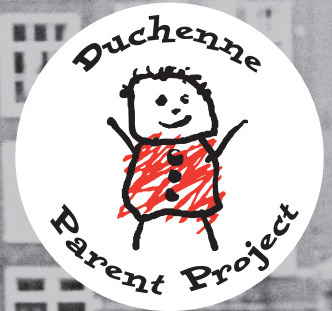


**The psychology**

**of Duchenne Muscular Dystrophy**

Jos Hendriksen, Ruben Hendriksen,  
Justus Kuijer and Elizabeth Vroom





**The psychology**

**of Duchenne Muscular Dystrophy**

Information about intelligence, learning, behavior and other relevant issues of this muscular disorder for parents, family members, teachers, paramedics and other persons involved.

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Ruben Hendriksen  
Justus Kuijer  
Elizabeth Vroom

This booklet was written on the initiative of the Dutch Duchenne Parent Project. Aim of the Duchenne Parent Project is to accelerate research into a cure and treatment for Duchenne Muscular Dystrophy (DMD) and to provide information to everyone involved with DMD patients.

Parent Project Muscular Dystrophy is pleased to help support the English translation and production of this booklet.

In writing this booklet the authors have done their best to provide adequate and reliable information. They do not accept any responsibility for incomplete or incorrect information. Suggestions for improvements to the text are welcome.

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## Foreword

When parents are informed that their son has Duchenne muscular dystrophy, it is difficult for them to understand its implications for the future. Lots of questions may arise: What will his life look like? How can we give him the best care and education? What is best for him? In what respect is he normal and in what respect different? How will he perform at school? What do we need to take into account? How do we explain Duchenne to others? Who do we tell what and when? How do we protect him from negative influences from outside?

Teachers may ask the same questions when a boy with Duchenne is put in their class: What do we need to take into account? How can we best accompany him? How and what should we tell his classmates?

This booklet aims to answer some of the above questions. Furthermore, we hope to give some general guidelines and advice to those living with DMD boys: at home, at school, during therapy and so on.

In reading this booklet, it is important to realize that there is no such thing as an average boy with DMD. Each boy is unique, and therefore the topics dealt with in this booklet are not always applicable to every DMD boy.

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## Introduction

During their developmental process, boys and adolescents with DMD are confronted with lots of physical problems . Recent research has shown that the boys and their parents learn to cope fairly well with these problems<sup>1</sup>.

Besides physical problems, primarily characterized by muscle weakness, it is now well known that boys and adolescents may have to deal with learning and/or behavioral problems. The consequences for learning and behavior are often not known to family members, friends or even professionals in education and care, and may therefore result in these consequences of DMD being dealt with inadequately.

### BOX 1: Misunderstandings.

“As long as you as a parent don’t understand, you may react inappropriately. When he was 3 years of age I started to doubt: was it my way of doing or his way of behaving? ..... He didn’t react when I talked to him, although I assumed he understood me. The same happened in kindergarten... The teacher in primary school once

In this booklet we want to inform parents, teachers and other professionals involved in caring for these boys about the psychological aspects of DMD. With this knowledge in mind, they can more adequately adapt the education, learning and therapy of the boys and their families. What will be described is applicable to a variety of situations: home, family, school and therapy.

By giving information on psychological aspects of DMD we try to explain why boys with DMD behave as they do.

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<sup>1</sup> It is now known that in exceptional cases Duchenne can also occur in girls.



In this way we hope to contribute to a better understanding of the boys and their needs. Normal development is always our starting point. We want to stress that we do not intend to describe areas of problematic functioning only: we aim to describe the boys in their overall functioning.

We give a description of the average boy with DMD: his strengths and weaknesses. But remember: the average boy with DMD does not exist. Five important domains of functioning will be dealt with successively in this booklet:

1. Physical functioning,
2. Cognitive functioning (cognitive functions and intelligence),
3. Learning at school,
4. Psychosocial functioning and
5. Behavioral functioning.

We will describe the possible problems boys with DMD may encounter in each of these domains. When possible, we will give practical advice and guidelines. In Chapter 6, for example, we will discuss talking about Duchenne with peers, teachers and so on. Chapter 7 will shortly describe the future perspectives of DMD men. In the final Chapter 8 we will present some useful websites providing background information and so on.

In order to support the information given in this booklet, we use boxes with additional information such as examples or quotations. Every chapter ends with a short summary of the main information presented.

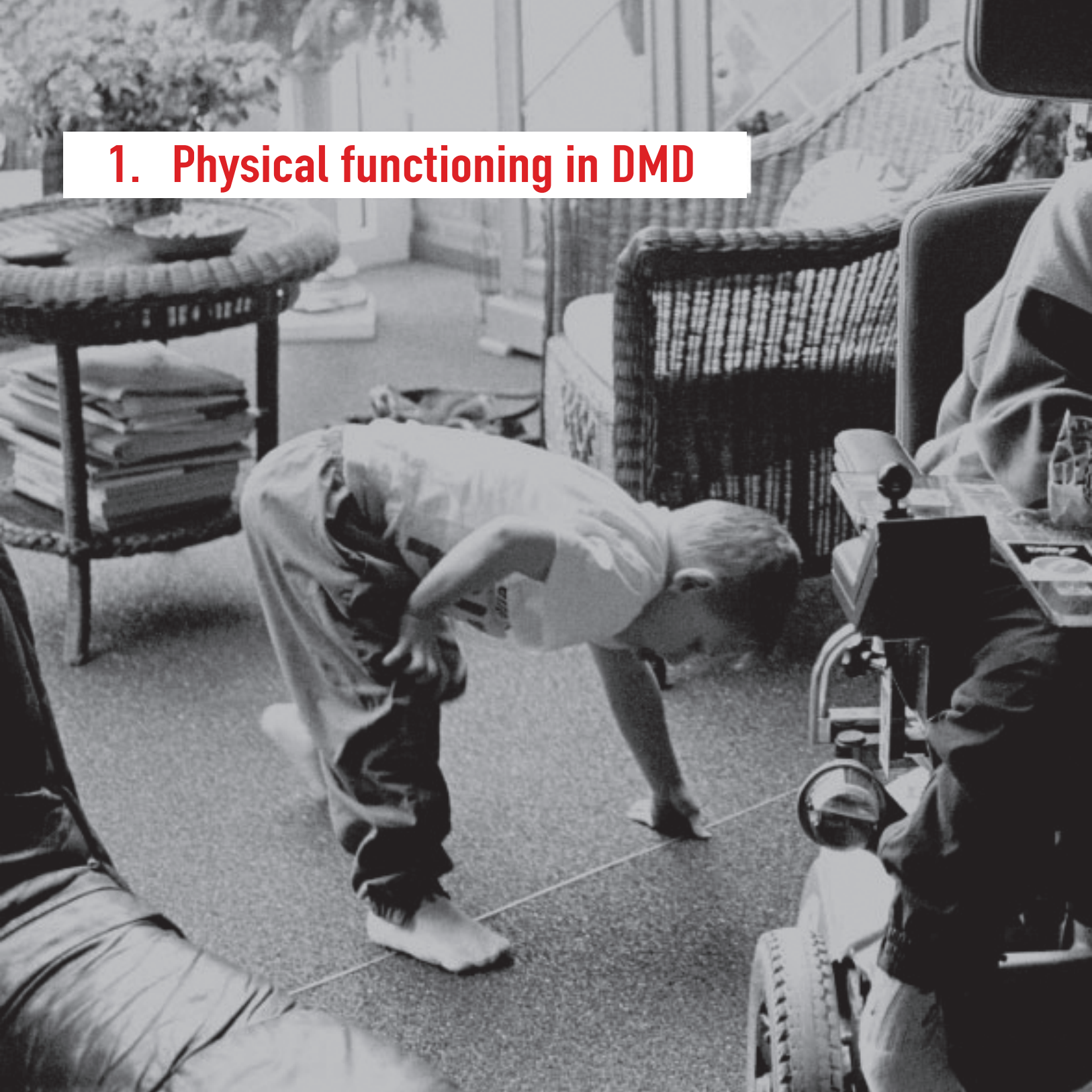
It is of utmost importance that DMD boys are given the same chance for normal and healthy development. Since they cannot rely on their physical functioning, it is important that they get the opportunity to develop other talents and use them as optimally as possible. This helps them to develop self-confidence in their future. In order to achieve this, they have to rely on their communication and social skills, their cognitive abilities, and appliances such as computers, which are very important to them.

Knowledge of one's weaknesses provides the opportunity to develop one's strengths. Although DMD boys have been robbed of their muscle strength, they can never be robbed of their independence, their desire to learn, to have pleasure and optimism in their lives. Unrealistic expectations should be avoided, as they can be a major pitfall. Parents, family members, teachers and other professionals can contribute significantly to the quality of life of the boys and their families. DMD boys have dreams for their future just like anyone else.

#### BOX 2: Greatly improved life expectancy.

Medical care has improved significantly over the last decades. Since 1990 life expectancy has increased by 10 years. In 1990, the average life expectancy was 19 years; currently it is 30 years. As a result we now have a new and unique patient population: adult men with DMD (Rahbek et al, 2005). Furthermore, much research is being done into a cure for DMD, with innovative results coming from gene therapy. These developments emphasize the importance of the psychological aspects of DMD.

# 1. Physical functioning in DMD



## 1.1 introduction

Duchenne muscular dystrophy is characterized by problems in physical functioning. These problems are caused by muscle weakness resulting from a genetic defect in the gene that codes for dystrophin. Because the weakness is progressive in nature and the physical consequences play a central role in daily functioning, it is important to shortly address this topic. We describe the course of the illness, the physical complaints encountered by the boys, and also discuss the role of corticosteroids. We end this chapter with a quotation from a teacher with a DMD boy in his regular classroom. For more detailed medical information we refer to the websites mentioned in Chapter 8.

