Fear and uncertainty can go hand-in-hand. This is especially true when it comes to confronting hereditary medical conditions such as heart disease or cancer or malignant hyperthermia (MH). But if you remove uncertainty from the equation, fear can be managed, or even overcome.

You’re not alone in confronting your fears when it comes to a genetic test for MH. There are qualified and experienced genetic counselors available to help you come to terms with genetic testing, both before and after the results.

“The most common fears and concerns that patients have are whether their children are at risk for MH. They also want to know if they need to take precautions with surgery,” says Deanna Steele, a genetic counselor with Magee’s Center for Medical Genetics at the University of Pittsburgh Medical Center.

“I try to advise the patient that results may help to answer their questions about their risk for MH. Based on the results, we can help them to take proper precautions, and that in educated hands, an MH-susceptible patient can have successful surgery.”

Ms. Steele has been a genetic counselor for 22 years and has counseled quite a few patients that are interested in MH testing. She’s had a number of face-to-face meetings with patients, but most occur over the phone.

“Patients are calling from all over the U.S.A. for testing and counseling,” she says.

Jenny Geurts has been a genetic counselor for 15 years and currently counsels patients at St. Luke’s Medical Center in Milwaukee, Wisconsin. She generally meets with patients for one to two sessions, with the first session taking place prior to the test and the second following the test results.

“Some may choose not to have the test after the first session,” she says.

Ms. Geurts emphasizes that molecular genetic testing for MH susceptibility does not replace the muscle biopsy test, which is still the “gold-standard” for diagnosis of MH. A genetic test for MH is not especially sensitive, meaning approximately only 30% of those that are MH-susceptible will yield a positive test result.

However, although a genetic test for MH is not very sensitive, it is very specific, meaning a positive report listing a documented causative RYR1 mutation virtually confirms the patient of being at risk for MH. As well, if a mutation is found in a family member, the other family members with that same mutation are considered MH-susceptible.

For some patients, painful emotions can resurface when considering a genetic test for MH.

“Maybe they lost a relative years ago,” says Ms. Geurts, “and they wonder, ‘Could this happen to me?’”

Ms. Geurts can relate to this concern. Her uncle suffered an MH episode. Fortunately, he survived.

“I explain that every family has something, whether it’s cancer, heart disease or MH. I try to stress prevention, because we don’t want history repeating itself. I also talk about the ‘silver...”

continued on page 3
Lack of MH Preparedness (and Dantrolene) a Concern in Developing Countries

The North American Malignant Hyperthermia Registry (NAMHR) recently received a case from the Middle East that highlights the lack of MH preparedness in the developing world.

An adult female was undergoing plastic surgery on her nose when she developed sinus tachycardia and excessive bleeding, with muscular rigidity, masseter spasm, heart rhythm disturbance, elevated temperature and cardiac arrest, known signs of MH. The medical doctor terminated the procedure, but dantrolene was not immediately available. It was necessary to obtain dantrolene from a nearby hospital, but that source only provided three vials. Fortunately, the patient survived.

"That MD deserves a lot of credit. First, for rescuing the patient in these adverse circumstances and secondly for reporting the case to the MH Registry," said Dr. Barbara Brandom.

Dr. Henry Rosenberg, MHAUS President, feels MHAUS should be more active in providing education and information about MH to other countries. But he recognizes the funding constraints, as well as the fact that MHAUS is a United States-based association, whose first objective is promoting MH awareness and preparedness in this country.

"I realize that Europe, Canada, Australia, New Zealand and perhaps Japan are pretty up-to-date concerning MH, but obviously many other countries are not," he said. "Some countries do not have dantrolene at all, others only in limited supply."

MHAUS has translated some of its educational material into Spanish and provided this information to organizations like "Operation Smile," a non-profit organization that provides free reconstructive surgery in developing countries for children with facial deformities such as cleft lips, cleft palates, burns and tumors. For its part, P&G Pharmaceuticals, which first introduced dantrolene into the market 28 years ago, has donated Dantrium® IV to Operation Smile so that doctors venturing into these countries are prepared should an MH crisis arise.

"Last year, P&G donated close to 5,000 bottles of Dantrium® IV to our organization," said Dena Liston, manager, Development and Gifts-in-Kind. "They have supported Operation Smile with in-kind donations since 1993."

Operation Smile (www.operationsmile.org) was founded by Dr. William P. Magee, a plastic surgeon, and his wife, Kathleen, a nurse and clinical social worker. In 1982, the Magees traveled to the Philippines with a group of medical volunteers to repair children's cleft lips and cleft palates.

