

# A TEACHER'S GUIDE TO

# NEUROMUSCULAR DISEASE



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**MDA Publications Department**





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## AN INVALUABLE LESSON



Angela Wigglesworth  
Houston  
Klenk Elementary School  
Member, MDA's National Task Force on  
Public Awareness  
Ms. Wheelchair Texas, 2004

Dear Friends and Colleagues,

I've never had a student in my class with a neuromuscular disease. So why, you may ask, am I writing an introductory letter to this informative booklet? Because, although none of my students uses a wheelchair, I happen to roll through the door of my third-grade classroom every day.

When I was 16 months old, doctors told my parents I had spinal muscular atrophy (SMA), and since then I've been living a successful life with muscles that are slowly weakening throughout my body. These weak muscles prevent me from using a chalkboard, opening marker caps, or even tying the shoelaces of a clumsy first-grader tripping down the hallway. Despite these obstacles, the students in my classroom learn an age-appropriate curriculum, and — perhaps more importantly — an invaluable lesson of compassion toward all of humanity.

If you're reading this booklet, you probably have the unique opportunity to teach a child with a neuromuscular disease. Just like me, your student will have limitations. The challenges posed by these limitations will lead you to consult this guide, exercise your teacher's intuition, and even ask the child directly what to do (thus inadvertently teaching the skill of self-advocacy). Regardless of how you solve issues that will come about, I ask that you have as much vision for your student as my teachers had for me.

I recently came across a pile of letters that my third-grade teacher had my classmates write me during one of the many hospital stays of my youth. They are a priceless compilation of overused crayons, misspelled words and sincere get-well wishes. They remind me that I was once a part of a little community that missed my presence, my positive influence and my example that we should always search for the abilities within our disabilities.

Your student with a neuromuscular disease will be all of these things to your classroom community, and you and the other children will be enriched for having known him or her.

As you and your students meet the challenges before you, keep in mind that MDA is always there to help with resources and support. You aren't alone, and — if my life is any proof — your efforts aren't in vain.

With best wishes and gratitude,

Angela Wigglesworth

## BELIEVE THAT ANYTHING IS POSSIBLE



*Amy Dunaway-Haney  
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Teacher of Spanish,  
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Recipient, MDA National Personal  
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Jiffy Lube Excellence in Teaching Award, 2000*

Dear Fellow Teacher,

Schools face ever-increasing pressures to provide safe, enriching environments for children, in which they can flourish into well-adjusted, highly educated citizens of our world.

It's daunting to imagine the funding, training and commitment necessary to individualize a program for every student, regardless of learning style, home environment, learning disability and exceptionality — but that's exactly what we must do as educators. We must look at each of our students as being capable of tremendous success.

I have limb-girdle muscular dystrophy, which first manifested when I was 8 years old. I had difficulty getting up from the floor, and I fell frequently. In fact, my second-grade teacher is the one who finally convinced my parents to seek medical attention, which led to my diagnosis.

I feel blessed to have had some of the best teachers at the public schools I attended in Kettering, Ohio. I was a student during the 1970s and '80s, before special education was a widely known concept. Nonetheless, my teachers encouraged me to develop my talents and get the best education possible. They even modified gym class and other physical activities for me before the law said they had to, because they were dedicated and creative and understood the key to educating kids: Help

them believe they can do anything they set their minds to do.

Now, as a teacher myself, I strive to continue that teaching philosophy. We all must follow a commonsense approach to working with people who have disabilities or other exceptional qualities. The key is not to do everything for them. The key is to help them see that with creativity, a strong will and a good work ethic, anything is possible! As Sharon Christa McAuliffe, teacher and astronaut, said, "Any dream can come true if you have the courage to work for it."

This booklet, along with other educational materials and resources from the Muscular Dystrophy Association, is a good tool designed to help you achieve the daunting — but doable — goal of dedicated teachers everywhere: to provide the best education possible for all children.

Thank you from my heart for everything you do!

Amy Dunaway-Haney



## INTRODUCTION

If you're a teacher of a student with a neuromuscular disease, this booklet will help you better understand the challenges faced by children and adolescents affected by neuromuscular diseases. This guide suggests general strategies to enhance your student's school experience, both academically and socially, and addresses school issues from kindergarten through high school.

Learning about the range of problems associated with neuromuscular diseases can feel overwhelming. The good news is that these students also come with their own unique set of deep strengths. And your role in encouraging, supporting and motivating them draws on many skills you're already using with your diverse students.

In addition, your local MDA office is a valuable resource of information and help. You also can call (800) 572-1717 or visit MDA's website ([www.mda.org](http://www.mda.org)) for free access to a huge library of publications and other material about specific neuromuscular diseases, disability information, research and resources.



