



Malignant Hyperthermia

“Hot Off the Press”

2014 PGA, NYC



Ron Litman

litmanr@email.chop.edu

Relevant Disclosures and Conflicts of Interests



*Dedicated to
Patient Safety*



MHAUS Guidelines Changed: New Recommendations for Preparation & Treatment

By DAVID KIRKMAN

Editorial

WALSHVILLE, Ill., July 1 — Since Farley made headlines, the only job the state had should be to clean off the dirt and have it sent on at longer than 10 minutes, otherwise just ignore it and open up the right eye.

"Damaged goods," Mr. Farley describes himself, meaning how the recently purchased is what happened to his mother about whether the "sprayed body" was much about.

For about five years, Mr. Farley,

46, used strongest what a

few other workers,

spray gun

paint,

glue together

boards

or chairs and tables

and other stuff.

He

hired,

HENRY ROSENBERG

2015 ASA DISTINGUISHED SERVICE AWARD WINNER

By DAVID KIRKMAN

DETROIT, Mich. — Henry Farley makes a living. The only job the 60-year-old could be sure of when he started out here is to stand on an integer that is 10 minutes, otherwise pure, unbroken, unbroken speech and right logic.

"Distinguished goods," Mr. Farley says when I meet him, meaning how the recently announced a chief competitor in his industry about whether the "supplied body" has much about.

For about five years, Mr. Farley

has worked alongside about a dozen other workers, spray gun

hand, glue together their

skins for chairs and couches

and other small items like

chairs, lamps, lamps and

chairs. Farley, from the

time he started a pension fund in

the plant, and Mr. Farley's

colleagues say that knowing there

is eventually the way of his

industry's endgame, resulting in what

the and his workers call

"the end."

A chemical she handled —

isomers or isopropyl bromide, or iPB — is also used by tens of thousands of workers in auto body shops, dry cleaners and high-tech electronic manufacturing plants across the nation. Medical researchers, government officials and some chemical companies that have monitored iPB have learned for over a decade that it causes neurological damage and affecting when inhaled or even hours over long periods, but the evidence grows thinner in the past six years.

Most recently demonstrating the difficulty, though, is a series of all but, of showing that Americans can breathe clean air in the following as workers after surface fall of months from the Occupational Safety and Health Administration that shows that managers at the auto plant, Detroit, where Mr. Farley was employed repeatedly exposed others to iPB levels that exceeded federal safety standards, failed to provide respirators and turned off the plant to save money.

But the story of the use of iPB and the decline of Mr. Farley's health is much more than the fate of one company, or another chapter in the ongoing debate over the need for more, or better, governmental regulation. Instead, it is a parable about the law of unintended consequences.

It starts here at Environmental Protection Agency, program



American Society of
Anesthesiologists®



ECONOMIC FEARS HAMPER

Government Is Pressed
to Increase Taxes and
Cut Subsidies

By DAVID B. KIRKMAN

CAIRO, Egypt — A fuel shortage has helped send food prices soaring. Electricity is stacking out more before the summer. And gasoline gaslights have killed at least five people and injured dozens over the past two weeks.

The root of the crisis, economists say is that Egypt is running out of the fuel refining it needs for fuel imports. The shortage is raising questions about Egypt's ability to keep importing what it needs to make refined fuel supplies, offering hints of an economic catastrophe at a time when the government is already struggling to quell inflation pressure by the political crisis.

Egyptians already took fuel for the journey that complete their diets, and they say they fear they will not have enough for the months to map their usual fuel purchases to the fuel.

