
Impact of hand and face disabilities on global disability and quality of life in systemic sclerosis patients

S. Maddali-Bongi¹, A. Del Rosso¹, S. Mikhaylova¹, B. Francini¹, A. Branchi¹,
M. Baccini², M. Matucci-Cerinic¹

¹Division of Rheumatology, Department of Experimental and Clinical Medicine, University of Florence;

²School of Physiotherapy, University of Florence, Italy,

Susanna Maddali Bongi, MD*

Angela Del Rosso, MD, PhD*

Svetlana Mikhaylova, MD

Benedetta Francini, Physiotherapist

Arianna Branchi, Physiotherapist

Marco Baccini, Prof. in Physiotherapy

Marco Matucci-Cerinic, MD, PhD

*These authors made an equal contribution to the study.

Please address correspondence to:

Angela Del Rosso, MD, PhD,

Division of Rheumatology,

Department of Experimental

and Clinical Medicine,

University of Florence,

viale Pieraccini 18,

50139, Firenze, Italy.

E-mail: angela.delrosso@fastwebnet.it

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ABSTRACT

Objective. In systemic sclerosis (SSc), the frequent involvement of hand and face leads to their disability. We aimed to assess influence of hand and face disability on global disability and Health-related Quality of life (HRQoL).

Methods. 119 SSc patients were assessed for global disability by HAQ, HRQoL, by SF36; hand disability by HAMIS, CHFDS, fist closure and hand opening measures; face disability by MHISS and mouth opening measure.

Results. Diffuse SSc (dSSc) patients present higher HAQ, lower Summary Physical Index (SPI) of SF36, major hand disability at hand (higher HAMIS, CHFDS, fist closure, lower hand opening) and face (lower mouth opening, higher MHISS) than lSSc patients ($p < 0.05$).

SPI of SF36 is negatively correlated with MHISS, CHFDS, HAMIS and positively correlated to mouth and hand opening ($p < 0.05$). Summary Mental Index (SMI) of SF36 is negatively correlated with MHISS ($p < 0.05$). HAQ is negatively correlated with mouth opening and positively correlated to MHISS, HAMIS, CHFDS ($p < 0.05$). By hierarchical multiple linear regression, SPI of SF36 is significantly associated with total MHISS ($B = -0.34$; $t = -3.78$; $p < 0.001$) and CHFDS ($B = -0.27$; $t = -3.01$; $p = 0.003$), together, explaining 22% of SPI variance. SMI of SF36 is significantly associated only with MHISS total score ($B = -0.22$; $t = -2.41$; $p = 0.017$), explaining 4% of its variance. HAQ is significantly associated with CHFDS score ($B = 0.61$; $t = 7.90$; $p < 0.001$), explaining 36% of HAQ variance.

Conclusion. dSSc patients present higher global and local disability, and lower HRQoL in SPI than lSSc patients. Local disabilities, assessed by CHFDS and MHISS, are independently related to global disability and HRQoL.

Introduction

Systemic sclerosis (SSc) is characterised by microvascular impairment, fibrosis at internal organs and at subcutaneous tissues and skin, leading to tissue retraction and atrophy, and by joint and muscle impairment (1).

In SSc, impairment of microcirculation and musculoskeletal and peripheral nervous system, fibrosis of skin and tendons and at inter- and metacarpophalangeal joints, leading to claw-type deformities, cause severe hands disability. The resulting impairment in movements and in daily activities reduces Health-related Quality of life (HRQoL) (2).

Despite its frequency, hand disability in SSc is measured with few specific instruments, such as Cochin Hand Function Disability Scale (CHFDS), also known as Duruoz Hand Index (DHI) (3), and Hand Mobility in Scleroderma (HAMIS) test (4, 5), both reliable and valid in assessing district-specific handicap and in following-up disease evolution and treatments (6).

