

The North American Malignant Hyperthermia Registry

**MALIGNANT HYPERTHERMIA
BIOPSY
and
DIAGNOSTIC CONSULTATION
REPORT**

INSTRUCTIONS FOR USE

This form is only to be used by the staff of an MH diagnostic center.

1. Use this form for each patient referred to you for MH evaluation, if they undergo muscle biopsy.
2. If any adult relatives wish to be registered by name, separate consent forms for participation in the Registry must be signed by that relative. If you wish to register a minor under the age of 18, a consent form must be signed by one of the minor's parent or guardian.
3. The Center Director must review and sign this form verifying accuracy before it is submitted to the Registry.
4. Please make a photocopy of the completed form for your records.
5. Submit original completed form to:
The North American Malignant Hyperthermia Registry
University of Florida
Department of Anesthesiology
1600 SW Archer Road, PO Box 100254
Gainesville, FL 32610

I certify that the information contained in this report is complete and accurate.

Biopsy Center Director Signature

_____/____/____
year month day

MH BIOPSY AND DIAGNOSTIC CONSULTATION REPORT

Version 9.6

May 2014

Complete this form for each patient referred for MH susceptibility evaluation. The MH muscle biopsy center director must review the completed form before it is returned to the NAMHR.

1. MH muscle biopsy center code number:
see final page for code numbers

PATIENT IDENTIFICATION

2. North American MH Registry Number for this patient (if previously assigned)

3. Any previous North American MH Registry numbers associated with the patient. That is, AMRA, RSR, (formerly AKA), close relative's reports, etc.

- a. _____ Comment _____
- b. _____ Comment _____
- c. _____ Comment _____

4. Patient's Initials

first middle last

5. Has consent been obtained to enter patient's name into the Registry?

check one

() yes

() no

If yes, please complete a-g on following page.

Note: DO NOT COMPLETE IF CONSENT HAS NOT BEEN OBTAINED

- a. Patient's name. This is the primary subject

last first middle

b. Patient's previous name

last first middle

c. Patient's maiden name _____

last

d. Patient's Address

street address

city state/province zip/postal code

country

e. Phone number

(Home) (____) ____ - _____

(Work) (____) ____ - _____

f. Patient e-mail address _____

g. Date of patient's birth

____ - ____ - ____ \ ____ \ ____

year month day

DEMOGRAPHIC INFORMATION

6. Sex

check one

() male

() female

7. Weight

____.____ kilograms OR ____ lbs

8. Height

_____ cms OR _____ ft _____ inches

9. Year of patient's birth

10. Race:

check as many as apply

- Caucasian
- African
- Hispanic
- East Asian
- African-American
- South Asian
- Native American
- Middle Eastern
- Hawaiian or Pacific Islander
- other (*specify*): _____

11. Body Build

check one

- Normal
- Lean
- Muscular
- Obese
- Postpartum
- Other (*specify*): _____

12. State or province of patient's residence

FAMILY IDENTIFICATION

13. Does the primary subject have minor children or siblings under the age of 6 and does this minor child's parent or guardian consent to the child being in the Registry?

check one

- yes
- no

If yes, please complete below for all children under the age of 6

a. **name**

_____ last _____ first _____ middle

Date of birth

____ \ ____ \ ____
year month day

Is this the child or the sibling of the biopsied patient?

check one

child

sibling

b. **name**

_____ last _____ first _____ middle

Date of birth

____ \ ____ \ ____
year month day

Is this the child or the sibling of the biopsied patient?

check one

child

sibling

c. **name**

_____ last _____ first _____ middle

Date of birth

____ \ ____ \ ____
year month day

Is this the child or the sibling of the biopsied patient?

check one

child

sibling

d. **name**

_____ last first middle

Date of birth

____ \ ____ \ ____
year month day

Is this the child or the sibling of the biopsied patient?

check one

child

sibling

e. **name**

last first middle

Date of birth

____ \ ____ \ ____
year month day

Is this the child or the sibling of the biopsied patient?

check one

() child

() sibling

14. Has consent been obtained to enter the names of children or siblings ages 6 through 17, or ages 18 and over, of the biopsied patient into the Registry?

NOTE: CONSENT MUST BE OBTAINED FROM EACH CHILD/SIBLING OVER 18 YEARS OF AGE FOR WHOM YOU ENTER THIS DATA (If the child/ sibling is deceased, the following data may be entered with the consent of the next of kin*. If the child is under 18 years of age, consent must be obtained from the child's parent or guardian).

* check your local/state regulations regarding the definition of next of kin

check one () yes

() no

If yes, complete below for all individuals for whom consent has been obtained

a. **name**

last first middle

Date of birth

____ \ ____ \ ____
year month day

Is this the child or the sibling of the biopsied patient?

check one

() child

() sibling

b. **name**

last first middle

Date of birth

_______________\ _____\ _____\ _____\
year month day

Is this the child or the sibling of the biopsied patient?

check one

child

sibling

c. **name**

_______________\ |_______________\ |_______________\
last first middle

Date of birth

_______________\ _____\ _____\ _____\
year month day

Is this the child or the sibling of the biopsied patient?

check one

child

sibling

d. **name**

_______________\ |_______________\ |_______________\
last first middle

Date of birth

_______________\ _____\ _____\ _____\
year month day

Is this the child or the sibling of the biopsied patient?

check one

child

sibling

15. Has consent been obtained to enter the names of the parents of a biopsied patient? *check one* yes

no

If yes, complete below

NOTE: CONSENT MUST BE OBTAINED FROM EACH PARENT FOR WHOM YOU ENTER THIS DATA (If the parent is deceased, the following data may be entered regardless of consent status.)

