



MYOTONIC
DYSTROPHY
FOUNDATION

Care and a Cure



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CONSENSUS-BASED CARE RECOMMENDATIONS: WHAT DO THEY MEAN FOR YOU?

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MDF CLINIC VISIT PLANNER

LIFE THREATENING SYMPTOMS

 ANESTHESIA	Must inform your doctor about anesthesia precautions - see www.myotonic.org/mdf-anesthesia-guidelines . Must inform healthcare personnel involved in any surgeries or other procedures needing anesthesia or pain control
 RESPIRATION	#1 cause of death – tell your doctor about any chest infections; request vaccinations as recommended; request a flu shot every year; request a pulmonary function test at least once a year
 CARDIAC	#2 cause of death – irregular heart beats or arrhythmias and conduction issues can be unexpected and progress quickly; tell your doctor about any dizziness, light-headedness, chest pain, rapid or irregular heart beat; request an ECG immediately if you experience these symptoms; request a cardiac evaluation annually or as recommended in the Consensus-based Care Recommendations

SEVERE SYMPTOMS

 MUSCLE WEAKNESS	Tell your doctor about any falls, changes in function or mobility, e.g. things you can no longer do; if you are becoming more reliant on a caregiver; things you have had to give up doing; report speech and swallowing difficulties or changes; any myotonia or muscle cramping
 EXCESSIVE DAYTIME SLEEPINESS	Tell your doctor whether your sleep has changed; let your doctor know whether you are using your bi-pap or c-pap machine regularly; bring the machine or chip to the appointment so your doctor can record your measurements
 GI	Be sure to update your doctor about changes in status of reflux, bloating, abdominal pain, bowel movements
 EYES/VISION	Request an annual cataract check-up; let your doctor know if you are experiencing eye irritation or weakness, or eye-lid drooping
 TUMORS	Mention any new or painful growths or lumps; ask your doctor to follow normal cancer screening guidelines, particularly for cancer of the breast, testes, cervical or colon
 PSYCHOSOCIAL	Let your doctor know if you're experiencing financial need, having problems related to daily living, work or school, or if you are feeling unsafe; request referral to social services or support programs
 ENDOCRINE	Request a screening blood test for diabetes once a year and other endocrine-related blood tests
 THINKING & BEHAVIOR	DM is also a brain disorder – let your doctor know if you are experiencing any issues with or changes in mood, thinking, attention, memory, planning and organizing, motivation, resolving problems, or fatigue

Consensus-Based Care Recommendations for Adults with Myotonic Dystrophy Type 1

Quick Reference Version

The studies & rigorous evidence needed to drive the creation of an evidence-based guideline for the clinical care of myotonic dystrophy type 1 (DM1) patients have not yet been executed for all affected body systems & manifestations. In order to improve & standardize care for this disorder now, more than 60 leading myotonic dystrophy (DM) clinicians in western Europe, the United Kingdom, Canada & the United States have created the *Consensus-Based Care Recommendations for Adults with Myotonic Dystrophy Type 1*.

Summary recommendations from the Consensus-Based Care Recommendations are below. The full compendium of recommendations by body system & their disease manifestations is available here <http://www.myotonic.org/clinical-resources>.

LIFE THREATENING SYMPTOMS – CLINICAL CARE RECOMMENDATIONS

Surgery, anesthesia & pain

- See Myotonic Dystrophy Foundation's Practical Suggestions for the Anesthetic Management of a Myotonic Dystrophy Patient for anesthesia risks & recommendations before any surgeries or procedures requiring anesthesia <http://www.myotonic.org/clinical-resources>
- DM1 patients are far more likely to have adverse reactions to medications used for anesthesia & analgesia; interactions of the cardiac, respiratory, muscle & central nervous systems in each DM1 patient can lead to a variety of untoward responses before, during & after surgery
- Serious adverse events can occur throughout the course of DM1 & have been reported in patients whose DM1 symptoms were mild

- Behavioral & cognitive abnormalities need careful assessment & management preoperatively (if time permits & if it is possible) since these manifestations along with hypersomnia & preoperative sleep deprivation can complicate the patient's immediate postoperative care & long term recovery
- Most serious complications occur in the post-anesthesia period
- See full recommendations at <http://www.myotonic.org/clinical-resources>

Respiratory symptoms

- Pulmonary complications are the leading cause of death in DM1 patients. Clinicians must monitor issues such as recurrent pneumonia at baseline & serially with pulmonary function tests, at least forced vital capacity (FVC)
- See full recommendations at <http://www.myotonic.org/clinical-resources>

Fig. 1 Respiratory Care Recommendations Flowchart

