

The Symptom Burden Index: Development and Initial Findings from Use with Patients with Systemic Sclerosis

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ABSTRACT. Objective. Our study had 3 aims: (1) to evaluate the functioning of the Symptom Burden Index (SBI) in patients with systemic sclerosis (SSc); (2) to determine the amount of burden per problem experienced by patients as well as the number of patients experiencing each measured problem area, and the number of SSc problems per patient; and (3) to characterize the burden profiles of problem area-specific subgroups of patients.

Methods. We developed the SBI to determine the effect of problems in 8 major symptomatic areas of importance to patients (skin, hand mobility, calcinosis, shortness of breath, eating, bowel, sleep, and pain).

Results. Sixty-two patients with SSc completed questionnaires on current disease-related problems, physical functioning, and health status. On average, patients were 53.4 years old and had had SSc for 8 years. Patients were mainly women (87%), English-speaking (87%), with diffuse SSc (63%), white (69%), married (61%), and lived with 1 or more additional household members (84%). Only 26% were employed full-time. The 3 most widely reported problem areas were pain, hand, and skin, experienced by 92%, 89%, and 88%, respectively. About one-third reported experiencing 0–5 problems and one-third 7–8 problems; individual patients experienced, on average, 5.7 problems.

Conclusion. Psychometric evaluation determined that (1) summarizing SBI problem area item sets to report burden scores per problem measured is justified; (2) the 8 proposed problem areas are independent and deserve separate evaluation; and (3) burden scores correlate as expected with the Health Assessment Questionnaire-Disability Index and the Medical Outcomes Study Short-Form 36 questionnaire. The number of problems experienced and the degree of problem-associated burden that patients with SSc bear are substantial. Use of the SBI's patient-focused measurements may aid physicians in resolving problems most directly affecting patients' quality of life. This approach to measuring symptomatic burden in patients with chronic disease could be extended to other conditions. (First Release June 1 2010; J Rheumatol 2010;37:1692–8; doi:10.3899/jrheum.090504)

Key Indexing Terms:

SYSTEMIC SCLEROSIS
PATIENT CARE MANAGEMENT

SELF-ASSESSMENT
PAIN MEASUREMENT

Patients with systemic sclerosis (SSc) experience a multi-system disease affecting many aspects of their daily lives.

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Supported by a grant from the Scleroderma Foundation. Dr. Suarez-Almazor is the director and principal investigator of the Houston CERT, one of the Centers for Education and Research on Therapeutics, a multicenter research program funded by the Agency for Healthcare Research and Quality.

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Managing care to achieve the best possible quality of life for these patients involves close and careful monitoring of disease status. Care provision can be complex, with individual patients presenting with distinct complications and disease-related problems varying in type, duration, and severity. Several measures have been proposed to monitor disease progression and response to treatment in patients with SSc, most being objective organ-specific assessments or physical function assessments^{1–6}. Patient-reported outcomes have also been used in this population, including the Health Assessment Questionnaire-Disability Index (HAQ-DI), which provides measures for dressing, arising, eating, walking, hygiene, reach, grip, activities, and pain^{7,8}, and the Medical Outcomes Study Short-Form 36 questionnaire (SF-36), which provides measures for physical functioning, role-physical, bodily pain, general health, vitality, social functioning, role-emotional, and mental health. These

self-reported measures correlate with objective functionality, effect of disease, and quality of life. For patients with SSc, disability and depression are also common problem areas and correlate with quality of life⁹⁻¹². In fact, subjective views of function and disease-related problems have been shown to be highly reliable in measuring the effect of disease on patients' quality of life¹³. Yet patients with SSc have a myriad of symptoms and functional impairments related to the unique features of the disease (e.g., skin tightening) that may not be adequately addressed by the currently used instruments, which are quite generic and do not systematically identify multisystem symptomatic burden. Our qualitative work suggests that patients with SSc are highly burdened by symptoms of their disease, and we have identified 8 major symptomatic or problem areas of high importance to patients: skin problems, hand mobility problems, calcinosis-type problems, shortness of breath, eating problems, bowel problems, sleep problems, and pain¹⁴. Our work also suggests that the burden of any 1 problem can be multifaceted and depends not only on its intensity but also on its frequency, on the amount and frequency of interference with daily activities, and on the perceived importance of the problem to the patient. We have therefore developed the Symptom Burden Index (SBI), which ascertains these disease-related problems across multiple structures to determine their total effect on patients.