Today, Operation Smile coordinates more than 30 medical mission sites in 26 countries annually. Last year, medical volunteers provided free surgeries for 9,221 children.

Operation Smile also provides a framework for its partner countries to come together to share knowledge, technology and skills through the use of programs customized to each country's specific medical infrastructure. The annual Operation Smile Physicians' Training Program (PTP) brings surgeons from around the world to the United States for training in specialized surgical skills.

Through organizations such as this, MHAUS and P&G Pharmaceuticals are working to promote MH awareness and preparedness. But there's still a long way to go.
Continued from front page

lining’ and that is, if the test comes back negative, at least we know now.”

With the uncertainty removed, fear is gone.

All patients who have had a positive muscle contracture test should consider the molecular genetic test. This is because a large fraction of such patients will return positive test results and, once a causative mutation is found within a family, then propagation of this finding throughout the family can be achieved for only a fraction of the cost of the muscle contracture test.

As well, all families with a clear history of MH should consider the molecular genetic test. This is regardless of whether some members of the family have already received the muscle contracture test. It is very important, however, that a member within these families who either has had a clear MH event or has had a positive contracture test result should be the first one tested. Testing members of these families first, who do not exhibit any evidence of MH susceptibility, does not appear to be productive.

A genetic test can provide the protection of knowing whether you’re MH-susceptible ... and helps the medical community better understand malignant hyperthermia.

The genetic test for MH is simple and relatively inexpensive. Patients have blood drawn in their local doctor’s office and then shipped to a testing lab. Presently, there are two genetic testing sites for MH: PreventionGenetics, LLC, (715) 387-0484, www.preventiongenetics.com, and the University of Pittsburgh Medical Center, (412) 648-8519, http://path.upmc.edu/divisions/mdx/diagnostics.html

A perfect example is the recent case report published in the August 2007 issue of “Anesthesiology” of a 73-year-old man who experienced a delayed MH episode following laparoscopic and open resection of a rectal tumor, colostomy, and lysis of adhesions. This patient, his wife, and their daughter chose to donate blood for examination of the RYR1 gene.

This is the first report of an individual with a compound heterozygous RYR1 mutation in North America. His wife had no variations within the portions of her RYR1 gene examined. All of their offspring must be considered MH susceptible because it is likely that each has one of their compound heterozygote father’s RYR1 variants.

MHAUS urges anyone tested to have their results recorded in the North American MH Registry (NAMHR). MHAUS supports the NAMHR, which contains clinical information on more than 2,000 individual cases of MH or suspected MH together with muscle biopsy contracture test results.

MHAUS is working diligently to promote the development of genetic testing for MH and has sponsored several meetings of scientists interested in the genetic diagnosis of MH. In 2006, MHAUS authorized a grant to Dr. Sheila Muldoon, Khishge Sambuughin and Barbara Brandom for the in-depth study of DNA changes in 100 MH-susceptible patients. As well, MHAUS has provided seed money to the clinical laboratories to begin testing patients for known genetic changes related to MH.

MHAUS anticipates rapid advances in the genetic diagnosis of MH in the next few years.
Planning for the Unexpected MH Crisis: Special Concerns for the Ambulatory Surgery Center

Editor’s note: A probable MH death four months ago of a patient with six previous uneventful anesthetics, and the death of a 21-year-old woman during elective plastic surgery at an ambulatory center one year ago, show that MH remains a very real threat. MHAUS continues to stress vigilance and preparedness, whether in a hospital, ambulatory surgery center (ASC), or transporting a patient. The following article first appeared in “Outpatient Surgery Magazine” in May 2007 and bears reprinting in these pages as a heed to constant vigilance.

by Henry Rosenberg, MD

When you think of testing for malignant hyperthermia (MH), you probably think of the pre-operative interview. It's standard to ask the patient about her history with anesthesia, follow with some questions about the family's experiences with anesthesia and determine whether there is a family history of unusual muscle disorders or malignant hyperthermia syndrome. This may catch the most obvious red flags, but for practical purposes, it's really not that effective for predicting a patient's risk for developing MH. Even patients who have low risk factors may later present with an MH episode.

Regardless of whether you're working in a hospital or a freestanding facility, if you're administering anesthesia you need a plan to recognize and treat this potentially fatal syndrome as quickly as possible. Here's what you and your staff should know about dealing with MH. The information provided has been developed over many years by those associated with the Malignant Hyperthermia Association of the United States (MHAUS), a not-for-profit patient advocacy group led by laypeople, physicians and scientists with an interest in MH and allied syndromes.