### BOX 3: The dystrophin gene.

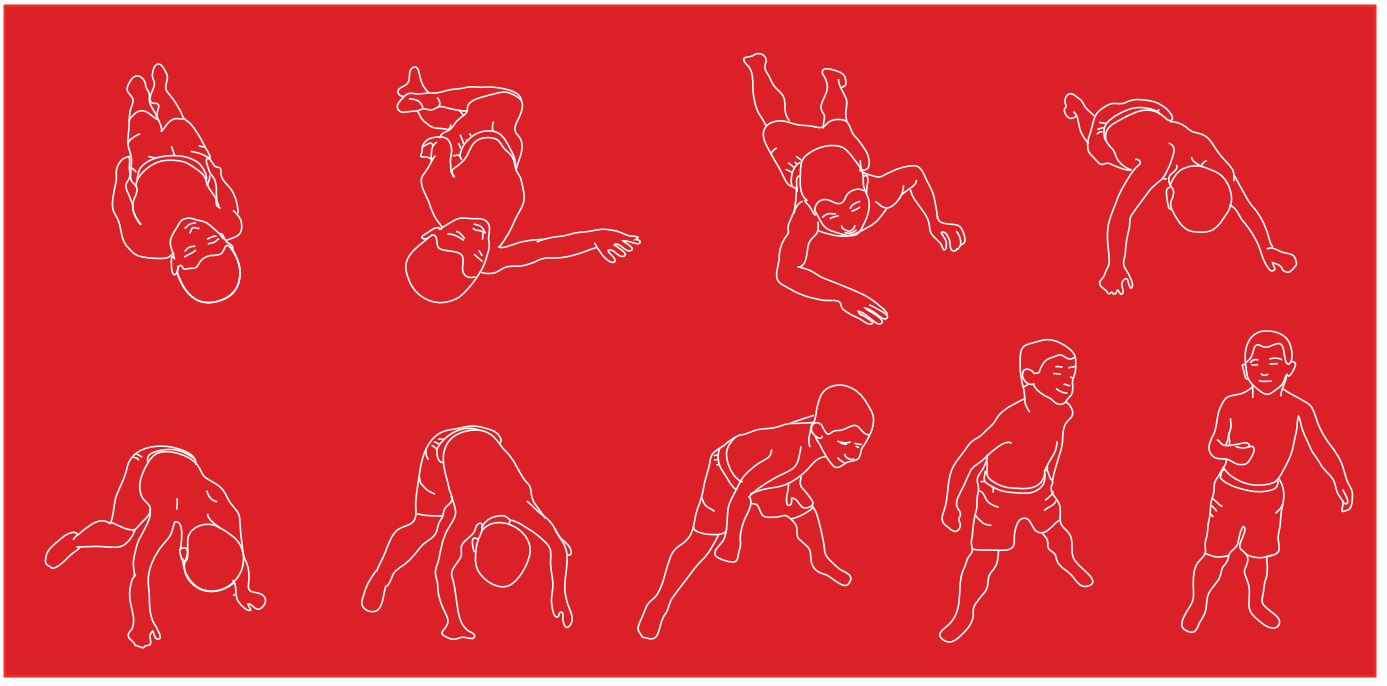
Dystrophin is the longest gene known on the DNA level, localized on the short arm of the X-chromosome at Xp21. It was discovered in 1986 and has a length of 0.84 mm. It has 79 exons that encode for a protein of over 3,500 amino acid residues.

## 1.2 characteristics and course of the muscle disease

Muscle weakness manifests itself in DMD boys between the age of 18 months and 3 years. Yet, it may be much longer before the diagnosis is made: usually between 4 and 6 years of age. The motor development of DMD boys is slower: they are usually able to walk without help at a later age than their peers. They have enlarged calves as a result of the muscle weakness. The weakened muscle tissue is replaced by connective tissue. The muscles look stronger from the outside, but in actual fact this is a sign of weakness. The first sign of muscle weakness is that the boys do not have such a strong impetus to move around. They get tired quickly and ask to be lifted up and carried. The boys are unable to stand from a

sitting position with their arms outstretched. Their method of standing up from the floor, also known as the Gower's sign, is striking. The boys use their arms to push themselves erect by walking their hands up their thighs. This enables them to assume a standing position from kneeling. Furthermore, they have difficulties running, climbing stairs and jumping.

Box 4: Picture of Gower's sign.



Between the age of 6 and 10 years muscle power decreases progressively. They first experience losing the ability to walk alone. They fall down more often and are not able to get upright on their own. This may cause feelings of anxiety, especially in company of other playing children. They cannot walk as quickly as they would like to, and so cannot compete with other children on the physical level and get tired quicker. Lifting big objects is more difficult, as is opening a heavy door, for instance. The boys are very well able to indicate when they are tired and need to be allowed to rest and take a time out of activities. Watch out for signs of tiredness, as fatigue is an important signal that should be avoided as much as possible.

Big muscles such as the leg muscles deteriorate at an earlier stage than small muscles like those in the fingers. For this reason DMD boys are perfectly able to use computers and so on, which are important appliances in school, for leisure activities and independence, until a much later age. However, even at an early stage they have less power in their hands. The facial muscles are also involved at a later age. This is contrary to other muscles diseases where facial muscles are involved at early stage.

DMD boys naturally compensate less muscle power by adopting a different body stance. On the one hand this is favorable because this particular body stance enables them to move. On the other hand, it is detrimental because this stance requires more energy and promotes muscle shortness. For example, walking on their toes with their shoulders pushed out backwards results in contractures of the muscles. To prevent contractures and asymmetric shortening of muscles it is important that the boys have a good seating position with enough support for the upper body and both feet flat on the ground. In addition, their knees should be kept straight and not bent inwards or outwards.

**BOX 5: Some practical guidelines for gross and fine motor functioning.**

- Use suitable pens and pairs of scissors, e.g., ones with big handles;
- Physiotherapy is important to prevent an unfavorable body stance;
- Do not use heavy chairs or play material in order to avoid overload and small accidents;
- Appliances are necessary to compensate for the consequences of muscle weakness. For example long leg braces or a wheelchair;
- At school it is sometimes necessary to provide an adjustable or adapted table and chair.

Most boys become wheelchair dependent before the age of 14 years. This may vary, however: one boy may be wheelchair dependent before his 10th birthday, while another may not reach this stage until his 13th. Even when boys are able to walk by themselves, they need something for longer distances to avoid fatigue: a large buggy, a wheelchair or an electric scooter.

A wheelchair may have several consequences. The house has to be adapted; there has to be enough space available in the classroom. Although a wheelchair is a symbol of being handicapped, it may also be a symbol of freedom: the boy is now able to move without too much effort. The boys gain more freedom of movement and independence and are safer because other children cannot push them over unexpectedly. Furthermore, by using a wheelchair they save energy, resulting in less fatigue. It also enables the boys to participate in sports at school. Teachers have to be creative in adapting these lessons so that the boys can participate optimally. For example, they can use a lighter ball, a lower basket and so on.

After some time hand function also decreases. This is experienced as a new and significant loss of function: the boys can no longer lift their hand upwards against gravitation. This has enormous consequences for their independence: independent eating, computer use, and use of mobile phones, for example, are not possible anymore.

Between 12 and 18 years of age there is a further increase in muscle weakness, resulting in heart and respiratory problems. Because boys with DMD are constantly in a wheelchair, they risk developing a scoliosis (an abnormal curve of the spine). For further information on these medical topics we refer to Chapter 8.

## 1.3 physical complaints due to muscle weakness

Recently, we asked the parents of 289 American and Dutch Duchenne boys to describe the physical complaints of their sons. We asked the same questions to the parents of 100 healthy Dutch boys. The results are described in Table 1. It can be concluded that DMD boys show a wide variety of physical complaints ranging from fatigue to intestinal problems and headaches. It is important to realize that these complaints can have an influence on daily functioning. For instance: fatigue may give rise to attention problems.

Table 1: Percentage of physical complaints.

Physical complaint	DMD boys	Control group
Fatigue	28 %	0 %
Sleeping problems	25 %	1 %
Incontinence	10 %	2 %
Intestinal complaints	12 %	0 %
Pain	15 %	0 %
Headaches	7 %	0 %

In spite of the increasing muscle weakness, several bodily functions show no deterioration. For instance, smooth muscles, which are responsible for the contractility of hollow organs such as blood vessels, the gastrointestinal tract or the bladder, remain intact. Problems with digestion are therefore not very common in DMD. The risk of being overweight, on the other hand, may be a real problem, as the boys sit in a wheelchair all day and have reduced body movement.

The boys' tactile functions also remain intact. They can feel with their hands as others feel, since nerve functions are normal in boys with DMD. Their sexual functions are also the same as their peers'. Finally, it should be mentioned that speech function in itself does not decrease with age, in contrast to other muscle diseases where this may be the case.



## 1.4 use of corticosteroids: benefits and side-effects

Because DMD cannot be cured yet, treatment focuses on symptom relief, deceleration of physical symptoms (muscle weakness and its consequences), and improvement of the boys' quality of life. One of the medications used is corticosteroids.

Prednisolon and deflazacort are the ones most used. Scientists do not yet agree at what age treatment with this drug should be started, and in which dosage. Most commonly this treatment is started when the first symptoms become manifest between 4 to 6 years of age. Several investigators show a significant positive effect of corticosteroids on physical functioning (Biggar et al, 2004, 2006). Boys using corticosteroids are able to walk for 3 to 5 years longer than those not on the drug. In addition, the manifestation of scoliosis is significantly retarded by using this drug: of the boys on corticosteroids 50% develop a scoliosis around the age of 15 to 17, compared to 90% of boys not on the drug. Heart and lung function are also better in those using corticosteroids.

Corticosteroids may have side effects varying in intensity:

- Weight gain and increased appetite;
- Cushing, resulting in excessive fat throughout the body and a large round face (puffy cheeks);
- Hypertension;
- Decalcification of the bones;
- Delayed puberty;
- Reduced height growth;
- Behavioral problems and temper tantrums.

**BOX 6: Should we use corticosteroids or not?**

"I believe the steroids sometimes result in irritable behavior in my son. But the advantages of using it are much more important than the disadvantages and negative behavior."

## 1.5 a boy with DMD in my class: a teacher's story

A teacher in primary school wrote down the following experiences:

“For six years there was a boy with DMD in my class. When he was 6 years of age his muscle weakness was not very noticeable: he was active and lively. When he left school he was wheelchair dependent. After a while some things were getting more difficult for him: climbing stairs, walking and running. But the boy himself was not bothered at all. As his teacher I didn't experience him as an extra burden. Except now and then when the stairs were too steep. He wanted to do everything by himself.

The most difficult period was during 5th grade, when walking became more difficult but he insisted on doing everything himself. Going to the toilet by himself, picking up his work by himself, cleaning his desk, going outside and so on. His classmates knew about his condition and were extremely caring. But as normal children, they were sometimes overexcited and inattentive. My greatest worry at that time was this pupil falling down due to an unintentional action of another child and that he would hurt himself. Whenever he stood up, I secretly kept an eye on him and cleared the way for him when everybody wanted to go outside to play. Always someone was available to give me a hand, as I had to take care of 26 other children as well.

When he was unable to walk anymore in 6th grade, times were more tranquil again. There was no more fear of falling down, not for him, not for me and not for his classmates. Most places were accessible for an electric wheelchair. During class time he used his wheelchair and during playtime I helped him onto his electric scooter. Because our school has several floors, the children took turns helping him in and out of the lift. He participated in sports lessons because he didn't want to miss anything. Several minor adaptations made it possible for him to participate. Special equipment was acquired for him (special hockey stick). Here also everyone was willing to do something for him”.

## 1.6 to conclude

Duchenne is a complex illness with drastic consequences for the boys, their family members and everyone else working with them. Adequate information on the illness and its consequences is of utmost importance.

## 2. Intelligence and cognitive function in Duchenne



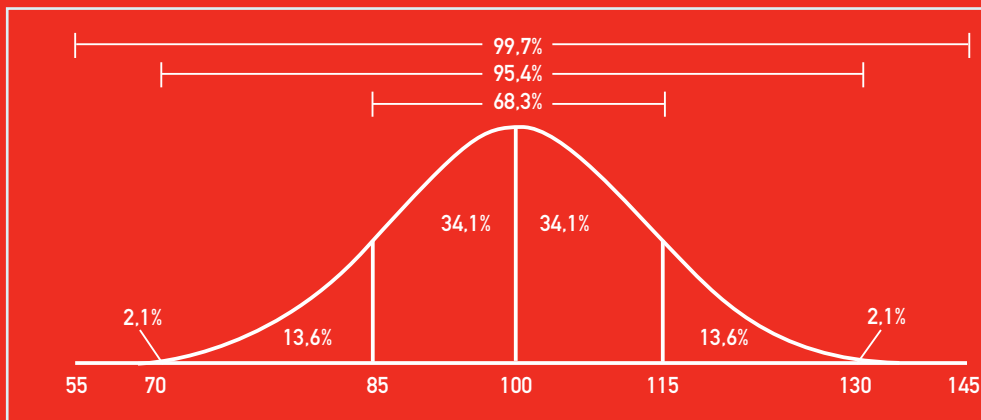
## 2.1 introduction

This chapter will discuss intelligence and cognition (thinking functions such as memory, attention, language and so on) in Duchenne. It is now well known that specific cognitive problems may be part of the muscle disease. We will show that the boys have strengths and weaknesses in their thinking processes. Where these problems come from will also be discussed to enable you to understand the causal factors. We also describe the consequences and how to deal with them.

