

ORIGINAL REPORT

## MYOTONIC DYSTROPHY: THE BURDEN FOR PATIENTS AND THEIR PARTNERS

Reinier Timman, PhD<sup>1,2</sup>, Aad Tibben, PhD<sup>2</sup> and Axel R. Wintzen, MD, PhD<sup>3</sup>

From the <sup>1</sup>Department of Medical Psychology and Psychotherapy, Erasmus University Medical Centre Rotterdam, <sup>2</sup>Department of Human and Clinical Genetics, Leiden University Medical Centre and <sup>3</sup>Department of Neurology, Leiden University Medical Centre, Leiden, The Netherlands

**Objective:** Dystrophia myotonica is characterized by progressive muscular weakness, myotonia, mental slowness and lack of initiative, which causes problems in daily life both for patients and for their spouses. Some couples seem to deal with these problems satisfactorily, while for others they are quite burdensome. The aim of this study was to describe the relationship of severity of dystrophia myotonica and psychological wellbeing in patients and partners.

**Methods:** Sixty-nine couples, in whom one partner had dystrophia myotonica, completed questionnaires on severity of dystrophia myotonica, marital satisfaction, anxiety and depression (Hospital Anxiety and Depression Scale), hopelessness (Beck Hopelessness Scale) and general psychological health (General Health Questionnaire-12).

**Results:** For patients, a worse view of the future, worse general wellbeing, more anxiety and more depression was associated with a greater need for help. For partners, worse general wellbeing and more anxiety was associated with a lack of initiative of the patient and less marital satisfaction. It is noteworthy that 40% of patients and particularly female partners had Beck Hopelessness Scale scores suggestive of clinically relevant depression.

**Conclusion:** Dystrophia myotonica places a heavy burden on patients, and especially on female partners. The need for help and dependency has more influence on the wellbeing of patients than the symptoms of dystrophia myotonica themselves. Marital satisfaction is a strong predictor of better wellbeing, both for patients and, even more so, for partners.

**Key words:** dystrophia myotonica; myotonic dystrophy; burden of disease; familial caregivers; psychological wellbeing.

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Correspondence address: Reinier Timman, Erasmus University Medical Centre Rotterdam – Medical Psychology and Psychotherapy, PO Box 2040, NL-300 CA, Rotterdam, The Netherlands. E-mail: r.timman@erasmusmc.nl

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### INTRODUCTION

Myotonic dystrophy, or dystrophia myotonica (DM), the most common hereditary dystrophic neuromuscular disorder, is characterized primarily by the occurrence of muscle weakness

and myotonia (1). Other common physical symptoms include cataracts, cardiac arrhythmia, male infertility, irritable bowel syndrome and hair loss. Symptoms vary greatly with regard to degree of severity and age of onset (2). As weakness often concerns facial and oropharyngeal muscles, a seemingly emotionless facial expression and dysarthria may interfere with social interaction. Deterioration over time is observed in activities of daily living that require finger and arm strength (3).

Cerebral symptoms associated with DM are excessive sleepiness and loss of initiative (4). Patients with DM have increasing problems in commencing activities, and they tend to postpone actions. Loss of initiative is related to mental slowness, not to muscle weakness (5).

It has been suggested that DM is associated with specific personality traits. Personality profiles of patients with DM are not abnormal, compared with healthy controls, but they seem to be abnormally homogeneous. The specific DM personality can be characterized by obsessive-compulsive behaviour, rigidity, stubbornness and avoidance (6). Sistiaga et al. (7) observed that aggressive/sadistic and paranoid traits occurred more often in patients with DM than in control subjects. Other personality characteristics more common in patients with DM were harm avoidance, persistence, self-directedness and cooperativeness (8). Patients with DM did not present significant depressive or anxious symptomatology, but rather an emotional deficit, which may be explained by an adaptive reaction to the threatening implications of the disease (9). Health-related quality of life is severely impaired in DM, depending on the severity and duration of the disease (10).

Given the broad variety of symptoms, DM is expected to cause many different problems in patients' daily lives. Clinical experience shows that patients may have many different ways of coping with these problems, although relatively small influence has been observed with regard to psychosocial dysfunction as assessed with the Sickness Impact Profile (3).