a. Mother of biopsied patient

last first middle

Date of mother's birth

____-____-____ \ ____-____ \ ____-____

year month day

Mother's maiden name _____

last

Father of biopsied patient

last first middle

Date of father's birth

____-____-____ \ ____-____ \ ____-____

year month day

16. Family History Table

Key to Family History table (below)

| Relationship to Patient | Known Medical Problems |
|---------------------------|---|
| a. child | 1. fatal MH |
| b. grandchild | 2. survived fulminant MH event |
| c. brother/sister | 3. possible MH event |
| d. half-sibling | 4. MH family history (use only for those relatives with CHCT results) |
| e. niece/nephew | 5. perioperative death - not thought to be MH |
| f. mother | 6. perioperative death - etiology undetermined |
| g. maternal grandparent | 7. S.I.D.S. or cot death |
| h. maternal aunt/uncle | 8. Sudden death - unknown cause, age 1.5 to 45 yrs |
| j. maternal first cousin | 9. heat stroke |
| k. maternal second cousin | 10. neurolept malignant syndrome |
| m. maternal - other | 11. myopathy |
| n. father | 12. idiopathic creatine kinase elevation |
| o. paternal grandparent | 13. CFIDS (Chronic Fatigue and Immune Dysfunction Syndrome) |
| p. paternal aunt/uncle | 14. muscle pain, weakness or fever with exercise |
| q. paternal first cousin | 15. episodic dark urine and muscle pain |
| r. paternal second cousin | 16. diabetes |
| s. paternal - other | 17. none of the above |
| t. relative by marriage | 18. unknown |

u. other blood relative

Please complete one row for each relative for whom relevant medical history is known.

| <u>Relative's Initials</u> | <u>Registry Number</u> <i>Leave blank if relative not registered. Insert "?" if relative registered but number not known</i> | <u>Relationship to Patient</u> <i>Select one letter from left-hand column above.</i> | <u>Sex</u> <i>M=Male F=Female</i> | <u>Medical Problems</u> <i>Select one or more numbers from right-hand or column above.</i> | <u>CHCT Test Result</u> <i>Write "pos", "neg", "equiv", "unknown" "not performed", "other"</i> | <u>Genetic Result</u> <i>Specify familial mutation or "neg", "not performed", or "other"</i> |
|----------------------------|---|---|--|---|---|---|
| _____ | _____ | _____ | _____ | _____ | _____ | _____ |
| _____ | _____ | _____ | _____ | _____ | _____ | _____ |
| _____ | _____ | _____ | _____ | _____ | _____ | _____ |
| _____ | _____ | _____ | _____ | _____ | _____ | _____ |
| _____ | _____ | _____ | _____ | _____ | _____ | _____ |
| _____ | _____ | _____ | _____ | _____ | _____ | _____ |
| _____ | _____ | _____ | _____ | _____ | _____ | _____ |
| _____ | _____ | _____ | _____ | _____ | _____ | _____ |
| _____ | _____ | _____ | _____ | _____ | _____ | _____ |
| _____ | _____ | _____ | _____ | _____ | _____ | _____ |

FAMILY HISTORY

16a. Before this episode, was the patient’s family history positive for:
check all applicable

- () malignant hyperthermia
- () masseter spasm
- () intraoperative death not thought to be MH
- () sudden infant death syndrome or cot death
- () sudden death from unknown cause at < 45 year >1.5 years
- () heatstroke
- () neurolept malignant syndrome
- () intolerance to heat
- () chronic muscle pain
- () frequent muscle cramps
- () chronic muscle weakness
- () exercise intolerance due to muscle pain, weakness or fever
- () episodes of dark urine and muscle pain
- () myopathies *specify type; write unknown if not known:* _____
- () idiopathic creatine kinase elevation

- diabetes
 - Type 1
 - Type 2
- none of the above
- unknown
- other specify _____

MEDICAL HISTORY

17. Does the patient have any of the following complaints?

check all applicable

- muscle weakness interferes with daily activity at least once/week
- muscle cramps interfere with daily activity at least once/week
- cola colored urine
- heat stroke or heat prostration
- oral (or rectal/axillary equivalent) fever $>38.6^{\circ}\text{C}$ or 101.4°F at least 6 times/year without medical cause
- recent generalized infection
 - If there was infection, how long ago was it? ___ (days)
- recent use of cholesterol lowering drugs
 - If so, which drug _____, and when was it last ingested? ___ (days)
- a regular regimen of physical activity?
 - If so, when was the last work-out? ___ (days)
- ingestion of any medicine to improve muscular performance
- intolerance to heat
- exercise intolerance due to muscle pain, weakness or fever
- diabetes
 - Type 1
 - Type 2
- other (*specify*) _____
- none of the above
- unknown

18. Has patient ever had physical findings of:

check all applicable

- increased muscle tone
- decreased muscle tone
- generalized muscle weakness
- myopathy *specify type; write unknown if not known:* _____
- ptosis
- strabismus
- hiatal hernia
- inguinal hernia
- umbilical hernia

- undescended testes
- clubbed foot
- joint hypermobility
- kyphoscoliosis (moderate or severe; curve >45°)
- pectus carinatum
- winged scapulae
- skeletal fractures (more than 2)
- gallstones
- kidney stones
- laryngeal papillomas
- other (*specify*): _____
- none of the above
- unknown

ANESTHETIC HISTORY

19. How many times was this patient anesthetized prior to this evaluation?

- ___ ___ unknown but > 0 unknown
Skip to question 35 if the response is zero

20. How many were general anesthetics?

- ___ ___ unknown but > 0 unknown

21. Indicate the number of anesthetics with the following agents

- ___ ___ volatile agents without succinylcholine
 ___ ___ volatile agents with succinylcholine
 ___ ___ succinylcholine without other known triggering agents

22. Year of most recent anesthetic (excluding present evaluation)

- ___ ___ ___ ___ unknown

23. Were unusual metabolic responses noted during prior anesthetics?

- check one* yes
 no
 unknown

23a. Were unusual metabolic or muscular responses noted during prior anesthetics?

