The North American Malignant Hyperthermia Registry

Report of Malignant Hyperthermia Research Subject

Already Known As

Malignant Hyperthermia Susceptible

("AKA Report")

INSTRUCTIONS:
This form is to be filled out by the person to be registered and their anesthesiologist/health care provider.

1. To register your name with the North American Malignant Hyperthermia Registry, sign the consent form for release of information by you and your physician to the North American Malignant Hyperthermia Registry and for release of information by the North American Malignant Hyperthermia Registry to your future physicians. If both parents of a child who experienced an episode of MH wish to be registered, then separate consent forms must be signed for each parent.

2. You can answer all questions except possibly 17-20 and 29-40. You may need to consult with your anesthesiologist or other physician responsible for diagnosing you as MH susceptible for assistance.

3. Send this AKA report to the anesthesiologist or other physician responsible for diagnosing you as MH susceptible. Please ask this physician to complete the rest of this form (questions 17-20 and 29-40). If this physician is not available, fill out as much of the form as you can.

4. Send this form and all signed consent forms directly to the North American MH Registry (address at bottom of this page). You will need to call the NAMHR office at (888) 274-7899 and speak to Dr. Brandom or one of the co-investigators to confirm your consent by conversation over the phone. Each person who signed a consent form will need to call the North American MH Registry to confirm that consent by conversation on the telephone as well.

5. Information sent to the North American MH Registry (NAMHR) will remain confidential. Patient specific information will only be accessible by people specifically designated by the research subject.

Return original completed form to:

The North American Malignant Hyperthermia Registry
UPMC Mercy Hospital
8th Floor, Ermire Building (B)
Room 8522-3
1400 Locust Street
Pittsburgh, PA 15219
1-888-274-7899

AKA MH REPORT

(Version 7.5 May 2014)

PATIENT IDENTIFICATION *(to be completed by the research subject or subject's guardian)*

1. Name (this is the index research subject)

   _____ _____ _____
   last first middle

2. Previous name

   _____ _____ _____
   last first middle

3. Maiden name

   _____
   last

4. Address

   _____
   street address

   _____ _____ _____
   city state/province zip/postal code

   _____
   country

5. Phone number

   (_____) _____ - _____ *home*
   (_____) _____ - _____ *work*

6. E-mail address _____

DEMOGRAPHIC INFORMATION *(to be completed by the subject or subject's guardian)*

7. Sex

check one

( ) male

( ) female

8. Weight at the time of your MH episode:

____.__ kilograms OR ____ lbs

9. Height at the time of your MH episode:

_____ cm OR ____ ft ___ inches

10. Date of subject's birth

___ ___ ___ ___ \ ___ ___ \ ___ ___

year month day

11. Race:

check as many as apply

(data utilized for demographic purposes only)

( ) Caucasian
( ) Hispanic
( ) African-American
( ) Native American
( ) Hawaiian or Pacific Islander
( ) other *(specify):*_____

( ) African
( ) East Asian
( ) South Asian
( ) Middle Eastern

12. Any previous North American MH Registry numbers associated with the subject. That is, AMRA, AKA, close relative's reports, etc.

a. ____ ____ ____ ____ ____ Comment _____

b. ____ ____ ____ ____ ____ Comment _____

c. ____ ____ ____ ____ ____ Comment _____

FAMILY IDENTIFICATION *(to be completed by the research subject or subject's guardian)*

13. Does the 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 research subject?
check one
( ) child
( ) sibling

b. **name**

_____ _____ _____
last first middle

Date of birth
__ __ __ __ \ __ __ \ __ __
year month day

Is this the child or the sibling of the research subject?
check one
( ) child
( ) sibling

c. **name**

_____ _____ _____
last first middle

Date of birth
__ __ __ __ \ __ __ \ __ __
year month day

Is this the child or the sibling of the research subject?
check one
( ) child
( ) sibling

d. **name**

_____ _____ _____
last first middle

Date of birth
___ ___ ___ ___ \ ___ ___ \ ___ ___
year month day

Is this the child or the sibling of the research subject?
check one
( ) child
( ) sibling

e. **name**

_____ _____ _____
last first middle

Date of birth
___ ___ ___ ___ \ ___ ___ \ ___ ___
year month day

Is this the child or the sibling of the research subject?
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 index research subject 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 index research subject?
check one
( ) child
( ) sibling

b. **name**

_____ _____ _____
last first middle

Date of birth
__ __ __ __ \ __ __ \ __ __
year month day
Is this the child or the sibling of the index research subject?
check one
( ) child
( ) sibling

c. **name**

_____ _____ _____
last first middle

Date of birth
__ __ __ __ \ __ __ \ __ __
year month day
Is this the child or the sibling of the index research subject?
check one
( ) child
( ) sibling

d. **name**

_____ _____ _____
last first middle

Date of birth
___ ___ ___ ___ \ ___ ___ \ ___ ___
year month day

Is this the child or the sibling of the index research subject?
check one
( ) child
( ) sibling

