



Malignant Hyperthermia Association of the United States  
and the North American Malignant Hyperthermia Registry

Volume 17, Number 4  
Fall 1999

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# The Communicator

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*What's Next for MHAUS?  
The Prepared Patient  
How the Whitehills Tackle MH  
plus, Hotline Activity*

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 malignant hyperthermia.

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Malignant Hyperthermia (MH) is an inherited muscle disorder which, when triggered by potent inhalation anesthetics and some other drugs, 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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# President Henry Rosenberg, MD On What's Next for MHAUS



As I conclude my first year as President of MHAUS/North American MH Registry, I am pleased to report on the activities and future plans for the organization, but first I would like to answer the question, how does MHAUS work?

#### *"How Does MHAUS Work?"*

MHAUS is governed by a dedicated board of directors, all serving voluntarily with no compensation. The board meets quarterly and once per year for a strategic planning retreat (held in June of this year) to set priorities, goals and operational guidelines and discuss needs of the organization, of patients and anesthesia providers in relation to MH.

The everyday operations of MHAUS are conducted from our home office in Sherburne, New York. The staff is led by Janice Bays, our executive director. Our staff is dedicated to carrying out the missions and goals of MHAUS and take pride in their work to help patients and providers cope with the often confusing issues of this disorder. Many of the staff have been with MHAUS for many years. You may meet them, as well as some of the board members, at the MHAUS exhibit booth at one or more professional meetings.

#### *Activities and Programs*

The activities of MHAUS are divided into maintenance programs that provide regular information and guidance to health care professionals and patients and to special programs and projects. The "routine" programs consist of this newsletter, *The*

*Communicator*, published since 1982, fulfilling patient inquiries, producing and mailing informational brochures and updates, maintaining our fax-on-demand program, our life-saving hotline and recording MH events and patient specific information in the North American MH Registry. We also regularly scan publications and the internet for stories related to MH. We exhibit at major anesthesia conventions where we literally give away information and advice to all comers.

All of these programs fulfill the mission of providing education and information to anesthesia providers in order to prevent death and disability from MH and related syndromes.

Each and every one of these activities is noteworthy, but in the interests of space, I will pass over to more recent developments.

The first program I would like to mention is our Web site: [www.mhaus.org](http://www.mhaus.org). Many of the brochures, treatment protocols and other information are posted on this site which is reviewed and updated regularly. Most recently, I have posted an MH slide presentation. Thousands have visited our site and complimented the material we have made available.

The second program I would like to mention is our new MH identification tags. These tags are reasonably priced and indicate MH susceptibility (as well as other medical conditions as appropriate.) Inscribed on the tag is the phone number of our nurse triage service and hotline in order for the caller to get the most recent information concerning MH. This ID program has taken several years to develop and is now ready for operation.

A third program, one that has also taken much time and effort to develop is the Kristin Duell memorial program. Kristin

***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.***

was an outstanding young woman who succumbed to MH a few years ago. This program is aimed at raising funds to support research in the development of a molecular genetic (DNA) diagnostic test for MH -- a very complex and formidable task. The first element of the program is a new comprehensive brochure for patients regarding anesthesia and what every patient requiring surgery or a special invasive test needs to know about anesthesia and the risk of MH. The brochure is titled "The Prepared Patient." Other elements of the program, including media attention for MH will follow shortly.

An extremely exciting program that we have recently completed is a procedure manual for management of MH. This comprehensive detailed protocol allows hospital personnel to train for and be prepared to manage an MH event. It was developed initially by Dr. Darlene Mashman of Emory University and consists of a videotape, a protocol and a quiz. AstraZeneca has generously underwritten the costs of this project. We will be developing a similar program for ambulatory surgery centers.

#### *Commitment to Mission Goals*

In order to further the goal of development of a molecular genetic test for MH susceptibility that will be less invasive and more accurate than the biopsy test, we will be awarding a competitive research grant for \$25,000 per year for a study that addresses the development of such a test in the population of the United States.

New programs are constantly under development to meet the ever-changing demands and challenges of this killer disorder.

For example, we are greatly concerned about the rapid growth in office-based surgical procedures. There have already been several MH-related deaths during surgery conducted in offices. We plan to modify our programs to address how best to prevent and treat MH in such locations.

With increasing frequency, patients are being denied insurance coverage for the benefits of a diagnostic biopsy for MH by insurance companies who know little about MH. We plan to provide support for biopsy centers and educational programs for insurance companies and HMOs concerning the problem of MH and that given proper guidance the MH-susceptible is

at no higher risk for anesthesia and surgery than any other member of the population at large.

