# Development and validation of a selfadministered systemic sclerosis questionnaire (SySQ)

# J. Ruof, P. Brühlmann, B. A. Michel and G. Stucki<sup>1</sup>

Rheumaklinik und Institut für Physikalische Medizin, Universitätsspital, Zürich, Switzerland and <sup>1</sup>Klinik für Physikalische Medizin und Rehabilitation, Ludwig-Maximillian Universität, München, Germany

# Abstract

Objective. To develop a self-administered systemic sclerosis questionnaire (SySQ) covering condition-specific functional limitation and symptoms.

Methods. An initial item pool was generated by open patient interviews. A preliminary questionnaire was devised using 62 systemic sclerosis (SSc; scleroderma) patients. Factor analysis was used for further selection and grouping of items into distinct scales. The retrieved scales were tested for internal consistency and test–retest reliability. Spearman's rank correlation and Wilcoxon's rank sum test were used to examine hypothesized associations of the SySQ with various clinical and laboratory features.

Results. Altogether 32 SySQ items were selected and aggregated into 12 scales addressing 'pain', 'stiffness', 'coldness', 'complex functions', 'strength of hands', 'rising', 'walking', 'shortness of breath', 'upper airway symptoms', 'eating', 'swallowing' and 'heartburn/ regurgitation'. Internal consistency ranged from 0.93 ('complex functions') to 0.73 ('heartburn/ regurgitation'); Spearman's correlation coefficient for test–retest reliability ranged from 0.93 to 0.73 (P < 0.001). While the scales were associated with corresponding functional impairments, there was generally less association with morphological impairments.

Conclusion. The SySQ is a valid and reliable condition-specific measure in patients with SSc. Individually applicable scales cover a wide spectrum of general and organ-specific SSc symptoms and functional limitation. After further validation with respect to its ability to measure change, it may be used in clinical, health services and epidemiological research.

KEY WORDS: Scleroderma, Questionnaire, Symptoms, Function.

Systemic sclerosis (SSc; scleroderma) is a connective tissue disease affecting a wide variety of organs [1], and placing a tremendous burden on both patients and society as a whole. Patients suffer from a wide spectrum of general and organ-specific symptoms, as well as functional limitation. Therefore, outcome assessment needs to address the different aspects of the disease comprehensively, both in terms of clinical measures and patient-perceived disease impact. Unfortunately, there is a lack of appropriate instruments allowing the study of treatment efficacy in SSc [2]. Few self-administered patient questionnaires have been validated for use in SSc. The Health Assessment Questionnaire (HAQ), which was developed to measure disability in rheumatoid arthritis patients, does not comprehensively address functional limitation of SSc patients [3–5]. The condi-

Submitted 22 June 1998; revised version accepted 29 January 1999. Correspondence to: G. Stucki, Klinik für Physikalische Medizin und Rehabilitation, Ludwig-Maximillian Universität, Marchioninistrasse 15, 81377 München, Germany.

tion-specific functional questionnaire devised by Guillevin and Ortonne [6] has not been tested for its reliability and validity characteristics. Silman et al. [7] recently published an 11-item SSc-specific disability assessment instrument. Although this instrument allows a simple method of measuring functional limitation in SSc, it showed poor agreement between patient assessment and physiotherapist (occupational therapist) assessment of functional performance. Additionally, it does not address the various aspects of organ involvement in SSc. An approach that includes such aspects in a patient questionnaire was presented by Steen and Medsger [5]. They suggested the use of visual analogue scales (VAS) to cover typical organ symptoms. However, the VAS do not cover the whole spectrum of SSc symptoms and, as previously shown by Ferraz et al. [8], the reliability of individual VAS was somewhat limited (test-retest correlation coefficient of pain and gastrointestinal scale < 0.7).

The objective of our study was to develop a comprehensive set of self-administered questionnaires covering

specifically the large spectrum of functional limitation and symptoms encountered in SSc patients.