We had 3 objectives: (1) to evaluate the functioning of the SBI in patients with SSc; (2) to determine the amount of burden per problem experienced by patients as well as the number of patients experiencing each measured problem area, and the number of SSc problems per patient; and (3) to characterize the burden profiles of problem area-specific subgroups of patients.

MATERIALS AND METHODS

Instrument development. The SBI is configured as an 8-problem area index and includes the following SSc-related areas: skin, hand, calcinosis, shortness of breath, eating, bowel, sleep, and pain. Each problem area is measured independently by 5 items: (1) How much of a problem was __? (2) How often was __ a problem? (3) How much did __ interfere with daily activities? (4) How often did __ interfere with daily activities? (5) How important a problem was __? Responses are given using a 0 to 10 anchored response scale (Table 1). Left-side anchors use the descriptors Not a problem, Never a problem, or Not important, with 0 indicating no burden or burden importance. Right-side anchors use the descriptors Very severe problem, Very frequently a problem, or Very important, with 10 indicating the greatest amount, frequency, or importance of a burden.

Table 1. The Scleroderma Burden Index (SBI): “shortness of breath” items.

Problem Area	Shortness of Breath												
1. How much of a problem was shortness of breath?	Not a problem	0	1	2	3	4	5	6	7	8	9	10	Very severe problem
2. How often was shortness of breath a problem?	Never a problem	0	1	2	3	4	5	6	7	8	9	10	Very frequently a problem
3. How much did shortness of breath interfere with daily activities?	Not a problem	0	1	2	3	4	5	6	7	8	9	10	Very severe problem
4. How often did shortness of breath interfere with daily activities?	Never a problem	0	1	2	3	4	5	6	7	8	9	10	Very frequently a problem
5. How important a problem was shortness of breath?	Not important	0	1	2	3	4	5	6	7	8	9	10	Very important

Patient sample and survey questionnaire. The Baylor College of Medicine Institutional Review Board approved our study. A list of patients was obtained from the Genetics versus Environment Outcome in Scleroderma Study (GENISOS), a local study group sponsored by The Scleroderma Foundation and the University of Texas Health Science Center at Houston. Patients were recruited in person during their regular visits with the GENISOS study or were mailed information regarding our study, asking them to participate.

Study participants completed a self-response questionnaire including sociodemographic information (age, sex, ethnicity, language, marital status, number of household members, education, and occupation), the SBI, the HAQ-DI, and the SF-36; basic information about patient SSc status was obtained from medical chart review. Patients did not undergo examinations at the time of questionnaire completion, as their study participation period, i.e., the time during which they provided responses to questionnaires such as the SBI, occurred between regularly scheduled physician visits.

Psychometric evaluation of the SBI. The SBI provides as its primary measurements the amount of patient burden associated with each of 8 potential problem areas. SBI score, reflecting burden amount, is measured uniformly across problem areas, using common item content (i.e., there are 8 each of the How much, How often, How much interfere, How often interfere, and How important items). The SBI can also provide 2 additional measurements: (1) the number of patients experiencing each SSc-related problem, and (2) the number of problems experienced by each patient.

