Quality of life in dermatomyositis

Renato Goreshi, MD,[a,b] Monika Chock, MD,[c] Kristen Foering, BS,[a,b] Rui Feng, PhD,[d] Joyce Okawa, MS,[a,b] Matt Rose, MS,[a,b] David Fiorentino, MD,[c] and Victoria Werth, MD[a,b]
Philadelphia, Pennsylvania, and Stanford, California

Background: Quality of life (QoL) for patients with inflammatory skin disease can be significant, but has been evaluated in just one study in dermatomyositis (DM).

Objective: We sought to examine the relationship between the Cutaneous Dermatomyositis Area (CDASI) and Severity Index, a DM-specific cutaneous severity instrument, and various QoL study instruments and to determine the impact of DM on QoL.

Methods: Skin-specific QoL instruments, the Skindex and the Dermatology Life Quality Index, and global medical QoL instruments, the Short Form 36 and the Health Assessment Questionnaire-Disability Index, were used. Pruritus was evaluated by a visual analog scale and a 0-to-10 scale in DM and cutaneous lupus erythematosus (CLE) populations, respectively.

Results: There was a significant correlation between the CDASI and all skin-specific QoL scores (lowest $P = .0377$). Using the Short Form 36, DM population was found to have significantly worse QoL scores than the general population with the exception of bodily pain (all subscore P values $< .01$). Furthermore, DM had a significantly lower vitality score, representing energy level, compared with CLE, hypertension, diabetes, and recent myocardial infarction scores (lowest $P = .003$). There was a significantly lower mental health score, representing overall mood, to all compared diseases except CLE and clinical depression (P values $< .01$ when significant). We found that DM produces more pruritus than CLE ($P < .0001$).

Limitations: A larger patient population needs to be studied to further assess QoL in patients with DM.

Conclusion: We conclude that DM has a large impact on QoL, even when compared with other diseases, and that DM skin disease activity correlates with a poorer QoL. (J Am Acad Dermatol 2011;65:1107-16.)

Key words: autoimmune disease; clinical research; connective tissue disease; cutaneous lupus; dermatomyositis; itch; pruritus; quality of life.

Dermatomyositis (DM) belongs to a group of diseases called idiopathic inflammatory myopathies.[1] Patients with amyopathic and hypomyopathic DM demonstrate absent or clinically insignificant muscle findings.[2] Recently, a tool called the Cutaneous Dermatomyositis Area and Severity

From the Philadelphia Department of Veterans Affairs Medical Center[a]; University of Pennsylvania School of Medicine[b]; Stanford University, School of Medicine[c]; and Center for Clinical Epidemiology and Biostatistics, University of Pennsylvania.[d]

This material is based on work supported by the National Institutes of Health (NIH), including NIH K24-AR 02207 (Dr Werth) and training grant NIH 5-R25-HL084665-04 (Dr Goreshi). This work was also partially supported by a Merit Review Grant from the Department of Veterans Affairs Veterans Health Administration, Office of Research and Development, Biomedical Laboratory Research and Development.

Disclosure: Dr Fiorentino received honoraria from Amgen, Centocor, Chemocentryx, and MedImmune; received grants from Centocor, Abbott Laboratories, and Amgen; and filed a patent (US patent application serial number 12/694,980 for "Profiling for Determination of Response to Treatment for Inflammatory Disease" filed on January 27, 2010). Drs Goreshi, Chock, Feng, and Werth, Ms Foering, Ms Okawa, and Mr Rose have no conflicts of interest to declare.

Accepted for publication October 10, 2010.

A poster and abstract of this study, with a smaller number of study patients, were presented at the Society of Investigative Dermatology Annual Meeting on May 10, 2010 in Atlanta, GA and published in the *Journal of Investigative Dermatology* 2010;130(Suppl):S54, respectively.

Reprint requests: Victoria Werth, MD, Department of Dermatology, Perelman Center for Advanced Medicine, Suite 1-330A, 3400 Civic Center Blvd, Philadelphia, PA 19104. E-mail: werth@mail.med.upenn.edu.

Published online July 4, 2011.
0190-9622/$36.00

doi:10.1016/j.jaad.2010.10.016

Index (CDASI) was validated as a useful means to measure cutaneous findings in DM quantitatively.[3,4]

Cutaneous autoimmune diseases have a negative impact on quality of life (QoL).[5-8] Up to now, there has been only one published report on the impact of DM on a patient's QoL.[9] Now that new skin evaluation instruments and comparable data between DM and other diseases are available, studies on QoL in DM can be introduced into the literature.

Recently we performed a prospective study on a cohort of patients with DM and skin involvement using the CDASI to elucidate the impact of the disease on their QoL and to demonstrate the need for more effective therapies that may include the use of off-label medications when standard less expensive options are ineffective. A substantial percentage of patients with DM have psychiatric disease, especially depression and anxiety, compared with the healthy population, the prevalence ranging from approximately 20% to 40% of patients with DM, compared with 11% to 30% of healthy persons, respectively.[10-17] Only one study has examined patients with cutaneous DM using a 5-point physician global assessment (PGA) and skin-specific QoL measures.[9] No studies on DM have been performed using global health QoL measures. DM is a systemic disease, and thus it is important to study its effects on QoL using global medical QoL measures in addition to dermatologic QoL measures.

Comparing the QoL in DM relative to other dermatologic and nondermatologic diseases is important to evaluate more completely its impact on QoL. Our previous work with a large cohort of patients with cutaneous lupus showed that patients with lupus have a significantly lower QoL than patients with common skin diseases such as nonmelanoma skin cancer (NMSC), acne, and alopecia.[5] Cutaneous lupus erythematosus (CLE) had a greater effect on QoL relative to nondermatologic diseases such as hypertension (HTN), type 2 diabetes mellitus, congestive heart failure, and recent myocardial infarction. There was no significant difference between lupus and DM using the Skindex-29 in a DM group.[5] Further research into comparisons of QoL between patients with DM and patients with cutaneous lupus is needed.