- check one* no
 yes
 unknown

24. Was there delayed awakening from previous general anesthetics?

check one yes

no

unknown

25. How many anesthetics were suspect for possible MH (director's opinion)?

— —

26. How many fulminant MH episodes occurred (director's opinion)?

— —

Skip to question 35 if the answer to questions 25 and 26 are zero

27. If the patient experienced possible or fulminant MH, answer questions a-w.

Report the anesthetic that was most suspect for MH

a. Date of possible or fulminant MH episode: _____ \ _____ \ _____
year month day

() unknown

NOTE: If consent for the patient has not been obtained only enter the year.

b. Patient weight at time of incident

____.____ kilograms OR _____ lbs

c. Height

_____ cms OR _____ ft _____ inches

d. State or province of patient's residence at time of incident.

— —

e. Location of incident

i. Hospital _____

ii. City _____

iii. State or Province _____

f. Type of procedure scheduled

check all applicable

() cardiothoracic

() thoracoscopic surgery (thoracic)

() dental

() oral surgery

() ear, nose, or throat

() orthopedic

() eye

() plastic surgery

() general surgery

() radiology

() laparoscopic surgery

() obstetrics

a) abdominal

b) pelvic

c) other (specify) _____

() gynecology

() urology

() neurosurgery

() vascular

transplant – Transplant type _____
 unknown other (*specify*): _____

g. Was the procedure an emergency?

check one

yes

no

unknown

g (a). Was the procedure performed outside a hospital?

check one

no

yes

ambulatory surgery center

office

unknown

g (b). Did this adverse reaction occur without exposure to anesthetic?

check one

no

yes

unknown

g (c). Was the environment hot when this reaction occurred?

check one

no

yes

If yes how hot? ___ . ___ C or ___ . ___ F

h. Was any infection present at the time of this surgery?

check one yes

no

unknown

i. If infection was present, what organisms were known to be present?

j. Premedication and anesthetic agents utilized (before reaction occurred):

check all applicable

sodium citrated citric acid (Bicitra)

atropine

cimetidine (Tagamet)

glycopyrrolate (Robinul)

famotidine (Pepcid)

lansoprazole (Prevacid)

ranitidine (Zantac)

scopolamine (Hyoscine)

metoclopramide (Reglan)

dolasetron (Anzemet)

omeprazole (Prilosec)

droperidol (Inapsine)

- hydroxyzine (Vistaril)
- ondansetron (Zofran)
- promethazine (Phenergan)
- diphenhydramine (Benedryl)
- clonidine (Duraclon)
- dexmedetomidine
- ketorolac (Toradol)
- acetaminophen (Tylenol)
- diazepam (Valium)
- lorazepam (Ativan)
- midazolam (Versed)
- etomidate (Amidate)
- ketamine (Ketalar)
- propofol (Diprivan)
- alfentanil (Alfenta)
- fentanyl (Sublimaze)
- fentanyl and droperidol (Innovar)
- meperidine (Demerol)
- morphine
- remifentanyl (Ultiva)
- sufentanil (Sufenta)
- hydromorphone (Dilaudid)
- unknown
 - NO potent volatile anesthetic**
- sevoflurane (Ultane)
- desflurane (Suprane)
- isoflurane (Forane)
- nitrous oxide
- other (specify): _____
- nalbuphine (Nubain)
- naloxone (Narcan)
- atracurium (Tracrium)
- cisatracurium (Nimbex)
- rocuronium (Zemuron)
- vecuronium (Norcuron)
- pancuronium (Pavulon)
- other NMB
- IM** succinylcholine (Anectine)
- IV** succinylcholine (Anectine)
 - NO succinylcholine**
- edrophonium (Tensilon)
- neostigmine (Prostigmin)
- physostigmine (Antilirium)
- bupivacaine (Marcaine)
- levo-bupivacaine
- chloroprocaine (Nesacaine)
- cocaine
- etidocaine (Duranest)
- lidocaine (Xylocaine)
- mepivacaine (Carbocaine)
- prilocaine (Citanest)
- procaine (Novocain)
- ropivacaine (Naropin)
- tetracaine (Pontocaine)
- epinephrine
- ephedrine
- neosynephrine

k. Anesthesia induction time
 ___ . ___ (in hours, express parts of an hour using decimal points)
 (example – 3 minutes = 0.05)

1. General anesthetic induction method
 check one
 inhalation

- intravenous
- other (*specify*): _____
- not applicable

m. Anesthesia duration
 __ __. __ __ (*hours and minutes since induction*)

- n. Type of anesthetic prior to adverse metabolic or muscular reaction to anesthesia
check all applicable
- monitored anesthesia care (local standby)
 - regional anesthesia
 - spinal anesthesia
 - epidural anesthesia
 - general anesthesia **without** endotracheal intubation
 - general anesthesia **with** endotracheal intubation
 - tourniquet use
 - tourniquet use
 elapsed time after the start of anesthesia tourniquet was inflated
 __ __. __ __ (*hours and minutes since induction*)
 elapsed time after final release of tourniquet
 __ __. __ __ (*hours and minutes since induction*)

