Disease and Symptom Burden in Systemic Sclerosis: A Patient Perspective

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ABSTRACT. ***Objective.*** Studies of systemic sclerosis (SSc) have enlisted measures of physical function and generic health-related quality of life in order to determine health status. However, the measurements obtained may not discriminate other essential quality of life (QOL) domains important to patients with SSc. Our study used qualitative methods to evaluate patients' assessment of disease and symptom burden in SSc.

Methods. We conducted 3 focus groups and 5 in-depth interviews of patients with SSc. Guiding questions were based on 5 themes: patient awareness, SSc-related problems, disease activity and progression, symptoms, and expectations. Thematic analysis was conducted using qualitative, grounded theory methodologies.

Results. Symptoms such as pain (localized or generalized), fatigue, and malaise were reported to have major influence on daily activities and QOL. Gastrointestinal symptoms were among the worst prevalent and disruptive physical problems. All participants reported significant disruptions in their social lives, a burden considered by many as the worst consequence of their disease. All expressed major effects on their overall well-being because of emotional distress, including depression, low self-esteem, concerns with physical appearance, and uncertainty about future outcomes.

Conclusion. Patients with SSc report significant symptomatic and emotional burdens, which, in turn, affect their QOL and psychological well-being. Additional research and fuller awareness of the disease and symptom-related burdens experienced by patients with SSc may lead to additional relevant outcome measures and more effective overall treatment programs. (First Release July 1 2007; J Rheumatol 2007;34:1718–26)

Key Indexing Terms:
SYSTEMIC SCLEROSIS QUALITATIVE RESEARCH QUALITY OF LIFE

Systemic sclerosis (SSc) is a chronic multisystem disease that carries a serious prognosis and has a major influence on quality of life (QOL). From the patients' perspective, changes in functional ability and appearance may be more distressing and debilitating than changes in objective measures of disease that preoccupy physicians. However, there is insufficient information as to which subjective domains may be most important to patients because many outcome measures used in SSc report either generic QOL, or single domains (e.g., physical function)[1-4]. Commonly used measures such as the Scleroderma Health Assessment Questionnaire (SHAQ) include visual analog scales for pain and for patient estimates of how individual organ problems [i.e., Raynaud's, digital ulcers, gastrointestinal (GI), breathing, global assessment] interfere with their lives; however, they may lack other constructs relevant to patients with SSc[3,5-8].

We evaluated patients' assessment of disease and symptom burden in SSc using patient-centered qualitative methods. We conducted focus groups and interviews of SSc patients in order to determine patients' perceptions of disease activity, disability, and QOL and to identify areas most distressful and concerning to patients. Ultimately, we wanted to identify subjective domains that need to be measured in order to improve patient-centered outcome measurement for SSc.

MATERIALS AND METHODS

We recruited study participants using 2 approaches: (1) advertising through the Scleroderma Foundation (Blue Bonnet Chapter, Texas); and (2) direct requests at a study investigator's clinical practice (MM). Our objective was to recruit candidates of varying disease duration using a 10-year categorization scheme. Patients who responded were contacted by telephone to confirm participation; they were then given a choice to participate in a focus group or an individual interview.

We conducted 3 focus groups: 2 comprised patients with disease duration of 10 years or more (consisting of 2 and 7 participants, respectively) and one group of 5 patients with SSc of more recent onset (under 10 years). In addition we conducted hour-long individual interviews with 5 patients having a disease duration less than 5 years. Since all participants preferred speaking either English (89%) or English and Spanish (11%), we delivered focus group

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and interview questions in English only. Diffuse SSc was defined by skin involvement of the trunk, face, and proximal and distal extremities; limited SSc was defined by involvement of skin distal to the elbows and knees, and the face[9].

Participants completed sociodemographic questionnaires in which they reported their disease duration, occupation, marital status, preferred language, education, and ethnicity. We did achieve sufficient data saturation, insofar as the final transcripts analyzed yielded no new codes that had not emerged from the interviews and other focus groups[10]. Although one of our groups was small, focus group size has not been shown to affect data validity; individual interviews and larger groups may generate more ideas than smaller groups, but group size has no bearing on the significance or applicability of the ideas themselves[11,12].

We stratified the groups by disease duration for the benefit of participants and the discussion. Homogeneous groups tend to bolster confidence of individual group members to voice their opinions[13]. Further, our purpose was not to divide the group for "between" comparisons during data analysis[14], but to create a taxonomy of areas of difficulty and concern for patients with SSc.

The conceptual framework used for the focus groups is shown in Figure 1. Based on a review of the literature and study investigators' clinical experience, we developed a list of guiding questions regarding effects of disease on QOL, perceptions of disease progression, overall symptom burden, and specific organ symptoms (Table 1). We used the same script of questions for both the interviews and focus groups. The script followed semistructured interviewing, insofar as questions were open-ended but geared to elicit responses focused around 5 themes: (1) patient awareness of SSc, (2) SSc-related problems, (3) disease activity and progression, (4) symptoms, and (5) expectations. For Theme 3, we created 4 different graphs depicting disease progression to assess participants' perceptions of their own clinical course (Figure 2).