The North American Registry represents an ongoing source of MH information concerning the manifestations of MH. Because of changes at its home site, Geisinger-Penn State Hershey Medical Center, we are reevaluating the operations of the Registry and updating the computer support (including Y2K compliance ) of this important database.

Being a leader in the battle against MH in the US has led us to provide guidance and mentorship to MH organizations in other countries, particularly those in Latin America. We plan to assist those organizations in educating anesthesia providers and also making dantrolene available where currently, because of licensure restrictions, dantrolene is not available.

We are also working with such accrediting organizations as the Joint Commission for Accreditation of Health Care Organizations, the Accreditation Association for Ambulatory Healthcare and the American Association for Accreditation of Ambulatory Surgery Facilities, to require that dantrolene and preparedness for MH are available wherever general anesthesia is administered.

These life-saving programs would not be possible without the dedication of the MHAUS hotline consultants, members of its Professional Advisory Council, the MHAUS staff, and its board members. The support and encouragement of our members and corporate sponsors, particularly Procter and Gamble Pharmaceuticals, are critically important in enabling us to fulfill our goals of no more deaths or disabilities from MH and MH-like syndromes. I thank them for their generosity. I look forward to alerting you to more exciting and life-saving programs.

If you wish to communicate with me or any member of the board, hotline or PAC, please e-mail us at: [mhaus@norwich.net](mailto:mhaus@norwich.net).

**New: An MH Slide Show on the Internet at [www.mhaus.org](http://www.mhaus.org), MH Training Manuals and The Prepared Patient Brochure**

***For more information or for materials on malignant hyperthermia or MHAUS' programs, call 1-800-98-MHAUS; write MHAUS, 39 East State Street, Box 1069, Sherburne, NY 13460; or visit us on the internet at [www.mhaus.org](http://www.mhaus.org).***

# Prepare and Persist: What Your Family Can Do About MH

As if waiting for a loved one in surgery weren't scary enough, when the news comes that something "rare" has happened, something that most doctors will see a tiny handful of times, if at all, in their careers, anyone would feel helpless.

"It's a very emotional time," says MHAUS Regional Representative Mary Ann Whitehill. "The first thing you want to do for someone when

## New Brochure Outlines Questions Patients Should Ask

A new brochure from MHAUS, entitled "The Prepared Patient," details questions all patients and their families should ask prior to having outpatient or inpatient surgery in hospitals, physician's offices or surgical sites.

Sponsored by the newly formed Kristin Duell Memorial Fund, the brochure will be administered by MHAUS because MHAUS feels that better-informed patients can help ensure safer surgeries.

Also included are reasons why it is so critical to be as complete as possible in the information you give to your caregivers.

Last minute schedule changes, for example, can sometimes mean different personnel than you thought. For this reason, you should be sure to wear your MH ID tag at all times to ensure that operating and recovery room anesthesia providers will be able to correctly identify and treat any complications.

To request your copy of this new brochure, please call MHAUS at 1-800-98-MHAUS.



*From the cover: The Whitehill family from left to right: Earle, David, Lois Clark, William, Anna Elizabeth Searight-Whitehill (their mother,) Joseph, and Ruth Hrizo.*

they find out is reassure them that information is available, that support is there."

But there are many things patients and families can do when they find out about MH. Mary Ann says the first advice she gives is to educate yourself with every piece of information that can be found.

"Call MHAUS, become a member. Get copies of every pamphlet you can get. The amount of information on MH now available at our fingertips is tremendous. If we as families can make sure the doctors know and have that information, there should be no MH crises."

The next step is to pass the information on to your families.

"My sister-in-law (RN Ruth Hrizo) and I have become the centers for MH information in our family," Mary Ann says, recalling one instance where a family member called, worried over her husband's pending surgery.

"I urged her to sit down with the

anesthesiologist prior to the surgery and ask all her questions, voice all her fears."

Make sure they all have MH ID tags, too, she says.

"The tags provide a good safety net to the families," Mary Ann points out, "because knowing they are wearing them gives some piece of mind."

Mary Ann became a Regional Representative for MHAUS because she felt she needed more official "clout" to get the attention and responsiveness of doctors and medical professionals who might someday be treating one of her children or her husband David, from whose side the susceptibility is believed to originate.