CAPSULE SUMMARY

- There is only one published study examining the impact of dermatomyositis (DM) on patients' quality of life.
- Patients with DM have worse quality of life on all subscales of the Short Form 36 compared with the general population and score worse on vitality and mental health compared with several chronic diseases. Patients with DM experience more pruritus than patients with cutaneous lupus.
- The presence of pruritus may help to distinguish between DM and cutaneous lupus.

Our study examined the importance of clinical signs or symptoms in DM relative to cutaneous lupus to aid in assessing therapeutic efficacy. Pruritus, for example, is a common symptom experienced by many patients with DM.[18] Although it has been suggested that pruritus can be used as a distinguishing marker between DM and cutaneous lupus, it is also a prevalent symptom in both juvenile and adult DM. There have not been any studies showing a difference between the pruritus experienced by patients with DM and patients with cutaneous lupus.[19-21]

METHODS

Patient population

All patients with clinical or pathologic evidence of DM seen at the Hospital of the University of Pennsylvania and Stanford University Dermatology Clinics were invited to participate in the study regardless of whether or not they were currently undergoing treatment. A total of 55 patients from the University of Pennsylvania and 65 patients from Stanford University were enrolled in the study. Of the 42 patients from the University of Pennsylvania who were able to participate in a study visit, 42 had their skin assessed by the CDASI, 42 completed the Dermatology Life Quality Index (DLQI), 41 completed the Skindex-29+3, 40 completed the pain visual analog scale (VAS), 40 completed the itch VAS, 40 completed the patient global assessment (PtGA), 42 completed the Short Form 36 (SF-36), and 39 completed the Health Assessment Questionnaire (HAQ). Of the 42 patients from the University of Pennsylvania, 8 were male, 34 were female, 14 had classic DM (CDM) with active muscle involvement, and 28 had no active muscle involvement. Of the 55 patients from Stanford University who were able to participate in a study visit, 55 had their skin assessed by the CDASI, 50 completed the HAQ, 50 completed the DLQI, 50 completed the pain VAS, 49 completed the itch VAS, and 7 completed the SF-36 fully (10 patients completed all but the role-emotional [RE] aspect of the SF-36). The differences in the number of completed questionnaires are a result of noncompletion of the study questionnaires by error by the study patients. The large discrepancy in the Stanford population completing the SF-36 is a result of their later introduction of the SF-36 into the study

Abbreviations used:

CDASI:	Cutaneous Dermatomyositis Area and Severity Index
CDM:	classic dermatomyositis
CLE:	cutaneous lupus erythematosus
CTCL:	cutaneous T-cell lymphoma
DLQI:	Dermatology Life Quality Index
DM:	dermatomyositis
HAQ:	Health Assessment Questionnaire
HAQ-DI:	Health Assessment Questionnaire-Disability Index
HTN:	hypertension
MH:	mental health
NMSC:	nonmelanoma skin cancer
PGA:	physician global assessment
PtGA:	patient global assessment
QoL:	quality of life
RE:	role-emotional
SF-36:	Short Form 36
SLE:	systemic lupus erythematosus

documents for their patients. There was no selection bias to which patients ultimately completed or did not complete the questionnaires. Of the 55 patients from Stanford University, 15 were male and 40 were female. The subtype of DM was not recorded in the Stanford population at the time of the research visit.

In all, 194 patients seen at the Hospital of the University of Pennsylvania Dermatology Clinic with clinical and/or pathological evidence of lupus were invited to participate in the study. Of these, 123 patients had CLE without systemic involvement and 71 patients had systemic lupus erythematosus (SLE) with cutaneous disease. In all, 178 of the 194 patients with lupus completed the Skindex-29. In all, 161 and 162 patients completed the pruritus and pain assessment scales, respectively. Of the 123 patients with CLE, 112 completed the SF-36 and 65 of the 71 patients with SLE completed the SF-36. The study was approved by the institutional review board. All patients were aged 18 years or older and were enrolled after signing institutional review board–approved informed consent and Health Insurance Portability and Accountability Act forms.

Study instruments

A number of study instruments were completed at the same visit. To assess QoL among patients with DM, two skin-specific QoL measures, the Skindex-29+3 and the DLQI, and two global health QoL measures, the SF-36 and the HAQ-Disability Index (HAQ-DI), were completed by patients. In addition, patients answered questions regarding their demographics and disease history. To assess disease severity, the physician completed the CDASI and PGA measures.

Skindex-29+3. The Skindex-29 is a validated skin-specific measure of QoL.[22-25] Each question has 5 choices, scored as 100, 75, 50, 25, or 0. Scoring is categorized to different subscales. These include functioning, symptoms, emotions, and photosensitivity. Three questions were added to represent DM-specific effects: namely two questions for photosensitivity and one question for alopecia. The maximum score in each category is 100, indicating maximum disability.

Dermatology Life Quality Index. The DLQI was the first validated dermatologic QoL study instrument to be described.[8,26] It measures the effect of dermatologic disease on QOL and consists of 10 questions, each of which can be answered by marking one of 4 choices. These are scored from 0 to 3, with a maximum total score of 30, indicating maximum disability. The DLQI has been previously shown to be reliable and valid.[8,26]

Short Form 36. The SF-36 is a global medical QOL study instrument that previously has been shown to be reliable and valid.[27,28] The instrument consists of 36 items answered by marking from 2 to 6 options. Scoring ranges from 0 to 100, with 0 indicating maximum disability. Scores are categorized to the following: physical functioning, role-physical, bodily pain, general health, vitality, social functioning, RE, and mental health (MH) (Table I). The first 4 scores can be summed to create the physical composite score, whereas the last 4 can be summed to create the mental composite score. The SF-36 was used to compare QoL of DM with lupus, congestive heart failure, recent myocardial infarction, depression, and HTN using the SF-36 Health Survey and values obtained from our previous work, except for SLE and CLE values, which were obtained during this study.[29]

Health Assessment Questionnaire-Disability Index. The HAQ-DI is a global medical study instrument found to be reliable and valid.[30-32] The combined responses to questions indicating the difficulty in performing 20 different instrumental activities assess the level of disability of the patient. Each is scored from 0 to 3, with 3 representing maximum disability. The HAQ-DI consists of 24 questions which assess 8 instrumental activities: (1) dressing and grooming, (2) arising, (3) eating, (4) walking, (5) hygiene, (6) reach, (7) grip, and (8) activities.

