



# The Communicator

## World Congress of Anaesthesiologists Leads To New International Cooperation

Over 10,600 people attended the 13<sup>th</sup> World Congress of Anaesthesiologists in Paris on April 18-23, 2004. The meeting, which is held every four years, focused this time on "Progress for All," a concept the organizers say was meant to look past world events and "to gather the anesthesia community around our values to promote a humanist concept of our specialty."

Those organizers are The French Society of Anaesthesia and Intensive Care (SFAR). They worked to bring the meeting together as well as to translate the sessions, posters, and presentations into the different languages represented by attendees. They also built an extensive website to allow the anesthesia community the opportunity to plan their visit. For a complete schedule of events and presenters, you can visit the site at [www.wca2004.com](http://www.wca2004.com).

Hundreds of speakers hailing from six continents presented sessions over the course of the meeting on subjects ranging from pharmacology to sepsis. MHAUS President Dr. Henry Rosenberg spoke on the clinical manifestations of MH in a two-hour panel related to MH.

Dr. Rosenberg also moderated a panel on post-operative fever and hyperthermia and Dr. Richard Kaplan, a longtime MH Hotline Consultant, spoke at another session as well.

Other speakers from the international MH community included Helle Ording (Denmark), Albert Urwyler (Switzerland) and Phil Hopkins (UK).

"In addition, there were three other panels related to MH during the meeting, two in French and one in Spanish," Dr. Rosenberg reports. "This demon-



Jo Nichols and Dianne Daugherty greeted visitors at the MHAUS Exhibition Booth with information about MHAUS' latest programs.

strates the importance of the subject to the community. I understand that each panel was well attended."

### Posters

Several of the posters discussed various aspects of MH. The group from Marseille had an abstract demonstrating that many people who de-

*continued...*

### In This Issue:

Paris World Congress	1-3
Profile of MH Lab	4-5
FASA Meeting	5
Promotional Campaign	6
Honor Your Anesthetist	7
Hotline Activity	8-9
Heat Stroke Info	10
Membership Info	11
Happenings	12

*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 Procter & 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.

#### EDITOR

Katherine O. Riess

#### Editorial Advisory Panel

Scott Schulman, M.D.  
Henry Rosenberg, M.D.  
Barbara Bandom, M.D.  
Cynthia Wong, M.D.  
Lena Sun, M.D.  
Ronald J. Ziegler  
Ingrid Skillings

#### FOR MHAUS

Henry Rosenberg, M.D.  
*President*

Sheila Muldoon, M.D.  
*Vice President, Scientific Development*

Stanley Caroff, M.D.  
*Vice President -- Director of NMSIS*

Christina Duetsch, MS, RN, CS-P  
*Vice President -- Assoc. Dir. of NMSIS*

Ingrid Skillings  
*Vice President -- Patient Affairs*

Ronald J. Ziegler  
*Secretary*

John L. Blair  
*Treasurer*

Dianne Daugherty  
*Executive Director*

Gloria Artist  
*Hotline Coordinator*

Josephine Nichols  
*Business Manager*

Cynthia L. Solyian  
*Administrative Coordinator*

Fay Kellogg  
*Office Assistant*

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.

Copyright 2004 by MHAUS

velop exertional heat stroke are also MH-susceptible based on biopsy as well as nuclear magnetic resonance biochemical marker techniques, which they use regularly.

There were two abstracts from the University of Toronto (Julian Loke). One demonstrated the presence of a mutation in an MH family, the other was a technique presentation showing a procedure for searching for up to 95 mutations in the genome for about \$300. The group is working on cleaning up the assay. This would make it easier to screen for the many mutations associated with MH. Another paper was from Norway demonstrating that some patients with the neuromuscular disorder McArdle's syndrome may be MHS.

Joel Lunardi (Grenoble) gave an excellent talk on the molecular genetics of MH and Central Core Disease and how the French biopsy centers are working together to identify families where molecular genetic investigative principles would be applied to the clinical situation to avoid contracture testing with muscle biopsy. His comprehensive talk showed the abnormalities that occur in the ryanodine receptor protein as well as other proteins associated with excitation contraction coupling in MH patients.

### ***Meeting Others Interested in MH***

There were many people in attendance from different parts of the world who had an interest in MH.

Executive Director Dianne Daugherty and Business Manager Josephine Nichols greeted visitors at the MHAUS Exhibition Booth and promoted awareness of MHAUS and its available programs.