Due to fibrosis of skin and soft tissues, SSc leads also to a typical involvement of the face and oral tissues, known as “sclerodermic face”, with amimia, disappearing of cutaneous wrinkles, vertical furrows around the mouth and sharpening of the nose. Face and mouth changes including teleangectasia, sicca syndrome, microcheilia and microstomia, lead to difficulties in eating, speaking, oral hygiene and dental treatments (7-9). Face involvement, often under-rated, is assessed by Mouth Handicap in Systemic Sclerosis scale (MHISS), a specific tool quantifying mouth disability and oral HRQoL (10), that explores problems not assessed by Health Assessment Questionnaire (HAQ) and Short Form 36 (SF 36), evaluating global disability and HRQoL.

In SSc, the involvement of hands and

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face may lead to notable concerns about disfigurement (11) and personal self-esteem (12), and may become one of the patients' greatest complaints (13, 14), potentially causing disability and impairing HRQoL. Skin deformities were also shown as core stressors of the disease, only preceded by fatigue (14).

In SSc, disability is usually assessed by HAQ, also regarded as a prognostic index. High scores are predictive of reduced survival in diffuse SSc (dSSc) (15), while low scores are associated with a better disease outcome in early dSSc patients (16). Moreover, HAQ scores are higher in dSSc than in patients suffering from intermediate and limited SSc (lSSc) (17).

In SSc, HAQ as well as Summary Physical Index (SPI) of SF36, is adversely affected by joint involvement, muscle tenderness, gastrointestinal involvement and high skin score (18, 19, 20). HAQ is also higher in patients with digital ulcers (21) and reduced mouth opening (20).

According to many studies, in SSc, HRQoL evaluated by SF36, is lower than in healthy controls (18, 22-25), with significant differences in many SF36 health dimensions in dSSc in respect to lSSc (22, 24, 18). A review demonstrates that dSSc patients have significantly lower scores in SPI but not in summary mental index (SMI) of SF36 (26) than lSSc patients.

In SSc, correlations were shown between the number of gastrointestinal symptoms, such as pain and bloating (23, 27), pain of whichever origin (24), dyspnoea (28), skin score (27), as well as age (22), and impaired HRQoL, especially SPI. Digital ulcers seem to influence negatively SMI but not SPI (29).

In SSc, the contemporary presence of impaired HRQoL and global disability with face and hand disability was previously shown, but a survey assessing the presence and the correlation between these items, as assessed by specific instruments, is still lacking.

The aim of our study is to assess, in SSc, HRQoL and local and global disability; to verify if they could be different according to subsets (dSSc and lSSc); to assess if HRQoL and global disability

could be affected by local disability at hands and face.

Materials and methods

One hundred and nineteen SSc patients [14 men and 105 women; 74 with limited SSc (lSSc) and 45 with diffuse SSc (dSSc) (30), whose age and disease duration were 59.46 ± 13.87 years; and 10.74 ± 7.42 years, respectively] fulfilling ACR criteria (31) were consecutively enrolled from the outpatient clinic and Day Hospital of the Division of Rheumatology, Department of Experimental and Clinical Medicine, University of Florence.

The patients signed a written informed consent, the procedures were conducted in accordance with the 1975/83 Helsinki Declaration. The study was approved by local ethics committee.

Clinic evaluation

SSc patients underwent an extensive clinical workout. The skin involvement was evaluated by modified Rodnan Skin Score (32). The presence of interstitial lung disease was assessed with high-definition computed tomography, pulmonary function tests (with DLCO) and bronchoalveolar lavage; pulmonary hypertension by echocardiography, right heart catheterisation and echocolor Doppler. Cardiac involvement was defined in the presence of pericarditis, arrhythmias, or right congestive heart failure. The oesophageal involvement was identified by hypomotility of oesophageal transit, assessed by radiography with barium swallow and/or manometry. Kidney involvement was evaluated by assessing serum creatinine concentration, creatinine clearance, blood and urine (24 hour) uric acid, sodium, potassium concentrations, microalbuminuria and 24-hour proteinuria.

The presence of Sjögren's syndrome secondary to SSc (33), as well as the presence of antinuclear (ANA), anticentromere (ACA) and anti-topoisomerase I (Scl70) antibodies were recorded.