15. Has consent been obtained to enter the names of the parents of the research subject? If the index research subject is your child, your information goes in this section.
check one
( ) yes
( ) no
If yes, complete below

<u>NOTE:</u> CONSENT MUST BE OFTAINED 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 the index research subject

_____ _____ _____
last first middle

Date of mother's birth
___ ___ ___ ___ \ ___ ___ \ ___ ___
year month day

Mother's maiden name

_____
last

b. Father of the index research subject

_____ _____ _____
last first middle

Date of father's birth
___ ___ ___ ___ \ ___ ___ \ ___ ___
year month day

16. Family History Table

Key to Family History table (below)

Relationship to Subject	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. none of the above
s. paternal – other	17. unknown
t. relative by marriage	
u. other blood relative	

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

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

PHYSICIAN'S IDENTIFICATION *(to be completed by the physician)*
Optional-for Registry use only

17. Anesthesiologist's or other physician's name

_____ _____ _____
last first middle

18. Hospital name

_____

19. Hospital Address

_____
street address

_____ _____ _____
city state/province zip/postal code

_____
country

20. Physician's office telephone number
(__ __ __) - __ __ __ - __ __ __ __

FAMILY HISTORY

21. Family history is 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
( ) none of the above
( ) unknown
( ) other *(specify)*_____ .

MEDICAL HISTORY

22. Does the subject have any of the following?
check all applicable
( ) muscle weakness interferes with daily activity at least once/week
( ) muscle cramps or pain interfere with daily activity at least once/week
( ) cola colored urine
( ) heat stroke or heat prostration
( ) oral (or rectal/axillary equivalent) fever>38.6°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
( ) none of the above
( ) unknown
( ) other *(specify):*_____

23. Has the subject 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

24. How many times was this subject anesthetized prior to this evaluation?

__ __

( ) unknown

Skip to question 28 if the response is zero or unknown.

25. How many were general anesthetics?

__ __

( ) unknown

26. Indicate the number of anesthetics with the following agents:

__ __ volatile agents without succinylcholine
__ __ volatile agents with succinylcholine
__ __ succinylcholine without other known triggering agents

27. Subjects's anesthetic history is positive for:

check all applicable

( ) clear-cut clinical MH episode(s)
( ) possible MH (not clear-cut MH)
( ) masseter muscle rigidity only
( ) delayed awakening from general anesthesia
( ) positive caffeine halothane contracture test
( ) positive calcium uptake test (performed in Boston)
( ) other *(specify):* _____
( ) none of the above
( ) unknown

28a. Date of possible or clear-cut MH episode
answer for anesthetic most suspect for MH

__ __ __ __ \ __ __ \ __ __
year month day

28b. Year of most recent anesthetic (excluding present episode).

__ __ __ __
year

( ) unknown

29. Pre-medication and anesthetic agents utilized during possible /clear cut MH:
check all applicable

( ) sodium citrated citric acid (Bicitra)
( ) cimetidine (Tagamet)
( ) famotidine (Pepcid)
( ) lansoprazole (Prevacid)
( ) ranitidine (Zantac)

( ) metoclopramide (Reglan)
( ) omeprazole (Prilosec)

( ) atropine
( ) glycopyrrolate (Robinul)
( ) scopolamine (Hyoscine)

( ) dolasetron (Anzemet)
( ) droperidol (Inapsine)
( ) hydroxyzine (Vistaril)
( ) ondansetron (Zofran)
( ) promethazine (Phenergan)
( ) diphenhydramine (Benedryl)

( ) clonidine (Duraclon)
( ) 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 anesthetics*
( ) other *(specify):*_____

( ) sevoflurane (Ultane)
( ) desflurane (Suprane)
( ) isoflurane (Forane)
( ) nitrous oxide

( ) 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)
( ) pyridostigmine (Mestinon)

( ) bupivacaine (Marcaine)
( ) levo-bupivacaine
( ) choroprocaine (Nesacaine)
( ) cocaine
( ) etidocaine (Duranest)
( ) lidocaine (Xylocaine)
( ) mepivacaine (Carbocaine)
( ) prilocaine (Citanest)
( ) procaine (Novocain)
( ) ropivacaine (Naropin)
( ) tetracaine (Pontocaine)