Patient Monitoring Utilized

- o. Monitoring utilized (before reaction occurred):
check all monitoring used
- blood pressure monitor
 - electrocardiograph
 - stethoscope
 - arterial catheter
 - central venous catheter
 - pulmonary artery catheter
 - end-tidal PCO₂
 - pulse oximeter
 - bladder (Foley) catheter

temperature probes:

- axillary
- bladder
- esophageal
- nasopharyngeal
- rectal
- skin - electronic
- skin - liquid crystal
- tympanic
- other monitoring (*specify*): _____

- p. If a liquid crystal temperature probe was used, did it accurately trend with core temperatures?

check one

yes

no

- q. Was a forced air or I.V. warming device in use?

check one

yes

no

unknown

Documentation of the Reaction

- r. Abnormal signs judged to be inappropriate by the attending anesthesiologist or other physician:

RANK in order of appearance. NUMBER do not check. WRITE ZERO if sign did not occur. (a number may be used more than once if signs were noted simultaneously)

___ masseter spasm: mouth cannot be fully opened but intubation possible

___ masseter spasm: teeth clamped shut, intubation via direct visualization impossible

___ generalized muscular rigidity

___ cola colored urine

___ tachypnea

___ hypercarbia

___ cyanosis

___ skin mottling

___ sinus tachycardia

___ ventricular tachycardia

___ ventricular fibrillation

___ elevated temperature

___ rapidly increasing temperature

___ sweating

___ excessive bleeding

___ hypertension > 20% of baseline

___ other (*specify*): _____

- s. Signs: Maximum values and times

fill in the blanks

___ . ___ ___ time first adverse sign noted (***after induction***)
(*hours and minutes since induction*)

___ . ___ ___ time second adverse sign noted (***after induction***)
(*hours and minutes since induction*)

___ . ___ . ___ maximum temperature noted (°C) **OR**
 ___ . ___ . ___ maximum temperature noted (°F)
 ___ . ___ . ___ time maximum temperature noted (*after induction*)
 (*hours and minutes since induction*)
 ___ . ___ . ___ maximum end-tidal PCO₂ noted (mmHg)
 ___ . ___ . ___ time maximum end-tidal PCO₂ noted (*after induction*)
 (*hours and minutes since induction*)

t. Type of ventilation used at the time hypercarbia was first observed:

- check one*
- () spontaneous
 - () assisted
 - () controlled
 - () not applicable
 - () unknown

u. Laboratory Evaluation used during the reaction.

Fill in the blanks for all lab tests obtained

Most abnormal arterial blood gas after MH was suspected:

___ . ___ . ___ FiO₂

___ . ___ . ___ pH

___ . ___ . ___ PCO₂

___ . ___ . ___ liters/minute ventilation at the time
of this blood gas.

___ . ___ . ___ PO₂

___ . ___ . ___ BE (mEq/L) (specify ±)

___ . ___ . ___ Bicarbonate (mEq/L)

___ . ___ . ___ Time (*after induction*)
(*hours and minutes since induction*)

peak lactic acid

___ . ___ . ___ mmol/L

peak K⁺

___ . ___ . ___ mEq/L or mmol/L

peak post-op creatine kinase*

first creatine kinase*

last creatine kinase*

___ . ___ . ___ U/L

___ . ___ . ___

___ . ___ . ___

___ hours after induction

___ hrs after induction

___ hrs after induction

***(recommended intervals for creatine kinase determination are 0, 6, 12, 24 hours after adverse reaction)**

serum myoglobin

___ , ___ ng/ml

___ hours after induction

urine myoglobin

___ , ___ mg/L

___ hours after induction

| | | |
|----------------------------------|-----|-----------------------------------|
| PT (prothrombin time) | INR | PTT (partial thromboplastin time) |
| __ __ seconds | __. | __ __ seconds |
| laboratory upper limit of normal | | laboratory upper limit of normal |
| __ __ __ seconds | | __ __ __ seconds |
| platelet count | | fibrinogen |
| __ __ __ , __ __ __ | | __ __ __ __ mg/dl |

v. Monitoring utilized (**after** reaction occurred):

check all monitoring used

- | | |
|---|---|
| <input type="checkbox"/> blood pressure monitor | <input type="checkbox"/> end-tidal PCO ₂ |
| <input type="checkbox"/> electrocardiograph | <input type="checkbox"/> pulse oximeter |
| <input type="checkbox"/> stethoscope | <input type="checkbox"/> bladder (Foley) catheter |

- arterial catheter
- central venous catheter
- pulmonary artery catheter

temperature probes:

- axillary
- bladder
- esophageal
- nasopharyngeal
- rectal
- skin – electronic
- skin - liquid crystal
- tympanic

other monitoring (*specify*): _____

w. Treatment given for possible or fulminant MH

check all treatments utilized; fill in the blanks

- Volatile anesthetics discontinued
- __ __. __ __ Time (**after induction**)
(*hours and minutes since induction*)

- Anesthesia machine changed
- Anesthesia circuit changed
- Hyperventilation with 100% oxygen
- Dantrolene (type)
 - Dantrium
 - Revonto
 - Ryanodex