All focus groups and interviews were audiotaped and transcribed verbatim. Transcripts were imported into Atlas.ti©, software used to organize and classify codes and quotations and to enable searching transcripts for specific word components and generating analyses in the form of frequencies and summaries. Thematic analysis was conducted using qualitative methodologies akin to open and selective coding from grounded theory[15-17]. We parsed transcripts line by line, developed codes for statements, and mapped codes to the study's guiding themes. Content categories beyond the original codes emerged during subsequent rounds of transcript review. Initial coding was independently conducted by 2 investigators who coded each transcript. Final coding was agreed upon by consensus of the 2 initial coders and a third investigator. The three-member group reached consensus on the natural language statements and codes to be included in the analysis.

RESULTS

Nineteen patients with SSc took part in the study. Although our 2-part recruitment effort lasted over 3 months and yielded 34 total potential participants, incomplete contact information prevented us from contacting 5 of the 34. Of the 29 remaining, none refused to participate; however, one was ineligible because no physician had officially diagnosed her with SSc. Of the 28 willing and eligible respondents, 7 were unable to participate because of scheduling conflicts or personal reasons, and 2 others missed their scheduled session. We cannot compare participants and nonparticipants because Health Insurance Portability and Accountability Act (HIPAA) restrictions prohibited us from obtaining clinical data on nonparticipants. Table 2 gives demographic characteristics of participants. The entire group's median age was 49 years; 17 (90%) were female and 13 (69%) were Caucasian; 18 (95%) had diffuse disease. Mean disease duration was 8 years, with disease onset defined as the date of the first non-Raynaud's phemonenon manifestation of scleroderma.

Common concerns emerged from the group discussions and individual interviews. While our preset themes anticipated many issues ultimately broached in the focus groups and interviews (27 substantive codes), other unexpected issues outside the scope of our original codes also emerged. Table 3 illustrates the most recurrent codes overall, both preset and emergent. The results are organized per theme below.

Theme One: patient awareness

Only a few participants used medical terms to describe their illness. The majority of participants defined their illness symptomatically: "I know that with scleroderma, my body does not release toxins like a normal person does because I don't sweat... I don't do anything automatically." Still others had only vague notions of the disease. "I know that...it's not a well-known disease, ...That it's got to do with, uh, the system.

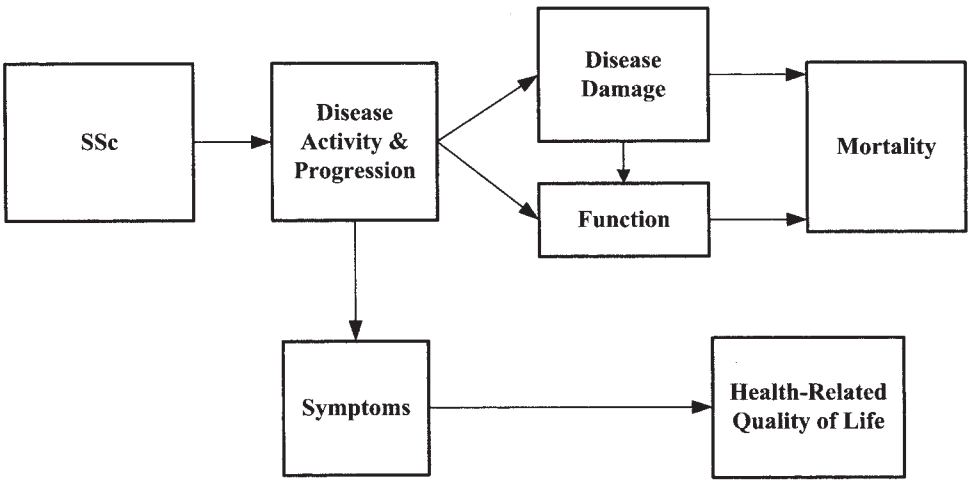

Figure 1. Conceptual framework for conducting and analyzing focus groups.

Table 1. Script of guiding questions asked during focus groups and interviews.

Theme 1 — Patient Awareness
1. What do you know about scleroderma?
2. What do you think caused your scleroderma?
3. How serious is scleroderma?

Theme 2 — SSc-Related Problems
1. What bothers you the most about your scleroderma?
2. How has your life changed since having scleroderma?
3. What kind of physical problems do you experience from your scleroderma?
4. What kind of emotional problems do you experience from your scleroderma?
5. What are the kinds of things or activities that you cannot do anymore because of your scleroderma?
6. What would you say are the three worst problems caused by your scleroderma, and why do they bother you so much?

Theme 3a — Disease Activity
1. Can you tell when your scleroderma is changing, getting better or worse? How can you tell, what changes do you feel?
2. Do you feel at times that your disease is flaring, becoming more active? How can you tell?
3. When you feel that you are having a flare, how long does it last?
4. Are there times when your disease seems to become stable and not progress? How can you tell?
5. Does your disease ever get better? If so, what symptoms go away that make you think you are getting better?

Theme 3b — Disease Progression
(Patients were shown the graphs in Figure 2, depicting disease progression, and asked the following question.)
Which of these patterns resembles your scleroderma the most?

Theme 4 — Symptoms
1. What bothers you the most about your skin?
2. When your disease is becoming active, what do you feel in your skin?
3. Do your joints ever bother you? What do you feel when they bother you?
4. What bothers you the most about your fingers?
5. Do you ever have stomach problems because of your scleroderma? What kinds of problems? What bothers you the most?
6. Do you have any problems eating your food because of your scleroderma? What kind of problems?
7. Do you have any problems with your bowels because of your scleroderma? What kind of problems?
8. Do you have any problems with your lungs because of scleroderma? What symptoms do you get?
9. Do you have any problems with your heart because of scleroderma? What symptoms do you get?
10. Do you generally feel energenic?
11. Do you ever feel tired or worn out because of your scleroderma? How so?
12. How much trouble do you have sleeping or resting because of your scleroderma? How does this affect you?
13. Has scleroderma affected your sexual life?
14. What other symptoms or problems of scleroderma that we have not discussed may be important to you?

