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COGNITIVE FUNCTIONING AND AUTONOMY OF PATIENTS WITH DUCHENNE MUSCULAR DYSTROPHY

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SUMMARY

Background: Cognitive problems and a deepening dependence on one's immediate environment inherently accompany Duchenne Muscular Dystrophy (DMD). The disease is progressive, and the size of the dystrophin gene determines the extraordinary complexity of the causes of this disease at the genetic and molecular level. The aim of the study was to characterize the cognitive problems and the extent of independence of patients with genetically confirmed DMD. An attempt was also made to reconstruct the patient's life history in three periods: before the appearance of the first symptoms, during the search for a diagnosis and after confirming the diagnosis of DMD.

Material/ Methods: The study group consisted of male patients between 10 and 13 years of age (N = 14). The *Diagnosis of Cognitive Functions Battery – PU1* and an experimental tool for studying autonomy were used. Information on the condition of the attention, memory and executive functions of patients was obtained. The study of autonomy measurably supplemented knowledge in terms of the degree of the patients' dependence on the environment with regard to everyday functioning.

Results: The best functioning component of attention in the examined patients is *orientation* (o) (13 patients achieved average results in this aspect). *Selectivity* (s) turned out to be the weakest component, as only five patients obtained average results (the others obtained low results) in this aspect. *Autonomy* results (AU) indicate group diversity and inter-subject variability in the disease progression (subjects scored from 6 to 47 points).

Conclusions: The clinical picture of the disease is not homogeneous. Patients, despite their similar age, differ in the progression of the disease and the resulting effects. This induces the need for an individual approach to each patient and the preparation of a unique set of therapeutic interactions for each of them.

Key words: rare disease, attention, memory, executive functions

BACKGROUND

Duchenne muscular dystrophy is a rare genetic disease (the incidence of 1/3500 male births). It is inherited in a recessive manner on the short arm (p) of the X chromosome (Jorde, Carey, Bamshad, 2010). The gene whose mutation causes dystrophy is one of the longest known genes to date. It consists of approximately 2.4 Mb base pairs and contains 78 introns. DMD binds to a mutation in the gene that encodes the dystrophin protein. Dystrophin is found in many body tissues, but mainly in skeletal and brain muscles (Emery, Muntoni, Quinlivan, 2015, Li et al., 2015, Nakamura et al. 2013).

The first symptoms of the disease appear around the age of 3, at which time the child experiences difficulties with locomotion and postural control (Jorde et al., 2010). Muscular dystrophy causes gradual loss of muscle functions, starting from the lower limbs and immobilization of the iliac rim, ending at the upper limbs and immobilization of the shoulder girdle (Li et al., 2015). Around the age of 6, patients develop so-called *Goverse syndrome*, which consists in rising to a standing position by supporting oneself with one's own body (Emery et al., 2015). At this stage of the disease you can also notice gradual hypertrophy of the calves, a shortening of the Achilles tendons or curvature of the spine (scoliosis, kyphosis, lordosis) (Jorde et al., 2010). Most patients around the age of ten can no longer move independently (Thomas, Rajaram, Nalini, 2014; Emery et al., 2015). In addition, there are respiratory and cardiac problems of varying intensity that impede daily functioning, forcing patients to be treated with the need for frequent hospitalization (Raman et al., 2017, Deconick, Dan, 2007, Quinlivan, Lewis, Marsden, 1996).

