

Clinical Study

Patient Preference Assessment Reveals Disease Aspects Not Covered by Recommended Outcomes in Polymyositis and Dermatomyositis

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Received 12 January 2011; Accepted 2 March 2011

Academic Editors: A. M. Huber and E. Tchetina

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Objectives. Polymyositis (PM) and dermatomyositis (DM) are characterized by impaired muscle function with a majority of patients developing sustained disability. The aim of this study was to evaluate the patient's individual priorities (patient preference) of disabilities most important to improve in PM/DM using the MacMaster Toronto Arthritis Patient Preference Disability Questionnaire (MACTAR), to correlate the MACTAR to myositis outcomes and to evaluate its test-retest reliability. **Methods.** Twenty-eight patients with PM/DM performed recommended outcomes as well as the MACTAR, which was performed twice with one week apart. **Results.** Sexual activity, walking, biking, social activities, and sleep constituted the predominating disabilities. Seventy-two and 33% of the identified disabilities were not covered by items of the Health Assessment Questionnaire and the Myositis Activities Profile. Correlations between the MACTAR and health-related quality of life measures were $r_s = -0.67-0.73$, correlations with measures of activities of daily living and participation in society were $r_s = 0.51-0.60$ with lower correlations for other outcomes. Intraclass correlation (ICC) and weighted Kappa (K_w) coefficients were 0.83 and 0.68, respectively, for test-retest reliability of the MACTAR. **Conclusions.** The MACTAR interview had promising measurement properties and identified patient preference disabilities in PM/DM that were not covered by recommended outcomes.

1. Introduction

Polymyositis (PM) and dermatomyositis (DM) are chronic inflammatory conditions characterized by muscle weakness, characteristic skin rashes in DM, and in about 85% of cases interstitial lung disease [1, 2]. Medical treatment consists of high-dose glucocorticoids, immunosuppressive treatment, and exercise [1, 3]. Although treatment usually results in diminished muscle inflammation and improved muscle function, the majority of patients develop sustained disability [4]. Patients with PM and DM have poorer perceived health than healthy individuals [5].

The International Myositis Assessment Clinical Study Group (IMACS) has validated outcome measures for disease activity and disease damage and recommends the SF-36 to

assess health-related quality of life [6–8]. In addition, two myositis-specific outcome measures have been developed assessing muscle endurance [9] and activities of daily living [10]. None of these measures reflect patient preference.

A patient preference outcome serves to identify individual disease-related disabilities. Studies indicate that predefined outcomes as fixed item questionnaires might include items not relevant for all patients and might not include items important to the individual patient [11–13]. The MacMaster Toronto Arthritis Patient Preference Disability Questionnaire (MACTAR) is valid and responsive for patients with rheumatoid arthritis (RA) [11, 14] and systemic sclerosis (SSc) [15, 16].

The objective of this study was to evaluate patient preference in PM and DM by use of the MACTAR, to compare the

MACTAR to recommended and disease-specific outcomes in PM and DM and to evaluate test-retest reliability of the MACTAR.

2. Methods

2.1. Patients. Patients with PM/DM are seen annually by the myositis team at the Rheumatology unit at Karolinska University Hospital, Stockholm, Sweden; all patients seen between February 2005 and January 2006 were invited to participate in the study. Inclusion criteria were (a) diagnosis of PM or DM according to Bohan and Peter criteria [17], (b) diagnosis duration > 6 months, and (c) speaking and understanding the Swedish language. Twenty-nine patients were invited, of whom one declined participation for unknown reasons; thus, 28 patients were included. Their level of education was categorised. Characteristics of all the 28 patients are presented in Table 1.

All patients were given written and oral information and signed informed consent. The study was approved by the local ethics committee at the Karolinska University Hospital.

2.2. Assessments. The Dutch modified MACTAR is a semistructured interview, including questions on different aspects of disability during the last week, general health, quality of life, physical function, social function, and emotional function [14]. In addition the interviewer asks the patient to identify disabilities due to their PM/DM. To help the patient to identify disabilities, the interviewer mentions different categories: household work, dressing and grooming, outdoor activities or activities related to work, moving around, sexuality, relations, social activities, recreational activities, sleeping, and maintaining food and eating habits. The patient ranks the identified disabilities in accordance to importance for improvement, and the five highest ranked disabilities are recorded. The MACTAR varies between 19 and 39, where 39 indicates no disability [14].

