

Predictors of health-related quality of life in adult ambulatory independence neuromuscular disease patients

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ABSTRACT

الأهداف: تقييم الصحة المتعلقة بجودة الحياة (HRQoL) مع مرضى العصب العضلي القادرين على المشي بشكل مستقل (NMD) مقارنة مع الأفراد الأصحاء النظراء من نفس العمر. وتقدير العلاقة بين المتغيرات الاجتماعية وجودة الحياة HRQoL في مرضى NMD القادرين على المشي بشكل مستقل.

الطريقة: اشتملت هذه الدراسة على 99 مريض بالغ تم عرضهم وتشخيصهم بأمراض عصبية عضلية NMD - قسم العلاج الطبيعي - كلية العلوم الصحية - خلال الفترة ما بين 2007م وحتى 2009م. كان المقياس الوظيفي (FIM) وملف نوتينغهام الصحي (NHP) هما المؤشرات الرئيسية لتقييم مستوى الاعتماد على الذات في النشاطات اليومية وجودة الحياة.

النتائج: تم قياس درجات (HRQoL) بواسطة (NHP) وكانت عالية (سيئة) في مرضى مجموعة التحكم الأصحاء. كان الاختلاف بين المجموعتين مهم للطاقة، والنشاط الجسماني، والدرجات الكلية. حقق الأفراد المرضى العاملين والمصابين ب (NMD) درجات أقل (أفضل) من الناحية الإحصائية في مقياس (NHP). ولم تظهر أي اختلافات مهمة بين الجنس، ومدة المرض بين جميع درجات البعد.

خاتمة: لدى جميع مرضى (NMD) درجات ضعيفة في مقياس الصحة المتعلقة بجودة الحياة (HRQoL) عن الأفراد الصحيين بخصوص الطاقة، و أبعاد النشاط الجسماني، والدرجات الإجمالية. إضافة إلى ذلك، يعتبر العامل الأساسي الذي يؤثر على (HRQoL) في مرضى العصب العضلي البالغين القادرين على المشي (NMD).

Objectives: To evaluate the health related quality of life (HRQoL) of totally independent ambulatory neuromuscular disease (NMD) patients in comparison with age matched healthy control subjects, and to assess associations between socio demographic variables and HRQoL in totally independent NMD patients.

Methods: Ninety-nine adult patients with a diagnosis of NMD referred to the Physical Therapy Department of the Health Sciences Faculty of Hacettepe University, Ankara, Turkey between 2007 and 2009 were included in the study. The Functional

Independence Measurement and the Nottingham Health Profile (NHP) were the main outcomes to assess independence level in activities of daily living and quality of life.

Results: The HRQoL score as measured by NHP was high (worse) in patients than healthy controls, and the difference between the 2 groups was significant for energy, physical mobility, and total score. Employed NMD patients scored significantly lower (better) than those unemployed in the majority of NHP domains. The genders and duration of illness displayed no significant difference in all dimension scores.

Conclusion: All NMD patients had a poorer HRQoL than with healthy subjects with respect to energy, physical mobility dimensions, and total score. Furthermore, occupation was found to be a main factor that affects HRQoL in adult ambulatory NMD patients.

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Quality of life (QoL) 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.¹ 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.² In recent years, there has been a growing awareness that a comprehensive assessment of health outcomes must involve patient-reported judgments of physical, mental, social,

and psychological functioning as well as traditional indicators of disease severity and functional status.³ Many neuromuscular diseases (NMD) are chronic and slowly progressive. Patients have permanent, residual disability caused by non reversible pathologic alteration, so they require special training for rehabilitation or may be expected to require a long period of supervision, observation, or care.^{4,5} Although different types of NMD patients display different symptoms, the main common symptom is as the disability increases, the individual's functional problems, activity limitation, and social participation will decrease. Beside physical symptoms like progressive muscle weakness, shortness, fatigue, decrease in aerobic capacity, demographic factors such as age of the patient, and duration of the illness can limit daily activities and decrease QoL by causing deficiency in functional activities.^{6,7} In recent years, there has been an increase in the studies that assess the influence of independence in daily living activities to QoL. It is suggested that there is a correlation with disability level of the patient and their QoL. An example is a study in which 77 adults with Duchenne muscular dystrophy were assessed. They concluded that increased dependence on others and difficulty in performing activities of daily living (ADL) have a weak to moderate correlation with lower QoL.⁸ This implies that a decline in QoL can only partly be explained by a decline in ADL-ability. Beside physical condition, psychological states, interpersonal relationships, social, and economic state have an important effect on the HRQoL.⁹ Stressors that negatively affect QoL such as complications, depression, and care problems are the main predictors of QoL.¹⁰ Previous studies combined data from patients with rapid progressive NMD disease. Although there are several reports examining QoL in severely disabling, rapidly progressive, and fatal neurological diseases such as ALS, the mild disabled patients are not examined truly regarding HRQoL. Patients included in these studies had severe problems of independence in daily living activities. Less is known about HRQoL of ambulatory independent patients. Furthermore, previous research mostly tended to focus on HRQoL of children with NMD. As the requirements and demands of daily living activities of the children and adult patients are different, it is difficult to interpret this knowledge in an adult population.¹¹ As NMD patients need long duration rehabilitation programs, identifying and preventing variables that can effect HRQoL before residual disability occurs will be beneficial to maintain and enhance QoL.¹² Therefore, the aim of this study was 1) to evaluate the HRQoL of totally independent ambulatory NMD patients in comparison with same age matched healthy control subjects. 2) To assess associations between socio demographic variables and HRQoL in totally independent NMD patients.