Clinical involvement of the hands was defined in the presence of arthralgia, arthritis (presence of joint swelling and tenderness), finger flexion contractures; calcinosis, Raynaud's phenomenon and digital ulcers were also registered.

Clinimetric evaluations

During a routine visit, SSc patients were administered with tools and tests evaluating global disability, HRQoL, hand, face and mouth involvement.

Assessment of global disability and Quality of Life

Short Form (SF36), assessing HRQoL, is organised in 8 scales evaluating physical functioning (PF), role limitations due to physical problems (PP), bodily pain (BP), general health perceptions (GH), vitality (V), social functioning (SF), role limitations due to emotional problems (EP), and general mental health (MH), combined into summary physical and mental indexes (SPI and SMI) with higher scores corresponding to better HRQoL (range 0-100) (34).

Health Assessment Questionnaire (HAQ) assessing disability (35), is organised in 20 items divided into 8 domains of activities of daily living and rated from 0 (no difficulty) to 3 (unable to do).

Assessment of hand function

Cochin Hand Functional Disability scale (CHFDS) or *Duruoz's Hand Index (DHI)*, is a self-report questionnaire that measures hand ability in performing 18 manual tasks. Total score ranges 0-90, with higher scores meaning higher disability (3). Validity of the Italian version of CHFDS in SSc patients was shown (36).

HAMIS (Hand Mobility In Scleroderma) test is a performance-based test evaluating hand function in SSc, composed of 9 items, related to movements that are part of daily occupations, with a total score of 27, in which higher scores indicate greater disability (4, 5). HAMIS score at right and left hand, as well as mean HAMIS score, were calculated. Validity of the Italian version of HAMIS in SSc patients was demonstrated by our group (37).

Fist closure and hand opening. Fist closure is valued as the distance between the fingertip of the third finger and tenar eminence, and hand opening as the distance between the fingertip of the third finger, when extended, and the table. Both measures, reported in centimeters and as mean of 2 consecutive

measurements, were assessed at right and left hand, and as mean between the 2 of them.

Assessment of face and mouth

Mouth Handicap in Systemic Sclerosis Scale (MHISS) assessing the handicap associated with mouth disability in SSc, consists in 12 items (with a total score ranging from 0 to 48) divided in 3 subscales: subscale 1 examines handicap related to reduced mouth opening; subscale 2 assesses handicap related to sicca syndrome; subscale 3 examines aesthetic concerns (10). Validity and reliability of the Italian version of MHISS was reported by our group (38).

Mouth opening is the distance between the tips of upper and lower incisive teeth (mean in centimetres of 2 consecutive measurements), measured at the maximal oral aperture.

Statistical analysis

Data are presented as mean $\pm$ standard deviation for continuous variables and as numbers for binomial variables. Continuous variables were tested with the Kolmogorov-Smirnov test for normality.

Clinical characteristics of groups were compared by Fisher's exact test or χ^2 test for binomial variables and with *t*-student and Mann-Whitney tests for continuous variables, when appropriated.

Bivariate associations were assessed by Spearman's correlation. Variables assessing local disability that significantly correlated with HAQ and with SMI and SPI of SF36 were entered in hierarchical multiple regression models where HAQ, SMI and SPI scores were the dependent variables.

For all tests, the significance was set at a *p*-value <0.05 . All analyses were performed by using SPSS version 20.0 for Windows (SPSS, Chicago, IL, US).

Results

Comparison between ISSc and dSSc

Clinical, clinimetric and laboratory characteristics of SSc (whole group) and ISSc and dSSc patients are reported in Table I. dSSc patients have higher skin score and higher incidence of clinical hand involvement and Scl70 positivity, whilst ISSc patients have higher

Table I. Clinical and clinimetric characteristics of SSc, ISSc and dSSc patients.

