Prospects For An Alternative To MH Trigger Drug Succinylcholine

by MHAUS President Henry Rosenberg, MD

One of the most useful drugs in the armamentarium of anesthesia providers and emergency medicine physicians is succinylcholine (also called Anectine). Succinylcholine is a drug that produces intense paralysis within seconds of injection, which lasts (in most patients) for about 5-8 minutes. This is just perfect for procedures that required intense paralysis for only a few moments, such as intubation of the trachea, orthopedic manipulation, or brief relaxation to assist the surgeon. As such, the drug gained widespread acceptance after it was introduced in 1952 especially since the alternative drug, curare, had many problems in predicting response and duration.

It is well known that every medication has some undesirable side effects. This is especially true for succinylcholine. Within a few years of the introduction of succinylcholine, clinicians noted that in some patients paralysis would last for hours instead of minutes. If the patient were not anesthetized, s/he would be awake yet unable to move or communicate. Researchers discovered that the reason for the long duration of action in these patients was the presence of an inherited defective enzyme responsible for the degradation of the molecule. After a while, clinicians began to recognize this problem and, when it occurred, would sedate the patient until the drug wore off. However, routine screening for the altered enzyme was not economically feasible, so one in 2500 patients receiving succinylcholine experience muscle paralysis for an hour or more. Parenthetically, the person who worked out this problem was a pharmacologist, Dr. Werner Kalow, who later in his career would work with Dr. Beverly Britt in describing the muscle biopsy contracture test for Malignant Hyperthermia.

Then in the 1960s another life-threatening problem was noted with succinylcholine. Some patients who were burned or had spinal cord injury would experience a cardiac arrest after receiving succinylcholine. Researchers found that these patients developed a massive rise in serum potassium. Normally, potassium levels are about 4meq/L, but in many of these patients the level quickly rose to 8, 9 or more. When potassium levels are so high, the electrical conduction system of the heart is effected, throwing off the normal heart rhythm and

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“It is a mistake to try to look too far ahead. The chain of destiny can only be grasped one link at a time.” — Winston Churchill

Dianne Daugherty
MHAUS Executive Director

Over the past several months, the Malignant Hyperthermia Association of the United States (MHAUS) has been extremely busy with many positive directions being pursued, and a few challenges, to be sure.

While MHAUS remains dedicated to promote optimum care and scientific understanding of MH and other heat-related disorders, we also are becoming more and more aware that our scope of influence continues to broaden. There has been discussion about a number of new topics, including whether to infuse more energy into determining whether our focus should actively pursue the area of heat-related disorders, such as heatstroke (unfortunately occurring in all levels of the athletic arena). We need to further define whether MH continues to be our “largest” area of concern or is it only a “part” of an even larger problem. We’ve grappled with this question for a while now and will be “unpeeling the onion” as we move into the future.

To date, we have been able to continue our objective of applying quality improvement techniques to enhance products designed to assist health care professionals Be Prepared for MH. We listened to our customers in order to better understand their needs for the future and have offered products to help them achieve their MH preparedness goals. For instance, we have revised the MH In-service to incorporate a format that is easier to use, arranged in chapters that can be viewed at will – and repeatedly – to ensure under-standing of the material. The final product took two years to complete, includes short video clips on our website at www.mhaus.org for a sample of the content, and met an MH training need to ensure correct management of the MH-susceptible patient for many years to come.

Additionally, we have been combining efforts with the Society for Ambulatory Anesthesia (SAMBA) to develop a plan of action for an unforeseen MH emergency in an office-based facility. It was understood that office-based surgical facilities may possess varying levels of staff and equipment, and the Emergent Transfer Plan (ETP) with accompanying ETP Checklist were developed to allow necessary internal adjustments for each facility’s specific needs. This project took over 18 months to complete and involved much give-and-take between the Task Force members who were determined to produce a solid resource for their counterparts to use. The jointly-owned product is available to purchase via the MHAUS website as a pre-planning tool to devise a plan of action to efficiently move a patient suffering unexpectedly from MH to a nearby hospital’s ER and keep them safe.