# Patients and methods

## Item generation

As a conceptual framework for the design of the selfadministered systemic sclerosis questionnaire, we used Nagi's taxonomy of disability [9]. According to his definition, both impairment and functional limitation involve major disease symptoms; in impairment, reference is to the level of tissues and organs; in functional limitation, reference is to the level of the organism or the person as a whole. Adapting this concept for application in SSc, three of the authors (PB, BM and GS) identified major areas of concern in SSc patients: general disease symptoms, organ-specific symptoms (i.e. cardiopulmonary and gastrointestinal) and musculoskeletal symptoms with a focus on functional limitation of upper (i.e. hands and fingers) and lower extremities. The identification of these areas and of items likely to assess such symptoms was based on the experience of the authors and on the review of 15 patient charts of the University Hospital in Zurich. Further items were generated by interviewing 12 selected patients manifesting a wide spectrum of disease activity, severity and duration. While conducting these interviews, we also asked the patients for appropriate wording of the items. All identified items were grouped into a preliminary questionnaire which consisted of 113 questions.

Likert response scales similar to the HAQ were used to score the items and referred to the ability to perform an action (0 = without difficulty; 1 = with some difficulty; 2 = with much difficulty; 3 = unable to do), described the intensity of symptoms (0 = no; 1 = some; 2 = moderate; 3 = very intensive) or described the frequency of symptoms (0 = never; 1 = sometimes; 2 = frequently; 3 = always).

#### Patient selection and examination

Our study sample was derived from the 'Scleroderma Patient self-support group of the German-speaking part of Switzerland'. Sixty-two patients voluntarily participated in the examination. They were invited to attend an additional scheduled visit between September 1994 and April 1995 at the Rheumatology Outpatient Clinic of the University Hospital in Zurich. After completing the preliminary questionnaire, the patients were examined clinically and laboratory tests were carried out: pulmonary function tests, echocardiography, ultrasound of kidneys, chest X-ray, and radiographic investigations of oesophagus, hands and feet. The clinical examination was performed according to a standardized protocol by a rheumatologist at the University Hospital in Zurich and included assessment of total skin score [10], grip strength (Vigorimeter), fingertip-to-palm distance, range of motion of upper (phalangeal joints, wrists, elbow) and lower (i.e. toes, ankles and knees) extremities, and assessment of digital ulcers. During clinical examination, the plausibility of patients' perception of some functional aspects of the questionnaire was reviewed (e.g. are the patients able to rise from a chair without arms or put on their socks?). Oral manifestations of the disease were assessed by a specially trained surgeon at the Department of Oral Surgery at the University Hospital in Zürich.

#### Item selection

The item selection was performed by means of: (1) factor analysis; (2) test for internal consistency; (3) consideration of content and face validity. A factor analysis was used to group intercorrelated items into distinct clusters (factors or scales, respectively). For each of the four main categories (general and cutaneous, musculoskeletal, cardiopulmonary and gastrointestinal symptoms), a separate factor analysis was performed. Factors were only extracted if Kaiser–Meyer–Olkin Measure of Sampling Adequacy was >0.6 for single items with a summary value >0.75 and Bartlett's Test of Sphericity revealed highly significant results (P < 0.00001) [11]. The criterion for determining the number of factors was that the eigenvalue be >1 [12]. After extraction of factors, the factor loading matrix was examined for the distribution of variance, factor loading patterns and unipolarity. Finally, a rotation (which makes factors independent of each other statistically) was performed. Internal consistency of the retrieved scales was assessed using Cronbach's coefficient  $\alpha$ , which summarizes the inter-item correlations among all items in a scale. The whole procedure of scale development was performed independently by two of the authors (JR and GS). Retrieved scales and selected items were compared afterwards. While selecting and grouping of items into scales, aspects of content and face validity were repeatedly considered, i.e. each selected item was assessed for its relative importance according to the purpose of the scale. Additionally, we considered whether the scales revealed clinically meaningful and interpretable results.

Scale scores were calculated as the mean of non-missing values of remaining items. If more than one item per patient and scale revealed a missing value, the scale value was scored as missing. Categorical scores were calculated as the mean of the respective scale values.

#### Reproducibility

Test-retest reliability was examined on a subsample of 53 patients, between two time points, 2 weeks apart. To exclude a systematic bias, we visually assessed the histograms of difference (scale × test – scale × retest) for each of the scales following the approach suggested by Bland and Altman [13]. The strength of the linear association of test-retest scales was tested by Spearman's rank correlation coefficient.

