MHAUS Remembers
Malignant Hyperthermia Pioneer
Michael Denborough, MD

MHAUS mourns the death of one of the pioneers in the study of Malignant Hyperthermia, Dr. Michael Denborough (July 11, 1929 to February 8, 2014) of Australia.

Dr. Denborough and colleagues were the first to describe the disorder that was later named Malignant Hyperthermia. He did seminal work in describing the clinical and genetic basis for human MH. He also worked with pigs who developed “porcine stress syndrome” and showed how those animals could be a model for human MH.

Later in his life he worked hard as an advocate for anti-nuclear proliferation.

Says MHAUS President Henry Rosenberg, MD, “He was a great scientist, and a wonderful human being.”

In 1961, Michael Denborough was one of two doctors who described a young man in Melbourne, Australia, with a fractured tibia who was more concerned about receiving general anesthesia than about his fractured leg because 10 of his family members had developed uncontrolled hyperthermia and death during general anesthesia with ether.

The anesthesiologists, therefore, decided to proceed with general anesthesia using halothane, a recently introduced general anesthetic agent. After 10 minutes of halothane anesthesia, the patient became tachycardic, hypotensive, hypoxemic, and hyperthermic, and the halothane was discontinued. He was packed in ice and subsequently recovered uneventfully.

The patient’s family was investigated by Dr. Michael Denborough, an internist and research fellow with an interest in genetics, who noted that the pattern of deaths appeared to follow an autosomal dominant inheritance. He later determined that serum creatine phosphokinase (CK) levels were elevated in many of the family members.

Denborough’s publication was the first to suggest that an inherited syndrome could be the cause of an anesthetic-induced fatal hyperthermic crisis.

Subsequent cases

continued on page 6

On the inside

Meet MHAUS’ New Membership Coordinator ..... 2
Dr. Dirksen To Chair PAC .................................. 3
Support MHAUS When You Shop ........................ 3
A Partnership To Save Lives .............................. 6
Investigators Seek Your Help ............................. 6
The Cost Effectiveness Of Stocking 36 Vials ..... 7
2013 Hotline Summary .................................... 8
Glossary Of MH Terms .................................. 10
Happenings .................................................. 12
Malignant Hyperthermia (MH) is an inherited muscle disorder which, when triggered by potent inhalation anesthetics and succinylcholine, may cause a life-threatening crisis. The incidence of MH is low, but, if untreated, the mortality rate is high. Since the advent of the antidote drug, dantrolene sodium, and with greater awareness of the syndrome, the mortality rate has decreased. Great advances in our understanding of MH have been made since it was first recognized in the early 1960s, but the nature of the fundamental defect(s) is still unknown.

MHAUS advocates that all surgical patients undergoing general anesthesia should receive continuous temperature monitoring, that adequate supplies of dantrolene be stocked near the OR and that thorough family histories be obtained.

MHAUS Welcomes Membership Coordinator Kecia Funaro-Burton

As we head into the spring season, I am focused on nature’s new growth and promises of fulfillment.

MHAUS is also poised for internal growth - from the roots of the organization – our members, all of you!

Membership is the backbone of who we are and why we are here – to protect patients who might experience the devastation of an MH crisis and inform and educate all healthcare professionals who watch over and care for them during surgery. Part of the support we receive from our members ensures we can continue to provide the MH Hotline to healthcare professionals who may unexpectedly find themselves dealing with an MH event and need a lifeline to knowledge from our MH experts who give of their time and experience freely – the hotline consultants remain invaluable to us and countless others!

Along with other programs and products we provide for our members and the public-at-large, we focus on what we can offer our members with value to them and can make their lives just a little easier.

Active members are the roots feeding our organization, and if we can make the roots strong, we can make the entire organization resilient and a force to be reckoned with.

Toward that end, we have recently hired a Membership Coordinator on February 10, 2014 to focus on and strengthen our “roots”. Her name is Kecia Funaro-Burton. Kecia has a solid background in communications and has been involved in customer service and strategic planning. She may have already reached out to you in an email message or perhaps you have spoken with her on the phone. She will be initially focused on reaching out to our present members and those who have lapsed - to obtain feedback and insight as to where we are meeting your needs and what things we might do to enhance your membership experience.