United States officials were of



MHAUS Hotline: 800-MH-HYPER



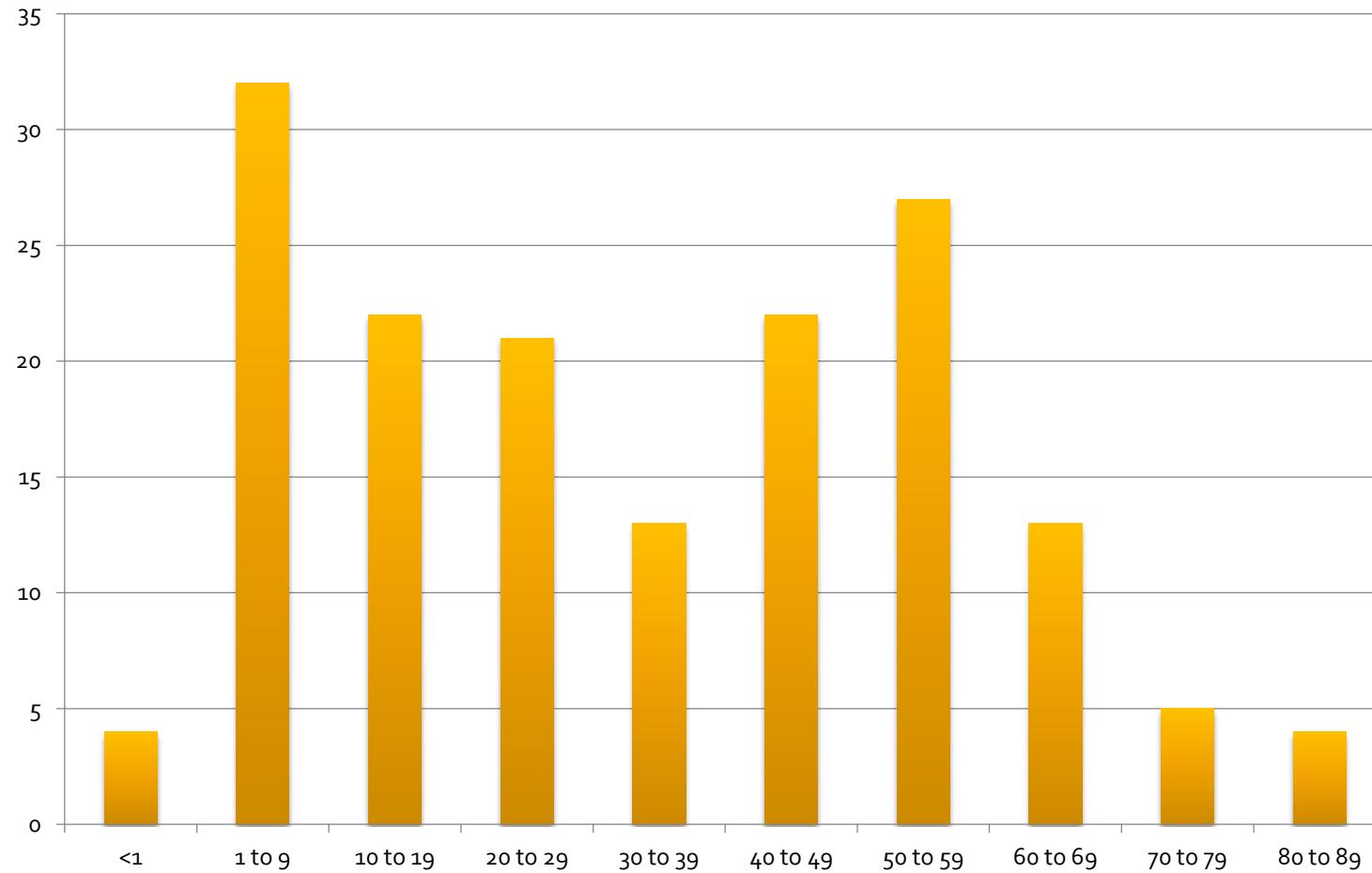
MHAUS Hotline Stats 2014

- 31 Hotline consultants
- > 400 calls thus far 2014
- > 200 website queries



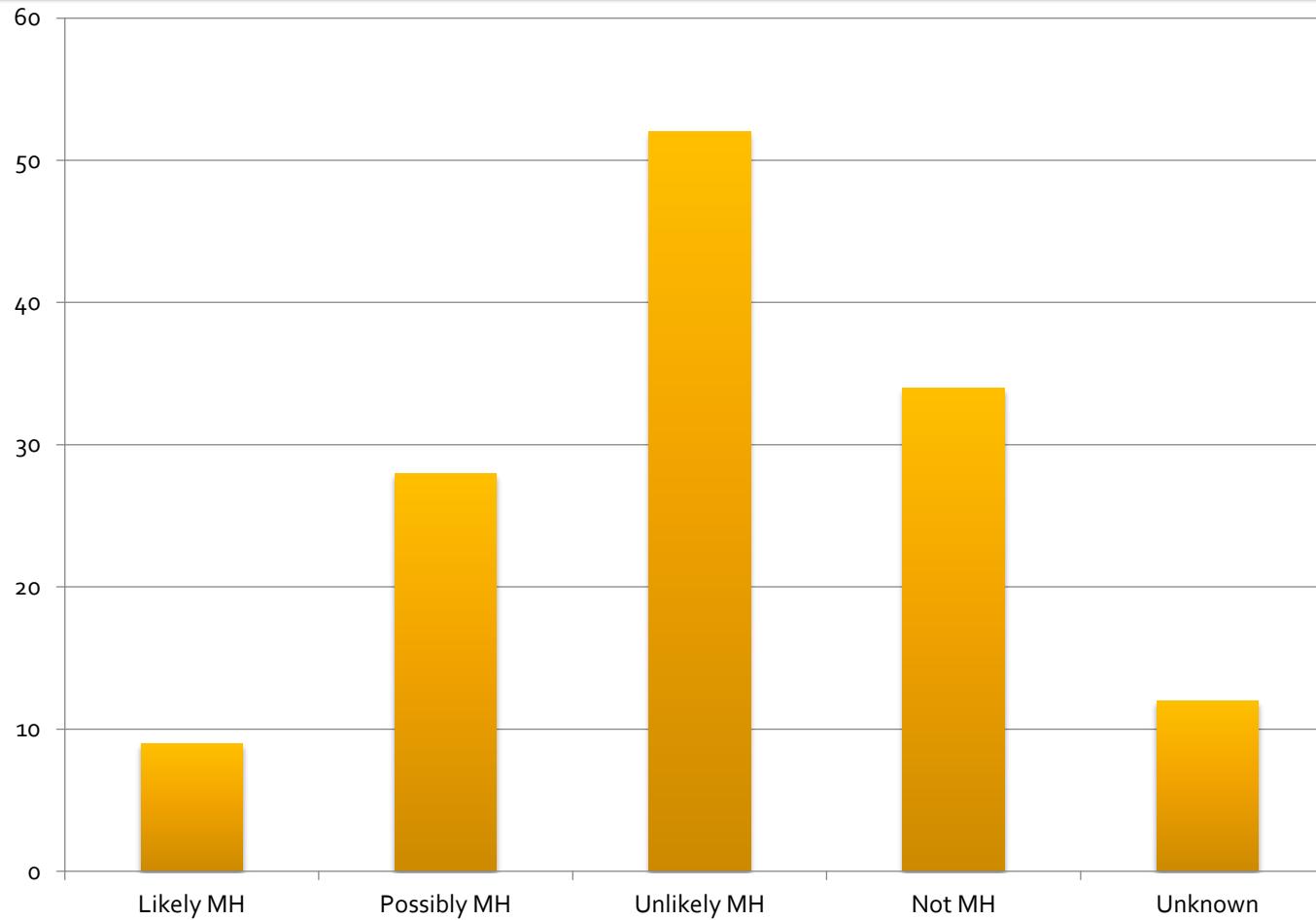
MHAUS Hotline Stats 2014

Ages of pts



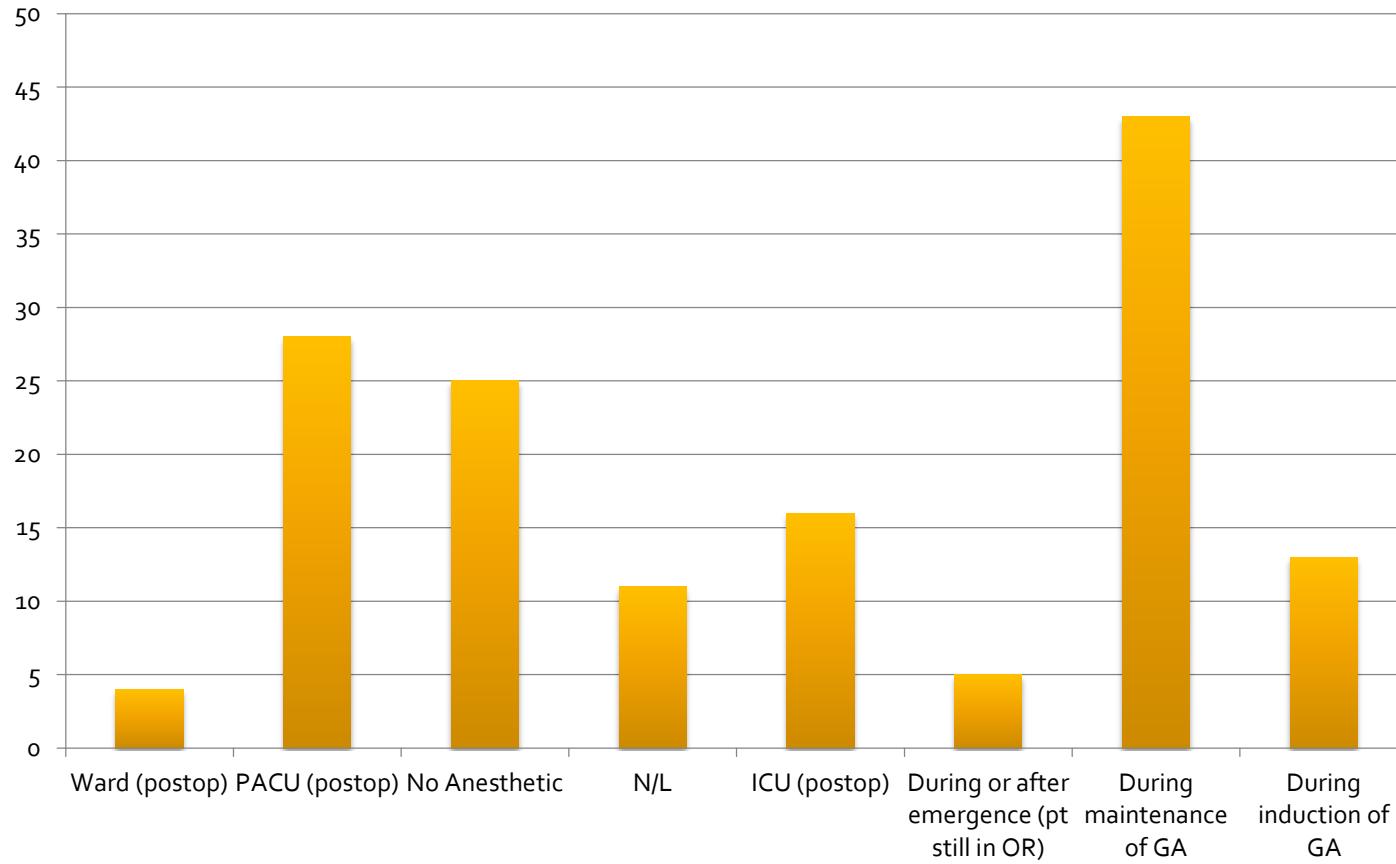
MHAUS Hotline Stats 2014

Diagnoses

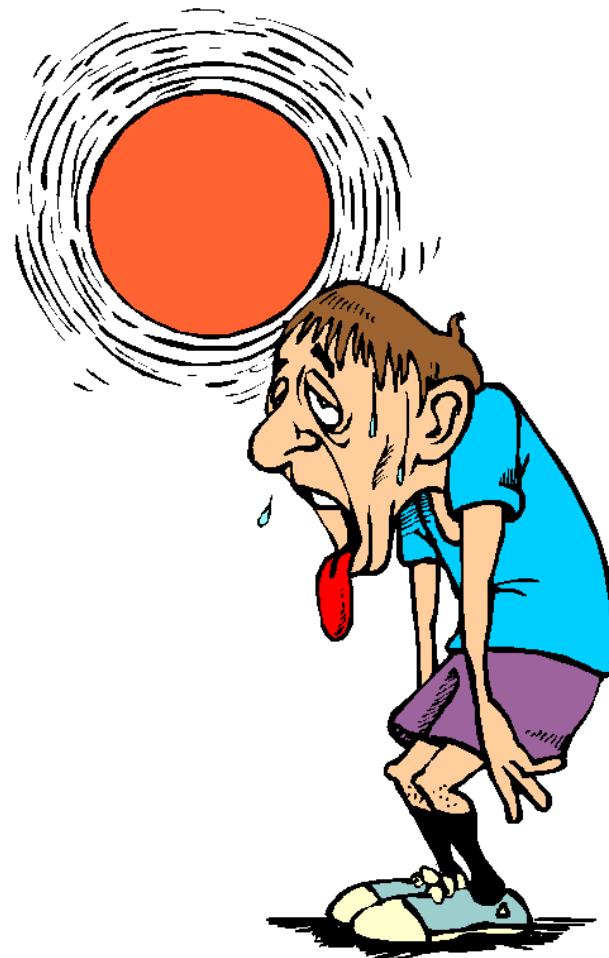


MHAUS Hotline Stats 2014

Location Episode Began



Important MH Principles



