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DISABILITY AND QUALITY OF LIFE IN INDIVIDUALS WITH MUSCULAR DYSTROPHY

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ABSTRACT. In the county of Örebro, Sweden, 32 individuals with myotonic disorders and 25 with other types of muscular dystrophy were examined. Disability was assessed with functional tests and standardized observations of muscle function (mainly based on those proposed by Dr. Brooke), a new self-administered questionnaire regarding the Activities of Daily Living (ADL) and the ADL staircase (based on Katz ADL index). The results of the different tests of disability were highly correlated. The Sickness Impact Profile and the Kaasa test were used for assessing the quality of life, and no significant differences were found between the groups of muscular dystrophy. In an explanatory factor analysis three main factors of disability were found. The factors "walk and move" and "finger function" were fair to good associated with the quality of life. This study offers an approach for research on the consequences of muscular dystrophy using established as well as new methods.

Key words: activities of daily living, disability, dystrophia myotonica, multimethod validation, muscular dystrophy, quality of life.

INTRODUCTION

During the last decade, a strictly medical viewpoint has been supplemented with psychosocial considerations in the endeavour to take a multidimensional view of what it means to be stricken with a disease (8). This change has created an interest in methods of measuring which focus on the patient's perception of being chronically ill (8, 41).

In this study of individuals with muscular dystrophy (MD), the consequences of the disease are described in terms of disability and quality of life (QoL). Disability reflects functional performance and activity, and represents disturbances at the level of the person (45). In the present study, functional performance is assessed by standardized tests of the muscle function. A common way to assess disability is also by rating the ability to perform the activities of daily living (ADL) (14, 23, 28, 30, 40). To our knowledge, there are no other studies published on individuals with MD where variables of ADL are included in the concept of disability.

QoL has emerged as a concept which incorporates a multitude of subjective experiences perceived as important for individuals (44). The definitions of QoL differ according to the theoretical point of view on which they are based. Most of the definitions can be placed in one or, usually, several, of the following four major domains: (i) physical status and functional abilities, (ii) psychological status and well-being, (iii) social interactions, and (iv) economic status and factors (41). In this study we used measuring instruments which cover the first three domains.

In a previous study, spirometry and arterial blood parameters were found to be pathological in 58% and the ECG abnormal in 37% of the 57 individuals afflicted with MD in a population survey. The result also indicates a weak association between objective and subjective ill-health (3).

The main aim of the present study was to assess the impact of the disease MD in terms of disability and QoL. Also, the measurement instruments were reviewed in order to assess reliable and valid measurements suitable for MD.

METHODS

Subjects

In a population survey of neuromuscular diseases in the county of Örebro (approx. 270,000 inhabitants), all individuals afflicted with different types of MD or myotonic disorders were identified (4). Fifty-seven out of 67 subjects (85% of all eligible adults) in the age range 16–64 years agreed to participate in all parts of the study. Three individuals were excluded because of incomplete participation. The design and protocol were approved by the regional ethics committee. The distribution by diagnosis was 32

Table I. *Factor loadings of items in the self-report ADL scale (n = 57)*

	Factors (categories)			
Items >0.50 in only one factor	Ambulation	Finger strength	Arm strength	Finger subtle function
Go downstairs	0.88			
Go upstairs	0.87			
Walk in the forest	0.82			
Cycle	0.80			
Walk indoors	0.78			
Walk outdoors	0.77			
Rise from the floor	0.67			
Vacuum-clean the floor	0.66			
Wash the floor	0.66			
Rise from a chair	0.60			
Remove screwcaps from jam pots		0.77		
Move frying pan from cupboard to cooker		0.77		
Remove screwtops from bottles		0.76		
Carry bags with food		0.76		
Cut meat, bread, etc.		0.65		
Put on ear-clips			0.86	
Comb hair			0.77	
Hang up the washing on a clothes line			0.72	
Put on necklace			0.71	
Tie shoe-lace			0.67	
Put plates in the wall-cupboard			0.63	
Cook in oven			0.63	
Pick up things from the floor			0.57	
Take plates out of the wall-cupboard			0.54	
Turn on taps				0.81
Button				0.79
Put on gloves				0.70
Turn on electric-stove controls				0.57