## Criterion validity

To examine criterion validity, we analysed the association of the scales with disease parameters including parameters suggested in a preliminary SSc severity index [14]. For dichotomous parameters, Wilcoxon's rank sum test was used; all other item validations were

Table 1. Clinical characteristics of the study sample grouped into four categories: early diffuse disease (<3 yr duration from diagnosis), late diffuse disease (>3 yr), early limited disease (<5 yr), late limited (>5 yr)

	Number of patients/SSc category				
Variables	Early diffuse	Late diffuse	Early limited	Late limited	
Total number of patients	8	11	17	26	
Sclerodactyly	7	8	13	17	
Digital ulcers or pitting scars	4	6	2	14	
Anti-topoisomerase antibody	4	6	2	17	
Erosions and/or marginal erosions of hand and/or feet	6	7	16	20	
Osteolysis of hand and/or feet	6	6	9	18	
Hypomotility at cine-oesophagography	5	9	9	18	
Pulmonary fibrosis on chest radiography	2	8	6	9	
Clinical evidence of 'primary' pulmonary arterial hypertension	0	2	3	4	

performed by Spearman's correlation coefficient. All P values are reported two-tailed; P < 0.05 was considered significant.

#### Results

#### Patients

All 62 patients satisfied the 1980 preliminary criteria of the American College of Rheumatology (formerly American Rheumatism Association) for the diagnosis of SSc [15]. The age range was 24–77 yr (mean  $\pm$  s.d.: 53.8  $\pm$  13.8 yr); 51 patients (82.3%) were female, 11 (17.7%) were male. The mean time period since the diagnosis of SSc was  $8.0 \pm 7.3$  yr (minimum 25 days, maximum 31.9 years). Clinical characteristics are shown in Table 1.

#### Scale analyses

Twelve scales with 2–4 items each were identified. Thirty-two out of a total of 113 items were finally selected for inclusion in the scales. Seven of the selected items were

Table 2. SySQ scales and items. Bold items are derived from the HAQ. All items are scored from 0 to 3. Answering categories: A, ability to perform activities (0, without difficulty; 3, unable to do); B, intensity of symptoms (0, no symptoms; 3, very intensive symptoms); C, frequency of symptoms (0, never; 3, always)

Number and name of scale	Content of items	Answering category
General symptoms		
(1) Pain	Painful hands	C
	Painful fingers when touching or holding objects	В
(2) Stiffness	Stiffness of hands	В
	Stiffness of arms	В
	Stiffness of legs	В
(3) Coldness	Coldness of hands	C
	Painful hands if they are cold	В
	Painful feet if they are cold	В
Musculoskeletal symptoms		
(4) Complex functions	Ability to cut meat with a knife	A
. , ,	Ability to wash and dry oneself	A
	Ability to put on socks	A
	Ability to cream one's own body	A
(5) Strength of hands	Ability to turn a water tap on and off	A
	Weakness of hands when holding objects	C
	Frequent dropping of objects when holding them	C
(6) Rising	Rising from a chair without arms	A
	Ability to lie down and get up from a bed	A
(7) Walking	Ability to walk on a flat street	A
	Ability to climb stairs	A
Cardiopulmonary symptoms		
(8) Shortness of breath	Shortness of breath when walking along a street	В
(*)	Shortness of breath when climbing stairs	B
	(two flights of stairs with 10 steps each)	
	Shortness of breath when dressing	В
(9) Upper airway symptoms	Coughing	В
	Expectoration	В
	Limited inspiration	В
Upper gastrointestinal symptoms		
(10) Eating	Ability to eat an apple	A
(10) Latting	Ability to eat large pieces of food without cutting them up	A
(11) Swallowing	Difficulty with swallowing	Č
(11) Swanowing	Pain when swallowing	C
	When swallowing, food becoming stuck in the throat	C
(12) Heartburn/regurgitation	Heartburn	C C C
(12) Heartourn/regurgitation	Regurgitation	C
	reguigitation	C

derived from the HAQ. Thirty of the selected items only loaded on one of the 12 scales (factor loading on other factors <0.3). Two items loaded on more than one factor. Table 2 shows the 32 items, their content, and the grouping into 12 scales and four categories.

The psychometric properties for the individual and categorical scales are shown in Table 3. All scales have a satisfactory internal consistency. All inter-item correlations within each scale were statistically significant (P < 0.05). Cronbach's  $\alpha$  scores for each of the 12 individual scales were greater than the 0.65 required for group comparison.

