Parent Project
Muscular Dystrophy

LEADING THE FIGHT TO END DUCHENNE

OUR MISSION

To improve the treatment, quality of life, and long-term outlook for all individuals affected by Duchenne muscular dystrophy through research, education, advocacy, and compassion.

Parent Project Muscular Dystrophy
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Learning and Behavior in Duchenne Muscular Dystrophy

for parents and educators

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contents

HOW TO USE THIS GUIDE:

You are probably reading this because you are concerned about your son's behavior or learning, or because you want to be proactive in making his academic career as positive as it can be. In either case, you are looking for information, guidance and assistance on how to best serve his needs so that he has the same opportunities for success as his peers. The purpose of this module is to provide specific information on learning and behavioral issues that may arise in the classroom and at home.

You should not automatically assume that all of the information in this module will be relevant to your child. Like any other group of children, among children with Duchenne you'll find very different temperaments, behavioral patterns, and learning styles. Our hope is that this module will increase awareness of potential problem areas, so that any difficulties can be identified early and addressed quickly. Research, clinical, and educational experience provide clues about behavior and learning issues that are more common in boys with Duchenne. This module reviews those issues, and potential interventions to improve your boy's experience in the classroom (and maybe at home).

The best way for your child to have his needs met is for parents and professionals to come together in a collaborative manner. Please use this module, together with the other Education Matters materials, as a starting point for assessing the needs of every young person with DMD and as a means of enhancing communication between everyone involved.

After you read this guide and gather other information, consider setting up a meeting between parents and teacher (and other school personnel). This should ideally take place before the school year starts. During the meeting, parents should take time to review relevant behavioral or learning issues with the teacher. Point out relevant learning and/or behavior issues and discuss how they are relevant to your child. If your child has multiple needs in the classroom, consider working with the teacher to prioritize what should be addressed first.

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Section 1:

Introduction to Learning and Behavioral Concerns More Common in Children with Duchenne Muscular Dystrophy

In this section:

Dystrophin is usually present in the brain as well as in the muscles, and scientists think that when dystrophin is missing (as in DMD), brain neurons may not function as efficiently as they should. This is probably why there is an increased chance for developmental, cognitive, learning, and behavioral difficulties among boys with DMD. Not every child with DMD is going to have problems with learning or behavior.

1.1

Learning and Behavior Summary

1.1 Learning and Behavior Summary

- Similar to other kids, boys with DMD can have a broad range of IQ scores (including well above average), and most boys are in the "normal" range of intellectual and learning ability. However, boys with DMD are more likely to have somewhat lower IQ and academic test scores than other children their age. There is an increased, though low, risk of mental retardation (which means an IQ below 70 and a reduced ability to live independently).

- Problems with short-term verbal memory (also called working memory) are the most common finding, and can be present even in children with a high IQ. This has an impact on the amount of information boys can take in and hold in mind at any one time.

- Language development can be problematic for children with Duchenne, including language comprehension and expression. Basic vocabulary is less likely to be affected.

- Weaknesses in phonological (the distribution and pattern of speech sounds) awareness/processing, combined with problems in short-term verbal memory, can increase the chance for a learning disability. This is probably why the rate of dyslexia (reading problems) may be as high as 40% in boys with Duchenne.

- Writing and mathematics disorders are less well studied, but evidence suggests that they can also be problematic.

- Boys with Duchenne may have problems with motor planning, which is separate from issues related to their muscle weakness. Problems with motor planning may cause a child to appear clumsy, uncoordinated, or as having poor hand-eye coordination.

- There is also an increased chance for neurodevelopmental problems such as attention-deficit/hyperactivity disorder (AD/HD), autistic spectrum disorders, oppositional/defiant or aggressive behavior, and sensory processing disorder.

- Some boys may appear to have obsessive-like features, and may have difficulty with mental flexibility (being able to shift their thought process to accomodate changes in expectations or circumstances).

- Boys with Duchenne also have an increased chance for emotional problems such as anxiety or depression, similar to what is seen in other chronic medical conditions.

- About 1/3 of boys with Duchenne may have significant social problems (e.g., making and maintaing same-age friendships, social maturity, etc.). This may be due in part to physical limitations, but may also be because of cognitive weaknesses such as language problems, impulsivity/hyperactivity, or deficits in social judgement, social reciprocity, and perspective taking.

The following section is broken into age and grade categories.

1.2

Cognitive & Behavioral Issues at Different Stages

pre-k, kindergarten & early elementary (ages 1–7)

Cognitive:

Parents and teachers may notice concerns about language development during this age period. The problem may be obvious, such as a lack of basic vocabulary, or may be subtle, in that they have a normal vocabulary but problems remembering or processing larger amounts of verbal information or expressing themselves. Weaknesses in phonological awareness and verbal memory place boys with Duchenne at increased risk for developing learning disabilities, especially dyslexia.