## 2.2 intelligence and Duchenne

David Wechsler, an important American psychologist, defined intelligence as “an individual’s ability to adapt and constructively solve problems in the environment”. Intelligence can be assessed by means of an intelligence test. The Intelligence quotient (IQ) in the normal population is 100 with a standard deviation of 15. This means that around 70% of the population has an IQ between 85 (100 minus 15) and 115 (100 plus 15)). The normal distribution of IQ scores is illustrated in figure 1. It shows that 2 ½ % of the population has an IQ below 70 (100 minus twice the standard deviation) and another 2 ½ % has an IQ higher than 130 (100 plus twice the standard deviation).

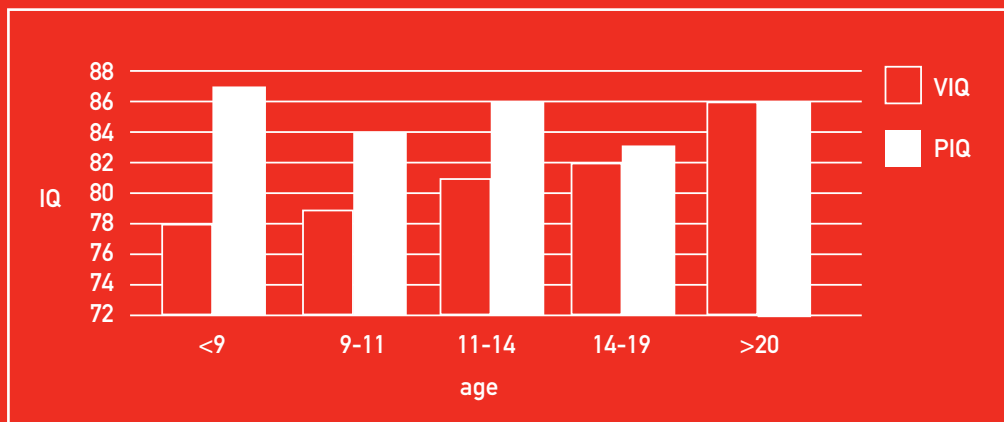
Figure 1: The normal distribution of intelligence.



In a large-scale review study on intelligence and DMD, Cotton summarized the results of intelligence testing in nearly 1200 boys with DMD reported in literature over the last 40 years (Cotton, 2001). She found that the mean IQ in DMD boys is 80. An IQ score of 80 is called “low average”. This also implies that around 30% of all DMD boys have an IQ below 70, which is called “mental retardation”. This study also revealed a very high variability in IQ scores in DMD boys: their IQ ranged from 14, which is extremely low, to 134, which is a very high score. So remember that DMD boys can be very bright and have a high IQ.

The Total IQ (TIQ) can be distinguished in Verbal IQ (VIQ) and Performance IQ (PIQ). Verbal IQ has to do with language, for example: “ How are a piano and an guitar similar?” In this example you have to find the similarity between two concepts, since both are music instruments. Performance IQ has to do with visual thinking and motor performance, as for instance in a subtest called block design, where you have to copy a block design using blocks. In the same study, again by Cotton, it was found that DMD boys younger than 9 years of age showed the biggest difference between Verbal IQ and Performance IQ, with Verbal IQ being lower. When boys grow older, the difference decreases because their Verbal IQ increases somewhat. It can be concluded that verbal thinking is more problematic for younger boys (Figure 2). It seems then, that as the boys grow older they grow out of language related problems. This is what is called “ growing out of deficit” .

Figure 2: Developmental profile of intelligence at different ages: VIQ increases, as boys grow older.



To summarize: generally speaking, boys with DMD have a low average IQ. Remember that in contrast with the progression of muscle weakness, intelligence does not decrease, as they grow older: IQ remains stable over time. In contrast, their verbal IQ even increases somewhat as boys grow older.

**BOX 7: Not all boys have a low average intelligence.**

“My boy really loves to read and he is very good at reading. When he is not challenged intellectually at his level, he occupies himself with computers. He has even developed a computer program for easy calculation of arithmetical computations.”

## 2.3 profile of strengths and weaknesses

Expressive language may be a first weakness in the cognitive profile of DMD boys, implicating that they have problems producing speech quickly and on command. These problems are not related to muscle weakness of the speech organs, as might be expected. A delay in speech development may even be manifest before the muscle weakness is present in DMD boys. English researchers (Essex & Roper, 2001) found that in 40% of the DMD boys without a family history of Duchenne there were concerns about speech development before the diagnosis of Duchenne was made. Thus speech problems may precede muscle weakness in Duchenne.

The language problems have to do with expression of speech, in other words fluent and quick production of speech. Problems that are more often reported are:

- Late onset of speaking;
- Problems with word finding (quickly finding and using words), so called “word finding problems”;
- Difficulty with fluent language production;
- Deficits in auditory working memory;
- Non-fluent speech with restarts, fillers such as “eh”, and pauses;
- Speech understanding is much better than speech expression

These problems reflect what is known in the literature as “specific language impairment” (SLI). SLI is known to be a risk factor for later reading problems, which we will discuss in chapter 3.

A second area of cognitive functioning that may present problems has to do with short-term memory. In this memory information that is not needed permanently is stored for a short time. People use this memory system very, very often every day. We use this system to understand language or to plan our behavior. When there are problems with short-term memory, it is difficult to store and retrieve recent information. Memory problems in DMD boys involve problems with both visual memory (what they see) and auditory memory (what they hear). This may have important implications for daily functioning.

#### BOX 8: Consequences of memory problems.

When I ask him to go to the living room, check whether the television is off, and also check for the post, he only gets part of the message. Part of the message has not been stored and so he can't perform it.

Concentration is a third area of possible cognitive problems. In our own research we found that 18% of the parents of DMD boys report that their boy has serious disadvantages because of his concentration problems. In a control group of boys, we found this percentage to be 8%. These concentration problems can give rise to serious school and learning difficulties. It is expected that physical fatigue, which is common in DMD, can contribute to concentration problems. It is important to understand this relationship between fatigue and concentration.

To summarize then: There is still debate on the specific cognitive profile in boys with Duchenne. It seems that expressive language, working memory and concentration processes are relatively weak. But DMD boys also have numerous strengths. They have good visual perceptual skills (that is: they can

discriminate visual patterns, recognize incomplete pictures and construct puzzles well). They are also strong in learning and rote memory. Remember their strengths and use them in their learning process. Take care not to overestimate them when looking at their strengths alone, or to underestimate them when looking at their weaknesses.

#### **BOX 9: When is psychological testing necessary and how is it done?**

When there are questions about the intellectual capacities of a DMD boy or when the boy has learning problems at school, this may be a reason for further psychological assessment. Ask your child's medical specialist to start the procedure. From the age of 2 ½ years the Wechsler preschool intelligence test (WPPSI) is used, from 6 ½ years on the Wechsler children's intelligence test (WISC) is used.

Cognitive function tests for memory and attention, for example, are available from the age of 6 years. Children mostly find the testing fun, and the assessment seldom causes unnecessary stress. Its duration depends on the specific question and varies from 2 hours for younger children (under 6 years) to a whole day (2 sessions of 3 to 4 hours) for older children.

In concluding this paragraph we want to stress that these strengths and weaknesses are not typical for all Duchenne boys. They simply have a higher risk for cognitive problems. If you have any doubt consult your physician and let your son be tested.

## **2.4 what causes cognitive weaknesses?**

What is the cause of these cognitive strengths and weaknesses? Most probably there is a relation to brain function. It is now known that dystrophin is normally also present in other tissues as well as muscle tissue, including the central nervous system. The lack of brain dystrophin might therefore play a role in the cognitive functioning of the boys. The dystrophine isoforms Dp427-C and Dp427-P would normally be distributed to the hippocampus and purkinje cells in the cerebellum respectively, contributing to greater post-synaptic density. The cerebellum and hippocampus are part of an integrated network that

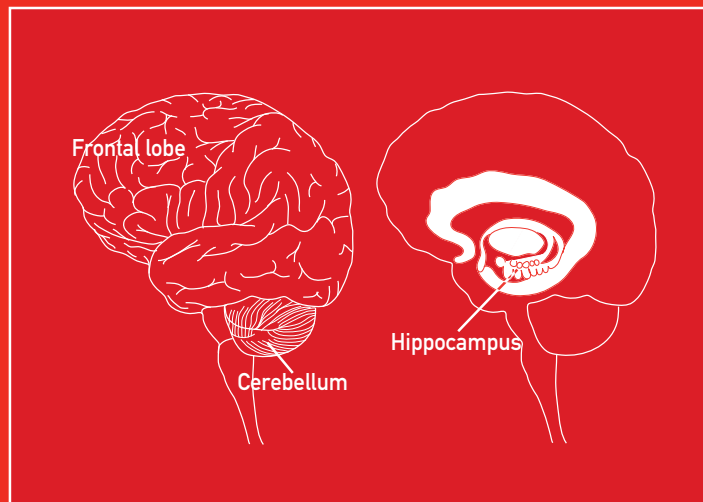


includes connections with the frontal brain regions. This altered postsynaptic plasticity may hinder efficient memory, automation and planning/organization. The cerebellum (small brain) in particular is an important structure. It is now known that the cerebellum (which means “small brain” in Latin) has up to four times more neurons than the cerebrum: so the term cerebellum is wrong and confusing.

Table 2: brain areas in which dystrophin has been found.

Brain area	DMD boys
Hippocampus	Memory
Cerebellum	Automatization
Frontal lobe	Planning & Organization

Figure 3: The hippocampus, the small brain and frontal lobe (anatomy of the brain).



More research needs to be done into the role of dystrophin in the brain. It is clear that dystrophin works differently in muscles to its function in the brain.

## 2.5 consequences of cognitive weaknesses

Language, memory and concentration can cause problems and have consequences for automation, planning and learning. Because of these cognitive problems Duchenne boys more often show difficulty with:

- Completing tasks;
- Time orientation;
- Initiative: they do not know how to start an activity;
- Resistance to change: they have more difficulty adapting to change and novelty;
- Switching from one activity to another.

### BOX 10: Unable rather than unwilling?

Teachers often complain to me that they don't understand why the boys stop working and do not continue with a second task when they have finished the first one. This is not a motivation problem as one might think. It might have to do with not being able to switch their attention quickly and carry on with another task. They are unable to switch, not unwilling to do so

## 2.6 coping with weaknesses: some guidelines

What can teachers and parents do to reduce or compensate the hinder of the weaknesses. First of all, be aware of this profile of strengths and weaknesses. Realize that it has to do with brain function and not with physical functioning or a motivation problem. The boys are not to be blamed.

