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Further evidence for the reliability and validity of the Fatigue and Daytime Sleepiness Scale

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ABSTRACT

Background: Myotonic dystrophy type 1 (DM1) is an inherited neuromuscular disease causing, among other symptoms, fatigue and excessive daytime sleepiness, which are frequently undifferentiated by patients and/or clinicians. The Fatigue and Daytime Sleepiness Scale (FDSS) has been devised to measure these two overlapping symptoms as a single clinical entity.
Objective: To further examine the reliability and the construct validity of the FDSS in patients with DM1.
Methods: The scale was administered to 48 DM1 patients on two occasions at a 2 week-interval. Intra-rater reliability and internal consistency were established by calculating the intraclass correlation coefficient (ICC) and Cronbach's alpha, respectively. Construct validity was assessed by using the known-group method. More precisely, the mean FDSS score of patients with and without subjective complaints of fatigue and/or sleepiness was compared.
Results: The FDSS showed good intra-rater reliability (ICC = 0.83) and acceptable internal consistency (Cronbach's alpha = 0.6). Also, the FDSS was able to distinguish between patients who had fatigue and sleepiness complaints from those who did not (mean FDSS score of 10.6 vs 8.0, $p = 0.01$), suggesting good construct validity.
Conclusion: Overall, the present study supports the continued use of the FDSS as a reliable and valid instrument to measure fatigue and daytime sleepiness in patients with DM1 for either clinical or research purposes.

1. Introduction

Myotonic dystrophy type 1 (DM1), an autosomal dominant disorder, is the most common adult-onset form of muscular dystrophy [1]. DM1 results from an unstable CTG repeat expansion in the 3′ untranslated region of the chromosome 19q13.3 [2]. DM1 is not only a muscle disease but a multisystemic disorder affecting the central nervous, ocular, respiratory, cardiovascular, digestive, endocrine and reproductive systems [3,4].

Fatigue and excessive daytime sleepiness (EDS) are common and significant clinical features of DM1 which occur in 62.5% and 30–39% of patients, respectively [5–7]. These symptoms are associated with reduced social participation and quality of life [8–10]. Fatigue is characterized by a subjective lack of physical and/or mental energy and is more common in DM1 than in other neuromuscular disorders [11]. On the other hand, EDS in DM1 tends to occur when attention is not being sustained, rather than during activity. Also, EDS cannot be entirely explained as a consequence of sleep-disordered breathing, chronic hypercapnia, or depression [12–15]. However, fatigue and EDS have common, overlapping features and they have been shown to be associated in DM1 patients [7,11].

There are currently some promising approaches for clinical trials in DM1 (e.g. trial on the IONIS-DMPKRx drug, Clinical Trial ID NCT02312011 [16]) but one essential step to be completed consists in identifying the best outcome measures that can capture any change occurring during such targeted therapies. In this regard, the paucity of data concerning the reliability and validity of outcome measures in the DM1 population is a barrier to these future clinical trials. Launched in 2011, the *Outcome Measures in Myotonic Dystrophy type 1* (OMMYD) international initiative aims to address this problem [17,18]. As part of the OMMYD initiative [18], the Special Interest Group addressing the symptoms of sleepiness, fatigue, and apathy identified the Fatigue and Daytime Sleepiness Scale (FDSS) for DM1 [19] as an interesting tool to take in consideration for natural history study and clinical trial but some metrological properties still need to be documented. The FDSS was devised specifically for DM1 patients [19]. The questionnaire

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contains 12 items taken from the Epworth Sleepiness Scale [20], the Daytime Sleepiness Scale [5] and the Fatigue Severity Scale [21]. It provides interval measures of fatigue and EDS on a single continuum addressing the same health construct. Each item is rated on a three-point scale where 0 means "Seldom or never", 1 means "Sometimes", and 2 means "Almost always". The raw sum scores varying from 0 to 24 (24 indicating more symptoms of fatigue and daytime sleepiness) can be transformed into interval measures that can be used for parametric statistical analyses. In its validation study, the FDSS fulfilled all Rasch model expectations. The hierarchy of items did not vary across DM1 patients according to age, gender, disease type, level of education, and use of psychostimulants, nor between patients of different countries (Canada and the Netherlands). The scale was shown to measure a single construct which combines aspects of sleep propensity and behavioural consequences of fatigue, suggesting good factorial validity. Also, the internal consistency was shown to be acceptable (person separation index = 0.80) and a trend was observed for patients with more severe muscular impairment to have higher levels of fatigue and sleepiness [19]. Although it was designed specifically for the DM1 population, its reliability and construct validity must be further documented.

The purpose of the present study was to gain further evidence of the psychometric properties of the FDSS (intra-rater reliability, internal consistency, the precision (standard error of measurement (SEM)), the minimum detectable change (MDC) and construct validity) in patients affected by DM1.

