Duchenne: An overview for Educators

Duchenne muscular dystrophy is a genetic linked neuromuscular disease which causes progressive loss of muscle function and independent functioning. It results from a defective gene responsible for producing the key muscle protein, dystrophin. Children with Duchenne muscular dystrophy are usually diagnosed before the age of five. The disease leads to a loss of muscle mass, breathing difficulties and decreased heart function. Muscles are prone to extensive damage when overused and fatigued. This muscle damage causes the disease to progress faster.

Duchenne muscular dystrophy, classified as a rare or orphan disease, affects approximately one out of every 5,000 boys. Currently, there are 15,000 individuals living with Duchenne muscular dystrophy in the U.S. alone and 300,000 worldwide. About a third of individuals living with Duchenne muscular dystrophy have some degree of learning difficulty, perhaps because of dystrophin abnormalities in the brain.

Duchenne muscular dystrophy can sometimes be referred to as a “hidden” disease as its progression is not always seen with the naked eye. Often, observed behavioral changes, misconstrued as defiance, are a result of the weakened physical and mental muscle function.

Typical Progression

Early Ambulatory	Mid-Ambulatory	Late Ambulatory/Transitioning	Non-Ambulatory
Moves slower than same age peers	Increased difficulty walking	Increased fatigue	Increased breathing difficulties
Appears clumsy/falls often	Weakened trunk area	Continued progression of loss of muscle function	Decreased heart function
Enlarged appearance of calves	Awkward gait	Activities involving upper body and lower body will require assistance	Requires use of a wheelchair for mobility
Energy conservation throughout the day is important	Difficulty rising from the floor	Walking has become unsafe and cumbersome	Reliance on assistive technology and adaptive equipment
	Necessary to begin assistive technology plan		

Treatment

While there is no known cure for Duchenne muscular dystrophy, there are standards of care across the U.S. being utilized to lessen symptoms and improve quality of life. Corticosteroids are used to slow muscle loss and increase strength, however, there are side effects that may impact behavior and academic performance. Psychological effects can include but are not limited to: difficulty with concentration, sleeping and controlling emotions. Additionally, impairments in reading, thinking, and coping skills can lead to depression and/or aggression.

Accommodations and Modifications for Energy Conservation

Cure Duchenne™ CARES

Advanced Planning

- Do not begin an activity that cannot be stopped
- Help in gathering all material prior to starting a project
- Build in rest breaks into daily schedule. Sometimes a few minutes will be enough whereas, at other times, a longer nap may be required
- Avoid rushing
- Schedule adequate time for activities
- Alternate tasks throughout the day- balance between level of physical intensity (low/high) For example, first snack time then PE
- Build in time for campus transitions. If ambulatory, allow for rest stops along the route
- Avoid stairs as stairs increase the stress on the muscles and hasten muscle damage
- Avoid overscheduling activities between home and school (maintain communication lines). When scooter/wheelchair is part of equipment in use, utilize this for distances over 100 feet

Classroom Set-Up

- Adjust workstation so that elbows, hips, knees, and ankles are positioned at 90 degrees
- Keep items within easy reach to avoid over-reaching and bending
- Keep heavier items at arm level
- Place desk straight in front of instruction (Avoid side-facing)
- Use of a chair instead of sitting on the floor. Consider having all students in chairs so as not to isolate the individual with Duchenne

Set Priorities

- Reduce tasks that are not necessary
- Eliminate tasks that involve busy work, i.e. homework
- Allow oral test taking instead of written to aide with energy conservation
- Stop an activity BEFORE becoming exhausted

Eliminate Extra Physical Effort

- Have child sit in a chair instead of on the floor
- Decrease the weight of items carried, i.e. lunch box, backpack
- Use adaptive equipment, i.e. rolling backpack
- Use pencil grips or thick pencils/pens to avoid application of excessive pressure
- Encourage good posture, i.e. use back supports on chairs
- Encourage sliding objects when possible to avoid lifting/carrying
- Consider assigning a buddy to help, i.e. to pick up items from the floor; tying shoes

Classroom Challenges

Common accommodations for individuals with Duchenne muscular dystrophy

Cure Duchenne™ CARES

Rising from a seated position

- Ensure that the individual has access to help always
- Either provide a hand or ensure that the individual has something to grab onto
  ***Especially important in the bathroom**
- Consider this challenge in the classroom, across the school campus, on field trips, etc.

