Quality of Life and Functional Status in Systemic Sclerosis Compared to Other Rheumatic Diseases

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ABSTRACT. ***Objective.*** To assess clinical factors associated with disability and physical health in patients with systemic sclerosis (SSc) compared to psoriatic arthritis (PsA), systemic lupus erythematosus (SLE), and rheumatoid arthritis (RA) and healthy controls.

Methods. Eighty-two patients with SSc, 82 with PsA, 74 with SLE, 42 with RA, and 60 controls were recruited from various rheumatology clinics and underwent physical examination, tender point count, Health Assessment Questionnaire Disability Index (HAQ-DI) and Short Form-36 Health Survey (SF-36) assessments.

Results. SSc patients were younger and had shorter disease duration than the comparator groups. SSc patients with joint involvement had significantly poorer HAQ-DI scores than patients with PsA (1.43 vs 0.84; $p < 0.05$), and had higher visual analog scale pain scores than RA patients (1.37 vs 1.01; $p < 0.05$). The SF-36 Physical Component Summary and HAQ-DI score in SSc patients were adversely affected by joint involvement ($p < 0.01$, $p < 0.001$, respectively), ≥ 11 tender points ($p < 0.01$, $p < 0.001$), gastrointestinal (GI) involvement ($p < 0.01$, $p < 0.01$), and high skin score ($p = 0.02$, $p < 0.001$).

Conclusion. Physical health relating to quality of life is adversely affected in patients with SSc. Disability is associated with the presence of ≥ 11 tender points, a high skin score, and joint and GI involvement. Joint involvement in SSc is more disabling than joint involvement in PsA; and patients with SSc experience more severe pain than patients with RA. (First Release April 15 2006; J Rheumatol 2006;33:1117–22)

Key Indexing Terms:
SCLERODERMA SYSTEMIC SCLEROSIS DISABILITY
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Systemic sclerosis (SSc) is a multisystem disease classically characterized by inflammation, fibrosis, and a diffuse vasculopathy. The main effect of the disease is on the skin, resulting in thickening and tightness. Joint pain may occur as a result of fibrosis of the joint capsule, thickened tendons, and/or erosive arthritis[1]. Raynaud's phenomenon (RP) and gastrointestinal (GI) manifestations are common, while pulmonary, cardiac, and renal involvement adversely affects prognosis[2]. Clinicians readily recognize that the biologic and physiologic changes of SSc lead to the development of symptoms in affected patients. These symptoms may lead to impairment of functional status. It has only been in the past 2 decades that clinicians have begun to appreciate the impact that a decline in physical function can have on one's general health perceptions. Together, a decline in functional status, an increase in pain, and an alteration of one's general health perceptions may adversely affect a patient's perceived physical health and ultimately quality of life (QOL)[3,4].

The Health Assessment Questionnaire Disability Index (HAQ-DI) is an instrument widely utilized to measure disability in patients with rheumatic disease. Patients self-report the amount of difficulty experienced performing 8 domains of activity. A composite HAQ-DI score is reported, falling between 0 and 3 on an ordinal scale. The HAQ-DI also contains a visual analog scale (VAS) where patients report the amount of pain experienced in the past week[5]. The measurement properties of the HAQ-DI have been evaluated among patients with SSc[6-11], rheumatoid arthritis (RA)[4,5,12], systemic lupus erythematosus (SLE)[13,14], and psoriatic arthritis (PsA)[15].

A commonly utilized measure of QOL is the Medical Outcomes Study Short Form-36 (SF-36)[16]. It is a general health status questionnaire that has the advantage of measuring patient centered outcomes rather than biological or disease centered outcomes perceived by the clinician. The composite QOL score can be broken down to separate components pertaining to physical health (using the physical component summary index) and mental health (using the mental component summary index)[17]. The SF-36 has been used to assess QOL in the rheumatic diseases, and has been validated in patients with RA, PsA, SLE, and SSc[11,18-22].

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Using the HAQ-DI, pain VAS, and physical component summary index of the SF-36, we assessed clinical factors associated with disability, pain, and physical health effects on QOL in patients with SSc, and compared them with PsA, SLE, and RA patients and healthy controls.