Theme 5 — Expectations
1. How do you think your scleroderma will change over the next few years?
2. Do you think scleroderma can be cured?
3. How satisfied are you with the treatment for your scleroderma?
4. What are your worries about the side effects of your scleroderma treatment?
5. Do you think your treatments improve your scleroderma? What is your opinion?

Um, I don't know what the proper words are....” Surfing the web and attending support groups were means by which some participants gathered disease information.

Many participants attributed the cause of their disease to heredity. “We have cousins and family members that have lupus on both sides of the family.” Others felt that stress had precipitated the disease. Still others attributed SSc to environmental factors. “I had worked as... a new construction clean-up person and I had my hands in chemical, daily scrubbing windows and things.”

Theme Two: SSc-related problems

Participants described the effects of SSc on their lives mostly in terms of emotions, appearance, and lifestyle. They often judged the severity of their own illness in comparison to themselves before SSc, or to other patients with SSc. “I can't use my hands the way I used to... I can tell my lungs have gotten weaker.... [compared] to the next person... I feel healthy....I'm the lucky one.”

Emotional distress. Physical symptoms contributed to emotional distress. “When I'm real tired it's easy to cry and stuff...to be sensitive to things that I'm not sensitive to when I've got more stamina.” Fear, depression, denial, low self-esteem, and uncertainty about future outcomes affected participants' overall well-being. “It's always lurking in the corner. What's going to come next?” Frustration with disability

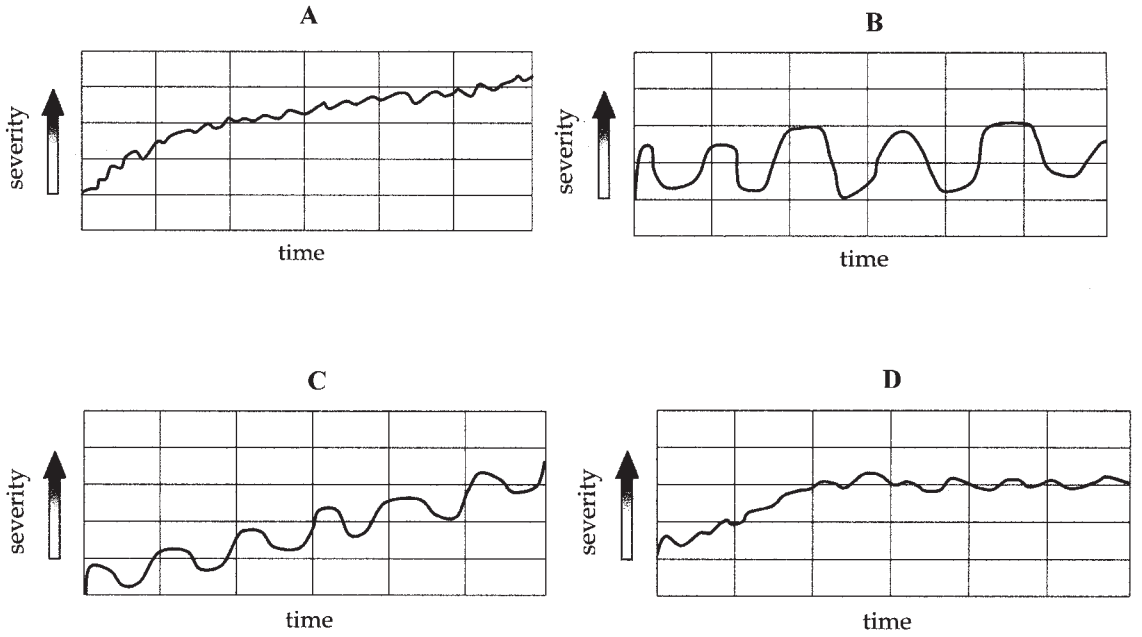

Figure 2. Disease progression graphs. Participants were asked whether they felt their SSc was getting better or worse and whether they could tell when symptoms would flare. A: Slowly progressing. B: Flares. C: Progressing with flares. D: Rapid progress, then stabilization.

Table 2. Demographic characteristics of focus group and interview participants.

Patient Characteristics	No. of Patients (%)
Sex	
Female	17 (90)
Male	2 (10)
Race	
Caucasian	13 (69)
Hispanic	4 (21)
African American	2 (10)
Education	
Less than high school diploma	1 (5)
High school diploma	2 (11)
Trade or technical school	3 (16)
Some college	8 (41)
Bachelor's degree	3 (16)
Advanced degree	1 (5)
Age, yrs, mean (SD)	49 (10)
Occupation	
Full-time	4 (21)
Part-time	2 (11)
Unemployed or disabled	7 (37)
Retired	3 (15)
Homemaker	2 (11)
Student	1 (5)
Disease duration, yrs, mean (SD)	8 (7)
Scleroderma type	
Systemic	18 (95)
Limited	1 (5)
Marital status	
Single	2 (10)
Married	13 (68)
Separated	1 (5)
Divorced	3 (16)

depressed many participants. "When I'm in physical therapy, I hate it. It's just a lot of work and it depresses me." Side effects, physical health, and uncertainties about the future of their condition also worried participants. "It's just another bunch of pills and they...help you with one thing and bother the other." Worries and responsibilities weighed on participants as an overall burden.

