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Evaluation of activities of daily living in patients with slowly progressive neuromuscular diseases

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ABSTRACT

Slowly progressive neuromuscular diseases include but are not limited to: facioscapulohumeral muscular dystrophy (FSHD) and limb-girdle muscular dystrophy (LGMD), hereditary motor and sensory neuropathy (HMSN) and spinal muscular atrophy type III (SMA3). The purpose of this study is to present an evaluation of basic and complex activities of daily living in patients suffering from these diseases.

The study was conducted on a group of 58 Polish patients: 25 patients with HMSN, 19 with LGMD and FSHD and 14 with SMA3. The research instrument consisted of two parts: a specially designed questionnaire and Nottingham Extended ADL Index. The survey was voluntary, anonymous and self-administered.

In our study the highest scores on the NEADL scale were achieved by HMSN patients, and the lowest by patients with SMA3. The research revealed statistically significant differences between all the groups in the total number of points achieved on NEADL scale.

The study revealed that for most respondents the most difficult tasks were those in the area of 'mobility'. It is consistent with reports in the literature, which confirm that out of the slowly progressive neuromuscular diseases included in this research, SMA3 is a disease leading to the biggest limitations in performing the activities of everyday life.

1. Introduction

Most neuromuscular diseases have a progressive character and lead to different degrees of impairment in physical performance. Among them we can distinguish a group of slowly progressive neuromuscular diseases which are characterized by usually milder course and therefore allow the patients to maintain independence for a longer time. These include disease entities such as: facioscapulohumeral

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Table 1 – Characteristics of somatic parameters of the studied groups.

Group	N	Age [years]	Height [cm]	Weight [kg]	BMI
HMSN	25	39.28 ± 11.35	171.4 ± 9.64	70.50 ± 18.12	23.87 ± 5.21
FSHD/LGMD	19	36.21 ± 13.21	168.9 ± 12.35	64.74 ± 15.80	22.36 ± 4.41
SMA 3	14	36.64 ± 10.72	163.3 ± 7.83	60.64 ± 17.03	22.63 ± 5.94

muscular dystrophy (FSHD) and limb-girdle muscular dystrophy (LGMD), hereditary motor sensory neuropathy (HMSN) and spinal muscular atrophy type III (SMA3) [1–5].

These diseases display common symptoms during their course, such as: muscle weakness, fatigue, problems with walking and with performing activities of daily living [6]. For most patients maintaining independence in the basic activities of daily living (ADL), such as moving around or self-feeding, reflects the desire to remain autonomous. There are a lot of scales used to evaluate ADL [7]. Numerous studies in patients with neuromuscular diseases evaluated ADL on Barthel scale (Barthel Index-BI) [1,8–11]. However, it was noted that Barthel scale may not be adequate to evaluate ADL in patients with slowly progressive neuromuscular diseases. It is too general and might not be accurate enough for the assessment of patients with milder forms of neuromuscular diseases [1,8]. Moreover, there is often a need to supplement the evaluation of basic ADL with the assessment of complex activities of daily living. For this purpose instrumental scales were created, also called Extended Activities of Daily Living (EADL) scales. These scales, used in the evaluation a patient's condition, are a link between functional scales and scales used for the measurement of quality of life [7,12].

Stubgen observes that it seems improper to believe that the scales should be specific for a particular disease entity. Probably more important is to use scales and tests that will be targeted to the nature of the problems that occur in patients [11]. Nottingham Extended ADL Index (NEADL) is a scale which was created and validated for the assessment of people with stroke [7]. It evaluates both basic and complex activities of daily living and can be an alternative for the evaluation of patients with slowly progressive neuromuscular diseases. Supplemented with a specially designed questionnaire it seems to be a good instrument for evaluation of this group of patients.

The purpose of this study is to present an evaluation of basic and complex activities of daily living in patients with slowly progressive neuromuscular disease.

2. Material and methods

The research was based on an anonymous self-administered online survey in which patients participated voluntarily. It consisted of two parts. The first was a specially designed questionnaire containing 35 closed questions. The second part was based on the Nottingham Extended ADL Index (NEADL).

The study was conducted in Polish patients with neuromuscular diseases. Survey questionnaires were distributed to people with neuromuscular diseases in electronic form and 96 completed questionnaires were sent back. The patients were divided into three main groups according to the type of disease. Finally 58 questionnaires were chosen for the analysis – 25 from patients with HMSN, 19 from patients with LGMD and FSHD and 14 from patients with SMA3.

Table 1 presents the characteristics of the groups in terms of age, height, body weight and BMI.

