

ACTIVITIES OF DAILY LIVING AND QUALITY OF LIFE IN PERSONS WITH MUSCULAR DYSTROPHY

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The study concerns 77 adults with muscular dystrophy (mean age 49 years) in two counties in Sweden. The purpose was to investigate activities of daily living, quality of life and the relationship between these. Data collection was performed with “the Activity of Daily Living Staircase”, “the Self-report Activity of Daily Living” and the Quality of Life Profile. The results indicated that over half of the subjects were dependent on others, chiefly in activities requiring mobility. Muscular dystrophy had mostly negative consequences, and nearly half stated that life would have offered more without it. Few significant diagnosis-related (no gender-related) differences emerged regarding activities of daily living and quality of life. Lower quality of life can only partly be explained by greater disability ($r = 0.30-0.54$). Therefore quality of life as a measurement of rehabilitation outcomes might be based both on physical status, disability and psychosocial factors in terms of positive and negative consequences.

Key words: muscular dystrophy, quality of life, activities of daily living, disability, chronic disease.

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INTRODUCTION

Few studies have focused on the consequences of muscular dystrophy (MD) in adults. MD is a term designating different types of hereditary, primary and incurable muscular diseases characterized by progressive muscular weakness, often with a slow progress. The particular diseases involve different clinical courses depending on whether they primarily affect proximal or distal muscle groups (1, 2). The muscular weakness impedes mobility, often involving loss of the ability to walk and other restrictions concerning daily life and leisure activities. The decline in mobility causes the person to become dependent on the help and support of others and on technical aids (3–5).

During the past two decades, there has been a change from a strict focus on physical health towards quality of life (QoL) as a measure of rehabilitation outcomes (6). In clinical investigation and patient care, QoL is a reflection of the way that patients perceive and react to their health status and to other subjective

experiences perceived as important (7, 8). Being afflicted with a progressive disease has received little attention in research on quality of life. In order to meet this lack of research, the current study focused on persons with MD and investigated what it means in terms of QoL to be afflicted with these progressive diseases over time with repeated losses of activities of daily living.

The instruments available for assessing quality of life are directed towards the person's current life-situation. A particular instrument has been developed (9), and in contrast with earlier instruments, it takes account of the person's evaluation over a long period. The present study had the following purposes: to describe activities of daily living in adults with MD and these persons' experienced QoL, to investigate whether there are any differences depending on gender or specific diagnosis, and to investigate whether ability to perform activities of daily living is a predictor for QoL.

METHODS

The study is a questionnaire inquiry and constitutes part of a research programme on MD (3). The research project has been approved by the research ethics committees of Örebro Medical Centre Hospital and Linköping University Hospital.

Subjects

The subjects were 77 adults with MD, the average age was 49 (range 24–74) years and 61% of them were women. Thirty-eight were living in the county of Örebro (Group A), the other 39 in the adjacent county of Östergötland (Group B), Sweden (Table I). The subjects were divided into three groups in accordance with type of MD (categorized in accordance with clinical course): (1) myotonic dystrophy (MyD) with in the first place distal muscular weakness and with a certain degree of general muscular weakness, (2) myopathia distalis tarda hereditaria (MDTH) which almost exclusively afflicts distal muscle-groups, and (3) Proximal MD, covering different diagnoses involving in the first place proximal muscle groups (Fascioscapulohumeral MD, Becker MD, Limb-girdle MD, Proximal MD without a definite diagnosis) (1, 10).

Group A were recruited from the neurology clinic at Örebro Medical Centre Hospital. All 38 persons were willing to answer the questionnaires and to participate in a rehabilitation programme during the period of the study (11).

Group B, who were in the same age-range and had similar diagnoses, were selected from the file of patients at the Department of Neurology, Linköping University Hospital, in the county of Östergötland. All of the 40 selected were willing to participate, but one became seriously ill, leaving 39 subjects for the study.

Instruments

Group A answered the ADL Staircase and the Quality of Life Profile by way of structured interview questions at hospital and Group B at home. The Self-report ADL was administered as a mail questionnaire.

