MHAUS Happenings, Events and Notices

THANKS! MHAUS thanks the following State Societies of Anesthesiology — Alabama, Connecticut, Illinois, Maryland, Michigan, Pennsylvania, 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!

Two Book Recommendations

Basics of Pediatric Anesthesia by MHAUS MH Hotline Medical Director Ron Litman is available as an e-book on iTunes.

Condolences
It is with sadness MHAUS learned of the recent death of Dr. Mary Lehan, a founding member of the European Malignant Hyperthermia Group (EMHG) and clinician of the Cork MH Unit. We remember Mary from EMHG meetings that she attended and from various international MH workshops. Our condolences go out to her family.

Congratulations Steve Howard
Steve Howard, MD, Veterans Affairs Palo Alto Health Care System, Palo Alto, CA, and a member of MHAUS‘ Professional Advisory Council, has been made Chair of the Anesthesia Patient Safety Foundation (APSF) Scientific Evaluation Committee.

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.

We Want To Hear From You
How can we better serve you? Call 607-674-7901 or email info@mhaus.org. Your comments and suggestions are important.

MHAUS Board Of Directors Holds Strategic Planning Meeting, Elects New Officer, Sets Goals

by Henry Rosenberg, MD
MHAUS President

In August the MHAUS Board held its annual strategic planning meeting followed by election of officers. I am pleased to report that Bonnie Denholm, MS, BSN, RN, CNOR, of the Association of Operating Room Nurses was elected to the position of vice president, succeeding Mr. Ron Ziegler, who retired after many years as a faithful Board member.

Bonnie was appointed to the Board in 2010 and brings to MHAUS the views and expertise of operating room and recovery room nurses, both of whom are wonderful supporters of MHAUS.

The two-day planning meeting reviewed the previous year’s activities and developed plans for the coming year, including several new programs and further enhancement of the MHAUS website. Many projects are on the drawing board for the coming year including a revised guide for managing MH in the ambulatory surgery center, and a program called “MH Genetics 101” that will provide an easy-to-understand explanation of the complexities of diagnosing MH susceptibility through genetic testing as well as practical issues related to getting the test done and paid for by insurance companies. Yet another program under development will establish a site visit program by one of our experts to all types of facilities (hospital, ASC, or outpatient center) to evaluate preparedness for an MH episode, combined with an update on managing MH.

All of this is in addition to the ongoing “MH mini-conferences” (see page 9) and preparation and updating of our printed material. Furthermore, there has been extensive reconfiguration of the MH website. Michael Wesolowski, our development coordinator and PR person as well as our de facto staff webmaster, has been reconfiguring the MHAUS website in order to make information easier to find, create the capabilities of creating groups and chapters, and further developing extensive social networking capabilities. A small group of individuals with diverse levels of MH interest, from patients to MH Hotline consultants, have been working with Dr. Sheila Muldoon, web-site oversight committee chair, and spent considerable time reviewing the website with Wesolowski.

On the inside
Remember MHAUS .................. 2
Ways To Contribute ............... 3
Five Honored At Recognition Reception 4
Director Outlines MH Hotline Changes 5
MH Scientific Conference A Great Success 6
“Let’s Save A Life” .................. 9
Happenings ....................... 12
Remember MHAUS When Planning Your Year-End Charitable Contributions

As we begin to prepare for the upcoming New Year, enjoy the holiday season, the Malignant Hyperthermia Association of the United States (MHAUS) Board of Directors and staff would like to take this opportunity to thank you for your enthusiasm and personal efforts to remain prepared for MH throughout the past year.

We embark this new year addressing internal financial challenges as are many others in this time of uncertainty and adjustment. We need your help to continue to provide lifesaving patient safety programs, such as the MH Hotline that assists healthcare professionals 24/7/365 with MHAUS’ volunteer MH experts’ advice and guidance as they face an MH event (and the possible loss of life).

Please consider making a donation to MHAUS as part of your generous charitable contributions this year!

Read below what others have to say about the important role MHAUS has played in saving lives.

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.

❑ Visa  ❑ MasterCard  ❑ Discover  ❑ American Express

Name on card: ____________________________________________

Credit Card Number: _____________________________________

Expiration Date __________________________

Signature: __________________________

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: __________________________

Please clip out this handy coupon, or feel free to photocopy if you prefer to keep your issue intact, then mail to: MHAUS, PO Box 1069, Sherburne, NY 13460-1069

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Ron Litman, DO, Professor of Anesthesiology and Critical Care at the Hospital of the University of Pennsylvania and the Children’s Hospital of Philadelphia, University of Pennsylvania School of Medicine, and Medical Director of MHAUS’ MH Hotline.

“It was a great honor and privilege to be named the Medical Director of the MHAUS MH Hotline this year.