"I try to touch as many of the surgery centers and recovery rooms as I can to share the MH information available... I've even gotten called back to quite a few of them this past year when they bring on new nurses and staff."

*continued next page...*

# Hotline Review Committee Reports its Findings at the ASA in October

The MH Hotline Review Committee has completed another year of activity. The Committee Chair, Dr. Barbara Brandom, will present the results of their findings in October at the ASA as “Diagnostic consensus in MHAUS Hotline consultations.”

Formed to review the consultation reports provided by the volunteer Hotline anesthesiologists, the goals of the review are to identify controversial clinical problems and to increase consistency in the advice given by the consultants. Although the Hotline has been in operation for many years, and consultants often communicate with each other regarding questionable cases, this is the first time there has been a quality control process used for Hotline activities. This process will be ongoing.

The Review Committee consists of eight

experienced Hotline Consultants. Every three months, a random selection of 10 reports from the Hotline calls for help with patient management are sent to each committee member. The reports are blinded so that the reviewer does not know who the consultant is. They answer these six questions for each:

1) Based on the information given in the report, do you feel the advice given was correct? 2) Could the consultant have given additional advice? 3) Do you feel the diagnosis was correct? 4) Was enough data presented to support the diagnosis? 5) Did you have trouble reading the form? 6) If it was determined that MHS was possible, was referral given to a biopsy center?

Reviewers often add comments to explain opinions. These results will also be available in full at [www.mhaus.org](http://www.mhaus.org).

*continued from page 4...*

She and her sister-in-law Ruth also organized a mini-conference with MHAUS in the Pittsburgh area. Together, they worked to find a location, develop an invitation list, and bring in speakers.

Because of the size of their family and the 6 or 7 documented MH events, the Whitehills recently agreed to participate in a genetic study with the Registry.

“Persistence is the most important thing,” Mary Ann says of her commitment to educating everyone she meets about MH.

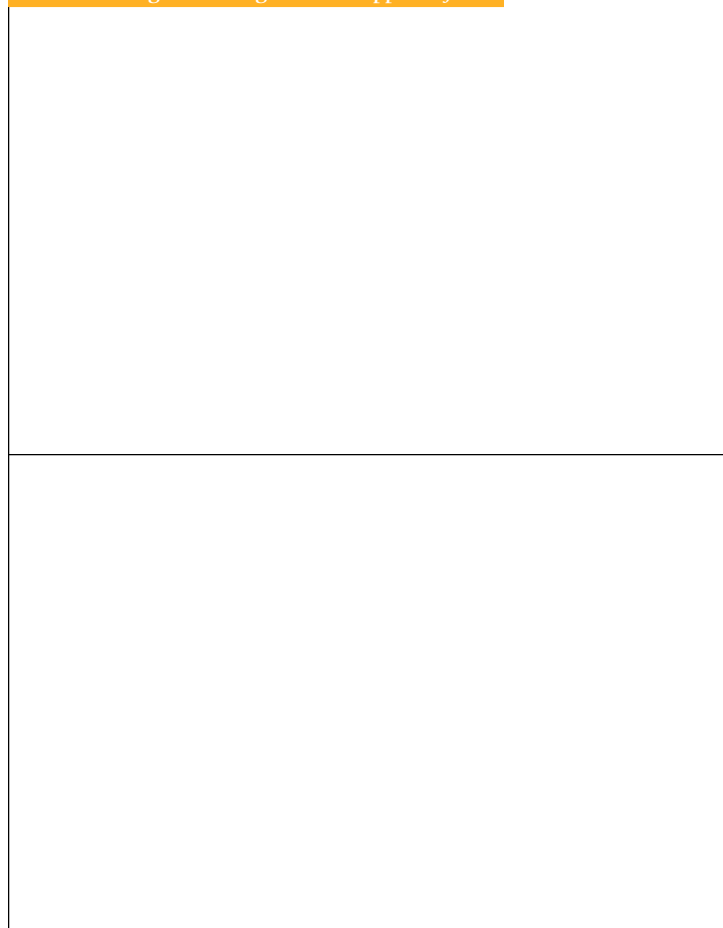
“Ruth and I got involved because of our children. We wanted everyone to be educated so that our children wouldn’t die in an operating room because of someone’s ignorance.”

Mary Ann suggests that MHS families build support networks of their own, starting with telling their stories in venues like *The Communicator*.

