initial treatment for any suspected MH episode. The recommended initial dose of dantrolene is 2.5 mg/kg bolus with repeated boluses as needed until hypermetabolism is controlled.

Acetaminophen, especially now that it is available intravenously, is increasingly used for analgesia and treatment of fever. However, its effectiveness in treating hyperthermia caused by MH has not been determined. Similarly, the role of nonsteroidal anti-inflammatory drugs as antipyretics during MH remains unknown. Nonetheless, the high body temperature associated with MH is due to excessive heat production by skeletal muscle; there is thus no theoretical basis, much less evidence, to suggest that antipyretic drugs will help control hyperthermia during MH episode. Noninvasive treatments of hyperthermia include strategic ice packing, forced air cooling, circulating cool water blankets, cold intravenous fluids, and ice-water immersion.

Cold intravenous fluid is effective: in healthy volunteers, 40 mL/kg infusion of 4°C or 20°C fluid, core temperature transiently decreased 2.5 ± 0.4°C and 1.4 ± 0.2°C, respectively. Cold fluids should be kept available and should typically be the initial cooling measure during an MH episode, especially since hydration is usually appropriate to limit the risk of renal injury from myoglobinemia. The method is limited by the amount of intravenous fluid that can be safely administered, typically about three liters in adults.

Ice packing (neck, groins and axillae) is effective, although prolonged direct skin exposure may provoke tissue injury. Convective cooling with forced air at ambient temperature is easy to implement and essentially risk-free. However, the method is nearly ineffective and little better than simply removing all covers and exposing the patient to ambient air. Ambient air temperature should be lowered to the extent practical. Circulating cool water blankets set to low temperatures such as 4°C absorb considerable heat, but are not available in all operating rooms and positioning water blankets or mattresses during an MH crisis may be complicated and distracting. As with any surface cooling method, efficacy is a linear function of surface area used. Some temperature management systems use circulating water with pads that feature thin hydrogel coating. They are more effective blankets than conventional circulating-water because they contact the skin well and have low thermal resistance. Ice water immersion is by far the most effective external cooling method, but is limited by the equipment required and need to move patients. In practice, immersion is not an approach that can be organized and implemented safely in the midst of an MH crisis.

Invasive strategies include bladder, rectal, gastric or peritoneal lavages, esophageal heat exchangers, intravascular heat exchange devices, and cardiopulmonary bypass. Gastric lavage is neither effective nor safe due to low return of aspiration of the injected fluids. Bladder lavage is ineffective due to small contact surface area and a relatively low bladder perfusion. Although not studied, rectal lavage may have similar limitations. Peritoneal lavage is highly effective because the peritoneum has a large contact surface area and is highly perfused. However, it should be noted that this method is invasive and requires special apparatus and skills (often available in emergency departments).

An esophageal heat exchanger is a new device which is inserted much like a standard orogastric tube. It has additional connectors designed for standard water blanket chillers/heaters. The device provides heat exchange via the blood circulation surrounding the esophagus. The system extracts about 50 watts which

continued on page 10
is relatively small compared to potential heat production during a severe MH crisis. Furthermore, the device is not yet commonly available. Lastly, cardiopulmonary bypass is by far the most effective cooling device, but its invasiveness and technical challenges are an obvious deterrent to recommending its application during an MH crisis unless bypass is required to treat hyperkalemic cardiac arrest. Furthermore, the degree of cooling is very rarely required.

Conclusions:
Cooling should never distract from dantrolene administration and hyperventilation. Most patients treated promptly with dantrolene and hyperventilation do not become seriously hyperthermic or necessitate active cooling. Active cooling should be used with care since there can be a substantial after-drop, depending on the cooling technique, duration of application, and body heat distribution; cooling should thus be discontinued when core temperature decreases to 38°C.

External cooling methods such as circulating-water mattresses or ice packs should be considered first. If external cooling is insufficient, an easy, effective, and safe next cooling strategy is to infuse 20 mL/kg of refrigerated intravenous fluid. Other treatments should rarely be necessary, but peritoneal lavage is probably the safest and most effective of the invasive approaches if the peritoneum is already open or the patient is in an emergency department with the requisite equipment and skills.

References:

Test your MH knowledge with the MH Knowledge Bank Quizzes available on the MHAUS website at: https://www.mhaus.org/healthcare-professionals/professional-development/quizzes-and-tests/

Can An MHS Person Have Surgery?