## WHAT IS A NEUROMUSCULAR DISEASE?



**N**euromuscular diseases are rare acquired or inherited (genetic) conditions that affect some part of the neuromuscular system:

- the muscles
- the peripheral motor nerves (in arms, legs, neck and face)
- the neuromuscular junction where the nerves and muscles meet
- the muscle-controlling nerve cells (motor neurons) in the spinal cord

More than a million people in the United States are affected by some form of neuromuscular disease, and about 40 percent of them are under age 18.

All neuromuscular diseases are progressive in nature, and all result in muscle weakness and fatigue. Some diseases are present at birth, some manifest in childhood, and others have an adult onset. The disease may be passed down through family genetic lines, and in some cases the student may have an affected sibling, parent or other relative. At other times there isn't a family history and the disease is the result of a spontaneous genetic mutation, an abnormal immune response or an unknown cause.

Life expectancy varies by disease and severity, from very short to normal length. Heart and respiratory problems, which are secondary effects of muscle deterioration, often are the cause of death.

Although muscle wasting isn't painful, the resultant weakness can cause cramping, stiffness, joint deformities, chronic aches and pain, and sometimes the tightening and freezing of joints (contractures).

In almost everyone with a neuromuscular disease, bladder and bowel control are normal, although students may need help in the bathroom as their physical abilities weaken. Sexual functioning and skin sensation usually are normal.

Strengthening exercises don't prevent muscle decline in neuromuscular diseases, and in some cases can further damage fragile muscle cells if done too vigorously. Moderate and light exercise or standing exercises, undertaken under the guidance of a physical therapist (PT) or doctor, may help maintain muscle tone and flexibility, as well as combat obesity and bone thinning. Be sure a doctor or PT has approved your student's exercise plan.

For information about a specific neuromuscular disease, see "Neuromuscular Disease Descriptions," page 24.





## DO NEUROMUSCULAR DISEASES AFFECT THE ABILITY TO LEARN?

Like other students, children with neuromuscular diseases show a range of mental abilities, and many have higher-than-normal intelligence.

However, muscle weakness and fatigue can make it hard for students to keep up with the physical demands of handwriting, completing assignments, organizing materials, etc. In winter, the increased susceptibility to (and life-threatening consequences of) respiratory infections may cause students to miss many days of school.

A few neuromuscular diseases bring a higher-than-average risk of learning disabilities and mental retardation. This may affect learning to read or understanding math concepts, being able to attend to a lesson or comprehending classroom concepts.

Unlike muscle weakness, cognitive effects don't get progressively worse over time. Students benefit from

appropriate accommodations for learning disabilities. For more on cognitive effects, see "Neuromuscular Disease Descriptions," on page 24.

Students with neuromuscular diseases usually qualify for special education services based on their orthopedic disabilities, their learning disabilities or both. Throughout their school careers, most are placed in mainstream classrooms with supports.

These students may have an IEP (Individual Education Program) outlining educational goals, strategies and supports, or a Section 504 accommodation plan.



## HOW ARE NEUROMUSCULAR DISEASES TREATED?



**A**t present, there's no cure for most neuromuscular diseases — although the day is rapidly approaching when genetic and drug therapies will change that situation. For immune-mediated disorders, drug therapies can be very effective for treating or reversing symptoms (see “Neuromuscular Disease Descriptions,” page 24).

Medical interventions have increased the life span and improved quality of life for many medically fragile children. These interventions focus on treating or delaying symptoms, enhancing physical mobility and social interactions, and preventing heart and lung complications.

Some common interventions include:





## ASSISTIVE EQUIPMENT

- Communication devices allow students with weak speech muscles to convey their needs and thoughts.
- Computer adaptations and software allow those with limited movement to type and surf the Internet.
- Small adaptations help with everyday tasks: special feeding utensils and cups, straws, a foam rubber cylinder for grasping pens and pencils, tape recorders, etc.
- Standing frames allow nonambulatory students to continue to bear weight on their legs, promoting healthier bones, better circulation and a straighter spine.
- Transfer boards and mechanical lifts make it easier and safer to move a nonambulatory student.
- Walkers, wheelchairs and foot, ankle and leg braces keep children safely mobile and part of the social scene.

## CARDIAC CARE

When the muscle layer of the heart weakens and doesn't pump effectively (cardiomyopathy), children may experience fatigue and lethargy, swelling in the legs and feet, cold extremities, digestive problems and other symptoms of poor circulation. Drug treatments available through a cardiologist may enhance heart muscle function. Some children benefit from a pacemaker, and some may even undergo a heart transplant.

## NUTRITIONAL SUPPORT

When the muscles used in swallowing and chewing are weakened, there's a risk of dehydration, malnutrition, choking or respiratory infections caused by inhaling food or liquid into the lungs (aspiration).