The impact of the progressive disease on the familial caregiver may be considerable. The patients' loss of initiative and apathy may lead to alienation between spouses and induce feelings of loneliness in partners. Additionally, physical deterioration of the patient can cause a decline in financial income and a gradual and steady increase in caring burden. This can result in worse general well-being and a depressing or frightening view of the future.

The aim of this study was to examine the impact of DM on patients and their partners. We hypothesized, firstly, that the psychological wellbeing of patients and their spouses declines with the severity of the disease; and, secondly, that marital satisfaction is associated with better psychological wellbeing. The final objective was to determine whether the burden of DM is different for men and women in both patients and partners.

## MATERIAL AND METHODS

### Participants

Eligible participants were adults with either a clinical or a genetic diagnosis of DM, who had an intimate relationship at the time of the study. Members of the Dutch Patients Association for DM were informed about the study by means of notification in their annual magazine and were invited to participate. At the time of the mailing, the association had 755 members. Eighty-two couples responded to the invitation to join the study. Most couples visited the outpatient clinic to participate in the study, while 14 couples were visited by the researcher in their homes because they were unable to travel. Patients and partners completed the questionnaires separately.

The inclusion criteria were: the ability to understand the Dutch language and to complete the questionnaires. Thirteen patients were excluded from the study, one because the diagnosis of DM was withdrawn, one patient was illiterate with insufficient knowledge of the Dutch language, the other excluded patients did not have a stable relationship, or the spouse did not give permission to cooperate. There was not a specific duration set for classifying a relationship as stable. Sixty-nine couples met the inclusion criteria. The study was approved by the medical ethics committee of Leiden University Medical Centre. All participants gave informed consent prior to inclusion.

### Data

**Demographics.** Information was gathered regarding sex, marital status, age, education level, employment, children, and duration of marital relationship.

Data on the general severity of DM were collected using a self-completed questionnaire that was constructed according to a list of symptoms formulated by the clinical neurological experience of one of the authors (ARW). This questionnaire comprised:

- age at onset and age at diagnosis, duration of the disease (years elapsed since first symptoms);
- 29 questions on DM symptoms (Appendix I);
- DM-related limitations on performing daily activities, 26 items, theoretical range 0–26 (Appendix II);
- need for help (a list of 6 activities of daily living in which the patient needs no, little or complete help), range 0–6 (Appendix III);
- use of walking aids, such as a manual or electric wheelchair.

Marital satisfaction was measured in two ways: (i) on a visual analogue scale (VAS) (11), participants were asked to rate the quality of their relation by marking a 100-mm line, ranging from “the worst conceivable marriage” and to “the best conceivable marriage”. The scores were determined by measuring the distance in millimetres from the left; (ii) by giving a number between zero (worst conceivable marriage) and 10 (best conceivable marriage). In addition, the hypothetical quality of the marriage if DM had not affected their lives was rated similarly. In the same way they were requested to rate the quality of the relationship at the beginning of the marriage retrospectively. Patients and partners completed questionnaires separately.

### Psychological outcome variables

The burden of disease was operationalized with 3 questionnaires, measuring general well-being, anxiety and depression, and future perspective.

**General psychological health: General Health Questionnaire-12 (GHQ-12).** The GHQ-12 (12) does not provide a precise psychiatric diagnosis, but identifies individuals with a probable mental health problem. A low score indicates a good general wellbeing. Overall, the GHQ-12 has been extensively validated (13) and is found to be a reliable instrument when used in a general population with relatively long intervals between applications (14). The GHQ-12 has a range of 0–12. A cut-off score for the Dutch version of the GHQ-12 was determined by Hoeymans (15) at  $\geq 2$ .

**Anxiety and depression. Hospital Anxiety and Depression Scale (HADS).** The HADS (16) is a 14-item questionnaire. Seven items measure anxiety and 7 measure depression. Both subscales range theoretically from 0 to 21. In a reliability study of the Dutch version conducted by Spinhoven et al. (17) no cut-off score was calculated for the Dutch version. For the English version Honarmand (18) and for the Norwegian version Olsson (19) suggest a cut-off score of  $\geq 8$ . We adopted these cut-off scores for the Dutch version.