Although the SBI functions largely as an index, the 5-item sets have the potential to display scale characteristics; therefore, we conducted item-level and score-level evaluations of item set functioning. For item-level evaluations, we first evaluated the 8 item sets' scale characteristics by examining each set's inter-item correlations and item-total score correlations. Next, we estimated internal consistency reliability (coefficient α) per set. We then compared average, minimum, and maximum item scores across the 8 reported-on problem areas, identifying the item content of minimum-scoring and maximum-scoring items and obtaining observed score ranges per problem area. Finally, we compared minimum and maximum item scores per items of common content (e.g., the 8 “How much of a problem was __?” items). We identified the problem area associated with each minimum-scoring and maximum-scoring item and obtained observed score ranges per common-content item. For the score-level evaluations, burden scores per SSc problem area were obtained by averaging responses to each problem area's 5 items. We first correlated burden scores with each other to examine the nature and magnitude of interproblem relationships and to assess the extent to which burden scores were independent. We then correlated burden scores with HAQ-DI scores (total score) and SF-36 scores [Physical Component Score (PCS) and Mental Component Score (MCS)] to evaluate anticipated relationships and to provide evidence of burden score validity.

Patient burden assessment using the SBI. Using patient responses to the SBI, we determined (1) the amount of burden per problem area experienced by a patient; (2) the number of patients experiencing each of the 8 targeted SSc problem areas; and (3) the number of areas experienced per individual patient. We used burden scores to characterize the amount of burden experienced across all patients and across specific patient sociodemographic and disease-status groups. Burden scores serve as the primary measurements to be derived from the SBI. These SBI scores are continuous in

nature and can range from 0 to 10. Unique burden scores are available per SSc problem area and are obtained by averaging the responses to each problem area's 5 items (e.g., shortness of breath items 1–5). Potential patient subgroup differences were examined using ANOVA. In addition, “restricted” burden scores were calculated per SSc problem; these scores included only patients who had reported experiencing some amount of burden caused by the specific problem in question.

For each SSc problem area, we classified patients into 2 groups: “no burden present” (responded “0” to items 1–4) and “burden present” (responded from “1” to “10” to 1 or more of items 1–4). Using this approach, all patients reporting some level of burden due to a specific problem area can be identified. Therefore, a “burden present” designation will not reflect amount of burden (e.g., mild, moderate, severe) but instead that a patient is, to some extent, experiencing symptomatic burden from a specific problem area. We then used the burden presence classification to determine the number of sampled patients experiencing specific problems (i.e., the distribution of patients per problem area).

We then created a “number of problems” variable ranging from 0 to 8 to reflect the number of SSc problems currently experienced by each patient (i.e., the distribution of problems per patient). We investigated the number of problems experienced per patient and per patient by sociodemographic and disease-related status. Potential patient subgroup differences were examined using ANOVA.

RESULTS

Sixty-two out of 70 patients (89%) with SSc agreed to participate, signed an informed consent document, and then completed survey questionnaires about their current disease-related problems, physical functioning, and health status. Patient sociodemographic and disease status characteristics are presented in Table 2. On average, patients were 53.4 years old (range 21–79) and had SSc for 8 years (range 2–36). Patients were predominantly women (87%), English-speaking (87%), and with diffuse SSc (63%). They tended to be white (69%), married (61%), and to live with 1 or more additional household members (84%). In general, patients were highly educated, with 41% having a bachelor's degree or higher. Only 26% of patients with SSc indicated they were employed full-time.

Psychometric evaluation of the SBI. Item-level evaluation by problem area: for the 8 problem area item sets (5 items per set), average inter-item correlations ranged from 0.69 to 0.93, average item-total score correlations ranged from 0.78 to 0.95, and internal consistency reliabilities (coefficient α estimates) ranged from 0.91 to 0.98. Highest average inter-item correlation, average item-total correlation, and reliability estimate were all associated with calcinosis; lowest average item correlations (inter-item and item-total) and reliability estimate were all associated with skin.

The highest average burden item score across problem areas was 5.1 (pain); the lowest average burden item score was 1.4 (calcinosis). Maximum individual item scores per problem area ranged from a low of 1.6 (calcinosis) to a high of 5.9 (pain); minimum item scores ranged from a low of 1.2 (calcinosis) to a high of 4.5 (hand). Observed item score ranges across SSc problems were as follows: 3.7 (average item scores); 4.3 (maximum item scores); and 3.3 (minimum item scores). Within-problem item

Table 2. Sociodemographic and disease-related status of patients with SSc (n = 62). One patient did not provide responses to all demographic items.

