Myotonic Dystrophy FOUNDATION

Practical Advice for Anesthesia for Individuals with Myotonic Dystrophy and Their Families

The mission of the Myotonic Dystrophy Foundation is Community, Care, and a Cure.

We support and connect the myotonic dystrophy community.

We provide resources and advocate for care.

We accelerate research toward treatments and a cure.

Thank you
MDF is grateful to the following writers and reviewers of this document:
Annalise Kalmanoff, Lorraine Dressler, Dr. Marla Ferschl, Dr. Michael Gropper, Dr. Stephen Bolt

This guide is meant to accompany the Myotonic Dystrophy Foundation publication *Practical Suggestions for the Anesthetic Management of a Myotonic Dystrophy Patient"*

Disclaimer: This guide was created to help educate you about anesthesia.
This guide does not replace any advice from your doctor and is meant to be educational only.

A publication of the Myotonic Dystrophy Foundation

Chief Executive Officer: Tanya Stevenson, EdD, MPH

Publication Design: Julie Mills, Designpony

11/24

Practical Advice for Anesthesia for Individuals with Myotonic Dystrophy and Their Families

For individuals living with myotonic dystrophy (DM), undergoing general anesthesia can pose higher risk than other people. According to the Christopher Project[1], around half of respondents were not aware of general anesthesia risks associated with DM and/or had not discussed these risks with a medical professional. This guide will help you understand potential risks, ways that you and your medical team can make the process safe, and how you can advocate for the best care possible.

Types of Anesthesia

Anesthesia is a medical intervention preventing patients from feeling pain, moving, or forming memories during surgery (or any painful or stress inducing procedure).

There are a wide range of anesthetic medications which can be given through inhalation and/or directly into veins. Because of the technical complexity and variability with these medications, medical specialists called anesthesiologists help create a plan based on your medical history and the type of procedure you're undergoing.

There are three main types of anesthesia:

1. **General**
   Under general anesthesia, the patient is unconscious. This is used for major surgeries such as heart surgery, abdominal/chest, and spine surgery.
2. **Regional/Neuraxial**

   Regional anesthesia causes numbness over smaller body regions and is useful for surgery on any/all extremities, and is especially useful for extended post-operative pain management without opiates/narcotics.

   Neuraxial anesthesia (spinal/epidural) will numb major body regions and is useful major chest or abdominal surgery, surgery on the hips or legs, and for childbirth.

   Both regional and neuraxial anesthetics are frequently combined with intravenous sedation during both the placement of the block and to provide amnesia during the surgical procedure.
3. **Local**
   Local anesthesia numbs a small part of body while the patient is conscious, such as when stitches are applied.

NOTE Refer to the Myotonic Dystrophy Foundation Clinical Care Guidelines for more information for for individuals with myotonic dystrophy and their families here: **www.myotonic.org/toolkits-publications**

Because of the sensitive nature of anesthesia and because the person with DM may be unconscious during procedures, it is recommended to have an informed family member and/or caregiver who can advocate on your behalf. This person should be familiar with the risks of anesthesia, your unique health history, and what steps your medical team should be taking. They should also attend your pre-operative assessment and any other related appointments with you.

[1] *The Christopher Project: Report to the Myotonic Dystrophy Community, 2019 https://www.myotonic.org/sites/default/files/pages/files/Christopher_Project_Full_Report.pdf.*

Anesthesia - particularly general anesthesia - can be dangerous with DM because it can worsen muscle weakness and affects a wide range of body systems.

Body System	Potential Risk with Anesthesia
 **Heart**	• Rhythm or electrical problems called "arrhythmias". • For more information on DM and the heart, refer to MDF's Myotonic Dystrophy and the Heart: A Community Guide found here: https://www.myotonic.org/toolkits-publications.
 **Lungs**	• Weak respiratory muscles can lead to weak cough or breathing, lung collapse "atelectasis", low volume of air inhaled "poor inspiratory capacity", infrequent breathing "alveolar hypoventilation", and pneumonias (lung infections). All can cause low oxygen "hypoxia" & high carbon dioxide (CO2) "hypercapnia" in the blood, which is very dangerous. • Respiratory risks are higher during surgeries around the upper abdomen, chest, spine, and for older individuals.
 **Muscles**	• Inability to relax muscles "myotonia" triggered by cold, low potassium, cautery (tool to stop bleeding), & electrical stimulation. • Increased sensitivity to muscle relaxant medications used during anesthesia. • Succinylcholine (a commonly used depolarizing muscle relaxant) should be avoided in patients with DM. • Non-depolarizing muscle relaxants may be necessary to allow the surgeon to have adequate conditions for your type of surgery. • The degree of muscle relaxation is constantly monitored during surgery by the anesthesiologist. S/he will also administer a medication to reverse the effects of the muscle relaxant at the conclusion of the operation.
 **Nervous System**	• People living with DM are more sensitive to sedative medications and opiates (type of painkiller) can cause a variety of problems throughout the body.
 **Airway**	• Aspiration pneumonia caused by weak airway muscles and an incompletely empty stomach, difficulty swallowing "dysphagia", and sleep apnea (breathing stops and starts repeatedly.)
 **Digestive Gastrointestinal (GI) System**	• Issues with GI muscles can cause reduced contractions called "gastrointestinal dysmotility" that can result in total shutdown called "ileus". This can lead to nausea, vomiting, constipation, reaching fullness easily, and a distended stomach.
 **Brain**	• Confusion called "encephalopathy" and difficulty waking up.
 **Endocrine**	• Problems with glucose and insulin metabolism that can lead to abnormal blood sugar.