Table I. Distribution of the study group in two counties and by diagnosis, gender, age, duration of disease

	Total group (n = 77)	Group A (n = 38)	Group B (n = 39)	MyD (n = 34)	MDTH (n = 12)	Proximal MD (n = 31)
Men/women n (%)	30/47 (39/61)	16/22 (42/58)	14/25 (36/64)	11/23 (32/68)	2/10 (17/83)	17/14 (55/45)
Mean age (range)	49 (24–74)	51 (25–71)	47 (24–74)	45 (24–62)	64 (50–71)	48 (25–74)
Disease duration, mean years (range)	23 (4–69)	23 (7–46)	23 (4–69)	22 (8–47)	25 (10–41)	24 (4–69)

MyD = myotonic dystrophy, MDTH = myopathia distalis tarda hereditaria, Proximal MD = proximal muscular dystrophy.

Group A = County of Örebro, Group B = County of Östergötland.

Disability

The ADL Staircase. This instrument assesses independence/dependence in 10 activities of daily living. It is based on the Katz ADL Index, and has six items concerning personal care (P-ADL): bathing, dressing, toileting, transfer, continence and feeding (12). The ADL Staircase comprises four complementary instrumental ADL (I-ADL): cleaning, shopping, transportation and cooking (13,14). ADL performance is ranked according to a manual in cumulative ADL grades (14): Independent, grade 0; Dependent in I-ADL, grades 1–4; Dependent in I- and P-ADL, grades 5–10. In the case of persons to whom none of these grades applies, there is employed the category Other, signifying dependence in two or more activities but not classified as above. A prerequisite for a reliable result is that this category does not exceed 5%. The instrument has been found to have good reliability and validity in respect of several patient-groups (12–14).

The Self-report ADL. This instrument has been developed inductively from interviews with persons with MD (3,4). It comprises 29 items concerning difficulties regarding activities of daily living. In the revised version the subject marks one of the following answers: “No difficulty” or “Can do it but don’t” (0.0), “Sometimes difficulty” (0.33), “Always difficulty” (0.66) and “Fails” (1.0). The results are given in the form of a percentage of the maximum number of points with regard to each of the following indices: Ambulation, Arm strength, Finger strength, Finger subtle function and Total index. The higher the percentage, the greater the difficulty. The instrument has been tested and has been found to be valid (3,4,9).

Quality of life

The Quality of Life Profile. The instrument has been developed inductively (3), is designed for persons with progressive long-term illness, and can be regarded as a health-related quality of life instrument. The self-assessment questionnaire has 44 items, grouped as follows: *Life-picture* (4 items), *Life-areas* (19 items), *Problems* (15 items) and *Acceptance* (6 items). *Life-picture* involves a global assessment of the consequences of the disease. The respondent ticks one of the four items, which best fits his or her situation. *Life-areas* offers four alternative answers: “in a positive direction”, “in a negative direction”, “don’t know in what direction” and “has not been important”. The 19 items concern the following: choice of occupation, educational opportunities, opportunities of gainful employment, marrying and settling down, relationship with spouse or equivalent, sex life, socializing with friends and neighbours, meeting new people, emotional support from other people, choice of dwelling, being or becoming a parent, bearing responsibility for family and relations, standard of living, being free and independent of others, extent of worries and problems, being able to make decisions and be in control of my everyday life, personal goals and ambitions, leisure activities, everyday freedom of movement. *Problems* has items concerning mobility, fatigue, pain, sleep and leisure. *Acceptance* has six items concerning how the person and those closest to the person accept the restrictions caused by the disease. In the case of both *Problems* and *Acceptance* the person can tick two or more items if this corresponds to his or her situation. *Life-areas*, *Problems* and *Acceptance* have been assigned both a positive and a negative index. The indices represent the sum of the responses “in a positive direction” or “in a negative direction”.

The instrument has been developed under the inspiration of Nordenfelt’s theory (15) of people’s vital goals and quality of life.