Skin severity. The CDASI, a validated disease severity score, was used to determine the severity of cutaneous disease. Total scores range from 0 to 132, which are divided into (1) activity and (2) damage, with scores ranging from 0 to 100 and 0 to 32, respectively.

Table I. Skindex-29 scores of dermatomyositis and other dermatologic diseases

	Sample size	Symptoms, mean (SD)	*P* value	Emotions, mean (SD)	*P* value	Functioning, mean (SD)	*P* value
Vulvodynia	280	50 (17)	.09	50 (20)	.91	44 (22)*	<.0001
DM (University of Pennsylvania)	41	44.9 (24.3)	-	50.4 (26.1)	-	28.2 (26.6)	-
CLE	178	41.3 (23.8)	.385	49.1 (27.8)	.785	28.4 (25.6)	.964
Epidermolysis bullosa	75	49 (25)	.39	35 (26)*	.003	31 (24)	.56
Eczema	102	48 (23)	.47	41 (27)	.06	26 (26)	.65
Pemphigus	126	37 (22)	.05	37 (22)*	.001	33 (23)	.26
Psoriasis	44	42 (21)	.55	39 (27)	.05	23 (27)	.37
Acne vulgaris	63	30 (19)*	.0007	41 (25)	.07	16 (16)*	.004
CTCL	95	32 (23)*	.004	29 (18)*	<.0001	22 (22)	.16
Rosacea	29	33 (20)	.03	33 (20)*	.004	16 (18)	.03
Alopecia	7	31 (24)	.17	27 (33)	.04	14 (23)	.19
Vitiligo	245	13.9 (14.6)*	<.0001	35.9 (23.6)*	.0004	16.7 (19.5)*	.001
NMSC/AK	136	29 (20)*	<.0001	20 (19)*	<.0001	9 (14)*	<.0001
Without skin disease	107	14 (12)*	<.0001	9 (13)*	<.0001	4 (8)*	<.0001

Diseases listed in descending order of aggregate mean of all Skindex-29 subscores.
AK, Actinic keratoses; *CLE*, cutaneous lupus erythematosus; *CTCL*, cutaneous T-cell lymphoma; *DM*, dermatomyositis; *NMSC*, nonmelanoma skin cancer.
*Significant finding ($P < .01$).

Pruritus and pain. Pruritus and pain were measured using a VAS. For pruritus, the VAS has been used extensively and has been found to be useful in a clinical research setting.[25,33,34] The VAS has been known to be a gold standard for assessing pain.[35-38] The VAS is a 100-mm horizontal line with one end representing maximum itch or pain and the other expressing no itch or pain. Patients with lupus used a 0-to-10 scale to represent the amount of pruritus and pain with 10 indicating maximum itch or pain and 0 indicating no itch or pain.

PGA and PtGA. The physician and patients were asked to rate the severity of various aspects and overall disease of DM using a VAS from 0 to 10, where 10 indicates maximum severity.

Statistical analysis

Descriptive statistics were obtained for each of the QoL measures for the DM population. Using the Skindex-29, mean DM scores (one score for each of the 3 subscales) were compared with mean scores of other dermatologic diseases obtained through our laboratory results for CLE, data obtained from Chren et al,[22-24] or literature review.[39-42] Using the SF-36, mean DM norm-based scores (one score for each of the 8 subscores) were compared with those of 7 other diseases and the general population norm-based scores. SLE and CLE data were obtained from our laboratory whereas other compared diseases were obtained from a previous article evaluating QoL in CLE.[5] Because SDs were not available for compared groups other than SLE or CLE, it was assumed that the SD of DM was nonsignificantly different from other groups (SLE and CLE SDs were used because they were available). Skindex-29 and SF-36 means were compared by using a two-tailed *t* test at an adjusted significance level of .01. Mean HAQ-DI and SF-36 subscores were compared between CDM and amyopathic/hypomyopathic DM using the Mann-Whitney U test at an adjusted significance level of .01. The adjusted significance level was used to minimize experiment-wise error rate. Mean itch, pain, and photosensitivity scores for CLE and DM were compared using the Mann-Whitney U test at a significance level of .05. To determine the correlation between disease severity and QoL, CDASI activity scores were correlated with the Skindex-29 and the DLQI, using Pearson correlation. Because there is no straightforward mean to test the difference between two Spearman rho, Pearson correlation was used to determine the correlation between SF-36 subscores and HAQ-DI scores to the PtGA-Overall Disease and the emotional subscale of the Skindex. To determine the significance of statistical differences among correlated correlations, we used the Z test.[43] Nonparametric tests were used because of the nonnormality of certain variables.

