MUSCULAR DYSTROPHY

What is it?

Muscular Dystrophy (MD) is a group of more than 30 genetic diseases characterized by gradual, progressive muscle weakness and muscle degeneration. Abnormal gene mutations interfere with the production of proteins necessary to form healthy muscle. There are many types of MD and they differ in terms of distribution of muscle weakness, severity of symptoms, and age of onset. Some types of MD affect children and others do not appear until adulthood.

In children, the most common form of MD is Duchenne Muscular Dystrophy (DMD). It presents in children between the ages of 3-5 years and is more common in boys than girls. DMD progresses rapidly. DMD is caused by a deficiency in dystrophin, a protein that helps strengthen muscle fibers and protect from injury. Weakness commonly begins in the upper legs and pelvis and most individuals with DMD are unable to walk by age 12. They may further require ventilator support. To date there is no cure for DMD but there are medications that can slow the progression of the disease that affects the heart, lungs and muscles.

The other major types of MD include Becker, Emery-Dreifuss, Facioscapulohemeral, Limb-girdle, Myotonic, Distal, Oculopharyngeal, and Congenital. MD is a life-long condition that is not correctable therefore management includes focusing on preventing or minimizing deformities and maximizing the child's functional ability at home and in the community. The child is seen by multiple specialists and therapist to help improve the quality of life.

What are the signs and symptoms?

Affected children will demonstrate a delay in walking, frequent trips and falls, and difficulty with stairs or getting up from a lying down or sitting position. Other symptoms include tip-toe walking, inability to hop or jump, leg pain, facial weakness, inability to close eyes, shoulder or arm weakness, enlarged calf muscles, pelvic weakness, muscle stiffness, and joint contractures.

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. They first pull up to their hands and knees. The child walks his/her hands up their legs to brace themselves as they rise to a standing position.

Another classic symptom of muscular dystrophy is known as the "slip through" symptom. Your child's provider puts their hands around the sides of the child's chest, right up under the arms. As the child is lifted up, the shoulders weaken and move upward, almost allowing the child to slip through your child's provider's hands.

By the age of 12 most children will need a wheelchair for mobility. Progression of the diseases also affects the respiratory and cardiac systems, leading to weakening of the heart muscles and a weakening of the diaphragm and lung muscles making breathing difficult. The child might first need oxygen and eventually will need to be supported by a tracheostomy and ventilator.

A few individuals with DMD will have serious cognitive disability but about one third will have some degree of learning disability. Doctors believe that dystrophin abnormalities in the brain may have subtle effects on cognition and behavior. Learning problems in DMD occur in three general areas: attention focusing, verbal learning and memory, and emotional interaction.

Kennedy Krieger Institute

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

Suggested school accommodations

Students who are affected by MD are able to attend school but will require ongoing accommodations as their condition progresses or they are returning to school after surgeries . Supporting students with this condition in the school require educators and parents/guardian to work as a team. Some accommodations to consider for a 504/IEP could include:

- PT/OT/SLP evaluations
- Modified or flexible school day
- Plan for absences and make-up work
- Adaptive technology
- Additional time for assignments and tests
- Bathroom pass
- Rest period
- Transition time/distance from classes
- Cafeteria assistance
- Preferential seating
- Classrooms close together
- Elevator key if applicable
- Extra set of books for home
- Clear and concise instructions
- Visual schedule
- Comfort of student
- Emotional support
- Staff education/training as appropriate
- Emergency Evacuation Plan (EEP)

Specific health issues for Individualized Healthcare Plan

- Diagnosis including type of MD, age diagnosed
- Child specific characteristics and symptoms
- Up to date medication list including respiratory treatments, PRN medications
- Baseline respiratory assessment including pulse ox parameters
- Nutrition order, oral motor protocol
- Aspiration precautions, position for meals, location of meals and snacks
- Adaptive equipment or orthotics, including hours of wear time and use
- Transfers, equipment involved for safety (slings, lifts)
- User manual for power wheelchair, battery life and/or charge accessibility
- Toileting protocol
- Communicate with school staff, parents/guardian, and provider any changes or concerns about the disease
- Emergency Care Plan(s) (ECP) related to medical needs in the school setting and staff education/training as appropriate for each

Resources & Manuals

Kennedy Krieger Institute: Center for Genetic Muscle Disorders
https://www.kennedykrieger.org/patient-care/centers-and-programs/center-for-genetic-muscle-disorders

Muscular Dystrophy Association (MDA)
https://www.mda.org/

MDA– Recommendations for school accommodations
https://www.mda.org/sites/default/files/Teachers_Guide_PT_flyer.pdf

Nemours– Kids Health
https://kidshealth.org/en/parents/muscular-dystrophy.html

Parent Project Muscular Dystrophy
https://www.parentprojectmd.org/