Characteristic	
Sex, n (%)	
Female	54 (87.1)
Male	8 (12.9)
Age, mean (SD) yrs	53.4 (12.5)
Ethnicity, n (%)	
African American	8 (12.9)
Spanish/Hispanic/Latino	7 (11.3)
White American	43 (69.4)
Other	4 (6.4)
Language, n (%)	
English	53 (86.9)
Spanish and English	8 (13.1)
Marital status, n (%)	
Partnered	38 (61.3)
Not partnered	24 (38.7)
Household members, n (%)	
Living alone	10 (16.4)
Living with others	51 (83.6)
Education, n (%)	
Up to high school	16 (26.2)
Trade school/some college	20 (32.8)
Bachelor's degree or higher	25 (41.0)
Occupation, n (%)	
Full or part-time employee	20 (32.8)
Homemaker/in school	13 (21.3)
Disabled	21 (34.4)
Unemployed/retired	7 (11.5)
Disease type, n (%)	
Limited	23 (37.1)
Diffuse	39 (62.9)
Disease duration, mean (SD) yrs	8.0 (5.8)

score ranges went from a low of 0.4 (calcinosis) to a high of 2.2 (skin).

Two items of specific item content received 7 of the 8 maximum item scores across SSc problem areas. The “How often” item received the maximum score for 4 areas (bowel, eating, pain, skin), while the “How important” item received the maximum score for 3 other problem areas (calcinosis, hand, shortness of breath). The “How much” item received the maximum score for 1 problem (sleep). For minimum scoring items, an item content pattern was also identified: 2 items of specific item content received all of the minimum scores per problem area. For 6 problems, the “How much interfere” item received the minimum score, while for the 2 remaining problems (bowel, sleep), it was the “How often interfere” item that received the minimum score.

Item-level evaluation by common-content item. Items were also summarized by the 5 common-content items (i.e., How much, How often, How much interfere, How often interfere, How important). Maximum item scores within common-content items ranged from 4.5 (How much interfere) to 6.0 (How often); minimum item scores ranged from 1.2 (How much interfere) to 1.6 (How important). Two problem

areas (hand, pain) received all 5 of the maximum common-content item scores: hand had the maximum score for 3 of the 5 items (How much interfere, How often interfere, How important); pain had the maximum score for the remaining 2 (How much, How often). One SSc problem (calcinosis) received all of the minimum common-content item scores. Score ranges within common-content items went from a low of 3.3 (How much interfere) to a high of 4.7 (How often).

Score-level evaluation. Table 3 presents the problem area burden score intercorrelation matrix. Except for 3 correlations involving shortness of breath (with calcinosis, pain, sleep), all interproblem correlations were statistically significant ($p < 0.05$), all positive and low to moderate in magnitude, and ranged from 0.25 (eating-calcinosis) to 0.66 (pain-sleep). No correlations were of such high magnitude to suggest that measurements associated with 1 problem area could adequately represent the measurements associated with another. Thus, burden scores across problems appeared independent.

Table 3 also presents correlations between burden scores and HAQ-DI and SF-36 (PCS, MCS) scores. Positive correlations between burden scores and HAQ-DI scores reflect the association of greater burden with greater HAQ-DI-specific problems, while negative correlations between burden scores and SF-36 component scores reflect the association of greater burden with worse physical or mental health status. Except for 2 correlations, again involving shortness of breath (with HAQ-DI, SF-36 MCS), all correlations were statistically significant ($p < 0.05$) and low to moderate in magnitude, ranging in absolute value from 0.26 (bowel-SF-36 MCS and eating-HAQ-DI) to 0.68 (pain-SF-36 PCS). Correlations between burden scores and SF-36 PCS and SF-36 MCS also provided a means of examining whether SSc problems were more physical or mental in nature. Five problems displayed stronger physical natures through their correlations with SF-36 component scores: bowel (−0.46 with PCS vs −0.26 with MCS), calcinosis (−0.38 with PCS vs −0.34 with MCS), pain (−0.68 with PCS vs −0.43 with MCS), shortness of