__ __ __. __ Initial dose (mg)

__ __. __ __ Time of first dose (**after induction**)
(*hours and minutes since induction*)

_____.____ Total dose (mg)
 _____.____ Time of last dose (*after induction*)
 (hours and minutes since induction)

- | | |
|---|---|
| <input type="checkbox"/> Active cooling | <input type="checkbox"/> Fluid loading |
| <input type="checkbox"/> Furosemide | |
| <input type="checkbox"/> Mannitol | <input type="checkbox"/> Bicarbonate |
| <input type="checkbox"/> Glucose, insulin | <input type="checkbox"/> Amrinone |
| <input type="checkbox"/> Bretylium | <input type="checkbox"/> Vasopressor |
| <input type="checkbox"/> Lidocaine | <input type="checkbox"/> Procainamide |
| <input type="checkbox"/> CPR | <input type="checkbox"/> Defibrillation |
| <input type="checkbox"/> other (<i>specify</i>) _____ | |
| <input type="checkbox"/> none of the above | |

28. Mark any of the following that were noted after dantrolene was given:

- Decrease in heart rate.
 Decrease in end-tidal carbon dioxide or carbon dioxide tension in blood.
 Decrease in temperature.

If none were noted, please skip to question 35

29. How many minutes after dantrolene administration was the maximum decrease in this sign noted and what was the magnitude of this change?

Heart rate

- (___) minutes
 (___) (beats/min)

Carbon dioxide

- (___) minutes
 (___) (mmHg or torr)

Temperature

- (___) minutes
 (_____.____ °C) or (_____.____ °F)

30. Were any problems noted with the dantrolene administration?

check one

- yes
 no

If no, please skip to question 32

31. What were the observed dantrolene complications?

check all applicable

- phlebitis
 excessive secretions
 gastrointestinal upset
 hyperkalemia
 muscle weakness

- respiratory failure
- other (*specify*): _____

32. Did the patient develop additional signs or symptoms after initial adequate treatment (recrudescence)? *check one*

- yes
- no

If no, please skip to question 35

33. What was the time of the recrudescence?
 ___ : ___ time (hours after anesthetic induction)

34. Signs of recrudescence that were noted:
 (judged to be inappropriate by the attending anesthesiologist or other physician)
RANK in order of appearance.
(a number may be used more than once if signs were noted simultaneously)

- ___ masseter spasm: mouth cannot be fully opened but intubation possible
- ___ masseter spasm: teeth clamped shut, intubation via direct visualization impossible
- ___ generalized muscular rigidity
- ___ cola colored urine
- ___ tachypnea
- ___ hypercarbia
- ___ cyanosis
- ___ skin mottling
- ___ sinus tachycardia
- ___ ventricular tachycardia
- ___ ventricular fibrillation
- ___ elevated temperature
- ___ rapidly increasing temperature
- ___ sweating
- ___ excessive bleeding
- ___ hypertension > 20% of baseline
- ___ other (*specify*): _____

ADVERSE METABOLIC REACTION TO ANESTHESIA (AMRA) REPORT

35. If an AMRA Report was submitted, did you review it after pertinent anesthesia records were obtained?
check one

- yes
- no

If no, skip to question 57

AMRA number (if known) ___ ___ ___ ___ ___

36. Were errors found in the AMRA Report?
check one
 yes
 no *If yes, specify* _____

LABORATORY EXAM:

Serum Creatine Kinase

37. Creatine kinase at the time of evaluation:
 ___ ___ ___ , ___ ___ ___ U/L
38. Laboratory upper limit of normal for creatine kinase
 ___ ___ ___ U/L

Muscle Biopsy

39. Was a MH diagnostic muscle biopsy indicated?
check one
 yes
 no
Note: If no, then skip to question 55
40. What was the reason for the MH diagnostic muscle biopsy?
check all applicable
 fulminant MH
 possible MH event (may include MMR), AMRA Report completed
 possible MH event (may include MMR), AMRA Report not completed
 family history of MH
 control
 negative genetic test
 location _____
 date _____
 exons examined _____

- other (*specify*): _____
41. Date of muscle biopsy
 ___ ___ ___ \ ___ ___ \ ___ ___
 year month day
42. Time of anesthetic induction for muscle biopsy
 ___ ___ : ___ ___ (military time)

43. Time muscle was excised

___ ___ : ___ ___ (military time)

44. Which muscle was biopsied?

check one

vastus

rectus abdominus

gracilis

other (*specify*): _____

45. Were any medications being taken at the time of biopsy?

check one

yes

no

If yes, specify type of medication:

| <u>Type of agent</u> | <u>Name of Drug</u> | <u>Hrs. before biopsy</u> |
|-------------------------|---------------------|---------------------------|
| calcium channel blocker | _____ | _____ |
| neuroleptic agent | _____ | _____ |
| adrenergic agent | _____ | _____ |
| lipid lowering agent | _____ | _____ |
| other | _____ | _____ |

