

## A cross-sectional comparison of three self-reported functional indices in scleroderma

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**Objectives.** In scleroderma, outcome measures such as skin score provide only limited information about the functional impact of the disease. The requirement for validated and convenient instruments that reliably reflect disease morbidity is now recognized. This study compares the Disability Index of the Health Assessment Questionnaire (HAQ-DI) with two more recently developed scleroderma-specific tools: scleroderma-visual analogue scales (scleroderma-VAS) and the UK scleroderma Functional Score (UKFS). In addition, the use of clinical and laboratory measures as predictors of disease severity have been examined.

**Methods.** One hundred and fifteen consecutive patients were studied. Subjects completed the 20-item HAQ-DI, the scleroderma-VAS and a questionnaire related to hand and muscle function (UKFS). Clinical details, measurement of maximal hand-spread, fist-closure and investigations for internal organ involvement were recorded.

**Results.** Over 68% of patients with diffuse disease had moderate to severe disease on the UKFS, compared with 44% with limited disease. The mean UKFS in diffuse disease was 14.7 (S.D. 9.1) and 10.6 (S.D. 8.5) in the limited subset ( $P=0.02$ ). The mean HAQ-DI in diffuse disease was 1.23 (S.D. 0.77) and 0.79 (S.D. 0.75) in the limited subset ( $P=0.005$ ). The HAQ-DI showed significant correlation with UKFS ( $r=0.9$ ;  $P < 0.001$ ). Several clinical and laboratory measures were associated with higher HAQ-DI and UKFS.

**Conclusions.** This is the first comparative study of the UKFS and the HAQ-DI. These data show a strong correlation between assessment methods. Higher scores correlated with clinical and laboratory indicators of severe disease. Used together, these inexpensive tools assess general and organ-specific symptoms, as well as functional limitation.

KEY WORDS: Scleroderma, Functional indices, HAQ-DI, UKFS.

Scleroderma is a multisystem connective tissue disorder characterized by overproduction and deposition of collagen in the skin, involvement of the microcirculation and with the propensity to affect a number of visceral organs [1]. It is well recognized that internal organ involvement is common in scleroderma and that the extent of involvement determines mortality. Although it is recognized that scleroderma is a disorder with considerable morbidity, the degree of functional impairment

has not been well characterized. While earlier studies suggested 50% mortality at 5 yr [2], improved survival rates in scleroderma have been seen over the past few decades [3]. Therefore, functional capacity represents an increasingly important clinical problem. Bryans *et al.* [4] recently reported a 93% 5-yr survival, in the absence of an elevation in erythrocyte sedimentation rate (ESR), impaired lung function or proteinuria. This suggests that morbidity rather than mortality

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would appear to be the more relevant issue in many patients.

With the development of clinical trials to assess the effectiveness of new therapies for use in scleroderma, the requirement for validated and convenient instruments that reliably reflect disease morbidity in scleroderma is now recognized [5]. Outcome measures must be appropriate to the disease being evaluated. Traditionally, studies have chosen as outcomes the measurement of changes in the extent of pathology, such as skin score [6]. Mortality and its surrogates, including end-stage organ involvement, are frequently chosen as endpoints. However, patients with scleroderma have variable disease presentations and these measurements may not be of great value in evaluating the relative effectiveness of different therapies. Indeed, disability may well be the most important category of outcome in scleroderma. Barst *et al.* [7] have recently illustrated that in the treatment of primary pulmonary hypertension with continuous ambulatory prostacyclin, benefit might be measured according to improvement or maintenance of functional capacity. They suggest that changes in the measurements of impairment of physiological function may be insensitive markers.