Why do we do this without financial remuneration? Because the intellectual and charitable satisfactions are satisfying beyond measure. There is no other similar organization that provides this vital service to anesthesiology community and our patients.

Healthcare professionals can support MHAUS to ensure experts are always available to answer your questions about MH, help your patients and their family negotiate the diagnostic process, and guide you through the management of an acute life-threatening MH event.”

Lisamarie Avallone-Groetzner, Advocate for Central Core Disease and Malignant Hyperthermia Awareness and Education.

“When our son Vincent died suddenly on June 14, 2010, we had no idea what happened or why. He was fine one moment and the next complaining of stiffness in his legs, extremely rapid heart rate, and increasing body temperature. After rushing him to the Emergency Department, after they continued on page 3
attempted to secure an airway with succinylcholine, he went into cardiac arrest; resuscitative efforts failed. I would not rest until I understood what took my son’s life just two weeks before his 7th birthday. After six weeks, the medical examiner advised she was 99% sure that Vincent had a Malignant Hyperthermia episode. Further testing was needed to prove it.

“After talking with MHAUS, we coordinated transfer of Vincent’s tissue sample for testing. We had no choice but to go forward in the hope of getting answers for our three other children. We could not lose another. Vincent’s DNA showed an RYR1 variance, which is directly linked to MH. My husband and two of the children have the same variance.

“I cannot stress enough the importance and value of genetic testing. Without it, we would not have known how to proactively care for our children to prevent this from happening again. Knowledge is power and if having gone through all this can save even one life, it will have been worth it.”

Henry Rosenberg, MD, CPE, Director of Medical Education and Clinical Research at Saint Barnabas Health, and President of MHAUS.

In about 600 cases the provider calls the MH Hotline and gets rapid, specific advice from a knowledge consultant 24/7, 365 day a year. AT NO CHARGE.

“For a relatively small organization, MHAUS has made a huge impact on anesthesia patient safety and on raising awareness of MH and MH-like disorders. We appreciate the generosity of our many members and donors and hope that our lifesaving programs can continue to be offered, expanded, and with the help of modern technology, made available to patients and professionals at a moment’s notice.”

“Please demonstrate your recognition of the voluntary efforts of the MHAUS experts through your contribution.”

Your support will not only help us provide this lifestyle but also continue to provide products and programs specifically designed as resources to our customers – using their feedback and suggestions as our initial impetus. From this process, we have worked with other disciplines to develop pre-emptive transfer plans for patients involved in unexpected MH events while in ambulatory or office-based settings and MH mock drills designed to shorten response time in an MH event. We are presently working on a introduction of a product to address the needs of a patient diagnosed with MH, needs of their family members, and needs of their healthcare providers as well, so they all understand the options available for testing and life-management choices.

“We continue to update our website as it shares important resources and data for all. We truly appreciate any and all support, and thank you for considering MHAUS and our ultimate mission of saving lives!”

Sincerely,

Dianne Daugherty,
MHAUS Executive Director

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!
Five Honored At MHAUS Annual Reception

by Henry Rosenberg, MD
MHAUS President
At the recent American Society of Anesthesiologists’ meeting, MHAUS hosted its annual recognition reception. The reception was attended by about 50 people.

Dr. Harvey Rosenbaum, MD, UCLA Medical Center, Los Angeles, CA, faithful MH Hotline consultant for 22 years, received the Hotline Partnership award for his advice concerning an MH case that started in an ambulatory surgery center. The anesthesiologist, Dr. Edward Gronka, MD, Piedmont Plastic Surgery Center, Fayetteville, GA, and Dr. Rosenbaum were in communication throughout the treatment and movement of the patient to the hospital. I am pleased to state that the patient survived without problems due to early recognition and prompt treatment.

Each year MHAUS offers a manuscript award, the Daniel Massik Award, named for Daniel Massik who died from MH many years ago: the award is generously supported by the Massik family. The award is given to either a trainee or anesthesiologist who has recently given to either a trainee or anesthesia provider who has recently given to the Hotline Partnership award for their manuscript, in peer-reviewed journals. I am hopeful this one will as well.

The MHAUS special recognition award was made to Lisamaria Avallone-Groetzner of Mount Dora, Florida, whose young son died after experiencing an awake episode of MH. Groetzner described the circumstances of the tragedy and the agony it caused her family. At first, no one could explain why her six-year-old son, whose only medical problem was a curvature of the spine, suddenly developed rigid muscles, high fever, and increased heart rate. When he was brought to the emergency room because of these signs, the physicians administered the paralyzing drug, and MH trigger, succinylcholine, in order to open his mouth to insert a breathing tube. That rapidly resulted in a fatal heart rhythm.