46. Premedication and anesthetic agents utilized (for biopsy):

check all applicable

- | | |
|--|--|
| <input type="checkbox"/> sodium citrated citric acid (Bicitra) | <input type="checkbox"/> clonidine (Duraclon) |
| <input type="checkbox"/> cimetidine (Tagamet) | |
| <input type="checkbox"/> famotidine (Pepcid) | <input type="checkbox"/> dexmedetomidine |
| <input type="checkbox"/> lansoprazole (Prevacid) | <input type="checkbox"/> ketorolac (Toradol) |
| <input type="checkbox"/> ranitidine (Zantac) | <input type="checkbox"/> acetaminophen (Tylenol) |
| <input type="checkbox"/> metoclopramide (Reglan) | <input type="checkbox"/> diazepam (Valium) |
| <input type="checkbox"/> omeprazole (Prilosec) | <input type="checkbox"/> lorazepam (Ativan) |
| <input type="checkbox"/> atropine | <input type="checkbox"/> midazolam (Versed) |
| <input type="checkbox"/> glycopyrrolate (Robinul) | <input type="checkbox"/> etomidate (Amidate) |
| <input type="checkbox"/> scopolamine (Hyoscine) | <input type="checkbox"/> ketamine (Ketalar) |
| | <input type="checkbox"/> propofol (Diprivan) |
| <input type="checkbox"/> dolasetron (Anzemet) | |
| <input type="checkbox"/> droperidol (Inapsine) | <input type="checkbox"/> alfentanil (Alfenta) |
| <input type="checkbox"/> hydroxyzine (Vistaril) | <input type="checkbox"/> fentanyl (Sublimaze) |
| <input type="checkbox"/> ondansetron (Zofran) | <input type="checkbox"/> fentanyl and droperidol |
| <input type="checkbox"/> promethazine (Phenergan) | (Innovar) |
| <input type="checkbox"/> diphenhydramine (Benedryl) | <input type="checkbox"/> meperidine (Demerol) |

- | | |
|---|---|
| <input type="checkbox"/> morphine | <input type="checkbox"/> neostigmine (Prostigmin) |
| <input type="checkbox"/> remifentanyl (Ultiva) | <input type="checkbox"/> physostigmine (Antilirium) |
| <input type="checkbox"/> sufentanil (Sufenta) | <input type="checkbox"/> pyridostigmine (Mestinon) |
| <input type="checkbox"/> hydromorphone (Dilaudid) | |
| <input type="checkbox"/> unknown | <input type="checkbox"/> bupivacaine (Marcaine) |
| <input type="checkbox"/> nitrous oxide | <input type="checkbox"/> levo-bupivacaine |
| <input type="checkbox"/> flumazenil (Romazicon) | <input type="checkbox"/> chloroprocaine (Nesacaine) |
| <input type="checkbox"/> nalbuphine (Nubain) | <input type="checkbox"/> cocaine |
| <input type="checkbox"/> naloxone (Narcan) | <input type="checkbox"/> etidocaine (Duranest) |
| | <input type="checkbox"/> lidocaine (Xylocaine) |
| <input type="checkbox"/> atracurium (Tracrium) | <input type="checkbox"/> mepivacaine (Carbocaine) |
| <input type="checkbox"/> cisatracurium (Nimbex) | <input type="checkbox"/> prilocaine (Citanest) |
| <input type="checkbox"/> rocuronium (Zemuron) | <input type="checkbox"/> procaine (Novocain) |
| <input type="checkbox"/> vecuronium (Norcuron) () | <input type="checkbox"/> ropivacaine (Naropin) |
| pancuronium (Pavulon) | <input type="checkbox"/> tetracaine (Pontocaine) |
| <input type="checkbox"/> other NMB | |
| <input type="checkbox"/> IM succinylcholine (Anectine) | <input type="checkbox"/> epinephrine |
| <input type="checkbox"/> IV succinylcholine (Anectine) | <input type="checkbox"/> ephedrine |
| <input type="checkbox"/> NO succinylcholine | <input type="checkbox"/> neosynephrine |
| <input type="checkbox"/> edrophonium (Tensilon) | |
| <input type="checkbox"/> other (<i>specify</i>): _____ | |

- 46a. Type of anesthetic used for biopsy:
check all applicable
- monitored anesthesia care (local standby)
 - regional anesthesia
 - spinal anesthesia
 - epidural anesthesia
 - general anesthesia **without** endotracheal intubation
 - general anesthesia **with** endotracheal intubation
 - general anesthesia with a face mask
 - general anesthesia with a laryngeal mask airway

HISTOLOGY

47. Was muscle histology performed?
check one
- yes
 - no
- If no, skip to question 48*
48. The muscle histology result was:
check one
- normal
 - abnormal
 - equivocal
- If normal, skip to question 48*
49. What were the abnormal histologic findings?
check one
- diffusely distributed internal nuclei
 - other (*specify abnormality, write pending if results not available*)
-
50. Was muscle histochemistry performed?
check one
- yes
 - no
- If no, skip to question 51*
51. The muscle histochemistry result was:
check one
- normal
 - abnormal
 - equivocal
- If normal, skip to question 51*

52. Specify results of muscle histochemistry:
check one
 moth-eaten fibers
 cores
 other (*specify abnormality, write pending if results not available*)
-

CONTRACTURE TESTS

53. In your lab, when muscle is exposed to 3% halothane, what is the minimum contracture indicating MH susceptibility?
0. ____ ____ grams
54. To date, how many control patients has this lab evaluated with the 1989 Biopsy Standards protocol? ____ ____ ____ ____

55. MH Diagnostic Muscle Biopsy Results

check one

- positive -- MH susceptible
- negative -- not susceptible to MH
- equivocal -- MH susceptibility not determined
- control biopsy