**Future perspective.** The Beck Hopelessness Scale (BHS) (20) measures pessimism and future expectations. The BHS consists of 20 “true” or “false” items (score range 0–20). A score of 9 or more predicted suicidal behaviour in psychiatric inpatients (21).

### Statistics

Differences in continuous outcome variables between groups were analysed with paired sample *t*-tests and multivariate analysis of variance. Intraclass correlation coefficients (ICC) were calculated for differences between patients and partners scores on DM severity and marital satisfaction. A principal component analysis (PCA, Appendix I) with a varimax rotation was performed on the 29 DM severity items. Wellbeing measures, BHS, GHQ, HADS anxiety and depression were entered as dependent variables in repeated measures analyses of covariance. Partners were defined as the repeated measures of the patients. Gender, age, years since first complaints, principal components of the 29 questions on DM symptoms, weakness, limitations on performing activities, needing help, years of healthy relationship and marital satisfaction were entered as covariates. In order to avoid multi-collinearity, for covariates that had significant ICCs between patients and partners, the mean of patient’s and partner’s scores was used. For covariates that did not have a significant ICC between patient and partner, the patient scores as well as the partner scores were entered. Insignificant covariates (*p*-out > 0.10) were removed backwardly from the analyses. Outcome variables and covariates were z-transformed to obtain standardized regression weights. Statistical analyses were performed with SPSS-16 (Chicago, IL, USA).

## RESULTS

### Demographics

Sixty-nine couples participated in the study. One couple comprised two women. The mean duration of the relationships was 22.9 (standard deviation (SD)=9.7) years (Table I). At the time of the study 38% of the patients and 70% of the partners were employed, whereas 41% of the patients and 9% of the partners received some form of disability pension. Seventy-eight percent of the couples reported having one or more children, and in 54% of the families children were living at the patient’s home.

### Severity of DM

The mean period that had elapsed after onset of DM was 19.5 (SD=9.4) years. Patients and partners often disagreed on the

Table I. Biographic and general characteristics of patients with myotonic dystrophy (MD) and their partners

Characteristics <sup>1</sup>	Patients		Partners	
	Men (n=34)	Women (n=35)	Men (n=34)	Women (n=35)
Age (years), mean (SD) [range]	47.5 (7.7) [31–63]	43.6 (8.7) [28–58]	47.6 (10.2) [29–70]	45.2 (7.4) [30–59]
Duration of MD complaints, mean (SD) [range]	19.4 (9.5) [1–44]	19.5 (9.5) [4–42]		
Duration since diagnosis, mean (SD) [range]	13.0 (8.4) [2–45]	12.5 (8.2) [0–31]		
Relationship duration, mean (SD) [range]	24.7 (10.1) [8–43]	20.9 (9.2) [6–37]		
Pre-morbid relationship duration mean (SD) [range]	8.5 (9.4) [0–32]	5.8 (7.4) [0–26]		
Education more than high school <sup>2</sup> , n (%)	11 (33)	8 (23)	18 (55)	12 (35)
One or more children <sup>3</sup> , n (%)	28 (82)	25 (71)	25 (74)	29 (83)
Children living at parents' home <sup>3</sup> , n (%)	20 (59)	17 (49)	18 (53)	20 (57)
Working status,				
Working full-time <sup>4</sup>	8 (24)	0 (0)	28 (82)	4 (11)
Working part-time <sup>5</sup>	9 (27)	9 (26)	2 (6)	14 (40)
Unemployed	0 (0)	9 (26)	0 (0)	1 (3)
Disability pension <sup>4</sup>	15 (44)	13 (37)	1 (3)	5 (14)
Retired	1 (3)	0 (0)	2 (6)	0 (0)
Other <sup>5</sup>	1 (3)	0 (0)	1 (3)	10 (29)
Unknown	0 (0)	4 (11)	0 (0)	1 (3)
Adjustments at work, n (%)				
No adjustments	9 (27)	6 (18)		
Shorter hours	5 (15)	2 (6)		
More help from co-workers	1 (3)	1 (3)		
Special furniture	1 (3)	0 (0)		
Other adjustment	2 (6)	2 (6)		
More than one of the above	3 (9)	2 (6)		
Not applicable	13 (38)	20 (61)		

<sup>1</sup>One lesbian couple is included.