- Gastrostomy tube (g-tube, feeding tube or PEG) is a tube permanently placed through the stomach wall, so a liquefied diet can be fed directly into the stomach. Those who use a g-tube because eating is difficult and time-consuming still can have food and drink by mouth for pleasure and extra nutrition. But those who use a tube because of choking and aspiration probably shouldn't eat anything by mouth.

## PHYSICAL AND OCCUPATIONAL THERAPY

Keeping the body flexible, upright and mobile combats some of the side effects of neuromuscular disease. Such therapy also benefits academic performance by allowing children to do more and stay healthier. Your student's IEP may specify that a certain time must be set aside each day for him or her to work with a physical or occupational therapist. PT or OT interventions include:

- Range-of-motion exercises and stretches prevent contractures or freezing of the joints of the knees, hips, feet, elbows, wrists and fingers.
- Hand splints keep the wrists and fingers in a good position.
- Swimming pool exercise, especially in a warm pool, is often easier for children with weak muscles.
- Proper body mechanics ensure safe transfers of nonambulatory students by aides, teachers and other caregivers, so neither party gets hurt.



## MEDICATIONS

Although only a few drugs are approved for use against the effects of neuromuscular disease, ongoing clinical trials are constantly seeking to expand that number.

One medication that has proven effective in some neuromuscular diseases is prednisone. This steroid may be taken by children with Duchenne muscular dystrophy (DMD) and some other neuromuscular conditions because it slows the loss of muscle function and increases muscle strength, providing for a few more months to a few more years of leg and arm use.

Prednisone's undesirable side effects include weight gain, loss of bone mass, thinning of the skin, raised blood pressure and blood sugar, depression, and difficulties with thinking, sleeping and controlling behavior.

It's important to carefully monitor the diets of children on prednisone, and to be aware of its behavioral effects.

## RESPIRATORY CARE

When weak respiratory muscles make it difficult to effectively move air in and out of the lungs, children may experience headaches, mental dullness, difficulty concentrating or staying awake, and nightmares. In addition, weak chest muscles make it hard to cough effectively, leaving the lungs more susceptible to infection. In some children, a simple cold can rapidly progress into pneumonia.

- Assisted ventilation: Help with breathing may be given either through an external mask or "sipper" tube, or by way of a tracheostomy, in which a tube is inserted directly into the airway to deliver air to the lungs.
- Cough assist machines: These machines help bring up lung secretions to keep the bronchial system free of infection.

## SURGERY

- Heel cord and foot joint surgeries are used to treat ankle contractures and joint deformities in order to prolong walking. Students usually are out of school less than two weeks.
- Scoliosis surgery is performed on older children and adolescents with serious spinal curvature. Timing of the surgery hinges on the student's growth, lung function, discomfort level and how fast the curve is worsening. Metal rods with hooks are inserted into the spine, and students can spend 6 to 10 weeks out of school. (When possible, these surgeries are scheduled during the summer.)

After scoliosis surgery, students sit much straighter and have fewer problems with breathing. They often require a new wheelchair and adjustments in classroom desk height. Unfortunately, some abilities, such as reaching, may be negatively affected by this surgery.

## HOW DOES A NEUROMUSCULAR DISEASE AFFECT CHILDREN AND FAMILIES?

**B**ecause strong school-family communication and cooperation are essential resources for kids coping with muscle diseases, it's useful to understand some of the day-to-day challenges of neuromuscular disease.

### FAMILY LIFE

Many adults with disabilities credit their success in life to their family's insistence on treating them like other kids, with responsibilities and dreams just like everybody else's.

Neuromuscular disease lays a heavy load on a family medically, emotionally, socially and financially. Parents often feel worried, overwhelmed and isolated, and siblings may feel overlooked and ignored. As the disease progresses, so does parent-child dependency. Family life can revolve around caring for all the child's physical needs, right down to waking every few hours to turn him or her in bed.

Faced with a life-shortening disease, families may focus strongly on the "here and now." They may have little patience with school systems that seem to emphasize discipline and long-term planning. Due to the unusual demands of living with neuromuscular disease, a parent may appear to teachers to be coddling the child, or to be overinvolved in his or her care.





## ELEMENTARY YEARS

Children may be blithely unaware of their neuromuscular disease, or they may express curiosity, anger, guilt, hurt, sadness or fear about the differences they see between themselves and other kids. They may get angry at “my stupid legs” when they fall. Research has shown that many children with serious chronic illnesses experience strong feelings of isolation, inadequacy and worthlessness.

At the same time, children’s natural resiliency allows them to bounce back from these negative emotions, especially if they feel accepted, supported and secure. Helping children verbalize their feelings, focus on their strengths and make friends with peers can enable them to incorporate a realistic awareness of their disabilities into their self-concepts.

