



The Communicator

New In-Service Video Now Available

MHAUS is starting its 25th Anniversary year with a new and improved model of the ever-popular In-Service video. Viewers of the video (DVD and VHS) will be brought into the new millennium with a more entertaining, story-like presentation covering information needed to be prepared to recognize and treat an MH incident, including a mock incident. The

inclusion of a DVD was added to more readily meet the need for the variety of mediums used in medical facilities today.

Soon you will be hearing a new name associated with this particular item: the **In-Service Kit**. The actual title will remain the same – “Malignant Hyperthermia: Knowing Your Role” – and the content remains true to the high standards demanded by MHAUS with review by the MHAUS Professional Advisory Council. There are additional features within the program such as suggested Dantrium® IV mixing instructions, risk management strategies, and the program will even touch upon the exciting area of webcasting.

Be prepared to give your patients the best care available. The educational update you have been asking for is now ready to be a part of your MH Plan of Action! We would appreciate your feedback on the presentation once you have had a chance to view it, as MHAUS continues to look for ways to improve the quality of our MH education.

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MHAUS accepts payment via credit card, purchase order or check. Please allow 2-4 weeks for delivery. Contact MHAUS, PO Box 1069, Sherburne, NY 13460. Tel: 607-674-7901 Fax: 607-674-7910 Website: www.mhaus.org

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The Communicator is published four times each year by the Malignant Hyperthermia Association of the United States (MHAUS) and is made possible by a generous grant from Proctor & Gamble Pharmaceuticals, manufacturers of Dantrium. The Communicator is intended to serve the information needs of MH-susceptible families, health care professionals, and others with an interest in MH.

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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.

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MHAUS Marks 25th Anniversary

Dear Friends,

The year 2006 will mark the 25th Anniversary of the founding of MHAUS.

It's hard to believe all that has transpired since 1981. Malignant hyperthermia (MH) was first described in 1960; by 1980 there still was not very much information available concerning the clinical presentation and treatment of MH, even in the anesthesia medical literature. As a matter of fact, the impetus for the creation of MHAUS was the death of a young man from the syndrome and a cousin's frustrating experience in trying to learn the implications of the diagnosis for her own upcoming surgery.

Although the FDA had approved dantrolene in 1979, few places stocked the drug and most did not know how to use it. Of greater concern, many anesthesia providers, surgeons and internists had never heard of MH or knew the signs, and without that information, MH was sure to continue to destroy lives.

MHAUS was formed to overcome those problems. Founders Suellen Gallamore, Owen Davison, George Massik and Bob Luckritz, with my

assistance, were convinced that education and training, along with the availability of dantrolene, would go a long way to save many lives. It was impossible for us to conceive of the changes that were to occur over the next 25 years, including:

- better understanding of the clinical presentations of MH,
- clarification of drugs that precipitate MH,
- insight into the inheritance of the syndrome,
- new devices to provide early warning of MH,
- understanding of how best to use dantrolene, and
- standardized testing for MH susceptibility.

As a result, at present, recognition and management of MH is taught regularly in medical and nursing schools, and it is almost impossible to find an anesthesia provider anywhere in the world who has not heard of MH. Most importantly, deaths from MH are very infrequent.

Sincerely,

Henry Rosenberg, MD, CPE
President

The Malignant Hyperthermia Association of the United States is a not-for-profit organization dedicated to reducing the morbidity and mortality of malignant hyperthermia and other heat-related disorders by: improving medical care related to MH; providing support information for patients; and improving the scientific understanding and research related to MH and other kinds of heat-related syndromes.

For more information or for materials on malignant hyperthermia or MHAUS' programs, call 607-674-7901; write MHAUS, PO Box 1069, Sherburne, NY 13460; or visit us on the Internet at www.mhaus.org.

MHAUS Introduces New Speakers Bureau



Getting the word out about MHAUS can be done in many ways: through professional trade and general media, The Communicator,

and the MHAUS web site. MHAUS is now offering a new program shown to be successful in educating health care professionals about the latest MH issues – the MHAUS Speakers Bureau.