"In the past few years," Dianne says, "MHAUS has devoted a good portion of its resources toward enhancement of the MHAUS website. Many of those visiting the booth had previously been to the website and were appreciative of the information available there. We spoke to physicians who have called our MH Hotline and emailed questions to us for help in solving a particular problem. We obtained their feedback on the quality of the information they received – it was overwhelmingly positive."

Dianne reports others voiced concerns about the availability of the MH antidote, dantrolene, in their particular country and asked if there was anything being done to help those unable to obtain this life-saving drug. Procter & Gamble

***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, 11 East State St., PO Box 1069, Sherburne, NY 13460; or visit us on the Internet at [www.mhaus.org](http://www.mhaus.org).***

Pharmaceuticals, the makers of dantrolene, are addressing this concern, and we are hoping for a resolution in the near future.

“Visitors to the booth included the Attorney General of the South African MH Group,” Dianne says, “as well as representatives from Saudi Arabia, Iran, and Lebanon. We made some wonderful contacts.”

One evening there was a gathering of people involved with MH from Australia, US, France, UK, Canada, Germany, Spain, Switzerland, and Holland to discuss current issues related to MH. MHAUS Executive Director Dianne Daugherty attended, calling it an “eye-opening experience” where 16 representatives came and discussed what they are doing in their respective countries to fight MH. They also discussed their testing protocols and the status of their biopsy testing centers. An anesthesiologist from Spain also attended who wishes to start an MH organization and was looking for advice.

Most centers in Europe are beginning to employ molecular genetic testing in conjunction with the contracture test to diagnose MH susceptibility.

### ***Taking the next steps***

Dr. Rosenberg introduced the concept of creating an international MH organization in a virtual format. He envisions this would consist of a website as well as moderated discussion lists related to MH similar to our Hotline discussion list.

On this website would be posted, country by country, activities related to MH such as biopsy centers, biopsy center directors, patient and professional organizations related to MH, contact names and addresses for people with MH traveling in those countries, as well as research activities related to MH conducted in the countries. It could include epidemiological information such as a compilation of numbers of biopsies done by center, number of molecular genetic tests, and the panels of genetic testing that are in use. Educational benefits could include URLs of websites related to MH, availability of teaching and educational material available and in what language, phone numbers of hotlines, names of hotline consultants, interesting/unusual presentations of MH, publications related to MH by country, topics of research activity that are in process, conferences related to MH by country, city, etc. and other informa-

### **MH Internet Addresses Around The World:**

**USA** • [www.mhaus.org](http://www.mhaus.org)

**Canada** • [www.mhacanada.org](http://www.mhacanada.org)

**New Zealand** • [www.midcentral.co.nz/hospital/mal-hyper/MalH-Main.htm](http://www.midcentral.co.nz/hospital/mal-hyper/MalH-Main.htm)

**UK** • [www.bmha.co.uk](http://www.bmha.co.uk)

**Japan** • [homepage3.nifty.com/JMHA/index.eng.html#Anchor-14210](http://homepage3.nifty.com/JMHA/index.eng.html#Anchor-14210)

**Switzerland** • [www.smhv.ch](http://www.smhv.ch)

**European MH Group** • [www.emhg.org](http://www.emhg.org)

tion that might be of interest to the physician and patient communities throughout the world.

For example, did you know there are three biopsy centers in Australia and one in New Zealand? One particular mutation is over-represented in Switzerland, another in the UK. The representative of Australia reported that there was a recent death from MH in that country. MH cases happen regularly and, unfortunately, some patients are still dying from the disorder.

“The cost for this organization would be minimal, but the benefits of having access to that information would be worth the investment,” Dr. Rosenberg points out. The next step would be to create a group to define the organization and keep track of its activities.

“After a number of days of enjoying conversations with attendees, taking in a bit of the breathtaking sights Paris has to offer,” Dianne adds, “we returned to the U.S. ready to continue our work and thankful for the opportunity to bring our message abroad!”



*Jo Nichols at the Arc de Triomphe >*

## Research Is Key For Dr. Kant and His Team at UPMC

**A**s we reported in our last issue, MHAUS has selected the molecular genetic diagnostic testing laboratory at the University of Pittsburgh Medical Center to introduce a clinical diagnostic test for malignant hyperthermia.

