

MUSCULAR DYSTROPHY

Background

Muscular dystrophy (MD) is a group of more than 30 genetic diseases characterized by gradual and progressive muscle degeneration and weakness. There are several types of MD that differ according to distribution of muscle weakness, severity of symptoms, and age of onset.

In children, the most common form of MD is Duchenne muscular dystrophy (DMD). DMD is caused by a genetic mutation in the gene necessary for making dystrophin, a key muscle protein for stabilizing and protecting muscles during use. DMD causes progressive muscle degeneration including:

- Weakness
- Loss of ambulation
- Motor delays
- Respiratory impairment
- Cardiac dysfunction

The diagnostic process typically begins in early childhood, before age 5, when suggestive signs and symptoms include weakness, delay in walking and clumsiness. A telltale clinical characteristic for DMD is Gowers' sign. Children with DMD find it very hard to get up from a sitting or lying position on the floor. The child will first pull up to their hands and knees before walking their hands up their legs for support as they rise to a standing position.

DMD selectively affects the limb muscles proximally close to the trunk before ones more distal. The legs are affected before the arms. By school age, difficult walking or toe-walking is common. Children frequently protrude their belly to support balance and may also have difficulty raising their arms. Most children will require a wheelchair by the age of 12. By the teen years, the child may require the use of mechanical ventilation related to trunk support, weakening of the diaphragm, and respiratory decline. Subtle effects on cognition and behavior can create varying degrees of learning disability. There is also increased risk for neurodevelopmental problems (e.g., attention deficit hyperactivity disorder, sensory processing disorder).



There is no cure for DMD, but medications are prescribed to help slow muscle loss and increase strength. Side effects of some of these medications, particularly corticosteroids, can affect behavior and academic performance. Management also focuses on preventing and minimizing the risk of progressive contracture and deformity.

Top Takeaways for School Considerations

Duchenne muscular dystrophy (DMD) is characterized by progressive muscle degeneration and weakness.

Muscle damage also causes faster disease progression. The child could require use of equipment including medical devices (e.g., feeding tube, tracheostomy/ventilator) and various assistive and mobility devices (e.g., orthotics, power wheelchair, standers).

DMD is associated with an increased risk for all three types of specific learning disabilities: reading, math, and written communication.

Physical changes can lead to psychological stress disorders such as depression and anxiety. The student, and their peers, should be supported.

Kennedy Krieger Institute's Specialized Health Needs Interagency Collaboration

The Specialized Health Needs Interagency Collaboration (SHNIC) program is a collaborative partnership between Kennedy Krieger Institute and the Maryland State Department of Education.



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Considerations for the Individualized Healthcare Plan (IHP)

- Nursing diagnosis of impaired physical mobility, fatigue, decreased cardiac output and ineffective airway clearance
- Current diagnosed health condition including date of diagnosis, progress of disease process and other chronic health conditions
- Current medication and treatment orders (consider schedule, equipment needs and side effects)
- Respiratory interventions and equipment needs (consider tracheostomy brand/size and downsize, suctioning brand/size, frequency of suctioning, ventilator brand, and settings); note location of suctioning, use of private duty nursing if applicable
- Nutrition interventions and equipment needs (consider brand/size of feeding tube, tube replacement, water flushes, fluid intake goal and supplements); note school district policy on tube replacement and consider keeping backup feeding tube kit at school if applicable
- Use of specialized equipment, adaptive equipment, and orthotics
- Activity, positioning, transferring (consider precautions and/or restrictions)
- Skin check, pressure relief techniques
- Equipment troubleshooting (consider equipment/device user manual, battery, charger)
- Consider emergency care plan(s) (ECP) and emergency evacuation plan(s) (EEP) as related to medical needs in the school setting, and staff education/training, as appropriate

Discussion Starters for Educational Team

1. Has the school staff been trained to implement the student-specific emergency plan?
2. Would the student benefit from evaluations or assessments in any of the following areas: physical therapy, occupational therapy, speech and language therapy, assistive technology, adapted physical education, functional behavior, psychology, hearing and vision?
3. Would the student benefit from additional academic support and/or modified education (e.g., copies of notes, extra time, reduced workload, simplified instructions, alternative formats for presentation of material, 504/IEP)?
4. Would schedule flexibility support the student?
5. Is the physical school environment safely accessible for the student's mobility needs (e.g., entry and exit, ramps, location of classes, access to elevator, doorways)?
6. Does the classroom environment support the student's needs and/or equipment (e.g., desk/seating options, maneuverability space, electrical outlets, flash pass for bathroom or nurse)?
7. Can rest breaks, safe spaces or reduced stimulation times be built into the student's schedule?
8. Does the student need additional adult support to access the academic curriculum in the least restrictive environment?

Resources

Kennedy Krieger Institute: Center for Genetic Muscle Disorders
kennedykrieger.org

Muscular Dystrophy Association (MDA)
mda.org/

Cure Duchenne
cureduchenne.org/



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