The speakers' presentations include topics such as MH Diagnosis and Treatment, a Basic Primer on MH, Lessons Learned from our MH Hotline Cases, and the new Molecular Genetics Test. Speakers include hotline consultants and members of our Professional Advisory Council. So far we have twelve who have signed up and are willing to share their

experience and expertise to further reduce MH morbidity and mortality!

The American Association of Oral and Maxillofacial Surgeons, the American Association of Nurse Anesthetists, and the Illinois Society of Anesthesiologists are already in the process of scheduling our speakers. Organizations are eager to educate their members during annual meetings or at smaller educational conferences. This allows us to not only inform medical professionals about MH, but also share information about our educational materials, such as our hospital, ambulatory surgery center, and office-based manuals, which prepare medical professionals for an MH episode. It also allows us an opportunity to speak one-on-one with our seminar participants, following our presentations, about their personal interest in the

MHAUS mission.

"Personal appearances are an important way to put a face on our organization," says Henry Rosenberg, MD, President of MHAUS and one of our MHAUS speakers. "Although many health care providers are aware of MH and how to respond, some are not up-to-date on the very latest treatment methods or how suspected MH patients and their family members should be evaluated. As we make our presentations, our audiences will get to know the many MHAUS educational offerings and will be able to optimize their ability to respond to an MH crisis and assist those with questions concerning MH."

You can find out more about our MHAUS Speakers Bureau by calling Al Rothstein at (866) 636-3342 or e-mailing mhaus@rothsteinmedia.com.

New Payment Code Approved For CHCT Test

The CPT for the caffeine halothane contracture test (CHCT) was approved and announced in the *Federal Register* in late 2005. The new code is 89049.

Representatives from the American Society of Anesthesiologists (ASA) and MHAUS were involved in preparing the list of equipment and personnel involved in the testing, and presenting the request for the new code and

its relative value to the American Medical Association (AMA) CPT Committee in February 2005. A representative from the Centers for Medicare and Medicaid Services (CMS) was also present to assist in guiding the proposal.

Now that this code is approved, it is no longer necessary to use the nonspecific neuromuscular physiologic testing code, which so often resulted in denials. Each payor will determine the amount of money they are willing

to pay the biopsy centers for their insured beneficiaries based upon the relative value of the code and the negotiations with each biopsy center (for example, in or out of coverage).

It is hoped this improved system of payments will more adequately address the real costs involved in providing this service and prevent more centers from closing from economic losses associated with lack of reimbursement for this service in the past.

Why They Give

by Al Rothstein

Twelve years ago Noah Small was playing competitive beach volleyball, had surfed the waves of Hawaii and crossed the Pacific on a sailboat. Not much made the 6'1", 185-pound agricultural consultant nervous, but that was about to change.

At that time, he was having routine surgery to remove gallstones, a problem that ran in his family. And he was suspicious about another genetic disorder as well – malignant hyperthermia.

"My uncle died during routine surgery on the operating room table in 1960, and I told my surgeon, anesthesiologist, and private physician before my surgery," says Small. "I told them I suspected that malignant hyperthermia could be a problem. Still, they gave me triggering anesthetics."

The result was an MH episode. Small was treated with dantrolene and recovered, but not before "waking up with violent

shakes and then not being able to move my limbs."

He believes that had his surgical care team been more educated, his episode would have been avoided. That's why he and his family are on a mission to educate others, both medical professionals and families.

They help educate by donating regularly to MHAUS, to support its educational materials, such as MH procedure manuals for the physician's office, the hospital, and ambulatory surgery center, and the In-Service Kit, which includes a video in VHS and DVD formats and a booklet with a test and continuing education credits. For more information on these and other MH educational materials for the medical professional and patient, visit www.mhaus.org.

MHAUS depends on the generosity of hundreds of individuals as well as a variety of businesses in order to alert the anesthesia and surgery community to the risks and dangers of malig-

nant hyperthermia. The Board and the staff of MHAUS are committed to eliminating death and disability from MH and MH-like disorders.