The Disability Index of the Health Assessment Questionnaire (HAQ-DI) is widely used to assess and monitor patients with rheumatic diseases; it measures physical disability by asking patients to rate their capacity to perform 20 activities of daily living, grouped into eight categories [8]. Over the past two decades its use in monitoring functional impairment in rheumatoid arthritis has been well documented. The literature delineating its use in scleroderma by comparison is relatively sparse [9]. Steen *et al.* [10] have supplemented the HAQ with visual analogue scales (VAS) to measure symptoms specific to scleroderma, these include pain, Raynaud's phenomenon, digital ulcers, gastrointestinal symptoms, pulmonary symptoms and overall disease severity. In this study Steen *et al.* [10] demonstrated that the modified HAQ had high face validity, good test-retest reliability and that the disability index of the HAQ predicted survival. More recently, a UK scleroderma Functional Score (UKFS) has been developed that focuses on disability caused by skin tightness in the upper limb and proximal muscle weakness [11]. Silman *et al.* [11] tested its reliability in 47 patients, good agreement was found between patients' first and second assessments, and between therapists. Nine of the 11 questions relate to upper limb function and as such have high face and content validity. The scleroderma-VAS and UKFS measure different aspects of functional impairment in scleroderma. The UKFS primarily measures hand function; whereas the visual analogue scales in addition determine the influence of internal organ involvement. The HAQ-DI is a global assessment tool that will therefore be influenced by both hand function and internal organ involvement, their relative contribution being difficult to discern. The UKFS is easier to use than the HAQ-DI and in addition it is more tailored

and discriminatory in assessing hand function in scleroderma.

Our purpose therefore was to correlate the HAQ-DI with two more recently developed scleroderma-specific tools: the scleroderma-VAS and the UKFS. In addition we have looked for predictors of disease severity.

## Patients and methods

### Patients

One hundred and fifteen consecutive scleroderma patients attending the connective tissue diseases clinic were studied. All patients had scleroderma satisfying the 1980 American Rheumatism Association criteria [12]. Demographic features including sex, age, presence of visceral organ involvement, the duration of Raynaud's phenomenon and the duration of scleroderma (taken as the date when skin changes were first noted by the patient) were documented.

### Diagnostic tests and organ involvement

Skin thickness was quantified using the modified Rodnan score [13] in which skin thickness was assessed clinically at standard sites on a 0–3 scale: 0=normal, 1=mild thickness, 2=moderate thickness, and 3=severe thickness. Maximal hand-spread was measured from the outermost aspect of the fifth finger to the outermost aspect of the thumb. Fist closure was recorded as the distance from the end of the right middle finger to the distal palmar crease during maximal fist closure (full closure was recorded as zero). The cut-offs chosen for variables such as hand spread and fist closure were taken as the median values; this is clearly an arbitrary choice as there are no accepted normal ranges for these variables. Reduced hand-spread was taken as less than 15.5 cm; reduced fist-closure as greater than 1.26 cm. The grade of Raynaud's phenomenon was assigned on a scale of 0–3: where 0=none, 1=possible, 2=probable, and 3=severe [14]. The presence of digital pitting, ulceration and gangrene was recorded. Haemoglobin, haematocrit, ESR, serum albumin, creatinine, creatinine kinase, C-reactive protein (CRP) and brachial blood pressure were measured in all patients.

Pulmonary function tests were performed, and the forced vital capacity (FVC) and carbon monoxide diffusion capacity (DLCO) recorded. Results were considered to be abnormal if they were <70% of the predicted value. The diagnosis of interstitial lung involvement was confirmed on high-resolution computed tomography. Pulmonary artery pressures were estimated by transthoracic Doppler echocardiography, an estimated peak systolic pressure of greater than 30 mmHg was taken as abnormal. Heart involvement was defined as a history of congestive cardiac failure, arrhythmia, pericarditis or moderate-to-large pericardial effusion. Renal involvement was defined by the occurrence of a scleroderma renal crisis or a serum creatinine level above the upper limit of normal. Muscle involvement was reported if two of the following criteria were present: symmetrical muscle weakness, biopsy evidence of myositis, elevation of serum creatinine kinase or an electromyographic pattern for myositis [15]. In addition, the finding of a raised creatinine kinase was recorded as muscle involvement, as it is well recognized that patients with scleroderma may have a mild subclinical myopathy.

Gastrointestinal involvement was based on a history suggestive of malabsorption or confirmed by investigations, such as hydrogen breath testing or barium studies of the small bowel. Oesophageal scintigraphy was used to quantify the degree of

dysfunction, ranging from grade 0 (normal) to grade 4 (severe abnormality). Oesophageal scintigraphy is a useful non-invasive screening test for the detection of asymptomatic disease, as it is well recognized that symptoms may be unreliable in judging the presence or extent of oesophageal disease in scleroderma. Grades 1 or 2 indicated mild oesophageal involvement, and 3 or 4 indicated severe involvement [16]. Anorectal disease was determined by history.