Ironically, children who are less disabled by neuromuscular disease can have a tougher time with peers. Children who seem “OK” and don’t use wheelchairs, but whose conditions cause them to be clumsy and weak, often suffer taunts, jibes and accusations of being lazy or not trying hard.

Fatigue is a very real symptom of muscle disease, especially in the afternoons. Teachers should realize that fatigue can masquerade as disinterest or noncompliance. Rest periods and small snacks are ways to address this problem.

## TEEN YEARS

Just as their friends start gaining independence, adolescents with some muscle diseases start losing it. At this most self-conscious age, these teens now may require help with eating and toileting, or need to wear leg braces, or have to ride special buses. As muscle wasting progresses, simple activities become increasingly difficult.

Many teens have a “take me as I am” attitude about their disabilities and participate as fully as possible in school life, finding creative ways to thrive and grow.

But some children withdraw emotionally as they enter their teens, preferring the world of video games, TV and online games to outside society. Others may deny the disease process and struggle on, resisting a wheelchair and pretending nothing is wrong.

If a child show signs of depression, parents should be notified so that appropriate treatment can be sought.



Again, kids with less obvious impairments can take a big hit in middle and high school, enduring brutal bullying and teasing because they look, walk or talk “funny.” One young woman with Friedreich’s ataxia (FA) (a condition that makes muscles progressively more uncoordinated and slurs speech) described herself as an “outcast” in high school, especially when her condition worsened during her junior and senior years, and she fought against using a wheelchair.

“I sometimes wonder if the kids in high school who called me names think of me now. I wonder if they remember how they tripped me, knocked my books out of my hands, slammed my locker shut while I was trying to open it, threw spit wads at me, and hit and bruised my legs,” she wrote at age 26. (“From a Cocoon to a Butterfly in Six Years,” MDA’s Quest magazine July-August 2004; available online at [www.mda.org](http://www.mda.org).)

## USING A WHEELCHAIR

Unless a bone has been broken, the transition from walking to wheelchair use takes place gradually, both physically and psychologically. Each child — and each neuromuscular disease — is unique. Some children never attain the ability to walk, and they use a wheelchair from early childhood. In other cases, the transition to a wheelchair may occur at any age.

When students get to the point of falling frequently, schools sometimes require them to use their wheelchairs at school in the name of safety, even though the student vigorously insists he or she wants to walk. There are advantages to walking for as long as possible, provided it’s safe to do so. Walking promotes bone strength, circulation and more similarity to peers. Using a

wheelchair full time at school may make it harder for students to maintain walking ability at home.

Many families let the child set the timetable for using a wheelchair, while making accommodations to minimize falling. At school, these can include providing help carrying books and lunch trays, allowing extra time to navigate the halls, appointing a designated runner during sports, and planning for classroom and playground safety.

Ironically, once children start to use a wheelchair full time, they often find they regain mobility, energy, freedom and confidence. To facilitate this transition, it’s vital that the school environment be wheelchair friendly and that teachers and school personnel accept the change positively.

# HOW WILL A STUDENT WITH A NEUROMUSCULAR DISEASE AFFECT MY CLASSROOM?



A strong family-student-teacher team is the best approach to managing neuromuscular disease's effect on education. The ever-changing symptoms of these diseases require ongoing communication between school and home, so problems can be quickly solved or prevented from arising.

## THINK BIG

Generally, even in cases of severe disability, it's best to educate students for a life of independence and self-support. Most of these children will grow up to have careers, families and contributing roles in their communities.

Increasingly, young adults with neuromuscular diseases graduate from college and want jobs. With an almost 70 percent unemployment rate among people with disabilities, a good education is an essential success tool.





## COMMON AREAS OF DIFFICULTY

Although each student has different needs, here are a few areas that may require particular attention and awareness:

- keeping up with classwork
- keeping up with homework
- difficulty with written language and writing tasks
- fatigue
- difficulty paying attention and concentrating
- math and reading difficulties
- participating in classroom activities
- participating in physical education
- being pulled away from academic instruction for therapy
- physical positioning and special seating
- health concerns, such as respiratory infections
- social skills and peer relations
- physical accessibility of the school campus, including doorway widths, stairs, heavy doors, water fountains, distances between classes, curbs, aisles between desks and accessible bathrooms

## COMMON ACCOMMODATIONS

These areas should be addressed in consultation with family, medical experts and school staff during the IEP process or as they arise:

- special transportation
- physical or occupational therapy
- medical care and avoidance of infection
- curriculum modifications
- provision of aides and note takers
- tutoring
- untimed tests
- adapted keyboards, software and calculators
- adaptive physical education