Henry Rosenberg, MD, President of MHAUS, says Noah and his family have experienced first-hand why the MHAUS mission is vitally important. To their credit, they have been dedicated to helping other families avoid the problems they have experienced.

"Nobody should have to suffer through the reaction if they don't have to," says Noah. "My kids and grandkids down the road need to be sheltered from the possible catastrophic effects of MH."

Small emphasizes that no matter how far advanced MH treatment is, there is still a need not just to have educational resources but to let people know where they can find them. "Before you can be your own advocate, you need information and people in your court," he says. "That's the value of MHAUS."

For example, during the birth of his now eight-month-old daughter, before the doctors gave his wife an anesthetic, he asked them to first call the toll-free MH Hotline (**USA and Canada: 800 644-9737, Outside the US: 315 464 7079**) as a precaution for MH. After the nurse did just that, she remarked that she had been unaware that so much information was available.

"It is amazing to me that doctors and nurses know about MH but don't know how to access

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P&G

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the information, for example, calling the MH Hotline," says Small. "It's all about information, and information costs money."

When Noah Small sees new advances in the field, made possible because of donations like his, he says "It's awesome when they can make strides to identify with confidence something that can prevent what happened to my uncle forty years ago. You can't put a price tag on that!"

"No amount of money should keep you from getting information to your loved ones."

Further up the western United States coast, the same beliefs about education spurred the Smith family to begin donating to MHAUS.

When Scott Smith was 18 years old in May of 1997, he went to the hospital for an appendectomy. Although he made it through the surgery, afterwards his temperature spiked to 106 degrees Fahrenheit. He was on a respirator temporarily because of fluid in his lungs, and his fever had to subside.

"Luckily our anesthesiologist recognized what was happening," remembers Scott's mother, Kim Smith. The hospital in the tiny coastal town of Florence, Oregon, used all of its dantrolene supply, along with that of other hospitals in the area, to save Scott's life.

The doctors at the hospital were convinced it was malignant hyperthermia, and the anesthesiologist told the Smiths about the Malignant Hyperthermia Association of the United States (MHAUS,

When Noah Small sees new advances in the field, made possible because of donations like his, he says "It's awesome when they can make strides to identify with confidence something that can prevent what happened to my uncle forty years ago. You can't put a price tag on that!"

www.mhaus.org). MHAUS then sent the Smiths basic information about MH and material such as a wallet card telling which anesthetics are safe.

That last item was critical because even though the family has had differing opinions from other doctors about whether Scott's episode was actually MH, neither Scott nor his family, which includes a sister and two children, have been able to get tested for it because the muscle biopsy test is expensive and it is hard for them to travel. But they are not taking any chances. Before any medical procedures, the family now asks surgeons to treat them as if they are MH susceptible.

"My daughter has had some surgery on her wrists, and because of the MH possibility they used local anesthetics instead of general," says Mrs. Smith.

The Smiths have been hoping for an easier test to detect MH, such as the new molecular genetics test, which was introduced in 2005. Because they have been hoping for such advancements, they began to donate money each year to MHAUS.

"We had never heard of malignant hyperthermia before Scott's incident," she says. "Then, when we found out how involved the muscle biopsy was, we were more than happy to start donat-

ing, in hopes that advances would be made. Also, at the time all of this happened to Scott, he had an infant daughter and there is no test for someone that small. The fact that our dollars help develop things like the molecular genetics test feels really good."

"With time, the molecular genetics test will make it easier for families like the Smiths to determine who in the family is at risk for MH," says Dr. Rosenberg. "Testing involves a simple blood test. The test is less expensive and more accessible. It is not at the stage where all families are eligible, and those who are, must qualify first. But it is an exciting advancement thanks to the generosity of families like the Smiths and the Smalls."

The Smiths are aware that only some qualify for the new molecular genetics test. To find out more, visit <http://www.mhaus.org/index.cfm/fuseaction/Content.Display/PagePK/MolecularGenetics.cfm>.