Behavior:

Problematic behaviors are quite common in all preschool children, and boys with DMD are no different. However, young boys with DMD may have more difficulty with impulsivity and emotional control than other children their age. They are also more likely to be rigid and inflexible in their thinking, which can result in noncompliance or arguing. They may have difficulty making transitions. They may also be taking steroid medication with side effects that can impact their behavior, such as making them more emotional and active than normal.

elementary & middle school (ages 7–11)

Cognitive:

Difficulty mastering academic material is a frequent concern during the primary years. Academic difficulties may be due to specific learning disabilities such as dyslexia or other cognitive deficits. Problems with short-term memory and attention may be problematic for boys with DMD at this stage, but may be overlooked by parents and teachers because they are not disruptive. As boys progress through the grades and more and more independence is expected in their work, problems with planning, organization, initiation and self-evaluation may become more apparent (these types of skills are collectively referred to as "executive skills").

Behavior:

Children with DMD tend to become more aware of their differences and limitations during this stage. Although most boys adjust well to their condition over time, there may be times of emotional distress because of DMD. Boys age 8 to 10 years may be most likely to have adjustment problems, as this is usually the time just prior to transition to regular use of a wheelchair.

junior high & high school (ages 12-18)

Cognitive:

As young men with DMD grow older and expectations for responsibility increase, problems with short-term memory and executive functioning can interfere with their ability to keep track of and efficiently complete assignments and projects. This is likely to become more problematic as the complexity of their work increases, and for this reason learning problems can happen for the first time during this age range.

Behavior:

As with any child, adolescence may be a difficult time for young men with DMD. Physical limitations and disruption in physical development that may occur with some medical treatment (e.g., short stature or delay of puberty from steroid treatment) may make adolescence more problematic. This can also be a difficult time for teens who are unable to establish their independence because they require more care and assistance from others, such as parents. As muscle weakness progresses, they are at risk for becoming more isolated or socially withdrawn. Parents and teachers should look for signs of chronic sadness, depression, or anxiety.

1.3

Assessment

1.3 Assessment

Given the increased risk of cognitive and emotional problems in DMD, we suggest early assessment and intervention. Some assessments should be done for all children with DMD (as described on right). Other assessments can be done if concerns arise. Given the complex nature of DMD, assessments should be done by people with expertise that is appropriate to the methods of assessment. Sometimes the expertise offered by your local school isn't enough for your child's initial assessment. If possible, it is best to visit a child neuropsychologist, but other child psychologists also offer appropriate assessment services.

Parents and Teachers – Please raise any concerns that you might have about the child's development, cognitive ability, academic progress, emotional adjustment, or behavior as soon as possible. Please do not wait to see if the child will grow out of it! In general, the earlier you start interventions the more successful they will be.

Recommended learning & behavior assessments for children with Duchenne

Screening for autism spectrum disorder

- Timing: between 2 and 3 years of age

Dyslexia screening

- Timing: If concerns arise

Comprehensive neuropsychological evaluation

- Timing: Kindergarten or 1st grade, or in response to concerns or change in school functioning
- Areas assessed may include, but are not limited to, IQ, language, visual processing, fine motor skills, memory, attention/impulse control, executive functioning (e.g., planning, problem-solving, initiation, mental flexibility), academic skills, academic processes (e.g., phonological awareness), and behavior/emotional adjustment.

Section 2:

Learning Issues in Duchenne

In this section:

While scientists are still figuring out the exact role of dystrophin in the brain, not having dystrophin seems to cause an increased risk for specific cognitive weaknesses and learning difficulties. This does **not** mean that all boys with Duchenne will have deficits in these areas.

Parents are STRONGLY encouraged to seek assistance from appropriate professionals if concerns arise regarding any of these problem areas.

2.1

Overview of Intellectual Ability

2.1 Overview of Intellectual Ability

Boys with DMD are at increased risk for developmental delays. The most common delays are in gross motor skills such as sitting, walking and running. However, boys with DMD are also at increased risk for delays in other areas such speech/language development, motor planning, and fine motor dexterity. Some boys are later than usual in potty-training and self-help skills.

Similar to the general population, IQ scores in DMD can range from well above average to below average. Most boys with DMD have an IQ that is in the normal range. However, they are also at increased risk for having an IQ that is below average or in the mental retardation range (IQ score less than 70). Boys with DMD may also have a specific pattern of cognitive strengths and weaknesses (as described below) that may not be accurately represented in an overall IQ score. Thus, educators and parents should take care when interpreting IQ scores for DMD boys.

Interventions for Developmental Delays:

Interventions for developmental delays (e.g., speech/language, physical, and occupational therapies) typically involve various therapies and should be targeted on improving specific skills. Children with intellectual impairment will need significantly more repetition, rehearsal, and practice to learn new academic information. Specific interventions are described in the sections that follow.

boys with Duchenne face an increased risk for developmental delays

2.2

Language

2.2 Language

Boys with Duchenne usually have a vocabulary that is in the normal range, but may have more trouble with other language skills. An area of weakness may be understanding complex verbal information. Because of this, boys with Duchenne may have difficulty understanding expectations or following directions, and may only get "part of the message". Boys with Duchenne may also have difficulty with expressive language. This can interfere with their ability to describe their ideas and ask questions. Teachers should note that, because their basic vocabulary is usually in the normal range, boys with Duchenne may give the impression that their level of understanding is higher than it actually is.

