ORIGINAL ARTICLE

The Italian version of the Mouth Handicap in Systemic Sclerosis scale (MHISS) is valid, reliable and useful in assessing oral health-related quality of life (OHRQoL) in systemic sclerosis (SSc) patients

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Abstract In systemic sclerosis (SSc), mouth and face involvement leads to problems in oral health-related quality of life (OHRQoL). Mouth Handicap in Systemic Sclerosis scale (MHISS) is a 12-item questionnaire specifically quantifying mouth disability in SSc, organized in 3 subscales. Our aim was to validate Italian version of MHISS, by assessing its test–retest reliability and internal and external consistency in Italian SSc patients. Forty SSc patients (7 dSSc, 33 lSSc; age and disease duration: 57.27 ± 11.41, 9.4 ± 4.4 years; 22 with sicca syndrome) were evaluated with MHISS. MHISS was translated following a forward–backward translation procedure, with independent translations and counter-translation. Test–retest reliability was evaluated, comparing the results of two administrations, with intraclass correlation coefficient (ICC). Internal consistency was assessed by Cronbach's α and external consistency by comparison with mouth opening. MHISS has a good test–retest reliability (ICC: 0.93) and internal consistency (Cronbach's α:0.99). A good external consistency was confirmed by correlation with mouth opening (rho: $-0{,}3869$, p: 0.0137). Total MHISS score was 17.65 ± 5.20, with scores of subscale 1 (reduced mouth opening) of 6.60 ± 2.85 and scores of subscales 2 (sicca syndrome) and 3 (aesthetic concerns) of 7.82 ± 2.59 and 3.22 ± 1.14. Total and subscale 2 scores are higher in dSSc than in lSSc. This result may be due to the higher presence of sicca syndrome in dSSc than in lSSc ($p = 0.0109$). Our results support validity and reliability in Italian SSc patients of MHISS, specifically measuring SSc OHRQoL.

Keywords Systemic sclerosis · Oral health-related quality of life · Rehabilitation · Disability · Mouth · Mouth Handicap in Systemic Sclerosis scale

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Introduction

Systemic sclerosis (SSc) is a connective tissue disease characterised by fibrosis at skin and internal organs (such as gastrointestinal tract, respiratory, renal, cardiovascular and genitourinary systems) and by joint modifications and muscle impairment [1], leading to tissue retraction and atrophy, and consequently to disability and impairment of health-related quality of life (HRQoL).

Thus, outcome measures reliably assessing handicap are mandatory to properly evaluate patients and to follow-up disease evolution and treatment efficacy. In SSc, HRQoL is assessed by Short Form 36 (SF 36) [2], and global disability by Health Assessment Questionnaire (HAQ) [3], and Scleroderma HAQ (sHAQ), more specific for SSc, as it adds to HAQ 5 visual analogue scales, evaluating Raynaud's phenomenon, digital ulcers, gastrointestinal and lung symptoms and overall disease severity [4].

Hand disability was studied by specific instruments, such as Cochin Hand Function Scale (CHFS) known also as Duruoz index [5] and Hand Mobility in Scleroderma (HAMIS) scale [6], both able to assess district-specific handicap and able to follow-up disease evolution and treatments [7, 8].

The involvement of the face and oral complications are typical features of SSc, leading to aesthetic changes and impairment of the patient's self-image. The face becomes amimic, the cutaneous wrinkles disappear, around the mouth vertical furrows (due to retraction of the skin) develop, and the nose becomes sharp. Face and mouth changes also include telangiectasia, sicca syndrome and thinning and reduction of mouth width (microcheilia) and opening (microstomia), also favoured by osteolysis of mandibular angles and by fibrosis of soft tissues [9–12].

Other than interfering with aesthetic self-image, these modifications affect oral health-related quality of life (OHRQoL), as they may hinder eating, by reducing the capacity to incise large parcels of solid food, cause a fatigue sensation in chewing and difficulty in speaking. The decreased oral aperture also renders daily oral hygiene measures and dental treatment more challenging. Although the involvement of face and oral tissues is commonly experienced by SSc patients, it is often underrated and not well managed in the daily practice and, in most severe cases, leads to surgical interventions, such as bilateral commissurotomies [9–12].

Recently, Mouth Handicap in Systemic Sclerosis scale (MHISS), a specific tool quantifying in SSc patients the handicap associated with mouth disability and the OHR-QoL, that explores problems not assessed by HAQ and SF 36, was developed and validated [13].

The aim of our study was to validate the Italian version of MHISS and to assess its test–retest reliability and internal and external consistency in Italian SSc patients.

Materials and methods

Forty consecutive SSc patients, fulfilling ACR criteria [14] [6 men and 34 women; 33 affected with limited SSc (lSSc) and 7 with diffuse Ssc subset (dSSc)] [15] (age: 57.27 ± 11.41 years; disease duration 9.4 ± 4.4 years),

were enrolled from the outpatient clinic and the Day Hospital of the Department of Biomedicine, Division of Rheumatology of the University of Florence and agreed by a written informed consent to participate in the study, which was approved by our institutional ethics committee and conducted in full accordance with ethical principles, including the World Medical Association Declaration of Helsinki.

Assessment

All patients underwent a clinical examination and were assessed according to international guidelines [16]. Interstitial lung disease was defined by the means of standard chest radiographs, high-resolution computed tomography, pulmonary function tests and/or BAL; pulmonary arterial hypertension by the means of color Doppler echocardiography and right heart catheterisation. Cardiac involvement was defined by the presence of pericarditis, arrhythmia or left ventricular congestive heart failure; oesophageal involvement was defined as hypomotility shown by barium radiography; muscle-skeletal involvement by the presence of arthralgias, arthritis, flexion contractures; sicca syndrome according to Vitali et al. [17]. Skin involvement was assessed by the modified Rodnan skin score [18]. The presence of active digital ulcers, telangiectasia and Raynaud phenomenon was recorded. Mouth opening was assessed in centimetres by measuring the distance between the tips of upper and lower right incisive teeth (mean of two consecutive measurements) (Table 1).