Patients characteristics	SSc (119)	ISSc (74)	dSSc (45)	<i>p</i> -value (ISSc vs. dSSc)
Age	59.92 $\pm$ 13.49	60.28 $\pm$ 13.25	59.31 $\pm$ 14.01	NS
Disease duration (years)	10.58 $\pm$ 7.42	10.28 $\pm$ 8.06	11.05 $\pm$ 6.39	NS
Sex (F/M)	106/14	66/5	40/9	NS
Skin score	11.27 $\pm$ 7.13	15.60 $\pm$ 8.80	8.70 $\pm$ 4.70	<0.001
Raynaud phenomenon	119/119	74/74	45/45	NS
Calcinosis	16/119	11/74	5/45	NS
Digital ulcers	69/119	40/74	29/45	NS
Clinical Hand involvement	84/119	57/74	27/45	0.03
Heart involvement	30/119	17/74	13/45	NS
Pulmonary hypertension	35/119	17/74	18/45	NS
Interstitial lung disease	69/119	41/74	28/45	NS
Kidney involvement	11/119	6/74	5/45	NS
Sjögrens syndrome	17/119	15/74	2/45	<0.05
Oesophageal involvement	42/119	26/74	16/45	NS
ANA	114/119	73/74	41/45	NS
ACA	65/119	62/74	3/45	<0.001
Scl70	42/119	13/74	29/45	<0.001
HAQ	0.63 $\pm$ 0.65	0.55 $\pm$ 0.70	0.76 $\pm$ 0.53	0.004
SF 36 Physical Functioning	67.27 $\pm$ 27.45	72.23 $\pm$ 26.86	59.11 $\pm$ 26.72	0.003
SF 36 Physical Role Limitation	47.27 $\pm$ 40.88	54.39 $\pm$ 40.41	35.56 $\pm$ 39.34	0.015
SF 36 Body Pain	51.26 $\pm$ 26.6	54.24 $\pm$ 25.61	46.36 $\pm$ 27.75	NS
SF 36 General Health Perception	39.93 $\pm$ 19.90	42.47 $\pm$ 19.68	35.76 $\pm$ 19.76	NS
SF 36 Vitality	52.44 $\pm$ 18.32	54.93 $\pm$ 17.51	48.33 $\pm$ 19.07	NS
SF 36 Social Functioning	69.66 $\pm$ 24.77	71.11 $\pm$ 24.00	67.29 $\pm$ 26.08	NS
SF 36 Emotional Role Difficulties	56.03 $\pm$ 41.70	60.89 $\pm$ 39.29	48.20 $\pm$ 44.67	NS
SF 36 Mental Health	60.20 $\pm$ 18.80	61.51 $\pm$ 17.36	58.04 $\pm$ 20.97	NS
SPI-SF36	41.69 $\pm$ 10.39	40.43 $\pm$ 10.39	35.76 $\pm$ 9.12	0.009
SMI-SF36	44.02 $\pm$ 10.61	44.72 $\pm$ 9.39	42.73 $\pm$ 11.89	NS
Right HAMIS	5.19 $\pm$ 5.36	3.82 $\pm$ 4.62	7.40 $\pm$ 5.77	<0.001
Left HAMIS	5.03 $\pm$ 5.12	3.58 $\pm$ 4.08	7.40 $\pm$ 5.77	<0.001
Mean HAMIS	5.11 $\pm$ 5.24	3.70 $\pm$ 4.30	7.40 $\pm$ 5.77	<0.001
CHFDs	10.68 $\pm$ 13.10	9.60 $\pm$ 15.36	12.51 $\pm$ 11.25	0.01
Right hand opening (cm)	3.40 $\pm$ 1.34	3.65 $\pm$ 1.29	3.01 $\pm$ 1.34	0.018
Left hand opening (cm)	3.28 $\pm$ 1.38	3.49 $\pm$ 1.44	2.95 $\pm$ 1.24	0.031
Mean hand opening (cm)	3.54 $\pm$ 1.40	3.57 $\pm$ 1.36	2.98 $\pm$ 1.29	0.02
Right fist closure (cm)	0.79 $\pm$ 1.40	0.53 $\pm$ 1.17	1.19 $\pm$ 1.61	0.009
Left fist closure (cm)	0.82 $\pm$ 1.42	0.51 $\pm$ 1.19	1.30 $\pm$ 1.62	0.002
Mean fist closure (cm)	0.86 $\pm$ 1.46	0.52 $\pm$ 1.18	1.24 $\pm$ 1.62	0.01
total MHISS	18.12 $\pm$ 11.21	16.26 $\pm$ 11.13	21.18 $\pm$ 10.77	0.024
MHISS mouth opening	8.14 $\pm$ 6.09	6.91 $\pm$ 6.31	10.18 $\pm$ 5.18	0.001
MHISS Sicca syndrome	6.96 $\pm$ 5.26	6.92 $\pm$ 4.91	7.02 $\pm$ 5.85	NS
MHISS aesthetic concerns	3.43 $\pm$ 2.93	3.08 $\pm$ 3.00	4.00 $\pm$ 2.76	NS
Mouth opening (cm)	3.89 $\pm$ 0.10	4.14 $\pm$ 0.88	3.47 $\pm$ 1.04	0.001