She will be involved with the various “chapter groups” beginning to set up shop across the nation with a mission to assure MH education and preparedness plans are in place in their home communities! These groups are also organizing fundraising events for MHAUS to help us assure we remain strong and viable for many years to come. Kecia can
Dr. Robert Dirksen To Chair MHAUS Professional Advisory Council

Robert T. Dirksen, PhD, Department of Pharmacology and Physiology at University of Rochester Medical Center has been newly appointed to Chair the Professional Advisory Council (PAC) of the Malignant Hyperthermia Association of the United States (MHAUS).

“I am pleased to announce that Dr. Robert Dirksen has been appointed as Chair to the Professional Advisory Council,” says MHAUS President Henry Rosenberg, MD, CPE.

Dr. Dirksen is an outstanding scientist who has focused much of his scientific efforts on understanding the underlying defects in skeletal muscle in patients with Malignant Hyperthermia, Myotonic Dystrophy and Central Core Disease among others. His particular focus has been on understanding the control calcium movements in muscle and the relation of calcium movements to abnormal muscle function. His work has been funded by the National Institutes of Health and the Muscular Dystrophy Association among others. He has published over 80 articles in leading scientific journals in addition to many abstracts, book chapters, and invited lectures.

As chair of PAC, Dirksen will work closely with the president, executive director, and MHAUS Board to assess strengths and opportunities for the PAC; assign specific topics or documents requested for review to the PAC; synthesize PAC comments; keep the Board of Directors updated on research which has the potential to enhance MHAUS’ mission; advise on parameters for PAC membership and terms of service on the PAC; and be a member of the organizing committee for future scientific conferences.

The PAC of MHAUS are healthcare professionals from throughout the world who are expert in Malignant Hyperthermia and together approve and generate all content published by MHAUS.

Shop 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. You may also want to add a bookmark to AmazonSmile to make it 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.
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.

*JHP's Dantrium® IV has a 36 month shelf life at manufacturing point. There is always some lag period between manufacturing date and when the product ships to the end user, which varies based on when the order is placed.
**Dantrolen® Intravenous (dantrolene sodium for injection)**

**DESCRIPTION:** Dantrolen Intravenous is a sterile, non-pyrogenic, lyophilized formulation of dantrolene sodium for injection. It is supplied in a vial containing 60 mg of dantrolene sodium, 65 mg of sorbitol and 0.5 mg of sodium metabisulfite in 3 mL of water for injection. Each mL of concentrate contains 20 mg of dantrolene sodium, 200 mg of sorbitol and 1.4 mg of sodium metabisulfite.

**CLINICAL PHARMACOLOGY:** In isolated nerve muscle preparations, Dantrolen has been shown to delay the development of the repetitive discharges characteristic of the myotatic reflex by a direct action on the motor end plates.

**Indications:** Dantrolen Intravenous is an effective, fast-acting skeletal muscle relaxant that appears to be useful in the management of malignant hyperthermia. Dantrolen is given intravenously for the treatment of malignant hyperthermia crisis in the operating room or emergency room setting.

**CONTRAINDICATIONS:** Dantrolen Intravenous is contraindicated in patients who are hypersensitive to the drug, or in those with a history of malignant hyperthermia.

**WARNINGS:** The use of Dantrolen Intravenous in the management of malignant hyperthermia crisis is not a substitute for prompt and proper supportive measures. These measures must be done by adequately trained personnel for the detection of malignant hyperthermia.

**PRECAUTIONS:** General: The patient should be observed for the development of malignant hyperthermia crisis until the entire course of the antimuscarinic drug therapy is completed. In patients with a history of malignant hyperthermia, the use of other antimuscarinic drugs (e.g., diphenhydramine, promethazine, and scopolamine) within 24 hours of the rechallenge is contraindicated.

**ADVERSE REACTIONS:** There have been reports of deaths following malignant hyperthermia crisis when treated with intravenous dantrolene. These cases are not fully described in the literature. In one study, a postoperative patient who received dantrolene for malignant hyperthermia developed a respiratory arrest and required intubation. Although the patient survived, the cause of death was not determined.

**DOSAGE AND ADMINISTRATION:** Dantrolen Intravenous should be administered at a dose of 2 mg/kg intravenously over 30 to 60 minutes, followed by a continuous intravenous infusion of 1 to 2 mg/kg per hour until the crisis is controlled. In patients who respond to a 2 mg/kg dose, the infusion rate may be reduced as the crisis is controlled.

