RECOMMENDATIONS FOR SURGERY AND ANESTHESIA

For Healthcare Professionals, detailed information is available at www.myotonic.org

Patients with myotonic dystrophy often exhibit adverse reactions to sedatives, anesthetics, and neuromuscular blocking agents. Serious complications and fatalities can be avoided by careful preoperative assessment, avoidance of certain drugs, careful monitoring and good postoperative patient care throughout their hospitalization.

It is especially important post-op to monitor heart, ventilatory function and airway clearance closely because of the prolonged effect of anesthesia and pain medication in patients with myotonic dystrophy. Complications are not proportional to the severity of the disease and can arise in seemingly mildly affected patients. Avoidance of general anesthesia is warranted if regional anesthesia or alternate approaches can be identified.

PREOPERATIVE:

1. **Cardiological assessment:** ECG essential, rhythm monitor (e.g., ZioPatch)
2. **Respiratory assessment:**
   a. FEV1 and FVC both lying and standing
   b. Chest x-ray, noting elevation of diaphragm or areas of atelectasis
   c. Arterial blood gases
3. **Premedication:** avoidance of opiates, and caution with benzodiazepines

INTRAOPERATIVE:

1. **Induction:** preferably gaseous; avoid hypnotic agents with slow metabolism such as Thiopentone. Adverse reactions have also been reported with Propofol; lower doses are likely to be required. Careful titration of intravenous induction agents to avoid hypotension.
2. **Relaxation:**
   a. avoid Suxamethonium chloride
   b. short-acting, non- depolarizing muscle relaxants are best used and may be needed in smaller doses; recovery from these may be prolonged
3. **Reversal:** neostigmine may produce ACH-induced depolarization blockade
4. Protection of airway to minimize risk of aspiration; tendency to a neuromuscular condition that may cause the following symptoms: temporomandibular dislocation - care needed in manipulating jaw
5. Neuromuscular and capnograph monitoring
6. ECG monitoring essential due to risk of arrhythmias
7. Monitor core temperature; to avoid postoperative shivering, maintain normothermia by using warming pads
8. Avoid Potassium (K+) containing fluids

POSTOPERATIVE: (FIRST 24 TO 48 HOURS)

1. Ensure respiration is fully re-established
2. Cardiac monitoring*
3. Respiratory monitoring: pulse oximetry, arterial blood gas analysis*
4. Use of a high dependency bed is preferable*
5. Early chest physiotherapy: these patients are especially prone to postoperative chest infections and atelectasis
6. Minimal use of opiates for analgesia; instead explore other methods, e.g., local anesthetic blocks or non-steroidal anti-inflammatory agents and, paracetamol

**The extent to which these precautions are taken will depend on the length and nature of the procedure.*

FOLD 1

PERSONAL DATA

Name

Address

City

State Zip

Phone

Cell

DOB Blood Type

EMERGENCY CONTACT

Name

Phone

Relationship

Contact #2 Name

Phone

Relationship

FOLD 3

PRIMARY PHYSICIAN

Name

Address

City

Phone

INSURANCE

Primary Company

Policy #

Secondary Company

Policy #

For additional information about myotonic dystrophy please contact:

Myotonic Dystrophy Foundation

www.myotonic.org

info@myotonic.org

+1 (415) 800-7777

FOLD 2

MEDICAL ALERT

The bearer of this card has **MYOTONIC DYSTROPHY**, a neuromuscular condition that may cause the following symptoms:

- Muscle weakness, stiffness and pain
- Balance difficulties
- Extreme fatigue and sleepiness
- Speech difficulties
- Swallowing difficulties
- Abnormal heart rhythm

PROBLEMATIC MEDICATIONS

- General anesthesia
- Benzodiazepines
- Liquid paraffin
- Neuroleptics
- Opiates
- Quinine, procainamide, tocainide

This list is not exhaustive and caution is recommended in the use of any medication that alter cardio/ pulmonary or muscular function. Consult with your physician prior to taking any medication.

MYOTONIC DYSTROPHY

MEDICAL ALERT AND HISTORY

NAME: _____

VITALS:

Wt._____ Ht._____ Baseline Temp._____ Date_____

Heart Rate_____ Date_____ Pulse_____ Date_____

ALLERGIES:
(medication, food & environmental)

_____

_____

_____

MEDICATIONS:

Medication: Dosage: Physician: Reason:

_____

_____

_____

_____

_____

_____

HOSPITALIZATIONS:

Date: Hospital: Reason:

_____

_____

_____

_____

_____

_____

PHYSICIANS:

Physician: Phone: Specialty:

_____

_____

_____

_____

_____

HOW MYOTONIC DYSTROPHY CAN AFFECT ONE'S HEALTH

- **CARDIAC PROBLEMS:** The impact of DM1 on the heart is mainly on the cardiac conduction system. Various tachyarrhythmias and bradyarrhythmias may cause palpitations, fatigue, chest pressure, dyspnea, syncope, presyncope or dizziness.
- **ANESTHESIA AND SURGERY:** Myotonic dystrophy can impair recovery from general anesthesia. The surgeon and anesthesiologist must know about the complexities of this diagnosis before any anesthesia or procedure.
- **RESPIRATORY PROBLEMS:** weakness of breathing and airway muscles, and impaired central respiratory drive results in hypoventilation, sleep-disordered breathing and aspiration with frequent lung infections.
- **DIGESTIVE PROBLEMS:** can lead to swallowing problems, reflux, bowel pains, gallstones, severe constipation, diarrhea and acute pseudo-obstruction.
- **FATIGUE:** is very common and often extreme with excessive daytime sleepiness. Heart problems: can cause abnormal rhythm and conduction problems requiring treatment and can sometimes be fatal.
- **MUSCLE WEAKNESS:** is variable and progressive, involving the face, eyelids, jaw, neck, forearms, hands, legs and feet. The muscles of speech are often affected.
- **MYOTONIA:** prolonged stiffness due to impaired relaxation of muscle after contraction.
- **OPTICAL PROBLEMS:** iridescent cataracts, droopy eyelids and corneal dystrophy.
- **COGNITIVE AND PERSONALITY CHANGES:** lack of motivation, avoidance, altered executive function, and attention deficit and autism spectrum changes in children.
- **OTHER PROBLEMS:** diabetes, thyroid dysfunction, male infertility, obstetrical issues including peripartum hemorrhage, premature labor, and other complications. Children may experience learning difficulties and inattention and ADHD among other problems.

For Healthcare Professionals, detailed information is available at www.myotonic.org and at the specific topic links below:

myotonic.org/Anesthesia
myotonic.org/Cardiology
myotonic.org/Pulmonology
myotonic.org/AdultsDM1
myotonic.org/AdultsDM2
myotonic.org/ChildrenDM1

Myotonic Dystrophy FOUNDATION

The information on this Medical Alert and History has been adapted from the Myotonic Dystrophy Care Card in the U.K. in 2010, with permission from Dr. Douglas Wilcox, Glasgow, for use within the U.S. medical system by the following members Myotonic Dystrophy Foundation and Medical and Scientific Advisory Committee:

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This Medical Alert and History was reviewed and updated in 2023 by the Myotonic Dystrophy Scientific Advisory Committee found here **www.myotonic.org/scientific-advisory-committee**