NEADL scale evaluates independence in four areas of everyday life: *mobility*, *kitchen*, *domestic activities*, and *leisure activities*. In these areas there are 6, 5, 4, and 6 points respectively. In total, the scale consists of 21 questions [13]. Full NEADL scale is presented in Table 2.

The authors have modified the scoring scale. Each point is reported on 1–4 score scale, where 1 means *I don't do it at all*, 2 – *I do it with help*, 3 – *I do it on my own with difficulty*, 4 – *I do it on my own easily*. The full scale value ranges from 21 to 84 points.

2.1. Statistical methods

The statistical analysis was performed using computer program Statistica (v. 12). Mean values and standard deviations of somatic data were calculated and the percent difference comparison was applied. For quantitative variables arithmetic mean and standard deviations (SD) were calculated. Shapiro-Wolf normality test was used to check whether the distribution of the surveyed characteristics is consistent with normal distribution. Analysis of variance (ANOVA) and post

Table 2 – Nottingham Extended Activities of Daily Living Index.

Mobility questions – Do you:
Walk around outside?
Climb stairs?
Get in and out of the car?
Walk over uneven ground?
Cross roads?
Travel on public transport?

In the kitchen – Do you:
Manage to feed yourself?
Manage to make yourself a hot drink?
Take hot drinks from one room to another?
Do the washing up?
Make yourself a hot snack?

Domestic tasks – Do you:
Manage your own money when you are out?
Wash small items of clothing?
Do your own shopping?
Do a full clothes wash?

Leisure activities - Do you:
Read newspapers or books?
Use the telephone?
Write letters?
Go out socially?
Manage your own garden?
Drive a car?

Table 3 – The detailed results obtained on NEADL scale.

NEADL	Max points	CMT	FSHD/LGMD	SMA 3	*p* values	Effect size *d*-Cohen
Mobility	24	20.88 ± 2.5	14.79 ± 5.6	9.79 ± 3.8	1 vs 2 0.00000	1 vs. 2 1.403
					1 vs 3 0.00000	1 vs 3 3.414
					2 vs 3 0.0009	2 vs 3 1.047
Kitchen	20	19.24 ± 1.2	15.84 ± 4.7	13.07 ± 5.3	1 vs. 2 0.005	1 vs. 2 0.985
					1 vs 3 0.00001	1 vs 3 1.589
					2 vs 3 0.04	2 vs 3 0.548
Home	16	14.52 ± 2.6	11.68 ± 3.9	9.79 ± 2.8	1 vs. 2 0.004	1 vs. 2 0.86
					1 vs 3 0.00003	1 vs 3 1.744
					2 vs 3 0.09	2 vs 3 0.555
Leisure Activities	24	19.28 ± 3.7	17.58 ± 3.9	16.43 ± 2.6	1 vs. 2 0.12	1 vs 2 0.447
					1 vs 3 0.01	1 vs 3 0.89
					2 vs 3 0.3	2 vs 3 0.345
Total on NEADL scale	84	73.92 ± 8.74	59.89 ± 13.98	49.07 ± 11.93	1 vs. 2 0.0001	1 vs. 2 1.203
					1 vs 3 0.00000	1 vs 3 2.376
					2 vs 3 0.009	2 vs 3 0.832

hoc Tukey's honest significant difference test (HSD) were used to check the significance of the differences in the results. Effect size (ES) was also measured to confirm the size of the statistically significant difference. The standard level of statistical significance $p < 0.05$ was adopted.

3. Results

72% of the patients with HMSN are professionally active, 12% declared that they received a disability pension due to illness, and 16% declared that they were not professionally active. In the group of FSHD and LGMD patients 37% are professionally active, 53% receive a disability pension, and 11% are professionally inactive. Half of the SMA3 patients claimed they received a disability pension due to illness, 43% said they were professionally active and the remaining 7% stated that they were inactive.

When asked about medical aids, some patients with SMA3 and dystrophy declared they used of a wheelchair – 71% and 21% respectively. In HMSN group 12% of patients used one crutch, and as many as 68% declared they needed an orthopedic appliance for their lower limb.

On NEADL scale the highest score was obtained by patients with HMSN – 73.92 ± 8.74, the lowest by people with SMA3 – 49.07 ± 11.93. Statistically significant differences ($p < 0.05$) were noted between the total scores obtained on NEADL scale by all the groups. Under the assumption of the effect size scale developed by Cohen [14] (Cohen, 1988), the difference between all the groups is large (ES > 0.8). The biggest difference is between HMSN group and SMA3 (ES-2, 376). The detailed results obtained on NEADL scale can be found in Table 3.

