A protocol to manage malignant hyperthermia with general anesthesia
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Introduction
Malignant hyperthermia (MH) is a disorder in genetically susceptible patients characterized by a hypermetabolic state triggered by agents used commonly during general anesthesia. The syndrome first was recognized in 1962 and is associated with tachycardia, muscle rigidity, increased oxygen demand, severe hyperpyrexia, metabolic acidosis, hyperkalemia, elevated serum enzyme creatinine phosphokinase (CPK), and myoglobinuria. Inheritance of MH susceptibility is by autosomal dominant transmission.

Depolarizing skeletal muscle relaxants, such as succinylcholine, and inhalation general anesthetic agents are the most prominently identified trigger agents. The onset can be sudden with or without apparent muscle rigidity and a rise in temperature of 1° C/15 min. Early recognition and prompt therapy often are required to prevent death.

Physiology and Diagnosis
The primary biochemical disorder causing MH has not been identified. However, present evidence implicates calcium regulation in the muscle cells.

A family history of difficulties related to general anesthesia should raise suspicions. There are no specific laboratory tests for determining MH susceptibility in patients, but examining the reaction of biopsied muscle tissue to halothane and caffeine has been reported to be the most reliable predictor of susceptibility. However, the test is not available at most hospitals and requires an invasive procedure to obtain the muscle tissue. Patients should be treated as being at high risk of susceptibility for MH if they have elevated resting serum CPK or inorganic pyrophosphate levels.

Anesthesia Protocol
A successful protocol to manage patients at-risk for MH requiring general anesthesia has been established at the University of North Carolina Hospitals. Contact with all known triggering agents, such as succinylcholine, halothane, ether, cyclopropane, and enflurane, is avoided. Nitrous oxide is used safely but must be delivered by a machine and equipment that is clean of triggering agents and dedicated for use with MH-susceptible patients. Intravenous (IV) barbiturates and narcotics are the agents of choice to obtain general anesthesia. Local anesthetics, both ester and amide types with vasoconstricting agents, can be used safely if required to supplement anesthesia or hemorrhage control.

Pretreatment with dantrolene, a muscle relaxant, is not recommended. Dantrolene blocks release of calcium from the sarcoplasmic reticulum but its use is reserved for when an actual MH reaction is suspected. Side effects of dantrolene include generalized muscle weakness and hepatic toxicity. Conscious patients may experience drowsiness, nausea, and vertigo.

Case History and Management
MDR was a 4-year, 1-month-old Caucasian female with a history of questionable fetal alcohol syndrome, failure to thrive as an infant, microcephaly, language delay, attention deficit syndrome, and persistent gastroesophageal reflux. She was not taking any medications and there was no history of drug allergies.

She had been hospitalized two months previously in a community hospital for dental care in the operating room under general anesthesia. The mandible became tight with both masseter muscles contracted upon administration of IV succinylcholine. The case was aborted due to a suspected early malignant hyperthermia reaction. There was no elevation in temperature or cardiac arrhythmias. The resuscitated patient was observed for several hours prior to discharge. Referral was made to the University of North Carolina for evaluation and treatment of suspected tracheal stenosis and malignant hyperthermia.

An elevated CPK (384 units/L; normal 30–125 units/L) was the only significant abnormal laboratory value. The parents refused to consent to a muscle biopsy for in vitro challenge with halothane and caffeine.

An IV line was established in the operating room and pentothal administrated for induction supported by nitrous oxide and oxygen. Anesthesia machine free of known trigger agents and dedicated to MH management was used to deliver all gases. IV fentanyl was added to maintain a state of anesthesia. Flexible fiber-optic bronchoscopy failed to detect any evidence of tracheal stenosis. Nasotracheal intubation assisted by administering a nondepolarizing muscle relaxant, vecuronium, was uneventful. The heart rate was maintained between 105–155.
SaO2 at 98–100%, and esophageal temperature in the 36.0–36.8°C range throughout the case. All planned dental restorations and extractions were completed and the patient was taken to the recovery room. As a precaution, the patient was admitted to the hospital overnight for observation. The postoperative course was uneventful and the patient was discharged the following morning.

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