Appearance. Altered physical appearance concerned many of the participants, who were most sensitive about disfiguration and discoloration of appendages and facial features. Participants described "smallness of the mouth and lips," blackened fingers, and bloated fingers that "look like sausages." Many feared the prospect and effects of amputations. "[T]he orthopedic surgeon wanted to amputate the whole leg...I wouldn't let him...I kind of lost it in his office!" "[R]eceding gum lines" and newfound gaps in teeth also affected participants' self-esteem. SSc altered participants' facial expressions. "[P]eople perceive you not being happy because you're not smiling...scleroderma gives you a mask-like appearance." Other people's perceptions mattered a great deal. "Children will ask, '...[W]hat happened to your fingers' and that type of thing...I kind of avoid being around children. Or, I [make a] fist all the time so they can't see my hands." Those participants without noticeable disfigurement expressed relief. "I know what I'm happy about is that I don't have...disfiguration in [my] face... My face doesn't look at all like a scleroderma patient's."

Lifestyle. All participants reported significant social and lifestyle disruptions, considered by many as the worst consequences of their disease. GI involvement, problems with cold

Table 3. Original and emergent codes in order from most to least recurrences.

	Original Codes	Emergent Codes
≥ 100 quotes		Pain (122)
		Self-efficacy/coping (100)
50–99 quotes	Energy/fatigue (94)	
	Problems skin (87)	
	Lifestyle (67)	Appearance (66)
		Compare over time (60)
	Problems eating (58)	
	Problems fingers (53)	
	Problems lungs (52)	
	Perception treatment improvement (50)	
25–49 quotes	Problems bowels (49)	Patient-doctor relations (49)
	Perception cause (49)	
	Problems stomach (43)	
	Problems joints (36)	
	Disease flaring (34)	
	Problems sleeping (32)	Compare others (32)
	Disease stable (29)	
	Lifestyle work (28)	
	Worst problems (28)	
	Emotional problems — fear (26)	
	Perception cause (26)	
	Bothers most about disease (25)	
	Physical problems (25)	
	Know (25)	Worries side effects (25)
		Worries uncertainties (24)
		Knowledge others (23)
		Burden of illness (23)
	Expectation cure (23)	
	Lifestyle sexual (22)	
	Disease change better (21)	
	Disease change worse (21)	
	Expectation disease change over time (20)	
	Physical problems — mobility (19)	Social issues — support (19)
15–24 quotes	Problems skin disease activity (19)	Social issues — family (18)
	Problems — heart (18)	Social issues others (18)
	Lifestyle — leisure (17)	Social issues — marriage (17)

Some participants included several aspects in a single response. Therefore, our totals exceed one response per each of 19 participants.

temperatures, fatigue, and physical appearance were all major determinants of social disruption.

Marital/sexual problems. SSc strained relationships with spouses; for some, it was a contributing factor to marriage dissolution. "My husband had to deal with my ups and downs...[H]e decided he wanted to be separated...I think that was very, very hard to deal with." Participants described how SSc complicates sexual intimacy. "There were times...when it wouldn't and then sometimes...I just couldn't do it because...I would just be so achy and everything." One participant decided not to have children because of SSc. Others praised their spouses for unconditional support and for providing basic aid and care. "I have a husband that doesn't mind mopping or sweeping or folding clothes." Nonetheless, participants felt guilty about increasingly having to depend on others. "My husband deserves more. And I can't fake it and I'm too dry to do anything about it."

Social issues. Although our original codes predicted lifestyle issues — daily activities of patients with SSc — they did not anticipate the effects on social relationships. Participants relied upon friends, support groups, and other networks. "Most of my friends are very good...For me, church has done a lot of good for me." SSc impeded friendships and social activity. "I can't keep up [with kids my age]...[It makes me feel] like I'm ruining everybody's time." Some participants also identified other people — and their perceived insensitivity — as a source of stress. Participants reported that others misunderstood and mistook their illness. "[If you need to park] in the handicapped area...[you have] to deal with people who come up to you and say, 'You don't look handicapped to me...Why are you parking there?'..." Pity bothered participants as well. "Not one relationship is the same because even friends... you wonder if they're sitting there...[feeling] sorry for you."

Table 4. Worst problems identified by participants.

Ranking	Main Code	Subordinate Codes	Quote Total
1	Lifestyle		24 total
		General	14
		Family	4
		Marriage*	3
		Work	3
2	System complaints		18 total
		Pain	7
		Bowel function	3
		Raynaud's*	2
		Skin tightness*	2
		Hands*	1
		Eating—esophagus*	1
		Lungs*	1
		Sicca symptoms*	1
3	Emotional		14 total
		Helplessness	6
		Uncertainty	5
		Fear	3
4	Energy		9 total
		Fatigue	5
		Lack of strength	4
5	Dependency*		4 each
	Finance*		
7	Appearance		3
8	Social life*		2 each
	Burden*		
10	Sleep*		1 each
	Public awareness*		
	Patient-doctor awareness*		

* Tie; some participants included several aspects in a single response. Therefore, some of our totals exceed one response per each of 19 participants.