We can observe similar distribution of the subscores on NEADL scale for *mobility* and *kitchen* – in both areas the highest score was obtained by people with HMSN and it was 20.88 ± 2.5 and 19.24 ± 1.2 respectively. The lowest score was obtained by SMA3 patients who received 9.79 ± 3.8 for *mobility* and 13.07 ± 5.3 for *kitchen*. Statistically significant differences were found between all groups. ES among all groups is big (ES > 0.8), the biggest between patients with HMSN and SMA3 (ES-3, 414).

In the area of *mobility* patients with HMSN as well as those with muscular dystrophies received the lowest number of points for the question '*Do you walk over uneven ground?*' and it was 3.12 ± 0.78 and 2.16 ± 1.17 respectively (Fig. 1). Statistically significant differences were found between the two groups. ES between the patients with HMSN and those with muscular dystrophies was 0.43, which means it is interpreted as average.

SMA3 patients got the lowest score for the question '*Do you climb stairs?*' – 1.29 ± 0.61 points - and statistically significant differences were found between all groups (Fig. 2). The biggest difference was between HMSN and SMA3 groups (ES-0.88). In one of the closed questions of the survey 79% patients with SMA3 declared that climbing and descending the stairs was impossible for them. The same answer was given by 26% of patients with muscular dystrophies.

In *kitchen* area for all patients the easiest activity was '*managing to feed myself*'. The lowest score for this activity was obtained by patients with dystrophies, who received 3.58 ± 0.69 for this question and statistically significant difference was found between this group and the group with HMSN. ES was 0.4 and is interpreted as average.

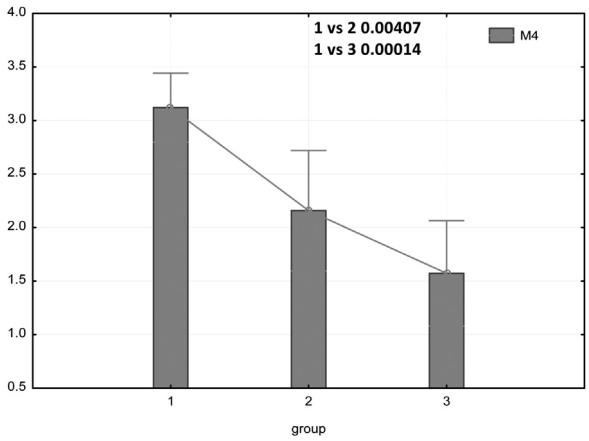

Fig. 1 – Interaction plot for the question 'Do you walk over uneven ground?' in the three groups. Group 1 – HMSN, Group 2 – FSHD/LGMD, Group 3 – SMA3.

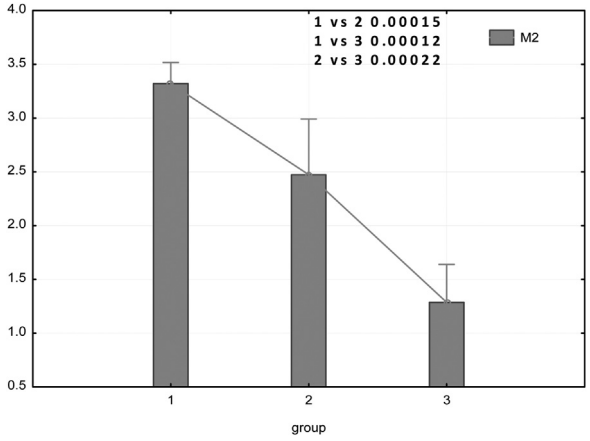

Fig. 2 – Interaction plot for the question 'Do you climb stairs?' in three groups. Group 1 – HMSN, Group 2 – FSHD/LGMD, Group 3 – SMA3.

When asked about access to rehabilitation, most patients replied that it is not sufficient. Such answer was given by 100% of patients with SMA3, 84% patients with muscular dystrophies and 72% with neuropathy.

4. Discussion

The purpose of this study is to present an evaluation of basic and complex activities of daily living in patients with slowly progressive neuromuscular diseases.

When reviewing literature, the authors found no study in which EADL in patients with neuromuscular diseases was evaluated using Nottingham Extended ADL Index (NEADL). It was also revealed that EADL measurement is usually only one of many endpoints, following the evaluation of muscle strength or functional tests [1,4,8,9,11,15].