The Smiths have dealt with other illnesses, such as cancer, and know that research has helped them survive. And they have advice for those who are thinking about donating money to MHAUS, but haven't done so yet.

"If there is no money, there is no cure, no solution," Kim says.

Of Mice And Malignant Hyperthermia

by Henry Rosenberg, MD

One of the ways that scientists learn about the diagnosis, treatment and natural history as well as other features of a disease or disorder, is to study a similar disorder in animals or to create an animal model of the disorder. In the case of malignant hyperthermia, the recognition that certain breeds of pigs developed signs of MH in a reproducible manner allowed scientists to identify the characteristic changes in skeletal muscle that result in MH in humans. Working with these animals, scientists developed the caffeine-halothane test used to diagnose MH susceptibility, unraveled the genetics basis of the disorder, and have demonstrated how dantrolene could be used to treat MH.

However, naturally occurring MH in pigs is not exactly the same syndrome as that which occurs in humans. For example, MH susceptibility in pigs is inherited in a homozygous manner (i.e., a double dose of the gene defect), whereas in humans it is inherited in a heterozygous manner (i.e., one genetic change being sufficient). MH susceptible swine trigger with stress, whereas it is not clear if that occurs in humans, or if it does, it is a very rare occurrence.

Since the discovery of the gene mutation that causes MH in pigs in 1990, enormous advances in molecular genetics of MH in humans has been made. In recent years it has become possible to incorporate selected segments of DNA into a developing animal so that the animal will express certain characteristics that the ordinary

(or wild type) animal does not. Such selective changes in DNA are very useful in producing animal models for human disorders or diseases.

In the recent issue of the *FASEB Journal*, investigators from Baylor College of Medicine, University of Rochester, and University of California, Davis describe the development of mice that harbor one of the genetic mutations that are known to produce MH in humans. The mutation that was introduced into the animal's genetic makeup is one of the DNA changes found in the ryanodine receptor gene that result in MH in humans. If the embryonic mice have a double dose of the gene (recessive) they do not survive; however, if they are heterozygous (as in humans), they are healthy and appear normal. However, when heterozygous mice are exposed to an inhalation anesthetic agent (isoflurane), they develop the characteristic signs of MH, namely, muscle rigidity, high body temperature and death. In addition, there is evidence of muscle breakdown and electrolyte changes similar to that observed following an MH episode in humans. Furthermore, isolated skeletal muscle obtained from heterozygous mice contracts abnormally in response to caffeine and other agents in a manner identical to that observed for skeletal muscle obtained from MH susceptible humans and pigs. This difference in caffeine responsiveness was found to arise from changes in the sensitivity of the muscle to release intracellular calcium stores in muscle derived from the genetically modified mice. Again, this is identical to

findings in MH-susceptible humans and pigs.

There are many other physiologic similarities between the MH-susceptible mice and naturally occurring mice. One of the interesting and thought provoking observations made in these animals is that they develop the typical MH changes when exposed to high environmental and body temperatures. Furthermore, the development of MH is delayed or prevented if the body temperature of the mouse is below normal. Whether this enhanced temperature sensitivity also occurs in human patients with this particular mutation or whether it only occurs in mice because they have different thermoregulation and genetic backgrounds than humans is to be established.

The creation of animals such as this is an important advance in our understanding of the MH syndrome and will provide us with insights in the pathophysiology and unusual presentation of this syndrome. I am sure that mice and other animals with DNA changes that are also known to produce MH will be produced in the near future. Our understanding of MH and our ability to help patients and practitioners is sure to improve.

Information about the article:

Heat- and Anesthesia-Induced Malignant Hyperthermia in an RyR1 Knock-in Mouse Mihail G. Chelu¹, Sanjeeva A. Goonasekera², William J. Durham¹, Wei Tang¹, John D. Lueck², Joyce Riehl³, Isaac N. Pessah³, Pumin Zhang¹, Meenakshi B. Bhattacharjee⁴, Robert T. Dirksen² and Susan L. Hamilton¹

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Honor Your Anesthesia Care Professional

MHAUS has developed a way you can recognize a particular anesthesia professional in print! Through our program called "Honor Your Anesthesia Professional," we offer the following: For a \$50 donation or more, your personal thank you message can be directed to your "special" anesthesia professional in our quarterly newsletter, *The Communicator*. We will print your personal message (25 words or less) as well as highlight the names of those professionals in a prominent location in *The Communicator* for a full year.