MATERIALS AND METHODS

Patient selection. Consecutive patients were recruited from the University of Toronto general rheumatology, lupus, psoriatic arthritis, and scleroderma clinics. Established in the 1970s, the rheumatology subspecialty clinics consist of prevalent, longitudinally followed cohorts of patients who underwent a standardized protocol assessment at regular clinic visits. Consecutive healthy controls were recruited from the waiting room of the family practice clinic. Control subjects were family members of patients in the clinic, who had no comorbid illnesses.

Clinical assessment. All patients and controls underwent a complete physical examination including a detailed joint examination and tender point count. Active joints were defined as joints [proximal interphalangeal (PIP) and metacarpophalangeal (MCP) joints, wrists, elbows, shoulders, hips, knees, ankles, and metatarsophalangeal (MTP) joints] with the presence of stress pain (pain in the extremes of range of motion), joint line tenderness, or swelling. Individuals with one or more active joints were categorized as having joint involvement.

Among patients with SSc, a modified Rodnan skin score assessment was performed[23]. Skin thickness was graded on a scale from 0 to 3, where 0 was normal, 1 denoted mild thickening including the edematous phase, 2 definite thickening with or without skin tethering, and 3 severe encasement. Skin was scored for thickening on the fingers, dorsum of the hand, forearm, arm, cheeks, chest, abdomen, lower leg, and feet bilaterally, giving a skin score range of 0 to 54. The skin score was analyzed as a continuous variable. Patients were considered to have limited disease if their skin thickening was confined to the distal extremities (distal to the elbows and knees). Diffuse disease was defined as skin thickening extending proximal to the elbows and knees. GI involvement was self-reported, and was defined as the presence of dysphagia, heartburn, constipation, or small bowel bacterial overgrowth (defined as the presence of steatorrhea or > 3 loose stools per day).

Quality of life and function measures. All patients completed the SF-36 English (Canada) Acute Version 1.0. It comprises 8 health dimensions: Physical Functioning, Role Physical, Bodily Pain, General Health, Vitality, Social Function, Role Emotional, and Mental Health. Each of the dimensions was separately scored using item weighting and additive scaling[24]. Summed data were transformed onto a 0–100 point scale. These 8 dimensions were combined into 2 health status measures: the Physical Component Summary (PCS index) and the Mental Component Summary (MCS index). For computation of the PCS and MCS, each dimension score was weighted in a 3-step process to produce a standardized T score. The population mean score was 50, with a standard deviation of 10[25].

The HAQ-DI includes 8 categories of functional activities, each of which had 2 or 3 component questions, adding up to 20 items. There are 4 possible responses for each item, ranging from 0 (without any difficulty) to 3 (unable to do). A mean score was calculated for each domain from 0 to 3 using the highest score in each domain, by summing the 8 scores and dividing by 8[4]. The use of devices, equipment, and assistance was included in the instrument as a component weighting factor[26].

As part of the HAQ-DI, a VAS was used to evaluate the amount of pain experienced in the past week. The VAS was a 15 cm line that was converted to a continuous scale from 0 to 3, where 1 cm was equivalent to 0.2 points. The anchors of the VAS were 0 (no pain) to 100 (very severe pain). A metric ruler was used to measure the distance in centimeters from the left anchor to the patient mark, and then multiplied by 0.2. The VAS pain score was not incorporated into the HAQ-DI composite score. Scleroderma, PsA, RA, and control patients completed the HAQ-DI and pain VAS. SLE patients did not complete the HAQ-DI or pain VAS.

Statistical analysis. Statistical analyses were performed using SAS software (SAS Institute, Cary, NC, USA). Data were analyzed using descriptive statistics, t tests, chi square, analysis of variance, and Pearson correlation coefficients as appropriate. P values < 0.05 were considered statistically significant.

RESULTS

The proportion of female patients was similar across all groups, ranging from 84% to 88%. Patients with SSc were slightly younger, with a mean age of 48.2 years, and had shorter disease duration at 7.1 years, than the PsA, lupus, and RA patient groups (Table 1). Among the SSc patients, 42 (51%) had limited and 40 (49%) had diffuse skin involvement.

All 4 patient groups had significantly poorer HAQ-DI, VAS pain, and SF-36 PCS scores than controls. Patients with SSc with joint involvement had poorer HAQ-DI scores than patients with PsA, with scores of 1.43 and 0.84, respectively ($p < 0.05$). SSc patients with joint involvement also had higher VAS pain scores than RA patients, 1.37 and 1.01, respectively ($p < 0.05$; Table 2).