### Assessment questionnaires

Subjects completed the self-administered 20-item HAQ-DI, the scleroderma-specific VAS and the 11-item UKFS. The HAQ-DI is divided into eight components: dressing and grooming, arising, eating, hygiene, reach, grip and activity. Each component is divided into two or three questions. Questions are scored from 0 (without difficulty) to 3 (unable to do). A disability index is calculated by dividing the summed component scores by the number of components answered. The HAQ-DI falls between 0 and 3 on an ordinal scale [8]. An 11-item UK functional questionnaire (UKFS) includes nine items relating to upper extremity function and two items relating to muscle weakness and lower extremity function. Each item is scored under four categories with both an integer and a descriptive heading, ranging from 0 (able to perform in a normal manner) to 3 (impossible to achieve). The UKFS is scored on an ordinal scale of 0–33 [11]. The scleroderma-VAS includes five scales that assess pain, vascular symptoms (Raynaud's and digital ulcers), gastrointestinal symptoms, shortness of breath and overall disease severity. Patients are asked to indicate how much these problems have interfered with activities. The anchors of the VAS are 0 (did not interfere) and 3 (very severe limitation) [10].

Functional status was also categorized for each assessment tool as mild, moderate or severe, by dividing the ordinal scale into three equal parts; where mild functional impairment corresponded to HAQ-DI <1, VAS <1 or UKFS <11; moderate functional impairment HAQ-DI ≥1 and <2, VAS ≥1

and <2 or UKFS ≥11 and <22; and severe functional impairment HAQ-DI ≥2, VAS ≥2 or UKFS ≥22.

### Statistical analysis

The data were analysed using SPSS software. The  $\chi^2$ -test was used to analyse functional category according to disease subgroup for the HAQ-DI and UKFS, and each individual VAS is analysed by the Mann-Whitney U-test. The correlation between the functional indices was analysed by Spearman's correlation coefficient. Independent Student's *t*-tests were used to examine for independent correlates of functional status. Multivariate analysis was used to determine independent correlates of the UKFS and HAQ-DI. We explored the distribution of residuals obtained in the regression analysis for HAQ-DI and UKFS and found them to be normally distributed. Covariates included in the multivariate model included: FVC, DLCO, ESR, CRP, pulmonary artery pressure, albumin, skin score, hand spread, sex, disease subset and disease duration. As the scleroderma-VAS was not normally distributed, the outcomes were dichotomized and analysed by logistic regression.

### Results

One hundred and fifteen consecutive patients were studied, 40 had diffuse cutaneous scleroderma and 75 had limited disease. Demographic and clinical features are detailed in Table 1. Subjects were categorized as having mild, moderate or severe functional impairment based on their scores on the HAQ-DI, VAS and UKFS questionnaires. Scores for each assessment tool and the percentage assigned to each category are summarized in Table 2. Over 68% of patients with diffuse scleroderma had moderate to severe disease on the UKFS, compared with 43.1% of those with limited disease. The mean HAQ-DI was 1.23 in those with diffuse disease

TABLE 1. Details of the demographic and clinical features for the scleroderma subgroups

	Limited cutaneous scleroderma (n=75)	Diffuse cutaneous scleroderma (n=40)
Age (yr) [mean (s.d.); IQ range]	57 (12.6); 49–66.7	49.3 (12.8); 40–57
Sex ratio (female:male)	66:9	36:4
Duration of Raynaud's phenomenon (yr)	16.6 (12.8); 8.5–20.5	8.7 (7.7); 3–14
Duration of scleroderma (yr)	10.38 (9.7); 4–14	6.5 (5.2); 2–12
Skin score	7.6 (4.8); 4–10	23.0 (13.6); 11–32.5
Visceral organ involvement (%) <sup>a</sup>		
Mild oesophageal disease	69.3	72.2
Severe oesophageal disease	22.7	25.0
Gastrointestinal disease	14.7	10.0
Renal involvement	28	17.5
Cardiac involvement	10.7	7.5
Myositis	20	37.0
Pulmonary hypertension	21.3	17.5
Pulmonary fibrosis	16	67.5
Maximal hand spread (cm)	16.1 (2.8); 15–18	14.5 (3.3)
Fist closure (cm)	0.75 (1.3); 0–1	2.2 (1.9)
Haemoglobin (g/dl)	12.6 (1.6); 11.7–13.9	12.3 (1.4); 11.2–13.4
Haematocrit	38.2 (4.4); 35–42	37.4 (3.8); 35–40
ESR (mm/h)	23.9 (19.4); 10–30	27.9 (23.5); 10–40
Albumin (g/l)	41.3 (4); 39–44	40.6 (4.8); 38.2–44.8
C-reactive protein (mg/l)	9.5 (16.3); 2–12.5	12.0 (15.2); 3.2–13.8
Diastolic blood pressure (mmHg)	75.9 (10.3); 70–80	75.9 (9.5); 70–80
Forced vital capacity (mean % predicted value)	88.5 (26.3); 81–105	74.2 (17.7); 61–90.3
DLCO (mean % predicted value)	67.3 (20.9); 57–81	55.1 (14.2); 44.5–64