RESULTS

Overview of QoL in DM

DM was found to have a mean (SD) DLQI score of 7.6 (6.5) and a HAQ-DI score of 0.795 (0.788). Each of the Skindex-29 and SF-36 subscores, pruritus, and pain score will be described below and in corresponding tables. The mean (SD) CDASI activity score in our study population was 19.5 (10.5). All of the

Table II. Short Form 36 norm-based scores of dermatomyositis and other diseases

	Sample size	PF, mean (*P*)	RP, mean (*P*)	BP, mean (*P*)	GH, mean (*P*)	VT, mean (*P*)	SF, mean (*P*)	RE, mean (*P*)	MH, mean (*P*)
SLE	65	39.8 (.959)	39.0 (.512)	43.0 (.010)	35.8 (.0004)*	40.1 (.110)	37.9 (.032)	40.1 (.035)	41.2 (.058)
CHF	216	35.1 (.050)	37.7 (.126)	46.8 (.165)	39.2 (.154)	44.0 (.784)	44.7 (.432)	43.9 (.549)	49.7 (.003)*
CLE	112†	49.6 (<.001)*	48.7 (<.001)*	53.6 (.046)	46.0 (.015)	49.0 (.003)*	47.4 (.025)	47.7 (.033)	47.7 (.193)
Clinical depression	502	45.2 (.001)*	40.5 (.987)	45.1 (.019)	42.0 (.848)	42.0 (.392)	38.5 (.014)	36.0 (.005)*	33.5 (<.001)*
DM	52‡	39.6	40.5	49.5	41.7	43.5	43.1	42.6	45.3
Recent MI	107	44.4 (.057)	42.5 (.366)	51.1 (.465)	44.9 (.088)	50.3 (.001)*	50.5 (.001)*	47.0 (.065)	50.3 (.013)
HTN	2089	46.0 (.002)*	45.5 (.006)*	50.9 (.520)	46.8 (.001)*	50.6 (<.001)*	51.4 (<.001)*	48.0 (.006)*	51.5 (<.001)*
Type 2 diabetes mellitus	541	43.6 (.066)	44.0 (.062)	49.3 (.901)	43.4 (.267)	49.4 (<.001)*	49.3 (<.001)*	47.6 (.013)	50.9 (.001)*
General population	2474	50.5 (<.001)*	50.9 (<.001)*	52.1 (.144)	50.9 (<.001)*	51.8 (<.001)*	49.9 (<.001)*	49.4 (<.001)*	49.7 (.007)*

BP, Bodily pain (how pain severity interferes with daily activities); *CHF*, congestive heart failure; *CLE*, cutaneous lupus erythematosus; *DM*, dermatomyositis; *GH*, general health (patient perception of health status); *HTN*, hypertension; *MH*, mental health (mood, especially sadness and anxiety); *MI*, myocardial infarction; *PF*, physical functioning (physical activities associated with daily living); *RE*, role-emotional (emotional state influence on work and other daily activities); *RP*, role-physical (physical health influencing work); *SF*, social functioning (emotional/physical problems interfere with social activities); *SLE*, systemic lupus erythematosus; *VT*, vitality (energy level of patient).

*$P < .01$.

†110 Patients with BP score.

‡49 Patients with RE score.

outcome measures had a mixed distribution except for the HAQ, which had a positively skewed distribution.

Skin-specific QoL of DM versus other dermatologic diseases

The Skindex-29 subscores were used to compare DM with 12 other dermatologic diseases and one negative control group (Table I). DM had among the highest mean subscores, with the emotional subscore being the most severely affected [mean (SD) = 50.4 (26.1)]. The emotional subscore in DM was significantly higher than in epidermolysis bullosa, pemphigus, cutaneous T-cell lymphoma, rosacea, NMSC/actinic keratoses, vitiligo, and the negative control group. DM had the highest emotional subscore among all compared groups with the exception of vulvodynia, which had an approximately equivalent emotional subscore. DM also showed a higher mean symptom subscore than all compared groups except for vulvodynia, epidermolysis bullosa (mixture of various subtypes), and eczema, and had a significantly higher score compared with acne vulgaris, cutaneous T-cell lymphoma, NMSC/actinic keratoses, vitiligo, and normal-appearing skin. Patients with DM were less severely affected in the functional realm with a mean (SD) score of 28.2 (26.6), significantly lower than vulvodynia but significantly higher than NMSC/actinic keratoses, vitiligo, and normal-appearing skin.

Global health QoL of DM versus other diseases

The SF-36 subscores were used to compare DM with 7 other diseases and the general population (Table II). When compared with the general population, DM was found to have significantly poorer scores in all domains, except for bodily pain. In the physical realm, DM was found to have significantly poorer scores than CLE, type 2 diabetes mellitus, clinical depression, and HTN, and a significantly better score than SLE either in some or all of the physical domains. DM had the poorest score in physical functioning with a mean (SD) of 39.6 (14.85). In the social/emotional realm, DM had significantly worse score(s) to congestive heart failure, recent myocardial infarction, type 2 diabetes mellitus, and HTN, although a significantly better score to clinical depression. DM had the poorest score in RE with a mean (SD) of 42.6 (13.71).

Global health QoL between different subtypes of DM

HAQ-DI and SF-36 scores were used to compare CDM with amyopathic/hypomyopathic DM.

Table III. Mean (SD) of Health Assessment Questionnaire-Disability Index and Short Form 36 scores in classic and amyopathic/hypomyopathic dermatomyositis

	HAQ-DI	PF	RP	BP	GH	VT	SF	RE	MH
CDM	0.8621 (0.8077)	36.6 (13.5)	40.1 (12.2)	47.4 (16.6)	38.2 (11.1)	40.3 (12.3)	44.0 (11.2)	41.8 (13.3)	45.4 (11.1)
AHDM	0.4450 (0.560)	43.5 (13.6)	41.3 (13.2)	50.4 (11.8)	42.8 (11.2)	45.6 (11.9)	43.2 (14.0)	41.4 (13.8)	43.7 (13.2)
P	.0818	.123	.670	.393	.229	.285	.978	.899	.779

AHDM, Amyopathic or hypomyopathic dermatomyositis; *BP*, bodily pain (how pain severity interferes with daily activities); *CDM*, classic dermatomyositis; *GH*, general health (patient perception of health status); *HAQ-DI*, Health Assessment Questionnaire-Disability Index; *MH*, mental health (mood, especially sadness and anxiety); *PF*, physical functioning (physical activities associated with daily living); *RE*, role-emotional (emotional state influence on work and other daily activities); *RP*, role-physical (physical health influencing work); *SF*, social functioning (emotional/physical problems interfere with social activities); *VT*, vitality (energy level of patient).