Phonological processing skills are an area of language that can be particularly problematic for boys with DMD. Good phonological processing is a prerequisite for the development of reading skills, and problems in this area place a child at risk for the development of dyslexia. [See the **Learning Disabilities: Dyslexia** section for more information about phonological processing.]

Interventions for language:

Speech and language therapy is usually recommended for a child with deficient language skills. The therapy should focus on improving skills that were shown to be weaker by the child's testing. Consider simplifying communication when interacting with a boy who has language delays. Children with weaknesses in this area may have difficulty with open-ended questions ("Why did you do that?"), and tend to do better with yes/no questions or limited response options.

Ways to Simplify Communication:

- use concrete and clear terms and phrases
- adjust the vocabulary you use
- give smaller chunks of information
- introduce only one new direction/concept at a time
- have them explain things in their own words

2.3

Attention, Listening, and Memory

2.3 Attention, listening, and memory

If given repeated exposure, boys with DMD are generally able to memorize the same amount of new information as other children. However, the amount of information that they are able to take in at any one time may be less than other children. This type of memory is called short-term memory, or working memory. Boys who have problems in this area may appear forgetful, have difficulty following directions, or seem to not listen. These memory weaknesses are particularly related to verbal information, but some boys also have difficulty with visual information. Even boys with DMD who have a high IQ and strong verbal skills can have this pattern of memory weaknesses.

Boys with DMD have an increased chance for problems with attention, concentration, and distractibility. They may also be "inefficient" in the amount of time it takes them to work through things (slow processing speed), and may have difficulty dividing their attention (multi-tasking). For related information, see the section on **ADHD.**

Examples of interventions for attention, listening, and memory problems:

- Provide seating near the instruction area.
- If the child can't sit still, allow him to stand when working.
- Instructions should be broken down into individual steps, which are provided one at a time.
- Break down new information into smaller chunks, and check for understanding before moving on.
- Consider eliminating time constraints for tests and give extra time for assignments.
- Provide a quiet, non-stimulating environment for test taking or completing assignments, outside of the classroom if necessary.
- Develop a reward system to aid in work perseverance. This should focus on rewarding positive on-task behaviors, rather than punishing off-task behaviors.
- Provide advanced warning of transitions (e.g. "Two minutes until…").
- Provide clear expectations and remain calm, firm, positive, and encouraging.
- Keep the classroom well organized, structured, and controlled.
- Summarize information at the end of each instructional lesson to assist with synthesis, organization, and retention of the information.

2.4

Executive Functioning

some boys have issues with goal-oriented behaviors and mental flexibility

2.4 Executive Functioning

Research studies have been inconsistent, but there is evidence that some boys with DMD have problems with executive functioning. This is a term used to describe skills that are needed to complete goal-oriented behaviors, including things like planning, organization, initiation, mental flexibility, and self-analysis. Mental flexibility in particular appears to be more problematic in DMD than in the general population. Boys with problems in this area will have difficulty adapting to changes in expectations or requirements, or in transitioning from one activity to the next. They can get stuck on one idea, and have a hard time shifting their thinking away from it, even when others are getting annoyed or angry with them. They may appear stubborn or hard-headed, but it is important to keep in mind that this reflects a cognitive weakness rather than a character flaw. Problems with executive functioning commonly occur in the presence of **ADHD and autism**.

2.5

Learning Disabilities

2.5 Learning Disabilities

Although research has predominantly looked at reading, there is evidence that suggests that children with DMD are at increased risk for all three types of specific learning disabilities:

- Dyslexia (reading disorder)
- Dyscalculia (mathematics disorder)
- Dysgraphia (written communication disorder)

Dyslexia

Dyslexia is **not a visual problem.** It is a problem with sound-segmentation and sound-blending while reading. Thus, the most telling sign of dyslexia is when a child has difficulty sounding-out words. Dyslexia is a specific learning disability that results in difficulty learning to read. Although there are a number of different factors that can play a role, research indicates that dyslexia is mostly caused by deficits in the following foundational skills:

Phonological processing:

Understanding that spoken words are made of up small sounds (phonemes) that are blended together, and that these sounds can be identified and manipulated.

Verbal memory span/working memory:

Verbal memory span, also called verbal short-term memory, represents the amount of new information you can hold in mind at one time. Working memory refers to the ability to keep information in mind and manipulate it.

Rapid naming:

This refers to how quickly and efficiently someone is able to find and retrieve information from memory. With sufficient practice, the retrieval of some information should become "automatic".

Research has demonstrated that specific deficits in verbal memory span/working memory and phonological processing are prevalent in DMD, and can occur across all IQ levels.