Functional assessment of patients focused on performing activities of daily living is especially important because the loss of function does not always go together with progressive weakening of muscle strength. Dunaway et al. describe a case of a patient with SMA3, where they demonstrate that it is a slowly progressive disease with a progressive loss of function with no measurable decrease in muscle strength [5]. Iannaccone et al. observed and assessed 73 children and adults with SMA type 2 and 3 over the period from 2 to 6 years. They noted that 10 of them experienced a loss of functional ability with the absence of deterioration of muscle strength. This means that the loss of function has to be the result of other factors, or that the functional assessment is more sensitive than the measurement of muscle strength for detecting small changes [5,16].

In the present study, the highest score on NEADL scale was obtained by patients with HMSN. Hereditary motor and sensory neuropathy, of which the most common form is Charcot–Marie–Tooth disease type 1A (CMT1A), is a chronic, mild and slowly progressive polyneuropathy, which is characterized by symmetric weakness and wasting of distal muscles, mainly in the lower limbs [17].

Our results are consistent with the research carried out by Menotti et al., in which patients with CMT1A eligible to take part in the assessment obtained 96.3 ± 3.8 points out of 100 on Barthel scale. In this study the authors observed smaller number and lower intensity in performing some ADL in patients with CMT1A compared to healthy individuals. It was noticed that they climbed fewer stairs – the differences were visible when climbing up and when descending the stairs. Moreover, patients with CMT1A reported fewer position changes – moving from sitting to standing position and the other way round. The authors speculate that people with neuropathy avoid the most demanding tasks, which require a lot of muscle work, both concentric and eccentric, in the lower limbs [10]. This is consistent with our results, where 48% of patients with HMSN confirmed that they have a problem with standing up from a squatting position and must support themselves against the items located near them. When asked about the stairs, 36% declared that both climbing and descending the stairs is difficult for them.

In the area of *mobility*, the most difficult task for people with HMSN was 'walking over uneven ground'. Perhaps it derives from the fact that one of the characteristic symptoms of hereditary motor and sensory neuropathy, in addition to weakening of distal muscles in the lower limbs, is proprioceptive dysfunction, which is noticeable even at the beginning stage of the disease [17]. Proprioceptors are one of sensory receptors which are involved in the control of balance and their dysfunction can lead to an impaired postural control [18]. It may be reflected in the problems patients experience with reduced mobility on uneven surfaces. In the future it would be useful to extend the study by adding a questionnaire on risk of falls in these patients.

Also for patients with muscular dystrophies the hardest thing to do in the area of *mobility* was '*walking over uneven ground*', but in comparison to HMSN group they obtained much lower score and the difference was statistically significant. Such difference is also noticeable in the question about performing a squat, where 37% patients with muscular dystrophies declared that a squat was impossible for them to perform, while in HMSN group it was not possible for only 4% of respondents. Similarly, climbing and descending stairs were perceived as impossible by 26% of patients with dystrophies.

FSHD is characterized by a specific distribution pattern of muscle weakness beginning from facial muscles, muscles which stabilize the scapula and arm muscles. Then weakening may involve peroneal muscles and pelvic girdle muscles. Quite often abdominal muscles can also be weakened [19]. LGMD is typically characterized by a progressive weakness of pelvic girdle and shoulder girdle muscles as well as proximal limb muscles [1,20].

In a study conducted by Lue et al. FSHD patients obtained 97.8 ± 4.7 out of 100 points on Barthel scale, while those with LGMD got only 85.6 ± 20.2 points. The difference between these patients was also detected on Vignos scale which evaluates functional activity of the lower limbs. 50% of patients with FSHD were on grade 1 of Vignos scale, which means that '*they walk and climb stairs without assistance*', but the percentage of patients with LGMD on this grade was much lower – only 14.8%. Much more often people with a limb-girdle

muscular dystrophy were placed on grade 3 of the scale – 29.6% could '*walk and climb stairs slowly with aid of railing*', and as many as 18.5% were on grade 9, which means moving around in a wheelchair. For comparison the lowest result for FSHD patients was grade 5 and it actually referred to only one person [9]. In another study carried out by Lue et al., 80% of patients with FSHD were classified on Barthel scale as '*completely independent*' and 15% as '*partially independent*' and the hardest thing for them was climbing and descending stairs [8]. And according to a study of Kilmer et al., 48% of people with FSHD were able to climb and descend stairs without assistance, 38% were placed on grades 2–3 of Vignos scale, but as many as 14% used a wheelchair [21].

