Assessment of health-related quality of life, depression, and anxiety in slowly and rapidly progressive neuromuscular disorders

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ABSTRACT

الأهداف: مقارنة جودة الحياة المتعلقة بالصحة (HRQoL) والخصائص النفسية في الاعتلال العصبي العضلي السريع (RPNMD) والبطيء التطور (SPNMD) في المرضى البالغين المعتمدين على أنفسهم لتحديد الاحتياجات الفردية في مجموعتين متقدمتين منفصلتين.

الطريقة: اشتملت الدراسة على 39 مريض مصاب باعتلال عصبي عضلي بطيء التطور (SPNMD) و46 مريض مصاب باعتلال عصبي عضلي سريع التطور (RPNMD). استخدمنا مقياس الاعتماد الوظيفي (FIM) لتقييم الحالة الوظيفية. ولتقييم الاكتئاب، والقلق، وجودة الحياة المتعلقة بالصحة (HRQoL). طلب من المرضى تعبئة مقياس استبيان بك للاكتئاب، ومقياس القلق، وملف نوتنغهام الصحي (NHP). أجريت الدراسة في قسم العلاج الطبيعي والتأهيل، جامعة هيستوب، كلية العلوم الصحية، أنقرة، تركيا خلال الفترة من أغسطس وديسمبر 2009م.

النتائج: لم يختلف المقياس الوظيفي (FIM) بين المجموعتين. كانت الطاقة فقط عالية (سيئة) بشكل مهم بين أبعاد ملف نوتنغهام الصحي في المرضى المصابين باعتلال عصبي عضلي سريع التطور (RPNMD). لم تختلف الأصناف الفرعية الأخرى بين مجموعتي المرضى. كانت مجموعة المرضى المصابة باعتلال عصبي عضلي بطيء التطور (SPNMD) أكثر اكتئاباً من المصابة باعتلال عصبي عضلي سريع التطور (RPNMD). كان مقياس القلق، ومعدل الحالة أعلى في المرضى المصابين باعتلال عصبي عضلي بطيء التطور (SPNMD).

خاتمة: لا يوجد صلة بين أعراض القلق، والاكتئاب مع التصلب الجانبي (ALS). يجب أن ينبه تشخيص الاعتلال العصبي العضلي البطيء التطور (SPNMD) الأطباء بالتحسن المماثل للتصلب الجانبي (ALS) ليكونوا على علم لاحتمال حدوث الاكتئاب، أو القلق، والاهتمام بالمرضى خصوصاً المصابين بالأمراض المزمنة من خلال التطور البطيء للمرض.

Objectives: To compare health-related quality of life (HRQoL) and psychosocial features in rapidly progressive neuromuscular disorders (RPNMD) and slowly progressive neuromuscular disorders (SPNMD) in adult ambulatory patients, to determine individual needs in 2 separate progression groups.

Methods: Thirty-nine SPNMD patients and 46 RPNMD patients were recruited. The functional independence measurement (FIM) was employed to evaluate the functional status. For the assessment of depression, anxiety, and HRQoL, patients were requested to fill out a Beck Depression Inventory, State-Trait Anxiety Inventory, and the Nottingham Health Profile (NHP). This study was performed at the Department of Physical Therapy and Rehabilitation, Hacettepe University Faculty of Health Science, Ankara, Turkey between August and December 2009.

Results: The FIM total score did not differ between the 2 groups. Only energy was significantly high (worse) among the dimensions of NHP in RPNMD patients. None of the other sub-items differed between the 2 patient groups. The SPNMD patients were more depressed than the RPNMD patients. The mean state and trait anxiety scores were significantly higher in SPNMD patients as well.

Conclusions: Significant depressive or anxious symptomatology is not associated with amyotrophic lateral sclerosis (ALS). The diagnosis of SPNMD should alert physicians in an equivalent promptness to ALS and possible depression or anxiety, and concerns of patients regarding the chronic, though slowly progressive course of the disease.