The items are based on 120 interviews with adults with MD (3). Furthermore, items included under *Problems* are based on the pattern of answers in a number of studies of MD where the Sickness Impact Profile (16) has been applied over a period of 5 years (17). The instrument was tested in a study about persons with post-polio syndrome, and has been found to have an acceptable discriminatory validity (9).

Data processing

In addition to the use of descriptive statistics, gender and group-related differences have been analysed by means of unpaired *t*-test (the Self-report ADL and the indices of the Quality of Life Profile) and the Mann-Whitney U test (the ADL Staircase). In order to compare the three diagnosis groups, Analyses of Variance (ANOVA) (the Self-report ADL and the indices of Quality of Life Profile) and the Kruskal-Wallis test (the ADL Staircase) were used. Correlations have been studied by means of Spearman’s correlation coefficient for ranked data (ρ) (nominal/ordinal data) and the Pearson product-moment correlation coefficient (r) (interval data).

RESULTS

Comparisons between the two county groups showed few differences. For this reason the groups have been merged ($n = 77$) for the presentation of the results. The significant differences found between the groups and also for gender are presented in the text.

The ADL Staircase. The results indicated that 52% (40/77) of the subjects were dependent in I-ADL (grades 1–4), and 18% (14/77) in both I-ADL and P-ADL (grades 5–10). Concerning the three diagnosis groups, the MyD group were significantly more dependent than the MDTH group ($z = 2.34, p < 0.05$). The degree of dependency on others differed significantly ($z = 2.71, p < 0.01$) between Group B (85% or 33/39) and Group A (58% or 22/38). One person was not classifiable according to the ADL Staircase.

The Self-report ADL. This showed that the MDTH group had the greatest disability concerning activities requiring distal muscular strength. The Proximal MD group had the greatest disability for *Ambulation*, while the results for the MyD group reflect both proximal and distal muscular weakness (Table II). The MyD and MDTH groups had significantly more difficulties with regard to *Finger strength* than the Proximal MD group (MyD/Proximal MD $t = 3.13, df = 63, p < 0.01$; MDTH/Proximal MD $t = 2.91, df = 41, p < 0.01$). With regard to *Finger subtle function* the greatest disability was to be found in the MDTH group (MDTH/Proximal MD $t = 8.03, df = 41, p < 0.001$; MDTH/MyD $t = 2.98, df = 44, p < 0.01$), whilst there was greater disability in the MyD group than in the

Table II. Disability assessed by the Self-report ADL with regard to type of muscular dystrophy

The Self-report ADL	Total group (n = 77) Score % Mean (S.D.)	MyD (n = 34) Score % Mean (S.D.)	MDTH (n = 12) Score % Mean (S.D.)	Proximal MD (n = 31) Score % Mean (S.D.)
Ambulation	41.9 (30.8)	41.2 (33.6)	29.5 (20.3)	47.5 (30.3)
Arm strength	33.4 (27.2)	30.5 (32.1)	42.9 (20.9)	32.8 (23.0)
Finger strength	40.1 (26.2)	47.8 (28.6)	49.1 (24.3)	28.4 (19.6)
Finger subtle function	18.1 (20.1)	19.7 (22.2)	39.7 (10.6)	7.9 (12.0)
Total index	33.8 (22.8)	34.5 (27.2)	38.6 (15.9)	31.1 (19.8)

MyD = myotonic dystrophy, MDTH = myopathia distalis tarda hereditaria, Proximal MD = proximal muscular dystrophy.

Proximal MD group (MyD/Proximal MD $t = 2.63$, $df = 63$, $p < 0.05$). In the case of the MyD group this disability can be attributed to the myotonia that afflicts the hand-muscles.

The Quality of Life Profile. Concerning *Life-picture*, the statement which drew the largest proportion of affirmative responses (44%, 31/71) was "Life would have had more to offer if the disease hadn't got in the way, but I don't go around thinking about it". Regarding the item "Life hasn't become worse because of the disease, I've a good life" the proportion of affirmative responses was 35% (25/71). Eleven per cent (8/71) gave an affirmative response to the statement "Of course I'm disappointed, since the disease hampers me", while 10% (7/71) gave an affirmative response to the statement "I've developed as a person: I probably wouldn't have had the personal strength I do have if I'd been in perfect health". Concerning *Life-picture* there were no significant differences related to diagnosis, gender or county groups.