#### Reproducibility and criterion validity

Visual assessment of the histograms of difference did not show a systematic bias. The strength of association was highly significant (P < 0.001) for all scales, ranging from 0.73 (strength of hands) to 0.93 (complex functions) (Table 3). The association of the scales with other disease parameters is shown in Table 4.

#### Excluded items

Eighty-one items from the preliminary questionnaire were excluded from the final version of the SySQ. Thirty-eight of these covered additional musculoskeletal symptoms and functional limitation, 16 covered general and cutaneous symptoms, nine cardiopulmonary symptoms and 10 gastrointestinal symptoms. Additionally, eight items on psychosocial consequences of SSc were excluded because they did not reveal a valid and reliable scale.

### Discussion

In contrast to most other rheumatic diseases, no comprehensive condition-specific questionnaire has been devised for use with SSc patients. Current concepts regarding the measurement of functional limitation in scleroderma patients mostly rely on the HAQ, which was developed to measure disability in rheumatoid arthritis [3-5]. Consequently, its content validity is limited when applied to SSc. Similar to the recently published disability assessment instrument [7], the SySQ has been developed for application in SSc patients and measures typical functional limitation. However, in contrast to the aforementioned disability assessment instrument, the SySQ includes distinct general, gastrointestinal and cardiopulmonary symptoms. As opposed to more general VAS covering these symptoms, as suggested by Steen and Medsger [5], the SySQ comprehensively and specifically covers a wide spectrum of organ involvement characteristic for SSc patients. While the limited reliability of individual VAS might influence their ability to capture treatment effects in clinical trials [8], the SySQ has been developed by aggregation of multiple items into highly reliable and internally consistent scales. Likert's fixed response answering system was used because of its advantages in terms of patient comprehension [16].

To test criterion validity of the SySQ, we examined the association with various clinical parameters. As expected, scales generally related more to functional than to morphological impairments. For example, the musculoskeletal symptom scales were associated with grip strength, finger-to-palm distance, and range of

Table 3. Psychometric properties of the 12 scales and the categorical scales. Test–retest reliability is reported with Spearman's rank correlation coefficient, all results were highly significant (P < 0.001)

Number and name of scale	Scale mean $\pm$ s.d.	Standard error of mean	Minimum/maximum of scale	Cronbach's coefficient α	Test-retest reliability
- Scale	mean ± s.b.	or mean	or scare	coemeient a	Tenaomity
General symptoms					
(1) Pain	$1.22 \pm 0.83$	0.11	0/3	0.75	0.81
(2) Stiffness	$0.98 \pm 0.83$	0.11	0/3	0.82	0.88
(3) Coldness	$1.67 \pm 0.82$	0.11	0/3	0.79	0.79
Categorical scale	$1.24 \pm 0.63$	0.08	0/2.58	0.78	0.91
Musculoskeletal symptoms					
(4) Complex functions	$0.61 \pm 0.72$	0.09	0/3	0.93	0.93
(5) Strength of hands	$0.88 \pm 0.63$	0.08	0/3	0.80	0.73
(6) Rising	$0.30 \pm 0.51$	0.07	0/2	0.83	0.82
(7) Walking	$0.49 \pm 0.53$	0.07	0/1.5	0.77	0.75
Categorical scale	$0.57 \pm 0.45$	0.06	0/2.06	0.75	0.88
Cardiopulmonary symptoms					
(8) Shortness of breath	$0.70 \pm 0.72$	0.09	0/2.67	0.82	0.82
(9) Upper airway symptoms	$0.67 \pm 0.80$	0.10	0/3	0.84	0.90
Categorical scale	$0.61 \pm 0.62$	0.08	0/2.50	0.69	0.91
Upper gastrointestinal symptoms					
(10) Eating	0.98 + 0.92	0.12	0/3	0.80	0.87
(11) Swallowing	$0.58 \pm 0.60$	0.08	0/2.67	0.84	0.87
(12) Heartburn/regurgitation	0.94 + 0.79	0.10	0/3	0.73	0.87
Categorical scale	0.84 + 0.59	0.08	0/2.89	0.65	0.89