In September 2012, MHAUS was advised by his family that Mr. Geoffrey Keller, who suffered MH at a young age, had passed away, possibly from MH. The Keller family felt strongly that MHAUS was an organization trying to make a difference in patient safety and believed it was their personal mission to support MHAUS and MH education. Thus, they began a solid dialogue with various MHAUS staff to determine how they could put a plan of action into place. Geoffrey’s father, Curt, is a minister and was particularly interested in holding organ concerts to benefit the mission of MHAUS. Soon, additional concert venues began to develop and the first (a blues-jazz type
producing a cardiac arrest. In some cases the patient was not able to be resuscitated. After the phenomenon was described and the cause noted, clinicians began to realize that succinylcholine should be avoided in burn and neurologically impaired patients (even those who had paralysis from a stroke).

Yet another life threatening problem was noted some years later. Some patients with an unrecognized or unappreciated muscle disorder would also develop a cardiac arrest due to elevated potassium after receiving succinylcholine. In these cases it was because the patient, most often a young male child, had an underlying, unrecognized muscle disorder such as Duchenne Muscular Dystrophy. Older patients who are at risk to high potassium levels after succinylcholine have a different form of Muscular Dystrophy. Again, after a while, most clinicians recognized that patients with muscle disease should not receive succinylcholine. However, there were always those patients, particularly young children, who had an underlying muscle disorder, but because they were too young to walk, the disorder was not recognized. Eventually, the FDA required that the manufacturers place a “black box warning” in the package insert indicating that succinylcholine should not be used without indication in young children for fear of high potassium levels resulting in cardiac arrest in those with undiagnosed muscle disorders.

Yet other significant problems were noted. In some patients, particularly children, succinylcholine would produce intense jaw muscle rigidity, making it almost impossible to open the mouth to insert an endotracheal tube. In those cases patients would also sustain significant muscle breakdown (called rhabdomyolysis). Furthermore, such intense jaw rigidity was noted to be a heralding sign of Malignant Hyperthermia.

Succinylcholine is also a trigger for MH. Animal studies and human experience demonstrated that succinylcholine could by itself be a trigger for MH. This meant that any anesthetizing location that has succinylcholine available – even for emergency use only – should also stock a full supply of dantrolene, even if they did not use the gas anesthetics that trigger MH (see President’s blog of May 2012). Despite some doubts, it is clear that this drug is a trigger for MH, particularly when used with the gas anesthetics that are MH triggers.

In addition to these life-threatening problems, there are a host of other unpleasant side effects, such as muscle aches and pains and in... continued on page 4

The Lila & Jerry Lewis Memorial Fund

There are many special people who take the time each year to remember their loved ones in a way that helps MHAUS. The people below have made gifts during FY 11-12 (October 2011 - September 2012) in memory of Lila and Jerry Lewis. We are most grateful for their support and special tribute gifts.

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In memory of Miriam Trustman - Marilyn Lewis Glassman
Continued from page 3

some non-MH patients, a mild degree of muscle breakdown.

So why is succinylcholine still used routinely? For several reasons, but most of all, the properties of the drug in most patients, namely short onset, intense paralysis, and short duration outweighed the other rare problems. Furthermore, no other drug matched these properties. Meanwhile, beginning in the 1980s a new series of muscle paralyzing drugs was introduced into anesthesia practice that had some of the desirable qualities of succinylcholine. Two such drugs were named vecuronium and rocuronium. These drugs do not lead to the life threatening increases in potassium as seen with succinylcholine, nor do they precipitate MH. If given in high doses they could produce rapid onset of paralysis just like succinylcholine; however, instead of the paralysis lasting about 5-6 minutes, it would last for more than 20 minutes. This is a problem if the procedure was a brief one.

One other important point about the alternative paralyzing drugs like vecuronium and rocuronium: The paralysis produced could be “reversed” if the clinician waited a long enough period of time after drug administration or with administration of a competitive reversal drug, i.e. neostigmine. (It is actually more complex than this, but the point is the same). However, in general, such reversal could not be effective in less than 20-30 minutes of administration.