Yes! Surgery can be safely performed on known MH-susceptible patients. However, only those anesthetics that do not trigger the MH reaction must be used. In addition, close monitoring of appropriate vital functions is necessary. When dealing with an MH susceptible, the anesthesiologist should:

- Avoid the use of MH-triggering anesthetics.
- Be familiar with the signs and treatment of MH, e.g., re-review the routine information.
- Continuously monitor the patient’s exhaled carbon dioxide concentration and minute ventilation.
- Continuously monitor the patient’s temperature (also during recovery). Skin temperature is not optimal in this situation.
- Have an MH kit or cart within the operating room suite stocked with an adequate supply of dantrolene.

The U.S. and Canada MH Hotline is 1-800-MH-HYPER (1-800-644-9737)
Outside the U.S., call 1-209-417-3722
Yes!

I want to support MHAUS in its campaign to prevent MH tragedies through better understanding, information and awareness.

A contribution of: $35  $50  $100  $250  $500  $1000 (President’s Ambassador)

or $ __________, will help MHAUS serve the entire MH community.

Please print clearly:

Name: ______________________________________________________________________
Address: _____________________________________________________________________
City: ____________________ State: _____________ Zip: ____________
Phone: __________________________ E-mail: ____________________

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Credit Card Number: _______________________________________________
CV Code: ____________________Expiration: _________________________

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your issue intact, then
mail to: MHAUS, PO
Box 1069, Sherburne,
NY 13460-1069

Did you know?

MHAUS offers a lifesaving Hotline, free-of-charge, for any healthcare professional who unexpectedly comes face-to-face with a malignant hyperthermia emergency and quickly needs help. The cost per call to MHAUS is $100.00, and includes the contracted service to transfer your call to a consultant, the costs associated with the MH Hotline Coordinator, who assures there are consultants ready every day on a 24-hour basis for you. Dedicated MH Hotline Consultants, all well-known MH Experts, freely volunteer their time to help their fellow healthcare professionals through an intense situation.

Consider making at least a $100.00 donation (to cover a single call) specifically to help us maintain this lifesaving tool provided by MHAUS to all healthcare professionals.

Enclosed is my tax-deductible contribution of $ ____________ in support of the lifesaving MH Hotline.

Please make checks payable to: MHAUS and send to PO Box 1069, Sherburne, NY 13460.

❑ Visa   ❑ MasterCard   ❑ Discover   ❑ American Express

Name on card: ___________________________________________________

Credit Card Number: _____________________________________________ Expiration Date_________

Signature: ______________________________________________________
MHAUS Happenings, Events and Notices

THANKS! MHAUS thanks the following State Society of Anesthesiology – New Jersey, Ohio, and Wisconsin – for their financial support. Call the MHAUS office to ask Gloria how your group can join their ranks.

EMHG Group Meeting, May 17-18
The European Malignant hyperthermia Group (EMHG) meeting is scheduled for May 17-18, 2018, in Ferrara, Italy. As of press, the EMHG has not released details of the meeting, but you can stay up to date on the upcoming meeting by visiting the EMHG website at www.emhg.org.

MH Memorial Ride and After Party, August 26
Come ride with us in MHAUS’ hometown of Sherburne, NY. All proceeds from this event will go to MHAUS’ education fund to allow us to continue to educate the healthcare community about early recognition to prevent any more unnecessary deaths from malignant hyperthermia. For questions, contact Tina Roalef at 607-674-7901 or by email at tina@mhaus.org.

Member Partnership Dues Increased to $30
MHAUS has increased member partnership dues to $30. This modest increase helps fund MHAUS’ mission to promote optimum care and scientific understanding of MH and related disorders. Member benefits remain unchanged and include a 30% discount on events and materials, access to member areas such as chapter groups, forums, pages, quizzes, surveys, the MH Hot Topics e-newsletter and The Communicator quarterly newsletter.

Is Your Dantrolene Expired? We’ll Take It! Do you need Expired Dantrolene? We Have It!
Dantrolene sodium for injection that has reached its expiration date is used for training purposes. Medical facilities wanting to train staff often contact MHAUS requesting expired dantrolene sodium for injection – to which we are happy to oblige. However, considering the demand, it’s not always possible to keep our shelves stocked, so if your medical facility has expired dantrolene sodium for injection, please donate it to MHAUS. In return, MHAUS will provide you a free year’s membership that includes a 30% discount on most MH educational materials, access to online forums, quizzes, webinars and plenty more! When shipping, please be sure the vials are wrapped properly to eliminate possible damage. Please ship to: MHAUS, 1 North Main Street, Sherburne, NY 13460.

MHAUS Seeks Blog Writers
MHAUS monthly blog is open to Board members, the Professional Advisory Council, staff, Hotline Consultants, and MHAUS members-at-large. The only conditions are that the topic relate to MH or MH-like disorders, not exceed 2,000 words, and be appropriate and respectful of all viewpoints. MHAUS invites those interested to comment on MH-related subjects or how MH has affected them and their family. If you have questions or want more information, please email info@mhaus.org.