The laboratory, under the leadership of Jeffrey A. Kant, MD, PhD, Director of the Division of Molecular Diagnostics in the Department of Pathology, currently tests for seventeen different inherited disorders, a number of which involve DNA sequencing. The lab also tests for a broad array of infectious agents and diagnostic changes in hematologic cancers.

Dr. Kant, who has been conducting research at the facility for over eight years, was recently elected a Fellow of the American Association for the Advancement of Science (AAAS) for his service to the field. He is looking forward to working with the researchers in Bethesda on the development of the clinical diagnostic test of ryanodine receptor (RYR1) muta-

tions that have been shown to cause MH. He is also looking forward to working with the leaders and members of MHAUS.

The laboratory will work closely with the Center for Clinical Genetics (CCG) at UPMC under W. Allen Hogge, MD. The CCG was established in February of 2003 as an initiative of the medical school with a purpose is to integrate elements of molecular genetic testing, genetic counseling and medical knowledge. The goal is to provide a comprehensive service to patients and physicians - to include outreach within the USA and overseas for rare disorders. Counselors work with physicians and patients to explain the significance of testing and obtain informed consent. The lab then does the testing and the results are disclosed to patients with medical advice and support on disease manifestations and treatment.

For MH, the key physician advisor will be Dr. Barbara Bandom, who is a Hotline Consultant, a pediatric anesthesiologist and the Director of the North American Malignant Hyperthermia Registry, also located at the University of Pittsburgh.

Dr. Kant points out that if patients have questions about the syndrome or susceptibility, they should direct those questions to their primary physicians, anesthesiologists, or the MHAUS office.

The biopsy center directors, the Hotline Consultants, the laboratory team, MHAUS board and staff members as well as members of the MHAUS Profes-

sional Advisory Council are meeting in June to work out many of the logistics. We will bring you a report of their plans in future issues.

While the agreement is in process and the details continue to be worked out, Dr. Kant and his team are busily planning the "intellectual" side of this effort.

In addition to the involvement of the researchers in Bethesda, John Uhrmacher, a developmental technologist at UPMC will work with Dr. Kant to help develop the test. Following development and validation against normal and abnormal samples, the test will then be transferred to the "routine" side of the clinically-licensed UPMC lab where it will become part of the lab's workflow of DNA sequencing assays.

"A primary question," Dr. Kant says, "will be determining when patients should get the test," and it will be important for people to remember that the test is still not perfect.

At this time, the genetic test is still only about 25% sensitive, meaning only 1 in 4 patients with a confirmed diagnosis will show a causative mutation in the RYR1 gene. For patients with a positive contracture test, it will be worth pursuing the genetic test to see if an RYR1 abnormality can be identified as the cause of that patient's susceptibility.

"But the contracture test will still be the gold standard."

Dr. Kant points out that the RYR1 is particularly complex by comparison to other genes.

"This is one of the biggest genes I've ever looked at, with



< Dr. Jeffrey A. Kant to lead MH genetic testing

## FASA Meeting Proves Fruitful

*Kant Team, continued...*

over 100 coding blocks. Based on findings to date, we will initially only be looking at 12 of those coding blocks for mutations associated with MH."

"Research is key," Dr. Kant says. "A particularly exciting feature of working with MHAUS on MH is the potential for future advances in genetics that allow identification of a larger fraction of patients. This can then be translated to expanded clinical testing. There are other blocks within the RYR1 gene as well as other mechanisms of mutation, and there are precedents in other diseases where other causes have emerged with further genetic study."

In Europe and Canada, this type of test has allowed a positive diagnosis of MH susceptibility to be documented for family members without a muscle biopsy and contracture test. This test can help us identify children at risk of MH. This will be the first time that a genetic clinical diagnostic test will have a direct impact on the practice of anesthesia in the United States. ■

**W**hen Dianne Daugherty, Executive Director of MHAUS, attended the Federated Ambulatory Surgery Association (FASA) meeting in Phoenix, Arizona, this past Spring, she was pleasantly surprised by the positive response she received to MHAUS programs, especially the Ambulatory Surgery Center version of the MH Procedure Manual.

"Those that have purchased the manuals are very pleased with their purchases," she reports. "They find the manual to be a helpful tool in preparing for a possible MH event, as well as an ongoing training tool for new hires and a regular review of the MH protocol."

The meeting took place May 5-7, 2004. Over 1600 ambulatory surgery professionals attended, with well over 100 of them visiting the MHAUS booth.

