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HEALTH-RELATED QUALITY OF LIFE IN MYOTONIC DYSTROPHY TYPE 1 AND ITS RELATIONSHIP WITH COGNITIVE AND EMOTIONAL FUNCTIONING

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Objective: **To evaluate the health-related quality of life in myotonic dystrophy type 1 and its relationships with clinical, genetic, neuropsychological and emotional factors.**
Design: **Case-control study of a continuous series of patients with myotonic dystrophy type 1.**
Patients and methods: **Twenty patients, and 20 age-, sex- and education-matched healthy controls underwent the MOS 36-Item Short-Form Health Survey (SF-36), an extensive neuropsychological battery and emotional functioning tests.**
Results: **Patients' SF-36 mean scores were lower than those of controls in all dimensions. The neuropsychological study showed a significant impairment in visuospatial and verbal abstract reasoning ($p = 0.001$), visuospatial memory ($p = 0.002$) and attentive functions ($p = 0.03$) in patients with myotonic dystrophy type 1. The emotional assessment showed significantly high scores in anxiety ($p = 0.002$) and depression ($p = 0.001$), which occurred in approximately 50% of patients. Both physical and mental SF-36 areas were inversely correlated with age, duration and grade of disease, depression and anxiety and positively correlated with attentive control. SF-36 areas were not correlated with cytosine thymine guanidine expansion.**
Conclusion: **Health-related quality of life is severely impaired in myotonic dystrophy type 1 and it is negatively influenced by severity and duration of disease as well as by specific cognitive deficits and changes in emotional functioning. Therapeutic intervention in this field could contribute to ameliorate health-related quality of life in myotonic dystrophy type 1.**

Key words: myotonic dystrophy, health-related quality of life, cognitive functioning, emotional functioning.

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INTRODUCTION

Myotonic dystrophy type 1 (DM1) is a progressive, disabling multisystemic autosomal dominant disorder (1), characterized by an unstable triplet cytosine thymine guanidine (CTG) repeat in the 3′ untranslated region of the protein kinase gene on chromosome 19, which appears to be excessively amplified (2–4).

In addition to neuromuscular symptoms, many patients with DM1 develop intellectual impairment and emotional disorders as a result of brain involvement. Recently, we demonstrated that non-congenital patients with DM1 have cortical atrophy, mainly located in areas involved in psychological and emotional functions (5). While mental retardation is commonly reported in the congenital form, patients with non-congenital DM1 frequently show subtler cognitive dysfunction. Moreover, personality disturbances and affective symptoms (particularly anxiety, lack of energy and motivation, apathy and social avoidance) have been reported since the first clinical descriptions of the disease (6, 7). Cognitive and emotional deficits are believed negatively to influence the quality of life of patients with DM1. A more thorough investigation of such deficits might lead to treatments that may improve the quality of life of patients with DM1.

The aim of this study was two-fold: first, to assess the health-related quality of life (HRQoL) and mental functioning (cognitive and emotional functioning) in a series of patients with non-congenital DM1; second, to assess whether patients' HRQoL and cognitive and emotional functioning are associated.

MATERIAL AND METHODS

Subjects

A continuous series of 20 patients with DM1 (11 men and 9 women, aged between 24 and 63 years, median age 37 years; years of education 5–18, median 13 years) gave their informed consent to participate in the study. No included patient had congenital DM1 or any other neurological disease that might affect muscle or cognitive function. Disease duration ranged from 1 to 43 years (median 12 years). Age at onset of disease ranged from 13 to 46 years (median 19 years). CTG triplet expansion, evaluated in all the subjects from leukocytes, ranged from 100 to 1570 repeats, mean 461 (SD 365). The severity of muscular involvement, measured according to the muscular disability rating scale divided into 5 grades (8), was: grade I (no clinical muscular impairment) in 2 patients; grade II (minimal signs: myotonia, facial weakness) in 7 patients; grade III (mild muscle weakness: no external help required in everyday tasks) in 5 patients; and grade IV (moderate muscle weakness: external help required in everyday tasks) in 6 patients. No patient had grade V of disease severity.