MHAUS's advice: Be prepared

Every minute counts during an MH episode as you and your team work to prevent a bad outcome, so the more preparation you've done in advance, the better. Training for MH is an important component. Everyone should be familiar with MH syndrome and the need for rapid and definitive action once it's diagnosed. Some of the tasks you need to delegate include:

- obtaining and mixing dantrolene;
- getting ice, hypothermia blankets and other devices to cool the patient;
- drawing blood samples for arterial blood gas;
- running electrolyte and coagulation tests; and
- alerting the nearest hospital to expect a patient who is experiencing an MH episode.

MHAUS' professional advisory council has recommended that every facility using general anesthesia with potent volatile agents should have a full supply of dantrolene immediately available stored with sterile water for injection USP without a bacteriostatic agent to reconstitute the dantrolene and a plan to handle the crisis.

To make sure everybody knows what to do, I suggest having a practice drill once a year. It's also wise to put the treatment plan in writing and post it near the ORs in case of an actual event. This will give everyone a quick reminder of what they have to do. You can include the MHAUS Hotline number, (800) MH-HYPER (644-9737) if there is a suspicion that the patient may be experiencing an MH episode.

If you administer gas anesthetics such as sevoflurane or isoflurane or the muscle relaxant succinylcholine, be sure to keep a full supply of dantrolene immediately available stored with sterile water for injection USP without a bacteriostatic agent to reconstitute the dantrolene. Having everything ready to mix lets your staff treat a patient with MH promptly, a valuable advantage, because every minute counts during an MH crisis.

Most offices and ASCs don't have the equipment to analyze arterial blood gases or electrolytes on-site; if that's the case in your facility, make sure you have arranged to rapidly transport specimens to an approved laboratory that reports results promptly.

It's also especially important for freestanding ASCs to have transfer policies and procedures to follow once a patient's condition is stabilized. In this context, stabilizing means administering enough dantrolene so that the patient isn't tachycardic, the end tidal carbon dioxide levels are declining to less than 50 mmHg or less, there is no rigidity and the core temperature is on the way down.

I also think that extra dantrolene (about 1 mg/kg), mixed and ready to use during the move, should be prepared. The receiving hospital should have dantrolene immediately available for administration upon the patient's arrival.

Intraop temperature

Except for very brief cases, you should monitor temperature during...
Continued from page 4

all general anesthetics. In many cases, temperature elevation is the confirmatory sign of MH, but it's usually a late symptom and, frighteningly, it doesn't present in all cases.

The rate of a temperature's rise often indicates the syndrome's severity. During an MH crisis, a person's body temperature can rise a degree every three to five minutes, so in a brief period of time, a patient can go from normothermia to 106°F. When temperature exceeds about 106°F, coagulation abnormalities often occur. If a patient develops such hyperthermically-induced coagulopathy, the likelihood of mortality is high. On the other hand, overly aggressive cooling may lead to marked hypothermia, which can also have adverse consequences.

The anesthesia provider should have and be familiar with the equipment to measure and monitor body temperature continuously during anesthesia. Esophageal, nasopharyngeal or oral temperature monitoring is the ideal way to monitor temperature, but this isn't always possible. It's acceptable to measure rectal, bladder or axillary temperature. However, remember that axillary temperature is often one to two degrees less than core temperature. A patient's forehead skin temperature bears a very rough correlation to the core temperature, and it has the virtue of being non-invasive. If there is a significant change in skin temperature, a more accurate temperature monitor, such as esophageal, should be used.

Temperature monitoring, personnel training and preparedness may seem like a lot of work for what's essentially a rather rare event. It's very likely that the MH plan you make for your facility is rehearsed more often than it's actually used. However, over the course of an anesthesia provider's career he is likely to either detect an MH episode, assist in management of a case or deal with a patient with a family history of MH. Therefore, all anesthesia providers should be aware of the syndrome and its symptoms, treatment and management. Without such preparation and training, an otherwise healthy patient may suffer needless morbidity or mortality. In other words, this knowledge may save a patient's life.

Did you forget something?

If you're moving, or planning to move, don't forget us! Please remember to contact MHAUS to provide us with your new address. Call (607) 674-7901 or send your address change to: MHAUS, P.O. Box 1069, Sherburne, NY 13460-1069.
A Reluctant Career Choice Marked by Profound Dedication

Editor's Note: This is the second in a series of biographies related to those who played an important role in our understanding MH.

Roderick Angus Gordon has been described as a “Renaissance Man” for his varied contributions and organizational skills that furthered the study of anesthesia in Canada.

Born in 1911 in Watrous, Saskatchewan, Canada, he started violin lessons at age six, playing in local concerts throughout his early school years. At age 16 he was awarded a scholarship to the Toronto Conservatory of Music, subsequently joining the faculty, teaching violin and playing in string quartets.

