



MYOTONIC  
DYSTROPHY  
FOUNDATION

Care and a Cure

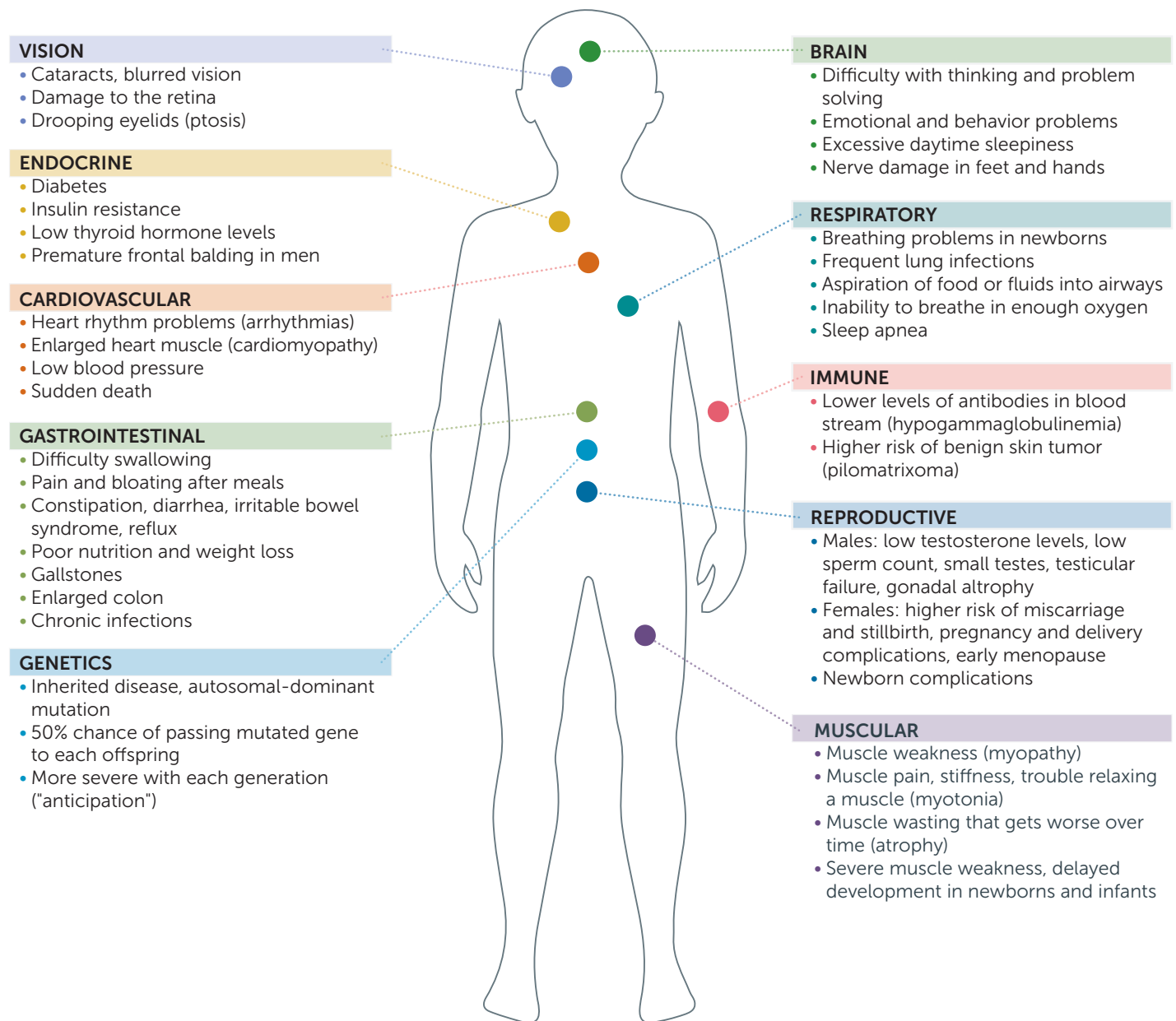
# A GUIDE TO MYOTONIC DYSTROPHY FOR SCHOOL PROFESSIONALS

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# A Guide to Myotonic Dystrophy for School Professionals

## How Myotonic Dystrophy Affects the Body

Myotonic dystrophy (DM) is a multisystemic disorder that can affect all age groups. Because of the range of systems affected, management requires a more expansive approach than most disorders and care is best provided by a coordinated, multidisciplinary team.