Furthermore, from research we now know that strategies you find for yourself are much more effective than strategies suggested by teachers or parents. So, help the boys find their own solutions to learning problems. Here are some more guidelines that can be used at home or in school or therapy:

### Guidelines for short term memory problems:

- Use short sentences without difficult words. Divide the information in small parts to be processed by the boys;
- Use visual cues to help them process the information (e.g. visualize instructions);
- Check whether he has understood what he has been told. This is called the principle of re-teaching: after you have completed your instructions, let him repeat the instructions in his own words. In this way he learns to control his own verbal memory;
- Summarize important information and repeat the main information;
- Introduce new information step by step and always repeat the previous step.

### BOX 11: Errorless learning.

The principle of errorless learning refers to teaching procedures that are designed in such a way that the learner does not have to make mistakes when learning new information or procedures. Errorless learning is in contrast to trial and error learning, in which the learner attempts a task and benefits from mistakes. The main thing is to prevent the boys from making mistakes.

One way to do this is to “pre-correct” answers. For example, when reading a sentence with a word in it that one doubts the boy can read, one can say: “ I see a tricky word in the next sentence starting with a P. Let me know if you need help”.

Essential to this technique of errorless learning is the avoidance of trial and error, which may result in learning the wrong thing and is difficult to unlearn later.

### Guidelines to improve attention:

- If necessary give extra time to complete the instructions;
- Use short time periods: e.g. instead of one long period of 20 minutes to complete a task, use two periods of 10 minutes;
- Use a room where the boy can work without being disturbed or distracted;
- Make control lists of “to do” activities he can plan himself and check himself, or even better, let him make his own control list;
- Finally, use agenda training to help him make his own daily plan of activities.

### 3. Learning in school and Duchenne



## 3.1 introduction

Learning disabilities occur more often in Duchenne boys. Learning disabilities are described as problems with reading, arithmetic or writing in persons with normal intelligence that have had sufficient education and instruction. The core problem is what is called automaticity, particularly in the areas of literacy and numeracy. This implies that a mathematical procedure, for instance, is performed quickly and routinely without thinking. Although children with learning disabilities have certain difficulties, they may also have certain strengths, such as in the area of analytic and creative thinking.

In short, a learning disability can be related to (1) recognizing letters and producing the right sounds, as in reading, (2) recognizing and producing numbers, as in dyscalculia and (3) producing letters in writing, as in dysgraphia. Actually, learning disabilities are about the 3 D's: dyslexia, dyscalculia and dysgraphia. We will describe two learning disabilities of relevance for Duchenne namely: dyslexia and dyscalculia. This chapter will be concluded with a discussion on school careers and choices to be made in DMD boys.

## 3.2 dyslexia and Duchenne

We live in a language oriented society in which reading is an important skill for independent living. Dyslexia (from the Greek “dys” meaning impaired and “lexis” meaning word) is a learning disability that has to do mainly with technical reading and/or spelling. It is now well known that around 3 % of schoolchildren have dyslexia (see: Learning Disabilities Association of America <http://www.lidaamerica.org>). Dyslexia can be defined as a neurologically based, often familial, disorder that interferes with the acquisition and processing of language. Varying in degrees of severity, it is manifested by difficulties in receptive and expressive language, including phonological processing, in reading, writing, spelling, handwriting, and sometimes arithmetic (revised definition from the International Dyslexia Association). Dyslexia is not the result of low cognitive ability and is not due to sensory impairment, poor learning opportunities or inadequate teaching. Children with dyslexia are characterized either by slow and accurate reading or quick and inaccurate reading. Although dyslexia is

a life-long problem, children with dyslexia frequently respond successfully to timely and appropriate intervention. They can learn strategies to cope with and compensate the deficit.

Boys with Duchenne are at a higher risk for reading problems. Together with Duchenne Parent Project in the Netherlands, we performed an extensive follow up research in 25 Dutch Duchenne boys (Hendriksen & Vles, 2006). We found that 20% of the boys showed serious reading problems and 20% moderate reading problems. So in total 40% of the boys had difficulty learning to read in spite of normal intelligence. As already stated, the percentage of serious reading problems in the general population is around 3%.

### 3.2.1 how do children learn to read, and what goes wrong if they are dyslectic?

When beginning to read, children must learn to recognize the visual characteristics of each letter (called a grapheme) and subsequently quickly and automatically pronounce the corresponding sound (called a phoneme). Beginners must learn to automatically produce phonemes (the smallest unit of spoken language) such as /a/ and /oo/. There are 48 phonemes in the English language (General American). Not everyone knows this and realizes the importance of teaching children the phonological alphabet, which is different from the 26 letters in the alphabet. The phonological alphabet has more letters than the normal alphabet: for instance, two-sound vowels such as /ea/, /oo/. Initially, reading is completed – often halfway through the first grade of elementary school – when a child knows and is able to convert all 48 signs (graphemes) quickly, automatically and correctly to the corresponding sound (phoneme), and combine them into words. Children with dyslexia have difficulties converting graphemes into phonemes automatically.

**BOX 12: Technical reading: making a grapheme-phoneme conversion quickly and automatically.**

Reading nonsense words such as the words mentioned in this box involves pure technical reading. The words have no meaning, but can be read phonologically. You cannot read these words using knowledge of vocabulary, because they simply don't exist.

foom

blayms

keamong

## 3.2.2 reading and Duchenne

According to current insights, a higher risk of dyslexia in boys with Duchenne is probably due to a lack of dystrophin in the cerebellum. In neuropsychology it is known that the cerebellum plays an important role in automatization (see also chapter 2).

For boys with Duchenne, reading is extremely important. It is therefore essential that teachers and parents are aware of the risk for reading problems to ensure early diagnosing and treatment. Reading is not only necessary for further education, but also because of the dependency on written words when communicating with the help of technical aids. Reading is a very important skill when using technical aids such as the computer and its software (word processing, drawing, e-mail, etc.). Until there is an optimal treatment for Duchenne, reading will have a very important role in the future of the boys.

## 3.2.3 strategies to compensating for dyslexia

Overall, early detection and early intervention is the most effective way to reduce problems resulting from dyslexia. The keyword is “early”. For boys with Duchenne, especially when their speech development progresses slowly, it is important that they are tested psychologically and by a speech therapist as early as possible - preferably around the age of 4 – to evaluate whether they do have a higher risk of developing a reading disorder.

Early detection also means early intervention, before they start learning to read in the first grade of elementary school. Reading aloud to your boy at an early age is very important and serves to increase phonemic awareness and expand vocabulary. In addition, pleasure in reading is enhanced.

Children who learn to read need to become aware of the sounds in the words and language. Phonemic awareness is the foundation for learning to read and can be trained early. If you train phonemic awareness, you are actually training initial reading skills. These strategies include learning to count the sounds in words, segmenting the sounds and

recognizing rhyming words. Various activities can be used, such as: rhyming words, adding, subtracting and deleting sounds, clapping syllables in names and words, or playing listening games. These exercises have proved to have a positive effect on learning how to read at a later age and thus help to reduce reading problems. This is also referred to as emergent literacy, refers to the pre-reading stage (age 4 to 6 years), and is aimed at training the prerequisites for reading

**BOX 13: Training phonemic awareness is useful.**

Phonemic awareness covers 48 phonemes of the English language. It can be trained by listening and letter games in which phonemes are pasted together to make a word, for example: /c/ /a/ /s/ /t/ /l/ /e/ combines to /castle/.

Reading is a pleasant and stimulating experience. Here are some practical suggestions:

- Make reading a special moment as well as a daily one;
- Show the boy that you enjoy reading aloud and actively involve him;
- Make him responsible for his reading by allowing him to choose his own books;
- Correct mistakes in a relaxed and enjoyable manner. Make sure you are a 'perfect example': pronounce the word and its phonemes correctly;
- Listening to books can be helpful to enhance the pleasure of reading

**BOX 14: Does practicing work?**

When learning to read, grapheme-phoneme conversion must be quick and automatic. Automatization works best with the following rule of thumb: it is better to practice 10 minutes each day (whereby we literally mean every day of the week, all 7 of them) than practicing for 35 minutes twice a week.



### 3.2.4 what can the teacher do?

If reading problems do not improve in spite of practice and stimulation, additional help is necessary. Remedial teaching by a teacher who is trained in reading problems can be valuable. Useful information can be found on the websites: <http://www.lidaamerica.org/> and <http://www.dyslexia-parent.com>. The next strategies can be helpful in the classroom, but also at home:

- Present information verbally instead of making the boy read;
- Use listening books when available;
- Let the boy answer tests or exercises verbally rather than in writing;
- Allow additional time for written assignments and tests;
- Use questions to stimulate the boy's understanding and interpretation of a text and to help him read it critically.

Trained professionals can identify dyslexia by formal evaluation. When dyslexia is diagnosed officially, the child can get a statement to say that he is dyslexic. Special help is possible through an Individualized Education Program (IEP, which each public school child who receives special education and related services must have) and/or a so-called 504 plan (legal document that outlines a plan of instructional services for students in the general education setting). Both have to do with the rules and regulations within the Individuals with Disabilities Education Act (IDEA) (see the website of the National Center for Learning Disabilities <http://www.ldonline.org/>).

Dyscalculia refers to a wide range of lifelong learning disabilities involving math. There is no

### 3.3 dyscalculia and Duchenne muscular dystrophy

single type of mathematical disability. Much less is known about dyscalculia than dyslexia, since reading plays a more significant role in our language-oriented society. The prevalence of dyscalculia is estimated at 2-4 percent of the general population. Our research (which has not been published yet) suggests that slightly more than 10 percent of Duchenne boys have serious difficulties with mathematics. Another 10 percent of the boys seem to cope with moderate problems in the area of numeracy. However, further scientific research is required to confirm these preliminary results.

BOX 15: illustration of typical faults in mathematics.

How much is  $64 + 13$ ?

A person with dyscalculia will find a different answer. Here is an illustration of their way of thinking: “First I have to see whether I have to add up or count down. OK, I have to add up. Now I work from left to right:  $6 + 4$  is 10 and  $1 + 3$  is 4. Then I have to add  $10 + 4$ . The answer is 14.”

$$\begin{array}{r} 64 \\ + 13 \\ \hline 14 \end{array}$$

### 3.3.1 definition of dyscalculia and its cause

Dyscalculia has been defined as: “a disorder that is characterized by severe problems with the learning and applying of mathematical knowledge (facts/rules) and skills”. Mathematical rules are recalled or applied too slowly, which is a great obstacle for people with dyscalculia. So children with dyscalculia comprehend mathematics, but again – as in dyslexia – they have difficulties with automatization.

Dyscalculia is not caused by low-quality math lessons at school or by the child’s unwillingness to learn to understand the principles of math. As in dyslexia, aberrant processes of information processing in the brain cause dyscalculia. What precisely goes wrong in the brain is still not known.

Dyscalculia can vary from person to person. Several abilities can be involved, and trained professionals can evaluate them:

- Ability with basic mathematical skills such as counting, adding, subtracting, multiplying and dividing;
- Ability to predict appropriate procedures based on understanding patterns knowing when to add, subtract, multiply, divide or do more advanced computations;

- Ability to organize objects in a logical way;
- Ability to measure — telling the time, using money;
- Ability to estimate quantities.

### 3.3.2 strategies to compensate for dyscalculia

An important means of assistance for children with difficulties in math is to visualize mathematical principles and key terms such as fractions and rows. This can be achieved in several ways: for example by cutting a round cake in pieces (quarter, half etc.). This way, fractions are visually represented and so can be explained easily. Mathematical rows can be explained the same way using a necklace, for example. Repetition is the best way to improve mental arithmetic because this has to become automatic. As noted earlier, it is better to practice 10 minutes a day than 35 minutes twice a week. There is plenty of computer software available to facilitate practicing and automatization of math skills, making learning a fun activity. Chapter 8 lists some websites offering these programs at no cost.