Table 3 gives details of the study cohorts and the numbers found to have joint involvement, ≥ 11 tender points, and GI manifestations. Among SSc patients, there was no significant difference in HAQ-DI score between male and female patients. However, the HAQ-DI score was adversely affected by the presence of ≥ 11 tender points ($p < 0.001$), joint involvement ($p < 0.001$), and GI disease ($p < 0.01$). Similarly, the SF-36 PCS score was not adversely affected by gender, but was adversely affected by the presence of ≥ 11 tender points ($p < 0.01$), joint involvement ($p < 0.01$), and GI disease ($p < 0.01$). The HAQ-DI was also correlated with the presence of a high skin score ($r = 0.50$, $p < 0.001$). There was no correlation between the presence of tender points and the SF-36 MCS score ($r = –0.04$, $p = 0.76$) or the mental health domain ($r = –0.02$, $p = 0.88$).

Table 4 compares outcome measures of SSc patients with limited disease to those with diffuse disease. SSc patients with diffuse disease had a significantly worse HAQ-DI score compared to those with limited disease (1.36 vs 0.59; $p < 0.001$). A significantly higher proportion of SSc patients with diffuse disease had joint involvement than those with limited disease (71% vs 32%; $p < 0.001$). SSc patients with diffuse disease had lower SF-36 scores than patients with limited disease, with borderline statistical significance (40.5 vs 32.4; $p = 0.01$). Similarly, patients with diffuse disease had lower PCS (32.4 vs 40.5; $p = 0.01$) and physical functioning domain scores (44.8 vs 64.1; $p < 0.01$) than patients with limited disease. There was no significant difference between SSc disease types and VAS pain, MCS score, physical role functioning, bodily pain domain, general health domain, vitality domain, social functioning domain, emotional role functioning, and mental health domain scores.

DISCUSSION

Patients with SSc vary in their disease manifestations but may

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Table 1. Patient demographics.

	Controls, n = 60	Scleroderma, n = 82	Psoriatic Arthritis, n = 82	Lupus, n = 75	Rheumatoid Arthritis, n = 42
Female, n (%)	52 (84)	71 (87)	72 (88)	64 (86)	36 (86)
Age, yrs	38.8 ± 10.1	48.2 ± 12.5	50.0 ± 13.8	52.3 ± 13.1	58.5 ± 15.6
Disease duration, yrs	NA	7.1 ± 5.9	16.2 ± 9.0	22.3 ± 6.7	12.9 ± 7.8

NA: not applicable.

Table 2. HAQ-DI, VAS Pain, and SF-36 Physical Component Summary (PCS) scores for patients and controls.

	Controls, n = 60	Scleroderma, n = 43/34*	Psoriatic Arthritis, n = 82	Lupus, n = 74	Rheumatoid Arthritis, n = 42
HAQ-DI	0.10 ± 0.24	1.43 ± 0.89	0.84 ± 0.79	—	1.15 ± 0.78
VAS Pain	0.27 ± 0.57	1.37 ± 0.90	1.2 ± 0.90	—	1.01 ± 0.73
SF-36†: PCS	54.5 ± 6.9	31.8 ± 13.2	36.1 ± 12.6	39.0 ± 13.0	34.1 ± 9.8

HAQ-DI: Health Assessment Questionnaire Disability Index. Range 0–3; 0 = better health. VAS: Visual analog scale. VAS pain range 0–3; 0 = no pain. † SF-36 subscales range: 0–100; 100 = better health. * Sample size for HAQ and pain measures/sample size for SF-36 subscales.

Table 3. Association of clinical variables with HAQ-DI and VAS Pain scores in patients with scleroderma.