cases with *myotonic disorders* (myotonic dystrophy 30 and myotonia congenita 2), eight cases with *myopathia distalis tarda hereditaria* (MDTH) and 17 cases with other types of a primary MD, designated "*Other MD's*" and comprising limb-girdle MD 5, myopathy with respiratory failure MRF 4 (19), facioscapulohumeral MD 3, Becker MD 2, Emery-Dreifuss MD 1 and two cases with a verified diagnosis of MD, although the exact type had not been established. Out of the 57 individuals, 45 were ambulatory and 12 were mainly confined to the use of a wheelchair. Thirty-five individuals had disability insurances (28 had partial or total early retirement pensions, 3 had partial or total temporary disability pension, and 4 had partial or total sickness cash benefit for more than three months).

Three independent measurements of disability

Brooke muscular examination scheme. All participants were examined by a neurologist (LGG) and their muscular function was assessed according to the functional evaluation suggested by Brooke (12). The examination procedure encompassed a standardized observation of the patient with regard to gait, stepping onto a footstool and onto a chair, and rising from the floor and from a chair (see Appendix A). All individuals were given standardized instructions. Their abilities were graded from 0 (normal) to 4 (fails). Each variable was evaluated separately and together with other variables, in order to obtain patterns of variables which discriminated between the individuals. Two variables in Brooke's observation scheme did not discriminate further between the individuals and were therefore excluded, namely "Compensatory lordosis" (Gait) and "Initial turn" (Rising from floor).

With regard to each of the functions—gait, stepping onto footstool/chair, rising from chair/floor, grip strength and graded functional evaluation—scores of relative disability were calculated, i.e. the percentage of disability in those variables constituting the function. Thus, zero implies normal function/strength and 100% an individual failing in all these tests. Grip strength was measured with a standard dynamometer, uncalibrated, scaled 0–70. Four timed functional tests according to Brooke (12) were performed, but these tests were excluded from the results since 12 participants did not perform them.

On the basis of extensive clinical experience Brooke has proposed that the level of function can be graded with regard to hips and legs, arms and shoulders and bulbar function (12). We used a slightly expanded variant of Brooke's grading system (Appendix B).

Table II. *Percent disability of muscular function assessed by Brooke's grading system and functional tests*

	Myotonic disorders $n = 32$	MDTH $n = 8$	Other MD's $n = 17$	All $n = 57$
Brooke's grading system				
Hips/legs	5.5	2.1	38.2	14.8
Arms/shoulders	6.3	4.2	33.3	14.0
Bulbar	18.0	0.0	4.4	11.4
Brooke's functional tests				
Mobility index:	23.9	24.4	65.1	36.4
Gait	11.2	15.7	44.1	21.7
Stepping onto footstool (20 cm)	18.6	21.1	62.1	31.9
Stepping onto chair (45 cm)	38.7	34.4	83.8	51.5
Rising from chair/floor	29.3	31.3	72.1	42.4
Grip strength	14.2	20.5	24.6	18.2

MDTH: myopathia distalis tarda hereditaria; MD: muscular dystrophy.

The ADL staircase. The Katz ADL index comprises performance of six personal activities of daily living important for self-care (P-ADL), namely bathing, dressing, toileting, transfer, continence and feeding (28, 29). The index has been translated into Swedish and the reliability and validity of this version have been tested (13). In the present study, we used a Swedish extended version, the ADL staircase, with four complementary instrumental activities important for independence of the environment (I-ADL): shopping, cleaning, transportation and cooking (24, 39). ADL performance is ranked in 10 cumulative ADL steps from total independence of other persons to complete dependence: 0 = independent in all 10 activities, 1 = dependent in one activity, 2 = dependent in cleaning and one further activity, 3 = dependent in cleaning, shopping and one further activity, 4 = dependent in cleaning, shopping, transportation and one further activity, 5 = dependent in all I-activities and one P-activity, 6 = dependent in I-ADL, bathing and one further activity, 7 = dependent in I-ADL, bathing, dressing and one further activity, 8 = dependent in I-ADL, bathing, dressing, going to the toilet and one further activity, 9 = dependent in I-ADL, bathing, dressing, going to the toilet, transfer and one further activity, 10 = dependent in all 10 activities, Others = dependent in two or more activities, but not classifiable as above. A prerequisite for a reliable result is that this category does not exceed 5%.