SSc: systemic sclerosis; ISSc: limited systemic sclerosis; dSSc: diffuse systemic sclerosis; ANA: anti-nuclear antibodies; ACA: anticentromere antibodies; Scl-70: anti-topo-isomerase (Scl70) antibodies; SF36: Short Form 36; HAQ: Health Assessment Questionnaire; HAMIS: Hand Mobility In Scleroderma Test; CHFDs: Cochin Hand Functional Disability Scale; MHISS: Mouth Handicap in Systemic Sclerosis Scale; NS: not significant.

incidence of sicca syndrome and ACA positivity ($p<0.05$ for all comparisons).

Quality of life, global disability

In SF36, the scores of SPI, PF and PRL subscales are significantly lower in dSSc than in ISSc patients ($p<0.05$ for all comparisons). The scores of SMI and the other SF36 subscales are similar in the 2 subsets.

Patients with dSSc present significantly higher HAQ ($p=0.04$) in respect to ISSc patients (Table I).

Hand and face disability and mobility

dSSc patients present higher scores in right, left and mean HAMIS ($p<0.001$ for all comparisons) as well as in CHFDs values ($p=0.01$) than ISSc subjects.

Table II. Correlations of HRQoL and global disability with measures of hands and face involvement in SSc patients.

		SPI-SF36	SMI-SF36	HAQ
Age	r	---	---	---
	p-value	NS	NS	NS
Right HAMIS	r	-0.267	---	0.385
	p-value	0.004	NS	<0.001
Left HAMIS	r	-0.268	---	0.377
	p-value	0.003	NS	<0.001
Mean HAMIS	r	-0.264	---	0.383
	p-value	0.004	NS	<0.001
CHFDS	r	-0.381	-0.156	0.670
	p-value	<0.001	NS	<0.001
Right hand opening (cm)	r	0.206	---	---
	p-value	0.030	NS	NS
Left hand opening (cm)	r	0.197	---	-0.185
	p-value	0.037	NS	NS
Right hand fist closure (cm)	r	-0.090	0.076	0.098
	p-value	NS	NS	NS
Left hand fist closure (cm)	r	---	---	---
	p-value	NS	NS	NS
MHISS total	r	-0.454	-0.210	0.413
	p-value	<0.001	0.022	<0.001
MHISS mouth opening	r	-0.383	---	0.447
	p-value	<0.001	NS	<0.001
MHISS Sicca syndrome	r	-0.353	-0.228	0.215
	p-value	<0.001	0.01	0.01
MHISS aesthetic concerns	r	-0.314	---	0.234
	p-value	<0.001	NS	0.01
Mouth opening (cm)	r	0.205	---	-0.254
	p-value	0.039	NS	0.007

SSc: systemic sclerosis; HAMIS: Hand Mobility In Scleroderma Test; CHFDS: Cochin Hand Functional Disability Scale; MHISS: Mouth Handicap in Systemic Sclerosis Scale; HAQ: Health Assessment Questionnaire; SF36: Short Form 36; HAQ: Health assessment Questionnaire; NS: not significant.