The results regarding *Life-areas* indicate that the subjects regarded the disease as having had few positive consequences. The commonest concerned "Emotional support from other people" (18% or 13/71) and "Choice of dwelling" (16% or 12/71). The most common negative consequences concerned "Everyday freedom of movement" (67% or 47/70), "Leisure activities" (60% or 42/70), "Opportunities for employment on the labour market" (35% or 25/71), "Extent of worries and problems in my life" (34% or 24/70) and "Being free and independent of others" (31% or 22/70). About a quarter of the subjects indicate a negative effect on "Financial standard of living" (29% or 21/71), "Personal goals and ambitions" (26% or 18/70) and "Being able to make decisions and be in control of my everyday life" (24% or 17/71). The subjects' uncertainty is greatest when it comes to "Emotional support from other people" and "Personal goals and ambitions" (both 20% or 14/71).

The results regarding *Problems* confirm the effect of the disease on mobility. No fewer than 70% (50/71) gave an affirmative response to the statement "I don't move around as much as other people, giving priority instead to what I most want to do or have to do". Slightly more than half (54% or 38/71) gave an affirmative response to the statement "I can't keep up the pace I used to, but it doesn't affect my everyday life to any real extent".

Concerning *Acceptance*, nearly half gave an affirmative response to the statements "I've entirely accepted the restrictions caused by the disease" and "I have to some extent accepted the restrictions caused by the disease" (Table III).

Again, from Table IV it can be seen that the most usual consequences of the disease were negative. There is a significant difference with regard to *Problems positive*, where a greater number of positive consequences were indicated by the MDTH group than by the other two diagnosis groups (MDTH/MyD $t = 2.8$, $df = 39$, $p < 0.01$; MDTH/Proximal MD $t = 2.7$, $df = 39$, $p < 0.05$). Regarding *Problems negative* Group B scored significantly higher than Group A ($t = 3.37$, $df = 69$, $p < 0.01$). The results indicate no other significant diagnosis- or county-related differences, and no gender-related ones at all.

Correlations between ADL and quality of life. Both the total group ($n = 77$) and the three diagnosis groups separately gave 3 times as many negative as positive responses. This irrespective of degree of dependence according to the ADL staircase. There was a weak to moderate correlation between on the one hand *Life-areas negative* ($\rho = 0.30$, $p < 0.05$) and *Problems negative* ($\rho = 0.51$, $p < 0.001$), on the other hand the *ADL staircase*. There was also a correlation between these two indices of the Quality of Life Profile and all four indices of the Self-report ADL. The results indicated a moderate correlation between *Life-areas negative* and both *Ambulation* ($r = 0.40$, $p < 0.01$) and *Arm strength* ($r = 0.42$, $p < 0.001$), and a somewhat weaker one between *Life-areas negative* and both *Finger strength* ($r = 0.35$, $p < 0.01$) and *Finger subtle function* ($r = 0.33$, $p < 0.01$). Further, there was a moderate correlation between *Problems negative* and *Ambulation* ($r = 0.52$, $p < 0.001$), *Arm strength* ($r = 0.54$, $p < 0.001$), *Finger strength* ($r = 0.50$, $p < 0.001$) and *Finger subtle function* ($r = 0.53$, $p < 0.001$).