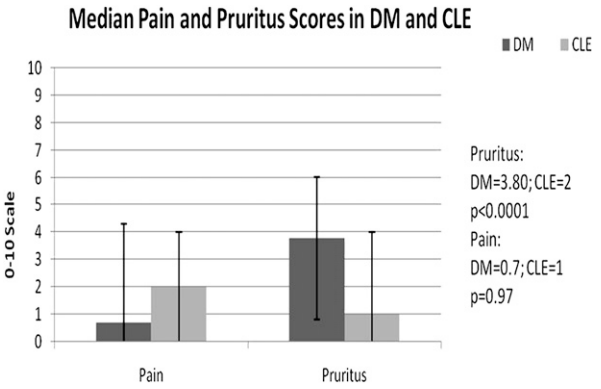

Fig 1. Comparison of pain and pruritus scores in dermatomyositis (*DM*) and cutaneous lupus erythematosus (*CLE*).

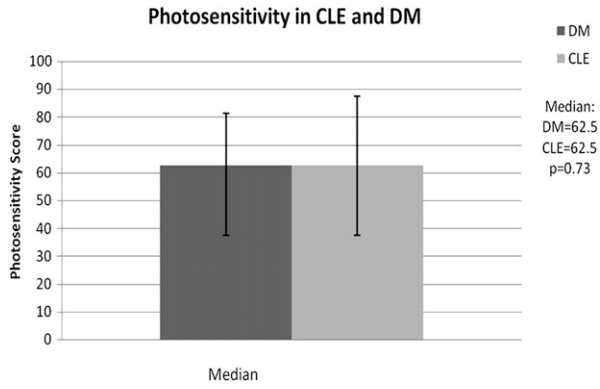

Fig 2. Comparison of photosensitivity scores in dermatomyositis (*DM*) and cutaneous lupus erythematosus (*CLE*).

Although mean scores in the physical subscores of the SF-36 and the HAQ-DI suggest poorer QoL in the CDM population, statistical significance was not reached (Table III).

Pruritus and pain in DM and CLE

DM was found to have a significantly higher median pruritus score than CLE (DM 3.80 and CLE 2) ($P < .0001$) whereas there was no significant difference between median pain scores (DM 0.7 and CLE 1). Fig 1 gives graphic representations of both findings.

Photosensitivity in DM and CLE

Median scores from the two photosensitivity questions added to the Skindex-29 showed no significant difference between DM and CLE (DM 62.5 and CLE 62.5) (Fig 2).

Correlations of skin-specific QoL instrument scores

Each of the Skindex-29 subscores significantly correlated with the DLQI scores (Skindex-29-Symptom $r = 0.6323$, Skindex-29-Emotion $r = 0.6744$, Skindex-29-Function $r = 0.8598$; all P values $< .0001$), implying that the DLQI and the Skindex-29 measure related constructs. The Skindex-29 scores were also found to correlate with one another (lowest $r = 0.5345$, highest $P = .0003$) (Table IV). In addition, the correlation between DLQI and Skindex-29-Function scores were significantly higher than the correlation between DLQI and Skindex-29-Emotion scores ($P = .004$).

Correlation between skin severity and skin-specific QoL instrument scores

All of the Skindex-29 subscores and the DLQI scores significantly correlated with CDASI scores ($r = 0.2729$, $P = .0377$), suggesting that increased cutaneous severity is correlated with a poorer QoL (Table V).

Correlation between pruritus/skin severity and skin-specific QoL instrument scores

Pruritus scores correlated significantly with Skindex-Symptom, Skindex-Function, and DLQI scores ($r = 0.6015$, $P = .001$; $r = 0.45520$, $P = .0036$; and $r = 0.3477$, $P = .0009$, respectively), but significance was not attained between pruritus and Skindex-Emotion subscores. The above results implicate that pruritus is associated with a poorer QoL, although it may not significantly affect a patient emotionally (Table V).

Table IV. Correlations (*P* values) among Skindex-29 subscores and Dermatology Life Quality Index in dermatomyositis

	Skindex-Symptoms	Skindex-Emotion	Skindex-Function	DLQI
Skindex-Symptoms	1			
Skindex-Emotion	0.5345 (<.0003)*	1		
Skindex-Function	0.6020 (<.0001)*	0.7620 (<.0001)*	1	
DLQI	0.6323 (<.0001)*	0.6744 (<.0001)*	0.8598 (<.0001)*	1

Rho/significance level.
DLQI, Dermatology Life Quality Index.
*$P < .01$.

Table V. Correlations (*P* values) between pruritus and Cutaneous Dermatomyositis Area and Severity Index and skin-specific quality of life instruments

	Skindex-Symptoms	Skindex-Emotion	Skindex-Function	DLQI
Pruritus	0.6015 (.0001)*	0.1937 (.2375)	0.45520 (.0036)*	0.3477 (.0009)*
CDASI-Activity	0.3257 (.0377)*	0.4623 (.0023)*	0.4413 (.0039)*	0.2729 (.0071)*

Rho/significance level.
CDASI, Cutaneous Dermatomyositis Area and Severity Index; *DLQI*, Dermatology Life Quality Index.
*$P < .05$.