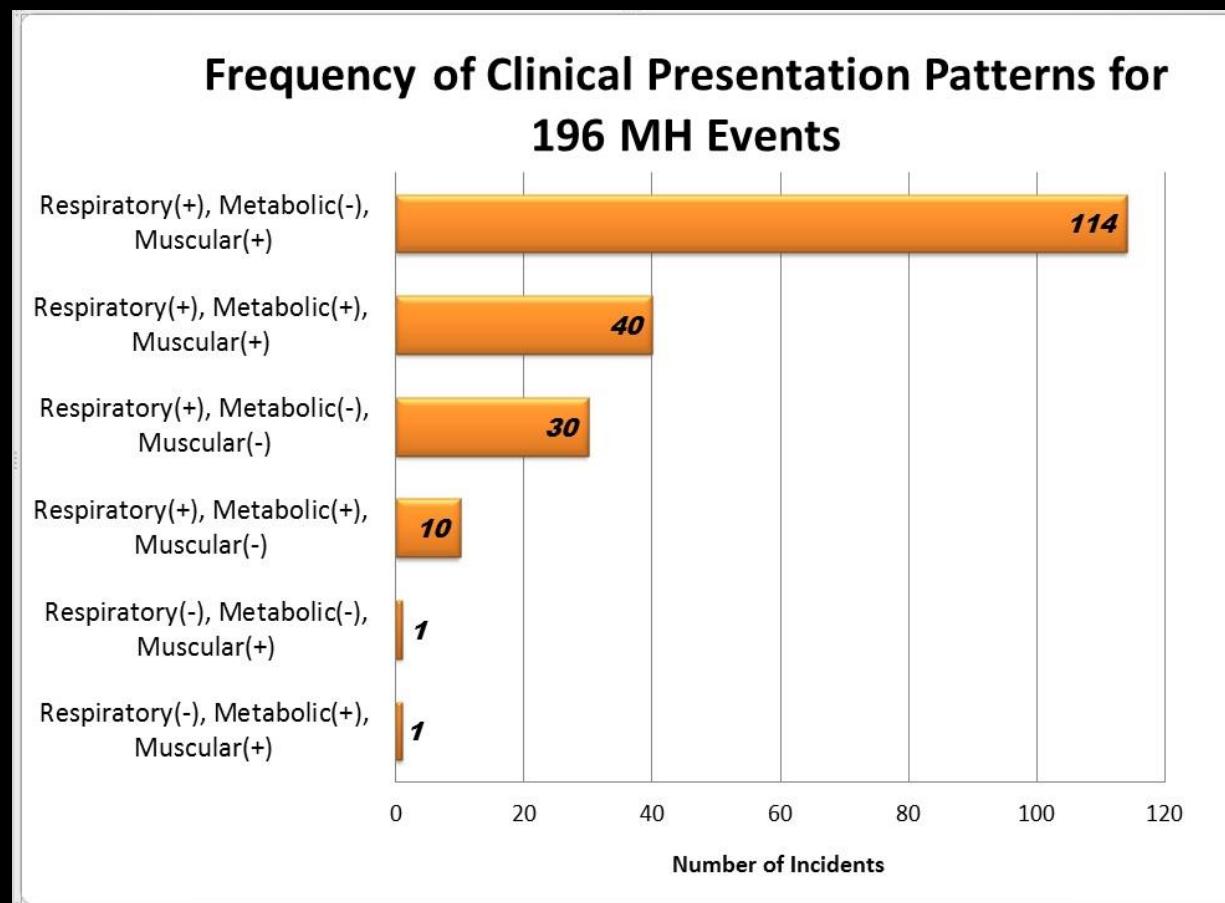
Clinical News

Clinical Presentation, Treatment, and Complications of Malignant Hyperthermia in North America from 1987 to 2006

Table 1. Anesthetic Drug Triggers for 284 Malignant Hyperthermia Events

Anesthetic drug	Frequency	%
+Succinylcholine, -Volatile	2	0.7
+Succinylcholine, +Volatile	153	53.9
-Succinylcholine, +Volatile	128	45.1
-Succinylcholine, -Volatile ^a	1	0.4

Clinical Presentation, Treatment, and Complications of Malignant Hyperthermia in North America from 1987 to 2006