Participants were also asked to rank the 3 worst SSc-related problems; we counted and categorized the most frequent responses into original and emerging codes. Some participants included several aspects in a single response; therefore, our totals exceeded 57 counts (or 3 responses per each of 19 participants).

Table 4 lists the 10 worst problems in order of frequency. Lifestyle concerns topped the list of worst problems. More participants (13 of 19) ranked it first among worst problems, above symptom complaints, which were the second worst problem. Emotional issues were the third worst problem — 3 of 19 participants ranked it their most troublesome problem. Other worst problems mentioned at least twice included lack of energy, dependency, financial problems, appearance, social life, and burden of illness.

Theme Three: disease activity and progression

Regarding disease progression, participants were shown graphs depicting various disease courses (Figure 2) and asked whether they felt their SSc was getting better or worse and whether they could tell when symptoms would flare. Most participants who responded (41%) chose graph C (progressive disease with superimposed flares). Graph A (slow, steady progression) and graph D (rapid progression and stabilization) were each chosen by 24% of respondents; 11% chose graph B (flares and remissions).

Participants reported knowing when their SSc was worsening moreso than when it was improving. Many attributed improvements to better treatment and measured recovery by improved physical ability and appearance. "My hands were black all the time and my feet...And now...look how nice and pink I am." Those for whom the disease worsened measured the downturn by symptom intensity and rate of decline. "Once I got Raynaud's then [the decline] seemed to start pretty quickly after that...My hands were swollen and tight. My skin was very shiny...[I]t is changing quickly for somebody that has worked out all their life." Participants reported that flares could last anywhere from 3 days to 3 months.

Theme Four: symptoms

We coded 447 reports of the following symptoms (general or organ-specific): pain (122), GI system (101), energy level (94), skin involvement (87), distal extremities (53), and lungs (52).

Pain. Most participants complained about pain (an emergent code). Many had widespread pain. "I pretty much had pain in most of my joints most of the time." Some reported localized musculoskeletal aches and pains. Still others described skin pain: aching, pinching, burning, tingling, and tightness. "I mean it's just like your skin is being ripped." Others reported pain associated with Raynaud's. "When they turn purple, it hurts and you feel like a tightening and a tingling sensation, like when your foot falls asleep." Some complained of GI and digestive pain.

GI system. Participants reported trouble swallowing and digesting particular foods. Constipation, diarrhea, pain, and bloating affected lifestyle (particularly in social interactions) and caused fear and embarrassment. "I ain't never went to the bathroom so much in my life. [I]f I'm participating in something, I need to be close to...a restroom."

Energy/fatigue. Participants reported experiencing both physical and mental fatigue. "[M]y skin is so exhausted it just sits and hangs on the bone...[T]hat is so totally different from fatigue when I was not with scleroderma. Fatigue back then meant, 'Oh, I don't feel like going out'...Now fatigue is honking [your horn when driving]." The emotional and social burden of the disease also drained energy. "You don't feel like going out. You don't feel like doing anything. I've become a couch potato."

Skin. Participants described stinging, swelling, discoloration, and tightness; skin-related pain was a recurrent theme.

Distal extremities. Tightness, calcinosis, and ulcers affected dexterity ("[I am] not able to grab... [Y]ou think you got the grip and it just slides.") and caused excruciating pain ("The calcinosis hurts...in my toes, I can't walk, I can't wear sandals

'cause I'm scared something will touch it..."; "With the gangrene...the pain that you felt in your fingers as they were dying was so excruciating that you almost begged to say please cut it off").

Lungs. Coughing and shortness of breath hampered daily activities. "When you're short of breath it just kind of limits you at work and any activities that you do."

Theme 5: Expectations

Participants expressed both optimism and pessimism about finding a cure for SSc. Most participants doubted that a cure would be found in the next 10 years. "I'm not real optimistic because...nobody knows about it...And the monies aren't out there." Some expected better treatments. Others expressed frustration that medications only treated symptoms of SSc rather than the disease itself. "I am not taking anything for scleroderma. I'm taking for symptoms — like for Raynaud's, ...medication for my blood pressure...stuff for my stomach." Almost half the participants expressed their concerns about the side effects of their medications on internal organs. "It's just another bunch of pills...they help you with one thing and bother the other." Others said that the benefits of the drugs outweighed the risks. "[E]ven with some of the side effects that you can get from them that they're not as bad as what I would feel if I wasn't on the medicine." Most simply hoped their disease course would remain stable. "I hope I continue to level...[I have] a good doctor and...they're coming out with newer things." Others expected their condition would deteriorate, given perceived lack of viable cures. "[My SSc will] get worse. Hopefully not, but continue affecting the internal organs."

Other emergent themes

Patient-doctor relations. Overall, participants were satisfied with their physicians. "I'm pretty happy...I think I'm seeing the right people." However, many reported encounters with physicians who were ignorant of the disease, which resulted in a prolonged diagnosis. "It is very frustrating to go to doctors...with all the symptoms that I've had and for them to tell you that there is nothing wrong with you." Participants also found it important to build rapport with pharmacists. "[My pharmacists] are a lot of help to me. I know if I go in and have a question about any of my medicines, you know they're there for me and they know me, which is another thing."