Neurosciences 2010; Vol. 15 (3): 177-183

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Received 21st December 2009. Accepted 9th March 2010.

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Neuromuscular diseases (NMD) are all characterized by progressive muscular impairment, but each of them has different clinical features. They may affect people of all ages. Most NMD such as, Becker muscular dystrophy (BMD), fascioscapulohumeral muscular dystrophy (FSHD), limb-girdle muscular dystrophy (LGMD), and myotonic dystrophy (MD) have slowly progressive reduction in muscular function with mildly reduced life expectancy. On the other hand, some of these disorders are characterized by a rapidly progressive course, such as amyotrophic lateral sclerosis (ALS), spinal muscular atrophies, and Duchene muscular dystrophy (DMD), in which muscle impairment worsens over months that results in death within a few years. Despite significant advances in pinning down the biological basis for NMD, few effective treatments are available, and no cure exists.[1,2] Common consequences of these diseases include fatigue, problems with locomotion, and loss of functionality in activities of daily living. Progressive muscular impairment may lead to loss of ambulation, being wheelchair-bound, and swallowing difficulties. Respiratory failure is the most common cause of mortality. Not surprisingly, psychological status and quality of life (QOL) may worsen in some individuals with NMD through their disease process.[3] Quality of life is defined as an individual's perception of their position in life in the context of the culture and value systems in which they live, and in relation to their goals, expectations, standards, and concerns.[4] Health related quality of life (HRQoL) can be distinguished from QOL. The World Health Organization defines HRQoL as a state of complete physical, mental, and social well-being, and not merely the absence of disease or infirmity; HRQoL is defined as QOL, in which a dimension of personal judgment over one's health and disease is added. It has been suggested that measures of HRQoL can be used as primary endpoints in clinical trials in ALS.[5] Psychological features, interpersonal relationships, social and economic state may affect HRQoL significantly, as well as physical and functional problems.[6] It is obvious that there is a need for more knowledge of the impact of the disease on psychopathology and QOL in order to develop evidence-based rehabilitation for adults with NMD. Terminal stages of NMD have been comprehensively evaluated with respect to HRQoL and psychopathology.[7] However, ambulatory patients with mild deficit have not been the focus of QOL research in NMD, and relevant studies are limited in number.[8] Populations in previous studies were heterogeneous with respect to age, rate of disease progression and functional severity. It is crucial to determine the goals in rehabilitation of these disorders, in which cure has not been achieved so far. To create the optimum conditions for rehabilitation, the requirements and emotional deficits of patients must be recognized. Scarce literature does not provide data clarifying how the rate of progression affects QOL and psychological status. The objectives of this study were to compare the HRQoL and psychological features of adult ambulatory patients, with rapidly progressive neuromuscular disorders (RPNMD) and slowly progressive neuromuscular disorders (SPNMD), and to determine individual needs in 2 separate progression groups.

Methods. Eighty-five consecutive patients with NMD who meet the inclusion criteria were recruited in this study. The SPNMD group (n=39) consisted of 21 LGMD, 5 MD, 3 FSHD, 2 BMD, and 8 other myopathies. The RPNMD group included 46 ALS patients. They were all outpatients who were referred by the Neurology Department and admitted to the Neurological Rehabilitation Outpatient Unit of the Department of Physical Therapy and Rehabilitation, Hacettepe University Faculty of Health Science, Ankara, Turkey between August and December 2009. The Hacettepe University local ethical committee approved the study. All patients provided written informed consent. Questionnaires and functional evaluations were applied by the same researcher for all subjects during the study. The self-report instruments that were all completed in the unit took approximately 30 minutes. Independent walking ability in the community, with a need of moderate assistance while performing activities of daily living were sought for inclusion. Patients were all living at home, but not in an institution. Individuals with other medically unstable major physical disease, cognitive impairment (with scores lower than 24 in Mini Mental Test), and without sphincter control were excluded. Patients were not ever diagnosed and treated, or had a family history of a major psychiatric disorder.