<sup>2</sup>Partners more often have an education above high school level,  $p=0.05$ .

<sup>3</sup>Discrepancies are caused by children from earlier marriages.

<sup>4</sup>Patient – partner  $p<0.001$ .

<sup>5</sup>Patient – partner  $p<0.05$ .

SD: standard deviation.

time of onset (intraclass correlation coefficient, ICC=0.56), although there was no consequent under- or over-reporting of patients or partners. The mean period that had elapsed since DM had been diagnosed was 12.8 years (SD=8.2) (ICC=0.95). Principal component analysis on the 29 DM symptom items resulted in 4 components. These components can be interpreted as: (i) weakness (ICC=0.79); (ii) lack of initiative and non-verbal expression (ICC=0.63); (iii) stiffness (ICC=0.61); and (iv) gastro-intestinal/weakness upper legs (ICC=0.56). The total explained variance was 36%.

DM-related limitations were experienced in half of the daily activities (Appendix II) (ICC = 0.83). Average “need for help” was 3.7 of the 6 activities of daily living (ICC=0.89). A manual wheelchair was used by 33.8% of the patients, an electric wheelchair by 32.4%, and 26.5% used both a manual and an electric wheelchair.

#### Psychological wellbeing

Mean scores on BHS, GHQ, HADS depression and anxiety of both patients and partners by sex are listed in Table II, and the significances of the differences are given in Table III. As the distributions of these outcome measures were skewed, these were root transformed in order to obtain a normal distribution. Patients and female partners had low expectations for the future. More specifically, 40–41% of the patients and 37% of the female part-

ners scored in the clinical range for the BHS. This is in contrast with the male partners, of whom only 9% scored in the clinical range. Female partners reported a much lower general wellbeing (higher GHQ scores), than male partners and patients. Male partners were less depressed than female partners (Table II).

Table II. Psychological wellbeing and marital satisfaction of patients with myotonic dystrophy and their partners by sex

	Patients		Partners	
	Men n=34	Women n=35	Men n=34	Women n=35
	Mean (SD)	Mean (SD)	Mean (SD)	Mean (SD)
>cut-off	>cut-off	>cut-off	>cut-off	>cut-off
	n (%)	n (%)	n (%)	n (%)
BHS	9.1 (4.9)	7.3 (3.8)	4.7 (2.9)	8.3 (4.2)
	14 (41%)	14 (40%)	3 (9%)	13 (37%)
GHQ	1.6 (2.0)	1.7 (2.9)	1.4 (1.9)	3.9 (3.7)
	13 (29%)	13 (37%)	11 (32%)	20 (57%)
HADS – anxiety	4.1 (3.3)	4.6 (3.5)	3.7 (3.1)	6.9 (5.1)
	5 (15%)	6 (17%)	6 (18%)	12 (34%)
HADS – depression	5.0 (3.6)	4.0 (3.4)	2.9 (2.8)	5.0 (4.2)
	4 (12%)	6 (17%)	3 (9%)	8 (23%)
Marital satisfaction	7.2 (1.5)	8.0 (1.3)	7.4 (1.3)	7.0 (1.9)

Significances are shown in Table III.

BHS: Beck Hopelessness Scale; GHQ: General Health Questionnaire; HADS: Hospital Anxiety and Depression Scale.

Table III. Standardized regression weights and p-values of repeated measures analysis of covariance of patients and partners on psychological outcomes

	Patients		Partners	
	$\beta$	<i>p</i>	$\beta$	<i>p</i>
BHS – future perspective				
Gender partner*	0.17	0.16	0.42	<0.001
Need for help	0.36	0.002	0.14	0.20
Marital satisfaction	-0.25	0.04	-0.22	0.06
GHQ – general wellbeing				
Gender partner*	0.01	0.92	0.25	0.03
Need for help	0.28	0.03	-0.01	0.93
Lack of initiative component	0.10	0.44	0.29	0.01
Marital satisfaction	0.04	0.73	-0.28	0.02
HADS Anxiety				
Gender partner*	-0.11	0.39	0.22	0.07
Need for help	0.38	0.002	-0.05	0.68
Lack of initiative component	0.12	0.35	0.25	0.04
Marital satisfaction	-0.03	0.79	-0.24	0.04
HADS Depression				
Gender partner*	0.18	0.14	0.24	0.05
Length of pre-morbid relationship	0.28	0.02	0.00	0.98
Need for help	0.39	0.001	0.15	0.19
Marital satisfaction	-0.10	0.40	-0.34	0.004

\*Gender partner: male=0, female=1.