The cause of death was thought to be related to meningitis, but that was ruled out very quickly. However, a very conscientious and astute medical examiner, Dr. Wendy Lavezzi, put the pieces of the story together and suggested that her son might have developed MH. After contacting Dr. Barbara Brandom and myself to discuss the likelihood that the cause of the problem might be MH susceptibility, the family was referred for diagnostic testing at the MH diagnostic testing center at the Uniformed Services University headed by Drs. Sheila Muldoon and John Capachione. The diagnosis was confirmed by genetic testing with the help of Dr. Khishge Sambughin and then Mr. Groetzner underwent a muscle biopsy contracture test, which was positive for MH susceptibility. Further diagnostic testing on continued on page 5

2013 MH Scientific Conference Presentations

The following were presentations at the 2013 Malignant Hyperthermia (MH) Scientific Conference held at the Chestnut Conference Centre, University of Toronto, Canada, in November, and sponsored by Malignant Hyperthermia Association of the United States (MHAUS) in conjunction with The University of Toronto.

Session I: RYR1 Physiology and Pathophysiology
- Filip Van Petegem, University of British Columbia, High Resolution Structures RYR1 Domains
- Clara Franzini-Armstrong, University of Pennsylvania, Impact of EC Coupling Proteins on Triad Structure
- Susan L. Hamilton, Baylor College of Medicine, Modification of RYR1 Function: Environmental and Pharmacological
- James J. Dowling, Hospital for Sick Children and University of Toronto, RYR1 Related Myopathies: Genotype-Phenotype Correlations and New Treatment perspectives

Session II: RYR1-Associated Disorders and Treatment
- Jerome Parness, University of Pittsburgh, Dantrolene Mechanism of Action: Uses in MH and Other Disorders
- Georgiina D. Vladutiu, University at Buffalo, RYR1, CACNA1S and Statin Myopathies
- Francis O’Connor, Uniformed Services University of Health Sciences, Heat/exercise-induced Rhabdomyolysis and MH: Clinical Perspectives
- John Capachione, Uniformed Services University of Health Sciences, Heat/exercise-induced Rhabdomyolysis and MH: Genetic Perspectives

Session III: Diagnosis of MH Susceptibility: Current and Future
- Sheila M. Muldoon, Uniformed Services University of Health Sciences, Contracture Testing: Updates and Standardization
- Khishge Sambughin, Uniformed Services University of Health Sciences, MH Genetic Testing in the Genomic Era
- Philip Hopkins, University of Leeds, MH Genetic Screening: RYR1 Variants, Exome Sequencing, New Genes

Session IV: Practical Clinical Cases
- Keynote Lecture: Dr. Wayne Chen, University of Calgary, “Mechanisms and Treatment of RYR-related Diseases”

Session V: Evaluating Risk in Patients with RYR1 Variants
- James Weber, Prevention Genetics, Commercial Laboratory Experiences with Genetic Testing of MH
- Mark Tamopolsky, McMaster University, Evaluation and Guidance of Patients with Myopathy

Session VI: Databases and How They Influence Clinical Care
- Donna Maglott, National Center of Biological Information, Role of Genetic Data in the MH Assessment and Resources for Scientists, Clinicians, Genetic Counselors, and Patients
- Barbara Brandon, University of Pittsburgh, Contributions of NAMHR to the Global Rare Diseases Registry
- Henry Rosenberg, Barnabas Health, Role of MHAUS in Gathering Data, Dissemination, and Education

“Let’s Save A Life” Mini-Conference
March 29th, 2014 Rochester, New York

MHAUS in conjunction with University of Rochester Medical Center (URMC) will present “Let’s Save A Life,” a one-day mini-conference on Saturday, March 29, from 10 am to 4 pm, at the URMC School of Medicine and Dentistry Entrance, Class of 62 Auditorium G-9425, 415 Elmwood Avenue, Rochester, NY 14642.

The conference is open to healthcare professionals, students, patients, and their families. Educational credits are available and lunch is included. Learn the basics of how to recognize, diagnose, and treat MH. Conference attendees will have the opportunity to talk directly with MH experts affiliated with MHAUS.

For more information contact Fay at fay@mhaus.org or call 607-674-7901
MH Scientific Conference
A Great Success
by Henry Rosenberg, MD
MHAUS President

The recent scientific conference in Toronto was a great success! I want to thank all those who participated (and there were a lot of people).

The organizing committee, especially Sheila Fiazi and Bob Dirksen and Sheila Muldoon along with the staff, organized a well thought out series of presentations, and the staff made sure all ran smoothly. I also would like to recognize the contributions of Dr. Brian Cavanaugh, Chair of Anesthesia at University of Toronto, who founded the funds to help out with the costs of the conference.

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The conference was about one in 350 random individuals, that the prevalence of pathologic sensitivity about the relation of MH to heat stroke and exercise-induced muscle breakdown in addition to a variety of rather uncommon muscle disorders.