DISCUSSION

The results indicated that muscular dystrophy has principally negative consequences for the subjects. Earlier studies have shown, similarly, that persons with MD are confronted with many disease-related problems in everyday life, most of these problems being caused by muscular weakness (3, 18, 19–22). Increased dependence on others and difficulty in performing ADL were found in the present study to have no more than a

Table III. Descriptive data on the Acceptance of the Quality of Life Profile in the study group

Acceptance	Number of agreement answers			
	Total group (n = 70) n (%)	MyD (n = 30) n (%)	MDTH (n = 11) n (%)	Proximal MD (n = 29) n (%)
1. My disease has not involved any restrictions	7 (10)	5 (17)	1 (9)	1 (3)
2. I have not yet accepted the restrictions caused by the disease	10 (14)	2 (7)	4 (36)	4 (14)
3. I have to some extent accepted the restrictions caused by the disease	29 (41)	9 (30)	6 (55)	14 (48)
4. I have entirely accepted the restrictions caused by the disease	31 (44)	16 (53)	1 (9)	14 (48)
5. I accept being restricted by the disease, but my closest relatives have not	6 (8)	6 (20)	0	0
6. My friends and workmates have not accepted the fact that I have certain restrictions due to my disease	6 (8)	3 (10)	1 (9)	2 (7)

MyD = myotonic dystrophy, MDTH = myopathia distalis tarda hereditaria, Proximal MD = proximal muscular dystrophy.

weak to moderate correlation with lower quality of life. This implies that a decline in quality of life can only partly be explained by a decline in ADL-ability, a finding that is in line with a recent study of persons with post-polio syndrome (9) and with previous research on MD (3, 4).

We have been unable to find in the literature any quality of life instrument specifically designed to assess what it is like to live with a progressive disease. The Quality of Life Profile measures positive consequences of the disease, and this feature is based on what persons with MD have said in the interviews (3). The present study showed comparatively few positive consequences of MD. The commonest concerned emotional support from other people, as was also found in the previous study (9). The MDTH group indicated the largest number of positive consequences, an explanation is that these persons have the least disability with regard to mobility and few were dependent on others in ADL. Some previous studies have focused on positive consequences of the disease (23–25). In the studies concerning the mobility-disabled approximately half of groups mentioned such advantages as challenge, goal or purpose, and the subjects had become more sensitive, tolerant and patient as well as living a less hectic life and having more contact with other people (23, 24). The explanation of the differences between the results of these studies and those of the present study may be the divergent

samples and methods. Further research is needed to focus on positive consequences in order to understand how they contribute to the overall quality of life associated with disability.

A third of the subjects indicated that their lives had not become worse because of the disease. One in 10 felt that they had developed as persons, the trials and tribulations caused by the disease having given them a personal strength, which they probably would not have had if they had been in good health. One in four of the persons with post-polio syndrome felt this way (9). These positive experiences may be attributable to the fact that the persons have had the disease for such a long time. There occurs a gradual adaptation to the change in the conditions of one's life (26).

Several studies have pointed to the subjective experience of having a high quality of life, irrespective of physical state of health (7, 27, 28). To some extent subjects may seek to present their life in a positive light in interviews and questionnaires (29). This means that in the interpretation of self-report data it must be borne in mind that such data can be imbued with a certain over-estimation of quality of life (27, 29).

It emerges that persons with MD are more dependent in ADL than the average elderly person in Sweden (14). Rather more than half of our subjects needed help in one or more I-ADL, whilst the proportion in the other study was 22% (14). Again,

Table IV. Indices of the Quality of Life Profile in the study group

Indices	(n answers)	Total group (n = 71) Mean (S.D.)	MyD (n = 30) Mean (S.D.)	MDTH (n = 11) Mean (S.D.)	Proximal MD (n = 30) Mean (S.D.)
Life-areas positive	(0–19)	1.2 (2.1)	1.5 (2.4)	0.9 (1.3)	0.9 (2.0)
Life-areas negative	(0–19)	4.6 (3.6)	4.4 (3.5)	4.5 (3.2)	4.8 (3.8)
Problems positive	(0–2)	0.8 (0.8)	0.7 (0.8)	1.4 (0.8)	0.6 (0.8)
Problems negative	(0–13)	3.8 (2.0)	4.2 (2.1)	4.1 (1.5)	3.2 (1.9)
Acceptance positive	(0–3)	0.9 (0.5)	1.0 (0.6)	0.7 (0.6)	1.0 (0.4)
Acceptance negative	(0–3)	0.3 (0.6)	0.4 (0.7)	0.4 (0.5)	0.2 (0.5)