Measures. Basic demographic information was collected including age, gender, education, duration of illness of patients, and information of patients on the nature of the neurological illness. The functional independence measure (FIM) was employed to evaluate the functional status. The instrument is an 18-item, 6-level scale that scores the care needs from 18 (complete dependence) to 126 (full independence). Six subscales are formed, including self-care, sphincter control, mobility, locomotion, communication, and social communication. The scoring scale includes 2 independent levels, and 5 helper levels. The need for supervision or assistance of a patient is rated. The highest level (7) indicates total independence and the lowest level (1) indicates total need for the assistance of 2 helpers to perform the activity. Normal range of the FIM score is between 18 and 126 (18-36: completely

dependent; 37-90: needed supervision and assistance while performing activities; 90-126: completely independent). It was adapted in Turkish language, and found to be reliable and valid in a spinal cord injury sample.[9] For the assessment of psychological status, patients were requested to fill out relevant self-report scales: Beck depression inventory (BDI) for depression, state-trait anxiety inventory (STAI) for anxiety, and the Nottingham health profile (NHP) for HRQOL. The BDI is a self-rating instrument developed by Beck et al[10,11] and psychometric properties were evaluated rigorously. It is used to identify potential cases of depressive illness, and to measure the severity of 21 depressive symptoms on a 4-point scale ranging from 0-3. The total maximum score is 63. The optimum cut-off point was found to be 16/17 in the reliability and validity study of the Turkish version in accordance with the original study.[12] The STAI is a self-report assessment scale, which provides a reliable measure of anxiety. It was adopted in the Turkish language, in which it was found to be reliable and valid.[13] It includes 2 subscales which measures state and trait anxiety. While state anxiety subscale explores how the subjects feel at that moment, the latter subscale explores how they feel generally. Each subscale consists of 20 items. Response to every item is assigned a score ranging from 1-4, indicating the severity of the symptom. Both subscales are scored from 20-80, with higher scores indicating higher anxiety level. Usually, a cut-off score of <40 and >40 is used in prior studies.[13] The NHP is a self-administered questionnaire, which assesses the subjective perception of the physical, emotional, and social aspects of health. The NHP evaluates 6 health dimensions: energy, pain, physical mobility, emotional reactions, sleep, and social isolation. The profile has dichotomized questions and the subjects may answer 'yes' or 'no.' Higher scores on the NHP represent worse quality of life. The NHP has been tested in both RPNMD and SPNMD patients, and it has been reported that it shows good reliability.[14] The Turkish version of these scales have been developed and thoroughly tested for reliability and validity in Turkey.[15]

Statistical analysis was performed using the Statistical Package for Social Sciences version 15 (SPSS Inc, Chicago, IL, USA). Data are expressed as mean (standard deviation), or frequency (percentage). Most of the variables including demographics, functional, and psychological measures, and QOL scores (including the 6 dimensions of NHP evaluated separately) of patients (except age, STAI, trait anxiety, state anxiety of patients) were not normally distributed. The Mann-Whitney U test, or student's t-test was used to assess intergroup differences. The association of gender with trait and state anxiety levels was analyzed using student's t-test. The association of gender with BDI and NHP scores was analyzed using the Mann-Whitney U test. A p-value <0.05 was considered statistically significant.