## Understanding the Neuropsychological Functioning Aspects of CDM and Childhood-onset DM1

The neuromuscular disorder myotonic dystrophy affects more than physical function. It is also a brain disorder that impacts thinking and behavior, especially in children with the congenital (CDM) and childhood-onset forms of the disease. Cognitive functioning, the mental processes necessary for learning, remembering, problem solving, and paying attention, may be mildly to severely impaired in children with CDM and childhood-onset DM1.

Changes in cognitive function may be subtle, occur gradually, and wax and wane due to environment

and psychological stressors, and they may be unique to the affected individual.

Cognitive function problems exhibited by children with CDM and childhood-onset DM1 are not issues of physical function, motivation, or unwillingness. Affected children are unable to complete tasks and follow directions in the same way as neurotypical children. The approach to addressing cognitive changes can be adapted to the child's level of functioning so that s/he can successfully complete tasks.

### Commonly Associated Cognitive and Psychosocial Symptoms that May Develop and Change with Disease Progression

- Attention problems
- Concentration problems
- Executive function impairment in one or more of the following:
  - Emotional control (emotional reaction to change)
  - Inhibition (stopping self from saying or doing something)
  - Initiation (difficulty starting a task)
  - Monitoring (underestimate/lack awareness of time it takes to complete tasks)
  - Organization of materials
  - Planning/organization
  - Shift (switching tasks, difficulty with transitions)
  - Working memory
- Intellectual disability
- Language problems
- Learning disorders
- Memory deficits (auditory, visual, working memory)
- Mood symptoms (e.g., anxiety, apathy, depression, flat affect)
- Motivation
- Perseveration
- Personality changes
- Problem solving or decision making (too many choices perceived)
- Processing speed (slowed)
- Thinking difficulty (thinking may be linear, circular, or disjointed)
- Visuoconstructional and visuospatial impairment

## Ways to Address Changes In Cognitive Function

- Determine individualized strategies that work for the child
- Introduce new information in steps
- Allow extra time for instructions and transitions
- Provide visual cues to help process information
- Help child initiate or start a project or task
- Create a system for organizing and planning (encourage child to be involved):
  - Use calendars (agenda planning)
  - Utilize check-lists for self-monitoring
- Address working memory problems:
  - Provide clear instruction
  - Use short sentences
  - Divide information and tasks into smaller sections or chunks
  - Ask for information to be repeated
- Check in: does the child understand?
- Repeat, repeat, repeat! (affected children may need to hear information many times and in different ways before they adapt their behavior)
- Reassess needs and strategies regularly

### **Cognitive functions can be impacted by emotions and behaviors.**

Children with CDM or childhood-onset DM1 will have normal emotional and/or behavioral reactions to disease progression. Affected children (and their families) may cycle through the stages of grief (denial, anger, bargaining, depression, and acceptance) as their disease progresses. Emotional and behavioral reactions may increase with an increase in physical and cognitive symptoms due to disease progression, and when affected individuals feel stuck because of their disease in comparison to their unaffected peers. Affected children may experience feelings of anxiety, hostility, depression and apathy, and may exhibit behaviors such as passivity or tantrums.