Self-efficacy, coping, and adaptation. Participants reported the importance of physical and psychological coping strategies — another emergent code. "I think we can do tremendous amount...by controlling our lifestyle, getting rest when you need it, not pushing ourselves, eating right." Accentuating positive aspects of life helped many cope. " [Y]ou give up some things but you need to focus on what can you do and what gives you pleasure...and joy." Humor helped coping. "I love my [handicapped parking] sticker... I covet my little advantage and I pull right into my handicapped space, rain or shine." Accepting a diagnosis played an important role. "I've figured out that this is going to be with me the rest of my life so I...just gotta live with it [and] move on." Religious faith comforted others. "I have faith in God...[who] has all power and if God wants this thing cured He usually can do it like that."

DISCUSSION

Patients with SSc report significant symptomatic and emotional burdens that affect their QOL and psychological well-being. We identified new areas of disease and symptom burden, including concerns about physical pain, coping skills, social pressures, physical appearance, and the patient-doctor relationship. These findings suggest that social, mental, and personal concerns — such as relationships with family and friends, emotional well-being, physical appearance, and coping skills — are as important to SSc patients as physiological issues.

A few previous studies support many of our findings. SSc-related pain has been shown to be a major symptomatic problem associated with low physical functioning and health outcomes[18-21]. Physical appearance has factored prominently as a concern for other study participants with SSc[22]. Severity of symptoms has been associated with coping, body image, and depression; anxiety and depressive symptoms have been associated with pain, feelings of helplessness, low working ability, low social activity, and minimal sense of coherence[19,20,23-30].

Joachim and Acorn performed another, smaller focus group investigating disease symptoms and QOL burden in patients with SSc[31]. In their study, participants reported difficulties with (1) the rarity of their disease, (2) shortness of breath and other physical manifestations, and (3) coping with physical abnormalities and social stigma. Our findings confirm and extend this work. Although both their and our studies uncovered similar patient concerns (e.g., physical appearance, disclosure to others, living normal lives, and facing the future), our study yielded more finely detailed analysis of these overarching categories. For example, participants in both studies were conscious of being different from others without SSc; our participants also stressed how they compared themselves to other patients with more and less severe SSc, sometimes as a coping mechanism. Participants in both the previous study[31] and ours encountered people in the general public who were unaware of SSc; our participants also dealt with physicians who were ill-informed about the disease. Social marketing campaigns and continuing education activities for primary care physicians might help increase public and provider awareness of the disease, and thereby relieve some of the burden SSc patients bear from the lack of recognition of their disease.

In the past decade, there has been increased emphasis on the use of generic QOL measures. While these have obvious advantages (particularly in comparing health across different disease populations), they may overlook some domains of

interest to patients with rare multisystem diseases such as SSc. Disease-specific measures primarily assess the severity of physical and functional impairment[32-39]. However, only one tool, the SHAQ, has evaluated symptom distress, but only using single-item visual analog scales. Other than pain, no other physical symptoms are generally assessed in generic instruments such as the Medical Outcome Study Short-Form 36. Some studies have correlated the effects of specific SSc symptoms with psychological well-being, including body image and depression[19,20,23,25,40], and coping and distress[41]. GI dysfunction and complications[30,42], physical pain[18], and SSc-related sleep deprivation[43] also contribute to patient depression. While these studies determined to what degree depression, coping, and other psychosocial dimensions do, indeed, occur in patients with varying degrees of scleroderma, they do not detail the lived experiences that contribute to negative psychosocial impact. Future studies might attempt to include a more holistic picture of SSc dysfunction by incorporating some of the domains of greatest distress identified here. Although controversy remains whether or not psychosocial and emotional issues impede or worsen treatment outcomes, many studies indicate that positive emotional and psychosocial influence can improve treatment adherence and outcomes — e.g., in patients with heart failure[44,45]. However, it is clear from our findings that functionality is affected by emotional and psychosocial impairments.

Our study has limitations. Most of our patients had diffuse disease. Patients of varying disease duration and severity experience different functionality and health, and therefore may have different perceptions of their illness and areas of difficulty and concern[21,46,47]. However, comparing diffuse and limited disease was outside the scope of our research. Our goal was to identify major areas of concern for patients, areas that are likely to be more encompassing and severe in patients with diffuse disease (the majority of our sample). Further, while ethnicity and socioeconomics may influence the presentation and immunogenetics of SSc[48-51] — which, in turn, might influence QOL — our small sampling of participants prevented us discussing how demographic factors played a part in code frequency. Future studies with larger samples can assess how ethnicity, gender, and socioeconomic background bear upon the identified recurrent and emergent themes, since the size of our study did not allow us to assess the effect of education, occupation, or marital status on patients' assessment of symptom burden, disease activity, quality of life, and disability.

Our findings extend the body of knowledge of SSc by identifying the importance of areas of disease burden that have not been adequately assessed in the past, such as pain and GI symptoms. Our study also uncovered the significance of coping skills, social pressures, and physical appearance, and the importance of the patient-doctor relationship. Our participants reported difficulty not only in accomplishing mundane tasks, such as grooming, but also in fulfilling longer-term personal choices, such as planning vacations and having children; their difficulties had an obvious influence on their subsequent happiness and sense of well-being. Our findings emphasize the need for an integrated interdisciplinary approach to the care of patients with SSc, involving physicians, physical and occupational therapists, psychologists, and social workers. The SHAQ and other function assessment tools have been used in conjunction with depression and pain scales to assess how individual organ problems disrupt quality of life[24,52,53]. However, our findings suggest that functional problems compound to create accumulative burden. Future work might include developing new and revised tools to measure cumulative impact on quality of life. Additional research is needed to develop, validate, and implement relevant patient measures that can more fully assess overall disease and symptom burden in SSc.