Some of the boys encounter difficulties in cognitive functioning. Dystrophin, whose production in these patients is abolished by a mutation in the gene, is also present in the brain, and takes on different forms depending on the exact place and type of mutation (Emery et al., 2015). Reduced glucose metabolism in structures that are rich in dystrophin in healthy people, explains the cognitive problems faced by some of these patients (Cyrulnik, Hinton, 2008). These structures include cerebellum, the temporal elements of the limbic system, the somatosensory cortex and the anterior part of the frontal lobes (Schara, Busse, Timmann, Gerwig, 2015), which are an important element of proper cognitive functioning. Many patients have problems with the following: language functions (Hoskin, Fawcett, 2014), reading and writing, memory capacity (Milic Rasic et al., 2014), spatial organization (Hendriksen, Vles, 2006), sequential processing (Schara et al., 2015); patients also display lower results in terms of general intelligence (Mehler, 2000). Researchers look for relations between the exact type and location of mutations and the presented difficulties in psychological functioning. Some of them suggest that the most serious intellectual problems can be expected when the mutation occurs in the distal parts of the described gene, and minor problems when it occurs in its proximal parts (Emery et al., 2015; Muntoni, Torelli, Ferlini, 2003). Impeded cognitive functioning is most likely re-

lated to the pleiotropic effect of the genetic mutation, but it is also associated with specific dystrophin protein isoforms within the nervous system. The following may be identified as the key isoforms: Dp427, Dp260, Dp140, Dp71 (also present in the brain) and Dp116 (present in Schwann cells). Research indicates a special role for the mental functions of Dp71 isoforms (Daoud, Angeard, Demerre, 2009) and Dp140 (Bardoni, Felisari, Sironi, 2000), the mutation of which will often cause mental retardation.

Our goal was to estimate the level of cognitive functioning in terms of attention, memory and executive functions as well as autonomy in everyday functioning, in a group of boys of similar age, diagnosed with DMD.

MATERIAL AND METHODS

The study group consisted of boys between 10 and 13 years of age (N = 14) with diagnosed DMD confirmed by genetic tests. The study was conducted at the Clinic of Paediatrics, Day Care Unit, at the Medical University of Gdańsk. The patients are provided with medical care at the Rare Diseases Centre of the University Clinical Centre's Genetic Counselling Centre. This care includes a number of examinations and consultations with specialists in various fields, including a cardiologist, rehabilitator, pulmonologist, ophthalmologist, neurologist, endocrinologist, dietician, and from the beginning of the research herein presented, also a psychologist. It allows one to provide DMD patients with comprehensive and specialist care.

Cognitive Functions (CF)

A single examination lasted for approximately 90 minutes. The cognitive functioning of the patients was diagnosed using the *Diagnosis of Cognitive Functions Battery - PU1*. This tool consists of 15 tasks, divided into three blocks: *Attention, Memory, Executive functions* (Borkowska, Sajewicz-Radtke, Lipowska, Kalka, 2015). This was used because of the possibility of obtaining detailed measurements of cognitive functioning indicators; e.g., the attention function was divided into such components as: selectivity, orientation and inhibition. Memory was associated with the visual-spatial processing and phonological loop.

Autonomy (AU)

Demographic data (age, sex, age at the time of the diagnosis, age at which the first symptoms of the disease were observed, etc.) was collected from the parents, together with information about the extent of independent functioning, which was obtained using an experimental tool (Anikiej, 2017). The parents' task was to determine the extent to which a child performs a given activity independently and in which he or she needs help or is totally dependent on the caretaker. In this way, the patient collected points determining his degree of autonomy (maximum 48 points). For each of the 16 statements he could receive from

0 (when he was completely dependent) to 3 points (when he was fully independent of the caretakers).

RESULTS

The table presents the individual stages of the child's disease development from the perspective of his parents. The first indicator WD (*without diagnosis*) shows the moment in the life of the child in which the parents noticed the first symptoms. Until that time, they had been unconscious of his child's illness. This period can last from the first months of the child's life (0.5 years) to even 9 years of age. The second indicator SD (*searching for a diagnosis*) is the time from the moment the first symptoms were noticed, to the diagnosis, i.e., the period when parents are looking for an answer to the question 'what is happening to our child?': a period full of questions, uncertainties and fear for the child's future. In the case of the analysed study group, this period lasted a few years (even up to 10-years of age). The last indicator D (*life with diagnosis*) is the time from the patient's diagnosis of DMD to the time of this examination. It was a time of adaptation, treatment and therapy.