For patients with muscular dystrophies as well as for those with HMSN the easiest thing to do in the area of *mobility* was '*getting in and out of the car*'. This is not surprising in the case of people with neuropathy, because weakness of the muscles located above the knee joints, which are heavily engaged in this activity, may not occur until the later stage of the disease [22]. Patients with muscular dystrophies also declared that it was the easiest activity in the *mobility* area, but their score was lower than the score obtained by HMSN patients and the difference was statistically significant. This is probably related to the fact that proximal muscles of the lower limbs and pelvic girdle muscles are affected at an early stage of this disease. The manner in which this activity was performed should also been taken into account. They might have, for example, supported themselves against the elements of the car.

The lowest result on NEADL scale was obtained by patients with SMA3, and the biggest difference was observed between them and the people with HMSN (EF-2, 376).

In a study conducted by Chung et al. SMA classification was adopted from Zerres and the patients with SMA3 were divided into two types. Type IIIa for those who got sick before the age of 3, and type IIIb when they got sick between the age of 3 and 30. The authors used on WeeFIM scale to evaluate the performance. Patients with SMAIIIa obtained 16.7 ± 5.6 out of 35 points in the area of *mobility* and people with SMAIIIb got 18.2 ± 12.1. The conclusion of the authors was that in performing most activities in the area of *mobility* patients with SMA needed help [4].

In our study people with SMA3 in the area of *mobility* obtained 9.79 ± 3.8 points out of 24. For them the most difficult activity in this area was '*climbing stairs*', which is consistent with the results of the study carried out by Chung et al., where it was also observed that the biggest obstacle for the autonomy of the patients was struggling with the stairs [4] (Chung).

Statistically patients with SMA3 begin to walk on their own at the age of 15 months and lose this ability at the age of 12 years. Patients for whom the biggest achievement was walking with assistance, usually lose this ability at the age of 7 [5]. Dunaway et al. presented the results of a five-year observation of a patient with SMA3. Just as it is described in the literature, the patient lost his motor functions over the course of time, however he was able to move with assistance for about 2 years longer than expected – until he was 9 years old [5]. In our study 71% of patients with SMA3 declared that they used a wheelchair. Russman et al. reported that in the moment SMA patients are no longer able to walk, they also lose the ability of raising their arms above the head [23]. When we asked a question: '*is raising your arms above the head difficult?*' 71% of patients with SMA3 gave affirmative answer.

For all patients the easiest activity in the '*kitchen*' area was '*managing to feed myself*'. The lowest score for this activity was obtained by patients with dystrophies. Perhaps this derives from the fact that in this group some of the patients suffer from FSHD and one of the first symptoms of this particular disease is weakness of the facial muscles, which can lead to difficulty in eating.

This study was conducted in patients with different slowly progressive neuromuscular diseases. Although each of these disease entities has a different genetic background and a different clinical course, they have several common characteristics. These include muscle atrophy, paresis and reduced muscle tension. Each disease has a distinct distribution pattern of muscle weakness and the order in which individual muscle groups are affected.

For most patients who took part in the survey the most difficult tasks were those in the area of *mobility*. The scores obtained here were for most patients much lower than the scores in other areas. What is interesting to note is that patients suffering from different diseases reported the same limitations in activities of daily living. This similarity is particularly reflected in the responses given by patients with HMSN and muscular dystrophies, for whom the easiest and the most difficult tasks in the area of *mobility* were the same. Of course patients with muscular dystrophies obtained a significantly lower score for these tasks, but the limited activity remained the same. The primary goal of physiotherapy is to adjust therapeutic procedures to the problems reported by patients, therefore an assessment of the limitations that patients experience in social life and in performing everyday activities is a valuable supplement to physiotherapeutical examination.

It seems that it might be particularly interesting to conduct a re-examination of these patients in order to evaluate and compare the progress of each disease.

Our study is consistent with reports in the literature, which conclude that out of the slowly progressive neuromuscular diseases included in this research, SMA3 is a disease leading to the biggest limitations in performing activities of everyday life. So far, the only available treatment for most patients with neuromuscular diseases is physiotherapy. When asked: '*Do you think that access to rehabilitation is sufficient?*' all patients with SMA3 gave negative response. The same answer was given by 84% of people with muscular dystrophies and 72% of patients with hereditary neuropathy. It is disturbing because physiotherapy is one of the basic pillars of medical care for people with neuromuscular diseases. Because of the chronic and progressive nature of these diseases patients should have access to systematic and repetitive rehabilitation. In the process of rehabilitation should be engaged a dedicated team, where, apart from a physiotherapist, the patient would have access to such specialists as an occupational therapist, a psychologist and a speech therapist.

Conflict of interest

None declared.

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