Methods. This study included 99 adult patients diagnosed with NMD by the Neurological Department of Hacettepe University Hospital, Ankara, Turkey and referred to the Physical Therapy Department of the Health Sciences Faculty between 2007 and 2009. Also 52 healthy subjects were included to the study as the control group. The control group was age matched healthy subjects who agreed to participate in the survey after being randomly selected from among a patient's community control. Written informed consent was obtained from all participating subjects. Hacettepe University Ethical Committee approved the study. The inclusion criteria were having a diagnosis of slowly progressive disease characterized by muscle and lower motor neuron lesion, able to attend the outpatient clinic, to be under supervision in a neurological rehabilitation clinic in terms of physical therapy and rehabilitation. Patients were excluded if they were suffering from another neurological, or orthopedic disease that might affect physical or cognitive function.

Measures. 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; the lowest level (1) indicates the total need for the assistance of 2 helpers to perform the activity. The normal range of the FIM score is between 18-126 (18-36: completely dependent, 37-90: needs supervision and assistance while performing activities, 90-126 completely independent).¹³ The FIM data may be reported in terms of FIM motor score, the sum of the first 13 items. The FIM motor scores range from 13-91. In our study we also used FIM motor scores. For each FIM item, we also calculated the percentage of patients requiring assistance (FIM score <5), supervision (FIM score = 5) and patients who are independent (FIM score >6) to indicate item difficulty, similar to Uchikawa.¹⁴ The FIM was adapted for Turkish language and found to be reliable and valid in a spinal cord injury sample.¹⁵ The Nottingham Health Profile (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; the subjects may answer as 'yes' or 'no'. Normal range of the NHP score is between 0-600 for total score, 0-100 for subgroups score. Higher scores on the NHP represents

poorer QoL. Turkish versions of these scales have been developed and thoroughly tested for reliability and validity in Turkey.¹⁶

The same researcher applied questionnaires and functional evaluations for all subjects during the study. The self-report instruments, which were all completed in the unit, took approximately 15 minutes. At the beginning of the study age, gender, disease duration, current status of employment, and role in the family were recorded.

Statistical analysis. Statistical analysis was performed using SPSS (version 15.0) statistical software. Continuous variables were described by mean (x) and standard deviation (SD). Categorical data are given as counts and percentages. Parametric tests were used for comparative analysis. The paired Student's t-test was used to compare quantitative variables. The level of significance was set at $p < 0.05$.

Results. We analyzed the response of 99 adult patients with NMD and 52 age matched healthy subjects in the study. Of the patients, 14.6% had limb girdle muscular dystrophy, 7.9% sensory motor neuropathy, 5.3% fascio-scapula-humeral muscular dystrophy, 4.6% had Becker muscular dystrophy, and 33.1% diagnosed as other myopathies. Disease-related demographics and clinical characteristics of patients and healthy subjects are summarized in Table 1. The median duration of signs was 89.85 months (range, 1-516 months). We found no significant differences in socio-demographic variables and age between the 2 groups. The FIM motor scores ranged from 46-91 mean 85.21 ± 9.25 . Figure 1 illustrates the percentages of patients requiring assistance (FIM score <5), supervision (FIM score = 5), and who were independent (FIM score >6) for each FIM item.

Table 1 - Demographic and clinical features of patients.