- rest time
- bathroom assistance
- field trip assistance
- extra time to pass between classes
- an extra set of textbooks to keep at home, to minimize carrying a heavy load
- classroom and school campus accessibility modifications
- help making/accessing friends and social relationships
- putting children in charge of their own care, especially as they reach middle school and high school — i.e., directing their aides, requesting accommodations, advocating for their needs, making choices and decisions — to help them develop emotional and social independence as their physical dependence increases
- allowing alternative ways to demonstrate understanding of a concept, such as making an oral rather than a written report
- assigning work buddies and using computer-assisted learning
- using special education strategies to compensate for learning, memory and language-processing difficulties
- implementing homebound (at-home) instruction for medically fragile children that features consistent support, communication, follow-through and high expectations
- inclusion of students with disabilities in social and extracurricular activities
- emphasizing the student's strengths and abilities, not just focusing on disabilities



## WHAT SHOULD I SAY TO THE STUDENTS ABOUT THE NEUROMUSCULAR DISEASE?

Children usually are best able to cope with their disease and treatment if they know as much as possible about it. Medical personnel usually tell parents and the child about the diagnosis, methods of treatment and progression, and some families go on to do their own research.

However, it isn't prudent to assume the child has been told everything about his or her disease. Some parents don't address such topics as life expectancy and increasing disability until the child asks about them. Talk to parents about the approach they're taking to informing the child, so you can support it at school.

When a teacher shows sincere interest and feels comfortable speaking with the child about his or her abilities and challenges, the child likely will feel comfortable asking questions and expressing feelings.





Accurate information also enables peers to cope with a classmate's disease. (However, sometimes a child is reluctant to have the disease discussed, and some parents may be hesitant for others to know about it. Discuss this preference with the family before giving information to classmates.)

It can help a class to know about the type of neuromuscular disease that affects their classmate; the nature of treatments such as physical therapy and prednisone; and the ways the disease may affect a child's appearance, abilities and behavior. With accurate knowledge, peers are less likely to tease and more likely to defend their classmate when teasing does occur.

When a student with a neuromuscular disease misses a lot of school due to surgery or illness, maintain his or her social connection with the class by having students send notes, get-well cards and other friendly tokens.

## DISABILITY EDUCATION ACTIVITIES

Be sure to discuss this topic with the child's family first, and invite them to take part if they wish.

Some classroom activities can enhance your students' knowledge of disabilities and their acceptance of people who are "different."

- experiential presentations (for example, a wheelchair obstacle course to show the importance of an accessible environment)
  - group discussions of stereotypes and attitudes toward people with disabilities
  - a lesson on "person first" language when speaking about disabilities (i.e., saying "a boy with a disability" instead of "a disabled boy" puts the person first, not the disability)
  - awareness-raising projects, such as walk-a-thons or carnivals, that also can be school fundraisers for MDA
- 
- a presentation by the student (and family members) about the disease and how he/she handles it
  - lectures and presentations of various types of adaptive equipment (your local MDA office may be able to help you with this)



## TEACHER TIPS

When teaching students with neuromuscular diseases, remember that:

- They face physical, academic and social challenges.
- Fatigue plays a big part in the student's school day.
- Learning disabilities are more prevalent in some neuromuscular diseases, but most students with these diseases have the normal range of intelligence.
- Students may need alternative methods (besides written work) of demonstrating academic comprehension.
- With imagination, almost any activity can be adapted to be accessible.
- Helping students make friends enhances their school experience and performance, and cuts down on bullying and teasing.
- Teachers should maintain high expectations of the student's school performance, no matter how severe the disability.
- Informed, cooperative, accessible schools and strong family-school teams are vital to helping students overcome challenges and get the most out of their educational experience.

As you encourage your student to develop his or her strengths, also remember to draw on your own strengths as a teacher: Be creative, empathetic, firm, flexible and — as much as possible — treat your student with neuromuscular disease just like everybody else.





## NEUROMUSCULAR DISEASE DESCRIPTIONS

**B**elow is a general overview of the characteristics of the neuromuscular diseases that affect children and teens. The disorders are grouped into six categories. For more detailed information about a specific disease, check out the Muscular Dystrophy Association's free "Facts About" disease booklets. The booklets are available online at [mda.org](http://mda.org) or through your local MDA office.

### **MUSCULAR DYSTROPHIES** (*involving the structure of the muscle cells*)

#### **BECKER (BMD)** • *Age of Onset: 2 to 16 years*

*Characteristics:* A milder, more slowly progressing form of Duchenne MD (see below).

#### **CONGENITAL (CMD)** • *Age of Onset: Birth*

*Characteristics:* Generalized muscle weakness with possible joint deformities. Progresses very slowly.