“Everyone’s experience with MH is different,” she says, “but at the same time, elements of your story may help others out there hearing for the first time the scary words ‘malignant hyperthermia.’ ”

**For more information on what you and your family can do about MH, call 1-800-98-MHAUS.**

MHAUS recognizes the generous support of



# Hotline Activity for January -March



During these three months, consultants received 68 calls to the MH hotline: 43 case consultations and 25 questions.

Two patients died and one other suffered cardiac arrest, though only one death had a possible association with MH. (See below for more about that death.)

Most calls came from the United States, representing 31 of the states. Two calls came from the province of Ontario, Canada. Of those identifying the patient's age, 69% concerned teens and adults, and 31% concerned young children.

Sixty-seven percent of the callers were anesthesiologists, 10% CRNAs, and 5% were internists and pediatricians. Two calls were from laypersons. The remaining calls were from nurses, surgeons, dentists, ER physicians, OR technicians, and general practitioners.

Ten of the case consultations concerned events that the consultants felt had a significant possibility of being MH. In several of the cases, increases in exhaled CO<sub>2</sub> were rapidly treated and the patients had no further problems. This made diagnosis less certain.

Today's better training, monitoring, and knowledge regarding MH has led to a significant increase in early recognition and definitive treatment with dantrolene while stopping the triggering agents. When a physician picks up the warning signs very early, treatment may start at such an early stage that definitive evidence of MH, such as severe muscle damage or blood gas abnormalities, may be lacking.

This is good for our patients, but then presents us with diagnostic quandaries, making referral for contracture testing even more important.

## ***“Classic” Cases of MH***

There were several cases of “classic” fulminant MH. One bears recounting, as the patient's response shows just how well prompt therapy can work. This was a 13-year-old boy with an ankle fracture, who was receiving a desflurane anesthetic. He developed a heart rate above 160 and exhaled CO<sub>2</sub> above 90, along with sweating and rapidly rising skin temperature. These all strongly suggested abnormally high metabolism, the hallmark of MH. The anesthesiologist promptly called for help, changed over to a non-triggering anesthetic, gave dantrolene, and placed monitoring lines.

The child's first blood studies (before dantrolene), showed a pH of 6.68, a pCO<sub>2</sub> of 126, and a potassium of 7.6. All these values were severely abnormal and immediately life-threatening. Only 10 minutes later, with appropriate care, the pH had risen to 7.21 and the pCO<sub>2</sub> declined to 60. And 20 minutes after that, the heart rate, temperature, and ventilation had normalized. He developed rhabdomyolysis (muscle breakdown) but recovered well, and went home 48 hours after the operation.

One patient who died may have had MH, though that is not clear. This patient was having a relatively minor operation. One and a half hours into the procedure there was a slight increase in temperature, exhaled CO<sub>2</sub>, and

heart rate. A potassium level drawn then was 6.8 (high).

The patient then had a cardiac arrest and was resuscitated, but not given dantrolene. He had severe rhabdomyolysis and eventually died. There was no definitive evidence of MH. It appears he suffered a cardiac arrest from the sudden potassium increase that can occur with rhabdomyolysis. There was no clear reason why he would have the problem then, though he had received succinylcholine 90 minutes prior.

Succinylcholine can cause rhabdomyolysis even in the absence of MHS, but the time course is against that here. There are some other causes to consider (e.g., interaction with certain lipid-lowering drugs) but, as with many of these calls, we are left with only a fragment of the whole story.

## ***Cases of Rhabdomyolysis***

Several other patients also had rhabdomyolysis. A healthy adult man had a tonsillectomy, receiving succinylcholine as part of the anesthetic induction. He went home. Late that evening he noted that his urine turned black. On return to the hospital, his CK (muscle enzyme) was measured at 50,000! (In normal circumstances, the CK level is below 200.) He had no muscle complaints, but obviously had severe succinylcholine-induced rhabdomyolysis.

A number of children have been killed by succinylcholine-induced rhabdomyolysis. The package insert for succinylcholine carries a boxed warning: “WARNING: cardiac arrest from hyperkalemic rhabdomyolysis”

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

and advises that it not be given in children unless an emergency exists or the rapid muscle relaxant action unique to succinylcholine is required.

Yet the drug continues to be routinely given to children by certain practitioners. In one call, a child had anesthesia induced with halothane and succinylcholine. He developed both trismus (very tight jaw) and hand rigidity. He was switched to a nontriggering anesthetic and the case continued.