Usually, the ADL staircase is based on observational data, but in the present study interviews in the homes of the participants were used. This method was chosen because the interviewer had not previously met the participants and therefore performing observations might have been considered objectionable.

The self-report ADL scale. Because available instruments for the assessment of ADL allowed only a fairly rough classification of individuals with MD, a new questionnaire was developed consisting of 34 items involving a variety of common activities of daily living. The response alternatives were "no difficulties", "succeeds but with difficulty" and "fails", which were assigned the values of 0.0, 0.5 and 1.0, respectively. The responses within each category were added and the sum was divided by the number of items and then multiplied by 100, thus deriving the disability score for each category expressed in per cent. The disability index for all items together was calculated accordingly.

After exploratory factor analysis (orthogonal varimax rotation), 28 items were retained and grouped into four factors. All these items had a factor loading greater than 0.5 in only one of the four factors (Table I). The factors were designated ambulation, arm strength, finger strength and finger subtle function. These categories accounted for 76% of the total variance (Table I).

Quality of life (QoL)

The Sickness Impact Profile (SIP). The SIP questionnaire consists of 136 items, grouped in 12 categories: sleep and rest, emotional behaviour, body care and movement, home management, mobility, social interaction, ambulation, alertness behaviour, communication, employment, recreation and pastimes and eating (10). Scores are calculated for each category using predetermined item weights based on estimates of the relative severity of each dysfunction. In summary scores, the physical index combines body care/movement, mobility and ambulation, and the psychosocial index combines emotional behaviour, social interaction, alertness behaviour and communication. The overall index is based on all 12 categories. The results are given as percentages of maximal dysfunction and higher scores mean a poorer health-related QoL. The SIP was translated into Swedish in 1981, and revised and evaluated for Swedish circumstances according to recommended standards for cross-cultural applications (42, 43). This version has since been used in several studies (6, 9, 11, 31, 35, 37, 38).

The Kaasa questionnaire. The Kaasa questionnaire consists of 12 questions about the individual's subjective evaluation of his/her life and situation. No questions are related to individuals' functional performance. The instrument is subdivided into three indices: psychosocial well-being, happiness and satisfaction. The psychosocial index consists of 10 items, with 5 positive and 5 negative statements in order to provide valid results. The 10 items are scored 1–5. The indices of happiness and satisfaction are scored 1–7 and each consists of only one item (25, 26).

Procedures

Participants were interviewed twice in their homes with an interval of 3 months, all by the same interviewer (GA). The items included in the ADL staircase were transformed into questions which were part of the first interview. Before the second interview, the participants completed the SIP. The self-report ADL scale was administered during the second interview. The interviews also included questions about coping, reported elsewhere (5). Between 2 and 3 months after the second interview, participants were examined by a neurologist for assessment of muscular function and confirmation of the diagnosis. Except for a few cases with myotonic dystrophy, all cases were verified with a muscle biopsy or electromyography (usually both).

Statistical analysis

Analyses of variance (ANOVA) were used to test differences between types of MD with respect to disability and QoL (F-test and Fisher PLSD). Correlations were calculated with the Pearson coefficient of correlation (r). In the case of data on nominal and ordinal scales Spearman Rank correlation (rho) was applied. Correlations were classified as poor (<0.40), fair to good (0.40–0.75) and excellent (>0.75) (20). The results of the latter analyses are present only when the coefficients were not within the same class. In order to map fundamental dimensions a factor analysis (principal component analysis with varimax rotation, orthogonal solution) was carried out on the variables of disability. A multitrait-multimethod matrix was applied in order to examine convergent and discriminate variables of disability between different methods (15).

RESULTS

Disability in muscular dystrophy

The graded disability assessed by Brooke's tests is shown in Table II. The profile was similar in the myotonic disorders and MDTH, except for bulbar function, which showed a significantly stronger reduction in the case of myotonic disorders than in the case of the other types ($F = 9.4$, $df = 2/54$, $p < 0.001$). In all other variables, with the exception of grip strength, individuals with myotonic disorders and MDTH were less impaired than the group of "Other MD's". The mobility index, all the variables included in it (except for "stepping onto a chair"), was strongly correlated to the variables "hips/legs and arms/shoulders" in Brooke's grading system ($r = 0.77–0.85$, $p < 0.001$). This indicates that these variables (included in Brooke's test) could be used interchangeably.