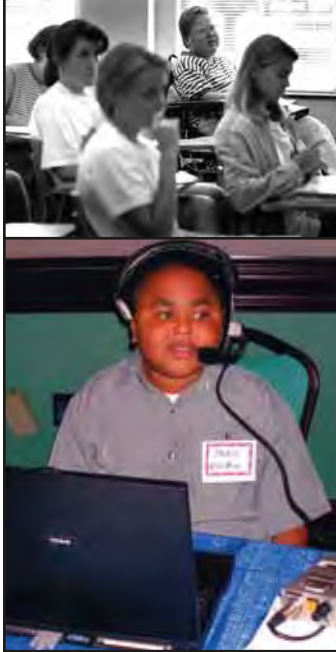
*Possible cognitive effects:* Some of the most serious brain effects in neuromuscular diseases are found among people with CMD, although not everyone is affected. Children with structural brain abnormalities and those with seizures are most at risk for a wide range of problems, from learning disabilities, to vision and reading difficulties, to severe mental retardation.

#### **DUCHENNE (DMD)** • *Age of Onset: 2 to 6 years*

*Characteristics:* General muscle weakness and wasting, beginning in upper arms and legs and eventually involving all voluntary muscles. DMD affects mainly boys but in rare cases may affect girls, who have a slower and less severe progression.

Boys in the primary grades may run more slowly, have trouble walking long distances, difficulty climbing stairs and getting up from the floor. By age 10, boys are likely to be using a wheelchair at least part-time, and their arms are weakened. Around age 15, the arms, legs and torso all are affected and wheelchair use usually is full-time. The student may need help writing and lifting, and may show early signs of respiratory and heart weakness.

*Possible cognitive effects:* About a third of children with DMD have some degree of learning disability, especially in three areas: attention focusing, verbal learning and memory, and emotional interaction.



Sometimes this impairment is mistaken for attention deficit disorder. DMD sometimes causes children to have poor social skills, be emotionally distant and moody, or inappropriately impulsive and lacking good social boundaries.

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**EMERY-DREIFUSS (EDMD) • Age of Onset: Childhood to early teens**

*Characteristics:* Weakness and wasting of shoulder, upper arm and shin muscles. Joint deformities are common, and heart complications can be serious.

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**FACIOSCAPULOHUMERAL (FSH) • Age of Onset: Childhood to early adulthood**

*Characteristics:* Childhood onset causes more severe symptoms than adult onset. Weakness and wasting affect face muscles, speech, eyelids, shoulders and upper arms. Progresses slowly with periods of rapid deterioration.

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**LEMB-GIRDLE (LGMD) • Age of Onset: Childhood to middle age**

*Characteristics:* Muscle wasting begins in the shoulder and pelvic girdles. Scoliosis and heart-lung problems are common. Progression rate varies. Therapy helps maintain mobility and avoid respiratory illness.

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**MYOTONIC (MMD) (STEINERT DISEASE) • Age of Onset: Birth to early childhood**

*Characteristics:* An inability to relax muscles (myotonia), combined with muscle weakness. Affects face, feet, hands and neck first. Progression is slow.

*Possible cognitive effects:* When MMD appears in infancy or childhood, about 75 percent of children have mental retardation, as well as severe facial weakness and speech abnormalities. Later-onset MMD (adolescence through adulthood) isn't as closely associated with mental retardation, but may cause teens to be overly sleepy during the day and to lack initiative and seem apathetic.

Medication helps students stay more alert, as does addressing any underlying respiratory or heart problems.

## **PERIPHERAL MOTOR NEURON DISEASES**

*(involving muscle-controlling nerve cells of the arms, legs, neck, face)*

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**CHARCOT-MARIE-TOOTH (CMT) DISEASE • Age of Onset: Childhood to young adulthood**

*Characteristics:* Weakness and atrophy of muscles and nerves of the arms from the elbows down and legs from the knees down. May involve foot deformities and some numbness. Ankle sprains are common. About



10 percent of children experience muscle cramping or burning nerve pain. Children may need leg braces, wrist braces and/or surgery, and may use a wheelchair for mobility.

### **DEJERINE-SOTTAS (DS) DISEASE** • *Age of Onset: Infancy*

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*Characteristics:* Slow development of early motor skills, leading often to loss of skill. Hands and legs are weak and may have impaired sensation. Severity and progression vary.

### **FRIEDREICH'S ATAXIA (FA)** • *Age of Onset: 7-13 years*

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*Characteristics:* Symptoms include shaky movements, lack of coordination, poor balance, slurred speech, muscle weakness and loss of sensation. Severity and progression vary. Often associated with diabetes and heart disease.

## **MOTOR NEURON DISEASES**

### **(involving nerve cells in the spinal cord)**

### **INFANTILE PROGRESSIVE SPINAL MUSCULAR ATROPHY (SMA TYPE 1) (WERDNIG-HOFFMANN DISEASE)** • *Age of Onset: Birth-6 months*

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*Characteristics:* Generalized muscle weakness, trouble swallowing and sucking, breathing distress, paralysis of legs and arms. Death often comes in very early childhood, but medical technology is expanding life span.