breath (−0.39 with PCS vs −0.20 with MCS), and skin (−0.48 with PCS vs −0.29 with MCS). Three problems displayed stronger mental natures: eating (−0.38 with MCS vs −0.31 with PCS), hand (−0.52 with MCS vs −0.36 with PCS), and sleep (−0.51 with MCS vs −0.39 with PCS).

Patient burden assessment using the SBI. To determine the amount of burden and association with patient characteristics, we investigated differences in average problem area SBI burden scores by patient sociodemographic and disease-status groups, using ANOVA. Patients differed statistically significantly ($p < 0.05$) by age, marital status, and occupation-related patient groups. For patient age groups, older patients (56+ years) reported being less burdened by eating (2.2 vs 4.0) and skin problems (3.5 vs 5.2) than younger patients (< 55 years old). For patient marital status groups, patients who were married or had a partner reported being less burdened by bowel problems (3.1 vs 4.8) and calcinosis (0.7 vs 2.6) than not-partnered patients (single, separated, divorced, or widowed). For patient occupation groups, employed or in-school patients reported being less burdened by calcinosis (0.3 vs 2.7), hand problems (4.2 vs 6.0), pain (3.8 vs 6.7), skin problems (3.6 vs 5.4), and sleep problems (2.4 vs 5.8) than unemployed, retired, or disabled patients. No statistically significant differences were observed in the amount of burden experienced by patients with up to 5 years' disease duration versus those with 6 or more years.

Table 4 presents restricted SBI burden scores (i.e., burden scores calculated per problem area based on patient subsamples including only patients reporting at least some level of burden from the particular problem of interest; patients reporting no burden were excluded from the problem's restricted burden score calculations). The highest, second highest, and third highest restricted burden scores within any burden profile were consistently associated with pain, hand, or skin. Pain was the worst burden for 7 of 8 profiles and second worst for the remaining 1; hand was the worst

Table 3. Burden score correlations between problem areas and with SF-36 (PCS, MCS) and HAQ-DI (n = 62). All correlations statistically significant ($p < 0.05$), unless otherwise noted.

	Bowel	Calcinosis	Eating	Hand	Pain	Breath*	Skin	Sleep
Calcinosis	0.39							
Eating	0.54	0.25						
Hand	0.31	0.31	0.35					
Pain	0.52	0.41	0.36	0.57				
Breath*	0.40	0.00 [†]	0.51	0.26	0.21 [†]			
Skin	0.43	0.44	0.42	0.59	0.49	0.34		
Sleep	0.40	0.48	0.32	0.43	0.66	0.18 [†]	0.50	
SF-36 PCS	−0.46	−0.38	−0.31	−0.36	−0.68	−0.39	−0.48	−0.39
SF-36 MCS	−0.26	−0.34	−0.38	−0.52	−0.43	−0.20 [†]	−0.29	−0.51
HAQ-DI	0.38	0.52	0.26	0.46	0.59	0.23 [†]	0.58	0.51

* Shortness of breath. [†] Nonsignificant. SF-36 PCS: Medical Outcomes Study Short-Form 36 Physical Component Score; MCS: Mental Component Score; HAQ-DI: Health Assessment Questionnaire-Disability Index.

Table 4. Burden profiles: burden scores when patients with SSc report specific problems (n = 62).