If only it were possible to reverse the paralysis of these newer drugs quickly, then the desired qualities of succinylcholine would be matched. Well a few years ago such a compound was developed. This agent, Sugammadex, could rapidly and completely reverse the paralysis of rocuronium in a unique manner. The reversal agent would actually “trap” the paralyzing drug molecule and block it from attaching to the cell surface where it exerted its effect. Furthermore, the basic molecular structure of this new agent was similar to a complex sugar, so it was not associated with organ toxicity. It demonstrates that Sugammadex was designed to trap the muscle-paralyzing drug in its “donut hole.” Sugammadex was approved for use in most European countries, Australia, and New Zealand in about 2008 and has lived up to its reputation of being a “clean drug” that reverses the action of rocuronium very rapidly from any level of muscle paralysis.

Patients do not become re-paralyzed with time either.

So, if this drug is so great and can replace succinylcholine, why is it not available yet? I don’t have all the answers, but it seems that after the drug was submitted to the FDA for approval, the agency wanted some additional studies to make sure there were no significant side effects when given in repeated doses as well as other studies on the possible side effects of the drug.

Recently, the drug company that acquired Sugammadex completed the studies requested and resubmitted the drug to the FDA for reconsideration. A decision may be made within the next few months.

In those countries where the drug is available, there is concern that routine use of rocuronium, because it can be reversed quickly with Sugammadex, would lead to increased drug costs since rocuronium is much more expensive than succinylcholine. So, it is unlikely that succinylcholine will be replaced entirely. Rather, in those cases where there is a question or suspicion that the patient may be at risk for MH, or where there is evidence of muscle disease, or where there are procedures that are very brief but intense paralysis is needed (such as procedures on the airway by ENT surgeons), the anesthesia provider will likely lean to use rocuronium in high doses to obtain rapid, deep paralysis, and then reverse the paralysis with Sugammadex.

In time, if the economic calculation of the cost of incurring a complication with succinylcholine exceeds the costs of using Sugammadex routinely, well then, succinylcholine may not be used as often. Even now without Sugammadex, succinylcholine is not used routinely by most pediatric anesthesiologists in the US because of the concerns about unexpected high potassium levels in a patient with undiagnosed muscle disorder, although it is reserved for special indications.

Where there is a choice of several drugs that produce similar effects, the clinician’s decision to use one or another is rather complex. I have simplified the explanation of the factors leading to the decision to use Sugammadex. However, the main point is that it is likely there will be an acceptable alternative to succinylcholine, thereby lessening the chances of precipitating MH or other undesirable side effects of succinylcholine. Time will tell.

Disclaimer: Dr. Rosenberg has no personal financial interest in Merck or any company that manufactures Sugammadex. Neither does MHAUS receive support from any manufacturer of the drug.
Patients with malignant hyperthermia susceptibility (MHS) experience skeletal muscle hypermetabolism characterized by contracture, heat and lactate production when exposed to certain volatile anesthetics and/or depolarizing muscle relaxants. Acidosis, tachycardia, hypoxemia, hypercapnia and rhabdomyolysis can develop unless quickly treated.\(^1\)

MHS is a complex genetic disorder typically inherited in an autosomal dominant pattern, but also described arising from spontaneous mutations.\(^2\) Genetic tests to determine which patients are at risk for developing this potentially lethal condition screen for characterized mutations in either the ryanodine receptor type 1 (RYR1) gene or the skeletal muscle dihydropyridine calcium channel gene (CACNA1S) gene.

Nearly 70% of MHS is associated with RYR1, in which more than 100 distinct mutations have been identified.\(^3\) Approximately 1% of known MHS is caused by CACNA1S mutations. Six different types of MHS are described based on the locus of the mutation, named MHS1-6. Given the large number of mutations identified for this disorder it is necessary to sequence both these genes to provide comprehensive genetic testing for MHS.