<sup>a</sup>Details of individual organ assessment are included in methods section.

TABLE 2. Functional scores and categories assigned according to diffuse and limited scleroderma (SSc) disease subgroups

	Limited SSc	Diffuse SSc	P value
HAQ-DI mean value (s.d.) <sup>a</sup>	0.79 (0.75)	1.23 (0.78)	0.005*
% in HAQ-DI functional category:			
Mild (0 to <1)	62.5	36	
Moderate ( $\geq 1$ to <2)	29.2	46	
Severe ( $\geq 2$ )	8.3	18	
UKFS mean value (s.d.) <sup>a</sup>	10.6 (8.5)	14.7 (9.1)	0.02*
% in UKFS Functional category:			
Mild (1 to <11)	56.9	31.4	
Moderate ( $\geq 11$ to <22)	27.7	45.7	
Severe ( $\geq 22$ )	15.4	22.9	
Individual VAS mean/s.d. <sup>b</sup> (expressed as percentage)			
Pain	39.7 (29.0)	47.4 (31.9)	0.25
Gastrointestinal	31.1 (29.9)	25.6 (29.9)	0.40
Shortness of breath	24.6 (28.9)	35.2 (30.8)	0.17
Raynaud's phenomenon	38.3 (31.0)	39.1 (30.0)	0.7
Digital ulcer	17.0 (27.3)	20.9 (30.9)	0.7
Overall severity	47.5 (31.9)	49.5 (28.9)	0.84

\* $P < 0.05$ .

<sup>a</sup>UKFS and HAQ-DI ordinal score analysed by unpaired *t*-test; UKFS and HAQ-DI functional categories analysed by  $\chi^2$ -test.

<sup>b</sup>VAS analysis by Mann-Whitney U-test.

compared with 0.79 in the limited group (95% CI 0.14, 0.76;  $P=0.005$ ). Likewise the UKFS was significantly higher in those with diffuse disease: the mean UKFS was 14.7 compared with 10.6 in the limited group ( $P=0.02$ ). The VAS for pain, gastrointestinal, respiratory and vascular symptoms and overall disease severity did not differ between the two subgroups.

When the assessment tools were compared, the UKFS and HAQ-DI showed excellent correlation,  $r=0.9$  (Fig. 1). The VAS for overall disease severity and pain showed good correlation with the UKFS and HAQ-DI, where  $r$  was  $>0.6$  (Table 3). Correlation between VAS vascular symptoms, digital ulcers, respiratory and gastrointestinal symptoms did not show good correlation with the UKFS and HAQ-DI. Correlations between the three functional indices are detailed in Table 3.

Several clinical and laboratory variables associated with higher UKFS and/or HAQ-DI scores. These variables were FVC  $<70\%$ , skin score  $\geq 15$ , fist closure

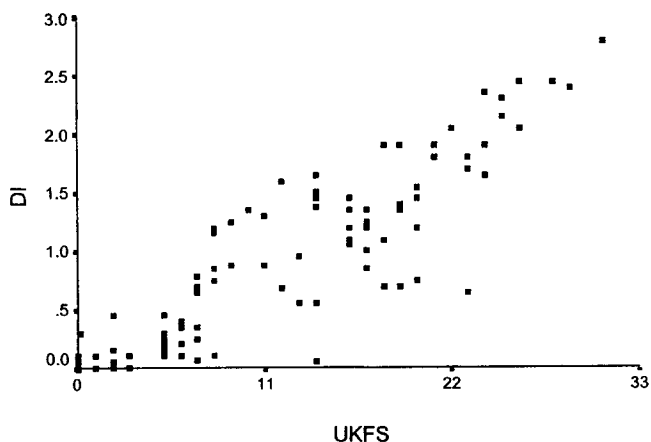


FIG. 1. Correlation of the UKFS and HAQ-DI. Pearson's correlation coefficient ( $r=0.9$ ).