"We have expanded our focus in the past few years to include ambulatory surgery centers and office-based surgery

centers in order to raise awareness about the risks MH presents outside of the traditional hospital settings," Dianne adds. "Those who visited the booth were very interested in learning about our organization. Some of them already knew of us and some had even used the Hotline."

Booth visitors were given stickers printed with the Hotline phone number, as well as membership information, flyers about the MH manuals, brochures, MH Protocol pocket cards and back issues of *The Communicator*.

Their comments included positive feedback about the procedure manuals, questions about the stocking of the MH antidote dantrolene, and information about a new CO<sub>2</sub> analyzer that is placed under the tongue.

Dianne feels this meeting was a worthwhile venture as many contacts were made that will help bolster MHAUS's efforts in this area. ■

### Looking for a way to make your gift last?

Lifetime Memberships to MHAUS are now available for a one-time cost of \$500 or more. MHAUS Lifetime Members receive a special membership card, no renewal notices, an uninterrupted subscription to *The Communicator*, and special acknowledgement in the MHAUS Contributor List each year.



MHAUS recognizes the generous support of



Makers of

**Dantrium® IV**  
(dantrolene sodium for injection)

To order Dantrium® IV, call 1-800-448-4878

## Susceptible Patients, Like Dennis Apgar, Spread Word

In our last issue, we announced that several governors nationwide had declared March “MH Awareness Month” as part of a promotional campaign to raise awareness about the dangers of MH. We salute those states for their part in this effort.

Many susceptible patients and families took part in this campaign in their own way, presenting their stories to their local television news programs, and many placed Letters To The Editors in their local newspapers.

One such patient was Dennis Apgar, Jr., whose letter appeared in the April 11, 2004 edition of *The Morning Call* which serves eastern Pennsylvania. His letter is reprinted here with the *Morning Call's* kind permission:

*“As someone who speaks from experience, I would like to urge people to find out as much as they can about their family’s medical history.*

*I am affected by a little-known genetic disorder called malignant hyperthermia. When exposed to common anesthetics, my muscles can break down and my temperature can soar to life-threatening levels. This can happen during or after surgery.*

*By proclaiming this past month Malignant Hyperthermia Awareness Month, New Jersey Gov. James McGreevey helped tremendously by encouraging public awareness for something that has previously received little notice. But March is over, and now it is our responsibility as citizens to follow the example set by our leaders.*

*If you have ever had a family member who has had an unusual reaction to anesthesia, please visit [www.mhaus.org](http://www.mhaus.org) to find out whether*

*malignant hyperthermia might affect you. It could save the life of someone in your family.*

*Dennis Apgar, Jr.,  
Phillipsburg, N.J.”*

### The Apgar Family’s Experience

Dennis Apgar is a 21-year-old man, recently married and the father of a 14-month-old baby girl.

Debbie Jasso, Dennis’ mom, remembers her son was just eight when he went into the hospital for surgery to remove his adenoids and have tubes inserted in his ears.

She says that despite the fact that it was a tiny hospital and a time when MH was still not widely known, the anesthesiologist recognized the signs immediately and began treating him for MH.

“Mid-surgery, his muscles became rigid and his temperature began to rise,” she recalls. “If it wasn’t for the anesthesiologist, Dr. Valsala Narayan, and her actions, I believe he would have died.”

Debbie says the anesthesiologist continued to monitor Dennis, kept him on ice for a day afterwards, and then counseled the family on what MH is and what its genetic nature could mean to Dennis’ relatives. She put Debbie in touch with MHAUS, and Dr. Rosenberg called her personally to answer her questions.

“He was very helpful to us when it happened. He gave us information about MH and how to identify him as susceptible. He also helped us to make the choice



*Dennis Apgar and his wife, Cassie*

about whether or not to have a biopsy, although we chose not to do it at that time because we felt the episode had been traumatic enough.”

She sent letters out to both sides of the family as well as to all the hospitals in her area. Dennis wears a medical ID necklace, but she still worries constantly about him.

“Every time he leaves the house in a car, I worry. Thank God he hasn’t needed any surgery since then because I live in fear.”

Dennis also experiences many unexplained muscle aches.

The other thing that concerns Debbie is the fact that they don’t know which side of the family Dennis from which he inherited his susceptibility. Debbie says her side does take it seriously though. They carry cards and mention it to doctors every time anyone needs a procedure. Still, she says, they are all looking forward to the day a reliable blood test becomes available so that they can be sure.