Gordon’s dream of continuing as a concert violinist was shattered by the Depression, and he entered medicine purely for financial reasons. He returned to Saskatchewan, graduating from the local university in 1934 and continuing at the University of Toronto (UT), where he received his MD degree in 1937. There were no openings in surgery, his choice of study; but an appointment in anesthesiology was available: a reluctant start to a career marked by his profound dedication to the specialty.

Following a three-year internship at Toronto General, Dr. Gordon was commissioned in the Royal Canadian Army Medical Corps in 1939. Sent to London, he furthered his learning by observing British anesthetists in a variety of medical centers. Then as casualties of the war mounted, Gordon, by then a Major, served as anesthetist to a team doing plastic reconstructive surgery, especially on injuries of the face. He also attended to civilians and was particularly interested in helping children with cleft palates.

Gordon served six years in England and, returning to Toronto, continued in the Reserve Forces, becoming Consultant in Anesthesia to the Canadian Armed Forces Medical Council, with the rank of Colonel.

While at UT, Dr. Gordon was devoted to influencing others to study anesthesiology. He developed a formal training for residents requiring a year of study and research in the Department of Pharmacology. Recruiting staff members and increasing the department’s funding were also priorities.

Dr. Gordon became interested in the attempts of Drs. Britt and Kalow of UT to examine the medical histories of malignant hyperthermia (MH) susceptible families, to determine the anesthetic agents that trigger MH crises in these families and to develop biopsy screening tests. He joined the researchers in a symposium in which he presented the history of MH. While there were few studies of the syndrome prior to 1966, he stated that hyperthermia as a reaction to anesthesia was first reported in 1937 by Dr. Arthur Guedel who had encountered six cases in a twenty-year period but never reported whether the patients were related. (Guedel, AE: Inhalation Anesthesia. New York, Macmillin, 1937).

The Canadian Anesthesia Society had been founded in 1921, and Dr. Gordon served as its Secretary-Treasurer from 1946 until 1961. His interests lay in the promotion of education, the quality and the economics of practice, including a fee-for-service arrangement. The Canadian Anesthesia Society later established the RA Gordon Clinical Research Award in honor of his giving a strong voice to the specialty in the medical community.

In 1954 Dr. Gordon founded the Canadian Anesthetists Society Journal (now the Canadian Journal of Anesthesia) in which he served as editor until 1982 and as editor emeritus until his death in 1998.

Dr. Gordon’s teaching commitments extended far beyond Canada. He established the Canadian Anesthetists Society’s Training and Relief Fund (now the International Education Fund) in 1964 which supports anesthesia education in developing countries. Especially interested in Nigeria, he helped develop the Department of Anesthesia at the University of Lagos.

He was Chairman of the Organizing Committee of the Second World Congress of Anesthesiologists in Toronto in 1960 and Vice President of the World Federation of Societies of Anesthesiologists from 1968 to 1972.

He was certified by the American Board of Anesthesiology. Fellowships included the Royal College of Physicians of Canada, Royal College of Surgeons in England and the American College of Anesthesiologists. Honorary memberships were also conferred by the Society of Anesthetists in Great Britain and Ireland and the Association of Surgeons of West Africa.

In 1939 Dr. Gordon married a nurse from Toronto General, Miss Ruth Breithaupt. In later years, they and their children and grandchildren vacationed in a self-built home on an island in Georgian Bay, where he instilled a love of music in his family and undertook a never-ending list of carpentry projects. At his death in 1998, Dr. Gordon was still counseling fledgling anesthetists and participating in his many endeavors related to his specialty.
NMSIS Strives for Development of Diagnostic and Treatment Guidelines for NMS

By Diane Van Slyke

NMSIS proposes to assemble a panel of worldwide experts to reach a consensus on diagnostic criteria and recommend treatment for NMS based on the best available empirical evidence.

Reliable guidelines are critical in raising awareness of NMS to assist practitioners in identifying symptoms quickly, thus reducing morbidity and mortality from NMS and other similar-presenting disorders.

Third Annual MH Mini-Conference Scheduled for September 27-28

Start making your plans now for the 3rd Annual MH Mini-Conference in Phoenix, Arizona, on September 27-28, 2008. The conference brings together medical professionals and MH-susceptible families in an open forum discussion of current MH research and testing, which includes molecular genetic testing.

The registration fee is yet determined, but it is expected to be minimal. Last year’s registration fee was $25.00. Visit the MHAUS website at www.mhaus.org for the latest updates and announcement of scheduled speakers.