56. Contracture Test Results

TENSION IN GRAMS MEASURED FROM ZERO OF MEASURING SCALE

| HALOTHANE AT 3% (<i>Required</i>): | <u>Strip 1</u> | <u>Strip 2</u> | <u>Strip 3</u> |
|--|------------------------------|------------------------------|------------------------------|
| Hours between excision to completion of test (h)..... | __ . __ __ | __ . __ __ | __ . __ __ |
| Stimulation: | | | |
| duration (milliseconds)..... | __ | __ | __ |
| frequency (Hz)..... | __ . __ | __ . __ | __ . __ |
| voltage (volts) | __ . __ | __ . __ | __ . __ |
| current (mA) | __ | __ | __ |
| Was a length/tension curve done? <i>check one</i> | <input type="checkbox"/> no | <input type="checkbox"/> no | <input type="checkbox"/> no |
| | <input type="checkbox"/> yes | <input type="checkbox"/> yes | <input type="checkbox"/> yes |
| Pre-drug twitch tension (g)..... <i>(measure from baseline for twitch tension only)</i> | __ . __ __ | __ . __ __ | __ . __ __ |
| Pre-drug tension 3% hal(g)..... | __ . __ __ | __ . __ __ | __ . __ __ |
| Low point tension 3% hal(g)..... | __ . __ __ | __ . __ __ | __ . __ __ |
| Contracture tension developed to 3% hal(g) | __ . __ __ | __ . __ __ | __ . __ __ |
| Do you consider the tension developed to be abnormal? <i>check one</i> | <input type="checkbox"/> no | <input type="checkbox"/> no | <input type="checkbox"/> no |
| | <input type="checkbox"/> yes | <input type="checkbox"/> yes | <input type="checkbox"/> yes |
| Length (cm)..... | __ . __ __ | __ . __ __ | __ . __ __ |
| Wet Weight (g)..... | . __ __ __ | . __ __ __ | . __ __ __ |

TENSION IN GRAMS MEASURED FROM ZERO OF
MEASURING SCALE

| | <u>Strip 1</u> | <u>Strip 2</u> | <u>Strip 3</u> |
|--|------------------------------|------------------------------|------------------------------|
| CAFFEINE ALONE (<i>Required</i>): | | | |
| Hours between excision to completion of test (h)..... | _ . _ _ | _ . _ _ | _ . _ _ |
| Stimulation: | | | |
| duration (milliseconds)..... | _ _ | _ _ | _ _ |
| frequency (Hz)..... | _ . _ | _ . _ | _ . _ |
| voltage (volts) | _ _ . _ | _ _ . _ | _ _ . _ |
| current (mA) | _ _ | _ _ | _ _ |
| Was a length/tension curve done? | | | |
| <i>check one</i> | <input type="checkbox"/> no | <input type="checkbox"/> no | <input type="checkbox"/> no |
| | <input type="checkbox"/> yes | <input type="checkbox"/> yes | <input type="checkbox"/> yes |
| Pre-drug twitch tension (g)..... (<i>measure from baseline for twitch tension only</i>) | _ _ . _ _ | _ _ . _ _ | _ _ . _ _ |
| Predrug tension 0.5mM (g)..... | _ _ . _ _ | _ _ . _ _ | _ _ . _ _ |
| Plateau tension 0.5mM (g)..... | _ _ . _ _ | _ _ . _ _ | _ _ . _ _ |
| Predrug tension 1.0mM (g)..... | _ _ . _ _ | _ _ . _ _ | _ _ . _ _ |
| Plateau tension 1.0mM (g)..... | _ _ . _ _ | _ _ . _ _ | _ _ . _ _ |
| Predrug tension 2.0mM (g)..... | _ _ . _ _ | _ _ . _ _ | _ _ . _ _ |
| Plateau tension 2.0mM (g)..... | _ _ . _ _ | _ _ . _ _ | _ _ . _ _ |
| Predrug tension 4.0mM (g)..... | _ _ . _ _ | _ _ . _ _ | _ _ . _ _ |
| Plateau tension 4.0mM (g)..... | _ _ . _ _ | _ _ . _ _ | _ _ . _ _ |
| Predrug tension 8.0mM (g)..... | _ _ . _ _ | _ _ . _ _ | _ _ . _ _ |
| Plateau tension 8.0mM (g)..... | _ _ . _ _ | _ _ . _ _ | _ _ . _ _ |
| Predrug tension 32.0mM (g)..... | _ _ . _ _ | _ _ . _ _ | _ _ . _ _ |
| Plateau tension 32.0mM (g)..... | _ _ . _ _ | _ _ . _ _ | _ _ . _ _ |
| CSC (mM)..... | _ _ . _ _ | _ _ . _ _ | _ _ . _ _ |
| % response at 2mM..... | _ _ . _ _ | _ _ . _ _ | _ _ . _ _ |

TENSION IN GRAMS MEASURED FROM ZERO OF
MEASURING SCALE

| | <u>Strip 1</u> | <u>Strip 2</u> | <u>Strip 3</u> |
|--|----------------|----------------|----------------|
| Do you consider the tension developed to be abnormal? <i>check one</i> | () no | () no | () no |
| | () yes | () yes | () yes |
| If yes, at what concentration? | ___ | ___ | ___ |
| If yes, at what CSC? | ___ | ___ | ___ |
| If yes, at what % response? | ___ | ___ | ___ |
| Length (cm)..... | __ . ___ | __ . ___ | __ . ___ |
| Wet Weight (g)..... | . ___ | . ___ | . ___ |
| HALOTHANE 1% & CAFFEINE (<i>Optional</i>): | <u>Strip 1</u> | <u>Strip 2</u> | <u>Strip 3</u> |
| Hours between excision to completion of test (h)..... | __ . ___ | __ . ___ | __ . ___ |
| Stimulation: | | | |
| duration (milliseconds)..... | __ | __ | __ |
| frequency (Hz)..... | __ . __ | __ . __ | __ . __ |
| voltage (volts) | ___ . __ | ___ . __ | ___ . __ |
| current (mA) | ___ | ___ | ___ |
| Was a length/tension curve done? <i>check one</i> | () no | () no | () no |
| | () yes | () yes | () yes |
| Pre-drug twitch tension (g)..... (<i>measure from baseline for twitch tension only</i>) | ___ . ___ | ___ . ___ | ___ . ___ |
| Pre-drug tension 1% hal(g)..... | ___ . ___ | ___ . ___ | ___ . ___ |
| Low point tension 1% hal(g)..... | ___ . ___ | ___ . ___ | ___ . ___ |
| Contracture tension developed to 1% hal(g).. | ___ . ___ | ___ . ___ | ___ . ___ |
| Predrug tension 0.25mM (g)..... | ___ . ___ | ___ . ___ | ___ . ___ |
| Plateau tension 0.25mM (g)..... | ___ . ___ | ___ . ___ | ___ . ___ |
| Predrug tension 0.5mM (g)..... | ___ . ___ | ___ . ___ | ___ . ___ |
| Plateau tension 0.5mM (g)..... | ___ . ___ | ___ . ___ | ___ . ___ |