Results. We analyzed responses of 85 patients (30 females and 55 males) with NMD in this study. Demographic and clinical characteristics of patients are summarized in **Table 1**. The age was statistically different between the 2 groups. The RPNMD patients were older than the SPNMD patients (t=-5.41, p=0.00). The 2 groups differed with respect to gender as well. There were more women in the SPNMD group (chi square=8.8, p=0.06). The age of onset differed between the 2 groups. The age of onset in the SPNMD patients was earlier (t=-7.71, p=0.00). The duration of illness was longer for the SPNMD group (t=-6.86, p=0.00). The FIM total score did not differ between the 2 groups (z=-0.88, p=0.38). However, the SPNMD group scored higher (better) on the self-care parameter than the RPNMD patients (z=-2.79, p=0.005). The HRQOL as measured by NHP was significantly high (worse) in the RPNMD patients (200.61 ± 98.7) than the SPNMD (147.77 ± 144.9) patients (Mann-Whitney U test: z=-2.92, p=0.004). Only energy was significantly high (worse) among the dimensions of NHP in the RPNMD patients (z=-4.32, p=0.00). None of the other items differed between the 2 patient groups. Normative data is not available for NHP scores. The BDI scores of the RPNMD patients (10.4 ± 8.8) were below, and the SPNMD patients (18.9 ± 11.6) scored above the cut-off point of 16/17. Fifty-nine percent of the SPNMD and 11.9% of RPNMD patients had BDI score above the cut-off point. The SPNMD patients scored significantly higher (more depressed)

Table 1 - Patients' characteristics (N=85).

Variables	SPNMD group (n=39)	RPNMD group (n=46)
Gender, n (%)		
Male	19 (48.7)	36 (78.3)
Female	20 (51.3)	10 (21.7)
	mean ± SD	
Age, years	36.15 ± 14.9	53.2 ± 11.07
Age of onset	25.75 ± 18.84	52.14 ± 11.58
Duration of illness, years	11.63 ± 9.9	1.2 ± 2.3
Total FIM Score	110.85 ± 19.13	109.65 ± 19.11
Subgroups of the FIM scores		
Self-care	36.87 ± 8.5	32.52 ± 11.56
Sphincter control	13.3 ± 2.3	13.95 ± 0.29
Mobility	18.1 ± 5.1	18.1 ± 4.77
Locomotion	10.36 ± 3.7	11.17 ± 3.5
Communication	13.0 ± 2.36	13.7 ± 3.7
Social integration	18.8 ± 4.28	20.5 ± 1.6

SPNMD - slowly progressive neuromuscular disorders, RPNMD - rapidly progressive neuromuscular disorders, SD - standard deviation, FIM - functional independence measurement

than the RPNMD patients (Mann-Whitney U test: Z=-5.02, p=0.00). For the RPNMD patients, state anxiety scores (36.81 ± 6.24) were below, trait scores (41.37 ± 5.35) were above, and for the SPNMD patients, both state (45.64 ± 5.5) and trait anxiety scores (47.72 ± 5.2) were above the cut-off point of 40. In the SPNMD patients, 90.2% scored above the cut-off point of state and 95.2% patients scored above the cut-off point of trait anxiety scales. In the RPNMD patients, 56.8% were above the cut-off point of state and 20.5% were above the cut-off point of trait anxiety. The mean state and trait anxiety score differed significantly between the SPNMD (t=6.86, degrees of freedom [df]=83, p=0.00), and RPNMD patients (t=5.51, df =83, p=0.00). As the SPNMD and RPNMD groups differed with respect to gender, the differences between the 2 patient groups with respect to NHP and BDI scores, state anxiety and trait anxiety were analyzed in male and female patient groups separately. The NHP scores were significantly higher (worse) in male RPNMD patients (177.15 ± 73.3) than in male SPNMD patients (114.95 ± 110.7), (z=2.5, p=0.012). Only energy was significantly higher (worse) among the dimensions of NHP in male RPNMD patients (68.8 ± 30.4) than in male SPNMD patients (32.7 ± 32.14) (z=3.49, p=0.00) as well. Again, none of the other items differed significantly between the 2 patient groups in males. Male SPNMD patients (20.4 ± 15.2) were significantly more depressed than male RPNMD (8.4 ± 5.7) patients (Mann-Whitney U test; z=-4.09, p=0.00). The mean state and trait anxiety score differed significantly between male SPNMD (t=6.1, df=52, p=0.00) and male RPNMD patients (t=5.38, df=52, p=0.00). Both state (45 ± 5.4) and trait anxiety levels (47.1 ± 4.9) were higher in SPNMD patients than in RPNMD patients (state anxiety: 35.5 ± 5.4; trait anxiety: 40 ± 4.5) in males. The reanalysis for females revealed more or less similar results. The NHP scores were higher (worse) in female RPNMD patients (275.24 ± 132.44) than female SPNMD (178.9 ± 168.2) patients as well, however, in this case there was only a trend toward significance (z=1.9, p=0.057). Again only energy among the dimensions of NHP was significantly higher (worse) in women with RPNMD (77.1 ± 35.9) than in women with SPNMD (39 ± 36.1) (z=2.5, p=0.01). None of the other items differed significantly between the 2 patient groups in females as well. Female SPNMD patients (17.3 ± 6.1) were significantly more depressed than female RPNMD (11.5 ± 6.3) patients (z=-2.3, p=0.02). The mean state anxiety score differed significantly between female SPNMD and RPNMD patients (t=2.3, df=29, p=0.03). State anxiety levels were higher in SPNMD patients (46.3 ± 5.7) than in RPNMD patients (41 ± 7.1) in females, as well. Although the difference was not found to be significant (p=0.24), trait anxiety levels of women with SPNMD (48.3 ± 5.6) were higher than women with RPNMD (45.8 ± 5.6).