**PRODUCT ADMINISTRATION:** Dantrolen Intravenous is supplied as a sterile, non-pyrogenic, lyophilized formulation. The lyophilized contents must be reconstituted with sterile water for injection to form a clear, colorless solution. Each mL of concentrated solution contains 20 mg of dantrolene sodium. Dantrolen Intravenous should be used only in conjunction with appropriate monitoring equipment and should be administered only by qualified personnel.

**SUPPLEMENTAL INSTRUCTIONS:** For adults, general supportive measures should be employed. Intravenous fluids should be administered in large quantities to prevent the possibility of fluid overload. An adequate airway should be maintained and arterial and respiratory monitoring should be used. Anesthesia should be maintained until complete voluntary muscle relaxation is achieved.

**Precautions:** Dantrolen Intravenous should be administered at a dose of 2 mg/kg intravenously over 30 to 60 minutes, followed by a continuous intravenous infusion of 1 to 2 mg/kg per hour until the crisis is controlled. In patients who respond to a 2 mg/kg dose, the infusion rate may be reduced as the crisis is controlled. Dantrolen Intravenous is supplied as a sterile, non-pyrogenic, lyophilized formulation. The lyophilized contents must be reconstituted with sterile water for injection to form a clear, colorless solution. Each mL of concentrated solution contains 20 mg of dantrolene sodium. Dantrolen Intravenous should be used only in conjunction with appropriate monitoring equipment and should be administered only by qualified personnel. Intravenous fluids should be administered in large quantities to prevent the possibility of fluid overload. An adequate airway should be maintained and arterial and respiratory monitoring should be used. Anesthesia should be maintained until complete voluntary muscle relaxation is achieved.

**Precautions:** Dantrolen Intravenous is supplied as a sterile, non-pyrogenic, lyophilized formulation. The lyophilized contents must be reconstituted with sterile water for injection to form a clear, colorless solution. Each mL of concentrated solution contains 20 mg of dantrolene sodium. Dantrolen Intravenous should be used only in conjunction with appropriate monitoring equipment and should be administered only by qualified personnel. Intravenous fluids should be administered in large quantities to prevent the possibility of fluid overload. An adequate airway should be maintained and arterial and respiratory monitoring should be used. Anesthesia should be maintained until complete voluntary muscle relaxation is achieved.
Continued from front page

Continued from front page

of anesthetic-related fulminating hyperthermia accompanied by rhabdomyolysis and muscle rigidity were reported in the 1960s. Because of its high mortality rate (> 70%) and consistent presence of elevated body temperature, this syndrome was referred to as “malignant hyperpyrexia” or “malignant hyperthermia.”

Since that time, our knowledge of the inheritance, pathophysiology, and treatment of MH has grown considerably. Anesthesiologists have become adept at prospectively identifying susceptible patients and diagnosing acute MH early in its course, which has led to a drastic decrease in mortality from MH, to less than 5% in developed countries.

Help Investigators By Registering In The Online CMD International Registry

MHAUS invites people with MH and muscle weakness to register in the online Congenital Muscle Disease International Registry (CMDIR) at www.cmdir.org. Registering means entering your contact information, taking an Intake Survey and signing a medical release.

The CMDIR is a global database run by 3 nonprofit foundations to:

· ensure that clinical trials in congenital muscle disorders are successful, enrolling early and completely;
· track how people with congenital myopathy are doing on an annual basis;
· and identify issues and complications unique to each muscle condition to help improve care.

The CMDIR is currently working with investigators who are planning a clinical trial for people with central core disease and RYR1 mutations. Registering allows CMDIR staff to ensure your information is complete and determine what additional testing may be needed to qualify for the trial.
MHAUS Backs Cost Effectiveness Of Stocking 36 Vials Of Life-Saving Drug Dantrolene

Editor’s Note: a full paper describing the basis for this conclusion will appear in the spring issue of Anesthesiology News.

Dantrolene sodium is the only specific, effective treatment for the syndrome of Malignant Hyperthermia (MH). MH is an uncommon inherited disorder that has no outward manifestation until the patient receives one of the potent gas anesthetics and/or the paralyzing drug succinylcholine.