TENSION IN GRAMS MEASURED FROM ZERO OF
MEASURING SCALE

| | <u>Strip 1</u> | <u>Strip 2</u> | <u>Strip 3</u> |
|--|----------------|----------------|----------------|
| Predrug tension 1.0mM (g)..... | ___ . ___ | ___ . ___ | ___ . ___ |
| Plateau tension 1.0mM (g)..... | ___ . ___ | ___ . ___ | ___ . ___ |
| Predrug tension 2.0mM (g)..... | ___ . ___ | ___ . ___ | ___ . ___ |
| Plateau tension 2.0mM (g)..... | ___ . ___ | ___ . ___ | ___ . ___ |
| Predrug tension 4.0mM (g)..... | ___ . ___ | ___ . ___ | ___ . ___ |
| Plateau tension 4.0mM (g)..... | ___ . ___ | ___ . ___ | ___ . ___ |
| Predrug tension 32.0mM (g)..... | ___ . ___ | ___ . ___ | ___ . ___ |
| Plateau tension 32.0mM (g)..... | ___ . ___ | ___ . ___ | ___ . ___ |
| HCSC (mM)..... | ___ . ___ | ___ . ___ | ___ . ___ |
| Do you consider the tension developed to be abnormal? <i>check one</i> | () no | () no | () no |
| | () yes | () yes | () yes |
| If yes, at what concentration? | ___ | ___ | ___ |
| If yes, at what HCSC? | ___ | ___ | ___ |
| Length (cm)..... | ___ . ___ | ___ . ___ | ___ . ___ |
| Wet Weight (g)..... | ___ . ___ | ___ . ___ | ___ . ___ |
| HALOTHANE AT 2% (Optional): | <u>Strip 1</u> | <u>Strip 2</u> | <u>Strip 3</u> |
| Hours between excision to completion of test (h)..... | ___ . ___ | ___ . ___ | ___ . ___ |
| Stimulation: | | | |
| duration (milliseconds)..... | ___ | ___ | ___ |
| frequency (Hz)..... | ___ . ___ | ___ . ___ | ___ . ___ |
| voltage (volts) _____ | ___ . ___ | ___ . ___ | ___ . ___ |
| current (mA) _____ | ___ | ___ | ___ |
| Was a length/tension curve done? <i>check one</i> | () no | () no | () no |
| | () yes | () yes | () yes |
| Pre-drug twitch tension (g)..... (<i>measure from baseline for twitch tension only</i>) | ___ . ___ | ___ . ___ | ___ . ___ |

TENSION IN GRAMS MEASURED FROM ZERO OF
MEASURING SCALE

| | <u>Strip 1</u> | <u>Strip 2</u> | <u>Strip 3</u> |
|--|-------------------|-------------------|-------------------|
| Pre-drug tension 2% hal(g)..... | ___ . ___ | ___ . ___ | ___ . ___ |
| Low point tension 2% hal(g)..... | ___ . ___ | ___ . ___ | ___ . ___ |
| Contracture tension developed to 2% hal(g).. | ___ . ___ | ___ . ___ | ___ . ___ |
| Do you consider the tension developed to be abnormal? <i>check one</i> | () no () yes | () no () yes | () no () yes |
| Length (cm)..... | __ . ___ | __ . ___ | __ . ___ |
| Wet Weight (g)..... | . ___ | . ___ | . ___ |

TISSUE AND BLOOD STORAGE

57. Has additional muscle tissue been stored?

check one

() yes

() no

If yes, specify Sample ID No: _____

Location _____

58. Has an additional blood specimen been stored?

check one

() yes

() no

If yes, specify Sample ID No: _____

DNA TESTING

59. Was a genetic exam performed?

check one

- yes unknown
 no

60. Where was the genetic test done?

60a. Is a sample of the DNA stored in the lab?

- yes
 no

61. When was the genetic test done?

62. Which of the RYR1 exons were examined?

_____ _____
_____ _____
_____ _____

If unknown, check here

63. Was any mutation associated with MH or central core disease present?

check one

- yes unknown
 no

If yes, specify _____

64. Were any other sequence variants identified?

check one

- yes
 no

If yes, specify _____

COMMENTS ON PATIENT (Optional)

Please mail original to:

The North American Malignant Hyperthermia Registry
University of Florida
Department of Anesthesiology
1600 SW Archer Road, PO Box 100254
Gainesville, FL 32610

MH BIOPSY CENTER CODE NUMBERS

| | |
|--|----|
| Wake Forest University..... | 06 |
| Toronto General Hospital | 05 |
| University of California at Davis..... | 07 |
| Uniformed Services University..... | 16 |
| University of Minnesota..... | 24 |