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REFERENCES

1. Kirwan J, Heiberg T, Hewlett S, et al. Outcomes from the Patient Perspective Workshop at OMERACT 6. J Rheumatol 2003;30:868-72.
2. Merkel PA, Herlyn K, Martin RW, et al. Measuring disease activity and functional status in patients with scleroderma and Raynaud's phenomenon. Arthritis Rheum 2002;46:2410-20.
3. Khanna D, Furst DE, Clements PJ, et al. Responsiveness of the SF-36 and the Health Assessment Questionnaire Disability Index in a systemic sclerosis clinical trial. J Rheumatol 2005;32:832-40.
4. Merkel PA, Clements PJ, Reveille JD, Suarez-Almazor ME, Valentini G, Furst DE. Current status of outcome measure development for clinical trials in systemic sclerosis. Report from OMERACT 6. J Rheumatol 2003;30:1630-47.
5. Furst DE. Update on clinical trials in the rheumatic diseases. Curr Opin Rheumatol 1998;10:123-8.
6. Furst DE, Khanna D, Mattucci-Cerinic M, Silman AJ, Merkel PA, Foeldvari I. Scleroderma — developing measures of response. J Rheumatol 2005;32:2477-80.
7. Poole JL, Steen VD. The use of the Health Assessment Questionnaire (HAQ) to determine physical disability in systemic sclerosis. Arthritis Care Res 1991;4:27-31.
8. Poole JL, Williams CA, Bloch DA, Hollak B, Spitz P. Concurrent validity of the Health Assessment Questionnaire Disability Index in Scleroderma. Arthritis Care Res 1995;8:189-93.
9. LeRoy EC, Black C, Fleischmajer R, et al. Scleroderma (systemic sclerosis): classification, subsets and pathogenesis. J Rheumatol 1988;15:202-5.
10. Zeller RA. Combining qualitative and quantitative techniques to develop culturally sensitive measures. In: Ostrow D, Kessler R, editors. Methodological issues in AIDS behavioral research. New York: Plenum Press; 1993:95-116.
11. Fern EF. The use of focus groups for idea generation: the effects of group size, acquaintanceship, and moderator on response quantity and quality. J Mark Res 1982;19:1-13.
12. Morgan DL. Designing focus group research. In: Stewart M, editor. Tools for primary care research. Thousand Oaks, CA: Sage Publishing; 1992:177-93.
13. Stewart DW, Shamdasani PN. Focus groups: Theory and practice. Thousand Oaks, CA: Sage Publishing; 1990.