Questions have been raised as to whether the cost of having a full supply of dantrolene (36 vials) is justified given the rarity of the syndrome, particularly for a free standing surgery center where resources may be limited.

Investigators from the Columbia University Department of Anesthesiology, Saint Barnabas Medical Center and MHAUS sought to answer that question by drawing on publicly available generally accepted epidemiologic data related to the prevalence of and the harm that may occur from the syndrome.

In the U.S. 15 million surgeries are performed in an outpatient facility. The incremental cost effectiveness of dantrolene compared to supportive care only in the management of an MH crisis is shown to be at least $196,000. Even varying assumptions of MH incidence, cost of dantrolene, and mortality with and without the drug, the incremental cost effectiveness is still substantial.

MHAUS, a patient advocacy, not-for-profit organization, has for many years recommended that all facilities where potent gas anesthetics and succinylcholine (the “trigger” drug for MH) may be used, maintain 36 vials of dantrolene be immediately available to treat the patient. This study adds an economic dimension to the humanitarian one concerning the availability of this life-saving drug however anesthetic trigger agents for MH are used.

Are You Prepared for a CRISIS?

- 36 month shelf life*
- Ready to administer in 20 seconds†

Visit www.revonto.com or call (877) 411-USWM (8796) to learn more about Revonto and to see the full Prescribing Information.

*from the date of manufacture for unit solution is clear

Important Safety Information

The use of Revonto in the management of malignant hyperthermia crisis is not a substitute for previously known supportive measures. These measures must be individualized, but it will usually be necessary to discontinue the suspected triggering agents, attend to increased oxygen requirements, manage the metabolic acidosis, institute cooling when necessary, monitor urinary output, and monitor for electrolyte imbalance. Patients who receive I.V. dantrolene sodium preoperatively should have vital signs monitored.

If patients judged malignant hyperthermia susceptible are administered dantrolene sodium preoperatively, anesthetic preparation must still follow a standard malignant hyperthermia susceptible regimen, including the avoidance of known triggering agents. Monitoring for early clinical and metabolic signs of malignant hyperthermia is indicated because attenuation of malignant hyperthermia, rather than prevention, is possible.

Despite initial satisfactory response to I.V. dantrolene there have been reports of fatalities, 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 have been reports of thrombophlebitis following administration of intravenous dantrolene. Tissue necrosis secondary to extravasation has been reported. Injection site reactions (pain, erythema, swelling), commonly due to extravasation, have been reported. Fatal and non-fatal liver disorders of an idiosyncratic or hypersensitivity type may occur with dantrolene sodium therapy.

To see the full prescribing information visit www.revonto.com.

© 2014 US WorldMeds, LLC. Revonto is a registered trademark of US WorldMeds, LLC.
In 2013, our 29 volunteer hotline consultants consulted on 481 calls. The calls originated mainly from the United States, some from Canada, and a few from outside North America, such as Cyprus and Germany. The pin map shows the locations of all the calls that originated from North America. These calls included 11 “mock” MH drills, which were handled by six hotline consultants. One of these MH drills came from South Korea.

Most calls originated from anesthesiologists, RNs, and CRNAs; however, many others came from different specialties such as surgeons, internists, pediatricians, and pharmacists. The complete list and the number of calls received can be seen in the graph to the right.

Incoming calls to the MH hotline were not restricted to normal surgical hours. Although most calls were received during the day, many were also received during the evening and overnight hours.

We categorized the types of calls into two major categories: simple inquiries and ongoing possible MH cases. The simple inquiries were classified as a preoperative preparation question (52 calls), an intraoperative question (99 calls), or a question related to a case that had already been completed (131 calls).

The ongoing possible MH cases were classified based on the diagnosis of the hotline consultant, as either “likely to represent MH” (30 calls), “unlikely to represent MH” (52 cases), or “suspicious case but not enough information to judge the likelihood of MH” (116 cases).

The vast majority of “unlikely to represent MH” cases concerned patients with postoperative hyperthermia without other concomitant signs and symptoms of MH.

Although unusual, postoperative hyperthermia can reach alarmingly high levels, even as high as 105°F. Without the other more typical signs of MH, such as rising end-tidal carbon dioxide levels, or muscle rigidity, this hyperthermia almost never represents MH. The exact cause of the high body temperature is rarely discovered, but may be due to a transient release of bacteria into the blood stream from the surgical site, or an effect of the general anesthetic altering the temperature regulating center of the brain. These patients should be treated with antipyretics (e.g., acetaminophen) and external cooling methods, such as tepid water baths.