Signs of Dyslexia:

Preschool
- Difficulty with rhyming
- Problems identifying the starting/ending sounds of spoken words
- Difficult or inconsistent identification of letter names

Elementary + Beyond
- Problems learning/remembering letter sounds
- Difficulty segmenting ("sounding-out") written words
- Difficulty blending sounds together to make words
- Looking at the first or last sound in a word and guessing at the rest
- Dropping word endings while reading ("slow" instead of "slowly")
- Difficulty with spelling
- Slow, laborious, or "dysfluent" reading
- Poor reading comprehension

Comments on Dyslexia Interventions

- Just telling a child with dyslexia to "read more" is not effective; research has shown that independent reading by itself does not correct the disorder. **Research also indicates that the most effective way to correct dyslexia is repetitive, intensive phonics instruction.**

- Some people think that sight-word memorization is a good strategy for children with reading problems (teaching them to recognize the word based on how it looks, not on sounding it out). The problem is that they tend to max out at around 3000-5000 words, which is far short of the number needed for fluent reading. Phonological awareness and phonics instruction must be an integral part of the process.

- Children with dyslexia will forget the reading skills they have learned if they are not reinforced or repeated. Thus, good reading programs build upon and reinforce previously learned skills, rather than switching to a different task or skill. Ongoing practice and review of previously learned steps will be necessary.

- Be patient with the child when attempting to teach reading skills. For children who struggle with dyslexia, it will be a long process that requires lots of repetition and encouragement to build confidence and skills.

Compensatory strategies for dyslexia

Despite intervention, some children may continue to have difficulty reading. Compensatory strategies do not "treat" the underlying problem like interventions do, but they may make it easier for a child who is experiencing dyslexia to succeed in the classroom. Below are modifications that teachers may need to make.

- Oral presentation of new information that the student would otherwise have to read

- Allowing the use of books on tape when available, or allowing someone to read the material to the child

- Oral presentation and responding during tests and assignments, either in person or via use of a tape recorder or voice recognition software

- Allowing extra time for the completion of assignments

- Removing time limits on tests

be patient with the child when attempting to teach reading skills

Dyscalculia (Mathematics Disorder)

There are two general areas of weakness that can contribute to problems with mathematics. One is a weakness in mathematical reasoning ability, which results in difficulty understanding math concepts. Children with problems in this area may have difficulty estimating amounts, understanding relative value (greater than, less than), or understanding abstract or symbolic concepts in math ("ten's place, hundred's place", money, fractions, etc.).

Another general area of weakness that can contribute to problems with mathematics is memory for arithmetic operations. Children with problems in this area have difficulty remembering number facts (e.g., multiplication tables), sequences/steps used in math problems, or computing easy calculations in their head. Some children may demonstrate problems in just one of these general areas, while others may have difficulty in both areas.

Interventions for Dyscalculia

Math Concepts

Children who are having difficulty understanding math concepts will benefit from an increase in the use of tangible objects and "real-life" examples when learning.

Numerical Operations

For a child to become proficient at numerical operations requires several things, including rote memorization of math facts (e.g., counting/sequencing for young children, multiplication tables for older children) and memorization of math procedures (e.g., strategies or steps to complete a problem).

Compensatory Strategies

Compensatory strategies do not "treat" the underlying problem like interventions do, but they may make it easier for a child who is experiencing dyscalculia to succeed in the classroom. Examples include:

- Provide plenty of workspace for each problem on tests and assignment sheets.
- Place steps for completing the problem at the top of the page or on an index card, and list steps vertically, from top to bottom.
- Allow use of a number line.
- Allow use of a math facts table or calculator.
- Allow extra time for tests and assignments.
- Consider reducing the number of problems; focus on quality vs. quantity if an assignment will be graded.

Dysgraphia (Disorder of Written Expression)

Writing is perhaps the most difficult academic task to master, as it requires the successful integration of a number of skills. Problems with muscle strength, fine motor dexterity, or motor planning can make the physical aspect of writing difficult and frustrating for some children. Children who have problems with language skills can have difficulties with spelling, grammar and syntax. Weaknesses in executive functioning can result in difficulty starting, planning and organizing longer written projects.

Interventions for Dysgraphia:

Fine motor dexterity or motor planning:

- Occupational therapy to improve pencil control skills
- Structured repetitive handwriting programs that teach strategies to form letters (for example, "Handwriting Without Tears")

Spelling Problems:

- These are usually related to weakness in phonological processing. Target interventions to the aspects of spelling that children are problematic for the child having problems.
  For example:
  - Instruction in clapping/counting syllables
  - Instruction in basic spelling rules (each word must have a vowel, "I before E, but not after C", etc.)
  - Instruction in word segmentation and blending
- Memorization of common words that are exceptions to spelling rules

Language Weaknesses:

- Limited vocabulary and difficulty with expression (such as grammar or sentence formation) may contribute to writing problems.

Compensatory Strategies

Compensatory strategies do not "treat" the underlying problem like interventions do, but they may make it easier for a child who is experiencing dysgraphia to succeed in the classroom. Examples include:

- Reduce paper and pencil tasks and decrease repetitive writing assignments, but do not change essential elements.
- For older students, accompany writing assignments with very specific steps and instructions, sequential outlines, etc.
- Provide copies of slides, teacher notes, and lecture outlines so the student is not missing important information while trying to take notes. When the student is given notes/outlines, require some activity of the student in order to maintain his active participation in the learning process, such as highlighting key words with a highlighter.
- Provide an opportunity for oral responses on tests or assignments and/or allow dictation.
- Do not provide a handwriting or neatness grade. Allow the use of cursive or printing, whichever is more legible.
- Allow for use of AlphaSmart or other keyboarding system.
- Do not grade for spelling, grammatical, or punctuation errors during one-sitting assignments.
- Modify test formats to increase structure. For example, instead of a general essay question, provide testing in multiple-choice, true-false, or fill-in-the-blank formats.