Discussion. In this study, we found that total HRQoL in the RPNMD group was worse, however, the difference was found to stem from the energy dimension. Otherwise, the 2 groups were comparable regarding HRQoL. No significant difference was found with respect to the other 5 dimensions (pain, physical mobility, emotional reactions, sleep, and social isolation). We can tentatively suggest that in the ambulatory stage of the NMD, the rate of progression does not affect HRQoL substantially. The intriguing results obtained in this study were the higher levels of depression and anxiety found in the SPNMD compared to RPNMD group. One of the reasons relevant to this difference between the 2 groups might be the gender issue. Female preponderance in the SPNMD group might have contributed to this difference. Prevalence of depressive and anxious states and disorders is known to be higher in women. What is more, it has been hypothesized that women would report more anxiety, distress, and complaints than men.[16,17] Nevertheless, levels of depression and anxiety were evaluated by self-report scales in our study. However, subsequent analyses of the male and female groups separately, with respect to psychological status and HRQOL revealed similar results excluding this probability regarding gender effect.

Only a few studies have examined the prevalence of depression and anxiety in patients with various NMD. In a recent study that included only SPNMD ambulatory patients, it was reported that 11% of patients presented current depression, and 32% of them had lifetime psychiatric comorbidity (depression being the most prevalent).[18] In our study, 59% of SPNMD patients had high depression scores (measured by BDI), and 90.2% have high state anxiety and 95.2% have trait anxiety. However, the reported prevalence of depression in ALS patients differs widely in previous studies. In a study exploring distress in ALS patients and their caregivers, it was reported that only 2% of patients with ALS have major depressive disorders, and 87% have no clinical depression.[19] Other series have reported frequencies of 11-75%.[20] In another study,[21] ALS patients were reported to have depressive (13-26%), and anxious symptoms (8-18%). The authors noted the lesser prevalence of depression in ALS, compared to other neurological disorders, which have slower progression rates such as, multiple sclerosis and Parkinson disease.[21] However, the measure that was used, or the associated symptoms of the patients might have affected the frequency of psychiatric symptoms. A total of 11.9% ALS patients

in the current study had depression (measured by BDI), 56.8% had state anxiety, and 20.5% had trait anxiety.