Malignant Hyperthermia in Canada: Characteristics of Index Anesthetics in 129 Malignant Hyperthermia Susceptible Probands

January 2014

Sheila Riazi, MSc, MD,*† Marilyn Green Larach, MD, FAAP, ‡ Charles Hu, BSc,*
Duminda Wijeysundera, MD, PhD, † Christine Massey, BSc,§ and Natalia Kraeva, PhD*

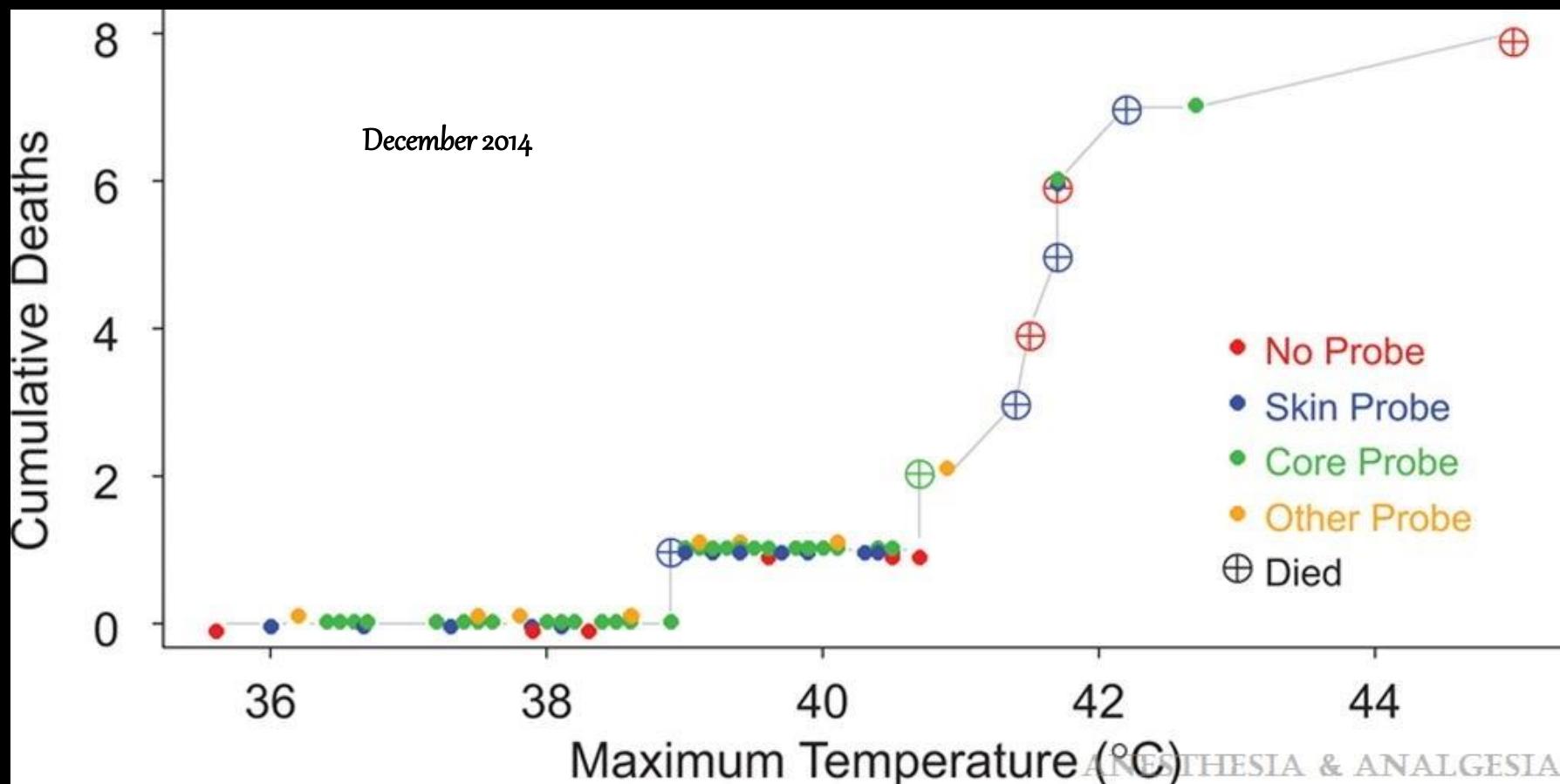


Table 1. Characteristics of Index Adverse Anesthetics in 129 Malignant Hyperthermia-Susceptible Probands

