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THE INFORMED PATIENT
By LAURA LANDRO



A Fresh Focus On a Rare Risk Of Anesthesia

April 30, 2008; Page D1

In the midst of surgery for a deviated septum, Catey Merriman's muscles began to go rigid and her temperature soared. As the surgical team realized she was reacting to the anesthesia, they halted the procedure, injected a powerful muscle relaxant and packed her body in ice. The Niskayuna, N.Y., teacher woke up in the hospital's critical-care unit and asked herself, "Did I die?"



Ms. Merriman didn't die, but she easily could have, had the medical team not acted quickly to reverse the onset of malignant hyperthermia, a rare but potentially

Anesthetics can trigger malignant hyperthermia, a rare but deadly disorder, in susceptible patients. MHA/US

deadly disorder. Malignant hyperthermia is caused by an inherited genetic mutation that triggers an uncontrolled rise of calcium levels in muscle cells when a patient inhales certain commonly used anesthetics. A patient's body temperature can jump to 108 degrees in minutes, followed by cardiac arrest and death.

The Malignant Hyperthermia Association of the United States, a nonprofit group started by patients, operates a 24-hour hot line to help medical professionals identify and treat malignant hyperthermia. The association fields about 650 calls a year, including one from the staff treating Ms. Merriman three years ago, according to anesthesiologist Henry Rosenberg, the group's president. Dr. Rosenberg, director of medical education at St. Barnabas Medical Center in Livingston, N.J., says incidents are likely underreported, and he estimates that there are about 1,000 cases a year.

Malignant hyperthermia can occur in any health-care setting. But the recent death of teen Stephanie Kuleba in a Boca Raton, Fla., surgical center where she was undergoing elective breast surgery has put a spotlight on the many doctors' offices and surgical centers where a growing number of surgical procedures are performed. Anesthesia administered in a hospital setting is considered safe because of the round-the-clock presence of trained anesthesiology professionals. In offices and surgery centers, there may not always be an anesthesiologist or nurse anesthetist with graduate training who can recognize and respond to danger signs.



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"As we've made anesthesia safer, one result is the attitude that it must be so easy even a caveman can do it," says Roger Moore, president-elect of the American Society of Anesthesiologists. "There are thousands of things that can go wrong that need the vigilance and training of an anesthesiology professional."

Patients undergoing procedures outside of hospitals may also face higher risk of complications from surgical infections, blood clots and blood-pressure problems that may require emergency transfers to hospitals. Studies show that the highest number of incidents occur with cosmetic-surgery patients, such as those undergoing liposuction in doctors' offices. Several have died after excess fluids were pumped into surgical sites or when they had a bad reaction to overdoses of the local anesthetic lidocaine.

At Risk

Signs of malignant hyperthermia under anesthesia

- **Intense muscle rigidity.** Jaw muscles clench during routine attempt to insert breathing tube
- **Rapid increase in body metabolism.** Indicated by carbon dioxide output and increased acid content in blood
- **Rapidly developing fever.** Could rise quickly to 108 degrees
- **Muscle breakdown.** Indicated by cola-colored urine
- **General signs.** Increased heart rate, potassium in blood, decrease in blood pressure, mottled appearance of skin

Source: Malignant Hyperthermia Association

Currently, 24 states have regulations aimed at protecting patients undergoing medical procedures in an office setting, and more are considering such measures. A spokeswoman for the Ambulatory Surgery Center Association, which represents outpatient centers, says patients should talk with representatives of a surgery center to learn about the quality standards in place and any other licensing or certification criteria that might apply.

Researchers are still studying malignant hyperthermia, and there are no precise statistics on the number of episodes world-wide. But recent studies indicate that as many as 1 in 3,000 people may have the genetic mutation that predisposes them to a malignant-hyperthermia reaction under inhaled anesthesia. Since the disorder was first identified more than three decades ago, death rates have fallen to less than 5% from more than 80%, thanks largely to the introduction of dantrolene, the muscle relaxant that can quickly stop the uncontrolled release of calcium cells.

But while virtually all hospitals have an adequate supply of dantrolene on hand, medical safety experts say doctors' offices and surgical centers often don't. The reason often given by doctors and surgical centers is the cost of the drug, which has a three-year shelf life. But Dr. Rosenberg says that isn't a good excuse: The cost of an adequate supply to treat an episode is about \$2,500, which is a relatively inexpensive

insurance policy against disaster.

Keith Metz, a member of the board of the Ambulatory Surgery Center Association, says that if inhalation anesthetics are being used, the most important questions for patients to ask are whether the center has an attending anesthesiologist and an adequate supply of dantrolene on hand -- and whether staffers know how to use it. Dantrolene can require preparing and administering as many as 36 vials in quick sequence. Dr. Metz, an anesthesiologist at a surgical center in Southfield Hills, Mich., says he has seen a case of malignant hyperthermia only once in his career. Still, he says, "It's the same as saying we are unlikely to have a fire, but it's important to have a fire extinguisher around."

Because inhaled anesthetics that trigger malignant hyperthermia are cheap, effective and easy to use -- including on children -- they are the drugs of choice in many surgeries. Uses range from ear-tube insertions, wisdom-teeth removal and elective cosmetic surgery to even more complex procedures like open-heart surgery. Drugs that trigger malignant hyperthermia include potent inhalation agents like halothane, isoflurane and sevoflurane, and older drugs like ether. Susceptible patients should be sedated instead with intravenous drugs such as propofol, barbiturates and benzodiazepenes, and short-acting narcotics like fentanyl.

Arizona's state medical board earlier this year banned physicians in office-based surgery centers from using any drug that could trigger malignant hyperthermia.

The Malignant Hyperthermia Association urges patients who have ever had a family member experience complications during anesthesia to discuss their own risks before surgery. The group offers detailed information on risks, prevention and treatment on its Web site, mhaus.org.

Those with muscular dystrophy in the family may be at risk. Indeed, Ms. Merriman's father had had the disease, and she says she informed the hospital of this before her surgery. Researchers also are studying whether there is a relationship between malignant hyperthermia and sudden deaths in young athletes and soldiers linked to heat stroke. Dr. Rosenberg says susceptible patients may have no problem under anesthesia in one surgery but may still be at risk in a future procedure. And patients can have a reaction after surgery in the recovery room, so close monitoring is important.

The most accurate test to determine predisposition to malignant hyperthermia is a specialized muscle biopsy that analyzes the response of a piece of muscle taken from the thigh to a triggering anesthetic. But it is available in only a handful of centers and can cost more than \$6,000. A genetic test is available that can be performed on a blood sample, but its accuracy still needs to be improved, Dr. Rosenberg says. Those who do have the genetic disorder or may be at risk because of family history should wear medical-identification bracelets. Teens and young adults, and especially highly athletic men and women, tend to be more vulnerable, perhaps because of their stage of muscle development or hormone levels.

Tom Kuleba, whose daughter Stephanie, a high-school cheerleader, died last month, says he had never heard of malignant hyperthermia and knew of no family history of problems with anesthesia. The surgery was performed by a board-certified plastic surgeon with an anesthesiologist present, and though dantrolene was administered, among the questions raised is the adequacy of the dose. The death is still being investigated by the state medical board. The family has started a Web site, stephaniejudekuleba.org, to educate consumers about the risks of anesthesia, especially in outpatient surgical centers. "We

need to raise awareness so no other parent has to suffer the horrific loss of a child," says Mr. Kuleba.

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