The group of DMD patients examined in this study presented a non-harmonious picture of the disease. At a similar age, likewise motor problems could be expected. Some of the patients, however, did not lose their ability to walk independently ($n = 4$), while the rest moved using a wheelchair. In addition, some imbalance was observed also in terms of the disappearance of upper limb motor function. Some of the patients coped well with tasks that required writing/drawing from them, but for the others it was difficult. These differences were demonstrated in the course of the assessment of *Autonomy*, where the caretakers could comment on the independence of their child's functioning.

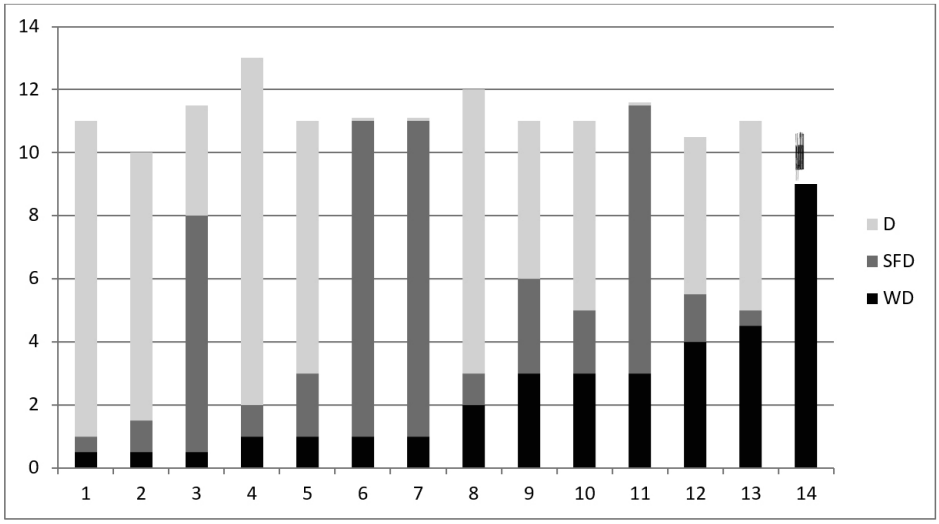

Figure 1. Stages of a child's life with DMD

Cognitive Functions

As far as cognitive functioning is concerned, patients achieved average and low results. None of them achieved high results. The collected data allow one to develop a cognitive profile of DMD patients , which is an important starting point for further treatment in improving their functioning in different environments.

Attention (A)

Three attention components were investigated:

Attention Orientation (o).

A(o) is a reaction to an emerging sensory stimulus with different modality. Almost all patients (n = 13) achieved average results in this function. One patient obtained a low result in this function.

Attention Selectivity (s).

A(s) is the ability to recognize the attributes of stimuli against a background. In terms of this component, 5 patients achieved average results while the others (n = 9) achieved low results.

Attention Inhibition (i).

A(i) is the possibility of extinguishing the response to the stimulus in case it is inadequate. The majority of patients (n = 9) achieved average results on this subscale, while the rest of the patients (n = 5) achieved low results.

Memory (M)

Two memory components were examined:

Phonological loop (pl).

The use of the construct M(pl) is based on the short-term maintenance of verbal information about, inter alia, items, sentences and numbers. In this respect, the patients obtained mostly (n = 10) low results, and the remaining patients (n = 4) achieved average results.

Visuo-spatial sketchpad (vss).

M(vss) is a memory component that allows you to maintain visual information and manipulate visual and spatial imaginations. In this respect, the patients mostly (n = 8) achieved low results, while the remaining patients (n = 6) achieved average results.