Demographics	
Male sex	79 (61.2%)
Age (y) at onset, median (range) [IQR]	23 (1–70) [10–33]
Caucasian	123 (95.3%)
Medical history	
Prior muscle symptoms	14 (10.9%)
Prior unremarkable anesthetics	17 (13.2%)
Prior unremarkable anesthetics (range) [IQR]	(1–5) [1–3]
Anesthetic triggers	
+Succinylcholine, –volatile, n (%) [95% CI]	20 (15.5%) [10–23]
+Succinylcholine, +volatile, n (%) [95% CI]	51 (39.5%) [31–49]
–Succinylcholine, +volatile, n (%) [95% CI]	58 (45.0%) [36–54]
–Succinylcholine, –volatile, n (%) [95% CI]	0 (0.0%) [0–3]

Malignant Hyperthermia Deaths Related to Inadequate Temperature Monitoring, 2007–2012: A Report from The North American Malignant Hyperthermia Registry of the Malignant Hyperthermia Association of the United States

Marilyn Green Larach, MD, FAAP,* Barbara W. Brandom, MD,*† Gregory C. Allen, MD, FRCPC,‡
Gerald A. Gronert, MD,§ and Erik B. Lehman, MS||



"All the News
That's Fit to Print!"

The New York Times.

Vol. XXXV, No. 21,894.

"Read Today!"

Copyright 1945 by The New York Times Company

NEW YORK, THURSDAY, AUGUST 9, 1945.

LATE CITY EDITION

Special with four additional pages.
Postage 10 cents. Postage 10 cents.
Subscription \$1.00 a year. Postage
10 cents a year. Postage 10 cents.

THREE CENTS

SHAFFER, DEXTER, BRULL DECLARE WAR ON SKIN TEMP

TRUMAN TO REPORT
TO PEOPLE TONIGHT
ON BIG 3 AND WAR

Half-Hour Speech by Radio to
Cover a Wide Range of Prob-
lems Facing the World

WE SIGNED PEACE CHARTER

AND THIS MAKES THIS COUNTRY
THE FIRST TO COMPLETE ALL
BATTLESHIPS REQUIREMENTS

At the announcement
of the Japanese signing of the
Instrument of Surrender, Mr. Truman
and Mr. Shaffer, his special
assistant on the Far Eastern
front, were interviewed on
the radio by Mr. Edward R. Murrow
in a three-part series.

The President's message
stated in this, and today, and
the speech, which probably were
the most dramatic of the day, showed
the pride and determination of the
American people to see the
whole of the world free of the
dictators.

Mr. Truman's speech on
the radio was followed by a series
of other news items which re-
mained during the meeting.

Foreigners
To Stay

Mr. Shaffer's speech
concerned with the
problem of the
Japanese and
other foreign
nationals who
have been
resident in
the United
States.

"The Japanese
are a very
responsible
people. In
the course of
the history of
the United
States, a
considerable
number of
Japanese
have been
resident in
the United
States. They
have been
responsible
citizens."

The closure of
the Japanese
embassy in
Washington, D.C., was
also mentioned
in the speech.

TANNERY
LAST OF
County Board
Executive Staff
Meeting P

*"If you don't
monitor core
temperature
routinely, start
today."*

IMPLANTED RUSSIA AIDS ALLIES

Join Pacific Struggle
After Spurning Foo's
Mediation Plea

SEEDS EARLY PEACE

Nolodoff Reveals Move
Three Months After
Victory in Europe



By ROBERT W. SHAFER
Mr. Shaffer is the new Far Eastern
assistant. Aug. 9.—Russia de-
clared war on Japan tonight, in a
decisive move which will end
the Japanese peace conference with
the U.S.S.R. Mr. Shaffer said
the declaration, which was made
in the name of Mr. V. M. Molotov
(S.P. W. New York Times).
In view of Japan's refusal to
sign the peace agreement, Mr. Shaffer said,
the Allies proposed that the Soviet
Union join the war against Japan
as soon as possible and that
the Allies would assist the
Soviet Union in its war against Japan.

Mr. Shaffer said that the
Soviet Union would assist the
U.S. in its war against Japan
and that the U.S. would assist the
Soviet Union in its war against Japan.

"All the News
That's Fit to Print!"

The New York Times.

Vol. XXXV, No. 21,894.

"Read Today!"

Copyright 1945 by The New York Times Company

NEW YORK, THURSDAY, AUGUST 9, 1945.

LATE CITY EDITION

Special with four additional pages.
Postage 10 cents. Postage 10 cents.
Subscription \$1.00 per year. Postage
10 cents. Postage 10 cents.

THREE CENTS

SHAFFER, DEXTER, BRULL DECLARE WAR ON SKIN TEMP

TRUMAN TO REPORT
TO PEOPLE TONIGHT
ON BIG 3 AND WAR

Half-Hour Speech by Radio to
Cover a Wide Range of Prob-
lems Facing the World

WE SING PEACE CHANTER

And That Makes This Country
The First to Complete All
Battalion Requirements

Mr. Truman and his
advisors are to speak to the
nation on the problems con-
cerning the world tomorrow
at 9:30 A.M. Eastern Daylight
Time in a half-hour broadcast on
the radio.

The President's speech
will be from the White House
and will cover a wide range of
problems, including the
atomic bomb, the Korean
conflict, and the like.

Mr. Truman's speech will
be the first of its kind in
the history of the United States.

Foreigners
To Stay

Mr. Truman and his
advisors are to speak to the
nation on the problems con-
cerning the world tomorrow
at 9:30 A.M. Eastern Daylight
Time in a half-hour broadcast on
the radio.

Mr. Truman's speech will
be the first of its kind in
the history of the United States.