Fortunately, in 2013, there were no deaths reported to the MH Hotline, as a direct cause of an MH event. The one death that was reported was directly related to the trauma of a motor vehicle accident.
Continued from page 2

offer assistance to begin a chapter in your community and works with our Development Coordinator, Michael Wesolowski, to handle the technical aspects of event coordination and promotion via the MHAUS website. She would be most happy to hear your ideas for products, programs, or fundraising events you would like to set up, as well as any quality improvement ideas you have to share.

We know that the members of our organization are focused and determined to help us reach our goal of 0 lives lost to MH! To make this a reality, we needed someone who could receive our members’ feedback and put a strategy into place to bring their ideas and thoughts to reality! MHAUS administration will benefit from the exercise as will MH-susceptibles and their family members.

Please join me in welcoming Kecia to the MHAUS Team and feel free to call her at 1-800-986-4287 or 607-674-7901 to welcome her to the group and/or share your story as to why you are a member of MHAUS!

Happy Spring!

Dianne Daugherty, MHAUS Executive Director
dianne@mhaus.org

I’ve Been Diagnosed as MH Susceptible – What Do I Do Now?

GET A MEDICAL ID TAG
The Medical ID Tag, developed by MHAUS and provided by MedicAlert Foundation, is engraved with an ID number, the words “MH-susceptible” and other conditions or allergies, as well as the 24-hour MH Hotline number. Medical professionals will have direct access to Hotline Consultants and important patient information in the event of an emergency. For more information, visit the MHAUS website, click on “Patients and Families,” scroll down to the FAQs and click on “ID Tags Program.”

SEND A LETTER TO YOUR LOCAL HOSPITAL(S)
MHAUS provides sample letters you can submit to your local hospital(s) alerting them of an MH-susceptible living in the area.

REGISTER WITH THE NAMHR
The North American Malignant Hyperthermia Registry of MHAUS maintains a central database of patients with MH susceptibility.

VISIT THE MH MESSAGE BOARD ONLINE
Communicate with other MH-susceptibles at www.mhaus.org.

LEARN ABOUT MH
Visit the MHAUS website for MH references material, FAQs, glossary of terms, and anesthetics information.

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 12-13 (October 2012 - September 2013) in memory of Lila and Jerry Lewis. We are most grateful for their support and special tribute gifts.

Life Benefactors
Dorothy Glassman
Gregory Lewis Glassman
Jacey Lila Glassman
Marilyn Lewis Glassman
Dr. Joseph Sugerman
Bob & Dianne Winters
Michele & Steve Lewis

Patrons
Brad & Julie Shames

Sponsors
Arline A. Hammer
Jacey Hayes
Neil Levy
Leonard Roberts
Les & Diane Surfas

Donors
Mr. & Mrs. Bill Rouse

Friends
Mitzi Birnbaum

Honoraria
In memory of Sharon Felder
Marilyn Lewis Glassman

Visit the MHAUS website to learn of the many avenues available to donate. You can choose how you want your donation dollars spent. Thank you!
Glossary of MH-related Terms

Contracture test – This is the test that is used to determine a patient’s susceptibility to MH. Muscle is taken from the thigh (about the size of a fingernail) and cut into strips of about one half inch long and mounted in a chamber and made to contract by electrical stimulation. When the anesthetic halothane is introduced in the chamber the muscle not only contracts but develops a contracture (a sustained contraction). This contracture is typical for MH susceptibles. The drug caffeine may also lead to an abnormal contracture, as may a variety of other anesthetics. Although the test is highly accurate, the inconvenience of the biopsy and the requirement for special technical expertise limits its use.