The University of Minnesota currently use the sequence capture and targeted sequencing of the RYR1 and CACNA1S genes using the next generation sequencing platform (HiSeq 2000, Illumina Inc.) to identify mutations in these two genes. This analysis is limited to the coding exons and immediately adjoining intronic splice/donor sequences of the analyzed genes. Mutations in non-coding regions outside the intronic splice/donor sequences will not be detected using this methodology. This analysis has not been validated for detection of insertion/deletion mutations larger than 18 bp in length. In addition, this analysis will not detect large structural variations, such as whole gene deletions. Results will be reported within approximately 8-10 weeks. This testing should be ordered by a physician and results will be reported to the ordering physician. Genetic counseling services can be provided upon request.

Contact the Molecular Diagnostics Laboratory at the University Of Minnesota Medical Center, Fairview for additional information regarding genetic testing for MHS.

**Dr. Bharat Thyagarajan**  
Director, Molecular Diagnostics Laboratory  
Department of Laboratory Medicine and Pathology  
University of Minnesota  
Phone: (612) 624-1257  
Email: thya0003@umn.edu

**Matthew Bower**  
Genetic Counselor  
Molecular Diagnostics Laboratory  
Phone: (612) 624-8948  
Email: mbower1@fairview.org

**References:**
2. Litman, R In UpToDate; Basow, D., Ed.; UpToDate: Waltham, MA, 2012.

In the U.S. and Canada, the MH Hotline is 1-800-MH-HYPER (1-800-644-9737)  
Outside the U.S., call 1-209-417-3722
Safety Information
Management of Malignant Hyperthermia (MH) crises requires various supportive measures individualized for the patient’s condition. Administration of Dantrium® IV is one component of therapy and should not be considered a substitute for these measures. Even when properly treated, an MH crisis can result in death. Adverse events with Dantrium® IV include loss of grip strength, weakness in the legs, drowsiness, and dizziness, thrombophlebitis, and tissue necrosis/injection site reactions secondary to extravasation. There have been rare reports of pulmonary edema, urticaria and erythema. Symptomatic hepatitis (fatal and non-fatal) has been reported at various dose levels of the drug. Fatal and non-fatal liver disorders of an idiosyncratic or hypersensitivity type may occur with Dantrium® therapy. In case of overdose, symptoms include, but are not limited to, muscular weakness, lethargy, coma, vomiting, diarrhea, and crystalluria. For acute overdosage, general supportive measures should be employed. Please visit www.dantrium.com for additional product information. For full prescribing information, please see reverse.
**Dantrolen® Intravenous**
(dantrolene sodium for injection)

**DESCRIPTION**
Dantrolen® Intravenous is a sterile, white, lyophilized, pharmaceutically formulated dantrolene sodium (dantrolene sodium) in a solution of 0.9% sodium chloride (NaCl) for injection (containing 70 L, 500 mg mannitol, and sufficient sodium hydroxide to yield a pH of approximately 7.0) as a ready-to-use, sterile, injectable injectable for intravenous use, reconstituted with 20 mL of sodium chloride solution, 300 mg NaCl, and sufficient sodium hydroxide to yield a pH of approximately 7.0. Solutions for injection are reconstituted with 50 mL, sterile, water for injection (USP) without a bacteriostatic agent.

**Dantrolen®** is classified as a direct-acting skeletal muscle relaxant. Chemically, Dantrolen® is hydroxy-2,3-dihydroxy-4(1H)-pyridone-1,1-dioxide. The molecular formula of dantrolene sodium is C13H12N2O5 and the molecular weight is 336.3. The structural formula for the hydrochloric acid is:

\[ \text{C}_13\text{H}_{12}\text{N}_2\text{O}_5 \]

**PHARMACOLOGICAL PROPERTIES**
Dantrolen® has been shown to be effective in patients with malignant hyperthermia, as demonstrated by the prevention of sustained muscle rigidity and the control of the myoglobinuria and rhabdomyolysis. In skeletal muscle, Dantrolen® abolishes the excitation-contraction coupling, possibly by interfering with the release of Ca²⁺ from the sarcoplasmic reticulum. The administration of Dantrolen® to human volunteers is associated with loss of muscle mass and urinary loss of muscle mitochondrial enzymes. (See PRECAUTIONS: Information for Patients). Information concerning the passage of Dantrolen® across the blood-brain barrier is not available.