In a study including moderately affected ALS patients who all were still able to walk, the authors examined how ALS patients assigned emotional valence. They investigated the responses to visual socio-emotional stimuli, and reported that patients with a tendency to minimize extreme emotions tended towards more positive attributions, and had a more balanced arousal state in the early stages of the disease.[22] In another study,[23] which examined the performance of ALS patients on neuropsychological tests involving emotional perception and memory, it was found that they did not show any preferential recognition for emotional stimuli relative to neutral stimuli. This finding suggests that they show a different pattern of cognitive performance with respect to memory for emotional material when compared to healthy adults.[23] These comprehensive studies suggest that there may be some unknown, possibly organic protective effect against depression in ALS. Alternatively, it may be the result of a psychological reaction to the changed situation in which the patient finds himself. In accordance with the above mentioned findings, our results contradict the assumptions of a generally negative impact of the ALS compared to SPNMD on the emotional disposition as well.

In this study, in order to describe the HRQoL and psychological status comparatively, we matched the 2 groups with respect to functional level. The 2 groups consisted of ambulatory patients with comparable physical functioning. Subgroup analysis of functional level revealed merely worse self care in RPNMD patients, but sphincter control, mobility, locomotion, communication and social communication were comparable between the 2 groups. Moreover, the self-care parameter, as well as all other subgroups of functional level did not correlate with the HRQoL (data not shown) in the whole group. There is limited data regarding the association between HRQoL and the severity of functional impairment in NMD. In previous studies,[24,25] there are conflicting results with respect to this issue. In a study exploring sickness impact in muscular dystrophy patients, it was reported that the level of disability is not a critical factor that significantly alters life satisfaction, which is similar to our results,[24] and comparable findings were reported by Kohler et al.[25] They report that HRQoL is not correlated with physical impairment in DMD patients. The results from Grootenhuis et al[26] indicate that neuromuscular patients with more severe disease do not necessarily have worse HRQoL. Simmons et al[27] and Robbins et al[28] report that HRQoL assessed in patients with ALS, does not correlate with measures of physical function and strength. We found that HRQoL was worse in the RPNMD group, but in fact the 2 groups were comparable with respect to other dimensions, except energy. A poorer score in the energy dimension may be related to fatigue, which is reported to be the most disabling secondary symptom accompanying all stages of ALS.[29] The ALS is a rapidly fatal NMD, and one may expect to find that life satisfaction is worse than SPNMD in all aspects of HRQoL, given the fact all patients in this study were provided adequate information regarding the nature of their diseases by their physicians. Therefore, comparable HRQoL in the 2 groups is of considerable significance. However, while interpreting this finding, it is important to keep in mind that ALS patients were all in the ambulatory stage of their diseases. As a matter of fact, a study comparing the ALS patients at different levels of their disease found that HRQoL was the best of all levels in level one, including recently diagnosed ambulatory patients with mild deficit, and decreases systematically with increasing severity of illness.[8]

In SPNMD, HRQoL has been studied in heterogeneous patient groups with respect to the severity of functional impairment.[26,30,31] The comparable HRQoL found in the SPNMD group to ALS in our study is difficult to explain clearly. In a study exploring psychosocial aspects of SPNMD, authors reported that although physical impairment was not severe, learning of the incurable, progressive, and hereditary disease was traumatic, and the subjects hoped the diagnosis was wrong. They felt uncertain of the future.[32] Every successive functional impairment will go along with periodic sorrow, and will cause anticipation anxiety regarding the next loss of a certain ability. Moreover, the authors declared that the limitations imposed by the disease led to more sedentary activities in the home, and they withdraw socially and from outdoor activities. Consequently, they feel hopeless and helpless, which is a common reaction in the case of NMD, and which may be an aspect of a depressive state.[32] Additionally, in a comprehensive study comparing SPNMD with absent or very mild muscular deficits (MD and FSHD patients) with controls, it was stated that the patients perceived physical anhedonia with the anticipation of future physical disability.[33] Given the fact that our moderately dependent SPNMD patients had substantial anxiety and depression and comparable HRQoL to ALS patients, we can state that SPNMD patients warrant attentive evaluation of HRQOL and mental health throughout the process from the beginning of their disease.