The disability estimated according to the ADL staircase is shown in detail in Table III. Fifty-six per cent (32/57) of all cases were dependent on other persons in one or more of the 10 activities of daily living. The degree of dependency differed significantly ($F = 9.04$, $df = 2/52$, $p < 0.001$) between those with MDTH (25% or 2/8) and those with "Other MD's" (88% or 15/17). No significant difference was found between myotonic disorders and MDTH.

Table III. *Number of individuals with muscular dystrophy (MD) graded by the ADL staircase*

Grades/steps	Myotonic disorders	MDTH	Other MD	All
Independent	17	6	2	25
1	6	1	5	12
2	1	0	0	1
3	3	0	1	4
4	2	0	1	3
5	0	0	0	0
6	2	0	1	3
7	1	0	1	2
8	0	0	5	5
9	0	0	0	0
10	0	0	0	0
Others	0	1	1	2
Total	32	8	17	57

MDTH = *myopathia distalis tarda hereditaria.*

Data from the self-report ADL scale are shown in Table IV. The results are in accordance with those reported above. The correlations between the four categories of this scale (ambulation, arm strength, finger strength and finger subtle function) and the ADL staircase were $r = 0.90$, 0.74, 0.54 and 0.41, respectively ($p < 0.001$). This indicates that the self-report ADL scale mainly contributed information about the finger function in addition to what was provided by the ADL staircase.

In order to study how the variables from different methods of measuring disability were associated, an exploratory factor analysis was performed. Three main factors were identified, namely "walk and move", "finger function" and "bulbar function". These factors together explained 82% of the total variance and individually 64%, 24% and 12%, respectively. The first two factors had a fairly positive correlation (0.53), but bulbar function was not correlated with either of the other two factors (0.02 and −0.09). Table V gives the associations between the different variables from different measuring methods.

There were no statistically significant differences between the groups of MD with regard to any of the indices of the Kaasa questionnaire and of the SIP. The means of the Kaasa questionnaire indices were for "psychosocial well-being" 2.1 (SD 0.7), "Happy" 3.1 (SD 1.1) and "Satisfied" 2.5 (SD 1.1). SIP indicated reduced QoL (high scores) for the "physical index" (mean 13.5, SD 16.7); the highest impact in this index

Table IV. *Disability assessed with the self-report ADL scale categories with regard to type of muscular dystrophy (MD) (n = 57)*

	Ambulation	Arm strength	Finger strength	Finger subtle function	Total index
	Score % Mean (SD)	Score % Mean (SD)	Score % Mean (SD)	Score % Mean (SD)	Score % Mean (SD)
Myotonic disorders	26.6 (29.0)	16.08 (23.0)	38.1 (25.3)	15.02 (15.5)	23.09 (21.4)
Myopathia distalis tarda hereditaria	28.1 (16.2)	38.9 (11.5)	33.8 (15.1)	37.5 (13.4)	33.9 (10.8)
Other MD	61.2 (32.3)	45.1 (26.0)	34.7 (26.3)	18.04 (25.8)	45.2 (25.4)
All	37.1 (32.4)	28.4 (26.1)	36.5 (24.2)	19.03 (20.1)	31.6 (23.3)

was that of ambulation and body care/movement. Higher QoL was found for the "psychosocial index" (mean 7.1, SD 10.9). Categories not included in these two indices were "home management", "employment", "sleep/rest" and "recreation/pastimes", and all these showed, as above, low QoL.

No variables of disability were significantly associated with the three indices of the Kaasa questionnaire. The variables related to mobility of the body showed good to excellent correlation to the overall and physical index of the SIP (Table VI), while most of the variables representing functions in distal muscles showed a considerably lower correlation. The association between the psychosocial index and

Table V. *Correlations between variables of disability measured with different methods*

	Walk and move Self-report ADL scale				
Walk and move	ADL staircase	Ambulation	Arm strength		
Brooke measurement					
Hips/legs	0.90***	0.85***	0.70***		
Arms/shoulders	**0.65*** **	**0.66*** **	0.75***		
Mobility index	**0.75*** **	0.87***	0.65***		
ADL staircase		0.90***	0.74***		
Self-report ADL scale					
Ambulation	0.90***		**0.77*** **		
Arm strength	0.74***	**0.77*** **			
				Self-report ADL scale	
Finger function				Finger strength	Finger subtle function
Brooke measurement					
Grip strength	0.37**	0.38**	0.34*	0.59***	**0.40** **
Self-report ADL scale					
Finger strength	0.54***	0.52***	0.61***		0.57***
Finger subtle function	0.41**	0.45***	0.57***	0.57***	

* $p < 0.05$; ** $p < 0.01$; *** $p < 0.001$.
Bold type = rho.