Twenty healthy subjects, matched with the patients with DM1 for age, sex and years of education, were used as controls.

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Assessments

Cognitive functioning. A battery of neuropsychological tests was administered to explore a broad spectrum of cognitive functions which included global cognitive status (Mini Mental State Examination, MMSE) (9), visuospatial (Raven's Standard Progressive Matrices, SPM) and verbal (Similarities) abstract reasoning (10, 11), verbal fluency (A-F-S verbal fluency test) (12), verbal short-term memory (Digit span) (13), immediate and delayed verbal recall and learning strategies (Rey's 15-word auditory learning test) (14), visuospatial and constructional abilities (Rey's complex figure copy, copying of drawing with landmark) (11, 15), delayed visuospatial recall (Rey's complex figure 15-minute delayed recall) (15), focused attention and visual scanning (Simple and Double barrage) (11), attention and ability to selectively suppress perceptual interference (Stroop Color Word Test) (16), ideomotor and buccofacial praxis (apraxia examination) (11). Raw scores were adjusted for age and education according to the Italian normative data (11).

Emotional functioning. Depressive and anxious symptoms were assessed using respectively the Hamilton rating scale for depression (HRS-D) 17-item version (17) and the Hamilton rating scale for anxiety (HRS-A) (18), both of which were administered after a psychiatric interview by an expert examiner (FS). Moreover, subjects were asked to fill out the state-trait anxiety inventory form Y (STAI–Y) (19), which provides a score for state anxiety (STAI-S) and a score for trait anxiety (STAI-T).

Quality of life. The HRQoL was evaluated by means of the MOS 36-item short-form health survey (SF-36) (20, 21), a multi-item scale that assesses 8 health concepts. The 4 domains of the SF-36 (physical area) that assess physical health concepts are: limitations in physical activities because of health problems (PF), limitations in usual role activities because of physical problems (RP), bodily pain (BP) and general health perception (GH). Four other domains (mental area) that assess mental health concepts are: vitality (VT), limitations in social activities because of physical or emotional problems (SF), limitations in usual role activities because of emotional problems (RE) and general mental health (psychological distress and well-being) (MH). Each domain is scored on a 0–100 scale, with a higher score indicating a better perception of health/functioning.

Statistical analysis

The statistical analysis was performed by a medical statistician.

Values of the SF-36 domains in controls were compared with data from a wide sample of an Italian healthy population ($n=608$) (22) using the 2 sample t-test to evaluate the representativeness of the control group. The scores of the SF-36 as well as those of the neuropsychological and emotional tests in both patients and controls were compared with the 2 sample t-test. Correlations between different variables in the DM1 group, controlled for age and both severity and duration of disease, as well intercorrelations between the eight SF-36 domains, were explored using Spearman's and Pearson correlation tests, as appropriate. Test results were evaluated using a p-value $=0.05$.

RESULTS

Neuropsychological and emotional evaluation

Table I shows the results of the neuropsychological and emotional evaluation in patients and controls.

Patients displayed a significantly lower score than controls on Raven's SPM, Digit span, Similarities, Rey figure recall, Stroop word card (W) and colour-word card (CW). The neuropsychological data did not correlate with the demographic (sex, age), genetic (CTG expansion) and clinical (disease duration, severity of muscular involvement) characteristics.

Patients scored significantly higher than controls in HRS-D, HRS-A and STAI-T. Mild depression (HRS-D score 8–17) was detected in 10 patients (50%), while marked anxiety symptoms (HRS-A $\geq$10) were observed in 8 patients (40%). If compared with the Italian normative data for STAI-Y (23), marked state anxiety and trait anxiety emerged in 9 (45%) patients. Trait anxiety was directly correlated with illness duration ($p<0.02$).