In the anesthetic-induced malignant hyperthermia syndrome, rhabdomyolysis points to an intrinsic, possibly mitochondrial basis of skeletal muscle tissue. In affected humans, it has been postulated that triggering agents (e.g., positive inotropic drugs and depolarizing neuromuscular blocking agents) produce a change within the cell which results in an accelerated rhabdomyolysis. The elevated cytoplasmic calcium affects acute cellular pathological processes that contribute to the malignant hyperthermia crisis.

It is hypothesized that addition of Dantrolen® to the “shocked” malignant hyperthermia muscle may retard the generation of cytoplasmic free calcium ions and mitigate the metabolic and biochemical changes associated with the malignant hyperthermia crisis may be reversed or attenuated by the intravenous administration of Dantrolen®. The specific mechanism of intravenous Dantrolen® effect of skeletal muscle in anesthetics has not been elucidated. Intravenous Dantrolen® administration is directed, while local concentrations never get near sufficient to the extent of the entire skeletal muscle. Clinical experience has shown that early, rapid and efficient tissue changes of malignant hyperthermia can appear during anesthetic administration and through the prophylactic use of Dantrolen® and other clinically accepted patient management practices. These effects are comparable to the therapeutic use of malignant hyperthermia and is associated with the administration of Dantrolen®. The administration of the recommended prophylactic doses of malignant hyperthermia to healthy volunteers was not associated with adverse outcomes. The effect of prophylactic doses of malignant hyperthermia on clinical outcomes is unknown.

Specific pathological pathways for the degradation and elimination of Dantrolen® in humans have not been elucidated. The major adverse effects of Dantrolen® are muscle weakness, rhabdomyolysis, and myoglobinuria. Major side effects in fluids are less than 1.5% of the intravenous dosage and adverse renal effects of Dantrolen® are not associated with renal damage. Another medical effect has been an atrial arrhythmia occurs related to the adler.

**INDICATIONS AND USES**
Dantrolen® is indicated for the prophylactic use of malignant hyperthermia susceptible and/or malignant hyperthermia susceptible genetically. The dose of Dantrolen® should be adjusted, along with appropriate supportive measures, for the management of the malignant hyperthermia crises of skeletal muscle or malignant hyperthermia crises in patients of any age. Dantrolen® intravenous should be administered if there is a possibility of malignant hyperthermia crisis. The onset of a malignant hyperthermia crisis is recognized by: (a) hyperthermia, tachycardia, tachyarrhythmia, and/or hypotension; (b) muscle rigidity, flushing, diaphoresis, and/or respiratory failure; (c) increased concentration of muscle enzymes in plasma or urine; and (d) death.

**INDICATIONS**
Dantrolen® is indicated for the treatment of patients with malignant hyperthermia (see PRECAUTIONS: Information for Patients). Information concerning the passage of Dantrolen® across the blood-brain barrier is not available.

**PRECAUTIONS: Information for Patients**
Information concerning the passage of Dantrolen® across the blood-brain barrier is not available.

**CONTRAINDICATIONS**
Do not use Dantrolen® Intravenous in patients with renal or cardiac insufficiency or those who are anemic.

**WARNINGS**
The use of Dantrolen® Intravenous is the management of malignant hyperthermia crisis is not a substitute for previously known supportive measures. These measures must be administered in addition to any possible life-saving emergency measures, such as administration of oxygen, resuscitation measures, and maintenance of adequate circulation and respiration. Dantrolen® should not be administered to patients with severe malignant hyperthermia crisis.

**ADVERSE REACTIONS**
Dantrolen® should be used in patients with severe malignant hyperthermia crisis. The administration of intravenous Dantrolen® to human volunteers is associated with loss of muscle mass and urinary loss of muscle mitochondrial enzymes. In patients with severe malignant hyperthermia crisis, Dantrolen® should be used only in cooperation with appropriate monitoring of skeletal muscle function including frequent determination of SGOT or SGPT.