Table VI. *Associations between disability and various indices of the Sickness Impact Profile (SIP)*

	SIP		
	Overall r	Physical index r	Psychosocial index r
Walk and move			
Hips/legs	0.58***	0.72***	0.27*
Arms/shoulders	0.60***	0.72***	0.35**
Mobility index	0.60***	0.74***	0.29*
ADL staircase	0.71***	0.81***	0.44***
Ambulation	0.71***	0.82***	0.41**
Arm strength	0.67***	0.71***	0.43***
Finger function			
Grip strength	0.41**	0.45***	0.29*
Finger strength	0.49***	0.51***	0.29*
Finger subtle function	0.47***	0.50***	0.34**
Bulbar function	0.10	0.01	0.2

*$p < 0.05$; **$p < 0.01$; ***$p < 0.001$.

variables of disability was in general low also in the SIP, and the highest correlation found was to the ADL staircase and to the variables Ambulation and Arm strength in the self-report ADL scale. Bulbar function was the only variable which was not correlated to the SIP indices. In summary, the results verified the relative independence of well-being (Kaasa) and functional abilities (SIP), which had been demonstrated in a previous study of this group of individuals (3).

DISCUSSION

In our study group identified in a population survey the degree of disability varied between the different types of MD. The myotonic disorders and MDTH were closely related with regard to degree of disability except for bulbar function. In contrast, the individuals with "Other MD's" were considerably more disabled. This result is in accordance with clinical experience, namely that myotonic disorders and MDTH in general are more mild diseases than those included in the group of "Other MD's" (12). However, two individuals with myotonic disorders and one with MDTH were using a wheelchair in everyday life. This fact stresses the importance of an individualized judgement and nursing based on every subject's unique situation rather than on generalizations.

In line with our findings, a measurement of disability in the case of individuals with MD should comprise variables for ambulation, arm strength, finger strength, finger subtle function and bulbar function. The results concerning disability constitute valuable tools for improving the communication about and understanding of the handicapping conditions.

The psychosocial well-being was evaluated using the Kaasa test and the results were similar to those of 95 patients with lung cancer (25). Every patient was given the questionnaire on eight different occasions, before radiotherapy or chemotherapy was instituted and during the year that followed (25). To our knowledge, the Kaasa questionnaire has not been applied to a healthy population (26).

In contrast, the health-related QoL of the study group can readily be compared with that of individuals with other diseases, since the Swedish version of the SIP has been used in studies of rheumatic disorders (1, 2, 42, 43), spinal cord injuries (31, 38), respiratory failure in scoliosis and neuromuscular disorders (37), stroke (34, 35), lung cancer (9), chronic renal insufficiency (11), chronic pain (6), tension headache (16) and osteoarthrosis (32).

The most relevant comparisons may be with data from rheumatoid arthritis and neuromuscular diseases. Two population studies of rheumatoid arthritis (1, 42) showed a psychosocial index (means 7.7, 8.0) similar to that found for MD (mean 7.1), but individuals with rheumatoid arthritis showed a higher QoL with regard to the physical index (means 7.7, 9.8 vs 13.5). A study which included 10 individuals with neuromuscular diseases under home ventilator treatment showed a somewhat poorer QoL with

regard to the psychosocial index (8.4 vs 7.1) and distinctly poorer on the physical index than in MD (19.4 and 13.5 respectively) (37).

In previous studies, SIP data have been compared with results from an age-stratified group of 112 healthy Swedish women (1, 6, 31, 37, 43). Individuals with MD have a poorer QoL compared with the results from this reference population (psychosocial index mean 7.1 vs 5.1 and physical index mean 13.5 vs 3.3).