#### BOX 16: Edutainment.

Edutainment is the new magic word: it refers to a form of entertainment that is designed to educate as well as amuse. Some interesting sites for training mathematics are: [www.schoolforge.net](http://www.schoolforge.net) and <http://edu.kde.org>.

As is true for dyslexia, assistance from somebody with a lot of knowledge about mathematical disorders can help compensate for dyscalculia. However, it is also true for dyscalculia that it is a lifelong condition that will never really disappear. This shows the importance of adapting the classroom-learning environment to a boy with Duchenne and mathematical problems. Here are some more strategies:

- Provide the boy with a map to help him solve mathematical calculations. This map should display the steps required to come to the solution. Such a map might be practical for homework. In addition, this map might include the answers to the

most frequently used multiplications (e.g. up to the product of 100). Such a map could easily be stuck onto his desk;

- The boy should be taught only one way of solving calculations. Too many methods increase his confusion. Work on finding different ways to approach mathematical facts; i.e., instead of just memorizing the multiplication tables, explain that  $8 \times 2 = 16$ , so if 16 is doubled,  $8 \times 4$  must = 32;
- Allow the use of tools such as calculators;
- Make 'memorizing sheets' of basic math knowledge;
- If a boy has severe difficulties, less homework should be considered: quality over quantity;
- Exercises that have not been completed in class should not be completed at home since this often results in extra frustration. Furthermore, the learning process can't be influenced sufficiently;
- Give extra time for the completion of exercises and tests.

Finally, as was true for dyslexia, a trained professional or child psychologist can perform a specific assessment in boys that seem at risk for dyscalculia. If dyscalculia is indeed diagnosed, the same rules and statements as for dyslexia apply: IEP and 504 plans, which among other things justify extra support and time.

### 3.4 school and DMD

School is extremely important for boys with Duchenne, probably even more important than for other children. At school they learn reading, writing, working on the computer and its programs and much more; skills that will occupy their lives. The latter goes for both work and hobbies. Besides, school is also important for the boys' social development. School is of essential importance for the shaping of their future. Therefore, the choice of the course of education to follow should be made carefully. Chapter 4 will elaborate upon the importance of having friends in coping with a disease like Duchenne.

### BOX 17: A description of the situation in USA.

In 2004 we did an extensive research project on Duchenne and school. Of the 277 USA boys with DMD, 22% were in special education, and 78% attended normal school. According to the parents, 22% had reading problems, 25% had problems in math, and 26 % had writing problems.

If possible, boys with Duchenne should visit mainstream schools. This is the best for both the boy himself and his environment. At a regular school the boy is in the middle of society and experiences how it works. In addition, he learns to get along with physically healthy boys and girls of his own age, which is important for further education and development. On the other hand, boys with Duchenne are special boys that can be of special interest and value to a class, as paragraph 3.5 will demonstrate.

Despite everything that has been mentioned about supporting tools and assisting devices (e.g. wheelchairs), schools should not be reluctant to welcome boys with Duchenne because they might mean extra work for both the school and the teachers. This is often not true. The opposite can also be true, as supported by the experience of the teacher reported at the end of chapter 1.

Sometimes, however, there may be valid reasons for a Duchenne boy to attend special education. One example could be if intelligence, learning problems or behavior result in slowing development. In this case the boy needs qualified, adjusted, flexible and individual education. Schools that teach children with psychological disabilities are trained in this kind of education and thus might be well suited for boys with Duchenne .

For a description of the American situation on special education needs we refer to the following website: [www.2ed.gov/osers](http://www.2ed.gov/osers) .

## 3.5 a boy with DMD in a regular classroom can be of great value for the whole class

This part of chapter 3 illustrates the importance of a boy with Duchenne attending regular school (i.e. no special education). For the school teacher this does not entail a great amount of extra effort. The story below, which was written by a Dutch teacher of a class that included a Duchenne boy, illustrates this.

Together, all the other children and I observed the development of his disease close up. When he came to my class, I knew from his parents what was going on and how his disease would develop both physically and educationally over the next years.

In the first 1.5 years he was a child just like the others; he liked to do everything himself, was funny and eager to learn. The expected learning difficulties (especially concerning automatization) appeared not to be of major significance. As he became physically more dependent and had to rely more on others, and the other children became more used to his disease as well, they were able to handle some of my usual activities; picking up something for him, grabbing something from the ground or taking him outside. As such, he and I had a class full of helpmates. It was nice to observe the willingness of the other children to help him when it seemed necessary. We regularly made trips to a museum and also went on a school trip over several days. A little creative thinking seemed sufficient to find an appropriate solution for potential problems (sleeping opportunities, concert halls). Only rarely activities remained impossible and had to be cancelled, but then we undertook other fun activities instead.

When writing became difficult, he got a notebook on his table so that he was able to continue with his schoolwork. Basically, the solution for anything seemed quite easy and hardly time-consuming. The major tasks for me as a teacher consisted firstly in transferring him from his scooter into his wheelchair and the other way around during the breaks, and secondly assisting him when he needed to visit the toilet. These tasks became daily routine to such an extent that they could hardly be seen as extra work. Moreover, during the last years of primary school his peers replaced me in many ways. For instance, taking off his coat, something he was not able to achieve independently anymore at this stage, was done

by classmates. He was always able to ask for help if necessary and there was always someone near him willing to help or cooperate.

Looking back at the years that he was part of my class, I can only value this period as beneficial. Children and adults always appeared willing to think in order to solve a particular problem or to help. In addition, the care of the other children positively influenced the atmosphere in the classroom. Apart from that, children learn the valuable lessons that it is not always the rule that your body just works, and the importance of caring. It is true that a boy with Duchenne in the classroom requires more effort and attention than an average pupil, but, as this story also illustrates, the boy offers the class a lot as well. In fact, some willingness and creative thinking are capable of transforming a boy with Duchenne to a child like all the other children.

### **3.6 summary**

Learning disorders (dyslexia and dyscalculia) appear more frequently in boys with Duchenne. These learning disorders can be subscribed to problems in automatization. Early detection and treatment is of great importance and professional and psychological examination should be considered. Overall, boys with Duchenne can keep up with their peers in the regular education program. However, sometimes special education might be required.

## 4. Psychosocial adjustment and normal behavior problems in Duchenne





## 4.1 introduction

Emotional and behavioral problems are a normal part of a healthy socio-emotional development. When growing up everyone has to cope with normal problems such as their first punishment and their first insufficient grade in school.

Research has shown that children with physical disabilities have an increased risk of developing behavioral and / or emotional problems. This is partly due to the way children adapt to the consequences of their disability (also called “psychosocial adjustment”). Emotional and behavioral problems may arise if the child does not adjust well to his disability.

In this fourth chapter, we will discuss normal emotional and behavioral problems which also occur in boys with Duchenne. First we will discuss the various aspects of psychosocial adjustment. Then we will discuss the theme of learned helplessness, which is a very important theme for Duchenne boys. Different strategies on how to deal with these problems will also be discussed.

## 4.2 psychosocial adjustment in boys with Duchenne

Psychosocial adjustment can be described as the way one adjusts to difficult and stressful events associated with Duchenne. In a large-scale questionnaire study conducted in cooperation with the Duchenne Parent Project (Hendriksen et al, 2008) in the parents of 289 boys and adolescents with DMD, it was shown that 17% of the boys with Duchenne had a low score for psychosocial adjustment. This means that the vast majority, 83% of them, are adapting well to the consequences of their dystrophy. It also shows that the adaptation scores of boys with Duchenne are not substantially different from those of boys with other chronic diseases (such as cancer and diabetes). However, it appears that adjustment problems are higher than in a group of healthy peers, which showed 2% low adjustment scores.

### BOX 18: Questionnaire on psychosocial adjustment (PARS-III).

The questionnaire on psychosocial adjustment that is now available (PARS-III) can help professionals to map and follow the socio-emotional development of a boy with Duchenne (this list is obtainable for professionals from Duchenne Parent Project).

In addition, this study revealed a number of other important findings that help us to understand why the boys behave in a certain way and may therefore be useful in dealing with the boys:

- As boys with Duchenne grow older, their adjustment scores improve. This means the boys learn to cope better with their disability;
- The period from age 8 to 10 years is characterized by the lowest adjustment scores. Clinically, this is the period of age in which the boys begin to realize the consequences of the disease, because walking gets more and more difficult and they start falling more frequently;
- Psychosocial adjustment is related to six domains of functioning, namely: relationships with friends, dependence, hostility, productivity, anxiety/ depression and withdrawal. It appears that the score for “relationships with friends”, unlike the other domains, gets lower as the boys grow older. Other studies have shown that parents and teachers need to encourage social relationships and social support. The scores for anxiety / depression and withdrawal remain stable as the boys grow older. Dependence, hostility and even productivity, however, improve the older the boys get.

### BOX 19: Stimulating relationships with friends.

“I deliberately let my son participate regularly in the scouts. This enables him to spend more time with classmates and other friends.”

## 4.3 learned helplessness in boys with Duchenne

An environment in which people perceive no control over what happens to them, can lead to what is called “learned helplessness”. This learned helplessness may play a role in chronically ill children, especially in children with a progressive muscle disease such as Duchenne. Understanding this mechanism of learned helplessness is an important guideline in dealing with boys with Duchenne in education, therapy, and social contacts.

A boy with Duchenne may experience learned helplessness when he goes through a lot of trouble for something, without achieving a direct result. This has to do with what is called “action-effect-affect” chains. The boy makes an effort (action), but experiences no effects (effects). For example: intensive physical therapy and walking with long leg braces does not lead to progress. Moreover, despite these efforts the boy still continues to lose functions! When an action does not have any impact, one is inclined to give up hope (affect; emotional reaction) and make no further effort.

Learned helplessness can lead to emotional reactions (e.g. apathy, depression) and behavioral problems (passivity). It is therefore important that parents, teachers and therapists deliberately build action-effect-affect chains in dealing with these boys. Parents and therapists need to choose activities that the boys can perform (action). If they experience an immediate consequence (effect), they get a positive feeling (affect). This lends them a sense of control. As a tangible result is achieved, they are motivated in different ways to keep on training. This applies to daily training exercises, but also to school.

### BOX 20: Building action-effect-affect chains.

	<u>Action</u>	<u>Effect</u>	<u>Affect</u>
<u>Negative:</u>	I do my best in physiotherapy	I am dependent on a wheelchair	Sadness / disappointment because I can not walk independently
<u>Positive:</u>	I practice driving my wheelchair	I am self-reliant and independent	I am happy and relieved about all the things I can still do

## 4.4 how to deal with emotional problems?

In the introduction, we already mentioned that growing up entails normal emotional and behavioral problems. For boys with Duchenne there are, besides the normal stress situations in which these emotions appear, even more stressful situations related to their disease (e.g. hospitalization, physical complaints, wheelchair dependency, etc.).

There are two age periods in which it is known that boys with Duchenne may have more emotional difficulties:

- When the boys start falling down and are unable to stand up on their own; this is a period when they often feel anxious;
- When they become wheelchair dependent. Because of their loss of independence, they respond with hostility or even depression. On the other hand, a wheelchair may also bring relief.

This confrontation with the consequences of the disease can lead to all kinds of normal emotional reactions and are part of the process of acceptance. It is important to respond the same way you would to any other child, particularly as boys with Duchenne indicate that they want to be treated and educated the same way as their peers and siblings.