*"Do it right, with a
continuous
electronic
measurement of core
temperature."*

UNPLATED RUSSIA AIDS ALLIES

Join Pacific Struggle
After Spurning Foo's
Mediation Plea

SEEDS EARLY PEACE

Nolodoff Reveals Move
Three Months After
Victory in Europe



Mr. Nolodoff, spokesman
for Moscow in the Far East
Soviet Union, Aug. 8.—Russia de-
clared war on Japan tonight, in a
decisive move against the
Axis. P. M. Foreign Minister
Nolodoff, who recently made his
declaration, which was received
in New York by H. P. M. Director
of the Foreign Service (P. M. New York Office).
In view of Japan's refusal to
surrender, Mr. Nolodoff said, the
Allies proposed that the Soviet
Union join the war against Japan
and its allies, Germany and Italy.
The declaration of war
was made by Moscow and
the Soviet Union, which
is the result of intense and
determined efforts.

"All the News
That's Fit to Print"

The New York Times.

卷之三

卷之三

2000-2001 THE BURGESS MUSICALS A 1000

Yunnan Chinese 2000

SHAFER, DEXTER, BRULL
DECLARE WAR ON SKIN TEMP

“This is better than explaining to patients, parents, or the next of kin, why you chose to save \$6.”

TRUMAN TO REPORT
TO PEOPLE TONIGHT
ON BIG 3 AND WAR

Foreigners To Stay

Half-Hour Speech by Radio to Cover a Wide Range of Problems Facing the World

THE THREE PLATE COUNTS

And Thus Stales This Country
The First Is Complete All
Ballot-box Requirements

At the same time, the *Washington Post* and *Times* report that the situation on the Potowatamie reservation is still unclear, with officials uncertain as to what the Indians had done. "There is a strong desire among

The (President) announced,
earlier to those, and today, was
the speech, which probably would
have been more detailed, had
he been present, dated from the
announcements made by the big
bosses of the town at the meeting
July 19.

TAMMANY
LAST CO.

County Comm.
Executive Secy.
Meeting R.

Mr. Green would be the
most likely to tell us what
kind of paper was used in
making money in Germany.

OMPLAISER RUSSIA AIDS ALLIES

Joint Pacific Struggle After Spurning Foe's Mediation Plan

SEEDS EARLY PEACE

Noktaff Reveals More Three Months After Victory in Europe

By John W. Thompson
In Volume 10, No. 1, January
1900. Aug. 10—Boston Ad-
mitted as an honor member. In

Japanese power companies had a 4.5% P. & E. Foreign Companies Committee at industry must bear the difference, which was passed by the new public at 10-17, M. 1945 (S. P. New York Used).

MH Mortality in ASC



FEATURING *iVillage* search MSNBC web MSN Home | Mail **msn**

People

Cheerleader's death highlights rare surgical risk

High school senior dies after undergoing cosmetic breast surgery

By Mike Celizic
TODAYShow.com contributor
updated 10:19 a.m. ET, Wed., March, 26, 2008

Stephanie Kuleba's friends called her "Sunshine" because that was the perfect nickname for the outgoing and bubbly girl who was everybody's friend, the cheerleader with the near-perfect grade-point average who was too nice and too perfect for anybody to resent.

"She was just the kind of girl that everyone loved," a friend, Dayna Mercer, told NBC News. "There was nothing bad about her."

But the 18-year-old high school senior, who was headed to college and then medical school, felt

Video



Launch

Most MH deaths from hyperthermia!



Heat Stroke and MH

- 12 year old child has acute MH in OR
- CHCT shows MH susceptibility
- 8 mos later: dies after football practice: T 108 F
- RYR-1 mutation detected in pt and relatives



Tobin et al. JAMA, 286, 2001

Awake MH?

Identical *de novo* Mutation in the Type 1 Ryanodine Receptor Gene Associated with Fatal, Stress-induced Malignant Hyperthermia in Two Unrelated Families

Linda Groom, B.Sc.,* Sheila M. Muldoon, M.D.,† Zhen Zhi Tang, Ph.D.,‡
Barbara W. Brandom, M.D.,§ Munkhuu Bayarsaikhan, Ph.D.,|| Saiid Bina, Ph.D.,#
Hee-Suk Lee, M.D.,** Xing Qiu, Ph.D.,†† Nyamkhishig Sambuughin, Ph.D.,‡‡
Robert T. Dirksen, Ph.D. §§

Anesthesiology 2011; 115:938

7 reported cases





To protect private
Patient Information

Only (1) family allowed at
the desk at a time

Last name appears on monitor
when prescription is ready

Thank You.

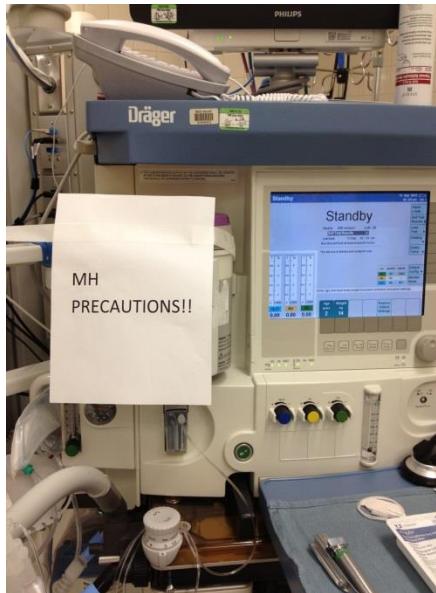
Anesthesia for Known MH Patients

- Non-triggering technique
- Carefully monitor etCO₂
- Normal discharge criteria
- ASC should be adjacent to hospital



Preparation of Anesthesia Workstations for MH Susceptible Patients

4 alternatives:



ASC Anesthesia for Known MH Patients



Vapor-Clean, Dynasthetics





Immediate Treatment of MH

Important Steps

- Mobilize personnel
- **DANTROLENE**
- Arrange transfer
- Continue active treatment
- **800-MH-HYPER (644-9737)**



RYANODEX

- Eagle Pharmaceuticals
- **250 mg/5 mL**
- Decreased time to preparation
- Less volume load
- **1** successfully treated case!
- **More data next year!**



RYANODEX

- Contains subclinical amounts of mannitol
- **Why is mannitol in older versions of dantrolene?**
- MHAUS changed website guidelines on treatment of myoglobinuria



Why Joan Rivers Matters



Why Joan Rivers Matters



Dantrolene & Ca Channel Blockers

- CCBs probably detrimental during dantrolene admin
- **Never withhold dantrolene in MH pts on maintenance CCBs.**
- MHAUS has retracted website statement