The QoL results suggest that psychosocial well-being is to some extent affected in a negative direction in MD, at least compared to "healthy individuals". The physical dimensions show distinct values of a poorer QoL. There was an impaired QoL irrespective of the type of MD, according to both tests used. These results reflect the fact that the data were derived from a population survey. A study on MD patients who seek medical care would probably have indicated a higher impact of MD on QoL, i.e. a reduced QoL.

Surprisingly, we found, no differences in QoL between the groups of MD, despite differences in disability. The result was the same irrespective of whether it was measured by the SIP or by the Kaasa questionnaire. Other studies have shown similar results, implying that a chronic disease does not automatically entail a deterioration in the individual's QoL. This fact prompts questions concerning the importance of the individual's coping for perceptions of illness and well-being (8). We intend to work further on these relations in forthcoming papers.

The variables included in the Brooke tests, the ADL staircase and the self-report ADL scale were highly intercorrelated and some of them could be used interchangeably. The results indicate that the newly developed self-report ADL scale is as valid as the functional grading according to Brooke which has turned out to be a standardized procedure with very high intra-rater reliability and low inter-rater variation (21). However, the self-report ADL scale has to be validated in further studies including patients with other diseases.

There are at least three ways of phrasing questions on functional disability. One can ask what a person can do (termed "capacity"), what he does do ("performance") or if he has "difficulty" in doing different activities of daily living. When asked about "capacity", the respondent may exaggerate his health, possibly by as much as 15–20% (36). A problem with "performance" is that there are things other than ill-health that restrict activities. For this reason "performance" questions tend to give lower scores than "capacity" questions. The present self-report ADL scale used the word "difficulty", which is placed between "performance" and "capacity" and therefore might give a more reliable estimation of disability (33).

The ADL staircase could reliably be adapted also to the individuals with MD in the present study, because only 3% of the participants could not be classified in the cumulative staircase. The fact that data were based on interviews only slightly influenced the results. Several ADL instruments have been used and published but only a few have been thoroughly validated and tested for reliability (18, 33). With the exception of the Katz ADL index, which is based on primary biological and psychosocial functions (27, 29, 33), most instruments have been developed empirically without a broader reference to a body of theory. The Katz ADL index has been extensively validated and tested for reliability (13, 27, 28, 29, 40) and has been used in several types of neurological and other diseases (7, 33).

There are only a few self-administered ADL instruments reported in the literature (33). Using the ADL level estimated by occupational therapists as a reference, self-assessed ADL has been validated, and a high degree of consensus was found (39). In the present study, there was a strong convergent validity between the ADL staircase and the variable ambulation (from the self-report ADL scale) as well as the other variables in Factor 1, walk and move. Clinical advantages of the self-report ADL scale are that it is self-administered, it is simple and fast to use, the results are easy to calculate, it focuses on the experienced disabilities and provides disability scores in four categories of activity as well as a summary score (total index).

All data on the different variables were primarily on ordinal scales, not suited for parametric statistical analyses. Using several observations, the results may well behave like interval data, thus justifying that they are analysed as such (22). However, we also performed correlation analyses with non-parametric statistics. With respect to Brooke's examination procedure we aggregated two or three tests for each variable (gait, etc.) and used only four grades of disability; these variables seemed to behave like interval data. The same is true for the self-report

ADL scale. Less suitable for this approximation was Brooke's grading system, which is not based on an aggregation of different tests, and therefore the correlations to this variable should be interpreted with great caution. These limitations must be kept in mind when the results are interpreted. For example, an individual with a disability score of 40% might not be twice as disabled as another individual with a disability score of 20% (17). It is also important to stress that the designations of the different variables and dimensions are based on empirical and clinical evaluations, and future experience may indicate a need for some revision.

The results of the Kaasa questionnaire showed only small variations between the individuals, irrespective of type of MD, and were similar to those found in the case of patients with lung cancer (25, 26). This might be interpreted as meaning that the questionnaire has a low discriminating capacity with regard to the QoL of these individuals (33).

The SIP instrument has been used for assessment of QoL in terms of illness-related dysfunctions based on the individual's self-report behaviour (10). The SIP instrument is used extensively (6, 9, 11, 31, 35, 37, 38) and has a good psychometric quality, and the Swedish version is validated and adjusted for cross-cultural applications (42). Most of the variables of disabilities in the present study were correlated with physical aspects of the SIP and markedly fewer of the psychosocial aspects of the SIP. Another factor that might have an impact on both ADL and QoL is coping, which has been studied with regard to MD's (5).

