Malignant Hyperthermia, MH

Malignant hyperthermia is a rare inherited disease. It causes a rapid rise in body temperature (fever) and severe muscle contractions when a person receives one of the common general (gas) anesthetics. People with this disease have no signs or symptoms except when receiving an anesthetic.

When people with MH receive a gas anesthetic, their muscle metabolism increases. This usually causes muscles to contract strongly. They remain contracted and become rigid. The gas is used to block the feeling of pain and help you sleep during surgery and other procedures. MH can also happen with one of the muscle relaxants (*succinylcholine, Anectine*) used to paralyze a person during surgery. MH is life-threatening without immediate treatment. Even when treated properly, MH can cause death. This is rare and happens in only 5% of cases. Survivors may rarely suffer:

- Brain damage.
- Kidney failure.
- Liver failure.
- Heart failure.
- Muscle death.
- Damage to other organs.

MH is passed on to children by one or both of the parents who are at risk for this disease. A child with one parent with MH has a 50% chance of inheriting this disease. This child is considered to be MH susceptible. Parents who are carriers are often not aware of this risk unless they or a family member develop a life-threatening crisis with anesthesia.

People who have this may never know unless they have a bad reaction to anesthesia during a surgery or have a parent with the problem. Specialized testing can be used to detect this. Usually, people with MH do not have other serious medical problems. Some inherited muscle diseases have a tendency toward malignant hyperthermia.

Not everyone who has a gene defect linked to MH will develop the MH crisis with each exposure to the triggering anesthetics. MH may be triggered in susceptible patients who have had no previous responses to the triggering agents.

SYMPTOMS

People at risk for this may develop muscle stiffness and breakdown of muscle fibers. This muscle spasm may cause:

- A high fever.
- Rapid heart rate.
- An increase of acid levels in the body (*acidosis*).

Fevers can go over 110° F (43.3° C). Blood pressure may become unstable. Heart arrhythmias may develop. The heart may even stop (*cardiac arrest*).

DIAGNOSIS

At this time there is no simple way to test for MH. So routine screening is not done. Genetic testing is only useful in families with a history of MH. It usually involves removing and testing a small piece of muscle tissue from the leg.

TREATMENT

The MH crisis must be recognized and treated early for a good outcome. The anesthesia provider must be attentive. This is important for diagnosis and treatment. The treatment must be continued for up to 3 days after an episode begins. Blood testing is done to follow the course of the crisis.

Your anesthesiologist will know which anesthetics are unsafe.

PROGNOSIS

Severe complications from MH include:

- Cardiac arrest.
- Brain damage.
- Muscle death.
- Internal bleeding.
- Failure of other body systems.

Death is usually due to a secondary cardiovascular collapse. This happens when large amounts of muscle die. They then release potassium into the blood stream.

PREVENTION

The best way to prevent MH is through detection of risk before having surgery. A family history of MH should be given to your caregivers. Any personal history of muscle disorders should be given. Ask if the facility where you or a family member is having surgery is stocked with enough dantrolene to handle a crisis. You should also know if a treatment protocol is available. MH-susceptible patients should always wear identification bracelets or materials to show their MH status.

IDENTIFICATION OF MH-SUSCEPTIBLE PATIENTS

All closely-related members of a family in which MH has occurred must also be considered MH susceptible. They need to be treated accordingly, unless proven otherwise. Those who have had anesthetics without problems may still be at risk. MH related deaths have happened even though a patient may have had earlier surgeries without problems. Any family with a history of anesthetic deaths or complications should make sure their anesthesiologist knows this history. They should also register their MH susceptibility. In the U.S. there is the North American MH Registry of MHAUS, Pittsburgh, PA. Call 412-692-5464 or toll free 888-274-7899.

MH-SUSCEPTIBLE PATIENTS CAN STILL HAVE SURGERY

Surgery can be safely performed in MH-susceptible patients. Non-triggering anesthetics are used. Special precautions and techniques are used as well. If you are a known MH-susceptible patient, make sure your anesthesiologist is familiar with and knows to:

- Avoid the use of MH-triggering anesthetics.
- Be familiar with the signs and treatment of MH.
- Monitor the expired carbon dioxide concentration during a procedure.
- Constantly monitor the temperature during the surgery and recovery. Skin temperature monitoring is not reliable.
- Have an MH kit or cart within the operating room suite. It should be stocked with an adequate supply of dantrolene.

Your anesthesiologist will not be offended by your asking these questions. You should make sure these precautions will be in place.

FOR MORE INFORMATION

You may contact the Malignant Hyperthermia Association of the United States at http://patients.mhaus.org/.

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