Our results are in accordance with the literature supporting the view that significant depressive symptomatology is not an inevitable or common outcome of ALS, despite the severe impact of this fatal disease on patients' lives.[34] Again, in accordance with the above-mentioned research in this issue, HRQoL is

not associated with the measures of physical strength and function.[20] Our data does not provide a clear interpretation of better mood status than, or comparable HRQoL to SPNMDs group in ALS patients. We can only tentatively suggest that some compensatory factors, whether psychological, existential/religious, or organic, stemming from the course of the disease degenerative in nature, implicated by the previous research may be the explanation. Or else, immediately planned rehabilitation appropriate for ALS, which is a common practice of clinicians, as soon as the diagnosis is established, might have been working. We must add that the diagnoses of SPNMD, does not alert the physicians, or rehabilitation staff in an equivalent promptness in Turkey, even though patients may experience repeated loss of several abilities, given the chronic and progressive nature of the SPNMD. Factors such as loss of independence, certain social roles, and one's identity as a healthy person should be taken into consideration. These factors should be identified on an individual-based approach, beginning from the early stages of the disease, even when the patients were mild to moderately dependent. The results regarding the levels of depression, anxiety, and HRQoL in SPNMD patients found in this study, may contribute to the relevant awareness of the rehabilitation staff. Considering the long-term psychological well-being of SPNMD patients, and the sickness impact on psychosocial areas in this group, as well as in ALS patients, may enable the clinicians to better understand the patient's needs for psychosocial support. Long-term goals in rehabilitation and treatment plans should involve the psychological well-being and life satisfaction of patients with NMD.

The most important limitation to this study is the lack of similarity between the 2 groups, with respect to demographic characteristics such as age, gender, and other socio-demographic variables. However, as a consequence of the matching of functional level in these 2 progression groups, these demographic variables differed. Our aim was to find out and describe the differences regarding HRQoL and psychopathology, based on progression rate. Gender effect was controlled by the separate analysis of male and female patients. We did not aim to explore the influence of certain other factors on psychopathology and HRQoL. In future studies, other factors involved in the HRQoL in addition to progression rate should be explored. Furthermore, careful follow-up of these 2 groups of patients, which have different progression rates, and repeated assessments of psychopathology and HRQOL should be undertaken periodically. The reevaluation and comparison of the psychological features and HRQoL in the later stages should be performed as well, when both groups of patients have become severely dependent on help and support. Similar differences revealed between 2 groups in the advanced stages, would obviously strengthen the findings of this study.

In conclusion, management of NMD is frequently grouped together in one rehabilitation center regardless of the progression rate. In the case of ALS, which has a rapidly fatal progressive course, there is frequently more concern on HRQoL and psychopathology, which gives rise to meticulous planning of management by clinicians. They try to establish more rigorously the education of the patient and family members, psychosocial support, medications, regulation of sleep, and so forth. However, the comparable alertness in planning of the rehabilitation of SPNMD patients is lacking. In accordance with this issue, current practice in Turkey may lead to problems in the rehabilitation of patients with NMD. Nevertheless, the results of this study show that patients with SPNMD, even in the ambulatory stage carry a comparable (if not higher) risk regarding HRQoL and depression. It seems that the assessment of psychological status and HRQoL of SPNMD warrants close and continuous attention, beginning from the early stages of the disease. Patients with poor psychological status should be referred for further evaluation and counseling. Clinicians should be aware of possible depression or anxiety, and concerns of the patients regarding the chronic, though slowly progressive course of the disease. The substantial anxiety and depression found in ambulatory patients with SPNMD in this study warrants precaution for the later stages, as well. Rehabilitation plans of people with NMD have to be performed with a lifespan perspective. The identification of specific needs, expectations, fears of what the future might hold, coping skills, adjustment strategies, and resources of the patients in each phase of the disease, and implementing the necessary interventions accordingly will contribute to the long-term success of rehabilitation and treatment. Awareness of the factors, which may have an impact on the psychological well-being and life satisfaction is crucial in this regard.

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