Section 3:

Behavior & emotional adjustment in Duchenne

In this section:

While scientists are still figuring out the exact role of dystrophin in the brain, not having dystrophin seems to cause an increased risk for specific cognitive weaknesses and learning difficulties. This does **not** mean that all boys with Duchenne will have deficits in these areas.

Parents are STRONGLY encouraged to seek assistance from appropriate professionals if concerns arise regarding any of these problem areas.

3.1

Coping with Duchenne

3.1 Coping with Duchenne

Most boys with Duchenne cope well with their medical condition and are described as emotionally well-adjusted. However, keep in mind that even well-adjusted boys may experience frustration, sadness, anger, or anxiety related to their Duchenne. These are normal reactions to a stressful situation, and are more likely to be triggered during important developmental periods or times of change.

Ways to help maximize coping

- Be available to talk with the child or with others about Duchenne.
- Help identify and address any specific cognitive or learning difficulty that is causing the child difficulty.
- Adjust the classroom setup and activities to maximize inclusion and participation of the child with Duchenne.
- Allow the child to be as independent as possible.
- Involve the child in decision making as much as possible in an age appropriate manner (including his education and healthcare/medical treatments).
- Help the child understand and express his emotions in a healthy manner.
- Active listening is important in helping a child, adolescent, or young man feel accepted and understood, and helps them "process their emotions". Several basic strategies help someone you know you are actively listening, including comments about how they appear to be feeling, summarizing what they are saying, and asking questions.
- Don't ask "Why..." questions, and don't give them advice or tell them that they shouldn't be upset, etc. Just listening to them and helping them clarify the situation and how they are feeling will enable them to work through their distress. Keep in mind that they typically just want to be heard and understood.

3.2

Depression and anxiety

3.2 Depression and anxiety

While most boys with Duchenne are not depressed or anxious, there is an increased chance when compared to other boys their age. Depression is different from normal feelings of sadness in that it is more pervasive, longer lasting (weeks to months instead of a day here or there), and powerful (significantly interferes with daily activities, relationships, and goals). Here are some signs that boys and young men with Duchenne may be experiencing depression and/or anxiety:

Signs of Depression

- Loss of enjoyment or interest in things they would typically enjoy
- Physical symptoms (e.g. headaches, stomach aches)
- Irritability, moodiness, or aggression
- Less patience or lower frustration tolerance
- Overly sensitive or tearful
- Feelings of sadness and/or hopelessness
- Suicidal thoughts
- Poor concentration, memory, or decision making
- Changes in work habits or schoolwork
- Changes in appetite or energy level

Signs of Anxiety

- Significantly worried or fearful
- Tense or uptight
- Jittery or trembling
- Problems separating from parents or other family members
- Experiencing chest pains, problem catching their breath, stomach aches, headache, or dizziness (though these may also be signs of a serious medical problem, so you should always notify a healthcare provider about any of these symptoms)

Younger children are more likely to show symptoms of irritability, aggression, over-sensitivity, or physical complaints, and do not always seem outwardly depressed. Most children have difficulty describing their emotions or identifying the cause of their distress. Therefore, they may not always be able to answer questions about how they are feeling or why.

Interventions for depression and anxiety:

Depression and anxiety can be very serious conditions and should be treated by mental health professionals. Mild to moderate symptoms of depression and anxiety may respond well to psychotherapy, though more severe cases may also require psychiatric consultation and possibly medication.

3.3

ADHD

3.3 ADHD

Boys with Duchenne are at increased risk for having an attention-deficit disorder. The formal name for this type of condition is Attention-Deficit/Hyperactivity Disorder (ADHD). The following symptoms may indicate the presence of ADHD:

Impulsivity

- Acts or says things without thinking
- Impatient or has difficulty waiting turn
- Interrupts/intrudes on others
- Talks excessively
- Acts too silly at inappropriate times

Hyperactivity

- Fidgets, can't sit still
- Often leaves seat
- Difficulty playing quietly, overly wound up

Inattention

- Does not seem to listen
- Avoids doing things that require sustained mental effort
- Loses or misplaces important things
- Forgetful, absent-minded
- Rushes through things, makes careless errors
- Doesn't complete work or turns things in incomplete
- Easily distracted, daydreams

Types of ADHD

Predominantly Hyperactive-Impulsive Type

Predominantly Inattentive Type

Combined Type

It is important to note that because of muscle weakness and physical limitations, symptoms of hyperactivity may be less obvious in boys with Duchenne. Thus, impulsivity may be the most obvious feature of ADHD for these boys. They may also have reduced sensory tolerances (see **Sensory Processing Problems**).

All types of ADHD may include weaknesses in executive functioning. Thus, children with ADHD are more likely to have problems getting started on things, and have difficulty with planning, problem-solving, and time management.