Variables	Patients (N=99)	Control (N=52)
	n (%)	
<i>Gender</i>		
Male	45 (45.5)	14 (26.9)
Female	54 (54.5)	38 (73.1)
Age (years, mean±SD)	37.53±15.14	43.00±10.04
Duration of illness (months, mean±SD)	89.85±109.32	-
<i>Status of employment</i>		
Yes	49 (49.5)	21 (40.4)
No	50 (50.5)	31 (59.6)
<i>Role in the family</i>		
Mother	21 (21.2)	17 (32.7)
Father	21 (21.2)	20 (38.5)
Child	44 (44.4)	12 (23.1)
Partner	9 (9.1)	2 (3.8)
Single	4 (4.0)	1 (1.9)

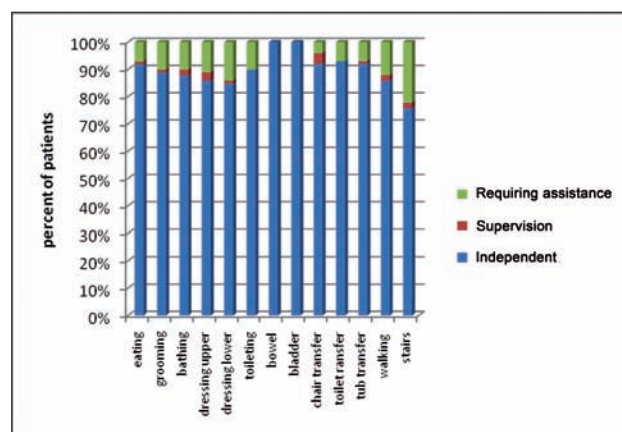


Figure 1 - Percentage of patients requiring assistance.

Table 2 - Comparison of NHP dimensions between the 2 groups.

NHP dimensions	Patients	Controls	P-value
Energy	48.44±34.58	28.04±29.91	0.000
Pain	21.29±29.81	14.49±24.41	0.137
Emotional reactions	29.83±28.82	29.14±29.51	0.892
Social isolation	21.41±28.14	19.84±24.22	0.723
Sleep	19.00±27.21	19.16±28.04	0.982
Physical mobility	39.87±23.74	11.96±19.46	0.000
Total	180.19±127.99	123.63±116.75	0.008

NHP - Nottingham Health Profile

The percentage requiring assistance was the highest for stair item (22.2%), followed in order by dressing lower body (14.1%), walking (12.1%), dressing upper body (11.1%), bathing (10.1%), grooming (10.1%), toileting (10.1%), eating (7.1%), tub transfer (7%), toilet transfer (7%), and chair transfer (4%). All patients were independent for bowel and bladder management (Figure 1). The results indicate that the HRQoL score as measured by NHP was high (worse) in patients than healthy controls, and the difference between the 2 groups were significant for energy, physical mobility, and total score (Table 2). The largest difference was in the energy and physical mobility dimensions, which were higher (worse) in patients than in controls.

With respect to the total study population, the lowest HRQoL was found in the dimension of energy and physical mobility. In general, the dimensions of pain, sleep, social isolation, and emotional reactions showed a markedly better profile than energy and physical mobility. Figure 2 shows the characteristics of the study and control group with regard to the dimensions of the NHP, energy, pain, emotional reaction, sleep, social isolation, and physical mobility. Table 3 shows the effects of the covariates: gender, age, employment status, and duration of illness on the dimensions of

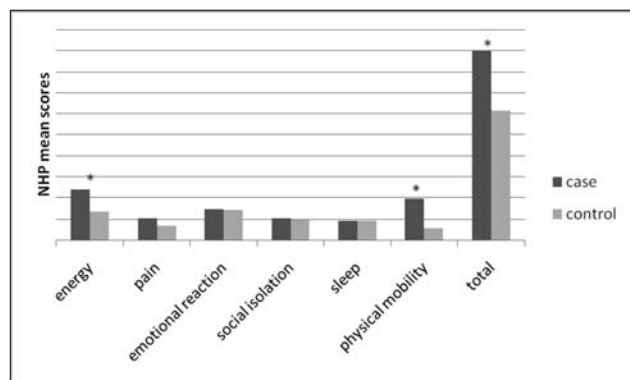


Figure 2 - Distribution of Nottingham Health Profile dimensions among neuromuscular disease patients and control subjects.

the NHP. Men showed higher (worse) scores on the NHP dimensions of energy, emotional reactions, sleep, and total NHP scores, however, the genders displayed no statistically significant difference in all dimension scores. Employed NMD patients displayed significantly lower (better) scores than those unemployed in pain, emotional reactions, social isolation, sleep domains, and total NHP scores. There was no statistically significant difference in all dimension scores and total NHP score between the patients with longer duration (>8 years) and the patients with a short duration (<8 years). There was also no significant difference in the total NHP score in patients with NMD older than 40 years and in patients younger than 40 years. The age >40 group showed significantly higher (worse) scores on the NHP in dimensions of physical mobility.