Problem	n	Burden Scores							
		Bowel	Calcinosis	Eating	Hand	Pain	Breath*	Skin	Sleep
Bowel	44	5.3 ³	1.8	3.8	5.7 ²	6.0 ¹	4.4	5.1	4.8
Calcinosis	16	5.3	5.4	4.6	6.6 ²	6.8 ¹	4.0	6.2	6.3 ³
Eating	39	4.6	2.0	5.0	5.8 ²	6.0 ¹	4.4	5.2 ³	4.6
Hand	55	3.9	1.6	3.3	5.7 ¹	5.6 ²	3.6	4.6 ³	4.2
Pain	57	4.0	1.5	3.4	5.4 ²	5.6 ¹	3.7	4.7 ³	4.4
Breath*	41	4.6	1.4	4.1	5.6 ²	5.8 ¹	5.4 ³	5.1	4.5
Skin	54	4.1	1.5	3.4	5.5 ¹	5.5 ¹	4.0	5.0 ³	4.2
Sleep	45	4.5	1.9	3.8	5.8 ²	6.3 ¹	4.3	5.2	5.5 ³

* Shortness of breath. ¹ Highest burden score per problem area. ² Second-highest burden score per problem area.

³ Third-highest burden score per problem area.

burden for 2 profiles and second worst for 6; skin was the third worst burden for 4 of 8 profiles. Patients reporting a calcinosis burden displayed the most challenging of burden profiles: these patients reported the highest restricted burden scores recorded for 6 of 8 SSc problem areas measured; they reported the second-highest restricted burden scores recorded for an additional problem area (eating).

Table 5 presents the number (and percentage) of patients experiencing each specific SSc problem. The 3 most widely reported problem areas were pain, hand, and skin, experienced by 92%, 89%, and 88% of sampled patients, respectively. The 3 problems least widely experienced were shortness of breath, eating problems, and calcinosis, which were experienced by 66%, 63%, and 26% of patients, respectively. That 16 patients (26%) reported experiencing some degree of problem with calcinosis reflects the perception of patients that they are having calcinosis-type problems; it is not an expression of a confirmation of a clinical diagnosis of calcinosis. Calcinosis was the only SSc problem experienced by fewer than half of the sampled patients.

Number of problems and associations with patient characteristics. The number of problems experienced by individual patients ranged from 0 to 8. Approximately one-third of patients reported experiencing 0–5 SSc problems, nearly one-third reported 6 problems, and about one-third reported 7–8 problems. The average number of problems experi-

enced by individual patients was 5.7. Differences in the average number of problems experienced by patient sociodemographic and disease-status groups were investigated using ANOVA. Patients differed statistically significantly ($p < 0.05$) only by marital status, with partnered patients experiencing on average fewer problems (5.2, $n = 37$) than not-partnered patients (6.4, $n = 24$). No statistically significant differences were observed in number of problems experienced by patients with up to 5 years' disease duration versus those with 6 or more years.

DISCUSSION

Our primary aim was to develop and test a patient-reported outcome instrument measuring burden of illness in patients with SSc. The instrument was developed on the basis of a qualitative study that identified 8 primary areas of symptomatic concern for patients with SSc.

The psychometric evaluation of the proposed measure, the SBI, suggests that the proposed areas for study, although associated with each other, are relatively independent and deserve separate evaluation. Item-level testing of the SBI also suggests it is reasonable to summarize item sets to report a single average burden score per problem measured. Inter-item and item-total score correlations per item set were all moderate to high, and internal consistency reliability estimates were high; these scale characteristics reflect the small to moderate item score ranges observed per item set: from 0.4 to 2.2. It is not coincidental that the highest reliability estimate was associated with the problem area having the smallest item score range (calcinosis), while the lowest estimate was associated with the problem area having the largest range (skin).

Patients with SSc responded to SBI items using the full range of item responses available (0–10). As a result, average item scores per problem area showed considerable variability, ranging from 1.4 to 5.1. Minimum and maximum item scores per problem also displayed variability and revealed the sources of average score variability, with minimum item scores ranging from 1.2 to 4.5, and maximum scores from 1.6 to 5.9.

Table 5. Number of patients per SSc problem (n = 62).

SSc Problem	Patients	
	n	%
Pain	57	91.9
Hand	55	88.7
Skin	54	88.5
Sleep	45	72.6
Bowel	44	71.0
Breath*	41	66.1
Eating	39	62.9
Calcinosis	16	25.8

* Shortness of breath.