TABLE 3. Details of the correlation between the three functional indices in this cohort of patients with scleroderma

	HAQ-DI	95% CI	UKFS	95% CI
Pain VAS	0.66***	0.53, 0.76	0.66***	0.53, 0.76
Gastrointestinal VAS	0.4***	0.22, 0.55	0.45***	0.28, 0.6
SOB VAS	0.51***	0.35, 0.64	0.53***	0.37, 0.66
RP VAS	0.46***	0.29, 0.6	0.52***	0.36, 0.65
Digital ulcer VAS	0.21*	0.02, 0.39	0.18*	-0.02, 0.37
Overall severity VAS	0.68***	0.56, 0.77	0.72***	0.6, 0.8

\* $P=0.07$ , \*\* $P < 0.01$ , \*\*\* $P < 0.001$ .

RP, Raynaud's phenomenon; SOB, shortness of breath. Analysis by Spearman's correlation coefficient.

$\geq 1.26$  cm, hand spread  $<15.5$  cm, albumin  $<40$  g/l, ESR  $\geq 20$  mm/h and CRP  $\geq 10$  mg/l. Disease duration, sex, creatinine level, haemoglobin, diastolic blood pressure, DLCO and pulmonary artery pressure did not predict higher UKFS or HAQ-DI scores. Results are shown in Table 4.

The VAS for shortness of breath was significantly higher for FVC  $<70\%$  ( $P=0.016$ ) and DLCO  $<70\%$  ( $P < 0.001$ ). The VAS for shortness of breath and overall disease severity were significantly higher with skin score greater than 15 ( $P=0.019$  and  $P=0.03$ , respectively). The VAS assessing overall disease severity and shortness of breath were significantly higher with albumin  $<40$  mg/l ( $P=0.013$  and  $P=0.03$ , respectively). Fist closure limited to distances  $\geq 1.26$  cm had significantly higher VAS scores in the overall disease severity scale ( $P=0.01$ ) and the VAS assessing pain ( $P=0.001$ ). Other clinical or laboratory variables did not differ significantly for the five VAS measured.

Multivariate regression analysis determined individual predictors of the HAQ-DI and UKFS (Tables 5 and 6). Elevation of CRP, impaired hand spread and fist closure were independent predictors of a higher UKFS. Fist closure and CRP were independent predictors of higher

TABLE 4. Clinical variables as independent correlates of functional status determined by independent *t*-test

Variable	HAQ-DI			UKFS			
	Mean (s.d.)	95% CI	<i>P</i> value	Mean (s.d.)	95% CI	<i>P</i> value	
FVC	≥ 70% predicted	0.85 (0.78)	0.09, 0.82	0.01	10.4 (8.5)	2.1, 10.3	0.004
	< 70%	1.25 (0.73)			16.1 (8.1)		
ESR	≥ 20 mm/h	1.05 (0.79)		0.07	13.6 (8.7)		0.03
	< 20 mm/h	0.73 (0.72)	-0.02, 0.66		9.4 (8.3)	0.24, 8.24	
Albumin	≥ 40 g/l	0.77 (0.7)	0.03, 0.8	0.04	10.4 (8.4)		0.01
	< 40 g/l	1.22 (0.74)			16.4 (8.4)		
Skin score	≥ 15	1.4 (0.9)	0.24, 1.0	0.002	18.1 (9.5)	3.6, 11.9	< 0.001
	< 15	0.8 (0.71)			10.4 (7.9)		
Fist closure							
Reduced	≥ 1.26 cm	1.5 (0.7)	0.53, 0.12	< 0.001	17.7 (7.2)	5.6, 12.3	< 0.001
	< 1.26 cm	0.63 (0.7)			8.8 (8.1)		
Hand spread	≥ 15.5 cm	0.7 (0.72)	0.28, 0.88	< 0.001	8.4 (7.6)	3.9, 10.7	< 0.001
Reduced	< 15.5 cm	1.2 (0.77)			15.6 (8)		
CRP	≥ 10 mg/l	1.6 (0.6)	0.74, 1.4	< 0.001	18.8 (7.3)	5.5, 13.7	< 0.001
	< 10 mg/l	0.6 (0.61)			9.2 (7.6)		

HAQ-DI. Logistic regression determined the individual independent predictors of each scleroderma-VAS; results are shown in Table 7.