Additionally, recognition will be highlighted on the MHAUS website reserved for this elite group. If an address is available, MHAUS will send a congratulatory letter to the anesthesia professional passing along the appreciation of one of his/her patients.

If you feel your anesthesia professional deserves your special thanks, please call or email the MHAUS administrative offices to express your appreciation of their outstanding care!

Honored Summer 2005

LCDR Alan Lovejoy is Chief Nurse Anesthetist aboard the USS Vinson

(CVN 70) in the Gulf war zone. Our son, Petty Officer Jonathan Pasquinelli, is assigned to this ship as part of the "air wing" and required an emergency appendectomy while on board.

He had an MH episode and LCDR Lovejoy "just happened" to know what it was, recognized it, and reversed it in time to save our son's life. Thank you, LCDR Lovejoy.

*J. Jeffrey Pasquinelli
M.A. Pasquinelli
Carson City, Nevada*

The Lila and 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 05-06 (Oct. 2005 - Feb. 2006) in memory of Lila and Jerry Lewis. We are most grateful for their support and special tribute gifts.

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Correction

In our last issue, there was an error in the FY 04-05 MHAUS Contributor List/insert. Gerald and Elaine Rosenberg as well as Ronald Ziegler were inadvertently omitted from the list. We sincerely apologize.

Have you visited us lately? Log on to www.mhaus.org to get the latest information on MH, order materials, post a message to the bulletin board or learn about the "Hotline Case of the Month."

MH Hotline Activity For April - August 2005



by James W. Chapin, MD

Malignant Hyperthermia Hotline Consultants answered calls referred by the Poison

Control Center nurses between April and August 2005. Consultants during that time were Drs. Bandom, Gronert, Herlich, Litman, Melton, Miller, Millman, Parness, Rosenbaum, Rosenberg, Tobin, Watson, Weglinski, Wong, and Chapin. Calls came from 32 states and Ontario, Canada. Of the 114 calls received, 65 were clinical reports and 49 were questions only. Of the clinical reports, 49 were judged unlikely to be MH and 16 were probable to definite MH. Eight of the unlikely MH group received dantrolene, and 12 of the 16 probable to definite MH group received dantrolene. Nine of the patients had trismus (jaw stiffness) following succinylcholine (a paralyzing agent used by anesthesia providers and is also an MH trigger). Only one of the trismus patients was thought to have an MH reaction and had a CK of 50,000 (enzyme from muscle that leaks out with an MH crisis; the normal value is about 200). The majority of the callers were anesthesiologists, the rest were CRNAs, adult and pediatric intensivists, RNs, and pharmacists.

The most frequent clinical sign prompting the call for a clinical incident was a fever, followed by elevated exhaled CO₂,

trismus, rigidity, and tachycardia (fast heart rate). The most frequent question was: I have a patient with a family history of MH, what technique should I use or should I send the patient to a bigger hospital for their anesthetic and operation? MHAUS feels that the anesthesia provider should be comfortable in caring for MH susceptibles, but a history of MH susceptibility does not mean that the patient's surgery care must be rendered at a hospital or a hospital that cares for very ill patients. Questions about the MH cart contents, how much dantrolene is needed, dosing of dantrolene, clean machine, safety of a tetracaine spinal anesthetic, were other inquiries. One question was about electroconvulsive therapy (ECT) for depression and the need for dantrolene immediately available since succinylcholine is used. The response was yes, since there have been reports of MH being triggered by succinylcholine alone without inhalational agents. Another caller was about to anesthetize a patient with periodic hypokalemic paralysis and read on the MHAUS website that it was associated with an MH-like response to anesthesia. The consultant response was to use a safe technique.