Measurements of disease activity and disease damage includes physician's assessment of global disease activity (MYOACT Global) and global extraskelletal muscle disease activity (MYOACT Extra), which includes assessments of 6 organ systems. The myositis damage index global (MDI Global) includes assessments of 11 organ systems [6–8], and serum levels of CPK were analysed. The Manual Muscle Test (MMT) measures isometric strength in 8 muscle groups, each scored from 0 to 10, with a score varying from 0 to 80 (where 80 indicates full strength). The Health Assessment Questionnaire (HAQ) comprises 20 questions divided into 8 categories [18]. The HAQ is scored from 0 to 3 (where 3 indicates unable to do). For assessment of Health-Related Quality of Life (HQL) we used the Short Form 36 questions (SF-36) comprising of 8 domains. Each SF-36 dimension is scored from 0 to 100, where 100 is optimal HQL [19].

The Functional Index 2 (FI-2) is a valid disease-specific clinical outcome measuring muscle endurance in patients with PM and DM [9]. It is scored as the number of correctly performed repetitions in seven muscle groups, five with a score varying from 0 to 60 and two with scores varying from

TABLE 1: Characteristics of 28 PM/DM patients.

	Value
Age, years, median (range)	57 (28–74)
Sex, female/male, <i>n</i>	15/13
Diagnosis, PM/DM, <i>n</i>	11/17
Disease duration, years, median (range)	9 (1–32)
Educational level, <i>n</i> (%)	
(i) Low	14 (50)
(ii) High	14 (50)
Employment status, <i>n</i>	
(i) Part-time/full-time job	18
(ii) Retired	7
(iii) Sick-leave	3
MYOACT Global, VAS, 0–100 mm	1 (0–5) ^b
MYOACT Extra, VAS, 0–100 mm	0 (0–6) ^c
MDI Global VAS, 0–100 mm	18 (8–23) ^c
CPK, $\mu\text{cat/l}$	1.8 (1.3–2.6) ^b
FI-2, mean (0–100%)	49 (27–56) ^b
MMT-8, 0–80	77 (72–80) ^b
MAP, 1–7	3 (2–4)
HAQ, 0–3.00	0.50 (0.13–1.00) ^a
MACTAR, 19–39	27 (23–29)
PGA, VAS, 0–100 mm	44 (26–59) ^d

Values are median (quartiles) unless otherwise indicated. ^aOne missing case; ^btwo missing cases; ^cthree missing cases; ^done case excluded. Low educational level; up to and including vocational training, and high educational level: higher vocational and/or university education. MACTAR: MacMaster Toronto Arthritis Patient Preference Disability Questionnaire, MAP: Myositis Activities Profile, FI-2: Functional Index 2, VAS: Visual Analogue Scale, PGA: patients' assessment of disease impact on well-being, HAQ: Health Assessment Questionnaire, MYOACT Global: physician's assessment of global disease activity, MYOACT Extra: physician's assessment of global extra skeletal muscle disease activity, MDI Global: myositis damage index global, MMT-8: Manual Muscle Test-in 8 muscle groups, and CPK: Serum levels of creatine phosphokinase micro cat/litre.

0 to 120 repetitions (60/120 indicating no limitations). The Myositis Activity Profile (MAP) measures activities of daily living consisting of 31 items divided into four subscales and four single items each scored from 1 to 7, 1: “no difficulty” to 7: “impossible” [10].

We added a measurement of participation in society using the patients' assessment of disease impact on well-being on a VAS, 0–100 (PGA), where 100 is severe impact on well-being.

2.3. Experimental Procedures. The Dutch modified MACTAR was first translated into Swedish by an authorized translator and then translated back into Dutch by a bilingual rheumatologist (RvV), according to the process described by Guillemain [20]. The translated MACTAR was then reviewed by three health professionals and pretested for relevance and comprehension in three patients with PM/DM. During a visit to the myositis team physical therapists, the FI-2 was performed followed by the MACTAR and the MAP. All patients with unchanged disease activity and medication for

three months were also invited to perform the 15-minute MACTAR interview once again one week later. All MACTAR interviews were performed by the same physical therapist (L. A. Munters) and the FI-2 was performed by any of two well-trained physical therapists (L. A. Munters or H. Alexanderson). The team nurse administrated the PGA, the HAQ, and the SF-36, and any of the two team physicians assessed myositis damage and disease activity (MDI Global, MYOACT Global, and MYOACT Extra) and CPK-levels within a week of the other measurements.