## BOX 21: Stages of acceptance (Kübler-Ross).

In dealing with emotions it is important to understand what five stages we go through when confronted with a severe stressor like loss of function. Dr. Elizabeth Kubler-Ross' in 1968 developed a theory of grief that can be used to describe the stages a person goes through when handling loss. They are also called the five stages of acceptance. Your boy but also parents go through these normal phases.

Phase 1: Denial: "no not me.."

Phase 2: Anger: "why me, it's not fair"

Phase 3: Bargaining: "Yes me, but I still want to ....."

Phase 4: Depression: "I'm so sad, why bother with anything..."

Phase 5: Acceptance: "I am going to have a worthwhile life".



An important strategy for reacting to the boys' emotional outbursts is positive parenting. This means (amongst other things) that the boys learn how to cope positively with stressful and emotional events. Emotion coaching is an important strategy that can be applied by parents, teachers and therapists. The name already explains the core strategy: one coaches or guides the emotions of the boy in an active manner. The following five steps are used in emotion coaching:

**Step 1:**

Pursue emotional awareness. You need to be aware of the boy's emotional feelings. In addition to fun as a positive emotion, three negative emotions can be distinguished: anger, sadness and fear. Awareness is the starting point and the key to change!

**Step 2:**

Think of emotions as learning opportunities. You can also learn from negative experiences; however unpleasant they may be, you can always learn from them. The question we must ask is: how can I teach him to do better/act differently next time?

**Step 3:**

Listen carefully to the boy. Listening well is the next step in the emotion coaching process. Good listening means that people immerse themselves in the emotions of others. If you want to understand the boy, it is very important to put yourself in his shoes and see the world through his eyes.

**Step 4:**

Give emotions a name. Help the boy to name his emotions. If he gets angry, afraid or sad, try to label this. It is useless to suppress emotions or pretend they are not there. Labeling emotions changes an uncomfortable feeling into something with a name, and makes it normal and negotiable.

**Step 5:**

Solve problems, but also set limits. When you have listened to the emotions carefully and labeled them together with the boy, you are better able to find possible solutions. Work out possible solutions together, or help him to choose a possible solution. Note: It is also important to set limits. Make clear that the feelings themselves are not the problem, but that the behavior accompanying them is not always acceptable.

For example: the boy may be angry with his sister, but he should not say nasty things about her. Feelings are acceptable, but not all kinds of behavior are. It is therefore important to set behavioral limits.

It is known from scientific research that emotion coaching is an effective and relatively easy way to make children resilient and teach them to deal with emotions and difficult situations. We also know that emotion coaching helps to build up confidence and self-esteem. It is therefore a powerful tool in raising boys with Duchenne.

However, do not use emotion coaching in all situations, because it can have the opposite effect. Do not use it if:

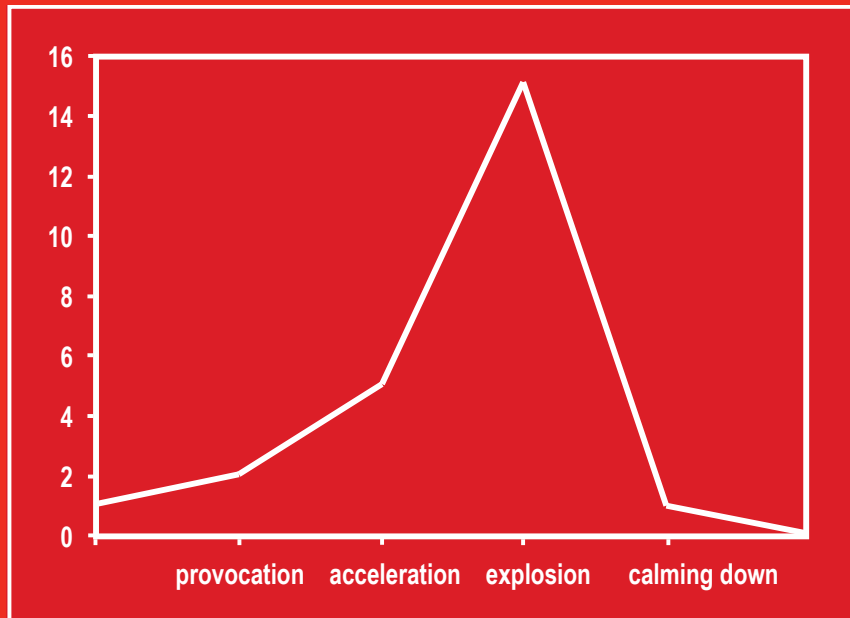
- You do not have the time or peace of mind;
- Others are present;
- You are angry, irritated or tired;
- You have already punished him for unacceptable behavior and restored order;
- He is in the middle of a tantrum (see paragraph 4.5 below);
- He is pretending to be emotional.

## 4.5 how to deal with (temper) tantrums and fits of anger

Tantrums are normal behavioral problems associated with the process of growing up, also for boys with Duchenne. They are a form of attention seeking behavior. Children and adolescents have tantrums because they want attention, and it does not matter whether that attention is positive or negative. They want 100% attention. If you lose your patience in such a situation, or start having a tantrum or outburst yourself, you are giving them exactly what they want: (negative) attention. However, you are also showing them that it is perfectly normal to have a tantrum; you have become equally angry yourself. It is best to ignore a tantrum, but that is difficult and not always possible.

In order to handle a tantrum, it is important to be aware of the normal course of a tantrum. A tantrum has four consecutive stages: there is always a reason, however small or meaningless; then the tantrum escalates to the point that it finally ends in an explosion; the last stage is the cooling down phase.

Figure 5: The course of a tantrum



During the first two phases it is important to respond clearly, to set limits and to try to prevent the tantrum escalating to the phase of explosion. Once a tantrum has reached the explosion phase, the boy will no longer be approachable and the tantrum is in fact unstoppable. Do not try to stop it, because this is counterproductive. Prevent further trouble and wait for the cooling down phase. Then the boy will be approachable again. You can now define the negative emotions and try to coach them: “I can see that you’re upset, but there’s no point in trying to get my attention by yelling. It is far better just to ask me.”

Sometimes you can use a ‘time out’ to help him cool down. Note: a time out is not a punishment! It’s a way of helping the boy to cool down. Let him know this. Moreover, a time out does not have to last longer than a few minutes. The time out is best done in a boring room with nothing to do (no computer or television), or in a place where other children cannot pay any attention to the boy’s behavior, such as in the hallway at school. This will guarantee peace and promote cooling down.



The best way to set boundaries is to take away something positive and make positive comments on things that are going well. For example: "I am taking away your play-station for a little while now, because I do not want you to hit your brother if you do not get what you want. When you have made up with your brother, you will get your play-station back." This is called positive feedback and enhances and encourages good behavior. Praising him when he has managed to cool down on his own after a tantrum can also do this.

Finally, tantrums are not usually a cause for concern because they generally diminish and eventually disappear by themselves (usually after infancy). However, when tantrums persist, worsen or happen more often you might consider involving a specialist, such as a child psychologist. It is best to discuss this with your son's medical specialist, who can refer you. Turning to a specialist may be considered if:

- You are not comfortable with your responses after the tantrum;
- You feel helpless in dealing with the tantrums and have given up hope of improvement;
- The tantrums cause many negative feelings and stress within the family;
- The boy is hurting himself or others during his tantrums.

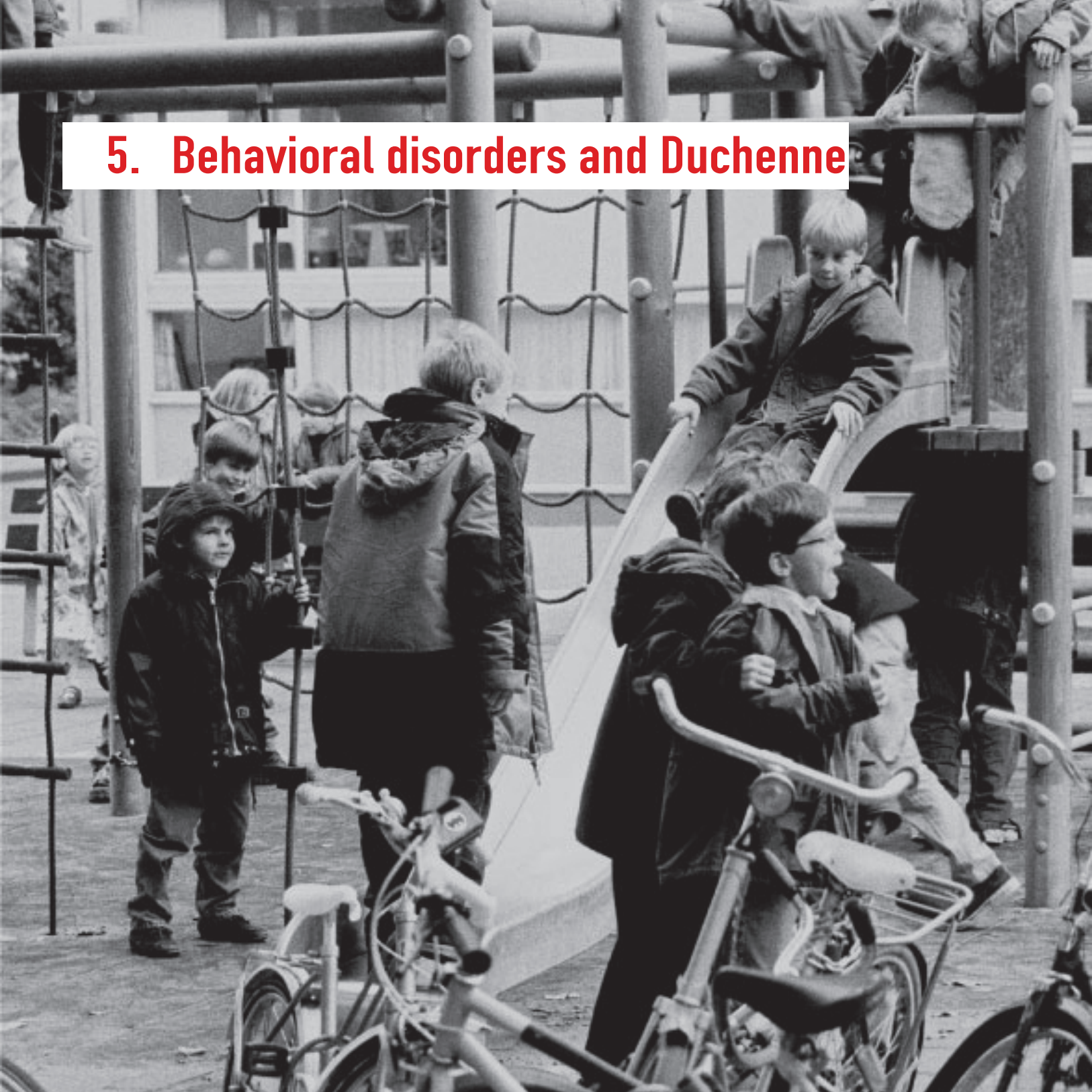
#### BOX 22: The adolescent brain.