Executive Functions (EF)

EF is a very important construct for regulating behaviour, creating plans and organizing an individual's activity. In terms of this component, patients obtained in most cases (n = 11) average results, with a few (n = 3) low results. None of the patients with DMD achieved any high results in any of the cognitive functioning components presented above.

Autonomy (AU)

The patients' functioning during everyday activities was analysed. Caretakers were asked to assess how many different activities, such as washing, dressing, eating or doing homework, their child did on their own, and to what extent they

needed their parents' help. This is an important element both for their functioning in the school area, and within the context of the failures and problems encountered in other environments. The maximum result that could be obtained was 48 points. The lowest result obtained was 6 points, and the highest was 47 points.

The motor problems of patients with DMD may cause a number of difficulties associated with diagnosing them. DMD patients examination requires some flexibility on the part of the diagnostician. It is important to take into account patients' motor problems. It is necessary to design the space around the patient to ensure full access to all test materials (patients are often unable to freely reach out for objects placed at a distance on the table). Remember that the position has to be convenient and does not hinder the tracking of tasks. It is also important that the table at which we perform the tasks is of the right height (the possibility of approaching it with a wheelchair). The examination should take place during periods of the patient's well-being. Patients may also experience diverse and extreme emotions. Some researchers emphasize, among others, the need to focus on a patient's positive traits, which may help to support concentration and prevent fatigability (Emery et al., 2015).

Patient motor slowdown should also be taken into account; this feature has been reported on by many researchers (Emery et al., 2015, Thomas et al., 2014; Nakamura et al., 2010). Some of the psychological diagnosis tools require time pressure, consequently they will not work in the diagnosis of DMD patients. In the test applied in this study (PU1), a few tasks required time measurement. In the case of tasks requiring only a recalling from long-term memory, certain time periods were introduced, while in the case of tasks in which patients were to draw or fill in something, this factor was not taken into account.

Table 1. Stages of life versus cognitive functioning and autonomy of patients with DMD

		Time of life from the parent`s point of view			AU	Attention*			Memory*		Executive functions*
ID	Age	WD	SD	D		o	s	i	pl	vss	
1	11	0.5	0.5	10	35	5	5	5	2	1	4
2	10	0.5	1	8.5	47	4	1	4	3	1	4
3	11.5	0.5	7.5	3.5	45	7	1	6	2	3	5
4	13	1	1	11	6	3	2	1	2	1	4
5	11	1	2	8	29	4	3	4	4	6	6
6	11	1	10	<1m	23	6	3	1	2	1	5
7	11	1	10	<1m	21	5	4	2	4	5	3
8	12	2	1	9	38	6	6	5	5	6	6
9	11	3	3	5	37	5	1	4	1	1	4
10	12	3	2	6	45	6	7	6	4	6	6
11	11.5	3	8.5	<1m	10	5	2	1	1	2	3
12	10.5	4	1.5	5	41	6	4	4	2	3	6
13	11	4.5	0.5	6	35	6	3	5	3	5	4
14	11	9	0	2	32	5	2	1	1	2	3

Note. WD – *time without diagnosis*, SD – *time of searching diagnosis*, D – *time with diagnosis*, AU – *autonomy*, o – *orientation*, s – *selectivity*, i – *inhibiting*, pl – *phonological loop*, vss – *visual-spatial sketchpad*.
*all of the results of attention, memory and executive functions presented in stens

During the study, there was no need for the permanent presence of a caregiver. Patients remained alone with the psychologist during the examination. Many patients even demanded that the carer stayed outside the room.

DISCUSSION

The cognitive development of 14 DMD patients aged 10-13 years was assessed. In addition, demographic data and indicators of the patients' daily functioning were collected (from the caregiver. It is worth noting that the examined patients formed a very homogeneous group due to their age.

