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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 muscular dystrophy (FSHD) and limb-girdle muscular dystrophy

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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

(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?

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