Adolescents have to take on more and more responsibilities. This also applies to Duchenne boys. The adolescent brain has received increasing attention in recent years because it is known that teenagers' brains are not yet mature; the adolescent brain is not complete! This is why teenagers have tantrums: they react more violently than adults to certain events and their ability to put the brakes on their behavior is either lacking or insufficiently active. Feelings can occur on all different levels, from very sad to extremely exuberant. We also know that the adolescent brain has more difficulty empathizing with others, so their behavior is often very self-centered (focused exclusively on themselves). For this reason adolescents - even more than other children - need structure and it is very important to set clear boundaries

## 4.6 summary

- Growing up brings normal negative emotions and behavior problems (e.g. tantrums);
- Boys with Duchenne have the same ability to adapt as boys with other chronic diseases;
- The risk of learned helplessness can be constrained by building action-effect-affect chains;
- Accept all of the child's feelings, but set limits on negative behavior: use the five steps of emotion coaching;
- Either respond immediately to tantrums or wait for the cooling down phase;
- Time out is not a punishment but a cooling down tool. Encourage desirable behavior by rewarding it.

## 5. Behavioral disorders and Duchenne



## 5.1 introduction

We have not known until recently that boys with Duchenne may develop behavioral disorders more frequently than their peers. As there has been little research on this topic, one should be cautious in drawing premature conclusions and, for instance, seeing behavioral disorders that are not present. If you have any doubts or questions, you should ask your specialist for advice

The behavioral disorders described in this chapter are essentially different from the normal behavioral problems described in the previous chapter. Behavioral disorders are about behavior that:

- Leads to stagnating development in the boy;
- Appears more frequently and more severely than average occurrence;
- Is not caused by specific circumstances;
- Has been present for a long time (at least more than 6 months)

In this chapter we will shortly describe the three behavioral disorders which are known to be relatively more common in Duchenne: (1) Autism Spectrum Disorder (ASD), (2) Attention Deficit Hyperactivity Disorders (ADHD) and (3) Obsessive Compulsive Disorders (OCD).

It is important that parents, teachers and other people involved are aware of the possible coexistence of behavioral disorders and Duchenne. This can improve their understanding of some of the boys with Duchenne. This chapter will only describe the fundamentals of the three disorders. Also remember while reading this chapter that every boy with Duchenne is unique and that behavioral disorders can accompany Duchenne, but do not necessarily do so. These behavioral disorders are not even present in the majority of Duchenne boys.

## 5.2 Autism Spectrum Disorders (ASD)

Autism spectrum disorders are about problems in three areas: (1) impaired social interaction, particularly with regard to reciprocity; (2) impaired verbal and nonverbal communication; and (3) imagination deficits. As a result, children with autism often show the following typical behaviors:

- A limited repertoire of activities and interests;
- Stereotype behavior (repetitive behavior);
- Having difficulty with change and seeking for sameness;
- Focus on details;
- Finding it difficult or impossible to empathize with others.

These behaviors make raising children with ASD more difficult, because educators / parents find it difficult to understand these children and thus to influence and stop them.

Autism is not curable and cannot be prevented. Someone with ASD will have these problems for the rest of his life. Treatment aims to keep it from getting worse and to prevent additional problems. An important starting point is to adapt the environment to the boy. Good guidance and information for parents, family members and other professionals is of the utmost importance. In Chapter 8 we present websites and other relevant material for further reading.

**BOX 23: Characteristics of ASD in behavior that need to be kept in mind.**

- Difficulty with change
- Focus on details
- Egocentric (inability to empathize with others)

Two studies show that ASD is relatively more common in boys with Duchenne (Wu e.a., 2004; Hendriksen & Vles, 2008). ASD has been diagnosed in approximately 3% of boys with Duchenne. A recent study in a U.S. metropolitan area estimated that 3.4 of every 1,000 children between 3 and 10 years old were autistic (source: National Institute of Mental Health: [www.nimh.nih.gov](http://www.nimh.nih.gov)). This means that ASD problems are more common in Duchenne than in the average population.

## 5.3 Attention Deficit Hyperactivity Disorder (ADHD)

Attention deficit hyperactivity disorder (ADHD) is one of the most common childhood disorders and can continue through adolescence and adulthood. Symptoms include difficulty staying focused and paying attention, difficulty controlling behavior, and hyperactivity (over-activity). ADHD has three subtypes: Predominantly hyperactive-impulsive, predominantly inattentive, and combined hyperactive-impulsive and inattentive. Common problems in children with ADHD are

- Emotional regulation (self-control);
- Controlling behaviors and activities by stopping them once they have started;
- Flexible thinking;
- Planning and problem solving;
- Learning from their own mistakes.

The fundamental problem of ADHD is a control and stopping problem. This means that children with ADHD have more difficulty stopping doing something they have started doing. Therefore, parents and teachers have to repeat their instructions more frequently. Children with ADHD often have difficulty listening, organizing and following up assignments, instructions and orders. Playing together with other children can therefore be difficult and cause problems. Children with ADHD may appear restless and sometimes find it difficult to sit still. The diagnosis of ADHD is usually made from 6 years on, the age at which children in grade three start with classroom instructions.

One of the first studies on the prevalence of ADHD in boys with Duchenne (Hendriksen & Vles, 2008) showed that 11.8% of boys with Duchenne were diagnosed with ADHD. Usually the percentage is between 3 and 6%. Although ADHD is more common in boys with Duchenne compared to healthy peers, only a small percentage of boys with Duchenne have ADHD. For further information we refer to the websites concerning ADHD (Chapter 8).

#### BOX 24: Treatment of ADHD problems.

To treat ADHD, which causes distress and/or stagnating development, a trial with Ritalin medication can be started. This medicine has long been used in the treatment of ADHD and is the only 'evidence-based' treatment to date. Ritalin stimulates the "stopping" centre in the brain and can thus contribute to better control over stopping behaviors. Ritalin has a positive effect on approximately 70% of patients with ADHD. The main side effects of Ritalin can be loss of appetite and difficulty falling asleep. As yet, there has been no research on the effect of Ritalin on boys with Duchenne. It is important to only use this medicine in boys with Duchenne on prescription from a medical specialist, who can best assess the risk of side-effects and discuss them with parents.

## 5.4 Obsessive-Compulsive Disorders (OCD)

Obsessive-Compulsive Disorder, OCD, is an anxiety disorder and is characterized by recurrent, unwanted thoughts (obsessions) and/or repetitive behaviors (compulsions). Repetitive behaviors such as hand washing, counting, checking, or cleaning are often performed with the hope of preventing obsessive thoughts or making them go away. Performing these so-called "rituals," however, provides only temporary relief, and not performing them markedly increases anxiety. OCD is more common in boys with a progressive physical illness, such as Duchenne. Research suggests that OCD is twice as common among boys with Duchenne. Usually, the percentage is around 2.3%. Despite this double risk, it still affects only a small percentage of Duchenne boys.

#### BOX 25: OCD in everyday life: A parent's statement.

Alex's most striking feature is that he increasingly wants to have everything under control. We believe that the more he loses control over his body, the more he wants control over everything and everybody.

## 5.5 conclusion

Although behavioral disorders are more common in boys with Duchenne, it appears that overall only a small percentage of boys manifests these disorders in addition to their neuromuscular disorder. The cause of this joint occurrence of a muscle disease and a neuropsychiatric behavior disorder is still not fully understood. A relationship with brain functioning is assumed: the dystrophin protein probably plays a role. In addition, psychological and psychosocial factors such as processing the diagnosis, fatigue, stress within the family and learning difficulties at school may contribute to the development of these disorders. We assume a relationship between genes (responsible for the lack of dystrophin in the brain), behavior and environment (psychosocial aspects). Further and more extensive investigation of these behavioral disorders in Duchenne is required.

If you want more information about ASD, ADHD or OCD, you can make use of the websites listed in Chapter 8. Note also that only a specialist such as a child and adolescent psychiatrist or child and adolescent psychologist can make a definite diagnosis of ADHD, ASD or OCD after examining the boy. The medical specialist treating your son should refer to one of these specialists.



## 6. Talking about Duchenne



## 6.1 introduction

When parents tell their son about Duchenne and its consequences, now and in the future, this can be highly emotional. To inform boys with Duchenne about their disease is not easy. However, it is very important and inevitable. Research dated from 1983 already showed that parents' biggest worry is "when to tell my son what?"

It is difficult to give a clear-cut answer to this question because it depends on different factors. Moreover, it has to do with the way parents have accepted the diagnosis themselves. Parents have difficulty talking to their son about his disease with full confidence or belief. This chapter contains some guidelines and recommendations that may help you as a parent in leading such conversations.

Besides the importance of parents talking to their son about Duchenne, it is also important that teachers and perhaps classmates are informed. This will be dealt with further on in this chapter (paragraphs 3 and 4).

## 6.2 informing your son

Do not just come out and tell the boy out of the blue that he is suffering from a disease. It is better to create an opening to a conversation, for example: "You may be wondering what all those examinations in the hospital are for". Then wait, listen to your son and answer any questions he may come up with. If he does not ask any questions, do not ask any or give any answers yourself. It is important to let your son take the lead and not to feel obliged to say something (Charlotte Thompson 1999). Keep in mind that children do not ask more than they are able to cope with: if they do not ask anything, then they are just not ready for it yet.

How questions that your son brings up are best answered depends on his age. The way he sees his disease also depends on his age. An important turning point is around the age of 7 (and varies from the age of 6 to 8). Boys under the age of 7 are, as psychologists call it, egocentric in their thinking. This means they look at the world from their own point of view

only. At this stage Duchenne is just one of the many parts of their life and they don't realize the (long term) consequences of it yet. Boys with Duchenne under the age of 7 are, like other children of this age, only interested in the here and now. They do not think about their future yet. The only thing they want is to play soccer, except that their legs are "naughty" or their muscles are "tired". Parents need to be conscious of their own point of view, which differs from their son's. Parents are more focused on the future. Parents do best in talking to their sons about Duchenne by following them in the here and now: support them and keep in mind that they do not want clear answers about the future yet. Questions about the future do not play any role in their thinking until later. A good reaction is what we call "mirroring". This means that you give back what your son is asking you in your own words, like a mirror. You do not need to give a clear answer to his questions. You only have to let your son know that you have heard his worries: His fear of falling or his grief at not being able to score when playing soccer.

When the boys get older (from age 7), they will become less egocentric and are more capable of thinking about their future. They can now think about their later lives. It is not until this turning point that they begin to accept their disease and its disabilities. As the boys get older, occasional short talks can help them to talk about the different aspects of their disease.

The following guidelines may help you talk about Duchenne with your son:

- Always be honest, but do not talk too much: provide the boy merely with the information he asks for. Keep in mind that children do not ask more than they are capable of coping with.
- Reassure him. You can do this by telling him, for example: "You will get anything you need, and we know smart doctors who will do everything they can to help us".
- Be sure that the boy understands that his disease is not his fault: he is not to blame!
- Anger, grief and other emotions can emerge during such talks (see box 21). This is completely normal, so do not try to avoid it. Instead show understanding and be a coach of the boy's emotions (see paragraph 4.4).
- Do not talk about his future if he does not ask about it. If he does ask, do not avoid the subject and always be honest. But never shut the door on new possibilities by saying, for example: "There are many doctors experimenting and searching for a cure, who knows what will be possible in a few years?"

### BOX 26: “Why are my muscles less strong?”

“He always asks me why his muscles are less strong. I always tell him that he was born this way and that other people are born with other disabilities like bad eyes or bad ears....”