Creatine kinase – An enzyme found in cells, especially muscle cells. Normal levels are up to about 200 iu/L. In cases of muscle membrane breakdown, the enzyme leaks out of the cell. This may occur from any type of muscle trauma, including malignant hyperthermia. After surgery CK levels may normally rise to 1,000 to 2,000 iu/L. When there is severe muscle damage the level may rise to 10,000 or more. At these levels, the muscle pigment, myoglobin, can be expected to be elevated in the blood as a result of muscle damage. In other words, elevated CK is a marker for leakage of myoglobin from the cell. Elevated levels of myoglobin can lead to temporary or permanent kidney damage. After an episode of MH the CK levels may be mildly or dramatically elevated depending in part on the promptness of treatment. In general, peak levels of CK occur about 24 hours after injury and may be elevated for days. Hence, in suspected cases of MH it is important to determine CK levels. In case of heart muscle damage, CK may be elevated, but this represents a slightly different form of CK. CK from regular muscle is termed CK MM, from heart muscle, CK-MB.

Dexmedetomidine – A selective agonist used as the hydrochloride salt as a sedative for patients in intensive care units.

General anesthetics – Compounds that produce loss of consciousness, pain relief and amnesia. General anesthetics are either gaseous agents such as halothane, sevoflurane, and desflurane (all triggers of MH). Nitrous oxide is often used as an adjunct to these agents. It is not a complete anesthetic, and also not an MH trigger. There are a variety of agents that are given intravenously that also may produce anesthesia such as the barbiturates (e.g. thiopental), propofol, and ketamine. None are MH triggers. A variety of other agents are often used during anesthesia such as the narcotics, benzodiazepines (e.g. Valium and Versed) which produce pain relief and sedation.

Hypercapnia – Excessive carbon dioxide in the blood.

Iatrogenic – Induced inadvertently by a physician or surgeon or by medical treatment or diagnostic procedures.

Local anesthetics – These compounds block transmission of nerve impulses involved in pain sensation. These are the “caine” drugs - novocaine, bupivacaine, lidocaine, mepivacaine. None trigger MH and are safe to use in the MH susceptibles. These drugs are commonly used by dentists, anesthesiologists, pain physicians and surgeons among others.

LMA – laryngeal mask airway – This device was introduced into practice only a few years ago. The device is often used when tracheal intubation is not needed, but control of the airway is desirable. It is a tube that is so constructed that it does not enter the tracheal but forms a seal around the entrance to the larynx (the glottis). Insertion of the LMA is not as traumatic as insertion of an endotracheal tube and does not require deep levels of anesthesiatics or forceful muscle paralysis.

Molecular genetics – Genetics is the study of inheritance. Molecular genetics is the study of how changes in DNA structure, such as mutations, affect the function of the genes. Molecular, because the study of DNA entails understanding of molecular or submicroscopic changes.

Muscle relaxants – These are drugs that are more properly termed paralyzing agents. There are two classes of muscle relaxants, non-depolarizing and depolarizing agents based on their mode of action. Typical non-depolarizing agents are vecuronium, pancuronium and rocuronium. None are triggers of MH. However, the one depolarizing agent, succinylcholine is a potent trigger of MH. These agents are administered intravenously and are therefore given by anesthesiologists, ER physicians and intensive care physicians.

Neuroleptic malignant syndrome – (NMS) This is a constellation of signs and symptoms marked by high fever, muscle breakdown, ac- dasis, muscle rigidity and other signs similar to MH. However, the syndrome is induced by drugs used in the treatment of major psychiatric disorders. These drugs include thorazine, haloperidol (Haldol), olanzapine and other potent antipsychotic agents. The syndrome is not inherited and does not predispose to MH. That is, there is no greater frequency of MH in those experiencing NMS or vice versa. Interestingly, dantrolene is effective in treating NMS. There is no diagnostic test specific for NMS susceptibility.

Opiate – A medication or illegal drug that is either derived from the opium poppy, or that mimics the effect of an opiate

Oxygen saturation – The main purpose of the blood is to carry Oxygen to the various parts of the body along with nutrients and to remove carbon dioxide and other byproducts of metabolism. The amount of Oxygen in a given quantity of blood is not easy to measure, however the saturation level of the hemoglobin in the blood that carries the Oxygen can easily be measured with an external probe attached to a pulse oximeter. Normal Oxygen saturation is above 98%. At levels below about 90% insufficient oxygen is delivered to the blood, which may lead to many problems.

Pseudocholinesterase – An enzyme that degrades the drug succinylcholine. In about one in 2500 patients this enzyme is deficient. Therefore succinylcholine which usually causes muscle paralysis for about 5 minutes leads to paralysis that may last several hours. It is not life-threatening so long as the patient is connected to a ventilator. Susceptibility to this problem is not related to MH.