For more information about this project, contact Al Rothstein toll free at (866) 636-3342 or [mhaus@rothsteinmedia.com](mailto:mhaus@rothsteinmedia.com).

# Honoring Your Anesthesia Care Professional: A New Program For MHAUS Members



Have you recently gone through surgery and were extremely pleased that your anesthesia professional was one of those individuals who made you feel that they were there for you? Their overall attitude and specific attention to your case made you feel much more comfortable about the level of overall concern for your health and welfare?

MHAUS has developed a way you can recognize that particular anesthesia professional and to express your appreciation 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 us at the MHAUS Administrative Offices to express your appreciation of their outstanding care!

## MHAUS To Offer Two Writing Awards

The Malignant Hyperthermia Association of the United States (MHAUS) is pleased to announce the availability of awards in the amount of \$2000 and \$1500 to the first place and second place authors, respectively, of manuscripts related to malignant hyperthermia (MH).

In order to promote awareness of MH and its various manifestations and to encourage continued study of the syndrome, Mr. George Massik, a founding member of MHAUS, has graciously offered to support two writers' awards. **The Daniel Massik Fund** at The Foundation for Jewish Philanthropies in Buffalo, NY was established by Mr. Massik in memory of his son who died from MH. These Awards will provide a stipend of \$2,000 for First Place and \$1,500 for Second Place to an anesthesia resident/fellow or an anesthesiologist who is within five years of ending his/her training to attend the annual meeting of the American Society of Anesthesiologists Meeting or another meeting of similar merit.

The Awards will be given to the primary author of the best manuscript concerning malignant hyperthermia. The format may be a case report, literature review or original study. The document should address a significant issue related to the problem of malignant hyperthermia. Those participating must currently be a resident/fellow in anesthesiology or an anesthesiologist who is within five years of ending his/her training. The paper must be a minimum of three double-spaced typed pages and a maximum of 10 pages. Author's CV should be included.

**Deadline** for receipt of the manuscript in the MHAUS office is **August 2, 2004**. The award will be presented at the annual MHAUS Recognition Reception at the annual meeting of the American Society of Anesthesiologists Meeting in Las Vegas, NV in October 2004. Winners will be notified by August 20, 2004 to allow for coordination of travel plans.

For further information regarding the application process for this Award, please contact MHAUS, attention Gloria Artist, either via regular mail at 11 East State Street, P.O. Box 1069, Sherburne, NY 13460, fax at 607-674-7910 or email gloria@mhaus.org.

### 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 03-04 in memory of Lila and Jerry Lewis. We are most grateful for their support.*

#### Life Benefactors

Marilyn Lewis Glassman  
David & Dorothy Glassman  
Gregory Lewis Glassman  
Jacey Lila Glassman  
Steve & Mickey Lewis  
Dr. Joseph Sugerman  
Bob & Dianne Winters

#### Sponsors

Gloria Leonard  
Lenny Roberts  
Mr & Mrs. Karl L. Sussman  
Erven & Beatrice Tallman

#### Donors

Mr. & Mrs. Larry Bloomingfeld  
Cindy & Jerry Kay  
Sy & Estelle Opper  
Mr. & Mrs. Steve Pollack  
Bill & Linda Rouse  
Stuart & Roberta Schoffman

#### Friends

Mitzi Birnbaum  
Mr. & Mrs. Doug Braun  
Larry & Shelly Kozak  
Dr. & Mrs. Jon Matthews  
Lois Soter  
George & Miriam Trustman  
Mr. & Mrs. Ron Tucker  
Jack & Toni White  
Ken & Debbie Winters

# MH Hotline Activity For October-December 2003



During the months of October, November and December 2003, Drs. Adranga, Bandom, Chapin, Gronert, Litman, Millman, Miller, Rosenbaum, Rosenberg, Weglinski, Wedel and myself provided hotline consultations which included 49 consults and 25 non-urgent questions forwarded from the MHAUS office or Poison Control Center.

Consultations were requested for preoperative assessment and planning of MH-susceptible patients, differential diagnosis of patients with suspicious symptoms or in the active management of a presumed MH episode, and follow up management of an MH episode.

## **Common Hotline Questions**

Common questions included reviewing drugs which could be administered safely. To our current knowledge, only inhaled anesthetic vapors and succinylcholine are triggering agents. Ketamine, local anesthetics, midazolam, nitrous oxide and opioids are all considered safe agents.

