

Dear Insurance Company Claims Adjuster,

The Malignant Hyperthermia Association of the United States (MHAUS) is a non-profit volunteer organization dedicated to reducing the morbidity and mortality of malignant hyperthermia (MH) by:

- improving medical care related to MH,
- providing support information for patients and
- improving the scientific understanding and research related to MH.

This letter explains the reasons for and the implications of the muscle biopsy requested. The reasons for the importance are summarized immediately below:

1. A patient armed with this vital information now has proof that the wrong type of medication will kill him/her.
2. If this patient should need to undergo surgery, his/her family and doctors will be aware of his/her true medical condition and a possible death or a long expensive stay in the hospital or ICU can be averted.
3. Prevention is better than a cure. Once armed with the knowledge resultant from this biopsy, a patient can determine whether his/her present or future children are at risk also.

An explanation of whom should undergo testing and the procedure is detailed below:

WHO SHOULD BE TESTED

- A muscle biopsy is indicated for anyone who showed any of the signs associated with MH during administration of an anesthetic.
- Because MH is a genetic disorder, all members of a family in which MH has occurred must also be considered MH susceptible and treated accordingly, unless proven otherwise through a muscle biopsy.
- Currently the muscle biopsy is the only reliable test. Tests based on the concentration of CK enzyme in the blood or a defect on chromosome 19 have been shown to be inconclusive.

PROCEDURE

I. The Muscle Biopsy:

A muscle sample will be removed from either the vastus lateralis or vastus medialis muscles of the patient. A non-MH-triggering anesthesia will be used for the procedure. This usually involves a block of the lateral femoral cutaneous nerve along with mild sedation. Children will usually receive general anesthesia using a non-triggering agent. The biopsy procedure will be performed on an outpatient basis. Anesthesia and surgical care will be performed according to standard outpatient protocols.

II. The Halothane-Caffeine Contracture Test:

The halothane-caffeine contracture test will be performed in accordance with the guidelines developed by the North American Malignant Hyperthermia Registry over two years. The test will be initiated as soon as the muscle sample is available. There are currently only a few test centers located throughout the US. This list is available at www.mhaus.org or upon request.

III. Test Results and Patient Counseling:

The results of the contracture test will be described to the patient in terms of the sensitivity of the patient's muscle sample to halothane and caffeine in vitro. Historically there have been no false negative test results.

The test result will be sent to the North American Malignant Hyperthermia Registry. The result will be explained to the patient and the following topics discussed:

1. The known genetic details of malignant hyperthermia and new genetic screening procedures;
2. the possible testing of other family members,
3. the monitoring of serum CK levels in potentially susceptible family members and
4. the clinical implications for future anesthetics in MH positive individuals.

All patients who had a positive contracture test will be advised to wear a medical identification tag. In addition, the possibility of performing genetic mapping studies will be discussed.

IV. Blood Samples:

Each patient will be asked to provide a blood sample for future genetic analysis. The North American Malignant Hyperthermia Registry recommends that each testing center attempt to obtain such samples from each tested patient.

For further information regarding malignant hyperthermia and bibliographic information concerning this test, please feel free to contact us at 607-674-7901.

Sincerely,



Henry Rosenberg, MD
President