Table 4. Association of SSc scales with functional and morphological impairments. Range LE and UE: absence/presence of range-of-motion deficits of upper and lower extremity joints, HAQ-DI (HAQ Disability index). Morphological impairments were assessed by X-ray of hands and feet (we separately assessed association of scales with three radiographic features: marginal erosions, osteolysis, and s.c. or periarticular sclerosis), TSS (total skin score [10]), chest X-ray and echocardiography (assessment for clinical evidence of pulmonary hypertension)

Number and name of scale	Functional impairments	Correlation	Morphological impairments	Significance
General symptoms				
(1) Pain	HAQ-DI	r = 0.55**	X-ray hand/feet	ns
(1) 1 am	Grip strength	r = 0.33 r = -0.30*	Digital ulcers	ns
	Fingertip-to-palm distance	r = 0.38**	TSS	r = 0.13  ns
	Range of motion LE	ns	155	7 0.15 115
	Range of motion UE	ns		
(2) Stiffness	HAQ-DI	r = 0.54**	X-ray hand/feet	ns
( )	Grip strength	r = -0.28*	Digital ulcers	ns
	Fingertip-to-palm distance	r = 0.30*	TSS	r = 0.18  ns
	Range of motion LE	*		
	Range of motion UE	**		
(3) Coldness	HAQ-DI	r = 0.38**	X-ray hand/feet	ns
(1)	Grip strength	r = -0.15  ns	Digital ulcers	ns
	Fingertip-to-palm distance	r = 0.19  ns	TSS	r = 0.19  ns
	i ingertip to puin distance	7 0.15 115	155	7 0.17 113
Musculoskeletal symptoms	- ·	,		
(4) Complex functions	Grip strength	r = -0.49***	X-ray hand/feet	ns
	Fingertip-to-palm distance	r = 0.67***	TSS	r = 0.41**
	Range of motion LE			
	Range of motion UE	**		
(5) Strength of hands	Grip strength	r = -0.43***	X-ray hand/feet	ns
	Fingertip-to-palm distance	r = 0.49***	TSS	r = 0.30*
	Range of motion LE	*		
	Range of motion UE	**		
(6) Rising	Grip strength	r = -0.22  ns	X-ray hand/feet	ns
	Fingertip-to-palm distance	r = 0.28*	Pulmonary hypertension	ns
	Range of motion LE	***	TSS	r = 0.12  ns
	Range of motion UE	ns		
(7) Walking	Grip strength	r = -0.30*	X-ray hand/feet	ns
	Fingertip-to-palm distance	r = 0.28*	Pulmonary hypertension	ns
	Range of motion LE	***	TSS	r = 0.26*
	Range of motion UE	ns		
Cardiopulmonary symptoms				
(8) Shortness of breath	Diffusing capacity	r = -0.24  ns	Pulmonary fibrosis	**
(-, -1101111100 01 01011111	Forced expiratory volume	r = -0.32*	Pulmonary hypertension	ns
	Vital capacity	r = -0.32 r = -0.17  ns	2 amonary hypertension	110
(9) Upper airway symptoms	Diffusing capacity	r = -0.17  Hs r = -0.37**	Pulmonary fibrosis	*
(,, oppor an way symptoms	Forced expiratory volume	r = -0.37 r = -0.40**	Pulmonary hypertension	ns
	Vital capacity	r = -0.40 r = -0.39**	2 differently hypertension	110
Upper gastrointestinal symptoms				at.
(10) Eating	Weight loss	ns	Cine-oesophagography	*
	Maximal oral aperture	r = -0.38**		
	Atrophy of chewing muscles	r = 0.48**		
(11) Swallowing	Weight loss	ns	Cine-oesophagography	*
	Maximal oral aperture	r = -0.22  ns		
	Atrophy of chewing muscles	r = 0.40**		
(12) Heartburn/regurgitation	Weight loss	*	Cine-oesophagography	*
	Maximal oral aperture	r = 0.15  ns		
	Atrophy of chewing muscles	r = -0.08  ns		

<sup>\*</sup>P < 0.05; \*\*P < 0.01; \*\*\*P < 0.001; ns = not significant.

motion of upper and lower extremities, while there was no association with radiographic changes of hands or feet. These findings are consistent with data reported by Blocka *et al.* [17], who found no correlation between clinical and radiographic changes in hands and feet of SSc patients. However, in comparison to other cohorts, our patients tended to manifest more radiographic changes. This may be due to the selection process

applied, whereby all patients were derived from the 'Patient self-support group of the German-speaking part of Switzerland'.