**DOSAGE AND ADMINISTRATION**
Dantrolen® should be administered to patients with severe malignant hyperthermia crisis at the following doses:

**POSSIBILITIES**
Dantrolen® should be administered to patients with severe malignant hyperthermia crisis at the following doses:

**SIDE EFFECTS**
Dantrolen® should be administered to patients with severe malignant hyperthermia crisis at the following doses:

**RECOMMENDATIONS**
Dantrolen® should be administered to patients with severe malignant hyperthermia crisis at the following doses:

**PHARMACOKINETICS**
Dantrolen® should be administered to patients with severe malignant hyperthermia crisis at the following doses:

**REFERENCES**
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**CLINICAL PHARMACOLOGY**
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During the months of September through December 2011 thirteen volunteer physicians answered 68 calls to the MH Hotline. Forty-three calls involved patients who were experiencing at least one symptom suggestive of MH and the caller was asking for help in determining whether or not the patient had MH (case consultations). The other 25 calls were solely questions about MH. Consultants working on the Hotline included Drs. Adragna, Chapin, Gronert, Litman, Melton, Kaplan, Rosenberg, Rosenbaum, Skoog, Tobin, Watson, Weglinski, and Wong. Four of the case consultations were deemed definitely MH and three were considered probably MH by the Hotline Consultant. There was one death (thought not to be MH related). Although not always recorded on the report form, one of the cases occurred in a non-hospital setting. Nineteen of the 68 calls involved children less than 18 years of age (seven of these calls were for children two years of age or younger).

The majority of callers were anesthesiologists from the United States (representing 28 states). One call came from Honduras. In addition to anesthesiologists, the Hotline was contacted by anesthesia residents, nurse anesthetists, emergency medicine physicians, a surgeon, urologist, pediatrician, ICU physician, hospitalist, and a pharmacist.

Of the four case consultations considered to be definitely MH, the most dramatic involved a 59-year-old male undergoing general anesthesia for a shoulder arthroscopy. The caller, an anesthesiologist, had anesthetized the patient twice before without difficulty. For this surgery, the patient received a volatile agent (anesthetic gas) and succinylcholine (muscle relaxant). Ten minutes after surgery began, the patient’s end-tidal carbon dioxide (CO2) rapidly increased to greater than 100 mmHg (normal is less than 50 mmHg). Carbon dioxide is produced by metabolism in the body, exhaled by the lungs, and measured continuously via the patient’s breathing tube during general anesthesia. An increase in CO2 can occur for a variety of reasons, but MH is one of the more prominent ones. After checking his anesthesia machine to make sure it was working properly, the caller converted the anesthetic to a non-triggering variety by turning off the anesthetic gas and starting an intravenous infusion of propofol. He also began hyperventilating the patient and giving dantrolene. After 6 mg/kg of dantrolene the patient appeared to be responding well, but then began exhibiting further signs of MH. His potassium increased to 6.5 mmol/L (upper limit of normal is 5.3 mmol/L) and his urine became tea-colored (suggesting muscle breakdown). The surgery was completed and the patient was transferred to the ICU asleep with his breathing tube still in place. His initial creatine kinase (CK) level was 12,000 U/L. It later rose to 41,000 U/L. A normal CK level for an adult male is less than 340 U/L. CK is an enzyme that leaks out of the muscle cell during muscle breakdown, such as can occur during an MH event. After receiving more dantrolene and careful monitoring, the patient recovered from the event. The patient’s impressive signs of muscle breakdown might partially be explained by the fact that he was described as a very muscular man who was a serious body builder taking testosterone to supplement his efforts. It was unknown if he was taking any other dietary supplements. This is an important point in that testosterone and some dietary supplements have been shown to possess calcium-releasing effects on muscle. There is no evidence, however, that testosterone will induce MH on its own. Nor is it known how it might enhance an MH reaction when given in combination with a triggering agent (Capaccione JF et al. Anesth Analg 2009;108:900-3).

Two of the case consultations involved young males (one was seven and the other was 19-years-old) who developed jaw and generalized body rigidity after receiving succinylcholine. Both of them subsequently developed signs of MH that were successfully treated with dantrolene. Isolated jaw rigidity can be due to a variety of causes such as an idiosyncratic reaction to
suxcinylcholine, temporomandibular joint disease, and neuromuscular disorders. However, generalized body rigidity is considered to be a fairly specific sign of MH, but is not always seen in every MH event.