Comparison of SF-36 and HAQ-DI as global health QoL assessment tools in DM

The R value correlation between each of the physical component SF-36 subscores and the HAQ-DI scores with the overall PtGA was determined. The emotional component of the Skindex was used instead of the PtGA since it was found that the RE and MH subscores of the SF-36 did not significantly correlate with the PtGA, inferring that the PtGA focuses on the physical aspect of a disease. Overall, the emotional components of the SF-36 correlated better with the emotional subscale of the Skindex than with the HAQ-DI. The social functioning subscore of the SF-36 correlated significantly more than the HAQ-DI with the emotional component of the Skindex. There was no significant difference between the individual physical components of the SF-36 or the HAQ-DI and the PtGA (Table VI).

DISCUSSION

The above results indicate that DM has a significant impact on QoL even when compared with various other dermatologic and nondermatologic diseases. Using the Skindex-29, DM was found have a significantly worse impact on the emotional realm of QoL compared with several other diseases. Based on the results of a global health QoL instrument, the SF-36, patients with DM were found to have a significantly worse MH, representing overall mood, than patients representing all of the compared diseases except CLE, SLE, and clinical depression, which all had significantly worse MH scores than DM. This is an important conclusion for multiple reasons. Firstly, although it is well established that psychiatric comorbidities are often present in dermatology, it has also been shown that dermatologists frequently do not identify patients with self-reported psychiatric disease.[10-14,16,17,44,45] Furthermore, a study of 1580 patients with psoriasis in 39 Italian dermatology centers showed that dermatologists do not tend to modify their clinical approach to include psychosocial interventions for patients when psychological distress is present.[46,47] This is especially alarming as patients who have suicidal ideations have significantly higher Skindex-29 subscores (subscore means ranging from 44.3-49.5) than those who do not have suicidal ideations (subscore means ranging from 18.6-32.1).[48] Consequently, it is important to realize that patients with DM may be an underserved population in terms of being evaluated by clinicians for psychological well-being. Secondly, there is increasing evidence in the field of psychoneuroimmunology supporting the role of psychological and social factors influencing the state of disease, particularly in psoriasis, through production of proinflammatory cytokines.[46,49,50] Further investigations are needed to assess how psychological and social factors may influence the disease state in the DM population.

Our results also show that there is a significant correlation between cutaneous disease severity and QoL. This is in accordance to a previous study using a 5-point Likert PGA scale.[9] This has also been shown in a variety of other disease states, including acne, pemphigus, psoriasis, cutaneous lupus, and vitiligo.[5,10,40-42,51]

Table VI. Correlations between global health quality of life instruments (HAQ-DI and individual components of the SF-36), overall disease patient global assessment, and the emotional subscale of the Skindex

Correlation	Correlation with Overall Disease PtGA	Correlation with Emotional subscale of the Skindex	Correlation difference rho-rho_0 (*P* value)
HAQ-DI	0.3453 (rho_0)	0.3451 (rho_0)	-
PF	0.3698	-	0.0245 (0.74)
RP	0.4212	-	0.0759 (0.47)
BP	0.296	-	−0.0493 (0.67)
GH	0.3367	-	−.0086 (0.94)
VT	-	0.2426	−0.1025 (0.43)
SF	-	0.6336	0.2885 (0.02)*
RE	-	0.5181	0.1730 (0.14)
MH	-	0.5418	0.1967 (0.21)

The HAQ-DI (Health Assessment Questionnaire-Disability Index) assesses mainly physical aspects of global QoL. The physical components of the SF-36 [*PF,* physical functioning (physical activities associated with daily living); *RP,* role-physical (physical health influencing work); *BP,* bodily pain (how pain severity interferes with daily activities); *GH,* general health (patient perception of health status)] and emotional compoents [*VT,* vitality (energy level of patient); *SF,* social functioning (emotional/physical problems that interfere with social activities); *RE,* role-emotional (emotional state influence on work and other daily activities); *MH,* mental health (mood, especially sadness and anxiety); *PtGA,* patient global assessment; Short Form 36 scores were multiplied by -1 to create positive correlations.

*$P < .05$, indicating significant difference in between two correlations.

It has been suggested that pruritus may be useful in distinguishing between patients with DM and lupus.[9,19,21] We have shown that prevalence of pruritus is significantly higher in DM than in the CLE population and that it correlates significantly with QoL (the latter finding being consistent with a previous study).[9] Although we have shown that the DM population experiences a significantly higher level of pruritus than the CLE population, further investigation is needed to determine its usefulness in distinguishing between the two diseases.

Although we were not able to achieve statistical significance, which likely was related to our small sample size, the data suggest that CDM is associated with a worse QoL with respect to physical functioning. Investigations that assess QoL among the DM subtypes could shed light on this result.

By comparing the two skin-specific QoL instruments, we were able to show that the DLQI significantly correlates with each of the Skindex-29 subscores and pruritus. Given that the DLQI is a simpler, shorter skin-specific QoL instrument, this finding suggests that the DLQI may be more feasible for clinical use in patients with DM where a more comprehensive outcome instrument such as the Skindex-29 is not needed (ie, where QoL is not a primary outcome variable). It is important to note, however, that the DLQI contains only one question examining purely the emotional impact of skin disease. Given the result that the DLQI correlates with the Skindex-29-Function significantly better than to the Skindex-29-Emotion, and because we have shown that DM creates a significant emotional impact on patients, study instruments examining emotion more thoroughly may be more informative.

In an article evaluating QoL instruments in psoriasis, the DLQI was noted to focus mainly on limitation, supporting our conclusion that the DLQI is an inadequate tool in the DM population because we have shown there is a significant emotional impact as well.[52] Comparing the two global health instruments, the SF-36 emotional subscores correlated better to the Skindex-Emotion than the HAQ-DI, suggesting that the SF-36 is a more useful instrument to assess the emotional components of QoL than the HAQ-DI. Because statistical significance was only achieved comparing the SF-36 social functioning score and the HAQ-DI, further research with a larger study population is needed to better demonstrate that the SF-36 is a better global medical QoL outcome instrument than the HAQ-DI to compare QoL in DM and other nondermatologic diseases. Furthermore, in our study population, 38% scored the minimal score on the HAQ-DI, compared with 0% on the SF-36. This may suggest that the HAQ-DI may not be sensitive enough to detect minor disease in the DM population. Interestingly, in another study, 25.3% of the study population in rheumatoid arthritis had the HAQ-DI minimum score, which may suggest that the HAQ-DI may create a positively skewed distribution in multiple diseases.[53] This conclusion, however, needs to be further assessed as the HAQ-DI has been used extensively, especially in arthritis populations.