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 06-07 (Oct. 2006 - Feb. 2007) in memory of Lila and Jerry Lewis. We are most grateful for their support and special tribute gifts.

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Honorary

MHAUS Welcomes Nicole Viera

MHAUS is pleased to announce the hiring of Nicole Viera as Administrative Assistant at our Sherburne, NY office. Nicole is responsible for database maintenance/quality control, organization of discussion group synopses, coordination of meeting arrangements and exhibit booth setup, as well as other general office duties.

Nicole studied Security and Law Enforcement and then joined the Norwich, NY Fire Department after high school, where she took firefighting and EMT courses and was certified in Self-Contained Breathing Apparatus (SCBA). She had made 1st Lieutenant before leaving the fire department and was recognized with a “Life Saving” pin for involvement in saving a person’s life. For the last eight years, Nicole has worked as a Special Education Teacher's Aide, where she worked closely with children with a variety of disabilities: Traumatic Brain Injury (TBI), Cerebral Palsy, Mentally Retarded (MR), Autism, Spinal Bifida, and Asperger's Syndrome.

“I started at MHAUS last August, and it took about a month or two to settle in,” she says. “I enjoy what I do here, and I am continuing to learn about MH.”
by Henry Rosenberg, MD, President MHAUS

During the three month period of January to April 2007, 15 Hotline consultants were called to handle 65 cases and address 17 inquiries related to MH. The consultants were Drs. Brandom, Chapin, Belani, Kaplan, Melton, Wedel, Rosenbaum, Miller, Weglinski, Skoog, Theroux, Tobin, Wong, Millman, and myself.

Calls derived from community hospitals, university hospitals, emergency rooms, surgery centers (5). The calls were placed by anesthesiologists, nurse anesthetists, internists, pediatricians, emergency medicine physicians and surgeons. They derived from all around the U.S.

There was one death clearly related to MH that began during a surgical procedure in a surgery outpatient center. The patient died in the hospital following transfer from complications secondary to high fever. This case has been reported previously in The Communicator (Vol 23, No. 3, Summer 2007).

In 16 cases, the consultants thought the problem was clearly MH or likely MH. In six cases the calls were related to increased muscle tone and rigidity not related to MH. Thirty calls were related to increases in body temperature, but the consultants did not believe that MH was the cause. Seven calls were due to increased carbon dioxide excretion during laparoscopic surgery (see below) and four calls were related to increased carbon dioxide levels not related to MH. The rest of the calls related to a wide variety of clinical situations.

Cases related to increase in body temperature
There were 30 calls related to patients who developed a high body temperature which the consultants thought were either definitely not or unlikely to be related to MH. One half of the calls concerned patients less than four years of age.

In these cases the consultants rely heavily on the results of the determination of the acid content and carbon dioxide tension in the arterial blood (i.e. arterial blood gases), the presence or absence of rigidity or other circumstances that make the diagnosis of MH unlikely. For example, if the patient has experienced an uneventful anesthetic and several hours later develops a high fever (three cases) this is not MH, as MH episodes occur either during the surgical period or within approximately one hour following the end of anesthesia.

In a few cases the cause of the fever was related to a preexisting infection, be it respiratory, ear or other evidence of infection. There were two cases of high fever after appendectomy. In one case the fever occurred well after the surgery and in the other case, the patient had an elevated body temperature prior to surgery.

Other sources of infection that contributed to post operative fever include dental or oral surgery (the mouth is a “dirty” area and when instrumented bacteria may enter the blood stream and cause fever) and the urinary tract. Fever after drainage of infected ears occurred most commonly in young children.

In some cases the temperature elevation was very striking (over 108 degrees) and dantrolene treatment, along with other forms of treatment of elevated body temperature, were employed.

In some cases the cause of the hyperthermia is related to aggressive patient warming. This occurs because there is concern that when a patient, particularly a child, becomes cold during surgery he/she may develop a variety of complications such as prolonged emergence from anesthesia and increased risk of infection. Hence, heating blanket devices are used to keep a patient warm. Careful attention is necessary, however, so that the patient is not warmed excessively. This was the case in at least one patient.

Here is an example of one related to body temperature elevation. An 18-month-old male had surgery for a repair of a urethral deformity. In the recovery area he was noted to have a temperature of 103 degrees. He was breathing rapidly. The blood gas test recommended by the consultant did not indicate changes of MH. The patient was treated with Tylenol, fluid and conservative measures and the temperature returned to normal. In addition, it was revealed that the child had an upper respiratory tract infection one week previously. The patient recovered completely. Rarely is the reason for the elevated temperature a faulty thermometer device, although that is also theoretically possible.