Talking with the boys about Duchenne helps them to clarify what is happening to them in their own minds. This lessens their fear of the unknown and enhances their sense of control over their own lives. In addition, they will be able to explain their disease to others, such as friends and classmates. So talking to the boys about Duchenne is of the utmost importance. As we have already mentioned, honesty is essential. If you are not honest with him, the boy may get confused, because he feels and sees that there is something wrong with him. Sooner or later, he will find out what is happening, on the Internet, for example. Apart from being overwhelmed by the sheer amount of sometimes erroneous information on the Internet, he will lose his faith in you. To enable him to accept his disease and learn to live with it, it is vital that the boy can fully trust the people who are closest to him.

Table 3: What words should you use and what words are better avoided when talking with the boys about Duchenne.

Avoid	Use
<ul style="list-style-type: none"><li>• Suffering</li><li>• Horrible, weakening disease</li><li>• Wheelchair bound</li><li>• Fatal, terminal, premature death</li><li>• Paralyzing</li><li>• There is something wrong with your muscles.</li></ul>	<ul style="list-style-type: none"><li>• The diagnose is</li><li>• Progressive muscle disease</li><li>• Support from a wheelchair</li><li>• A different life</li><li>• Disease which makes your muscles less strong</li><li>• Your muscles are special/ your muscles are different to others'/ tired muscles</li></ul>

## 6.2 informing your son

Parents and teachers should talk to each other before a boy with Duchenne starts school. This allows parents to provide the teacher with information such as what exactly is happening to their son, what Duchenne does to him, and how to handle certain situations. Together you can figure out a kind of strategy so as to be prepared for questions asked by the boy with Duchenne or his classmates (for example about the wheelchair or why the boy with Duchenne is not able to run so well). This helps to avoid potential misunderstandings about the boy with Duchenne within his class.

**BOX 27:** creating a justification for the things he can't do.

It may help to create a reason, which excuses the boy for the things he cannot do. All persons involved (teachers, professionals, grandparents, peers) should use the same justification: for example "weak muscles". Parents should take the lead in creating a justification that fits with their son.

In addition, parents could set up a so-called student profile. A student profile is a kind of handout that consists of only one or two pages providing the teacher with the most important information about their son. The goal is to focus not only on Duchenne Muscular Dystrophy, but also on helping the teacher to understand who the boy is as a person.

Lastly, it is very important that the teacher is informed about Duchenne Muscular Dystrophy as a disease. This helps the teacher understand what is going on in the boy and his family, not just on the physical level, but also on the level of acceptance and psychosocial adjustment to the disease and its consequences.

## 6.4 informing classmates, friends, and family members.

Of course classmates, friends and family members wonder what is happening to the boy with Duchenne. They need clarification as well. Research shows that classmates of a handicapped child will often defend the child instead of teasing him if they know what is happening to the handicapped child. Moreover, if classmates know what is going on, wild rumors will be less frequent.

Classmates should only be informed if the boy with Duchenne agrees himself. If the boy does not want his classmates to be informed, he should not be pushed. If he agrees, it is his own choice to be present or not when the teacher informs his classmates. The boy makes his own choice and his choice must be respected. Some guidelines that can be helpful when informing classmates are:

- Emphasize that Duchenne Muscular Dystrophy is a disease that he was born with; it is never the boy's own fault;
- It is not contagious; other children can not catch it;
- Adjust your explanation and instructions to the developmental stage of the class. A class of young pupils should get a simpler explanation than a class of older pupils. E.g., in a class of infants, your explanation and instructions can be, for example: "His muscles are tired"; "Do not push him"; "Be careful on the stairs"; and "Help him carry heavy materials";
- If the boy with Duchenne wants to explain his disease to his classmates himself or answer their questions, your role is to support him;
- Do not 'flood' the class with too much information;
- Answer questions from the class as honestly as possible.

Guidelines that parents and teachers have found useful when informing classmates of their son or pupils are:

- Place 1 kg weights on classmates who want to participate and let them run around. This is what it is like running if you have Duchenne; undoubtedly more difficult;
- Give examples of handicapped people who still made it and even became world-famous like Stevie Wonder and Beethoven;
- Show that the biggest part of the body (30 to 40%) consists of muscles by making

a simple drawing of the human body. More importantly, show that the brain does not consist of muscles. This will help make it clear that only the muscles of a boy with Duchenne are different, but that the brain and feelings, for example, are exactly the same as their classmates’.

Useful materials for informing classmates can be found on the website of Parent Project

**BOX 28: Make agreements with friends and family about how they will inform their children.**

“One of the first things I did was e-mail our friends to ask them if they would please not tell their children that he is ill, because we want to do that ourselves when the time is right. He was not yet attending school at that time so we had to make agreements in our private lives. Unfortunately, some friends had already informed their children, and when we visited them their children would ask me: “He is ill, right?” and “Is he able to do that?”.”

## 6.5 summary

Muscular Dystrophy: [www.parentprojectmd.org](http://www.parentprojectmd.org)

- Do not just come out and tell the boy out of the blue that he is suffering from a muscle disease. It is better to create an opening to a conversation by asking a question.
- Boys under the age of 7 are egocentric and do not think about their future yet. So give them support rather than explanations. Boys over the age of 7, on the other hand, need an explanation.
- By talking about their disease they obtain clarification of what is happening. Honesty is most essential.

## 7. The future of boys with Duchenne





## 7.1 introduction

What does my Duchenne son's future look like? Parents ask themselves this question regularly, but they do not know the answer. In this chapter, we do not pretend to give an answer. After all, nobody can see into the future. What we do want to illustrate in this chapter is what Duchenne boys can achieve in their lives, how worthwhile their lives can be and how they can attain a high quality of life. We will attempt this in the following two paragraphs on independence and guidelines to improving quality of life.

### BOX 29: Positive energy.

"The fact that Duchenne is an incurable disease overwhelms you at first. Nothing matters anymore. Membership in Parent Project Muscular Dystrophy and helping to collect money can give you positive energy. Besides, in our case, it made family and friends believe they can do something. They too feel powerless and are happy to support."

## 7.2 independence and Duchenne

Formerly, people thought that because of their poor prognosis boys and adolescents with Duchenne, would never be able to get and carry out a paid job. This has now changed. There are plenty of men with Duchenne who acquire paid jobs. This is due to the significantly increased life expectancy.

A significant number of men with Duchenne complete intermediate vocational training, higher vocational training, or even acquire university degrees. These opportunities to study and have a job are possible because of new technical adaptations and the financial means to draft out their future. With the help of technical adaptations they can use a computer until a late age, and respiratory equipment enhances their life expectancy. When

they have employment and thus earn their own money and experience satisfaction, men with Duchenne can function highly independently.

Young men with Duchenne can even live independently. Although this appears impossible, it now occurs more and more frequently. They can live on their own or in a residential home for physically disabled young adults. It is even possible for them to live on their own within a relationship or to start a family. Because of the significantly increased life expectancy (see Box 2), a new group of patients has evolved with its own unique problems: adults with Duchenne. Knowledge and experience in treating and taking care of this group has just recently begun to grow (Rahbek et. al., 2005).

#### BOX 30: His own car.

“Despite Duchenne, my son handles his disease well. He is doing voluntary work, thinks positively and so has a positive influence on others. He has a driving license. He drives independently in his adapted car, using a joystick and electronic control devices.”

## 7.3 guidelines to improving quality of life

Just like anyone else, boys with Duchenne have a whole life with possibilities ahead of them. Quality of life is more important than quantity. So when a boy with Duchenne, his family and the people around him make every effort to give him a good life and try to get everything out of it, this boy may have a better life than many other (physically) healthy people. Apart from the topics mentioned in the previous paragraph (employment and studying), the life of a boy with Duchenne can be qualitatively improved by making use of the following guidelines:

- Try not to look into the future; instead, try to enjoy every day and the little things that make a day good.
- Treat him the same way you treat other (physically healthy) children. Boys with Duchenne mention this as one of their biggest wishes.

- Try to let him do the same as other children as much as possible. Let him play outside, just like other children. To avoid muscle damage it is important to use lighter materials and tools.
- Stimulate him to undertake and keep up relationships with friends. This is vital for his well-being.
- For as long as possible, let him do as much as he can by himself. It will give him a feeling of independence and self-esteem.
- Look for a nice hobby together.
- Do enjoyable activities such as day trips. Do not stop doing activities because of Duchenne. If a certain activity doesn't work any more, try to find creative solutions.

#### BOX 31: A mother about the life of her son.

Despite Duchenne, Jasper had a good and beautiful life. Every day he was busy with friends and all sorts of activities. When Jasper's mother was asked how it was possible that Jasper was such a terrific and remarkable boy, she answered that she had treated him just like she treated her other children. She expected him to do his household tasks just like his brothers and sisters and to help them the same as they helped him. He was treated exactly the same as his brothers and sisters. She also said: "We did not look too far ahead, but tried to make the most out of each day."

## 8. Useful websites



On [www.parentprojectmd.org](http://www.parentprojectmd.org) , the American parent organization of DMD, presents information on DMD as well as information on all sorts of activities as organized by parent organization. The paragraph 'understanding Duchenne' deals with aspects that have been explained in chapter 1 of this booklet.

The website [www.uppmd.org](http://www.uppmd.org) is from the international United Parent Project muscular dystrophy organizations. One can register using the following website <http://www.duchenne-community.org>, and can find here the latest articles, good tips, or post questions. Furthermore there is an activity calendar, showing the different activities in each country. Also one can keep their own blog.

<http://www.bdadyslexia.org.uk/> is a British website with a vast amount of information on dyslexia. It informs you about a lot of different aspects of dyslexia in all stages of life. Additionally they give information on useful books, they keeps track of the latest news on dyslexia. Dyscalculia is also slightly touched.

The International Dyslexia Association (IDA) is a non-profit organization that is dedicated to the helping of individuals with dyslexia, their families and the communities that support them (<http://www.interdys.org/>). The research database on this website is very abundant. The website also includes amongst others a forum, an online bookstore and offers the possibility of registering to a newsletter.

A website providing information on dyscalculia is <http://www.dyscalculia.me.uk/>. It contains information for parents and teachers and also gives access to articles on dyscalculia as published by SEN, a UK magazine for special educational needs.

The website [www.softlearning.org](http://www.softlearning.org) provides exercises helping your child to become more confident with numbers en practice simple calculation skills. It helps improving skills and insight in math. It might be of useful assistance in exercising the basic skills as they were explained in chapter 3. Different kinds of simple calculations can be performed and automatised. All exercises are presented in a child-friendly manner and environment.

Although this website looks quite straightforward it contains a lot of information on ADHD. Different aspects that can be distinguished within the scope of ADHD and ADD are discussed: <http://www.nimh.nih.gov/health/publications/attention-deficit-hyperactivity-disorder/complete-index.shtml#pub1>. As the URL reveals it is part of the National Institute of Mental Health.

The latest news and developments on ADHD can be found on <http://www.adhdeurope.eu/> and more specifically on the header 'news'.

Additional information on autism can be found on <http://www.autism-society.org/>. It not only discusses how autism is caused and what it is, but also gives significant information on the living with autism. Autism is highlighted through the lifespan. Potential treatment options are provided as well as personal notes on autism. This website is regularly updated and is easy to use, which make it an appropriate source for additional information on autism.

Valuable information on OCD can be found on the website of the International OCD Foundation (IOCDF): <http://www.ocfoundation.org/index.aspx>. This website questions several aspects of OCD and gives answers to them. There is also the possibility of downloading brochures with useful information on OCD.

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LEADING THE FIGHT TO END DUCHENNE