This is the first study about muscular dystrophy including ADL variables, and further studies of disability and QoL are required to verify its results. There is also need for further development of measuring methods for QoL suitable and valid for muscular dystrophy.

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APPENDIX A

Manual for a standardized observation and grading of muscle function according to Brooke's examination procedure (12).

The same equipment was used in the case of all patients and the tests were administered in the same order. The patients were told to avoid hand support unless they really needed it. The outcome of each test was graded as: 0 = normal, 1 = slight weakness, 2 = moderate weakness, 3 = extreme weakness, 4 = fails to perform the test.

Gait		
Hip:	Normal	0
	Slightly reduced strength in "shock absorbers"	1
	Moderately reduced strength in "shock absorbers"	2
	Extremely reduced strength in "shock absorbers"	3
	Cannot walk	4
Thigh:	Normal	0
	Slight weakness shown by "back kneeing"	1
	Moderate weakness shown by "back kneeing"	2
	Extreme weakness shown by "back kneeing"	3
	Cannot walk	4
Lower leg:	Normal	0
	Slight foot drop	1
	Moderate foot drop	2
	Extreme foot drop	3
	Cannot walk	4
Stepping onto footstool (20 cm)		
Hip:	Normal	0
	Slight weakness shown by hesitation	1
	As the above and/or "hip dip"	2
	As the above and/or throws up the foot	3
	Cannot step up	4
Thigh:	Normal	0
	Brief support from one or both hands	1
	Protracted support from one hand	2
	Protracted support from both hands	3
	Cannot step up	4
Stepping onto chair (45 cm)		
	The same as for footstool	
Rising from armchair (45 cm)		
Hip/thigh:	Normal	0
	Forward lean but no hand support used	1
	Forward lean and brief support from one or both hands	2
	Forward lean and repeated or protracted support from one or both hands	3
	Cannot rise from chair	4
Rising from floor		
	Normal	0
	Brief support from one hand	1
	Brief support from both hands	2
	Repeated or protracted support from one or both hands	3
	Cannot rise from floor	4

APPENDIX B

A modified grading system of muscle function based on Brooke's grading system (12)

Hips and legs*

0. Walks and climbs stairs without assistance.
1. Walks and climbs stairs with aid of railing.
2. Walks and climbs stairs slowly with aid of railing (>25 sec for 8 steps or >3 sec for a single step).
3. Walks unassisted and rises from chair but cannot climb stairs.
4. Walks unassisted but cannot rise from chair or climb stairs.
5. Walks only with assistance or walks independently with long leg braces.
6. Walks in long leg braces but requires assistance for balance.
7. Stands in long leg braces but unable to walk even with assistance.
8. Needs a wheelchair and propels it without difficulty.
9. Needs a wheelchair but manages to propel it only short distances.
10. Needs an electrical wheelchair and drives it without difficulty.
11. Needs a wheelchair but does not even manage to drive an electrical one. Has to be pushed by an assistant.
12. Confined to bed.

Arms and shoulders

0. Starting with the arms at the sides, the patient can abduct the arms in full circle until they touch above the head. Can place a weight of 2 kg or more on a shelf above eye level with one hand.
1. Can raise arms above head as previously but cannot place a 2 kg weight on a shelf.
2. Can raise arms above head only by flexing the elbow or using accessory muscles.
3. Cannot raise hands above head but can raise a glass of water to mouth.
4. Can raise hands to mouth but cannot raise a glass of water to mouth.
5. Cannot raise hands to mouth but can use hands to pick up pennies from table.
6. Cannot raise hands to mout and has no useful function of hands.

Bulbar

0. Normal speech and swallowing.
1. Speech and/or swallowing is abnormal but presents no practical difficulty.
2. Speech is occasionally difficult to understand and/or the swallowing difficulty causes occasional choking (on a daily basis)
3. Speech can be understood by close friends or relatives but it is difficult for casual acquaintances to understand and/or swallowing difficulty is always present and prolongs mealtimes.
4. Speech is impossible to understand even by close friends or swallowing is impossible.

* Step 9, "Is in wheelchair" according to Brooke (12), was replaced with steps 8–11.