Total MHISS and MHISS mouth opening scores are higher in dSSc than in lSSc ($p=0.024$ and 0.001 respectively), with no differences in the MHISS subscales assessing sicca syndrome and esthetic concerns.

In patients with dSSc, right, left, mean hand opening and the maximal mouth opening are significantly lower and right, left, mean fist closure are significantly higher than in lSSc; ($p<0.05$ for all comparisons) (Table I).

Correlations between SF36 and HAQ with hand and face disability

SPI of SF36 is negatively correlated with total MHISS and with MHISS mouth opening, sicca syndrome and esthetic subscales ($p<0.001$ for all

statistics) and positively related with mouth opening ($p=0.039$). SPI is negatively related with CHFDS ($p<0.001$), HAMIS of right and left hands and HAMIS mean scores ($p<0.05$ for all statistics) and positively related with opening of the right and left hands ($p<0.05$ in both cases).

SMI of SF-36 is negatively correlated with total MHISS ($p=0.022$) and with MHISS sicca syndrome scores ($p=0.01$). HAQ is negatively correlated with mouth opening measure ($p=0.007$) and positively correlated with total MHISS, MHISS mouth opening ($p<0.001$ in both cases), MHISS sicca syndrome and esthetic subscales ($p=0.01$ in both cases), HAMIS left, right and mean score and CHFDS ($p<0.001$ in all cases).

No significant correlations were found between SPI, SMI and HAQ and other variables (Table II).

Independent predictors of Global Disability and Quality of Life

In the hierarchical multiple linear regression models, SPI of SF-36 is significantly associated with MHISS total score ($B=-0.34$; $t=-3.78$; $p<0.001$) and CHFDS ($B=-0.27$; $t=-3.01$; $p=0.003$), which, together, explain 22% of SPI variance.

SMI of SF-36 is significantly associated only with MHISS total score ($B=-0.22$; $t=-2.41$; $p=0.017$), which explains 4% of its variance.

HAQ is significantly associated with CHFDS score ($B=0.61$; $t=7.90$; $p<0.001$), which explains 36% of the variance in HAQ score.

Discussion

To the best of our knowledge, this is the first study assessing systematically HRQoL, global disability and district disability at hands and face by using specific clinimetric instruments and anthropometric measures in a group of SSc patients according to subsets, and showing their correlation.

According to our results, dSSc patients present higher levels of disability, as assessed by HAQ, lower HRQoL in SF36 physical domains and SPI, confirming data already published in literature (18, 22, 24, 26).

Our data show a higher clinical incidence of hand involvement in dSSc, substantiated by higher scores in CHFDS, HAMIS, reduced hand opening, and increased fist closure than in lSSc.

Previously, both CHFDS (36) and HAMIS (37) showed higher scores in SSc patients with hand impairment. In our survey, both scales have higher values in dSSc, showing a major clinical hand involvement, than in lSSc, thus demonstrating to be able in distinguish hand disability according to subsets.

The reduced hand opening and the increased fist closure (39) confirm a more important hand involvement in dSSc than in lSSc and indicate these measures to be helpful, as already shown by our group, to follow up, together with HAMIS and CHFDS, the efficacy of

rehabilitation programmes in patients with SSc (6, 40, 41).

In parallel with hand involvement, we have shown face and mouth involvement as more prominent in dSSc patients, presenting lower mouth opening, higher total MHISS and MHISS mouth opening scores than lSSc patients, who, however, show a higher incidence of sicca syndrome. These results confirm the major disability found by MHISS in a previous study in dSSc (38) and show a different facial and oral involvement according to the subsets.

Our data show that, in SSc patients, physical HRQoL and global disability are strictly related to disabilities of hands and mouth, that our study evaluates, for the first time, in a detailed and systematic way by a large panel of clinimetric tools and anthropometric measures, differently from previous surveys, which assessed local disability only by a few tools in each study (27-29, 36, 37, 42).