MH is known to occur most commonly in young adult males, so a case consultation regarding an 81-year-old female was unusual. The patient was undergoing a laparoscopic-assisted resection of part of her colon under general anesthesia. During surgery her end-tidal CO2 began to rise (up to 100 mmHg). Despite increasing her respiratory rate and volume of breaths with good lung and chest expansion noted, the anesthesiologist was unable to decrease her CO2. Laparoscopic surgery involves distending the abdomen by pumping in an absorbable gas, most commonly CO2, in order to allow visualization of abdominal structures by the laparoscope. An increase in end-tidal CO2 is common during laparoscopic cases due to some of the abdominal CO2 being absorbed into the bloodstream and then exhaled from the lungs. However, the increase is usually mild and easily managed. The caller also palpated the patient’s chest and neck to make sure no CO2 had spread under her skin. Soon thereafter the patient developed a heart rate of 140. An arterial blood gas showed a CO2 of 88 mmHg. At this point dantrolene was given, the patient was cooled, and surgery was aborted. Shortly after receiving the dantrolene, the patient’s heart rate slowed and her end-tidal CO2 decreased to 50 mmHg. The Hotline Consultant was contacted to offer his opinion as to whether or not this was MH (he agreed it was) and suggest further treatment and evaluation. As with any suspected episode of MH, the caller was urged to complete an Adverse Metabolic or Muscular Reaction to Anesthesia (AMRA) report to be submitted to the MH Registry of MHAUS.

Meet This Issue’s Hotline Consultant

Dr. Margaret Weglinski, Assistant Professor of Anesthesiology at the Mayo Clinic in Rochester, Minnesota, has been a Hotline Consultant since 1997.

“What I like best about my work with the Hotline is the chance to speak with health care providers from around the country,” she says. “Whether it’s answering a straightforward question about MH or trying to determine whether or not a patient is experiencing an MH episode, I find it rewarding to (hopefully) be of assistance.

In her spare time, Dr. Weglinski likes to travel, bike and hike, and says she tries to combine the three as often as possible.
Continued from page 2

of concert) was held in Atlanta, GA on March 13, 2013. The family has also been working with MHAUS staff to devote a spot on the MHAUS website to allow them to post events, provide visitors with donation and specific funding options, and converse back and forth within their particular group. They will use the system as a planning tool for upcoming events and to share insight, feedback, and questions with each other, MHAUS staff and MH experts. More Chapter Groups will be evolving in the future and will be able to accommodate healthcare professionals, patients and their families, and mixed groups, as needed.

MHAUS website online Chapter Groups have evolved through our transition to an online association management software which allows visitors to join MHAUS as one of various MEMBERSHIP options online, or to visit as a FRIEND at no charge. Membership of course, offers options such as 30% off MH educational materials and the ability to take advantage of future webinars and online courses at no charge; non-members will incur a charge.

In addition, we have been working with the Association of Operating Room Nurses (AORN) on multiple projects. We were pleased to be able to gain their assistance to secure accreditation for an MH course presently housed online at CME-Zone and CECity via a link from the MHAUS website. This course has had nearly 5,000 readers in the first year (available until December 2013) with nearly 50% successfully completing the test online to obtain their FREE credits. This program has given MHAUS additional insight as to customer needs through their feedback regarding topics of interest for future MH and other heat-related disorder topics.

We are also planning MH Mini-conferences in Orlando, Florida in June 2013, and Albany, New York in September 2013. These conferences bring together MH experts to share MH preparedness requirements for both healthcare professionals and patients and their families in joint presentations. The venue offers open communication in order to share concerns, insight and education between both groups in a non-confrontational manner. Specific time for questions and answers is allotted. Visit the MHAUS website for details.