### **INTERMEDIATE SMA (SMA TYPE 2)** • *Age of Onset: 6 months-3 years*

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*Characteristics:* Weakness in arms, legs, upper and lower torso, often with skeletal deformities. Lung disease is common. Rapid progression. Survival into early adulthood is common but respiratory problems are a constant threat.

*Possible Cognitive Effects:* Although not scientifically validated, high intelligence often is noted in people with SMA.

### **JUVENILE SMA (SMA TYPE 3) (KUGELBERG-WELANDER DISEASE)** • *Age of Onset: 1-15 years*

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*Characteristics:* A milder form of intermediate SMA, with slower progression. Weakness in leg, hip, shoulder, arm and respiratory muscles. Calf muscles often are enlarged. A wheelchair may not be required in youth.



### **SPINAL-BULBAR MUSCULAR ATROPHY (SBMA) (KENNEDY'S DISEASE) • Age of Onset: 15-60 years**

*Characteristics:* Occurs only in males, causing weakness in limbs and muscles involved in talking, chewing and swallowing. Some males experience breast enlargement. This disease progresses very slowly.

## **NEUROMUSCULAR JUNCTION DISEASES**

***(involving the site where nerves and muscles meet)***

### **CONGENITAL MYASTHENIC SYNDROMES (CMS) (SOMETIMES DIAGNOSED AS MYASTHENIA GRAVIS)**

*Age of Onset: Infancy to childhood*

*Characteristics:* Generalized weakness and fatigability of voluntary muscles, including those controlling mobility, eye movement, swallowing and breathing. Rest can help restore strength. Varies in severity and weakness can fluctuate. May be controlled with medication.

## **MYOPATHIES**

***(involving tone and contraction of muscles controlling voluntary movements; may include inflammation of muscles or related tissues)***

### **CENTRAL CORE DISEASE • Age of Onset: Birth to infancy**

*Characteristics:* Slow development of motor skills. Hip displacement common.

### **DERMATOMYOSITIS • Age of Onset: Childhood to age 60**

*Characteristics:* Symptoms include skin rashes, muscle pain and tenderness, fever, gastrointestinal distress, and progressive weakness, especially affecting the shoulders, upper arms, hips, thighs and neck muscles. Swelling of the upper eyelids also is common. Hard painful nodules may appear under the skin. Progression and severity vary by individual. Corticosteroid drugs and restricted diet may result in remission.

### **HYPERTHYROID/HYPOTHYROID MYOPATHY • Age of Onset: Childhood to adulthood**

*Characteristics:* Weakness in arms and legs. Stiffness and cramps common. Severity depends on success in treating underlying thyroid condition.

### **MYOTONIA CONGENITA • Age of Onset: Infancy to childhood**

*Characteristics:* Muscle stiffness and difficulty moving after periods of rest. With exercise, muscle strength and movement may return to normal.



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**MYOTUBULAR MYOPATHY (CENTRONUCLEAR MYOPATHY) • Age of Onset: Birth to infancy**

*Characteristics:* Drooping of upper eyelids, facial weakness, foot drop and some weakness of the limbs and trunk. Individuals usually have no reflexes. Slow progression.

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**NEMALINE MYOPATHY • Age of Onset: Birth to infancy**

*Characteristics:* Low muscle tone and weakness of arms, legs, trunk, face and throat muscles. Severe cases have respiratory weakness.

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**PARAMYOTONIA CONGENITA • Age of Onset: Childhood to early adulthood**

*Characteristics:* Muscle stiffness and difficulty relaxing muscles, especially after repeated use or exercise.

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**POLYMYOSITIS • Age of Onset: Childhood to age 60**

*Characteristics:* Weakness of neck and throat, shoulder, hip and thigh muscles, and generalized muscle swelling. Swallowing difficulties are common. Severity and progression vary. Corticosteroid drugs may help.

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**METABOLIC DISEASES OF MUSCLE**

**(involving errors in metabolism in producing energy in muscle cells)**

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**ACID MALTASE DEFICIENCY (POMPE DISEASE) • Age of Onset: Infancy to adulthood**

*Characteristics:* For infants, the disease is generalized and severe, impairing the heart and liver. Later-onset forms involve weakness of mid-body and respiratory muscles. Progression varies.

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**CARNITINE DEFICIENCY • Age of Onset: Early childhood**

*Characteristics:* Varied weakness of shoulder, hip, face and neck muscles. Often occurs with other metabolic conditions. Progression varies. Carnitine supplementation can be effective.

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**DEBRANCHER ENZYME DEFICIENCY (CORI OR FORBES DISEASE) • Age of Onset: 1 year**

*Characteristics:* General muscle weakness, poor muscle control and an enlarged liver with low blood sugar. Slow progression.