Explained variance BHS 0.23; GHQ 0.27; Anxiety 0.29; Depression 0.23.

BHS: Beck Hopelessness Scale, range 0–20, higher scores more hopeless; GHQ: General Health Questionnaire, range 0–12, higher scores less wellbeing; HADS Anxiety: Hospital Anxiety and Depression Scale, range 0–21, higher scores more anxiety; HADS Depression: Hospital Anxiety and Depression Scale, range 0–21, higher scores more depression.

Need for help, range 0–6.

Marital satisfaction, range 0–10.

#### Psychological outcome and disease severity

The first hypothesis was that the psychological wellbeing of patients and their spouses declines with the severity of the disease. This was confirmed as follows. For patients, the need for help was associated with a worse view of the future, worse general wellbeing, more anxiety and more depression. For partners, a lack of initiative and non-verbal expression was significantly related to a worse general wellbeing and more anxiety (Table III).

#### Marital satisfaction

Fifteen participants (11%) did not administer a score on the VAS scale, two participants did not rate the 0–10 scale. The correlation between the two measures was 0.73. We calculated a weighted mean of these measures in order to compensate for missing values. This weighted mean resulted theoretically in a score between 0 and 10. Patients as well as partners rated their marital relationship as satisfactory (mean 7.4). They also agreed reasonably well on marital satisfaction, ICC=0.69. Female patients were more satisfied than male patients (8.0 vs 7.2,  $t_{(65)} = 2.49$ ;  $p = 0.02$ , not in Table II), patients were more satisfied than their partners (7.6 vs 7.2,  $t_{(66)} = 2.13$ ;  $p = 0.04$ ).

Participants reported a loss of quality of their marriages since the beginning of their relationships (7.4 vs 8.1,  $t_{(135)} = 3.19$ ;

$p = 0.002$ ). This also held true for the quality of the marital relationship, comparing present quality with the hypothetical quality without DM (7.5 vs 8.1,  $t_{(123)} = 5.20$ ;  $p < 0.001$ ). There was no significant difference between estimated quality of the relationship in the past and the hypothetical relationship at present without DM.

It was further hypothesized that marital satisfaction was positively associated with psychological wellbeing. In patients it was confirmed that marital satisfaction was associated with a better view of the future, for partners this effect did not reach significance. In partners it was confirmed that marital satisfaction was associated with better general wellbeing, and less anxiety and depression. The analyses showed that a longer pre-morbid relationship was associated with more depression (HADS) in patients.

## DISCUSSION

MD is a severe disorder that inevitably affects the patient's partner. As the disease progresses, patients become increasingly dependent on their partners, who take on the role of caregiver. Substantial subgroups in our study showed anxiety and depression levels in the clinical range, which is in line with the findings of Bungener et al. (9). Bungener et al. reported higher depression and anxiety scores for patients with DM than for controls. Using Welch's *t*-test (22), these differences were significant at  $p < 0.05$  level. This observation may be explained by the increasing severity of the disease, which confirms our first hypothesis that the psychological wellbeing of patients and their spouses declines with the severity of the disease. This finding is in accordance with the findings of Antonini et al. (10), who observed that disease duration was negatively correlated with both physical and emotional areas of the SF-36, and these were inversely related to depression and anxiety. The final regression models showed that not so much the specific symptoms, but the impact, in particular the patients' increasing need for help, was positively related with all psychological outcomes, future perspective, general wellbeing, anxiety and depression.

Interestingly, the observation that the need for help was strongly associated with the psychological outcome measures implies that the consequences of specific DM symptoms are more burdening than the disease itself. Most stressful are the consequences of DM, the dependency on others for activities of daily living, such as personal hygiene, light and more arduous household tasks, cooking, shopping and mobility outdoors. This is in accordance with the findings of Gagnon et al. (23), who found that performing major household tasks was reported by the patients as the life habit that was most severely disrupted by DM. Our findings may also support the report of Boström & Ahlström, who found that the most obvious deterioration over time was in activities of daily living that require finger and arm strength. The influence of DM with regard to psychosocial dysfunction was relatively small (3).