MHAUS will also sponsor a Scientific Conference this year. Genetic research has developed to a level that condones holding this monumental event in Toronto, Ontario on November 1-2, 2013. Dr. Sheila Riazi, MH Diagnostic Testing Center Director in Toronto, will be hosting the event and the organizing committee has come up with two days of intense forward-thinking presentations by leading authorities in the areas of RYRI physiology and pathophysiology, heat and MH susceptibility, genetic testing, practical cases, RYR2 and RYR1 comparison, and RYR1 and other gene variants, as well as a final discussion on dantrolene insights. The proceedings will be written up for publication in the months following the close of the event. Mark this on your calendar as a premier event for 2013! Contact us for more details anytime.

The MH Mock Drill Kit was a natural segue to our newest program, the MH Prep Check. Are you positive that you have all systems in place for a possible MH event? Would it be helpful to have an MH expert looking over your shoulder during an MH Mock Drill to assure you have addressed all areas correctly and to offer suggestions to improve the quality of your response? Would an MH expert’s opinion on your MH preparedness plan offer a higher level of confidence?

Then the MH Prep Check may be for you! This program is being developed now and we plan to release it in the summer months of 2013. A minimal amount will be charged to cover some of the costs, but due to a strong supporter’s willingness to help get this program off the ground, much of the initial costs will be covered. The presenter will be advised of the purchaser’s specific questions prior to his/her visit and will leave the purchaser with an MH Mock Drill Kit, an Operating Room Protocol Poster, and other MH materials. These tools, in addition to the MH expert’s time and focused attention, will offer specific answers to each facility’s personal areas of concern regarding MH preparedness and help them fine tune their plan of action and areas to improve.

Visit our website at www.mhaus.org regularly or ask to have your email added to our e-newsletter to receive regular updates of what is new in the MH world! We hope to hear from you soon.
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Despite initial satisfactory response to i.v. dantrolene there have been reports of fatalality, which involve patients who could not be weaned from dantrolene after initial treatment. The administration of i.v. dantrolene is associated with loss of grip strength and weakness in the legs, as well as drowsiness and dizziness. There are rare reports of pulmonary edema developing during treatment, in which the diluent volume and mannitol needed to deliver the drug possibly contributed. There have been reports of thrombophlebitis following administration of intravenous dantrolene. Tissue necrosis secondary to extravasation has been reported. There have been rare reports of urticaria and erythema possibly associated with the administration of i.v. dantrolene. Injection site reactions (pain, erythema, swelling), commonly due to extravasation, have been reported. Symptoms which may occur in case of overdose include, but are not limited to, muscular weakness and alterations in the state of consciousness (e.g., lethargy, coma), vomiting, diarrhea, and crystalluria.
THANKS! MHAUS is grateful for the financial support of the following State Societies of Anesthesiology: Alabama, Maryland, Michigan, and Pennsylvania. Our appreciation also goes out to the following Associations of Nurse Anesthetists: New York, Michigan, and Tennessee. Call the MHAUS office to ask how your group can join their ranks!

Upcoming Events
Visit MHAUS at these upcoming events: AANA, August 10-13, Las Vegas, NV; ASA, October 12-16, San Francisco, CA.

Let your Voice Be Heard On The MHAUS Blog
MHAUS has decided to suspend the monthly President’s Blog in favor of a new forum open to Board members, the Professional Advisory Council, staff, Hotline Consultants, and MHAUS members at large.

The only conditions are that the topic should 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.

All submissions will be reviewed by MHAUS President Henry Rosenberg, M.D., a Board member, and a member of the Professional Advisory Council. If you have questions or want more information, please email info@mhaus.org.

MH Mini-Conferences
Make plans now to attend one of the two upcoming MH Mini-Conferences – the first at Orlando Med Center in Orlando, Florida, on June 29; and the second at St. Peters Health Partners Hospital in Albany, NY, on September 14. Email fay@mhaus.org for more information.

Congratulations John Skoog
MHAUS congratulates MH Hotline Consultant John Skoog who is now Chairman of Anesthesia at Mercy Medical Center in Des Moines, Iowa.

Visit MHAUS Website For Resource Links
You could spend hours searching the Web for the information you need, or you could simply visit the MHAUS website; there you’ll find 28 resource links for both professionals and MH-susceptibles alike.

We Want To Hear From You
Let us know how you think MHAUS can better serve you. Call 607-674-7901 or email info@mhaus.org. Your comments and suggestions are important.