The three scales of pain, stiffness and coldness cover general SSc features that often require symptom-modifying treatment. The fact that these scales were associated with functional rather than morphological impairments is consistent with reversible dysfunctions.

This set of symptom scales may be useful in the study of dynamic vascular dysfunction and for the evaluation of protective measures such as heating gloves.

Cardiac and pulmonary involvement are among the most important predictors of survival in SSc patients [18]. The two SySQ scales covering cardiopulmonary symptoms may be useful in early detection and monitoring. Further research is needed to clarify whether these two scales are related to the two different patterns of pulmonary involvement recently described by Morelli et al. [19].

The three gastrointestinal scales cover problems localized at the level of the oral aperture, the pharyngooesophageal passage and oesophageal-gastric passage. Management of manifestations at these sites is an important factor in the treatment of SSc patients [20]. These scales may be helpful in the screening and localization of problems. They may be used as specific outcome measures when studying therapeutic interventions. The preliminary questionnaire also included items covering lower gastrointestinal tract symptoms such as diarrhoea and flatulence. Although these items may be useful in monitoring lower gastrointestinal tract involvement, they did not reveal a stable and reliable scale, and were therefore excluded from the final version of the SySQ.

The use of the individual scales allows for detailed documentation of individual patients and may be advantageous as end points in studies on problems represented by a specific scale. In other settings, it may be preferable to use summary scales. We thus developed summary scales for the four categories: general symptoms, functional limitation and musculoskeletal symptoms, cardiopulmonary symptoms, and gastrointestinal symptoms. Since the scales of each of these categories represent distinct though related constructs, the use of categorical scales seems appropriate. However, if groups were compared in randomized clinical trials, for example, one would need to ensure equal patterns of scale involvement and test for differential responses of scales.

While the use of a global score would be simple, we

TABLE 5. Clinical and epidemiological aspects of the SySQ

Clinical aspects	Epidemiological aspects
Advantages of the SySQ Condition-specific questionnaire Inclusion of a wide spectrum of functional limitation and symptoms	Development by factor analysis High internal consistency High reproducibility
Future research agenda Discriminative capacity Raynaud's phenomenon and renal involvement Comparison of SySQ musculoskeletal symptoms scale with Silman's disability assessment instrument [7]	Discriminative capacity Examination of interval properties of scales and item difficulties Cross-cultural adaptation
Applicability (after further validation Clinical trials Clinical quality management	ion) Epidemiological research Health services research

do not recommend such a procedure because of the lack of association among individual scales across categories, and because of the unresolved problems of how to weight scales or categories. Similar to concepts suggested in systemic lupus erythematosus [21], we recommend reporting in terms of scales or categorical profiles.

The SySQ was developed as a result of major input from patients, rheumatologists familiar with SSc and clinical epidemiologists. Despite this interdisciplinary approach, some problems have yet to be solved completely, and have been added to the future research agenda (Table 5). First, it was not possible to develop a scale addressing Raynaud's phenomenon, a key feature of SSc. Although there was a good correlation of Raynaud's phenomenon with the items 'itching' and 'dysaesthesia of feet when walking' (r ranging from 0.43 to 0.49), factor analysis did not reveal an internally consistent and valid scale. In accordance with Steen and Medsger [5], we would suggest assessing Raynaud's phenomenon using a VAS.

Second, renal involvement, a relevant predictor of survival in SSc patients [18], was not addressed. We screened for renal involvement clinically by ultrasound, measurement of serum creatinine and examination of urine sediment. The vast majority of patients did not show signs of major renal involvement, which is probably explained by appropriate antihypertensive medication of the study sample.

Third, the SySQ has been developed according to a cross-sectional design. It remains to be shown whether the SySQ scales are sensitive to change longitudinally, and whether they can discriminate between active and placebo treatment in early disease patients, who are the ones most likely to participate in disease-modifying trials. Until such data are available, the SySQ cannot be used to measure change in clinical trials. Additionally, a cross-cultural adaptation of the original German language questionnaire has to be undertaken if the SySQ is to be used in an international multicentre setting.

In conclusion, the SySQ is a valid and reliable condition-specific measure in patients with systemic sclerosis. Individually applicable scales cover a wide spectrum of general and organ-specific SSc symptoms and functional limitation. After further validation with respect to its ability to measure change, it may be used in clinical, health services and epidemiological research.