The data collected from parents turned out to be a rich source of knowledge. There is a period of undisturbed development in the course of DMD, although interviews with close relatives indicate that even at this time they are concerned about certain types of their children's behaviour (the *floppy child* syndrome) (Emery et al., 2015). Usually, this period lasts for about 3 years, but the presented study showed lower (0.5 years) and higher values (4 years and more). This implies that the progress and dynamics of DMD changes can be varied. This may be related to the type of genetic mutation (Schara et al., 2015). Anomalies in the coding of the dystrophin protein may be caused by deletions in different axons and involve larger or smaller parts of the genetic material (Muntoni et al., 2003).

The case of a family who indicated that the first 9 years of a child's life had been an asymptomatic period, is particularly interesting. This can be explained in two ways. The first explanation is that it could be a time when the family expected a different diagnosis, saw the symptoms, but did not assign them to this particular medical condition. The second explanation is a complete rejection of the disease symptoms and a possible further sense of guilt because of the late start of the child's treatment.

Living with motor disabilities will have an impact not only on social development, and consequently the emotional, but also on cognitive functioning (Cyrulnik, Fee, DeVino, Goldstein, Hinton, 2007, Gerc, 2011). Problems in cognitive functioning can have many causes, including metabolic changes, cardiological problems (Magalhães Pereira, Areias, Areias, Dias da Silva, Peixoto, 2011; Maryniak et al., 2012), social isolation (Pąchalska, Góral-Półrola, Mueller et al. 2017) . Our goal was to find different patterns of boys' functioning in order to propose solutions to each of the families to compensate for these deficits.

Patients with DMD achieved low and average results in terms of attention selectivity and inhibition. Deficits in this area are often associated with impulsive behaviours, emotional retardation, problems with adapting to new situations, understanding the activities required from them, maintaining positive social relations, initiating and completing entrusted tasks (Borkowska et al., 2015).

The lowest results in terms of attention were obtained in the selectivity component. This function is associated with the ability to choose one stimulus against the other. Therefore, it is advisable to follow certain rules when presenting important information, both at school and at home. The patients demonstrate poorer selec-

tivity in terms of visual modality, so it is recommended that stimuli should also be presented using other modalities. In addition, relevant information should be expressive and distinguish significantly from the less important content. Large pictograms and verbally repeated instructions work particularly well.

Another important component, where we can expect some interference, is memory. Children having deficits in the working memory are characterized by such unwanted forms of behaviour as: getting lost in during tasks, abandoning tasks before their completion, frequent failures while solving school problems, confusing commands, mixing information. To facilitate their functioning in the social environment, it is important to divide instructions into smaller parts (a smaller range of elements kept in the short-term memory), but also to use different types of hints so that the patient can recall information on their own. For example, a good solution is to reproduce instructions using pictures that the patient can always relate to when he forgets what to do.

As far as EF is concerned, the patients mostly obtained average results, but these were close to the lower limit. It is worth taking this into account when working with patients, also remembering the deficits encountered in the other areas. The observation of this group of patients shows that they often lose track of instructions, have problems with continuing the tasks entrusted to them, which can be aided by introducing calendars and activity plans (daily, weekly). The patients keep a poor track of time, which can be compensated for by wearing a normal wrist watch.

Thanks to the simple solutions proposed by the authors, it is possible for these boys to function independently and effectively for a long period of time. The disease does not have to take away the possibility of attending school or participating in group tasks. This is an extremely important issue for their mental well-being.

The results in AU indicate a significant variation and non-harmonious progression of the disease. It is worth noting an interesting regularity, namely patient No. 4, who achieved the lowest score (6 points), and also obtained low results in the area of cognitive functioning. However, such a pattern was not observed in the case of all the patients. Patient No. 2, had the highest AU score, yet achieved low results in terms of memory functioning. On the other hand, there are also patients with high scores, where this relationship is completely the opposite. This brings to mind the need to expand research in this area and to look for new dependencies and factors affecting those investigated.

Researching this group allows one to optimize the psychological support process provided for a DMD patient , as well as their family. For the goal is to improve their quality of life.

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