Partners, however, do suffer from the lack of initiative which has also been found in studies on caregivers of patients with Alzheimer's disease and fronto-temporal disease (24).

These results confirm our second hypothesis, that marital satisfaction is associated with better psychological wellbeing. A good marital relationship is associated with better future perspective of the patients. Also, partners who reported a good relationship felt that they had better wellbeing and were less depressed. Although the causal connection between relationship quality and hope about the future needs to be established, we speculate that a good relationship might serve as a buffer against worse times, as has been observed in a study of partners of patients with fronto-temporal dementia (25).

No relation of age with psychological outcome variables was found. However, a positive relation was found between the duration of the relationship before the patient became ill and depression in patients. The psychodynamics of this observation need to be studied further, but it may be that a longer relationship is associated with more dependency on the spouse, which may cause feelings of vulnerability and, in the long-run, depression. For the spouse there was no relation between the duration of the premorbid relationship and depression.

Female partners have a much worse sense of wellbeing than patients and male partners. This is in line with previous research (26). Several explanations have been provided for the higher burden of the female caregiver: (i) because of the female social role, they provide more nurturing activities (27, 28); (ii) females reported more strain because their relationships with family members and others were affected due to their supportive role (29); (iii) men cope more efficiently with life stresses in general (30, 31); (iv) response bias, men just report less stress and burden (32); (v) male caregivers receive more support from informal and formal sources to ease the burden of care (33); and (vi) men are harder for than women, possibly because they rely more exclusively on the primary caregiver (34). Much of this research has been carried out with dementia patients. The passive symptoms of DM can provide an additional explanation through the differential role that is expected from men and women. It is more accepted for women that they are passive than it is for men. Men are more expected to play an active role within and outside the family. This difference between men and women is not observed in the patients themselves, it especially affects the partners; both female partners, for whom care may be imposed and who may lack family income; and male partners, of whom a number may feel comfortable with a submissive wife. In general it is more burdening for women to have a passive husband than it is for men to have a passive wife.

We found that 40% of the patients and 38% of the female partners report BHS scores in the clinical range ( $\geq 9$ ). According to Beck et al. (21) these scores on the BHS are indicative of suicidal ideation. Fourteen percent of patients and 16% of partners were found to have depression. These proportions are higher than reported by Kalkman et al. (35), who found a 1-month depression diagnosis in 2.5% of patients with DM in their sample. This percentage is similar to the 2.7% 1-month prevalence of depression in the general Dutch population that was reported by Bijl et al. (36). This difference can be explained by the severity of the cooperating patients. Kalkman et al. (35) included only ambulatory patients; in our study 14

patients were visited at home as they were not mobile enough to visit the hospital.

Limitations of this study concern the self-selected study sample. The majority of participants were members of the patient organization and therefore may not be representative of all couples with DM. Membership of a patients' association, through promoting contact with fellow-patients, may be beneficial to people with a chronic disease, especially to people who are relatively severely ill (37). A second bias may be caused by the inclusion criterion that only complete couples were invited to participate in this study. We have no data on the issues involved in divorced or otherwise separated couples, even not whether such a separation is the result of DM. Thus our sample may consist of couples that have better marital satisfaction than separated couples. The patient-partner relationship regarding marital satisfaction and other psychological outcomes may be different in separated DM couples. Thirdly, because of the inclusion criterion that patients should be able to complete the questionnaires, there is a selection of less severely affected patients. Hence generalizability is restricted to these patients and the findings may not hold for couples with more severe patients.

In this study we made use of generic questionnaires on wellbeing for the burden of DM. In particular, the BHS is used for the patient's and partners' view of the future. A more thorough and meticulous understanding of their future perspective, for example on the implications for the next generation, could be obtained by in-depth interviews. Also, the perspective and possibilities to articulate marital satisfaction can be quite different for patients and partners. Follow-up studies may provide more insight into this.