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**MITOCHONDRIAL MYOPATHY • Age of Onset: Early childhood to adulthood**

*Characteristics:* Severe muscle weakness. Progression and severity vary. In some cases the brain is involved, causing seizures, deafness, loss of balance and vision, and mental retardation. Other systems in the body also can be affected.



**Possible cognitive effects:** Some children have impaired cognition, especially if they experience seizures, strokes or high levels of lactic acid in the blood. But others have high intelligence, such as the late MDA National Goodwill Ambassador Mattie J.T. Stepanek, who was a New York Times best-selling poet.

**PHOSPHORYLASE DEFICIENCY (McARDLE DISEASE)**  
**PHOSPHOFRUCTOKINASE DEFICIENCY (TARUI DISEASE)**  
**PHOSPHOGLYCERATE KINASE DEFICIENCY**  
**PHOSPHOGLYCERATE MUTASE DEFICIENCY**  
**LACTATE DEHYDROGENASE DEFICIENCY**

*Age of Onset: Childhood, adolescence or adulthood*

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**Characteristics:** Children with these disorders may not appear to be impaired until they exert themselves physically, and so often are unfairly thought to be lazy. These metabolic conditions cause a low tolerance for exercise, with symptoms including cramps, muscle pain and weakness, nausea, vomiting, muscle damage and discoloration of the urine (due to muscle breakdown).

Rest usually helps restore strength. Severity varies, increasing with age. Children often are advised to avoid strenuous exercise.

## HELP FROM MDA

The Muscular Dystrophy Association can help teachers in several ways.

You can call your local MDA office to arrange for an MDA speaker to visit your classroom or school.

Or, you can learn more from an MDA video or publication about living with neuromuscular disease.

- Preparing for Emergencies
- Quest Magazine
- Travis: I Got Lots of Neat Stuff

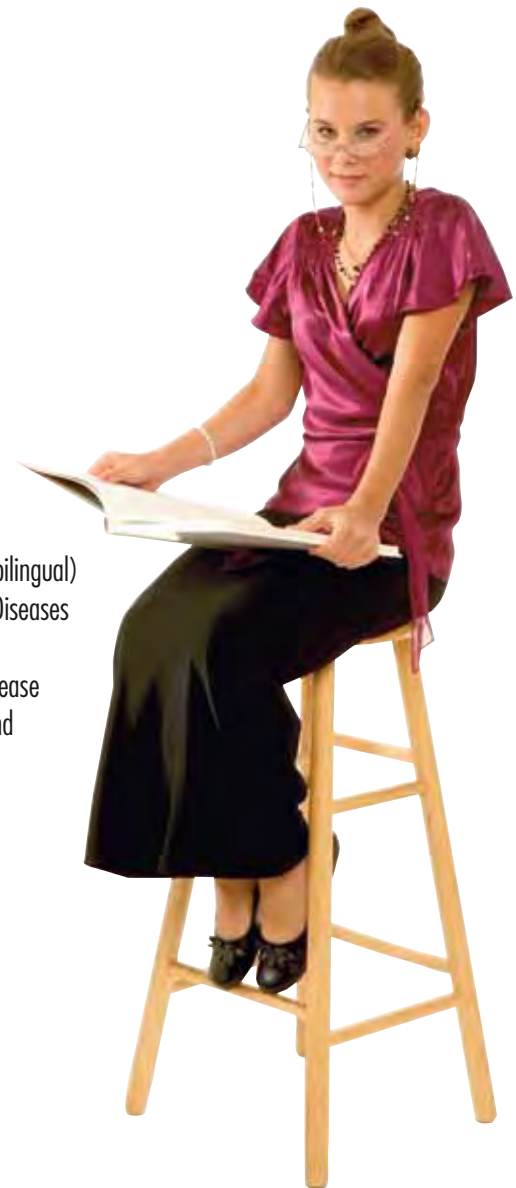
Just call your local MDA office for these materials, or visit the MDA website at [www.mda.org](http://www.mda.org).

### VIDEOS INCLUDE:

- Breathe Easy
- Family profiles
- Hispanic family profiles
- Hop-a-Thon Stories
- MDA Summer Camp
- Sarah's Wish
- Standing and Walking are Gifts
- Videos on MDA programs

### PUBLICATIONS INCLUDE:

- Breathe Easy: Respiratory Care for Children With Muscular Dystrophy
- Everybody's Different, Nobody's Perfect (bilingual)
- Facts About Genetics and Neuromuscular Diseases
- Hey! I'm Here, Too!
- Learning to Live with a Neuromuscular Disease
- MDA Services for the Individual, Family and Community



## A TEACHER'S GUIDE TO NEUROMUSCULAR DISEASE



[www.mda.org](http://www.mda.org)  
(800) 572-1717