( ) epinephrine
( ) ephedrine
( ) neosynephrine

30. Signs and abnormal findings during possible or fulminant MH

Abnormal signs noted 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 direct laryngoscopy is possible
____ masseter spasm: jaw clamped shut, intubation by direct visualization impossible
____ generalized muscular rigidity
____ cola colored urine
____ tachypnea
____ hypercarbia
____ cyanosis
____ sinus tachycardia
____ ventricular tachycardia
____ ventricular fibrillation
____ elevated temperature
____ rapidly increasing temperature
____ sweating
____ excessive bleeding
____ skin mottling
____ hypertension > 20% baseline
____ other *(specify):* _____
____ none of the above

31. Abnormal metabolic values during possible or fulminant MH

Most abnormal arterial blood gas after MH was suspected:

__.__ __ FiO_2
__.__ __ pH
__ __ __ PCO_2
__ __ __ PO_2
__ __.__ BE (mEq/L) (specify ±)
__ __ Bicarbonate (mEq/L)
__ __ __ time (hours after 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)**

32. Treatment given for possible or fulminant MH

check all treatments utilized

( ) Volatile anesthetics discontinued at time: __ __ __ (hours after induction)
( ) Hyperventilation with 100% oxygen
( ) Dantrolene (*type*)
  ( ) Dantrium
  ( ) Revonto
  ( ) Ryanodex
  __ __ __.__ Initial dose (mg)
  __ __ __ Time of first dose (hours after induction)
  __ __ __ __.__ Total dose (mg)
  __ __ __ Time of last dose (Hours after anesthetic induction)
( ) Active cooling
( ) Fluid loading
( ) Furosemide
( ) Mannitol
( ) Bicarbonate
( ) Glucose, insulin
( ) Bretylium
( ) Amrinone
( ) Vasopressor
( ) Lidocaine
( ) Procainamide
( ) Defibrillation
( ) CPR
( ) other *(specify):*_____
( ) none of the above
( ) unknown

33. Were any problems noted with the dantrolene administration?

check one

( ) yes
( ) no
( ) unknown

If no, please skip to question 35

34. What were the observed dantrolene complications?

check all applicable

( ) phlebitis
( ) excessive secretions
( ) gastrointestinal upset
( ) hyperkalemia
( ) muscle weakness
( ) respiratory failure
( ) other *specify*:_____

DNA TESTING (*to be completed by the physician)*

35. Was a genetic test performed?
check one
( ) yes
( ) no

36. Where was the genetic test done?

_____

36a. Is a sample of the DNA stored in the lab?
( ) yes
( ) no

37. When was the genetic test done?

_____

38. Which of the RYR1 exons were examined?

_____ _____
_____ _____
_____ _____

39. Was any mutation associated with MH or central core disease present?
check one
( ) yes
( ) no
*If yes, specify:*_____

40a. Were any other sequence variants identified?
check one
( ) yes
( ) no
*If yes, specify:*_____

40b. Did the subject survive the initial reaction?
check one
( ) yes ( ) unknown because of transfer of case during treatment
( ) no

40b. Did the subject survive any subsequent reaction (recrudescence) and recovery?
check one
( ) yes ( ) unknown because of transfer of case during treatment
( ) no

MH DIAGNOSTIC MUSCLE BIOPSY

Answer for caffeine halothane contracture test or European IVCT test only. These tests are only done at MH Biopsy centers, and are different from regular pathology biopsies.

41. Date of diagnostic muscle biopsy

___ ___ ___ ___ \ ___ ___ \ ___ ___
year month day

42. Results

check one

( ) positive—MH susceptible
( ) negative—not susceptible to MH
( ) equivocal—MH susceptibility indeterminate

43. Center which performed MH Biopsy (Caffeine Halothane Contracture Test)

check one

( ) Children's Hospital of Oklahoma
( ) Cleveland Clinic
( ) Hahnemann University
( ) Thomas Jefferson University
( ) Loyola University
( ) Northwestern University
( ) Mayo Clinic
( ) Ottawa Hospital- Civic Campus
( ) Presbyterian University Hospital (Pittsburgh)
( ) Toronto General Hospital
( ) UC-Davis
( ) UCLA
( ) Uniformed Services University
( ) University of Calgary
( ) University of Florida
( ) University of Iowa
( ) University of Manitoba
( ) University of Massachusetts
( ) University of Nebraska
( ) University of South Florida
( ) University of Texas-Houston
( ) University of Texas Medical Branch
( ) University of Washington
( ) University of Wisconsin
( ) Wake Forest University
( ) other *(specify):*_____

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

_____
(Signature of subject submitting this report)

___ ___ ___ ___ \ ___ ___ \ ___ ___
year month day

COMMENTS ON SUBJECT
Optional

_____
_____
_____
_____
_____
_____