# Acknowledgements

We would like to thank Dr P. van Leeuwen for his critical comments, and Dr J. Hodler and Dr S. Duewell for reading the radiographs.

## References

- 1. Seibold JR. Scleroderma. In: Kelley WN, Ruddy S, Harris ED, Sledge CB, eds. Textbook of rheumatology. Philadelphia: WB Saunders, 1997:1133-62.
- 2. Pope JE, Bellamy N. Outcome measurement in scleroderma clinical trials. Semin Arthritis Rheum 1993; 23:22-33.

- 3. Fries JF, Spitz PW, Young DY. The dimensions of health outcomes: the Health Assessment Questionnaire, Disability and Pain Scales. J Rheumatol 1982;9:789–93.
- 4. 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.
- 5. Steen VD, Medsger TA. The value of the Health Assessment Questionnaire and special patient-generated scales to demonstrate change in systemic sclerosis patients over time. Arthritis Rheum 1997;40:1984–91.
- 6. Guillevin L, Ortonne JP. Traitement de la sclerodermie. Ann Med Interne (Paris) 1983;34:754–65.
- Silman A, Akesson A, Newman J et al. Assessment of functional ability in patients with scleroderma: a proposed new disability assessment instrument. J Rheumatol 1998;25:79–83.
- 8. Ferraz MB, Quaresma MR, Aquino LRL, Atra E, Tugwell P, Goldsmith CH. Reliability of pain scales in the assessment of literate and illiterate patients with rheumatoid arthritis. J Rheumatol 1990;17:1022–4.
- Nagi SZ. Disability concepts revisited: implications for prevention. In Pope AM, Tarlov AR, eds. Disability in America. Toward a national agenda for prevention. Washington, DC: National Academy Press, 1991:309–27.
- Clements PJ, Lachenbruch PA, Ng SC, Simmons M, Sterz M, Furst DE. Skin score. A semiquantitative measure of cutaneous involvement that improves prediction of prognosis in systemic sclerosis. Arthritis Rheum 1990;33:1256-63.
- 11. Brosius G, Brosius F. Faktorenanalyse. In: Brosius G, Brosius F, eds. SPSS Base System and professional statistics. Bonn: Thomson Publishing, 1995:815–46.
- Norman GR, Streiner DL. Principal components and factor analysis. In: Norman GR, Streiner DL, eds. Biostatistics, the bare essentials. St Louis, Mosby, 1994:129–42.

- Bland JM, Altman DG. Statistical methods for assessing agreement between two methods of clinical measurement. Lancet 1989;i:307–10.
- 14. Medsger TA, Silman AJ, Steen VD, International Scleroderma Study Group. Development of a severity index for systemic sclerosis. Arthritis Rheum 1994;37 (suppl. 9):S260 (Abstract).
- 15. Subcommittee for Scleroderma Criteria of the American Rheumatism Association Diagnostic and Therapeutic Criteria Committee. Preliminary criteria for the classification of systemic sclerosis (scleroderma). Arthritis Rheum 1980;23:581–90.
- Joyce JRB, Zutshi DW, Hrubes V, Mason RM. Comparison of fixed interval and visual analogue scales for rating chronic pain. Eur J Clin Pharmacol 1975;8:415–20.
- 17. Blocka KLN, Bassett LW, Furst DE, Clements PJ, Paulus HE. The arthropathy of advanced progressive sclerosis: A radiographic survey. Arthritis Rheum 1981;24:874–84.
- Altman RD, Medsger TA, Bloch DA, Michel BA. Predictors of survival in systemic sclerosis (scleroderma). Arthritis Rheum 1991;34:403-13.
- Morelli S, Barbieri C, Sgreccia A et al. Relationship between cutaneous and pulmonary involvement in systemic sclerosis. J Rheumatol 1997;24:81–5.
- Cohen S, Laufer I, Snape WJ, Shiau YF, Levine GM, Jimenez S. The gastro-intestinal manifestations of scleroderma: pathogenesis and management. Gastroenterology 1980;79:155–66.
- 21. Stoll T, Stucki G, Malik J, Pyke S, Isenberg DA. Association of the systemic lupus international collaborating clinics/American College of Rheumatology damage index with measures of disease activity and health status in patients with systemic lupus erythematosus. J Rheumatol 1997;24:309–13.