Discussion. The results of this study revealed that although patients had higher functional level during ADL, NMDs have profound effects on HRQoL, when compared with healthy subjects with respect to energy, physical mobility dimensions, and total score. Furthermore, occupation was found to be a significant and independent predictor for HRQoL of

adult ambulatory independence NMD patients. As the traditional focus is on physical disability in the neuromuscular outpatient clinics, measuring of QoL is overlooked and generally stressors of HRQoL are not anticipated in the mild dependent group.¹⁷ Therefore, the results of this study can provide some clues to clinicians on the long term rehabilitation process of these patients.

Generally, NMD patients scored higher (worse) on all NHP sub parameters. The largest of the differences between NMD and controls were found for energy and physical mobility dimensions. These findings are in line with results obtained elsewhere.^{18,19} For example Antonini¹⁹ showed in his study that patients' SF-36 mean scores were lower than those of controls in all the dimensions of SF-36. Previous research shows that employed patients showed higher levels of QoL than unemployed patients.¹⁹ An earlier study on muscular dystrophy shows that patients had lower QoL regarding having no work or meaningful occupation.¹² Similar findings emerged in our study, the employed group of NMD patients had higher levels of QoL than the unemployed patients. Employment had a positive effect for patients. Therefore, encouraging working abilities, hobbies, meaningful activities, or occupation may be a fundamental part of the rehabilitation process in these patients.

Some studies found that advanced age has a negative impact on QoL in patients with NMDs.¹² Due to important psychosocial aspects generically linked to advancing age, this may be an expected result. Therefore, in this study the dichotomy in age is made to evaluate the effects of age. However, in this current study aging was not found to be associated with low HRQoL of the patients. Similarly, in this study, long duration of illness patients do not necessarily have a worse HRQoL. This is in line with findings of Boström.¹² He found that illness duration is not related with HRQoL. In slowly progressive types of NMD, patients are living with their disease for a long time from the beginning of childhood. There may be a gradual adaptation to the change in the

Table 3 - Mean scores of NHP by gender, employment status, duration of illness, and age.

NHP	Gender		Employment status		Duration of illness		Age	
	Men (n=38)	Women (n=33)	Employed (n=49)	Unemployed (n=50)	<8 years (n=67)	>8 years (n=32)	<40 years (n=58)	>40 years (n=41)
Energy	49.49	47.56	44.83	51.97	50.96	43.15	45.71	52.09
Pain	19.80	22.54	15.84**	26.64	19.56	24.92	18.89	25.82
Emotional reactions	30.10	29.60	21.48*	38.00	30.88	27.62	30.24	29.24
Social isolation	21.30	21.50	16.01*	26.70	21.63	20.95	17.89	26.39
Sleep	21.02	17.40	12.01***	25.94	16.72	23.92	17.84	20.75
Physical mobility	38.50	41.01	37.27	42.41	38.10	43.57	32.06	50.91**
Total	181.54	179.06	148.35*	211.38	178.73	183.23	163.03	204.45

* $p < 0.05$, ** $p < 0.01$, *** $p < 0.001$, NHP - Nottingham Health Profile

conditions of one's life. The results of the present study indicate that gender is not a factor that influences QoL. When examining gender differences, we found men showed significantly higher (worse) scores on the NHP dimensions of energy, emotional reactions, and sleep however, the genders displayed no significant difference in all dimension scores.

The perception of difficulty is a personal experience behind the main symptoms like muscle weakness. Local and general fatigue, socio-cultural and environmental factors might play a role in perceived difficulties.²⁰ In the current study, we tried to describe the difficulty pattern by calculating the percentage of patients requiring assistance. Although all the patients were independent according to total score of FIM, they reported difficulties in various activities. The results of our study confirmed previous findings since they showed that NMD patients had most difficulty in the stair item followed in order by dressing lower body and walking.

A limitation of the study is that a homogenous patient sample should be included. Further research must be carried out within the same homogenous subgroups defined by relevant parameters. Identifying homogeneous groups can provide insights into the characteristics of QoL.

As HRQoL is influenced mostly by physical activity and energy level it is obvious that exercise to maintain physical mobility and energy conservation techniques ought to be fundamental interventions. The physical, psychological, social, and medical problems of the patient change over time with progressive NMD. The literature revealed that the negative effects of parameters like physical, emotional, and social factors affect the QoL in the late stage of the disease. However, little study has been conducted on ambulatory NMD patients. Our results clearly indicate that ambulatory NMD patients had lower QoL in comparison with healthy subjects. Additionally, occupation was found to be a significant and independent predictor for HRQoL of adult ambulatory independent NMD patients, and it should be an essential part of rehabilitation. Thus, the factors associated with HRQoL should be evaluated and followed periodically in these patients. Greater knowledge means more opportunities to create optimum conditions for rehabilitation.

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