Previous data showed that SSc patients with clinical hand involvement, evaluated by anthropometric measures and grip strength, and with digital ulcers present high disability at HAQ (27-29). Concordantly, HAQ was related to hand and finger function and hand grip (27). A high HAQ score was correlated with reduced mouth opening, increased finger flexion, subcutaneous calcinosis and flexion contractures (28) and, most importantly, with hand extension, that, by multivariate logistic regression remained independently correlated with HAQ (28).

Our results, showing a negative correlations of SF36 SPI with HAMIS and CHFDS are in keeping with the impact that hand dysfunction in SSc has on HRQoL and are confirmed by the correlation of low scores of SPI with reduced hand opening.

The correlations of HAQ with HAMIS and CHFDS are somewhat expected, because HAQ contains items also evaluating hand impairment and, as well as the correlation of SF36 with HAMIS, are concordant with previous studies (36-37, 42).

By regression models, CHFDS remains independently associated with SPI and with HAQ. These results confirm and

strengthen the findings of Rannou *et al.*, showing that, in SSc, CHFDS contributed to 75% of the HAQ variance and underline manual disability as an independent predictor of physical HRQoL and global disability in SSc.

In our study, we found low SPI and high HAQ scores to be related to mouth handicap, as represented by high scores in total MHISS and in its 3 subscales, evaluating the complains due to mouth opening, sicca syndrome and esthetic aspect. Only low SPI, however, was related with reduced mouth opening, assessed as a clinimetric measure. SMI was correlated only with total MHISS and MHISS sicca syndrome scores.

The correlation of MHISS with Physical and Mental HRQoL and global disability may reflect the consequences of SSc not only on the physical sphere, but also on mental dimensions, caused by the frequent, although under-rated, mouth and face involvement, already shown to be related to disfigurement (11) and change of personal self-esteem (12).

How functional changes of mouth and face may affect HRQoL in SSc is confirmed, at the best of our knowledge for the first time, by the independent association of total MHISS with SPI and SMI of SF36 found by regression models. Face involvement, reported by SSc patients as a prominent concern, and evident since the earliest phases of the disease, is seldom evaluated or under-rated in daily practice. However, the influence of oral-related HRQoL (10, 38) on global HRQoL is not surprising, because face and mouth changes may lead not only to severe functional problems in the daily life (7, 8, 9) but also, by altering patient's self-image, ultimately, may reduce HRQoL (14, 43).

Recently, telangiectases, mainly present in the face of SSc patients, were found to be related with body image dissatisfaction, and with changes in behavior, self-image, emotional impact and appreciation of life (44).

Based on our data, we suggest that hand and face disability, as predicting and influencing HRQoL and global disability, could be evaluated (preferentially by MHISS and CHFDS) in surveys and in clinical trials assessing SSc subjects, as additional outcome measures of func-

tion. Moreover, the assessment of face and hand disability should be added to the tools used in outpatient clinics, in the context of a proper approach to the patient (45), not only to evaluate severity of SSc manual and facial involvement, but also to choose the most suitable techniques for their specific problems, to manage adequately, from the earliest phases of the disease (46), the different disabilities with specific protocols for hand (6, 41), face (47, 48), and global rehabilitation (40, 49), and, eventually, with psycho-educational programmes (50), and to follow-up the outcome of the therapies.

Conclusion

Our study shows that global disability and HRQoL are more impaired in lSSc than in dSSc, as well as face and hand disability and anthropometric measures of hand and mouth functions.

Bivariate correlations show that global disability and HRQoL are influenced by changes in hand and face disability and in anthropometric measures.

Hand and face disability as evaluated by CHFDS and MHISS are independent predictors of disability and HRQoL. Thus, these tests should be used to assess properly SSc patients, and to choose and to follow up the more adapt rehabilitative treatment in order, ultimately, to improve their conditions.

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