It is important to know that some of the cognitive patterns observed in Duchenne can lead to a child being misidentified as having ADHD. More specifically, language or discrete short-term memory deficits may cause a child to appear inattentive or forgetful, or to have difficulty following directions. Mental healthcare providers should consider these alternative possibilities prior to giving a boy with Duchenne a diagnosis of ADHD.

Interventions for ADHD

Treatment and intervention often has three components:

1. The use of medication is the most effective component for many boys. Approximately 80 to 90% of children with ADHD obtain some benefit from medication, although this is not always complete improvement. Stimulants are the most commonly prescribed medications. Most boys with DMD respond well to treatment with stimulant medication. However, stimulant medications should be used with extreme care in any child with heart problems, which are common in older boys with DMD. Thus, the doctor should closely monitor the cardiac status of boys with DMD who are prescribed stimulant medications. Some boys may have a minor increase in heart rate and blood pressure when taking these medications, even if they have not yet developed heart problems. It is unknown what potential impact this could have on their heart over the long run. There are non-stimulant medications that are also approved for treatment of ADHD that may be a good option for some boys, but they may also increase heart rate in some children.

2. Another component of ADHD intervention consists of psychosocial strategies. Behavior modification therapy may be helpful in mild to moderate cases of ADHD, and is most effective when focused on parent training/consultation. This can also help improve compliance and reducing arguing and temper tantrums. Behavioral therapies are usually the most effective before the child is 10 or 11 years old, so early intervention is key.

3. The third component of ADHD intervention usually involves modifying or adapting the child's environment to reduce the impact that ADHD has on their daily activities. This includes things like increasing structure and oversight during activities, implementing compensatory strategies at school or home, developing supports for memory and organizational weaknesses, and implementing routines that can be followed consistently.

3.4

Oppositional, Explosive, and Aggressive Behavior

3.4 Oppositional, Explosive, and Aggressive Behavior

Boys with DMD are at increased risk for having significant problems with arguing, not following directions, temper tantrums, or refusing to do what they are asked. Their oppositional and argumentative behaviour may be the result of problems with mental flexibility or inability to be adaptive in their thinking. Adopting an overly punitive discipline style (mostly focused on punishment) with children who have these types of cognitive weaknesses usually results in escalation of conflict, power struggles, and an increase in negative behaviors. Fatigue can contribute to behavior problems in DMD. Steroid medication can also contribute to temper tantrums and explosive behavior.

While analysis of parenting strategies is helpful, it is important to understand that behavior problems are not always the result of parenting style, and many children with behavior problems do not respond to "typical" parenting strategies. Behavioral therapies may assist parents in identifying certain situations that are likely to trigger negative behaviours, and assist in developing alternative strategies to try to correct them.

Interventions for Oppositional, Explosive, and Aggressive behavior

We recommend consultation with a mental health or behavioral therapist. Therapy with a professional for these types of behaviors should always include a large parent-training component, because this approach tends to be much more successful than individual "talk therapy" or "play therapy" with the child.

General Recommendations:

- Prioritize. You can't change everything, so focus on 1 or 2 of the biggest concerns and go from there.
- Develop and follow a routine as much as possible. Review what is going to happen during the day. Give advance notice of transitions, or changes in routine or expectations.
- Try to keep calm when a child is misbehaving. Angry parents and teachers tend to escalate the situation.
- Focus on the positive. Strategies that only focus on punishment do not promote positive behaviors, increase motivation, or change attitudes. Rewarding/praising/encouraging good behavior is more effective in the long run. Look for opportunities to say "yes" instead of "no." ("Yes, you can have a cookie, after you…").

Strategies for Dealing with Oppositional, Explosive, and Aggressive Behavior in Preschool and Early Elementary Children

At Home

- Ignore negative behaviors when the behavior is not aggressive or destructive.
- Praise positive behavior and create opportunities for positive interactions and success.
- Reward positive behavior.
- Break directions up into small manageable steps.
- Be specific and concrete when explaining expectations.
- Use time out for aggressive behaviors (that is, remove the child from any exciting activity and put in a time-out chair alone, until calm).

At School

Classroom management for preschool-aged children involves many of the techniques previously listed for home behavior management. Some teachers find reward charts helpful for children aged three and up.

Strategies for Older Children and Teens

At Home and School

Strategies to manage difficult behavior in older children typically fall into one of two categories. Depending on the characteristics of the child, one strategy or the other, or a combination of both, may be beneficial.

Behavior Modification Plan

This type of strategy has elements that are similar to the preschool strategies listed above. Essentially, the goal is to decrease negative behaviors and increase positive behaviors through the use of rewards. This type of plan makes the assumption that a child's negative behaviors result in some kind of gain (such as getting what he wants or avoiding responsibility), or are due to low motivation or a desire to gain control.

Step 1: Identify behaviors that should be reduced (e.g., angry outbursts), and/or behaviors that should be increased (e.g., time spent on homework). Prioritize and pick only one or two to start with.

Step 2: Examine when and where these behaviors do (or don't) occur to see if there are obvious triggers, circumstances, or surroundings that need to be changed.