Malignant Hyperthermia

MHAUS Hotline 1-800-644-9737

Surgery Centers: 911 and transport arrangements

Anesthesiologist

1. CALL FOR HELP!
2. Stop triggering agents and place charcoal filters in circuit
3. Hyperventilate with 100% O₂ @ 10L/min
4. Start TIVA
5. Give Ryanodex 2.5 mg/kg IV
6. Aggressive IV hydration
7. Active cooling if T > 38C
 - Bair Hugger to cool
 - Cold IVF 0.9% NS
 - Ice packs to head, groin, axillae
 - Cold lavage, cavity irrigation
8. Place lines
 - Arterial line
 - Foley catheter
 - NG tube
9. Obtain Labs
 - ABG, glucose, CK, lactate, electrolytes, CBC, serum myoglobin, coags, D-dimer, fibrinogen
 - Urine myoglobin

Nursing

1. Call Anesthesia techs for MH equipment
2. Obtain:
 - Supplies for wound irrigation and closure
 - Urinary catheter tray
 - Ice packs

Anesthesia Techs

1. Bring:
 - MH cart
 - Defibrillator
 - ISTAT, glucometer
 - Arterial and central line supplies
2. Prepare cold 0.9% NS IVF

Surgeon

1. Irrigate cavity with cold fluid
2. Stop procedure when appropriate
3. Place urinary catheter
4. Assist with line placement
5. Call ICU to arrange bed

**IN EVENT OF PVCs, DYSRHYTHMIA, OR ARREST
ASSUME HYPERKALEMIA**

RYANODEX

- Each vial contains 250mg
- Dissolve each vial with 5 mL **sterile water**
- Give 2.5mg/kg IV bolus
- Repeat dose if signs of MH persist (up to 10mg/kg)
-Ca Channel blockers are contraindicated after administration of Dantrolene!
-Dantrolene causes potentiation of NMBD

Treatment Goals

1. HD Stability
2. Temp <38 C
3. Prevent Rhabdomyolysis
4. Correction of Acidosis and Hyperkalemia
 - Aggressive IV hydration with 0.9% NS
 - Maintain urine >2ml/kg/hr
4. Correction of Acidosis and Hyperkalemia
 - Hyperventilation
 - NaHCO₃ 1-2 mEq/kg
 - CaCl or Ca Gluconate
 - Insulin 0.1U/kg with D50





Malignant Hyperthermia Association
of the United States

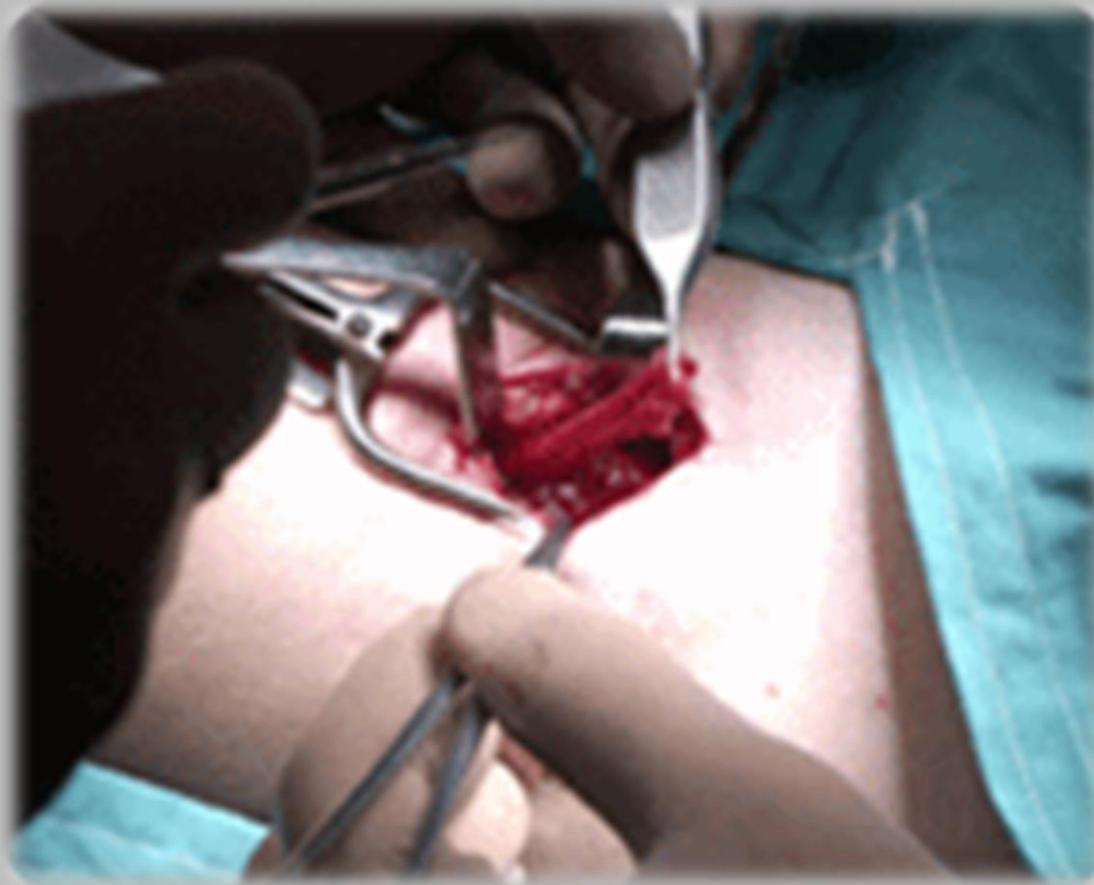
MHAUS GUIDELINES



**Testing for Malignant
Hyperthermia (MH)
Susceptibility:**

**How do I counsel
my patients?**

Muscle Contracture Testing (CHCT)



CHCT Testing, U.S. and Canada

University of Minnesota
Minneapolis, MN
Paul A. Iaizzo, PhD
(612) 624-7912 or -3959

Uniformed Services University
of the Health Sciences
Bethesda, MD
(Military & Civilian)
Sheila M. Muldoon, MD