**For A Limited Time Only! Until December 31, 2004, new and renewing MHAUS members at the \$35 level or higher can choose either a copy of the MH Grand Rounds or Knowing Your Role (for in-servicing) video, each a \$25 value, for FREE. Contact the MHAUS office for more information!**

Another common request for consultation included a number of patients with fever or other symptoms (high CO<sub>2</sub>) while receiving general anesthesia in obese patients (greater than 110 kilograms or 245 lbs). Although none of the consultants believed any of these cases were MH episodes, it is clear that we are doing a better job overall of keeping patients warm in the operating room, but these patients may present a diagnostic challenge.

Some consultations were requested by an anesthesiologist or nurse anesthetist who wanted reassurance that suspicious symptoms were not consistent with an MH episode.

An anesthesiologist may see a patient with a specific rare syndrome or diagnosis only once in their career. When patients with a rare muscle syndrome present for elective surgery, anesthesiologists are increasingly concerned regarding the potential susceptibility to MH.

## **Poland Syndrome**

Consultations this quarter included the potential susceptibility of patients with Poland syndrome, myoadenelate deficiency, inclusion body myositis, or preoperative elevation of creatine phosphokinase (CPK or CK). Although none of the four specific diagnoses listed above were considered to be associated with MH susceptibility (MHS), there was a divergence of opinion as to how to manage the patient with elevated CK (without identifiable reason) for

administration of general anesthesia. Drs. Weglinski, Wedel, and Engel published their experience with patients who had chronic elevations of CK and found a high percentage of them (49%, or 24 of 49 patients) were diagnosed as MH susceptible by the biopsy test (*Anesth Analg* 1997;84:1038-1041). Consultants sometimes recommended a nontriggering anesthetic technique, but others did not. We do not have consensus on this point. Hotline consultants deal with difficult issues and provide expertise regarding these concerns.

## **Masseter Muscle Rigidity**

Some consults were prompted by unanticipated masseter muscle rigidity, requesting advice for acute management, and referral for biopsy. Historically, these patients are considered to have a nearly 50% risk of being MHS, so these patients are routinely referred for biopsy evaluation.

## **Safe Delivery For MHS**

Interesting and difficult consults included management of laboring patients who now required general anesthesia for urgent Caesarean section. In three cases the mother was known or considered MHS, and in one consult the father of the baby was known to be MHS. The risks and benefits to the patient and the fetus were discussed regarding rapid sequence airway management, intravenous anesthetics for maintenance of anesthesia and the need for resuscitation personnel

*continued...*

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

for the infant since he/she may have neurologic or cardio-respiratory depression at birth.

### ***Post-Operative Fever***

Other calls included patients with presentation of fever in the operating room, recovery room or the ICU following surgery where sepsis or focal infection were considered an important concern.

Often there is sufficient data to exclude MH, but sometimes the data and presentation are difficult to rule out MH. In these circumstances, consultants provided advice (pros and cons) regarding use of dantrolene (if the caller has not already done so). However, consultants routinely educate the callers that referral for a biopsy evaluation is important and that a response (decrease in fever) to dantrolene is not specific for a diagnosis of MH.

### ***Suspicion of MH Episodes***

MH can strike unexpectedly and with no known history. More calls are coming from outpatient surgery centers with possible MH episodes or concerns about appropriateness of caring for a patient with known MHS. All suspicious MH episodes in these settings were appropriately dealt with, and every center that called did have dantrolene available and a back-up system to transfer to a

major hospital for continued care. We hope that all surgicenters will take a few moments to update their staff regarding a possible MH episode which becomes a medical emergency, and have a sufficient stock of dantrolene on site.

A few consults were requested because of postoperative muscle pain and elevated CK. This occurred in a patient who had a nontriggering anesthetic and the question of 'stress-induced' MH episodes was discussed. Although not considered likely by the consultants, this question is occurring with greater frequency.

Also, a few consults were obtained regarding the issues of positioning or propofol-induced rhabdomyolysis.