2. Method

A total of 425 patients with DM1 are currently followed at the Saguenay Neuromuscular clinic (Jonquière, Québec, Canada). Among these, 237 meet the following inclusion criteria: 1) being aged 18 years or older, 2) having a molecular confirmation of the DM1 diagnosis, and 3) having the late onset or the adult phenotype of the disease. Eligible patients were selected either consecutively during their annual medical check-up or by phone. Of the 58 patients who were invited to participate in the study, 48 have completed the study, 8 patients declined to take part in the study, and 2 patients withdrew from the study. All participants were able to provide an informed consent. A sociodemographic questionnaire was administered to collect information on age, sex, educational status, and level of mobility/autonomy. CTG repeat number was taken from patients' medical record. In order to document fatigue and daytime sleepiness complaints, patients were also asked: "Do you tend to feel tired?" and "Do you tend to be sleepy during the day?". The FDSS was administered on two occasions (T1, T2) at an interval of 2 weeks by the same trained PhD student in neuropsychology. The study was approved by the Ethics Review Board of the *Centre Intégré Universitaire de Santé et Services Sociaux du Saguenay Lac-St-Jean* (Québec, Canada) and written informed consent was obtained from all participants.

Data are expressed as mean ± standard deviation (SD) for continuous variables and as frequency and percentage for categorical variables. Statistical analyses were planned in accordance with COSMIN guidelines for methodological quality in studying the measurement properties of outcome measures [22]. Internal consistency was assessed with Cronbach's alpha reliability coefficients from the first questionnaire completion. The desired range for the alpha value is between 0.70 and 0.90 to be considered as good [23]. The intra-rater reliability was determined using the intraclass correlation coefficient (ICC (2,1)) to compare results at T1 and T2 [24]. An ICC above 0.75 is considered as indicating good reliability [25]. The SEM, which is defined as the standard deviation of the measurement errors, was calculated with this formula: $SEM = SD_{baseline} \times \sqrt{(1 - ICC)}$. The minimal detectable change (MDC), which is defined as the amount of change in a variable that must be achieved to reflect a true difference [25], was calculated with the formula: $MDC_{95} = 1.96 \times \sqrt{2} \times SEM$. The construct validity was assessed by using the known-group method [23]. More precisely, the mean score of the FDSS was compared between patients with and without complaints of fatigue and/or daytime sleepiness. The a priori hypotheses were that 1) the FDSS mean score should be higher in DM1 patient who reported experiencing symptoms of daytime sleepiness and fatigue and 2) the FDSS mean score should be similar across gender and CTG repeat class. Data were analysed using IBM SPSS Statistics for Windows, Version 24.0 (Armonk, NY: IBM Corp).

3. Results

Sociodemographic and clinical characteristics of participants are shown in Table 1. Participants' age ranged from 20 to 64 years. The large majority of participants have completed high school (98.0%) and reported not needing help to walk inside (85.0%) or for home maintenance (88.0%). Mean CTG repeat number was 715 (SD 480).

In addition, no significant difference in FDSS scores was found between men and women (mean (SD) = 8.4 (3.6) and 10.2 (3.4), $p = 0.10$) and between CTG repeat groups (mean (SD) = 10.5 (3.9) (50–200 CTG), 9.0 (3.5) (201–1000 CTG), and 9.5 (3.8) (>1000 CTG), $p = 0.70$). Yet, a moderate positive correlation was found between the mean FDSS score and participants' age ($r = 0.33$, $p = 0.02$).

The FDSS showed moderate/acceptable internal consistency (Cronbach's alpha = 0.60) and a good intra-rater reliability with an ICC of 0.83 (95% CI = 0.70–0.90). Based on this latter coefficient and the standard deviation of the baseline mean, the SEM and the MDC were 1.6 and 4.3, respectively.

Table 2 presents the comparison of mean FDSS score with participants' complaints of fatigue and daytime sleepiness. The mean FDSS score was significantly higher for those who only reported fatigue complaints ($p < 0.000$) and a trend was seen for higher FDSS scores in those who only reported complaints of daytime sleepiness ($p = 0.05$). More importantly, participants who reported both fatigue and daytime sleepiness complaints had higher FDSS scores than those without complaints ($p = 0.01$).

4. Discussion

Consistent with previous studies [26–28], data show that fatigue and daytime sleepiness complaints are prominent in patients affected by DM1, with 86% and 54% of participants reporting these symptoms, respectively. In agreement with our previous findings [7], virtually all patients with daytime sleepiness complaints also reported fatigue complaints (96.3%) while a much lesser proportion of patients with fatigue complaints also reported daytime sleepiness complaints (61.0%). In both patients with sleep disorders and neurological conditions, it is

Table 1
Sociodemographic and clinical characteristics of the 48 participants.

Age	Mean (SD)	44.2 (13.2)
	Range	20–64
Sex, n (%)	Men	22 (45.8)
	Women	26 (54.2)
CTG repeats, n (%)	50–200	6 (12.5)
	201–1000	28 (58.3)
	>1001	14 (29.2)
Educational level, n (%)	Elementary	1 (2.1)
	High school	31 (64.6)
	Vocational training	10 (20.8)
	College	5 (10.4)
	University	1 (2.1)
Indoor mobility, n (%)	No walking aid	41 (85.4)
	Cane/walker	1 (2.1)
	Wheelchair	6 (12.5)
Independent living, n (%)	Independent	42 (87.5)
	Partial assistance	5 (10.4)
	Complete assistance	1 (2.1)
FDSS score, mean (SD)	Baseline	9.4 (3.6)
	Follow-up	8.7 (4.0)