After the operation he developed signs of rhabdomyolysis and was transferred to a university hospital. His peak CK was greater than 110,000. Physicians were able to prevent further damage from the episode, and he went home two days later. He will be referred for testing.

Trismus alone is of concern, although perhaps 1% of children receiving succinylcholine with halothane will get it, far too many to believe MHS is the cause for many of those events. But trismus with total body rigidity is far more worrisome, and probably all such patients should get a full evaluation for MHS and other muscle diseases.

### ***Non-MH Cases of Interest***

Several non-MH related causes of hotline calls bear repeating, as they may represent recurring scenarios that create confusion with MH itself. In two cases, operations were cancelled due to children stiffening and jerking about during inhalation induction

with sevoflurane. Excitement during induction has been noted to be more frequent with sevoflurane than halothane (an older anesthetic commonly used for inhalation inductions) and at least one investigator saw seizure-like activity similar to that reported by one of our callers (Hobbhahn, J; Funk, W. [Sevoflurane in pediatric anesthesia] *Anaesthesist*, 1996 Feb, 45 Suppl 1:S22-7).

Seven of the case consultations were prompted by fevers due to infection, including three patients suffering from a perforated appendix.

Fever is not uncommon in patients after surgery, and these cases just represented unusually high fevers from a common cause,

rather than a manifestation of the rare syndrome of MH. In medicine, we commonly use this saying: "When you hear hoofbeats, expect horses, not zebras." Most of these cases were easily distinguished from MH; the fevers did not begin during the anesthetic, and other symptoms were relatively mild.

One would expect that by the time someone with MH developed a significant fever, other signs of the disorder would be quite prominent. Our consultants play an important role in these calls: reassuring the caregivers that MH is not a consideration and avoiding the erroneous labeling of entire families as MHS. □

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



The Hotline Activity was summarized by Rory Jaffe, MD, of the University of California at Davis where he is an Associate Professor of the Department of Anesthesiology and Pain Medicine, School of Medicine at UC Davis.

Dr. Jaffe lives in California with his wife and their two children. He has been a Hotline Consultant with MHAUS for a "long time," he says, 10 years.

"The Hotline is an extraordinarily useful service to physicians and patients alike," he feels, because MH is uncommon, "and though physicians may experience it only once in their careers, when it happens they won't have the time to go looking up the information they need. For this reason, the Hotline is very important." ■

# MHAUS Happenings, Events, and Notices

❑ **Congratulations** to MH Hotline Consultant Steven C. Hall, MD, Anesthesiologist-in-Chief at Children's Memorial Hospital in Chicago, and Co-Director of Northwestern University's MH Diagnostic Center. Dr. Hall was recently elected to the Board of Directors of the American Board of Anesthesiology. The ABA Board is responsible for the quality of education in anesthesia and offers "board" certification. For more information, visit [www.abanes.org](http://www.abanes.org).

❑ **Effective July 1, Dr. Gregory Allen**, Director of the MH Registry at the Penn State University Medical Center at Hershey, moved into private practice and relocated to Olympia, Washington. We at MHAUS would like to thank him for all his contributions and hard work while Registry Director. In the interim, Dr. Sheila Muldoon,

MH Registry Chair, has assumed the duties as Director until a replacement is found.

❑ **MHAUS Medical Identification Tags Available:** Do you want the peace of mind that the MH Hotline will be called FIRST if you or your MH-susceptible loved ones are in an emergency? The tags are imprinted with the MH Hotline number which allows your physician to access your complete medical conditions and be connected with an MH expert within minutes. Call 1-800-98-MHAUS for more information about the MH ID program.

❑ **From the Mailbag** Recently, MHAUS received a letter from Dr. J. Ernesto Rojas in Columbia, regarding his efforts to get dantrolene into his country. Here

is an excerpt from his letter: "Dear Dr. Rosenberg, It is a real pleasure to tell you that I just received a donation of 10 doses of dantrolene from Procter & Gamble. As you know, it is a long story about dantrolene in Colombia, but throughout a "research protocol," I got a provisional registry in my country and in that way those 10 doses arrived here.

(Also,) ... a Spanish translation of the article by Dr. Larach about the Grading Scale published in *Anesthesiology* 80:771-779 1994 will appear in the Colombian Malignant Hyperthermia Committee Web page: <http://www.geocities.com/~hipertermia>, and in the Colombian Anesthesiology Society Journal.

Thank you for your help."  
*J. Ernesto Rojas, MD*

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