			≥ 11 Tender Points		Joint Involvement		Gastrointestinal Disease	
	Male, n = 11	Female, n = 71	Present, n = 37	Absent, n = 39	Present, n = 43	Absent, n = 39	Present, n = 57	Absent, n = 24
HAQ-DI	0.82 ± 0.91	1.0 ± 0.87	1.40 ± 0.92	0.70 ± 0.70†	1.43 ± 0.89	0.50 ± 0.51†	1.13 ± 0.92	0.60 ± 0.60†
VAS Pain	0.81 ± 0.97	1.1 ± 0.94	1.61 ± 0.90	0.67 ± 0.80†	1.36 ± 0.90	0.80 ± 0.92†	1.27 ± 0.95	0.67 ± 0.83†
SF-36 PCS	38.0 ± 15.6	36.3 ± 12.8	30.8 ± 11.7	40.7 ± 13.1†	31.8 ± 13.2	41.3 ± 11.3†	34.2 ± 11.8	42.7 ± 14.6†

HAQ-DI: Health Assessment Questionnaire Disability Index, VAS: visual analog scale, PCS: Physical Component Summary score. † $p \leq 0.01$. SF-36 subscales range: 0–100; 100 = better health.

experience pain due to arthritis and digital ischemia from severe RP. Functional difficulties arise because of thickened and bound-down skin resulting in flexion contractures and restricted joint movement. Weakness can occur on the basis of inflammatory muscle disease or general debility. A reduced quality of life or impairment in function is therefore not unexpected in patients with SSc. However, very few studies have systematically evaluated QOL and function in SSc, and none have compared their findings across other rheumatic diseases[20,22,27,28].

We found that the QOL in patients with SSc, as indicated by their level of physical function, was significantly reduced compared to healthy controls, but similar across groups of rheumatology patients. This is consistent with work by Burckhardt, *et al*, who found a similarly reduced QOL between female patients with SLE and RA, measured using the Quality of Life Scale[29].

We also found that the degree of self-reported disability among SSc patients was adversely affected by the presence of a high skin score. This observation is discordant with the findings of Herrick, *et al*[30], but concordant with other investigators[6,20,27]. The discordance with Herrick, *et al* may be due to the use of different measures of disability in their SSc cohort. Previously reported correlates of a high HAQ-DI score have included diffuse cutaneous involvement, poor hand mobility, reduced fist closure, reduced hand spread, thrombocytosis, lung disease, older age, female sex, tendon friction rubs, and joint pain[7,22,27,31]. Poorer physical function in patients with higher skin scores is expected, as these are patients with more severe disease. Not only do these patients have more severe flexion contractures and loss of hand function but they are at greater risk for serious pulmonary, cardiac, and renal involvement.

We found that SSc patients with joint involvement had more disability than patients with PsA and suffered more severe pain than patients with RA. These observations are consistent with the report that patients with PsA have a lower pain threshold and fewer tender points than those with RA[32]. Studies also indicate that patients with SSc have greater disability than those with RA and SLE, and that disability subsequently has a significant influence on psychosocial adjustment[7,33]. However, disability has largely been attributed to major internal organ involvement, RP, or skin tightening (resulting in limited range of motion)[9,22]. In our study, dis-

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Table 4. Comparison of outcomes between limited and diffuse SSc.

Outcome Measure	Limited SSc, mean (SD) or n (%)	Diffuse SSc, mean (SD) or n (%)	p
HAQ-DI	0.59 (0.70)	1.36 (0.85)	< 0.001
VAS Pain	0.94 (0.98)	1.24 (0.90)	0.15
SF-36	59.91 (24.07)	49.1 (23.9)	0.05
PCS	40.5 (12.6)	32.4 (12.5)	0.01
MCS	44.5 (12.5)	45.8 (11.4)	0.66
PFD	64.1 (26.2)	44.8 (28.4)	< 0.01
PD	52.1 (44.3)	34.8 (41.4)	0.10
BPD	59.9 (28.8)	51.3 (24.9)	0.19
GHD	49.2 (23.6)	38.7 (25.6)	0.08
VD	44.6 (25.9)	37.1 (24.0)	0.22
SFD	64.8 (27.1)	54.4 (26.6)	0.12
ED	63.8 (42.3)	54.5 (46.3)	0.39
MHD	63.6 (22.6)	69.5 (18.8)	0.39
Joint involvement	13 (32%)	30 (71%)	< 0.001
GI involvement	30 (73%)	28 (68%)	0.63

HAQ-DI: Health Assessment Questionnaire Disability Index, SF-36: Medical Outcomes Study Short-Form 36, PCS: Physical Component Summary, MCS: Mental Component Summary, PFD: Physical Functioning Domain, PD: Role Functioning–Physical, BPD: Bodily Pain Domain, GHD: General Health Domain, VD: Vitality Domain, SFD: Social Functioning Domain, ED: Role Functioning–Emotional, MHD: Mental Health Domain, GI: gastrointestinal.

ability was associated with GI involvement. This is perhaps not surprising, as many of the GI manifestations, such as severe dysphagia and heartburn, gastric stasis with early satiety and malnutrition, small bowel bacterial overgrowth, constipation with pseudoobstruction, rectal prolapse, and fecal incontinence, can all have a devastating effect on the QOL.