2.4. Statistical Analysis. Analyses of content of the individual identified disabilities using the MACTAR were performed as comparisons to all items in the HAQ and the MAP. Spearman's correlation coefficient (r_s) was used for analyses of correlations between the MACTAR score and the other measures: r_s 0–0.25: “very little or little” correlation, r_s 0.26–0.49: “low”, r_s 0.50–0.69: “moderate”, r_s 0.70–0.89: “high”, and $r_s > 0.90$: “very high” correlation [21]. For analyses of random variation between the test and the retest interview, the linear Weighted Kappa coefficient (K_w) and intraclass correlation coefficient (ICC) were used. The Sign test was used to analyse systematic variance of the MACTAR score. The level of significance was determined to $P < .05$. Weighted Kappa coefficients between 0 and 0.20 indicate no or low agreement, between 0.21 and 0.40 as fair, between 0.41 and 0.60 as moderate, between 0.61 and 0.80 as substantial, and between 0.81 and 1.0 as almost perfect [22]. ICC coefficients < 0.75 indicate low-fair reliability and those > 0.75 indicate good-excellent reliability [23]. The disabilities identified in the first MACTAR interview and at retest were analyzed as descriptive comparison.

3. Results

3.1. Patient Preference in PM/DM. A total of 43 different disabilities were identified by using the MACTAR. Eleven patients identified sexual activity among the five disabilities that were most important to her/him to improve. This was the most commonly identified disability followed by walking ($n = 9$), social activities ($n = 9$), sleeping ($n = 8$), and bicycling ($n = 7$). Twenty-one percent of the 43 reported disabilities were completely covered by items of the HAQ, 7% were partly covered, and 72% were not covered. Thirty-two percent were completely covered by items of the MAP, 35% partly, and 33% were not covered (Table 2).

3.2. Correlations between the MACTAR and Recommended Outcomes. The MACTAR score correlated best with three dimensions of the SF-36: mental health ($r_s = -0.73$) ($P < .05$), social functioning ($r_s = -0.70$) ($P < .05$), and role Emotional ($r_s = -0.67$) ($P < .05$) and secondly with the PGA ($r_s = 0.60$) ($P < .05$).

There were lower correlations to the HAQ ($r_s = 0.57$) ($P < .05$), the MAP ($r_s = 0.51$) ($P < .05$), the MMT ($r_s = -0.46$) ($P < .05$), the FI-2 ($r_s = -0.29$) (NS), the MDI Global ($r_s = 0.11$) (NS), the MYOACT Global ($r_s = 0.13$) (NS), the MYOACT Extra ($r_s = 0.03$) (NS), and the CPK

($r_s = 0.14$) (NS). One patient was excluded due to the patient's misunderstanding of the PGA.

3.3. Test-Retest of the MACTAR Score. All 28 patients were invited to perform retest of the MACTAR. However, in six cases, the retest interview was not performed due to administrative problems, one patient declined invitation, and one patient was excluded due to an emotional crisis. Therefore, the results are calculated on the remaining 20 patients.

The median MACTAR score was 27 (19–33) for the test and retest interview. The weighted Kappa coefficient (K_w) was 0.68 without systematic variation, $P = .60$. The ICC was calculated to 0.83 with a measurement error of 3.28 (CV% = 12%). A median of 3 out of 5 (3-4) disabilities was identical in the MACTAR test and retest; however, they were not always in the same rank order. A median of 1 (0–2) disabilities out of five was ranked identically in the two MACTAR interviews.

4. Discussion

The MACTAR identified aspects of disability of high importance to the patients not covered by IMACS recommended outcomes and myositis-specific outcomes. The patients identified sexual activity, walking, biking, social activities, and sleeping as disabilities deemed most important to improve. The MACTAR seems to have promising measurement properties assessing patient preference in these patients.

Most disabilities that were listed by the patients were not covered by items in the HAQ, while a larger proportion were covered by items in the MAP. This difference could be due to the fact that the MAP is a disease specific outcome whereas the HAQ was originally developed for RA. In a previous study of the Dutch MACTAR in RA only 48% of the self-reported disabilities were covered by items in the HAQ which could be due to the fact that the HAQ only includes self-care and physical activities [14]. Our study confirms and extends these results to myositis because the patient-identified disabilities were also related to restriction in society and interpersonal interactions and relationships. The most commonly identified disability categories by RA patients in the Dutch study were moving around, household work, personal care, recreational activities, and hand function which partly diverges from our study. In a French patient population with systemic sclerosis the most commonly cited domains were mobility, domestic life, and community, social, and civic life [15]. These variations in prioritized disabilities may be explained by the differences among the rheumatic conditions and/or by personal and societal contexts. Strength of the MACTAR is the possibility to identify disabilities adapted to the environmental context of importance to the individual.