Although each caller may only call once in a career, the hotline provides an important service to the anesthesia care professional community and MHS patients. Callers commonly express sincere gratitude for the existence of this service, particularly when they are involved in a rare, but treatable, crisis situation. It is our pleasure to continue to be active members of the MH Hotline. ■

## ***Meet This Issue's Hotline Consultant***

The Hotline activity was summarized this issue by Dr. Joe Tobin, who is a pediatric anesthesiologist and intensivist at the Wake Forest University School of Medicine in Winston-Salem, North Carolina. He has been a Hotline Consultant for three years and assumed the role of Director for the biopsy testing center upon Tom Nelson's retirement.

"Tom has been a great friend and role model for over a decade," Dr. Tobin says of his mentor. "It was an honor to accept his invitation to keep the biopsy center going. Knowledge regarding MH is growing, as evidenced by the number of referrals and calls to our center. Hotline consulting is a critical part of the service we provide."

Dr. Tobin finds both his hotline and biopsy center work rewarding in many ways.

"It's an exciting time as we hope to develop genetic testing to reduce the number of patients who need to travel to have biopsies performed."



## Be An Advocate For Kids: Empower Your Coach!

By Ingrid Skillings, MHAUS Vice President For Patient Relations

As summer approaches thoughts turn to lakeside cabins and warm days. Ow! Unfortunately reality can hit hard. With the warm weather, the threat of heat-related illnesses increases. This summer we challenge you to become an advocate for the children in your neighborhood. Heat-related illnesses can strike anywhere and anytime.

Two young girls, one who happens to be MHS, recently helped prevent a tragedy. On a warm, but somewhat humid, spring day at a fifth grade track and field event, the athlete began to experience cramping and nausea after a 50m hurdle race, the soccer punt and helping her friend train for her relay race.

During a break before her own relay race, she mentioned to her friend that she wasn't feeling well. Together they decided that a water break and rest should do the trick.

As the announcement for the relay race rang out and she began to get up, a second wave of nausea hit her. Her friend, acutely

aware of her advocacy for MH and related heat illnesses, suggested she seek medical attention.

A nurse at the event could tell these students knew what they were talking about! Needless to say, a tragedy was avoided.

Another story that shows the impact one person can have on others is the case of a twelve-year-old athlete and the impact it would have on his lacrosse team. In this case it was the parent who caused a ripple.

As with most athletes, this boy was known to push himself beyond his limits. This, along with a tremendous stubborn streak and his natural teenage need to be "superman," was reason enough for his parent to alert the coach with the words "and if he complains of cramps and nausea while his face is bright red, just call 911." With that in mind, the coach decided on the following day to actively inform and sent an e-mail to every parent on the dangers of heatstroke and dehydration.

Countless stories like these prompted research into what coaches across the country are hearing this year as the warm weather sports season begins.

The National Athletic Trainers' Association (NATA) set forth a consensus statement on Exertional Heat-Related Illnesses which outlines the Factors that Influence Heat

Illness, General Considerations for Risk Reduction, Recognition, Treatment and Return-to-Play Considerations for Dehydration, Exertional Heat Stroke, Heat Exhaustion, Heat cramps and Exertional Hyponatremia.

There are two versions, one for healthcare professionals treating athletes on the field, and one for parents and coaches. For a complete list of the information, visit [www.nata.org](http://www.nata.org).

And the link between exertional heatstroke and MH, you ask? To date, there have been many studies but no formal link has been found. A recent study done in Marseille, France was presented at the World Congress of Anesthesiologists. Those researchers found that "up to 80% of 376 subjects displayed energetic metabolism, morphological, histoenzymological or IVCT anomalies. However, no defined myopathy was found. Although aetiopathogenesis is different between exertional heat stroke susceptibility (EHS) and MH, a failure of calcium homeostasis with regard to the triadic junction might be hypothesized. Considering IVCT anomalies, investigations should be performed to search for MH susceptibility in all EHS subjects. Without investigation, they should be considered at risk for any anesthetic procedure using halogen gas associated or not with succinylcholine."

MHAUS continues to keep watch for information that many link exertional heatstroke to MH. We published an article in the Summer 2000 *Communicator* detailing studies on the subject and will bring you the latest data as it becomes available.

MHAUS recognizes the generous support of



**AstraZeneca**  
A Business Unit of Zeneca Inc.  
1800 Concord Pike PO Box 15437  
Wilmington DE 19850-5437

**DIPRIVAN**  
INJECTABLE  
EMULSION  
propofol

## Slide Show Presentation For MH Risk Available

MHAUS offers a slide show kit (CD-ROM or 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.

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. Two CMEs are available.