A call to the hotline involved a methamphetamine (speed) overdose in a 15-year-old who had a high fever (107°F), rhabdomyolysis (leak of cellular contents from muscle cell), CK of 100,000 and heart, liver and renal failure. Hotline consultants recom-

mended dantrolene to help with the fever and muscle abnormalities, which resulted in some clinical improvement and lowering of temperature and CK levels. (Editor's note: there are some reports that dantrolene has helped in recovery from such a reaction, while others have found no difference in outcome. This depends on how rapidly the drug is administered and in what dosage.)

Another call involved an elderly patient with a definite MH reaction who was successfully treated with dantrolene, but required several days of mechanical ventilation partly due to weakness from dantrolene, which was continued for 24 hours. The Hotline consultant web chatline discussed the current recommendations of proper dosing of dantrolene and whether it should be changed. A recent article in the journal *Anesthesia Analgesia* by Podranski suggests an infusion regimen adjusted to initial bolus doses of dantrolene would give more stable serum concentrations. Maybe a lower dose for the elderly is indicated. (Editor's note: The Professional Advisory Council and the hotline consultants will be formulating a recommendation concerning dantrolene dosing after an MH episode.)

Several interesting questions related to the care of the pregnant patient were asked. One involved a parturient requiring a general anesthetic for an emergent C-section who received succinyl

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In the U.S. and Canada, the MH Hotline is 1-800-MH-HYPER (1-800-644-9737) Outside the U.S., call 1-315-464-7079

choline. After the C-section it was learned that the mother had a positive family history of MH. The concern was about the risk of MH in the baby. The consultant thought there was little risk since very little of the succinylcholine crosses the placenta. Another question centered on the drug Hemabate used to increase uterine contractility after a C-Section and to decrease blood loss. The question was if Hemabate is a trigger for MH. The consultant thought it was safe.

A patient with hyperthyroidism (overactive thyroid gland) was in recovery room following a removal of the thyroid gland and developed agitation, diaphoresis, and lower extremity rigidity that resolved with administration of a muscle relaxant. The consultant thought it was not MH but most likely Serotonin Syndrome (excess serotonin, a brain transmitter; signs of this syndrome closely resemble MH) due to the antidepressant medication the patient was taking. Another call that I took was one involving muscle rigidity in the recovery room that turned out to be Hyperventilation Syndrome with carpal-pedal spasm (hand and foot muscle spasm) and a CO_2 in the low 20 range (normal is 40). Since the signs of this syndrome relate to the patient breathing so rapidly and

deeply that they reduce carbon dioxide to very low levels, treatment involved having the patient breathe into a bag! I also received a call about a pediatric patient with tyrosine hydroxylase deficient dopamine responsive dystonia for a gastrostomy tube placement. (Editor's note: This is a very unusual genetic disorder not related to MH.) The patient was on chronic dantrolene for muscle spasticity. I researched the literature and found no correlation with MH, and recommended either inhalation or TIVA (total intravenous anesthesia) non-triggering technique. The patient did fine.

I am always amazed at how thankful and appreciative the callers are to have the MH Hotline and that we take the time to help them. I learn a lot about MH as a consultant and think we provide a great service.

Challenge Yourself With The MH Case Of The Month

Have you challenged yourself with the new MH Case of the Month? Visit www.mhaus.org and go to the *Home or Professionals' Info Center* pages to think about the correct way to proceed with these actual MH cases. Answers with narratives are provided for the previous month's cases.

Meet This Issue's Hotline Consultant



James W. Chapin MD, is a Professor of Anesthesiology, University of Nebraska Medical Center, Omaha, Nebraska. He has been on faculty for over 30 years. He has been a Hotline Consultant nearly 20 years. UNMC was an MH muscle biopsy center for many years. He worked with Dr. Dan Wingard on MH research. He is Director of Liver Transplant Anesthesia and Anesthesia Residency Program Coordinator.

Some Helpful Malignant Hyperthermia Terms

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.

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.

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.

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, emergency room physicians and intensive care physicians.

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.

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.