Health-related quality of life

The SF-36 mean score profile of our controls was not different from that of the healthy population. Patients' SF-36 mean scores were lower than those of controls in all the dimensions (Fig. 1).

Table II shows the correlation between the scores of each SF-36 domain and the demographic, clinical, neuropsychological and emotional scores in patients with DM1. The age of patients, as well as the duration and severity of disease, were inversely correlated with both physical (PF, RP, GH) and mental areas (VT, SF). No correlation was found between the SF-36 scores and sex or CTG expansion.

With regard to the cognitive data, only the performance in the CW card of the Stroop test, which tests selective attention and the ability to suppress an automatic response in the presence of interfering stimuli, positively correlated with SF-36 domains (PF, GH, VT). No correlation was observed between MMSE and SF-36 scores.

The scores in the emotional tests (HRS-D, HRS-A, STAI-T) were inversely correlated with both the physical (RP) and mental areas (VT, SF, MH).

Table III shows intercorrelations of SF-36 domains. Both physical and mental dimensions had several positive intercorrelations.

DISCUSSION

Our study shows that overall HRQoL is significantly lower in patients with DM1 than in either controls or the general healthy population. Previous investigations in groups of individuals with muscular dystrophy, including DM1, have similarly shown a low HRQoL, which was correlated with physical disability (24, 25) and changes in respiratory functions (26). However our study, analysing a homogeneous group of DM1 patients, gives specific information about the influence of cognitive and emotional functioning on HRQoL in this disease. In our patients, several aspects of HRQoL were correlated with the demographic and clinical characteristics. In particular, limitations in physical and social activities because of health problems, general health perception and vitality were negatively influenced by the patients' age, disease severity and disease duration. On the contrary there was no correlation between SF-36 scores and CTG expansion, which has been related to severity of disease (4). This leads us to hypothesize that factors, other than genetics, (i.e. disease duration, insight of disease, emotional reactions or deficits, capacity to cope with physical limitations etc.), might influence the effect of the severity of disease on HRQoL.

Table I. *Neuropsychological and emotional findings in 20 patients with myotonic dystrophy type 1 and 20 healthy controls*

	Patients Mean (SD)	Healthy control Mean (SD)	*p*-value
Neuropsychological tests			
MMSE	27.69 (1.64)	28.22 (0.92)	ns
Raven SPM	38 (13.43)	51.42 (8.92)	0.001
Digit span			
Forwards	5.94 (1.31)	7.05 (1.13)	0.008
Backwards	4 (1.20)	5.42 (1.54)	0.003
Similarities	16.84 (1.26)	18.71 (0.80)	0.001
Fluency	23.49 ± (8.13)	28.99 (8.61)	ns
Rey figure			
Copy	34.10 (2.60)	35.15 (1.21)	ns
Recall	18.31 (7.16)	23.57 (6.28)	0.021
Rey words			
Immediate recall	38.83 (7.34)	43.02 (8.06)	ns
Delayed recall	8.27 (2.04)	8.89 (2.40)	ns
Stroop			
W	92.95 (19.11)	113.95 (20.58)	0.002
C	68.79 (12.89)	73.79 (9.74)	ns
CW	38.16 (14.55)	47.21 (10.28)	0.033
Barrage			
Simple	59.89 (0.31)	59.94 (0.23)	ns
Double	11.74 (1.82)	11.78 (1.13)	ns
Copying of drawing with landmarks	20.58 (1.17)	20.89 (0.31)	ns
Apraxia			
Ideomotor	19.58 (0.61)	19.84 (0.37)	ns
Buccofacial	19.58 (0.69)	19.74 (0.56)	ns
Emotional tests			
HRS-D	8.42 (4.68)	3.68 (2.85)	0.001
HRS-A	8.84 (5.32)	4.05 (3.22)	0.002
STAI-S	38.16 (12.45)	34.26 (9)	ns
STAI-T	43.16 (8.69)	36 (10.57)	0.029

MMSE = Mini Mental State Examination; SPM = Standard Progressive Matrices; W = word card; C = colour card; CW = colour-word card; HRS-D = Hamilton rating scale for depression; HRS-A = Hamilton rating scale for anxiety; STAI-S = score for state anxiety; STAI-T = score for trait anxiety; ns = not significant.