Cost is \$125 plus \$5 for shipping and handling for either the slides or the CD. For both formats, the cost is \$135 plus \$5 for shipping and handling. Call 607-674-7901 or visit [www.mhaus.org](http://www.mhaus.org) to order.

## Every MH-Susceptible Should Wear a Medical ID Tag

MHAUS now has help available for the MH-susceptibles who have no insurance, or cannot afford to purchase a medical ID Tag.

The **Sandi Ida Glickstein Fund** was recently established for the purpose of providing free ID Tags for MH-susceptible patients who qualify.

To take advantage of this program, please send us a letter indicating why you would like MHAUS to provide you with a complimentary ID Tag.

The goal of the free ID Tag program is to ensure the safety of all MH-susceptibles during an emergency situation and to prevent a tragic outcome from MH.

For further information, please contact MHAUS at 11 East State Street, P.O. Box 1069, Sherburne, N.Y. 13460-1069; call 607-674-7901, or visit [www.mhaus.org](http://www.mhaus.org).

***Have you visited us lately? Log on to  
[www.mhaus.org](http://www.mhaus.org) and get the latest information on  
MH, post a note to the message board, order a new  
program, or just let us know what you think.***

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

A contribution of:  \$35 (Basic)  \$50  \$100  \$250  \$500  \$1000 (President's Ambassador) or  (other amount) \$ \_\_\_\_\_, 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: \_\_\_\_\_

Expiration Date: \_\_\_\_\_

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

# MHAUS Happenings, Events, and Notices

## Carry the card that cares

Thank you to our many members who are already using the MHAUS affinity credit card offered by MBNA, the only bank whose core business is affinity marketing. This support program, initiated in 1998, provides one more way for our members to help benefit MHAUS while receiving benefits themselves. MHAUS receives royalties from retail transactions.

Through a mutual agreement, MHAUS sends MBNA a random list of names and addresses (no other information is released) for the sole purpose of mailing an invitational letter offering the program. MHAUS does not sell addresses to MBNA nor are any names/addresses released when a member requests us not to do so, and all letters are approved by MHAUS.

Our goal is to provide a credit card program superior to existing premium cards that benefits you. With every purchase, your benefits include the earning of rewards that will give you the flexibility of selecting travel rewards or merchandise, all in one program. The rewards earn miles toward travel worldwide with no blackout dates. There is no annual fee. The credit line is available up to \$100,000 with an introductory 0% APR for one year. Additional benefits are outlined in the letter.

If you have not already received an offer through a special mailing from MBNA, watch for the next invitation to arrive. Or, we would be happy to assist you from the MHAUS office. We invite you to consider participating in this program which benefits all concerned.

❑ **A New MH Summary** written by Dr. Henry Rosenberg and Dr. Nyamkishig Sambuughin is now available at [www.genetest.org](http://www.genetest.org). This summary includes characteristics of the syndrome, diagnosis and testing information, indications for susceptibility and molecular genetics information. You can read this summary by selecting the genereviews choice at the top of the homepage, then typing "malignant hyperthermia" into the search prompt or by visiting the links section of the MHAUS website at [www.mhaus.org](http://www.mhaus.org) and clicking "Gene Tests."

❑ **Traveling With MH This Summer?** Call the MHAUS office at 607-674-7901 for the latest updates on MH experts in countries around the world. Because the information changes so quickly, we now have a print-on-demand sheet to bring you the most recent contact information we know of for experts in the countries you will be visiting.

❑ **THANKS!** MHAUS is grateful for the financial support of the following State Societies of Anesthesiology: **Arkansas, California, Florida, Idaho, Indiana, Maine, Michigan, New Hampshire, Ohio and Pennsylvania.** Our grateful appreciation is also expressed to the following state components of the American Society of PeriAnesthesia Nurses: **Arkansas, California, Hawaii, Missouri-Kansas, Nevada, Tennessee, Texas and Wisconsin.**

Call the MHAUS office today to find out how your group can help.

❑ **Errata.** In our last issue, in the article regarding the North American MH Registry on page 5, there was a typographical error. The item should have read "the American Society of Anesthesiologists" instead of "Association." We apologize for any confusion.

MHAUS  
11 East State Street  
P.O. Box 1069  
Sherburne, NY 13460-1069  
[www.mhaus.org](http://www.mhaus.org)

NONPROFIT  
US POSTAGE PAID  
PERMIT #10  
SHERBURNE, NY 13460