Behavior Modification Plan

Step 3: Set goals for change.

- Expectations for behavior should be very clear and highly specific (that is, don't say, "You need to be a good student right now.")
- Develop a reward system to implement when the child meets the expectations.
  - The reward should immediately follow the behavior. Long-range rewards (such as earning a prize at the end of the school year) are too abstract and distant from daily behavior and are not effective. Also, if a child does not meet expectations at the beginning of the program and loses the chance to earn the reward, there is little reason for him to keep trying.
  - Always give praise, but other rewards will likely be necessary. Younger children may need things like candy (not too much!), stickers, pennies, or whatever else will motivate them. Older children are more likely to respond to points or stars that can be traded in for bigger prizes.
  - Avoid strategies that focus only on punishment. This type of plan does not promote positive behavior, does not change attitudes, and emphasizes failure instead of success. One example of this is to start a child with a certain number of points or on a certain "behavior level". Points are then removed or his level is downgraded when he engages in negative behavior. This type of strategy will not be effective for most children who have behavior problems.
  - Do not give rewards before the expected behavior occurs (such as, "I'll give you your reward now, as long as you promise to not have a temper outburst later").
- Involve the child in developing the goals, expectations and rewards.
- Be realistic in the amount of change you anticipate. Set small goals initially so that everyone can experience some success. For example, if a child is having a temper outburst 10 times a day, it is unrealistic to expect that he can suddenly stop. A more realistic approach might be to identify one time period during the day (such after lunch, or when he gets home from school) that the behavior should not happen.
- Consistency by the parent or teacher is very important.

Step 4: Evaluate progress in meeting goals and adjust plan as needed.

Collaborative Problem-Solving Strategy

The goal of this strategy is to promote problem-solving skills. It is particularly helpful for children who are chronically rigid and inflexible in their expectations, and/or when limit-setting or punishment routinely escalate temper meltdowns. It assumes that children want to do well and get along with others if they can. They start arguing or having a temper tantrum because they have weaknesses in skills (such as controlling their frustration, taking another's perspective, generating alternatives, being flexible in their thought process, expressive language, etc.) that make it difficult for them to resolve difficult or unexpected situations. This approach attempts to improve their deficient skills, thereby resulting in more effective problem-solving and less negative behavior. The process is highly flexible and tailored to each child and the family/teacher, but here is a general overview.

Step 1: Empathy and Reassurance

Identify and understand the child's concerns and point of view (such as completion of homework, attending an event, or engaging in an activity).

- Validate how the child is feeling, and tell him his concerns are important. Ask questions to help clarify why the child is upset, but avoid "Why" questions ("Why are you so mad about this?"). Show you are listening by paraphrasing what he is saying. Don't argue or point out how he is wrong. The goal in Step 1 is to help him learn to express himself and to "feel heard", and can be a helpful "de-escalation" tool.

Step 2: Define the Problem

Identify and explain the adult's concerns on the same issue.

- Use words the child will understand. Present the adult's perspective as one point of view, not the point of view.

Step 3: Invitation

Brainstorm possible solutions.

- Emphasize that both perspectives are important (parent/teacher and child), and that the goal is to think of a solution that addresses both sides' concerns. You are a team that will work together to solve a problem, etc. Have the child think of at least one possible option/compromise. Make a list and discuss each one. Agree on the best one and try it out. See if it works, and adjust accordingly next time. Both the adult and child work together to think of possible solutions. The goal is to come up with a plan of action that is realistic and satisfactory for both the adult and child.

3.5

Autistic Spectrum Disorders

3.5 Autistic Spectrum Disorders

Research shows that there is an increased chance of Autistic Spectrum Disorder (ASD) in children with DMD. Children with ASD have severe problems in language development, social interactions, and restricted interests or repetitive behaviors. There may be a number of children who have mild symptoms or problems in these areas, but who are not severe enough to really qualify for a diagnosis of ASD. Problems with communication skills and socialization appear to be the most common features of ASD in children with DMD, but problems in these areas can also be due to other factors (e.g. ADHD, depression/anxiety, etc.), so it is necessary to have a careful evaluation.

Children who are suspected of having an ASD or who have features associated with ASD should be assessed by a mental health or behavior professional such as a psychologist, psychiatrist, neurologist or developmental pediatrician.

Interventions for ASDs

There are several specific interventions designed for children with an ASD. The most commonly recognized of these interventions include Applied Behavior Analysis (ABA), Floortime, and TEACCH. Other interventions are usually problem-focused, and designed to address specific problem areas.

Additional information on interventions for specific aspects of ASD's can be found in other sections of this manual (in particular the Language; Oppositional, Explosive, Aggressive Behavior; Sensory Processing Disorder; Obsessive-Compulsive Behaviors; Executive Functioning; and Social Interactions sections).

3.6

Sensory Processing Disorder

3.6 Sensory Processing Disorder

Although there are no research studies addressing this particular issue in boys with DMD, some parents report that their children with DMD have features of sensory processing disorder. They report children who are hypersensitive to tactile stimulation; for example, children who complain about seams or wrinkles in their socks, socks that are not on straight, or tags in their shirts. Other parents report that their sons are aversive to "messy" play or may be "picky eaters" regarding certain tastes, consistencies, and textures. Others report that their child has difficulty tolerating noisy environments.