14. Morgan DL. Focus groups. Annu Rev Sociol 1996;22:129-52.
15. Glaser BG, Strauss A. The discovery of grounded theory: Strategies for qualitative research. Chicago: Aldine; 1967.
16. Jeon YH. The application of grounded theory and symbolic interactionism. Scand J Caring Sci 2004;18:249-56.
17. Strauss A, Corbin J. Basics of qualitative research. Thousand Oaks, CA: Sage Publishing; 1998.
18. Benrud-Larson LM, Haythornthwaite JA, Heinberg LJ, et al. The impact of pain and symptoms of depression in scleroderma. Pain 2002;95:267-75.
19. Richards HL, Herrick AL, Griffin K, Gwilliam PD, Loukes J, Fortune DG. Systemic sclerosis: patients' perceptions of their condition. Arthritis Rheum 2003;49:689-96.
20. Richards H, Herrick AL, Griffin K, Gwilliam PD, Fortune DG. Psychological adjustment to systemic sclerosis — exploring the association of disease factors, functional ability, body related attitudes and fear of negative evaluation. Psychol Health Med 2004;9:29-39.
21. Sultan N, Pope JE, Clements PJ. The Health Assessment Questionnaire (HAQ) is strongly predictive of good outcome in early diffuse scleroderma: results from an analysis of two randomized controlled trials in early diffuse scleroderma. Rheumatology Oxford 2004;43:472-8.
22. Paquette DL, Falanga V. Cutaneous concerns of scleroderma patients. J Dermatol 2003;30:438-43.
23. Benrud-Larson LM, Heinberg LJ, Boling C, et al. Body image dissatisfaction among women with scleroderma: extent and relationship to psychosocial function. Health Psychol 2003;22:130-9.
24. Khanna D, Clements PJ, Furst DE, et al. Correlation of the degree of dyspnea with health-related quality of life, functional abilities, and diffusing capacity for carbon monoxide in patients with systemic sclerosis and active alveolitis: results from the Scleroderma Lung Study. Arthritis Rheum 2005;52:592-600.
25. Legendre C, Allanore Y, Ferrand I, Kahan A. Evaluation of depression and anxiety in patients with systemic sclerosis. Joint Bone Spine 2005;72:408-11.
26. Matsuura E, Ohta A, Kanegae F, et al. Frequency and analysis of factors closely associated with the development of depressive symptoms in patients with scleroderma. J Rheumatol 2003;30:1782-7.
27. Pincus T, Swearingen C, Wolfe F. Toward a multidimensional Health Assessment Questionnaire (MDHAQ): Assessment of advanced activities of daily living and psychological status in the patient-friendly Health Assessment Questionnaire format. Arthritis Rheum 1999;42:2220-30.
28. Roca RP, Wigley FM, White B. Depressive symptoms associated with scleroderma. Arthritis Rheum 1996;39:1035-40.
29. Van Lankveld W, Teunissen H, Vonk M, Van den Hoogen F. Coping and cognitions as predictors of psychological distress in scleroderma. Ann Rheum Dis 2005;64 Suppl:900.
30. Trezza M, Krogh K, Egekvist H, Bjerring P, Laurberg S. Bowel problems in patients with systemic sclerosis. Scand J Gastroenterol 1999;34:409-13.
31. Joachim G, Acorn S. Life with a rare chronic disease: the scleroderma experience. J Adv Nurs 2003;42:598-606.
32. Clements PJ. Measuring disease activity and severity in scleroderma. Curr Opin Rheumatol 1995;7:517-21.
33. Clements PJ, Wong WK, Hurwitz EL, et al. Correlates of the Disability Index of the Health Assessment Questionnaire: a measure of functional impairment in systemic sclerosis. Arthritis Rheum 1999;42:2372-80.
34. Clements PJ, Hurwitz EL, Wong WK, et al. Skin thickness score as a predictor and correlate of outcome in systemic sclerosis: high-dose versus low-dose penicillamine trial. Arthritis Rheum 2000;43:2445-54.
35. Clements PJ, Wong WK, Hurwitz EL, et al. The Disability Index of the Health Assessment Questionnaire is a predictor and correlate of outcome in the high-dose versus low-dose penicillamine in systemic sclerosis trial. Arthritis Rheum 2001;44:653-61.
36. Georges C, Chassany O, Mouthon L, et al. Validation of French version of the Scleroderma Health Assessment Questionnaire (SSc HAQ). Clin Rheumatol 2005;24:3-10.
37. Johnson SR, Gladman DD, Schentag CT, Lee P. Quality of life in scleroderma: Compared to psoriatic arthritis, SLE, rheumatoid arthritis and healthy controls. J Rheumatol 2004;31:1425.
38. Ostojic PS, Damjanov NS. The Scleroderma Assessment Questionnaire (SAQ); A new self-assessment questionnaire for evaluation of disease status in patients with systemic sclerosis. Z Rheumatol 2006 Mar 3. [Epub ahead of print]
39. Ruof J, Bruhlmann P, Michel BA, Stucki G. Development and validation of a self-administered systemic sclerosis questionnaire (SySQ). Rheumatology Oxford 1999;38:535-42.
40. Danieli E, Airo P, Bettoni L, et al. Health-related quality of life measured by the Short Form 36 (SF-36) in systemic sclerosis: correlations with indexes of disease activity and severity, disability, and depressive symptoms. Clin Rheumatol 2005;24:48-54.
41. Hansdottir I, Malcarne V, Furst D, Weisman M, Clements P. Relationships of positive and negative affect to coping and functional outcomes in systemic sclerosis. Cog Ther Res 2004;28:593-610.
42. Nietert PJ, Mitchell HC, Bolster MB, Curran MY, Tilley BC, Silver RM. Correlates of depression, including overall and gastrointestinal functional status, among patients with systemic sclerosis. J Rheumatol 2005;32:51-7.
43. Prado GF, Allen RP, Trevisani VM, Toscano VG, Earley CJ. Sleep disruption in systemic sclerosis (scleroderma) patients: clinical and polysomnographic findings. Sleep Med 2002;3:341-5.
44. Moser DK. Psychosocial factors and their association with clinical outcomes in patients with heart failure: why clinicians do not seem to care. Eur J Cardiovasc Nurs 2002;1:183-8.
45. Joynt KE, Whellan DJ, O'Connor CM. Why is depression bad for the failing heart? A review of the mechanistic relationship between depression and heart failure. J Card Fail 2004;10:258-71.
46. Gelber AC, Wigley FM. Disease severity as a predictor of outcome in scleroderma. Lancet 2002;359:277-9.
47. Steen VD. Treatment of systemic sclerosis. Am J Clin Dermatol 2001;2:315-25.
48. Arnett FC, Cho M, Chatterjee S, Aguilar MB, Reveille JD, Mayes MD. Familial occurrence frequencies and relative risks for systemic sclerosis (scleroderma) in three United States cohorts. Arthritis Rheum 2001;44:1359-62.
49. Callahan LF, Smith WJ, Pincus T. Self-report questionnaires in five rheumatic diseases: comparisons of health status constructs and associations with formal education level. Arthritis Care Res 1989;2:122-31.
50. Reveille J, Fischbach M, McNearney T, et al. Systemic sclerosis in 3 US ethnic groups: A comparison of clinical, sociodemographic, serologic, and immunogenetic determinants. Semin Arthritis Rheum 2001;30:332-46.
51. Reveille JD, Durban E, Goldstein R, Moreda R, Arnett FC. Racial differences in the frequencies of scleroderma-related autoantibodies. Arthritis Rheum 1992;35:216-8.
52. Georges C, Chassany O, Mouthon L, et al. [Quality of life assessment with the MOS-SF36 in patients with systemic sclerosis]. Rev Med Interne 2004;25:16-21.
53. Denton CP, Black CM. Scleroderma — clinical and pathological advances. Best Pract Res Clin Rheumatol 2004;18:271-90.