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 trachea (the glottis). Insertion of the LMA is not as traumatic as insertion of an endotracheal tube and does not require deep levels of anesthesia or muscle paralysis.

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.

Neuroleptic malignant syndrome (NMS)

This is a constellation of signs and symptoms marked by high fever, muscle breakdown, acidosis, 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 thiorazine, 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.

Reversal agents

There are several drugs that can antagonize or "reverse" the effects of other drugs. The drug, Narcan, or naloxone reversed 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.

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.

Triggering agents for MH

These are drugs that will lead to the onset of MH. These include all the potent gas anesthetics and succinylcholine.

Slide Show Presentation For MH Risk Available

MHAUS offers a slide show kit (CD-ROM and slides format) with lecture notes on "Managing Malignant Hyperthermia Risk in Today's Surgical Environment." This presentation reviews the risk of MH and assesses current trends in the management of MH in the inpatient and outpatient settings. Two CMEs are available.

This is a valuable tool to assist in developing standard of care practice guidelines and algorithms to ensure patients at risk will have access to appropriate interventions for treating MH. This program is arranged so that it can also be used as a self-study program to enhance individual knowledge of MH and the risks involved.

Cost is \$135 plus shipping and handling for the slides and CD. Call 607-674-7901 or visit www.mhaus.org to order.

Plan To Attend The Patient Mini-Conference Scheduled For July 15

Ever have questions, but forget them until after you left the doctor's office? Ever have a question you want answered, but don't wish to post it on the MHAUS message board for all to see? How about the basic questions regarding MH from a medical professional's perspective?

Well, here's your chance! You can get those questions and more answered by attending the upcoming MHAUS mini-conference for patients and medical professionals alike. The conference is scheduled for July 15,

2006, at Carroll Community College in Westminster, Maryland.

If you are interested in attending this very informative meeting and would like more information on how to get on the attendee list, please contact the MHAUS administrative office via phone at 607-674-7901 or email to info@mhaus.org.

Watch the MHAUS Website, so you don't miss this enlightening meeting with a chance to "ask the MH experts" your "burning" questions face-to-face.

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

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MHAUS Happenings, Events and Notices

□ **THANKS!** MHAUS is grateful for the financial support of the following State Societies of Anesthesiology: **Connecticut, Florida, Maine, Michigan, Ohio and Pennsylvania.** Our appreciation also goes to the following state components of the American Society of PeriAnesthesia Nurses: **Arkansas, Kansas, Missouri, New Hampshire, Texas, Vermont and Wisconsin.** Call the MHAUS office today to ask how your group can help.

□ **P&G Establishes Phone System For Ordering Dantrolene In The U.S. & International Markets** For information regarding ordering dantrolene sodium for injection, please contact P&G Pharmaceuticals Customer Service in the U.S. at 1-800-448-4878 or in Canada at 1-800-265-8676. Requests for information outside the U.S. and Canada can be sent

via fax to +49-6151-877-601 (in Germany). Dantrium® IV full prescribing and product information can be found on the Internet at www.pgpharma.com.

□ MHAUS/NMSIS 2006 Meeting Schedule

3/21-23 – Association of PeriOperative Registered Nurses (AORN), Washington, DC.

4/30-5/4 – American Society of PeriAnesthesia Nurses (ASPN), Orlando, FL.

5/17-19 – The Federated Ambulatory Surgery Association (FASA), Orlando, FL.

5/21-24 – American Psychiatric Association (APA), Toronto, Canada.

8/6-8 – American Association of Nurse Anesthetists (AANA), Cleveland, OH.

10/8-10 – American Society of Plastic Surgeons (ASPS), San Francisco, CA.

10/13 – Society of Pediatric Anesthesia (SPA) – 1 day meeting – Chicago, IL.

10/14-18 (exact exhibiting dates not yet received) – American Society of Anesthesiologists (ASA), Chicago, IL.

10/22 – New York Chapter PeriAnesthesia Nurses, RIT Rochester Institute of Technology, Rochester, NY.

10/29 - 11/2 – American Society for Healthcare Risk Management (ASHRM), San Diego, CA.

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