What to Expect with General Anesthesia

General, regional and local anesthesia require careful preoperative planning, close intraoperative monitoring and vigilant postoperative care.

Before Receiving Anesthesia

- In the weeks leading up to the procedure, you will have a team of specialists who will communicate with each other and with you.
  - This team may look different for everyone, but will likely include a surgeon, anesthesiologist, primary care physician, pulmonologist, cardiologist, neuromuscular specialist/neurologist etc. If you have specific health needs, make sure the associated specialist is notified (e.g., If you have respiratory issues a pulmonologist should be heavily involved and in good communication with the rest of the medical team)
  - Give every member of the medical team MDF's ***Anesthesia Guidelines*** (https://www.myotonic.org/sites/default/files/pages/files/MDF_PracticalSuggestionsDM1_Anesthesia2_17_21.pdf) and ***Quick Reference Guide*** (https://www.myotonic.org/sites/default/files/pages/files/MDF_PracticalConsiderationsAnesthesiaQG_1_21.pdf)
  - Organizational tools like medical binders, calendars, and reminders are useful for managing symptoms, tests, and specialists.
- You will have a pre-operative assessment meeting with an anesthesiologist to create an anesthesia plan and determine your specific risks.
  - Assessment should be early (2-4 weeks before procedure) and in-person (ask for an in-person meeting if you're offered phone call or video meeting)
  - Tell your anesthesiologist you have or may have DM, health conditions and/or specific health problems, if you've had anesthesia before, if you use a BiPAP/CPAP machine, and about any medications you take
  - Discuss an opiate-sparing postoperative pain management strategy with your anesthesiologist, such as one using regional/local anesthesia, IV or oral acetaminophen, NSAIDS (if allowed by the surgeon) in order to minimize potential complications due to DM.
  - Your anesthesiologist will decide, in conjunction with the surgeon, if general anesthesia is needed. If appropriate, general anesthesia may be combined with local/regional anesthesia, especially to provide extended pain relief in the postoperative period (regional and local may be safer than the use of opioid pain medications). Lastly, s/he should tell you your options, potential risks and benefits, and what the process may look like. Ask them about options if they do not bring it up.
- Your medical team will perform a physical examination, check your vital signs, and may run some common preliminary medical tests.

REMINDER Carry a **Myotonic Dystrophy Medical Alert Card** that indicates you have DM in case of an emergency surgery. www.myotonic.org/sites/default/files/pages/files/MDF-Medical-Alert-Card.pdf

During Your Procedure

- Anesthesiologists will give you anesthesia medication to put you to sleep ("induction") and monitor your condition throughout the procedure.
- Anesthesiologists will give the anesthesia through inhalation or intravenously (or both depending on the types of anesthesia medication being used).
  - During general anesthesia, doctors will manage your airway to prevent aspiration, usually with a breathing tube.
  - Anesthesiologists may use a wide variety of anesthesia medicine (there's no specific medicine preferred for people living with DM) but should avoid succinylcholine.
  - If you can, communicate your symptoms with the medical professionals around you - never assume what you are feeling is normal, and speak up if you are concerned.
- The anesthesiologist will monitor your vital signs (heart rate and blood pressure, oxygen and CO2 levels, and temperature), and ensure a warm environment (room and operating table) to minimize myotonia.

After Your Procedure

- Problems are most likely to occur after the operation but safety precautions during the procedure, a prolonged postoperative stay, and close monitoring can reduce the risk of post-operative complications.
- It is the anesthesiologist's and the medical team's job to monitor your condition after the procedure:
  - Monitor heart rhythm (with EKG) and oxygen (with a pulse oximeter.)
  - Watch for shallow or irregular breathing and use artificial airway support (e.g., BiPAP) if necessary.
  - Monitor CO2 levels (especially if you are feeling tired or have difficulty breathing normally).
- Doctors will minimize your aspiration risk caused by poor airway control and weak cough by elevating the head of your bed to at least 30 degrees (they may use cough assist machine or incentive spirometry to improve lung strength), monitor for confusion, manage pain safely (with NSAIDS/acetaminophen/local/regional anesthesia and minimize the use of opiates/sedatives/muscle relaxers), and encourage movement and physical therapy (once it is safe).

Advocacy

This is a summary of an ideal, safe anesthetic procedure for a person living with DM following anesthesia guidelines. It is the medical professionals' responsibility to keep you safe, ***HOWEVER*** if you (or whomever may advocate for you while you're unconscious or unable) believe the care you are receiving violates any of these guidelines and/or you feel something is wrong, **you have the right to speak up, ask questions, and say no. Remember that your comfort and safety are top priorities; communicate your needs and feel empowered to refuse care until those needs are met.**

Summary

Undergoing anesthesia can be risky for people living with DM, but there are a variety of things you and your medical team can do to make the process much safer. Make sure your medical team is informed about DM and anesthesia, and prepared to take the safety precautions recommended. It is important for you to understand that you (or a caregiver) can and should advocate for your health and safety throughout the process by speaking up when something feels and saying no when you feel uncomfortable.

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663 Thirteenth Street, Suite 100, Oakland, California 94612
415.800.7777 | info@myotonic.org | www.myotonic.org