



**Ambulatory  
Surgery Centers**

## Be prepared: Malignant hyperthermia

**S**uddenly, the word is out that surgery centers and other non-acute care facilities need to reassess their capability to respond to malignant hyperthermia (MH). And “suddenly” is how this rare but often fatal disease manifests. With an estimated occurrence of just 1 in 14,000 surgeries, the condition is not at the top of the checklist for most OR managers, surgeons, or anesthesiologists as a procedure begins. But when a MH occurs, minutes count. Without an action plan and immediate access to drugs and related tools, the patient will likely die.

The industry, as well as the public, got a reminder of that on March 21, 2008, when Stephanie Kuleba, a healthy 18-year-old cheerleader, died following cosmetic breast surgery in a physician’s office surgery suite in Florida. Based on her symptoms, which included a rapid temperature increase, she is believed to have contracted malignant hyperthermia in response to the general anesthesia.

### Speedy response means survival

An incident in 2001 resulted in a different outcome. Following surgery at the Peachtree Orthopaedic Surgery Center in Atlanta, a young man “was nonresponsive to verbal requests and painful stimuli,” according to the center’s administrator, Cheryl Fielder, RN. The anesthesiologist and staff immediately began the malignant hyperthermia (MH) protocol, administering the only known antidote, dantrolene sodium.

The patient recovered.

“While the ASC was able to report a successful patient outcome in this case, its postreview of this event indicated several improvements the ASC has since introduced to enhance its staff’s ability to respond if another MH crisis should ever occur,” Fielder said in a presentation posted on the web site of the ASC Association (ASCA), Alexandria, Virginia (<http://ascassociation.org>).

Fielder and David Strick, MD, the center’s anesthesiology director at the time, developed a set of guidelines for ASCs in case of MH symptoms, which are listed in the article.

Several organizations have issued guidelines on MH, including the Malignant Hyperthermia Association of the United States (MHAUS), the American Society of Anesthesiologists (ASA), and AORN. All stress the importance of preparedness, awareness, and immediate response to symptoms.

### A family affair

The condition first surfaced in Australia in 1960, where 10 members of the same extended family died during surgery with general anesthesia. An investigation showed that malignant hyperthermia appeared to be inherited as a dominant trait, and researchers subsequently have associated mutations in about 20 different genes with it.

In patients with the condition, under anesthesia their muscles become stiff rather than relaxed, breathing accelerates instead of slowing, irregular heartbeats follow while blood pressure falls, and body temperatures rise rapidly, reaching more than 108°F. The muscles release acid and potassium in large amounts that cause cardiac damage or death.

In 1975, experimenting on pigs, which tend to be susceptible to malignant hyperthermia, scientists found that dantrolene would relax muscles, lower temperature, and

### **MH crisis management**

**For live help, call the MH  
hotline:**

**1-800-MH-HYPER  
(1-800-644-9737)  
in US and Canada**

**Other resources available:**

- Emergency therapy poster
- Anesthetic choices for MH-susceptible patients
- Frequently asked questions, and more.

Contact Malignant Hyperthermia  
Association of the United States.  
[www.mhaus.org](http://www.mhaus.org)

halt acid production without damaging the heart. The primary triggers, they found, were halothane and succinylcholine. Following a clinical study in the late 1970s, the FDA approved dantrolene for human use for MH.

Explaining the condition in an article in *Breakthroughs in Bioscience*, Marilyn Green Larach, MD, FAAP, says, "Yet, 1 to 2 patients in the United States still die every year from malignant hyperthermia. Young, healthy patients often succumb because anesthesiologists or nurse anesthetists fail to diagnose MH early enough because of inadequate temperature monitoring, or because they diagnose MH appropriately but lack dantrolene to treat it.

"Many medical facilities outside of hospitals lack dantrolene, even as they continue to administer MH-triggering anesthetics. Patients receiving general anesthesia in surgical offices appear at highest risk. Surgeons and their office administrators sometimes fail to stock dantrolene because of the cost involved."

Indeed, the drug is costly, and the price has been rising steadily. Its primary US supplier is Procter and Gamble, and while it is sometimes discounted, the current list price \$86 per vial, according to a MHAUS source. Protocols call for administration of 36 vials, so keeping the drug on hand is expensive. According to MHAUS, the shelf life is 24 months, although some sources say it can be kept up to 3 years. While repeated purchases may end up being thrown away, when MH occurs, the dose on hand will prove its worth. (MHAUS says it will take expired dantrolene.)

### **Fingering the suspects**

Not all anesthetics trigger MH. According to MHAUS, the culprits are volatile inhalation anesthetics such as sevoflurane and halothane. Another trigger is the depolarizing muscle relaxant succinylcholine.

For patients determined to be susceptible to MH, there are safe alternatives, MHAUS says, including local anesthetics, nitrous oxide, propofol, and ketamine. Also safe are the non-depolarizing muscle relaxants such as pancuronium, and vecuronium.

Following the Florida fatality, MHAUS published a special April edition of its online newsletter, quoting president Henry Rosenberg, MD as saying, "The popularity of cosmetic surgery and the fact that more and more surgeries are being pushed out of the hospital setting and into office-based surgery suites and ambulatory surgery centers has raised concerns by many."

Screening candidates for elective surgery should go a long way toward identifying patients who are at risk for MH. In practice, however, there are limits. Other than relatives who have been stricken in the past, the best indicator is a muscle biopsy that only about 10 hospitals nationwide can perform.

To help assess risk, MHAUS maintains the North American Malignant Hyperthermia Registry and urges patients who have had MH episodes to submit their names and blood samples to aid in research.

Dr Larach and other researchers used the registry database to analyze MH outcomes between 1987 and 2006 and found that "modern US anesthetic practice did not prevent MH-associated cardiac arrest and death in predominately young, healthy patients undergoing low-to intermediate-risk surgical procedures."

The researchers called on anesthesiologists and nurse anesthetists to provide blood samples suitable for genetic research on all patients suffering cardiac arrest or death that could be traceable to MH.

MHAUS has a suggested protocol for anesthesiologists. A poster with a protocol for emergency management can be downloaded from [www.mhaus.org](http://www.mhaus.org). (Look under Medical Professionals.) It could be laminated and posted on the wall of each OR. Guidelines and other information are in the sidebar.

### **The nurse's role**

AORN has an extensive guideline in its 2008 *Perioperative Standards and Recommended Practices* manual. The guideline alerts nurses to both question and reassure patients before surgery. According to AORN, research indicates that between 1 in 30,000 and 1 in 37,000 people may be MH susceptible.

"Nursing care begins when the patient is scheduled for any procedure involving

general anesthesia," the guideline says. The nurse should begin by asking the patient and family about past experience, using such questions as:

- Has anyone ever told you that you had a "bad" reaction to anesthesia?
- Has anyone ever told you that you or a family member had a problem with anesthesia?
- Have you or a family member experienced a high fever while under anesthesia?
- Has anyone in your family died unexpectedly in the operating room?

If the answers indicate susceptibility to MH, the nurse should notify the surgeon and anesthesiologist.

In an ASC or other facility without critical care services, AORN recommends making advance arrangements for transfer to a hospital.

"Patients who know they are susceptible to this potentially lethal syndrome are likely to have high levels of preoperative anxiety," the AORN guideline continues. "They need continual reassurance that everyone is aware of the potential for problems and that an anesthesia treatment plan has been developed to avoid an MH occurrence." ❖

—Paula DeJohn

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