Specific item content was consistently associated with minimum and maximum scoring items per SSc problem. “How much interfere” and “How often interfere” were minimum scoring items for all measured problems; “How often” and “How important” were maximum scoring items for 7 of 8 problems, while “How much” was the maximum scoring item for 1 problem. The fact that interference items routinely received lower scores than amount, frequency, and importance items may reflect patient experience that the SSc burden itself remains the fundamental concern, and that burden interference, important and real as it is, is nevertheless secondary. The variety of observed maximum scoring items (How often, How much, How important) reinforces the idea that the SSc burden can be multifaceted.

Examination of minimum and maximum item scores per common-content item serves to confirm findings about item response variability and the distribution of the SSc problem burden. Minimum item scores per common-content item ranged from 1.2 to 1.6, and all were associated with calcinosis, the least widely experienced problem; maximum item scores ranged from 4.5 to 6.0 and were associated with hand and pain, the 2 most widely experienced problem areas. Score ranges within common-content items were considerably greater than score ranges within SSc problem (3.3–4.7 vs 0.4–2.2). This greater variability among common-content items accurately reflects expected greater differences across problems than within problems.

Item-level evaluations also suggest that the relationship between “How much” and “How often” items (and between “How much interfere” and “How often interfere” items) varies across problems and persons. For some problems, how much a problem is a burden may be a function of how often that problem is a burden. For patients burdened by calcinosis, there was a clear tendency to report similar burden levels across the “How much” and “How often” items: 11 of 16 patients (69%) reported identical burden levels on these 2 items. Skin provides a counterexample. For patients with a skin burden, only 18 of 54 (33%) reported identical burden levels across the 2 items. Therefore, while some SSc burdens may be more straightforward to measure, in that patients seem to either experience them or not (as appears to be the case with the burdens of calcinosis, hand, and shortness of breath), other burdens may be more complex, with their full weight formed through a combination of the amount and frequency of patient experience. Evidence here suggests that this is more likely the case with the burdens of bowel, eating, pain, skin, and sleep problems.

Score-level evaluations support the utility of the SBI. Burden score intercorrelations indicate that, while burden scores share some variability across problems, they are independent: no between-problem correlation was of sufficient magnitude to imply that measurement of 1 problem’s level of burden would represent measurement of another’s (maximum between-problem area correlation 0.66 for pain-

sleep). Score-level evaluations also provide initial evidence supporting SBI burden score validity. Correlations between burden scores and HAQ-DI, SF-36 PCS, and SF-36 MCS scores were in the expected direction and of anticipated magnitude to suggest a clear relationship between the level of specific SSc burdens experienced by patients and patient-reported physical functioning and health status. SSc burden does not equal physical functioning or health status, yet it will likely influence them. Thus, the range of correlations observed between burden scores and HAQ-DI (0.23 to 0.59), SF-36 PCS (–0.31 to –0.68), and SF-36 MCS (–0.20 to –0.52) are supportive of SBI score validity. Additionally, examining correlations between burden scores and SF-36 component scores (PCS, MCS) provides a means of investigating relationships involving burdens expected to be either more physical or mental in nature. The following relationships were observed as expected: sleep was more strongly related to MCS than to PCS; bowel, pain, shortness of breath, and skin were more strongly related to PCS than to MCS.

Burden scores represent the degree of burden experienced by patients with SSc. The near-ubiquitous pain, hand, and skin problems among patients with SSc are also the problems contributing the greatest amount of burden to these patients as a group. The observation of occupation group status differences in the calcinosis, hand, pain, skin, and sleep burdens might be considered less a finding than a known-groups validation of the SBI: the working able are expected to be less problem-burdened than the disabled.

Several marital status differences in the degree of burden (bowel, calcinosis) were observed. The partnered patient group reported being less burdened than the not-partnered group; social support realities may play a role. Observed differences between age groups revealed that the older patient group reported being less burdened by eating and skin problems than the younger group. These differences may reflect successful accommodation or may be related to natural disease regression.