MyD = myotonic dystrophy, MDTH = myopathia distalis tarda hereditaria, Proximal MD = proximal muscular dystrophy.

this comparison indicates that MD gives rise to a large number of difficulties in daily life, principally concerning mobility, personal care and transportation (3–5, 17, 30). Group B was significantly more dependent in ADL than Group A, and had a significantly greater number of problems experienced as negative. A possible explanation of these differences is that Group A were identified by way of a population study from 5 years before (2), whereby this group included rather more persons with a milder MD progress than in the case of Group B (who were only consecutive patients). Another possible explanation is that Group A had participated in a programme of recurrent rehabilitation during the past 18 months (11). Only a few persons in Group B had undergone rehabilitation within the conventional framework during this period. An explanatory factor may be that Group A had technical aids better adapted to their particular needs.

In the present study, almost half responded affirmatively to the statement that life would have had more to offer if the disease had not got in the way, while in the case of the post-polio group the proportion was somewhat lower (9). The persons experience difficulties attributable to their restricted freedom of movement, both in everyday life and with regard to leisure activities they formerly engaged in. The results also indicated that rather fewer than half of the subjects had arrived at complete acceptance, and that about the same number had arrived at partial acceptance. The majority of the subjects have had the disease for more than 20 years, and the results indicated that the temporal aspect could not alone explain acceptance/non-acceptance.

Research has indicated that the progressive course of the disease involves significant deterioration of functional capacity over a 5-year period (30). Similar results have emerged from Californian studies based on a 10-year period (19–22). Progressive functional impairment involves the loss of one important ability after another and demands renewed adaptation on the part of the person afflicted. Knowledge of the greater vulnerability of people who have experienced repeated losses has an important role to play in the professional encounter with the patient in respect of care and rehabilitation (31–33). Staff need to encounter patients in a spirit of openness and with sensitivity, bearing in mind what it is like for them to live with newly occurring functional impairment (11, 31–33). They should give coping support to patients who express helplessness, hopelessness and anxious preoccupation with the problems, such states of mind having in an earlier study found to be related to a decline in quality of life (18).

One person in three, in both county groups, indicated that the disease had reduced the chances of obtaining gainful employment. In an earlier study it emerged that rather more than half of the persons with progressive muscular dystrophy who were unemployed did not want a job (either a new one or the old one) (34). A possible explanation is that many jobs call for muscular strength, whilst the possibilities of workplace adjustment are often greatly restricted (35). In the case of persons with MyD a further possible explanation of reduced working capacity and

unemployment is cognitive impairment (36–38). Another thing is that the majority found it difficult to ask fellow-workers for help when there was some part of the job which they were unable to manage (11).

The three instruments used in this study have been tested and validated in earlier studies. The Quality of Life Profile can be regarded as a health-related instrument. Except in the case of the index constituting a summary of the numbers of answers, the answers of the Quality of Life Profile are on a low scale-level. This limits the possibilities of psychometric testing. Earlier research has demonstrated an acceptable discriminatory validity (9). Empirical validity is important when it comes to determining whether the instrument achieves an acceptable validity (13). Since the results of the present study do not disagree with earlier research results, we judge the empirical validity to be satisfactory. The instrument has a comparatively restricted range of application in that it presupposes that the persons got the disease at an early age and had lived with it for a long time. In future research, there is a need of other studies of appropriate patients rather than ones based on a normal population, in order to obtain data for comparison. Up to now there has been just one study of this type (9).

In conclusion, this study indicated that ADL would not be a good predictor for quality of life. Instead, measurement of rehabilitation outcomes in terms of quality of life should take a multidimensional view of physical status and disability, psychological status and well-being, social interactions and economic status (7). Another implication of this and also of previous results was that rehabilitation for persons with MD should be recurrent, provided by a multidisciplinary team and be focused on both negative and positive consequences for the subjects. Knowledge of patients' own perspective on their quality of life is of fundamental importance concerning rehabilitation for persons with MD.

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