Although we were able to make several conclusions from this study, a few limitations should be addressed. The study was cross-sectional and did not discriminate between those who had initiated treatment and those who had not. To fully explore the relationship between disease activity and QoL, future studies on the changes in QoL as disease activity changes are warranted. Secondly, the study is limited by the small study population. A larger patient population needs to be studied to further assess QoL in patients with DM, specifically to better compare DM subtypes and to better compare DM with the other diseases. Thirdly, two study populations were used, namely one population at the

University of Pennsylvania and the other at Stanford University. Thirdly, as both study centers are in referral centers, it is possible that our study population may not represent the DM population as a whole.

REFERENCES

1. Dugan EM, Huber AM, Miller FW, Rider LG, International Myositis Assessment and Clinical Studies Group. Review of the classification and assessment of the cutaneous manifestations of idiopathic inflammatory myopathies. Dermatol Online J 2009;15:2.
2. Gerami P, Schope JM, McDonald L, Walling HW, Sontheimer RD. A systematic review of adult-onset clinically amyopathic dermatomyositis (dermatomyositis siné myositis): a missing link within the spectrum of the idiopathic inflammatory myopathies. J Am Acad Dermatol 2006;54:597-613.
3. Gaines E, Werth VP. Development of outcome measures for autoimmune dermatoses. Arch Dermatol Res 2008;300:3-9.
4. Klein RQ, Bangert CA, Costner M, Connolly MK, Tanikawa A, Okawa J, et al. Comparison of reliability and validity of outcome instruments for cutaneous dermatomyositis. Br J Dermatol 2008;159:887-94.
5. Klein R, Moghadam-Kia S, Taylor L, Coley C, Okawa J, LoMonico J, et al. Quality of life in cutaneous lupus erythematosus. J Am Acad Dermatol 2011;64:849-58.
6. Jayaprakasam A, Darvay A, Osborne G, McGibbon D. Comparison of assessments of severity and quality of life in cutaneous disease. Clin Exp Dermatol 2002;27:306-8.
7. Zachariae R, Zachariae H, Blomqvist K, Davidsson S, Molin L, MØrk C, et al. Quality of life in 6497 Nordic patients with psoriasis. Br J Dermatol 2002;146:1006-16.
8. Finlay AY. Quality of life assessments in dermatology. Semin Cutan Med Surg 1998;17:291-6.
9. Hundley JL, Carroll CL, Lang W, Snively B, Yosipovitch G, Feldman SR, et al. Cutaneous symptoms of dermatomyositis significantly impact patients' quality of life. J Am Acad Dermatol 2006;54:217-20.
10. Picardi A, Abeni D, Melchi CF, Puddu P, Pasquini P. Psychiatric morbidity in dermatological outpatients: an issue to be recognized. Br J Dermatol 2000;143:983-91.
11. Hughes JE, Barraclough BM, Hamblin LG, White JE. Psychiatric symptoms in dermatology patients. Br J Psychiatry 1983;143: 51-4.
12. Wessely SC, Lewis GH. The classification of psychiatric morbidity in attenders at the dermatology clinic. Br J Psychiatry 1989;155:686-91.
13. Woodruff PWR, Higgins EM, du Vivier AWP, Wessely S. Psychiatric illness in patients referred to a dermatology-psychiatry clinic. Gen Hosp Psychiatry 1997;19:29-35.
14. Aktan S, Ozmen E, Sanli B. Psychiatric disorders in patients attending an outpatient dermatology-psychiatry clinic. Dermatology 1997;197:230-4.
15. Attah Johnson FY, Mostaghimi H. Co-morbidity between dermatologic diseases and psychiatric disorders in Papua, New Guinea. Int J Dermatol 1995;34:244-8.
16. Picardi A, Pasquini P, Abeni D, Fassone G, Mazzotti E, Fava GA. Psychosomatic assessment of skin diseases in clinic practice. Psychother Psychosom 2005;74:315-22.
17. Kessler RC, McGonagle KA, Zhao S, Nelson CB, Hughes M, Eshleman S, et al. Lifetime and 12-month prevalence of DSM-III-R psychiatric disorders in the United States: results from the national comorbidity survey. Arch Gen Psychiatry 1994;51:8-19.
18. Kovacs SO, Kovacs SC. Dermatomyositis. J Am Acad Dermatol 1998;39:899-920.
19. Sontheimer RD. Dermatomyositis: an overview of recent progress with emphasis on dermatologic aspects. Dermatol Clin 2002;20:387-408.
20. Peloro TM, Miller OF III, Hahn TF, Newman ED. Juvenile dermatomyositis: a retrospective review of a 30-year experience. J Am Acad Dermatol 2001;45:28-34.
21. Shirani Z, Kucenic MJ, Carroll CL, Fleischer AB Jr, Feldman SR, Yosipovitch G, et al. Pruritus in adult dermatomyositis. Clin Exp Dermatol 2004;29:273-6.
22. Chren MM, Lasek RJ, Quinn LM, Mostow EN, Zyzanski SJ. Skindex, a quality-of-life measure for patients with skin disease: reliability, validity, and responsiveness. J Invest Dermatol 1996;107:707-13.
23. Chren MM, Lasek RJ, Quinn LM, Covinsky KE. Convergent and discriminant validity of a generic and a disease-specific instrument to measure quality-of-life in patients with skin disease. J Invest Dermatol 1997;108:103-7.
24. Chren MM, Lasek RJ. Skindex 1.1: reliability, validity, and responsiveness of a refined instrument to measure the effects of skin disease on quality-of-life. J Invest Dermatol 1997;108:366.
25. Veraldi S, De Micheli P, Schianchi R, Lunardon L. Treatment of pruritus in mild-to-moderate atopic dermatitis with a topical non-steroidal agent. J Drugs Dermatol 2009;8:537-9.
26. Finlay AY, Khan GK. Dermatology life quality index (DLQI): a simple practical measure for routine clinical use. Clin Exp Dermatol 1994;19:210-6.
27. Ware JE Jr, Sherbourne CD. The MOS 36-item short-form health survey (SF-36), I: conception framework and item selection. Med Care 1992;30:473-83.
28. McHorney CA, Ware JE Jr, Raczek AE. The MOS 36-item short-form health survey (SF-36), II: psychometric and clinical tests of validity measuring physical and mental health constructs. Med Care 1993;31:247-63.
29. Ware JE Jr, Kosinski M, Gandek B. SF-36 health survey: manual and interpretation guide. Lincoln (RI): QualityMetric Inc; 2003.
30. Chatzitheodorou D, Kabitsis C, Papadopoulos NG, Galanopoulou V. Assessing disability in patients with rheumatic diseases: translation, reliability, and validity testing of a Greek version of the Stanford health assessment questionnaire (HAQ). Rheumatol Int 2008;28:1091-7.
31. Fries JF, Spitz P, Kramer G, Holman HR. Measurement of patient outcome in arthritis. Arthritis Rheum 1980;23:137-45.
32. Bellamy N, Boers M, Feldson D, Fries J, Furst D, Henry D, et al. Health status instruments/utilities. J Rheumatol 1995;22: 1203-7.
33. Wahlgren CF, Ekblom A, Hägermark O. Some aspects of experimental induction and measurement of itch. Acta Derm Venereol 1989;69:185-9.
34. Langner MD, Maibach HI. Pruritus measurement and treatment. Clin Exp Dermatol 2009;34:285-8.
35. Jensen MP, Karoly P, Braver S. The measurement of clinical pain intensity: a comparison of six methods. Pain 1986;27: 117-26.
36. Yarnitsky D, Sprecher E, Zaslansky R, Hemli JA. Multiple session experimental pain measurement. Pain 1996;67:327-33.
37. Myles PS, Urquhart N. The linearity of the visual analogue scale in patients with severe acute pain. Anaesth Intensive Care 2005;33:54-8.
38. Bergh IH, Stener-Victorin E, Wallin G, Mårtensson L. Comparison of the PainMatcher and the visual analogue scale for assessment of labor pain following administered pain relief treatment. Midwifery 2011;27:e134-9.