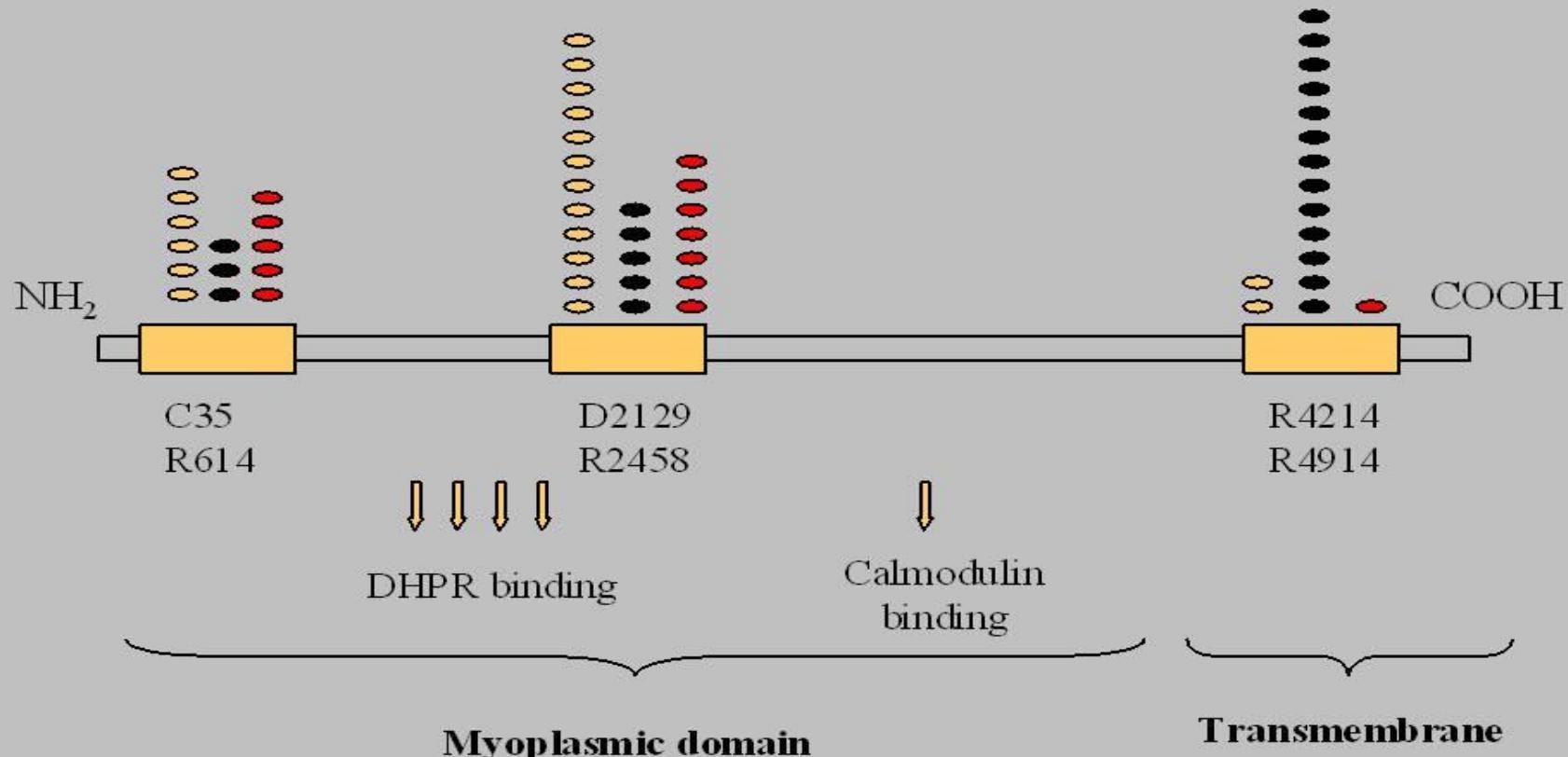
University of California
Davis, CA
Timothy Tautz, MD

Wake Forest University
Winston-Salem, NC
Joseph R. Tobin, MD

Toronto General Hospital
Toronto, Ontario
Julian Loke, MD, FRCPC

Genetic Testing: RYR1 Gene Sequencing

RYR1: gene structure and mutational spots





NEW YORK POST

Page
Six

LATE CITY FINAL



The next Jeter

BOMBERS' BIG
TRADE: SPORTS



**European MH
Group Finds NEW
MUTATIONS!**

36 MH-Causing Mutations!

ISITÄT BASEL

EMHG

European Malignant Hyperthermia Group

[HOME](#) [SITEMAP](#) [CONTACT](#) [LOGIN](#)

[EMHG](#) [MH UNITS](#) [DOCUMENTS](#) [GENETICS](#) [NEWS & EVENTS](#) [HONORARY MEMBERS](#)

**European Malignant
Hyperthermia Group (EMHG)**



Correspondence about the EMHG to:
Professor PM Hopkins
Secretary of the EMHG
Leeds Institute of Molecular Medicine
St James's University Hospital
GB-LEEDS
LS9 7TF

E-Mail: P.M.Hopkins-at-leeds.ac.uk

Correspondence about this website see
[contacts form](#).

EVENTS

11.06.2015 to 13.06.2015
[2015 Annual Meeting: Lille](#)
26.05.2016 to 28.05.2016
[2016 Annual Meeting: Ulm](#) (Dates
to be confirmed)

NEWS

12.12.2014
New diagnostic mutations
19.08.2014
33rd annual meeting abstracts on **BMC Anesthesiology**
12.06.2014
33rd EMHG Meeting in Würzburg, Germany
16.12.2013
Dr. Mary Lehane , FFARCSI
10.07.2013
Speaking about MH => here are some slides for you

QUICKLINKS

[:: EMHG](#)
[:: MH Units](#)
[:: Documents](#)
[:: Genetics](#)

MALIGNANT HYPERHERMIA SUSCEPTIBILITY NEXTGEN SEQUENCING (NGS) PANEL

[Clinical Features and Genetics](#)[Methods and Pricing](#)[How to Order](#)[Additional Information](#)

TEST METHODS

- ▶ [NEXTGEN SEQUENCING](#)
- ▶ [DELETION/DUPLICATION TESTING VIA ARRAY COMPARATIVE GENOMIC HYBRIDIZATION](#)

NEXTGEN SEQUENCING

Test Number	Test	Price	CPT Code(s)
1383	NextGen Sequencing (3 genes)	\$1,690	81479 (x2), 81408

In addition, Targeted Familial Mutation testing via Sanger sequencing is available for any gene in the panel:

Test Number	Test	Price	CPT Code
100	Targeted Familial Mutations - Single Exon Sequencing	\$250	81479
200	Targeted Familial Mutations - Double Exon Sequencing	\$370	81479
300	Targeted Familial Mutations - Triple Exon Sequencing	\$440	81479



Malignant Hyperthermia

“Hot Off the Press”



Ron Litman

litmanr@email.chop.edu