**When addressing psychosocial concerns, it is important to respond as you would to any child having an emotional reaction to change and/or circumstances.** Naming a child's emotional response to and feelings associated with the given situations helps to normalize and validate his/her reactions. Mourning disease progression and loss of function helps move toward acceptance. Working together to openly address emotions provides learning opportunities for affected children and parents/caregivers to engage in active listening, increase awareness of emotions and associated behaviors, and to problem solve.

**Planning activities that affected children can successfully complete creates a positive experience that may increase sense of control, motivation to continue, and healthy coping strategies.** Open discussion of disease symptoms, disease progression and impact is crucial to reducing associated feelings of anxiety and depression, and empowers affected children to control their emotional experience of living with CDM or childhood-onset DM1.

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**For more information contact the Myotonic Dystrophy Foundation**

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## Frequently Asked Questions About Myotonic Dystrophy

### ***What is myotonic dystrophy?***

Myotonic dystrophy (DM) is a multi-systemic inherited disease that affects 1 in every 2,550 people, or over 130,000 in the U.S. alone. Although often viewed as a muscle disease, individuals affected by DM may have skeletal muscle problems, heart function abnormalities, breathing difficulties, cataracts, issues with speech and swallowing (dysarthria and dysphagia), cognitive impairment, excessive daytime sleepiness, and diabetic symptoms. Any single individual is unlikely to have all or even most of these symptoms. Myotonic dystrophy is one of the most variable and complicated disorders known. The systems affected, the severity of symptoms, and the age of onset of those symptoms vary greatly between individuals, even in the same family.

### ***What is the prognosis for myotonic dystrophy diagnoses?***

Myotonic dystrophy is a progressive or degenerative disease. Symptoms tend to worsen gradually over several decades, though symptoms worsen at a different pace for different people. While no treatment currently exists that slows the progression of myotonic dystrophy, management of its symptoms can greatly improve patient quality of life. Early intervention can reduce or avert complications that sometimes arise. DM2 tends to be less severe than DM1 and has minimal impact on life expectancy. DM1 is much more variable and the prognosis for an affected individual is difficult to predict. Some people may experience only mild stiffness or cataracts in later life. In the most severe cases, respiratory and cardiac complications can be life-threatening even at an early age. In general, the younger an individual is when symptoms first appear, the more severe symptoms are likely to be. However, how myotonic dystrophy affects one individual can be completely different from how it manifests in another, even for members of the same family. It is impossible to predict how the disease will affect any single individual. Prognosis is as variable as the symptoms of this disease, thus it is difficult to make an accurate prognosis due to varied progression.

### ***Why do many patients have problems with diarrhea and constipation?***

Most problems are due to intestinal motility. Selection of foods is important. Appropriate amounts of fiber supplements may be useful, although overuse may produce impaction if the patient becomes constipated. Stool softener and non-irritant laxatives are useful to prevent constipation.

### ***What does one do when swallowing becomes a problem?***

Swallowing problems come from both oropharyngeal muscle weakness and abnormal motility of the esophagus. The patient should be evaluated by a speech pathologist and a gastroenterologist with fibroscopic and manometric testing. If patients are not able to do these studies, the modified barium swallow test should be done to assess the risk for aspiration. The speech therapist should be able to give useful advice to alleviate the problem. If the problem imposes high aspiration risks, G tube insertion should be considered. Chewing, drinking fluids, and pureed foods may help. If aspiration occurs, or may have occurred and fever starts, go to an emergency center for treatment.

### ***How should DM patients exercise?***

Overdoing is counter-productive, thus low intensity aerobic training may be useful, such as resistive exercises in water. For more information, read the exercise section of the physical therapy guidelines on the MDF website, <https://www.myotonic.org/sites/default/files/Physical%20Therapy%20FINAL.pdf>

### ***Is there a medication to help with daytime sleepiness?***

Modafinil is a common choice although it is expensive, but can often be covered by insurance when documentation meets the requirements. Sleep apnea contributes to the daytime somnolence but patients often continue to have daytime sleepiness after CPAP.