Elevated level of carbon dioxide excretion (end tidal CO2)
One of the earliest signs of MH is an unusually high level of carbon dioxide in the exhaled gases which are routinely measured during anesthesia. However, there are several other reasons for increased excretion of CO2.

Here is the primary one. One of the most significant advances in surgery has been the introduction of minimally invasive techniques to avoid large incision and subsequent pain and prolonged hospital stay as well as a variety of other complications. The technique which is most often used during surgery in the abdomen involves expanding the abdomen by insufflating carbon dioxide gas. This creates a space in the belly for the surgeon to introduce thin instruments through tiny skin incisions (often through the umbilicus or belly button). With these probes the surgeon can cut, suture, and remove diseased tissues. The technique is called laparoscopy and is used extensively throughout the U.S. and Europe for such procedures as gall bladder removal and gynecologic surgery. In some situations the carbon dioxide gas seeps out of the abdomen and spreads under the skin and/or is absorbed into the blood stream. When that occurs, the gas is exhaled and the anesthesia provider notices an increase in end tidal carbon dioxide. It then raises the suspicion that this might be an early sign of MH. Often it is obvious that the elevation is related to the carbon dioxide absorption, but not always. About 10% of all Hotline case calls are related to increased end tidal carbon dioxide during laparoscopy, often accompanied by another sign of MH, increased heart rate. The Hotline consultant must therefore carefully question the circumstances, the type of anesthesia being used, the presence of elevated temperature, or muscle rigidity to determine that the change is or is not related to MH. If the elevated CO2 is reduced by stopping the surgery and removing the gas in the abdomen the diagnosis is clear (i.e., not MH). However, in some cases, because the CO2 has infiltrated under the skin, it serves as a reservoir for excretion of CO2 and the exhaled CO2 levels remain elevated suggesting an MH episode. In two of the six calls for this problem, dantrolene was given, while in four others it was not. All the patients did well. None had post operative signs that they had experienced an MH event.

The anesthesia and surgery medical literature has not addressed this issue in a quantitative manner and there are...
In the U.S. and Canada, the MH Hotline is 1-800-MH-HYPER (1-800-644-9737) Outside the U.S., call 1-315-464-7079

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few guidelines to determine with accuracy whether the patient is experiencing MH or just rapid absorption of carbon dioxide. Our MH Hotline consultants bring their experience and knowledge to analyzing such cases and helping out the patient.

Why is CO2 used to inflate the abdomen? Because it is not flammable, inexpensive, is rapidly absorbed with few physiologic negative effects, and allows the surgeon a clear view of the abdominal contents.

Patients who experienced isolated muscle rigidity

Muscle rigidity on induction or during anesthesia is very unusual and is one of the important signs indicating possible MH. Six calls related to muscle rigidity without other signs of MH. The causes were: insufficient depth of anesthesia when the anesthesiologist attempted to open the mouth, a history of temporomandibular (TMJ) disease limiting mouth opening, increased muscle tone upon induction with the anesthetic gas sevoflurane. Increased muscle tone upon induction or awakening from sevoflurane anesthesia is an uncommon complication of the drug. Increased jaw muscle tone upon induction with the paralyzing agent succinylcholine is often a sign of MH, but only when there is great rigidity of the jaw limiting or preventing mouth opening. However, the anesthesia provider must consider this jaw rigidity as a sign of MH or impending MH until proven otherwise. Another cause of increased muscle tone is shivering or just increased muscle tone following anesthesia because the patient's body temperature has fallen during anesthesia or the patient is experiencing the early onset of an infection.

In one case of prolonged jaw rigidity after surgery in a six-year-old child, it was finally determined that the child refused to open his mouth because of the presence of a breathing tube in his nose and the tape inhibited mouth opening.

Not included in these calls is the occasional patient with a muscle disorder such as myotonia who develops rigidity with succinylcholine or on emergence from anesthesia. In these cases the Hotline consultant prevented needless labeling of the patient as MH susceptible.

Miscellaneous conditions

Anesthesia providers often call the Hotline concerning patients with rather unusual problems. There were 10 such calls. In one case a 29-year-old patient developed muscle contractures (rigidity) due to hyperventilation syndrome secondary to anxiety.

A 46-year-old patient who was anesthetized for a kidney transplant developed a cardiac arrest shortly after induction of anesthesia. No signs of MH were present. A variety of other possibilities were discussed, but the common causes were quickly eliminated. On autopsy the patient was found to have significant inflammation of the heart muscle, a condition called myocarditis that is often difficult to diagnose with routine tests.