Rising from the floor

- Consider eliminating and/or limiting the number of activities on the floor
- It is exhaustive for individuals with Duchenne to stand up from a seated position on the floor
- For activities conducted on the floor, the individual with Duchenne should be seated in a chair with arms
  ***Consider inclusion by restructuring the lesson, so some or all students sit in a chair**

Picking up objects off the floor, tying shoes, etc.

- Keep in mind that it is easy for an individual with Duchenne to lose balance without any warning, as though a rug was just pulled from under the legs
- Consider assigning a nearby buddy to help when needed

Walking long distances

- AVOID long distance walking when possible
- When walking for campus transitions, allow additional time and take rest stops as needed
- If the individual has a scooter/wheelchair, it is recommended to use this means of mobility for long distance transitions

Stairs

- AVOID use of stairs always
- Stairs increase the stress on muscles and can hasten muscle damage
- Use elevators and ramps instead of stairs

Recess and Physical Education

- Muscle weakness will contribute to difficulty in balance and climbing
- Collaborate with the school Physical Therapist/Adaptive Physical Educator to assist with accommodating/modifying the individual's recess and physical education plan

Activity Log

Date:	Activity/Seating: Manual/Power Chair Classroom Chair Floor	Power Chair Used for Campus Transitions? Yes or No	How do you feel?	Adult Check-In
Homeroom (Time Frame)	Activity: Seating:	YES NO		
Writing (Time Frame)	Activity: Seating:			
Reading (Time Frame)	Activity: Seating:			
Snack (10 minutes)				
Related Arts (Time Frame)	Activity: Seating:	YES NO		
Math (Time Frame)	Activity: Seating:			
Lunch (Time Frame)	Lunchbox: Buy:	YES NO		
Recess (Time Frame)	Activity: Seating:	YES NO		
Science (Time Frame)	Activity: Seating:			
Dismissal		YES NO		
End of Day	Reward:	Reward Earned? YES NO		

Activity Log Directions

6-Point Scale for Fatigue and Pain Measurement

Wong-Baker FACES Pain Rating Scale

 0 NO HURT	 2 HURTS LITTLE BIT	 4 HURTS LITTLE MORE	 6 HURTS EVEN MORE	 8 HURTS WHOLE LOT	 10 HURTS WORST
I feel good 0	I am a little tired, but I can still participate 2	I can do most activities with some rest breaks 4	I can participate in some activities with many breaks 6	I am unable to participate in most activities even with breaks 8	I am unable to participate in any activities because I am exhausted 10

The initial set up of the Activity log includes inputting the specific information relative to the individual child's schedule. The column entitled: "Power Chair Use" can be deleted if the child is not currently in need of this device. If the child is using alternate means of mobility, the child's team can determine the best use of this column as related to energy conservation. The "Reward" can be deleted as well should there not be a need for positive reinforcement for the use of the log. Determination of its use is based upon the present needs of the child by his/her school-based team. The columns entitled: "How do you feel?" and "Adult Check-In" are to remain as is.

An example row entry for the Activity Log

Homeroom **8:00-8:20 am**	Activity: **Morning Work** Seating: **Classroom Chair**	YES NO	Wong-Baker FACES Pain Rating Scale  0 NO HURT · 2 HURTS LITTLE BIT · 4 HURTS LITTLE MORE · 6 HURTS EVEN MORE · 8 HURTS WHOLE LOT · 10 HURTS WORST	Adult Check-In: **Teacher**

Step 1: Once the log is customized to the child and ready for implementation, it is suggested to complete the log for 3-5 consecutive school days.

Step 2: At the beginning of the school day, if the "reward" system is being utilized, the child selects the end of the day reward. The name of the reward is entered onto the log where indicated. The child then determines present level of energy/pain based on the 6-point fatigue and pain management scale rating where indicated. Should the "reward" system not be utilized, the only item to complete at the start of each school day is the energy/pain scale.

Step 3: Assessment of the child's energy/pain is captured where indicated throughout the day using the printed scale within the log.

Step 4: Upon completion of the pre-determined initial time frame (i.e., 3-5 consecutive school days), the child's school-based team (including parent) is recommended to review the completed logs to determine if there are any trends in the level of fatigue. Adjustments to the child's schedule should be made to assist with reducing the tasks/activities/seating/time allotment that may be contributing to increased fatigue levels.

Step 5: The activity log, once adjusted from the initial implementation, should be reviewed and modified as needed at a pre-determined frequency such as every 4-6 weeks.