## Discussion

It is well recognized that patients with scleroderma have multisystem involvement and hence disability in scleroderma can be expected to be multifactorial in origin. The clinical heterogeneity of scleroderma is exemplified by differences in the extent of skin involvement, which define the diffuse and limited subsets. Additionally the propensity for internal organ involvement varies considerably between individual patients. Therefore, manifestations of scleroderma can range from those that are moderately troublesome to those that are life-threatening. The focus of our attention when assessing patients with scleroderma is often based on physiological measurements, such as lung function, or the extent of pathology, such as skin score. Measures of end-organ damage and mortality have been the traditional tools used to assess patients with scleroderma both in clinical practice and as part of therapeutic trials [5]. These modalities do not necessarily relate to the true impact of the disease on a patient's lifestyle or functional capacity.

The HAQ was developed initially to measure disability in rheumatoid arthritis and its results correlate with changes in laboratory and physical examination measurements in RA [17], and have been shown to predict mortality [18]. The HAQ has been shown to be of use in assessing patients with scleroderma [9]. Steen *et al.* [10] modified the traditional HAQ by including five visual analogue scales related to symptoms of pain, vascular, respiratory and gastrointestinal involvement and a scale determining overall disease severity, with the goal of providing additional precision in assessing patients with scleroderma. The recent development of the UKFS was based on the recognition that traditional scoring systems inquired about global disability but did not necessarily focus on specific problems of relevance in scleroderma.

TABLE 5. Independent correlates of the UKFS determined by multivariate regression analysis

Variable	Coefficient (95% CI)	<i>P</i> value
Hand spread (reduced)	3.7 (0.5, 7.0)	0.025
Fist closure (reduced)	6.6 (3.0, 10.1)	< 0.001
CRP (elevated CRP ≥ 10 mg/l)	4.7 (1.0, 8.4)	0.014

TABLE 6. Independent correlates of the HAQ-DI determined by multivariate regression analysis

Variable	Coefficient (95% CI)	<i>P</i> value
Fist closure (reduced)	23.3 (0.97, 0.4)	< 0.001
CRP (elevated CRP ≥ 10 mg/l)	15.0 (0.27, 0.85)	< 0.001

This may be particularly true for patients with disease restricted to the hands, such as those in the limited cutaneous subset. The core questions were developed from data collected from patients and therapists. It is interesting that the areas identified as being problematic were mainly disability related to hand function. Nine of the questions in the UK functional assessment relate to hand function, while two examine proximal muscle strength. The UK functional assessment and the modified scleroderma-HAQ are therefore disease-specific instruments designed to capture the true impact of scleroderma in a more specific way than traditional assessment tools.

This is the first comparative study of the HAQ-DI, the scleroderma-VAS and the recently developed UK functional questionnaire. The mean overall disability index seen in the cohort was 0.94, when diffuse and limited subgroups were considered together. This is in keeping with the reported range of the HAQ-DI among scleroderma patients of 0.83–1.2 [19]. This range is similar to reported measurements among patients with rheumatoid arthritis and systemic lupus erythematosus. The significantly higher HAQ-DI found in our patients with diffuse disease compared with those with limited

TABLE 7. Independent correlates of the scleroderma-VAS determined by logistic regression analysis

VAS	Predictor(s)	Odds ratio (95% CI)	P value
Pain	Fist closure (reduced fist closure)	7 (1.9, 25.6)	0.003
	DLCO (reduced < 70%)	4.9 (1.2, 20)	0.026
SOB	Albumin (reduced < 40 g/dl)	5.2 (1.2, 23.2)	0.03
	Fist closure (reduced fist closure)	15.8 (1.9, 27.8)	0.009
Overall well-being	Hand spread (reduced hand spread)	3.9 (1.2, 12.9)	0.023

disease concurs with the findings of earlier studies [10]. The mean UKFS in our cohort was 11.9, compared with 9.8 in the original paper [11], which assessed the reliability of the questionnaire as a patient self-administered tool and when administered by a trained observer. A lower median UK functional score has recently been reported by Herrick *et al.* [20]; however, no correlation was found between functional score and the total Rodnan skin score, even when the skin of the upper limbs and digits was considered separately.