Importantly, in our study sexual activity was the most frequently identified disability. Limitations in sexual activity have not to our knowledge been described before in patients with PM/DM. Questions about sex life are usually excluded in predefined questionnaires as they can violate personal

TABLE 2: The highest prioritized MACTAR disabilities identified by 28 patients with PM/DM and their representation in the HAQ and the MAP.

MACTAR disabilities	n^a (men/women)	Disabilities included in the HAQ	Disabilities included in the MAP
Sexual activity	11 (6/5)	Not	Not
Walks/walking	9 (6/3)	Included	Included
Social activities	9 (4/5)	Not	Included
Sleep	8 (2/6)	Not	Not
Bicycling	7 (2/5)	Not	Not
Vacuuming	6 (1/5)	Included	Included
Getting up from ground/floor	5 (3/2)	Partly	Not
Relations to child/spouse	5 (3/2)	Not	Not
Carpentry	5 (3/2)	Not	Partly
Gardening	5 (3/2)	Not	Included

Disabilities identified by less than five individuals: driving a car, playing with child/grandchild, carrying/lifting heavy objects, walking uphill, climbing stairs, running/jogging, skiing ($n = 4$), opening jars/bottles, lifting a frying pan, holding a dish brush for longer time, putting up curtains, taking down a lamp from ceiling, washing hair, traveling ($n = 3$), swallowing/eating, dancing, getting out of a car/boat, putting on socks/shoes, playing golf, working activities, hiking, going to the movies/sports events ($n = 2$), doing buttons, stress management, taking up a boat from water, dressing, playing tennis, climbing a ladder, playing guitar, walking on icy ground, using public transportation, playing soccer, and fatigue ($n = 1$).

^a Number of patients identifying each of the listed disabilities.

Included: exact the same disability identified with MACTAR is also included in the questionnaires MAP or HAQ, Partly: the identified disability is partly represented in the MAP or HAQ, and Not: the disability is not at all represented in the MAP or the HAQ.

integrity. The original HAQ included a ninth domain including questions about sexual activity; however, the authors stated that a relatively low number of patients responded to this question and it was later on excluded [24]. In the Swedish version of HAQ, sexual activity is not included [18]. Maybe it is easier to answer questions about sexual activity in an interview than in a predefined questionnaire.

The MACTAR score correlated best with the SF-36 dimensions mental health, social functioning, role emotional, and the PGA. This could be due to that the MACTAR includes specific questions on general health, quality of life, social function, and emotional function.

Our findings of high correlations with only three domains of the SF-36 and lower correlations with the other myositis outcomes indicate that the MACTAR adds important new aspects of disability not captured in hitherto recommended outcomes for patients with PM or DM.

A good to excellent test-retest reliability was achieved for the MACTAR score. Considering the large potential variation of the score, the error of measurement of 3.28 (CV% = 12) is fairly small. There was a lower agreement than expected between the disabilities at test and retest. In some cases this appeared to be due to external changes such as seasonal weather changes. These results point out a possible limitation of the MACTAR in that disabilities once prioritized for improvement may lose relevance over time [11, 16]. Test-retest was not performed in eight patients. However, the missing cases did not differ statistically significantly from the 20 patients in the retest group regarding age, disease activity, disease duration, muscle function, or activities of daily living.

The low total number of patients included in this study is a limitation, and all patients had chronic, stable disease with low disease activity. However, patients of both gender,

various ages, disability, and education level were included; thus the external validity of this study seems reasonable for patients with chronic disease.

In conclusion, the MACTAR identified disabilities of high importance to the patients that were not covered by recommended and disease-specific outcomes for patients with PM and DM. The MACTAR has promising measurement properties assessing patient preference in PM and DM and might therefore be considered as a patient preference outcome in clinical trials and other clinical settings.

Acknowledgments

The authors would like to thank all participating patients, Maryam Dastmalchi for performing disease activity measures, Ingrid Lundberg for performing disease activity measures and manuscript input, Christina Ottosson for distributing the questionnaires, and Nina Brodin for help with ICC calculations.

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