With regard to cognitive functioning, the neuropsychological evaluation in our sample showed a significant failure in visuospatial and abstract reasoning, as well as in visuospatial memory and attentive functions. While the prevalence of dementia in DM1 varies greatly in different studies (discrepancies are probably due to the different types of patients studied), most evaluations of the neuropsychological profile in DM1 report a constant frequency of visuospatial and constructional disabilities, often associated with difficulties in frontal lobe tasks and, in more severe cases, with alterations in language and memory (27–30). This is in accordance with the finding that brain involvement in DM1 is characterized above all by regional, rather than global changes (5, 29).

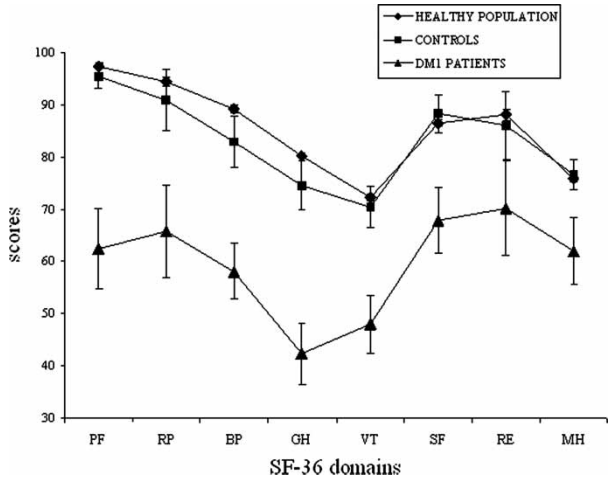

Fig. 1. SF-36 mean score profile in 20 patients with myotonic dystrophy type 1 (DM1), 20 healthy controls and general healthy population ($n = 608$). PF = physical functioning; RP = role limitations due to physical problems; BP = bodily pain; GH = general health perception; VT = vitality; SF = social functioning; RE = role limitations due to emotional problems; MH = general mental health.

The emotional assessment revealed marked anxiety characteristics and mild depression in approximately 50% of our patients. Affective symptoms, particularly atypical depression, are frequently reported in DM1, whereas few patients meet the criteria for a depressive disorder (31–33). Slowness, poor self-esteem, fatigability and irritability are the depressive symptoms most commonly reported (30). Similarly, a high incidence of anxious personality traits (avoidant, dependent, obsessive-compulsive, passive-aggressive) has been observed in DM1 (30, 34, 35). Some authors state that patients with DM1 "do not present significant depressive or anxious symptoms, but rather an emotional deficit, which may be an adaptive reaction to the threatening implications of the disease, or the effect of the central nervous system lesions" (33). The

Table II. *Demographic, clinical characteristics, neuropsychological and emotional scores significantly correlated with SF-36 dimensions (R of Pearson and ρ of Spearman as appropriate)*

	PF	RP	BP	GH	VT	SF	RE	MH
Demographic and clinical data								
Age	−0.63**	ns	ns	ns	−0.51*	ns	ns	ns
Disease duration	−0.62**	−0.52*	ns	−0.71**	−0.69**	−0.5*	ns	ns
Grade of disease	−0.67**	ns	ns	−0.51*	−0.62**	ns	ns	ns
Neuropsychological tests								
Stroop CW	0.63**	ns	ns	0.46*	0.50*	ns	ns	ns
Emotional tests								
HRS-D	ns	−0.55*	ns	ns	ns	ns	ns	−0.68**
HRS-A	ns	−0.56*	ns	ns	ns	ns	ns	−0.54*
STAI-S	ns	ns	ns	ns	ns	−0.61*	ns	−0.81**
STAI-T	ns	−0.55*	ns	ns	−0.55*	−0.66**	ns	−0.61*