Interventions for sensory processing disorders

A "Sensory diet" is a set of activities or environmental modifications that an occupational therapist develops to increase or reduce specific sensory stimuli, with the belief that this will result in improved sensory processing. The activities are tailored to the unique needs of the individual child. Children who have heightened sensitivity (hypersensitivity) may be exposed to activities that the therapist believes to be calming such as listening to relaxing music, gently rocking in a quiet dimly lit room, or removing auditory or visual distractions. Children with reduced sensitivity (hyposensitivity) may be exposed to strong sensations such as hugging, brushing, rubbing, and swinging, or activities focused on tactile stimulation such as sandboxes or finger painting. In addition, the child may be rewarded for their ability to tolerate activities they would normally avoid.

3.7

Obsessive- compulsive behaviors

3.7 Obsessive-compulsive behaviors

There may be an increased prevalence of obsessive- and compulsive-like behaviors in boys with DMD. Concerns related to compulsive-like behaviors including arranging or lining things up, checking, hand-washing, re-doing something until it feels "just right," and trouble with transitions.

In some cases obsessive-compulsive (OC) behaviors may be due to sensory sensitivities previously discussed (see **Sensory Processing Disorder** section) or due to deficits in mental flexibility or adaptability (see **Executive Functions** section). While many DMD children may have these tendencies, most would not be severe enough to receive an OCD diagnosis.

Interventions for OC behaviors

Cognitive-behavioral psychotherapy has been shown to be effective in treating symptoms of OCD, and medication can also be helpful. A combination of both usually works best. In addition, here are some general strategies for parents and teachers:

- A calm, supportive atmosphere is important to overcoming OC behaviors.
- If your child is struggling with significant OC behaviors at school, provide the school with a written summary of your child's OCD challenges and needs.
- School accommodations need to be individualized based on a student's unique pattern of OCD behaviors and coping skills.

Common strategies include:

- Keep routine predictable
- Anticipate and prevent triggers
- Anticipate and ease transitions
- Allow the child to take breaks to "take space" or regroup
- Create a classroom setting that is free from stigma and teasing

3.8

Social Interactions

3.8 Social Interactions

Social interactions and the development of social relationships have the potential to be particularly problematic for children with Duchenne, although many children with Duchenne have many positive social interactions.

Reasons for social interaction struggles

- Cognitive deficits previously mentioned in this guide (language problems, social judgment/perspective taking, mental retardation, ADHD, etc.) causing social difficulties
- Psychosocial factors such as anxiety or depression
- The physical limitations and fatigue caused by Duchenne making it difficult for the child to keep up with others during play activities, sports, or games

Because strong peer relationships improve quality of life and can help provide emotional support, we strongly encourage parents and teachers to play an active role in facilitating this as much as possible.

Interventions to improve social skills

Some boys may benefit from participation in a social skills training program. The goal of social skills training is to teach basic skills necessary for positive social interactions. This should be in a small-group format, and should emphasize modeling and practice of very specific skills: how to enter a group appropriately, how to respond to teasing, how to ask someone about their interests, etc.

Even if it is inconvenient, go out of your way to encourage and facilitate participation in opportunities for social interaction. Examples may include modified/adapted sports, special interest clubs, summer camps, youth groups/programs, art groups, equestrian and aqua therapies, use of service dogs, nature programs, appropriate internet/chat rooms, etc.

Educating peers in a developmentally appropriate manner will often make them more inclusive and protective of a child with Duchenne. Strategies for this can be discussed between teachers and parents, as well as school counselors if needed.

Recommended resources

Cognitive and Learning Issues:

Learning Disabilities

Learning Disabilities: From Identification to Intervention by J. Fletcher (Book)

Overcoming Dyslexia: A New and Complete Science-Based Program for Reading Problems at Any Level by S. Shaywitz (Book)

Explosive/Oppositional Behavior

The Explosive Child: A New Approach For Understanding and Parenting Easily Frustrated, Chronically Inflexible Children by R. Greene (Book)

www.Lostatschool.org

Attention/ADHD

Taking Charge of ADHD: The Complete, Authoritative Guide for Parents by R. Barkley (Book)

How to Reach and Teach Children with ADD/ADHD: Practical Techniques, Strategies, and Interventions by S. Rief (Book)

Autistic Spectrum Disorders

First Signs, Inc. (www.firstsigns.org)

The Gray Center for Social Learning and Understanding (www.thegraycenter.org)

Autism Speaks (www.autismspeaks.org)

Obsessive-Compulsive Disorder

The Obsessive-Compulsive Foundation (www.ocfoundation.org)

What to do When Your Child Has Obsessive-Compulsive Disorder: Strategies and Solutions by A.P. Wagner (Book)

Talking Back To OCD by J. March (Book)

Social Interactions

Last One Picked…First One Picked On by R. Lavoie (Parent and Teacher Guides)

APPENDIX C: DMD SCIENTIFIC REFERENCES

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