In conclusion, the burden of the disease for couples confronted with DM is exceptionally high. In healthcare, consideration should be given to the impact of DM on the partner relationship, but also to the potential that a good relationship can offer for dealing with worse times in the future.

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## APPENDIX I. Principal components and loadings of 29 symptoms of dystrophia myotonica

	Weakness	Lack of initiative non-verbal expression	Stiffness	Gastrointestinal
<b>Weakness</b>				
Weakness in forearms, e.g. when removing lid from jar	<b>0.64</b>			
Weakness in arms or legs when exercising	<b>0.64</b>			
Weakness in feet/lower legs, tripping/ankle sprain	<b>0.60</b>		-0.20	0.25
Weakness in neck, keeping head upright	<b>0.58</b>		0.22	0.23
Weakness in hands/fingers when writing or grasping	<b>0.51</b>	0.30		-0.22
Weakness in arms/shoulders, e.g. when combing hair	<b>0.51</b>		0.28	
Difficulty chewing properly	<b>0.49</b>		0.38	
Frequent choking	<b>0.46</b>			0.21
Weakness in upper legs, difficulty rising from chair	<b>0.45</b>			<b>0.46</b>
Stiff hand after firm grip	0.36	0.25		-0.30
Frequent constipation	0.32			
Frequent severe abdominal cramps	0.25			
<b>Lack of initiative/non-verbal expression</b>				
Difficulty undertaking actions		<b>0.72</b>		
Tendency to postpone actions		<b>0.71</b>		
Feeling as if curtains before eyes impair view		<b>0.49</b>		
From looking at my face others think I am not interested		<b>0.49</b>		0.33
Difficulty getting up in the morning		<b>0.47</b>	0.37	
Difficulty articulating		<b>0.41</b>		
From looking at my face others think I am tired		0.38		
Drooping upper eyelids in photographs of own face	0.27	0.28		
Naps during the day/after meals	0.30	0.29	-0.37	
<b>Stiffness</b>				
Cramp in jaw when chewing	0.25		<b>0.72</b>	
Stiff jaw when biting			<b>0.71</b>	
Stiff tongue when yawning			<b>0.61</b>	
Cramp in jaw/tongue when yawning		0.22	<b>0.52</b>	
Cramp in hand when grasping		0.28	<b>0.41</b>	-0.20
<b>Gastro-intestinal</b>				
Occasional faecal incontinence				<b>0.62</b>
Food can get stuck in throat	0.26	0.25	0.24	<b>0.48</b>
Frequent diarrhoea		0.28		<b>0.48</b>
Explained variance (%)	11.7	9.5	9.2	6.0

Loadings >0.40 in **bold**, loadings <0.20 have been blanked for clarity.

Loadings are comparable to correlations of the items with the latent components.

APPENDIX II. *Limitations in performing activities*

Activity	Patient (%)	Partner (%)
Standing	48	43
Standing without support/stability	54	57
Walking without support	37	33
Walking with steps normal length	66	63
Rising from chair without use of arms	66	52
Climbing stairs	65	62
Descending stairs	59	41
Rising from squatting position	71	65
Chewing tough food	57	56
Swallowing solid food	32	27
Swallowing without choking	37	37
Speaking articulated	65	61
Making gross movements arms (swinging, polishing)	35	28
Making delicate movements hands and fingers (writing, small buttons)	46	34
Making precise movements legs/feet (accelerator/brake of car)	24	15
Keeping head upright	30	32
Reaching with arms above shoulder height	42	34
Going up steps	48	39
Getting into a bus, train or tram	49	42
Carrying objects such as shopping bags	66	65
Hearing appropriately during conversation	25	20
Appropriate vision	42	39
Expressing oneself appropriately in speech	37	22
Expressing oneself appropriately in writing	27	18
Being able to concentrate	39	32
Being able to recall events for more than 5 min	8	4
Total	45	39

APPENDIX III. *Need for help*

	Patient		Partner	
	Some (%)	Complete (%)	Some (%)	Complete (%)
Personal hygiene	16	3	19	4
Light activities, dusting, dishwashing	30	13	24	18
More arduous tasks, making beds, vacuum-cleaning	24	43	35	31
Cooking	24	11	30	19
Shopping	38	18	45	16
Mobility outdoors	22	14	26	19
Total	26	17	30	18