Reversal agents – There are several drugs that can antagonize or “reverse” the effects of other drugs. The drug, Narcan, or naloxone reverses the effect of narcotics (including the analgesia from these agents). Some drugs, neostigmine and pyridostigmine and edrophonium, reverse the effects of the non-depolarizing muscle paralyzing drugs.

Rhabdomyolysis – When muscle is damaged and cells are disrupted, the intracellular constituents begin to leak into the blood stream. This includes creatine kinase, myoglobin and the electrolyte potassium. This is termed rhabdomyolysis. This breakdown may be manifested by muscle pain and in extreme cases dark or cola colored urine.

Subcutaneous emphysema – Gases that are introduced into a body cavity, for example as part of laparoscopic surgery may, in some cases migrate from the body cavity to the tissues under the skin. This is called subcutaneous emphysema. It is recognized because a cracking sensation is felt on touching the skin. The gases eventually are absorbed into the blood stream.

Tachycardia – A rapid heart rate, usually defined as greater than 100 beats per minute.

Tracheal intubation and mainstem intubation – In order to control gas exchange during anesthesia a plastic tube is often placed in the trachea (windpipe). This is done usually when the patient is first anesthetized. One end of the tube is connected to a ventilator or respirator to control ventilation. Since the windpipe bifurcates just below the neck line, if the tube is inserted too deeply, the end may go into one of the branches of the trachea (usually the right side) and therefore only one lung will be ventilated. This may lead to a decrease in oxygen in the blood, and rarely an increase in carbon dioxide as well.

Trendelenberg Steep head down position.
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 $35.00, and includes the contracted service to transfer your call to a consultant, but this cost does not include 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 $35.00 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.

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: ____________________
❑ I am MH-Susceptible      ❑ I am a Medical Professional
Please charge my   ❑ Visa   ❑ Mastercard   ❑ Discover   ❑ American Express
Name on card: ___________________________________________________
Credit Card Number: _______________________________________________
CV Code: ____________________Expiration: ____________________
THANKS! MHAUS thanks the following State Societies of Anesthesiology – Alabama, Connecticut, Illinois, Maryland, Michigan, Pennsylvania, Wisconsin, and Texas – for their financial support. 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!

Ron Litman Joins MHAUS Board Of Directors
MHAUS is pleased to report that Dr. Ron Litman has joined the MHAUS Board of Directors effective January 2014. Dr. Litman is Attending Anesthesiologist at The Children’s Hospital of Philadelphia where he also chairs the Pain and Sedation Committee. Dr. Litman has held numerous faculty appointments and editorial positions as well as membership in many professional organizations, including MHAUS in which he has been a member and Hotline Consultant since 2001. Ron is also Medical Director of the MH Hotline.

Updated Ambulatory Surgery Center Manual Available
MHAUS has updated the Ambulatory Surgery Center (ASC) Procedure Manual. The goal of the manual is to prepare all members of the Surgery Center staff for an MH crisis. This manual uses worksheets to assign specific tasks to surgery center staff, provides complete checklists, and emphasizes periodic mock drills to minimize the risk associated with MH. An instructional DVD which outlines a response plan is also included. The ASC Procedure Manual is very helpful to train your staff for an MH crisis. Visit the MHAUS website or call 607-674-7901.

EMHG Meeting May 15-17, Wurzburg, Germany
The European Malignant Hyperthermia Group (EMHG) meeting will take place May 15-17, 2014, in Wurzburg, Germany. A draft of the program as well as information on how to register will be available soon. To check status on the upcoming meeting, visit www.emhg.org.

Inaugural Geoffrey Keller Open Water Swim, June 21, 2014
The inaugural Geoffrey Keller Open Water Swim will take place at Lincoln Trails State Park, Marshall, Illinois, on June 21. The open water swim is held in honor of the late son of Curt and Kathy Keller to support Malignant Hyperthermia awareness. Presented by Marshall, IL, the Geoff Keller Group of MHAUS, and Illinois Department of Natural Resources, the open water swim follows a rectangular course of 1.2 and 2.4 miles marked by buoys, lifeguards, rowboats, and other safety resources. Awards will be presented to overall top three men and women finishers in each age group. For more on the event, visit the Get Involved page on the MHAUS website.