Our results indicate that joint involvement and pain are also factors contributing to disability in patients with SSc. It has been recognized that pain is common in SSc, and may be the result of digital ischemia and ulceration associated with severe RP or joint inflammation[34-38]. The traditional clinical signs of active joint inflammation are not usually seen in SSc because of the thick, bound-down skin, but an erosive arthropathy does occur with this disease[1]. However, 49% of our SSc cohort had ≥ 11 tender points. Fibromyalgia (FM) has previously been associated with SLE, RA, and PsA[32,39,40]. Gladman, *et al* reported the presence of FM in 21% of patients with SLE, and these patients had a reduced functional status and poorer sense of well-being than lupus patients without FM[41]. The high prevalence of patients with the required number of tender points likely indicates that FM is also associated with scleroderma. The etiology of these tender points is uncertain and may be related to local changes in the skin, tendons, and nerve fibers. There was no correlation between the presence of tender points and measures of mental health. This is an important observation that strengthens the association between pain and physical function and social adjustment in this population[34]. Appropriate pain management interventions may therefore improve physical function and health related QOL.

Comparisons between SSc disease subtypes indicated that patients with diffuse disease suffered more disability and impaired QOL than patients with limited disease. In our study, this may be related to a high proportion of patients with diffuse disease having joint involvement, which influences their PCS score and physical functioning on the SF-36. Together, increased joint involvement, increased disability, and impaired physical functioning scores have concurrent validity. However, one would expect that patients with diffuse disease would have impaired general health, social functioning, mental health, and vitality scores compared to patients with limited disease. This lack of discriminant ability suggests that the format of these domains in the SF-36 may be insensitive to some of the important determinants of QOL in patients with SSc. A disease-specific measure of QOL may have better sensitivity to these important determinants. Future investigators will need to compare the measurement properties of both a generic and a disease-specific measure of QOL to determine the optimal outcome measure in scleroderma trials.

There are a number of limitations to our study. First, examining for the presence of active joint inflammation is difficult in SSc because of the thickened overlying skin. Biopsy studies have shown that active synovitis does occur in SSc, but joint swelling and effusions are difficult to ascertain clinically[42]. Because of the high prevalence of patients with 11 or more tender points in this study, identification of actively inflamed joints, based primarily on the presence of joint tenderness only, may have resulted in an overestimation. A second limitation lies within the conceptual framework of QOL and the constraints of the SF-36 as a QOL measure. Although used extensively in the rheumatologic literature, it is a generic instrument that may not measure all domains that are important to rheumatologic patients. However, in PsA it was the one that most closely indicated changes in clinical status[43]. In our study the SF-36 as a whole was able to determine the limited QOL in SSc. A third limitation lies in the external validity of these results. All patients were assessed in university affiliated, hospital based clinics (these clinics serve as primary, secondary, and tertiary referral center). The mean age of the SSc patients was similar to the patients with PsA and lupus, whereas one may expect the lupus cohort to be younger than the SSc cohort. This reflected the fact that the patients had been followed at the lupus clinic longer than in the other clinics. Patients in the lupus clinic are similar to patients followed at other lupus clinics[44].

Despite these limitations, we believe the results of our study to be important. To our knowledge, this is the first study to evaluate variables that have a negative impact on disability, physical health, and QOL in SSc, compared with 3 other rheumatological populations and healthy controls. These results highlight the degree of pain and functional limitation SSc patients suffer compared to patients with RA and PsA. These associations have previously been underrecognized. The results provide the impetus for further inquiry into this area.

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Physical health relating to quality of life is adversely affected in SSc. Disability is associated with the presence of ≥ 11 tender points, high skin score, and joint and GI involvement. SSc patients with joint involvement are more disabled than those with PsA, and experience more severe pain than patients with RA.

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