APPENDIX: Fragebogen Sklerodermie

	ohne Schwierig- kieten	mit leichten Schwierig- kieten	mit großen Schwierig- kieten	unmöglich
Können Sie				
—das Fleisch mit dem Messer schneiden?				
—sich ganz waschen und abtrocknen?				
—Socken anziehen?				
—sich selbst die Haut/Körper eincremen?				
—einen Wasserhahn auf- und zudrehen?				
—von einem Stuhl ohne Armlehne aufstehen?				
—ins Bett gehen und aufstehen?				
—auf ebener Straße gehen?				
—Treppen steigen?				
—einen Apfel essen?				
—größere Lebensmittelstücke essen ohne zu zerschneiden?				

	nein	leicht	mäßig	stark
—Haben Sie Schmerzen in den Fingern beim Berühren oder Halten				
von Gegenständen?				
—Haben Sie ein Steifigkeitsgefühl in den Händen?				
—Haben Sie ein Steifigkeitsgefühl in den Armen?				
—Haben Sie ein Steifigkeitsgefühl in den Beinen?				
—Bekommen sie bei Kälte Schmerzen in den Händen?				
—Bekommen sie bei Kälte Schmerzen in den Füßen?				
—Bekommen Sie beim Gehen auf ebener Straße Atemnot?				
—Bekommen Sie beim Treppensteigen (2 Treppen a ca 10 Stufen) Atemnot?				
—Haben Sie Atemnot beim Ankleiden?				
—Leiden Sie unter Husten?				
—Haben Sie Auswurf?				
—Haben Sie Mühe beim tiefen Durchatmen?				
	nie	manchmal	häufig	immer
	inc	manemmai	naung	
	_	_		
—Haben Sie beim Halten von Gegenständen eine Schwäche in den Händen?				
—Fallen Ihnen Gegenstände, die Sie halten, aus der Hand?				
<ul><li>—Fallen Ihnen Gegenstände, die Sie halten, aus der Hand?</li><li>—Verspüren Sie Schmerzen in den Händen?</li></ul>				
<ul><li>—Fallen Ihnen Gegenstände, die Sie halten, aus der Hand?</li><li>—Verspüren Sie Schmerzen in den Händen?</li><li>—Haben Sie kalte Hände?</li></ul>				
<ul> <li>—Fallen Ihnen Gegenstände, die Sie halten, aus der Hand?</li> <li>—Verspüren Sie Schmerzen in den Händen?</li> <li>—Haben Sie kalte Hände?</li> <li>—Haben Sie Mühe beim Schlucken?</li> </ul>				
<ul><li>—Fallen Ihnen Gegenstände, die Sie halten, aus der Hand?</li><li>—Verspüren Sie Schmerzen in den Händen?</li><li>—Haben Sie kalte Hände?</li></ul>				
<ul> <li>—Fallen Ihnen Gegenstände, die Sie halten, aus der Hand?</li> <li>—Verspüren Sie Schmerzen in den Händen?</li> <li>—Haben Sie kalte Hände?</li> <li>—Haben Sie Mühe beim Schlucken?</li> <li>—Haben Sie Schmerzen beim Schlucken?</li> <li>—Bleiben beim Schlucken Speisen im Hals stecken?</li> </ul>				
<ul> <li>—Fallen Ihnen Gegenstände, die Sie halten, aus der Hand?</li> <li>—Verspüren Sie Schmerzen in den Händen?</li> <li>—Haben Sie kalte Hände?</li> <li>—Haben Sie Mühe beim Schlucken?</li> <li>—Haben Sie Schmerzen beim Schlucken?</li> <li>—Bleiben beim Schlucken Speisen im Hals stecken?</li> <li>—Haben Sie Sodbrennen?</li> </ul>				
<ul> <li>—Fallen Ihnen Gegenstände, die Sie halten, aus der Hand?</li> <li>—Verspüren Sie Schmerzen in den Händen?</li> <li>—Haben Sie kalte Hände?</li> <li>—Haben Sie Mühe beim Schlucken?</li> <li>—Haben Sie Schmerzen beim Schlucken?</li> <li>—Bleiben beim Schlucken Speisen im Hals stecken?</li> </ul>				