The staff and Board of MHAUS surprised me at the reception with a lovely plaque recognizing my accomplishments and dedication to the goals of MHAUS. It was not necessary, but I appreciate the recognition and thanks.

Dr. David MacLennan, our honoree and a pioneer in the study of the genetics of MH, attended the meeting, the dinner, and the keynote presentation by Wayne Chen, one of his former associates. We presented Dr. MacLennan with a plaque of appreciation, and I believe he was very pleased at the recognition. Those of you who were present were treated to some fantastic presentations on the biology and structure of the ryanodine receptor gene (RYR1). Recent research has shown that DNA changes in this gene are responsible for MH susceptibility in over 50% of MH-susceptible patients as well as a variety of muscle disorders, referred to as congenital myopathies.

It was coincidental a recent publication in Anesthesiology found that the prevalence of pathologic DNA variants in the RYR1 gene was about one in 350 random individuals. This is significantly higher than has been found previously.


It is quite obvious that MH is the tip of the iceberg of disorders associated with RYR1 mutations, so the anesthesia community should take note of the wide-ranging implications and be a leader in patient care beyond the OR. I think there will be more opportunities than ever before for partnerships with other organizations (such as AORN and the Muscular Dystrophy Association and the neurology community, to name a few).

As we had done in the past, there were several students/residents who took notes on the presentations and will provide a synopsis of the lectures. With presenters’ permission, we will post or distribute the slides of the meeting as well as the posters.

Watch for a more detailed summary of the Scientific Conference presentations in the next Communicator.

continued from page 4

their children showed that two of the three also have the genetic change found in MH.

Groetzner and her husband now devote themselves to raising awareness about MH among emergency medical professionals and EMTs. It is only now becoming apparent that many health care providers other than anesthesiologists and operating room personnel need education for preparedness for MH. MHAUS will need to expand our educational efforts significantly to raise awareness about the relation of MH to heat stroke and exercise-induced muscle breakdown in addition to a variety of rather uncommon muscle disorders.

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MH Hotline Medical Director Outlines Upcoming Changes

Medical Director of MHAUS’ MH Hotline Ron Litman, DO, outlined upcoming changes and improvements to the Hotline and MHAUS website at a breakfast meeting of Hotline consultants held during the recent American Society of Anesthesiologists’ annual meeting.

One of Litman’s top priorities is recruiting new, young Hotline consultants. “More importantly I would like to have more than three people on call at a time. So I encourage you to actively recruit young people,” Litman urged. “I already have two people interested that couldn’t make the meeting,” he added. “One of the discussions we will have is how to formalize the recruitment process.”

Litman praised the work of MHAUS Vice President of Scientific Development Sheila Muldoon, MD, for her work of updating the MHAUS website material. He encouraged consultants to visit the website and look over the improvements themselves. Litman said he has been collaborating with MHAUS Development Coordinator, Michael Wesolowski, on a searchable database of MH online discussions that can be posted on the MHAUS website for easy access. This is but one improvement in store for the website and the Hotline.

“I had asked everyone when I took over this position for ideas for improving the Hotline. I didn’t get a chance to respond (to everybody), but I did look at everyone’s suggestions and agreed with all of you,” he said.

Litman agreed that one of the most frustrating problems involves the difficulty following up after a call; he assured consultants that the process would improve in the future. He also told consultants that he has been working on developing a call calendar with a rotating schedule which would include time zones so consultants would be called based on the time zones where they live.

Litman also agreed that entering calls into the current database is cumbersome and said he has been investigating a better database called RedCap. As well, he admitted the current system doesn’t do well at capturing data from Hotline consultants.

“We have all the audio files (from Hotline calls) and we are currently in the process trying to figure out how to get a staff member trained to listen to the audio calls and take that information and put it in the database for us.”

Litman said, “We really only get about 600 calls a year. What we would like to do is capture the calls that are likely MH.”

As a means of encouraging AMRA reports, Litman reminded everyone that someone who reports an MH case receives a free MHAUS membership for one year. Litman also encourages callers to become MHAUS members.

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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, dizziness, and diaphoresis, and tissue necrosis at injection sites not secondary to extravasation. There have been case reports of pulmonary edema, urticaria and erythema. Symptomatic hepatitis and non-fatal renal failure 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 overdose, 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 date. There is always some lag period between manufacturing date and when the product ships to the end user, which varies based on the order placed.

Dantrium® Intravenous (dantrolene sodium for injection)

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, dizziness, and diaphoresis, and tissue necrosis at injection sites not secondary to extravasation. There have been case reports of pulmonary edema, urticaria and erythema. Symptomatic hepatitis and non-fatal renal failure 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 overdose, general supportive measures should be employed. Please visit www.dantrium.com for additional product information. For full prescribing information, please see reverse.

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