A most unusual case involved a 46-year-old, 260 pound body builder who is very muscular and admitted to taking a variety of performance enhancing medications along with a drug used to lower cholesterol (fenofibrate). The anesthesiologist used succinylcholine and propofol for induction and, although there was mild elevation of carbon dioxide, the body temperature was normal. Following the surgery for a tumor of the thyroid gland, the patient awoke complaining of shoulder pain and arm weakness. The anesthesiologist was concerned and had some blood tests done. The patient developed evidence of significant muscle breakdown (elevated creatine kinase, myoglobin in the urine) and evidence of kidney dysfunction. He eventually recovered. The anesthesiologist was seeking guidance as to whether this represented MH susceptibility. Very difficult to state, since the lipid lowering drugs are often associated with muscle breakdown and there is a possibility that the greatly enhanced muscle strength might also have predisposed the patient to muscle breakdown from succinylcholine.

A 31-year-old man had emergency surgery for a femur fracture with MH trigger agents. After the surgery it was learned that the patient had a family history of MH! He had evidence of significant muscle breakdown, but no other signs of MH. He was lucky!

Questions, Questions, Questions

Many questions requiring the expertise of the Hotline consultants were logged – thirty to be exact. These ranged from questions as to the likelihood of MH in a patient with myotonia (not likely) and a variety of other muscle disorders, dantrolene prophylaxis in an MH patient (not needed), precautions for a baby when the father is a known MH-susceptible, but the mother is not (no known problems during the delivery have been reported), proper anesthesia care for a woman with a history of MH during labor, advice on testing for MH-susceptibility, death of a patient in an emergency department without having had anesthesia (not continued on page 10
Continued from page 9

MH). A nurse called from a center that specialized in spine and joint surgery requesting information as to what laboratory testing should be available on site, just in case of MH. They do have the capability of basic laboratory testing and this was felt to be adequate. This is just a sample of the type of questions that we receive!

**Cases where the consultant thought that MH was certain or likely**

Sixteen calls were in this category. The one death almost certainly related to MH took place in an office surgery suite. The patient was having plastic surgery done using isoflurane. At the end of the case the forehead skin temperature was 104 degrees F, and the oral temperature 107, with increased end tidal carbon dioxide. The patient was treated with some amount of dantrolene and sent to the local emergency room where she received more dantrolene. Unfortunately though, the damage was done (most likely the very elevated body temperature) and the patient expired within 10 days. The tragedy is compounded since the patient was only 21-years-old and 10 days. The tragedy is compounded since the patient was only 21-years-old and otherwise healthy.

MHAUS is extremely concerned about the level of preparedness for treating MH in ambulatory centers and in emergency rooms where patients who develop MH are transferred. We will be developing guidelines for such management over the next several months.

Three of the other patients who developed MH were females. The youngest patient was 13, the oldest 57 years of age.

In one situation the anesthesiologist was treating a 31-year-old man who was undergoing ankle surgery who developed unexplained elevation of end tidal carbon dioxide and elevated potassium level along with acidosis requiring aggressive treatment.

Although some colleagues were not convinced that this represented MH, nevertheless dantrolene treatment was begun and the treatment guided by one of the Hotline consultants. His creatine kinase (indicative of muscle damage) was quite elevated. Eventually he was tested for MH-susceptibility and found to be positive. I don’t know why some people are skeptical that MH is occurring when just about all the signs point to MH and the treatment with dantrolene has few side effects.

A very similar case occurred involving a 13-year-old patient undergoing orthopedic surgery but his temperature went up to about 101 degrees F as well. Dantrolene was effective in treating the patient. His post operative creatine kinase was elevated. Testing for susceptibility was not carried out though.

It is of interest that in most of the other cases the signs of MH occurred after several hours of anesthesia. In these cases, the onset of the syndrome was marked by increased exhaled carbon dioxide that was not controlled by increased minute ventilation. Perhaps the reason for delayed onset is that one of the trigger agents (succinylcholine) was not used and perhaps the concentration of the anesthetic gases was reduced because of the concomitant use of intravenous narcotics that are not triggers of MH.

A second MH episode occurred in an ambulatory surgery center involving a 23-year-old undergoing elective plastic surgery. He was treated with dantrolene and the elevated end expired carbon dioxide levels that had been noted, declined. He was transported to the nearest hospital after being treated with the recommended dose of dantrolene and survived.

A 23-year-old man who was undergoing spine surgery with desflurane (a gas anesthetic and MH trigger) and sufentanil (a short-acting narcotic) anesthesia developed rapid increase in expired carbon dioxide, increased heart rate, and a rise in body temperature to 104 degrees. He was also somewhat stiff. He was treated with dantrolene acutely and after the episode, guided by the Hotline consultant, and did well. His creatine kinase indicated marked muscle breakdown since the value was 15,000 units (normal is about 250).