The extent of burden experienced by patients with SSc is remarkable. In sheer number of problem areas, nearly two-thirds of sampled patients (64%) reported currently experiencing a minimum of 6 SSc-related problems; more than a third (36%) reported experiencing at least 7 out of the 8 problems measured. Eleven patients (18%) reported being burdened by each of the SBI’s 8 measured problems. In terms of specific problem areas, pain, hand, and skin burdens are essentially ubiquitous among patients with SSc; indeed, no measured problem was found to be rare or uncommon among patients: 26% reported experiencing calcinosis, the least commonly experienced problem, while 63% to 92% of the sampled patients reported being burdened by the 7 other problems. Why partnered patients reported experiencing fewer burdens than not-partnered patients was not readily apparent from our data. However, it is reasonable to think that partnered patients experiencing

particular SSc-related problems may have considered themselves less burdened by the problems as a result of support provided by their partners.

Restricted burden scores are burden scores derived using only those patients reporting experiencing some degree of burden from the specific problem in question. Although not used for characterizing group-level distribution or experience of problems, these scores are helpful for characterizing the actual level of burden experienced by the patients who suffer from a specific problem. In addition, the status of these patients regarding other problems they experience and the burden they associate with those problems can be informative. Within each "problem-experienced" patient group, the 3 highest burden scores were identified. Across all 8 problem profiles, the 3 highest burden scores tended to be associated with pain, hand, and skin. For 7 of the 8 SSc problem areas, patients reported experiencing pain as their worst burden; for all 8 of the SSc problem areas, patients reported experiencing hand problems as either their second worst or worst burden; for 4 of the SSc problem areas, patients reported experiencing skin problems as their third worst burden. These findings correspond to the reported group-based information. Of special interest, however, are the results obtained when viewing the specific burden profiles (one at a time) associated with patients burdened by a particular problem. Calcinosis was identified as the least commonly experienced problem from among the 8 problem areas measured by the SBI. Yet calcinosis-burdened patients reported the highest level of burden recorded for 6 of 8 SSc problem areas (bowel, calcinosis, hand, pain, skin, sleep) and the second-highest level of burden recorded for 1 other problem area (eating). Although less commonly experienced, calcinosis, when it is experienced, is associated with a burden profile that here was the most severe of all 8 profiles.

Our study has several limitations. SSc is a rare disease and the restricted sample size constrained the types of psychometric and comparative analyses we could conduct. A second limitation concerns our sample's composition and the degree to which it is representative of the larger SSc population. Representativeness appears to be less of an issue regarding age and sex composition, but may be more so regarding education level and, in particular, ethnicity. Finally, many of our patients had high pain scores. But our study was not planned to evaluate concomitant causes of chronic pain, such as fibromyalgia. Indeed, symptom burden was high overall in this patient group. Because this is a preliminary study and our patient sample was small, we were not able to reliably detect whether reported symptoms were more likely to occur in patients with shorter disease duration or those with longer duration and organ damage.

The SBI was developed as a direct result of our qualitative work involving the study of the personal experiences of patients living with SSc. The 8 symptomatic or problem areas measured by the SBI are those identified by patients with SSc as the most relevant and most critical to establish-

ing and maintaining their quality of life. Initial psychometric evidence suggests that the SBI provides internally consistent burden scores, that burden scores are independent across SSc problems measured, and that these burden scores correlate as expected with measurements derived from 2 extensively studied and widely used instruments, the HAQ-DI and the SF-36. The number of problems experienced by patients with SSc and the degree of problem-associated burden that they bear is substantial. Use of the patient-focused measurements derived from the SBI may help physicians providing care to patients with SSc to know and act to resolve those problems most directly affecting their patients' quality of life. Moreover, this approach to measuring symptomatic burden in patients with chronic disease could be extended and applied to investigating the burden of patients experiencing other disease conditions.

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