A strong correlation between the UK functional score and the HAQ-DI has been demonstrated in this study. The HAQ-DI is historically considered to be a global assessment tool; the score is based on the ability of the individual to perform eight activities of daily living. The strong correlation seen between the HAQ-DI and the UKFS is not surprising because six of the categories examined in the HAQ are directly influenced by hand function. However, while the activities of dressing and grooming, grip, reaching, eating, personal hygiene and the ability to vacuum examined in the HAQ are strongly influenced by hand function, this is not their only influence. Upper and lower limb strength, the extent of skin contractures and general level of fitness, which would bear a direct relationship to the extent of lung involvement, would all be expected to affect the final HAQ-DI score. The UKFS, therefore, has the advantage of defining the influence of impaired functional capacity related predominantly to hand function. This functional assessment tool would therefore be of use in assessment of digital ulcers, which account for considerable morbidity in patients with scleroderma. Physicians can clearly diagnose the presence or absence of ulceration, indeed the development of new digital ulcers is now recognized as a valid end-point for clinical trials [21]. A gradual improvement in digital ulceration, albeit incomplete, is often accompanied by improved functional capacity. The UKFS would clearly be useful in providing a relatively selective assessment of the functional impact of digital ulcers, before and after therapeutic intervention, either in clinical practice or as part of a clinical trial.

The scleroderma-VAS were found to correlate with the UKFS and HAQ-DI only in the scales that examined overall disease severity, respiratory symptoms and level of pain. A significant association was seen between physiological measurements reflecting pulmonary involvement and the VAS assessing shortness of breath, suggesting good construct validity [22]. It is therefore clear that the three assessment modalities in part assess

common components of functional disability. Additionally, however, the UKFS allows a more precise assessment of hand function and the scleroderma-VAS likewise has the ability to assess organ-specific manifestations of the disease. The use of these tools is therefore complementary and adds little in terms of complexity or time when patients are assessed.

Several clinical and laboratory measures were seen to predict higher UKFS and HAQ-DI scores in our study. Higher scores were directly correlated with skin score, reduced forced vital capacity and indicators of impaired hand function, namely reduced fist closure and hand spread. Furthermore, elevated ESR and CRP (both acute-phase reactants) and reduced serum albumin (considered a negative acute-phase protein) correlated with higher UK functional scores. Multiple regression analysis to determine independent predictors for the UKFS found that impaired fist closure and limitation in hand spread predicted higher scores; these findings are not surprising but rather provide further support for the construct validity of this tool in the assessment of hand function. Several groups have demonstrated similar correlations between higher HAQ-DI scores and laboratory measures and higher skin scores [9]. Recent studies have also addressed the ability of the HAQ to change over time among patients experiencing changes in their disease activity [10]. The results showed that patients with diffuse scleroderma who died had a higher HAQ score at their first clinic visit compared with surviving patients with diffuse disease, and in addition demonstrated a significant worsening of HAQ score prior to their deaths. This study and an analysis of the HAQ-DI in patients participating in the high- vs low-dose D-penicillamine trial found that changes in the HAQ-DI correlated with changes in skin score [6]. The HAQ's correlation with clinical change and the ability to predict survival among patients with scleroderma has important implications for clinical trial design and clinical care, which might be further strengthened by additional scleroderma-specific indices including the UKFS.

In conclusion, the HAQ-DI, UKFS and scleroderma-VAS are self-administered questionnaires that scleroderma patients can complete easily and rapidly. Used together, these inexpensive tools assess general and organ-specific symptoms, as well as different aspects of functional limitation. The importance of obtaining measures of health-related outcomes such as physical disability in patients with chronic illnesses such as scleroderma is becoming widely accepted. These assessment

measures should form part of the standard assessment of patients with scleroderma by both clinical researchers and practising physicians.

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