Other MH cases:

- A 57-year-old woman anesthetized with sevoflurane developed increased carbon dioxide and increased temperature after three hours of anesthesia. She received dantrolene and did well. It was revealed afterwards that there is a family history of MH-susceptibility. She should have had an MH ID tag!

- A 32-year-old woman to undergo a C-section developed jaw muscle rigidity with succinylcholine followed by increased heart rate and increased carbon dioxide, increased potassium and acidosis, but no rigidity. She was treated with dantrolene and she and the baby recovered well.

- A 30-year-old man developed increased carbon dioxide levels on emergence from anesthesia with sevoflurane. He was treated with dantrolene and vital signs returned to normal.

A 37-year-old man undergoing knee surgery developed increased carbon dioxide levels and increased heart rate and temperature elevation to 101.2 degrees F after about six hours of anesthesia, along with elevated potassium. Fortunately, this occurred at the end of surgery. No dantrolene was given and he recovered. He was lucky. The consultant advised that the patient be tested for MH-susceptibility and be considered as MH-susceptible even though he did not progress into full blown MH.

A 57-year-old man undergoing neck surgery developed hypertension, increased heart rate and elevated carbon dioxide several hours after induction of anesthesia. His temperature was going up as well. He was treated with dantrolene with reversal of the signs. He was advised to obtain an MH ID bracelet and be tested for MH-susceptibility.

As you can see, although we are happy to provide experts to help in the management of MH and situations that seem to be MH, it is not always possible to confirm the diagnosis as the event is happening. Because the muscle biopsy contracture test is not available at many places and genetic testing is poorly understood by most practitioners, patients are often left in the dark as to their diagnosis. I hope these cases demonstrate the vital role that MHAUS plays in assisting clinicians in the care of patients who are experiencing potentially life threatening conditions. Although most cases turn out well, even one death is too many.

Your support of MHAUS and its programs is vital in continuing our mission.

Note: You can view a glossary of MH-related terms on the MHAUS website at www.mhaus.org.

**Meet This Issue’s Hotline Consultant**

Dr. Henry Rosenberg is Director of the Department of Medical Education and Clinical Research at Saint Barnabas Medical Center in Livingston, New Jersey, Professor of Anesthesiology and the current President of MHAUS. He has been a Hotline Consultant since the beginning of the Hotline in 1982.
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 ❑ (other amount) $ ___________, will help MHAUS serve the entire MH community.

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Every MH-Susceptible Should Wear A Medical ID Tag

MHAUS 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 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 MH-susceptibles during an emergency situation and to prevent a tragic outcome from MH.

For further information, please contact MHAUS at P.O. Box 1069, Sherburne, N.Y. 13460-1069; call 607-674-7901, or visit www.mhaus.org.

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

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

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

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

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THANKS! MHAUS is grateful for the financial support of the following State Societies of Anesthesiology: California, Connecticut, Florida, Illinois, Maine, Maryland, Michigan, Nevada, Ohio and Pennsylvania. Our appreciation also goes to the following state components of the American Society of PeriAnesthesia Nurses: Arkansas, Colorado, Delaware, DC, Illinois, Kansas, Maryland, Missouri, Nebraska, New Hampshire, New Mexico, North Carolina, Pennsylvania, Texas, Vermont and Wyoming. Call the MHAUS office to ask how your group can join their ranks!

Upcoming MHAUS Meeting Dates and Locations
APA, Washington, DC, NMS Meeting, May 3-8; AACN-NTI, Chicago, IL, May 5-8; ASPAN, Grapevine, TX, May 5-7; ASC Association, San Antonio, TX, May 14-17; AANA, Minneapolis, MN, August 9-13; MH Mini-Conference, Phoenix, AZ, September 27-28; ASHRM, Boston, MA, October 2-5; APNA, Minneapolis, MN, October 15-18; ASA, Orlando, FL, October 18-22.

27th Annual European MH Group Meeting
The 27th Annual Meeting of the European MH Group (EMHG) is set for May 28-30 in Lund, Sweden. For general information about the annual meeting, program information, abstract information, registration, Lund venues, and how to there, visit www.emhg.org and click on “meeting” on the left side of the page. See you there.

Speakers Bureau Brings MH Experts to Your Facility
MH experts can come to your facility or meeting and speak to your staff about malignant hyperthermia. MH experts are ready to offer insight and suggestions on the treatment regimen for MH should you find yourself facing a possible MH event. MHAUS offers this service as an important tool to help you keep your patients safe and well. Contact Diane at (607) 674-7901 or email diane@mhaus.org.

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