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Office of Medical Information
The University of Texas Health Science Center at Dallas
5323 Harry Hines Boulevard Dallas, Texas 75235
214/688-3404

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CONTACT: Emme Nelson
OFFICE: 214/688-3404
HOME: 214/528-2796

*****Malignant hyperthermia: nightmare for anesthesiologists, patients

DALLAS -- Malignant hyperthermia is the stuff of anesthesiologists' nightmares. Representing one of the very few diseases occurring only with anesthesia, malignant hyperthermia (MH) is a rare, life-threatening condition that causes body temperatures in its victims to skyrocket as high as 115 degrees.

MH is now being successfully treated with the drug dantrolene. The mortality rate from MH has decreased significantly since the introduction of dantrolene -- to less than 10 percent in adequately treated cases and 70 percent in untreated cases or those in whom treatment was delayed. Before the introduction of dantrolene treatment, extreme cases of MH could result in excessive accumulation of fluid in the brain, kidney failure and/or death by cardiac arrest.

Dr. Tillmann Hein, assistant professor of anesthesiology at The University of Texas Health Science Center at Dallas, has seen three cases of MH during surgery in the past eight years and has published several articles dealing with the condition. Hein says preconditions for MH include a susceptibility that is determined by genetic factors, a trigger substance -- which includes certain anesthetics -- and stress. One of the main triggering agents is halothane, a drug used for general anesthesia. Halothane, Hein says, is most often used in anesthetizing children, a possible reason for the higher incidence of MH in the youth population.

In North America MH occurs in about 1 in 50,000 administrations of general anesthetics to adults and 1 in 10,000 in children. The disease affects more males than females, apparently because MH affects the skeletal muscles and those in males are larger, stronger and more susceptible to the triggering influences.

MH has a higher incidence in cooler areas of the world. The disease is more common in Wausau, Wis., and eastern Switzerland. Although it has been observed in North American blacks, MH has never been reported in blacks in Africa. It is not certain whether these geographical differences are due to racial, climatic or reporting variations.

Hein speculates that climate plays a part in the incidence contrasts -- that some children playing vigorously outdoors in hot regions of the world (such as the southwestern United States) who subsequently develop high fever and die of what is presumed to be heat stroke are actually victims of MH. Since those children do not reproduce, the gene disappears in that area.

Hein says the first step in successful management of the disease is recognizing the susceptible patient prior to any surgical procedure. Because a practical, reliable and non-invasive test is not yet available, emphasis must be placed on carefully taken individual and family histories. Indicators of possible susceptibility within these histories include anesthesia-related complications in the individual or family; fatalities in the family related to sudden deaths such as heart attack, heat stroke and sudden infant death; or physical factors such as muscular weakness, frequent muscle cramps, congenital joint dislocations, hernias and all other physical symptoms of connective tissue weakness.

MH may occur as soon as the patient is anesthetized; some patients have exhibited symptoms while in the recovery room. Sometimes the initial symptom of MH in the operating room is lack of relaxation after the administration of succinylcholine, a muscle relaxant. The masseter muscle, which is exercised in chewing, goes into spasms, making the jaw lock, rendering oral intubation impossible. Rapid heart beat, muscle rigidity, bluish discoloration of the skin and sweating may follow. Body temperature and respiratory rate increase.

(More)

If a patient in surgery exhibits symptoms of MH, the first step is to stop administration of the anesthetic, or triggering substance, Hein says. Second, the operation should be aborted, unless it is a lifesaving procedure.

Therapy for MH -- which includes intravenous delivery of dantrolene and cooling of the body -- must be prompt, Hein says. "All hospitals should have a supply of dantrolene in their operating room suite," he says.

Dantrolene is expensive. The drug, which has a two-year shelf-life, costs about $1,000 for 36 vials, the supply necessary for one full-blown case of MH.

A recent informal telephone survey of the 23 Dallas institutions performing surgery showed that all major hospitals do have a supply of this lifesaving drug. However, four of the institutions did not have dantrolene on the premises, a fact that Hein found "appalling."

Hein says it is also beneficial to have a "clean machine," or an anesthesia-delivering machine through which triggering drugs have never traveled, nearby. These machines ventilate the MH victim with pure oxygen, which is crucial in order to prevent carbon-dioxide poisoning and because the victim's metabolism is running in overdrive.

Active cooling of the patient begins immediately with wrapping the patient in cooling blankets, inserting ice water enemas and flushing any open cavity -- such as the abdomen in an appendix operation -- with ice water.

"The crucial step is to begin intravenous dantrolene as quickly as possible," Hein urges.

Hein says the first mention of the disease was about 25 years ago. Of the three episodes he has seen, none developed into full-blown cases because the patient was found susceptible to the disease before surgery and was treated with dantrolene prophylactically.

"Within the last 25 years," Hein says, "we've seen the knowledge develop from total guessing as to what was causing the problem, to pinpointing the triggering agents and understanding it as something that takes places in the skeletal muscle, to discovering the subcellular site within the muscle cells where the disease process is located, to locating in the mid-'60s a suitable animal model in which we can study the disease. From that, we've found a drug that prevents the outbreak of the disease, treats the disease and works at that subcellular site. That drug is dantrolene."

Dantrolene has been advocated as the treatment of choice for MH for about five to six years, Hein says. A support group, known as the Malignant Hyperthermia Association of the United States (MHAUS), has been established for victims and family of victims and others interested in MH.

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NOTE: The University of Texas Health Science Center at Dallas comprises Southwestern Medical School, Southwestern Graduate School of Biomedical Sciences and the School of Allied Health Sciences.