* $p < 0.05$; ** $p < 0.01$.
PF = physical functioning; RP = role limitations due to physical problems; BP = bodily pain; GH = general health perception; VT = vitality; SF = social functioning; RE = role limitations due to emotional problems; MH = general mental health; CW = color-word card; HRS-D = Hamilton rating scale for depression; HRS-A = Hamilton rating scale for anxiety; STAI-S = score for state anxiety; STAI-T = score for trait anxiety; ns = not significant.

hypothesis that emotional traits, such as neuropsychological changes, are linked to neurobiological factors has also been raised on the basis of cerebral blood flow studies (30). Personality/emotional disturbances may, however, also reflect an adjustment to this progressively disabling medical condition (34). Indeed, disease duration largely influenced trait anxiety in our sample.

The second aim of our study was to assess whether HRQoL dimensions correlated with patients' cognitive and emotional performance. Our results revealed that attentive control, anxiety and depressive symptoms were the main factors correlated with HRQoL. In particular, the impairment of attentive control, as measured by Stroop Color Word, was correlated with the SF-36 dimensions that assess performance in physical activities, general health perception and vitality. The emotional components were correlated with the dimensions that assess the influence of physical health on work or daily activities, the quality and amount of social activities and the perception of mental well-being. In accordance with these findings, other authors have reported a high prevalence in DM1 of avoidant personality trait, a disturbance that interferes with the quality of life of these patients in their personal, work and leisure aspects (30). Moreover, an association between low quality of life and emotional reactions to stressful situations caused by this disease has been reported (36).

The intercorrelations between physical and mental domains of SF-36 suggest a reciprocal influence of physical and mental dimensions on wellbeing perception.

In conclusion, our findings provide further evidence of the existence of a cognitive profile and emotional disturbances in DM1, and suggest that both specific cognitive deficits and emotional changes affect HRQoL in this disease. However, we cannot exclude that emotional changes (e.g. depression) may directly affect the judgement of the perceived HRQoL, as measured with SF-36. Moreover, to some extent, it could also be possible that a low HRQoL produces or increases anxiety and depression. At this regard, different ways of assessment could contribute to obtain an evaluation of HRQoL less generic than SF-36. A psychological and/or a pharmacological intervention aimed at helping patients to cope with their disease by preventing or reducing anxiety and depression, as well as at improving attentive functions might enhance HRQoL in DM1. More extensive studies designed to evaluate the influence of neuropsychological and emotional factors on HRQoL and to explore the suitability of therapeutic intervention in patients with DM1 are warranted.

Table III. *Intercorrelations between the SF-36 domains (R of Pearson)*

	PF	RP	BP	GH	VT	SF	RE	MH
PF		ns	0.56*	0.69**	0.064**	ns	ns	ns
RP	ns		ns	0.66**	0.62**	0.68**	0.50*	0.62**
BP	0.56*	ns		0.49*	ns	ns	ns	ns
GH	0.69**	0.67**	0.50*		0.70**	0.71**	ns	0.78**
VT	0.64**	0.62**	ns	0.70**		0.67**	ns	0.61**
SF	ns	0.68**	ns	0.71**	0.67**		0.53*	0.77**
RE	ns	0.50*	ns	ns	ns	0.54*		ns
MH	ns	0.63**	ns	0.78**	0.55*	0.87**	0.74**	

*$p < 0.05$; **$p < 0.01$.
PF = physical functioning; RP = role limitations due to physical problems; BP = bodily pain; GH = general health perception; VT = vitality; SF = social functioning; RE = role limitations due to emotional problems; MH = general mental health; ns = not significant.

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