39. Tabolli S, Sampogna F, Di Pietro C, Paradisi A, Uras C, Zotti P, et al. Quality of life in patients with epidermolysis bullosa. Br J Dermatol 2009;161:869-77.
40. Paradisi A, Sampogna F, Di Pietro C, Cianchini G, Didona B, Ferri R, et al. Quality-of-life assessment in patients with pemphigus using a minimum set of evaluation tools. J Am Acad Dermatol 2009;60:261-9.
41. Sampogna F, Frontani M, Baliva G, Lombardo GA, Alvetreti G, Di Pietro C, et al. Quality of life and psychological distress in patients with cutaneous lymphoma. Br J Dermatol 2009;160:815-22.
42. Linthorst Homan MW, Spuls PI, de Korte J, Bos JD, Sprangers MA, van der Veen JPW. The burden of vitiligo: patient characteristics associated with quality of life. J Am Acad Dermatol 2009;61:411-20.
43. Meng XL, Rosenthal R, Rubin DB. Comparing correlated correlation coefficients. Psychol Bull 1992;111:172-5.
44. Sambhi R, Lepping P. Psychiatric treatments in dermatology: an update. Clin Exp Dermatol 2009;35:120-5.
45. Picardi A, Amerio P, Baliva G, Barbieri C, Teofoli P, Bolli S, et al. Recognition of depressive and anxiety disorders in dermatological outpatients. Acta Derm Venereol 2004;84:213-7.
46. Locala JA. Current concepts in psychodermatology. Curr Psychiatry Rep 2009;11:211-8.
47. Finzi A, Colombo D, Caputo A, Andreassi L, Chimenti S, Vena G, et al, PSYCHAE Study Group. Psychological distress and coping strategies in patients with psoriasis: the PSYCHAE study. J Eur Acad Dermatol Venereol 2007;21:1161-9.
48. Picardi A, Mazzotti E, Pasquini P. Prevalence and correlates of suicidal ideation among patients with skin disease. J Am Acad Dermatol 2006;54:420-6.
49. Tausk F, Elenkov I, Moynihan J. Psychoneuroimmunology. Dermatol Ther 2008;21:22-31.
50. Urpe M, Buggiani G, Lotti T. Stress and psychoneuroimmunologic factors in dermatology. Dermatol Clin 2005;23:609-17.
51. Jones-Caballero M, Pedrosa E, Peñas PF. Self-reported adherence to treatment and quality of life in mild to moderate acne. Dermatology 2008;217:309-14.
52. De Korte J, Mombers FM, Sprangers MA, Bos JD. The suitability of quality-of-life questionnaires for psoriasis research: a systematic literature review. Arch Dermatol 2002; 138:1221-7.
53. Linde L, Sørensen J, Ostergaard M, Hørslev-Petersen K, Hetland ML. Health-related quality of life: validity, reliability, and responsiveness of SF-36, 15D, EQ-5D [corrected] RAQoL, and HAQ in patients with rheumatoid arthritis. J Rheumatol 2008; 35:1528-37.