Questionnaires administration

During a routine visit, all SSc patients were administered with Italian versions of MHISS, SF36 and HAQ, and the mouth opening was assessed. To determine test–retest reliability of MHISS, all the patients were asked to answer the questionnaire a second time within 2 weeks since the first administration.

MHISS, assessing the handicap associated with mouth disability in SSc [13], consists of 12 items (each scored 0–4, with a total score ranging from 0 to 48) divided into 3 subscales: subscale 1 (5 items: 1, 3, 4, 5 and 6; range 0–20) examines handicap related to reduced mouth opening; subscale 2 (5 items: 2, 7, 8, 9 and 10; range 0–20) assesses handicap related to sicca syndrome; and subscale 3 (items 11 and 12; range 0–8) examines aesthetic concerns. The total score is obtained by summing the score for all items [13]. The English and the Italian versions are shown in Table 2.

HAQ, a self-report questionnaire, is organised in 20 items divided into 8 categories: dressing and grooming, arising, eating, walking, personal hygiene, reaching, gripping and other activities. Each item is rated from 0 (no

Table 1 Patients' characteristics

Number	40
Sex (F/M)	34/6
Age (years)	57.27 ± 11.41
Subset (lSSc/dSSc)	33/7
Disease duration (years)	9.4 ± 4.4
ILD	21/40
PAH	15/40
Cardiac involvement	20/40
Oesophageal involvement	13/40
Muscle-skeletal involvement	16/40
Sicca syndrome	22/40
Telangiectasia	10/40
Active ulcers	11/40
Raynaud	40/40
Mouth opening (cm)	4.00 ± 1.08
Skin score	11.7 ± 5.6
MSI (SF-36)	38.99 ± 9.33
PSI (SF-36)	37.32 ± 8.08
HAQ-DI	0.70 ± 0.80

ILD interstitial lung disease, *PAH* pulmonary hypertension, *MSI* mental synthetic index, *PSI* physical synthetic index, *HAQ-DI* health assessment questionnaire disease index

difficulty) to 3 (unable to do). A score for each category is the highest score for any question in the category. A disability index is calculated by adding the scores from each category and dividing by the number of categories answered and rated from 0 (less disabled) to 3 (more disabled) [3]. Validity of the Italian version of HAQ in SSc patients has been previously reported [19].

SF-36, a self-report questionnaire, consists of 36 items organized into 8 domains measuring 8 health concepts: physical functioning, role limitations due to physical problems, bodily pain, general health perceptions, vitality, social functioning, role limitations due to emotional problems and general mental health. In SF-36 domains, scores are rated so that higher values correspond to better conditions and lower scores to worse conditions (range 0–100). The 8 domains, weighted according to normative data, are also combined into a summary physical index (SPI) and a summary mental index (SMI), scored from 0 to 100, with higher values reflecting better HRQoL [2]. Validity of the Italian version of SF36 in SSc patients has been previously tested by our group [2].

Translation

MHISS was translated following a forward–backward translation procedure, with independent translations to Italian and counter-translation to English, according to

international methodology [20]. Two of the authors (ADR, SMB) independently translated the questionnaire from English to Italian, with a pooling in a common version. This version was sent to two native English speakers with good knowledge of Italian but without any knowledge of either questionnaire (the original in English and the Italian one). They back-translated the Italian version of the questionnaire into English. This version was almost identical to the initial document. The final Italian version was administered to 5 SSc patients to find out that no problems with acceptance and understanding of the questionnaire content or phrasing were present.

Statistics

Internal consistency was assessed with Cronbach's α coefficient. External consistency was evaluated by comparison with mouth opening by using Spearman's correlation.

The test–retest reliability was assessed, comparing the results of the first and second administration (at a 2-week interval), and was investigated by computing the intraclass correlation coefficient (ICC). Statistical analysis was performed using the SPSS package.

Results

All demographic and clinical characteristics of the SSc patients are shown in Table 1. Twenty-two out of 40 patients (55%) presented sicca syndrome [17]. Mouth opening, assessed as interincisor distance, was 4.00 ± 1.08 cm. SF36 mental and physical summary Indexes (SMI and SPI) were 38.99 ± 9.33 and 37.32 ± 8.08, respectively, and HAQ–DI was 0.70 ± 0.80.

The English and the Italian versions of MHISS are shown in Table 2. Total MHISS score in our patients was 17.65 ± 5.20, with scores of subscale 1 (handicap related to reduced mouth opening) of 6.60 ± 2.85 and scores of subscales 2 (handicap related to sicca syndrome) and 3 (aesthetic concerns) of 7.82 ± 2.59 and 3.22 ± 1.14, respectively.

Total score and score of subscale 2 (handicap related to sicca syndrome) were higher in dSSc than in lSSc patients (Table 3). This result may be explained by the higher prevalence of sicca syndrome in dSSc in respect to lSSc (7/7–100% vs. 15/33–45.5%-; 0.0109 by Fisher exact test).

MHISS had a good test–retest reliability (ICC: 0.93) and internal consistency (Cronbach's α: 0.99).

A good external consistency was confirmed by the significant correlation of MHISS with mouth opening (rho: $-0,3869$, *p*: 0.0137).

Table 2 Mouth Handicap in Systemic Sclerosis (MHISS) scale

	Never	Rarely	Occasionally	Often	Always
Mouth Handicap in Systemic Sclerosis (MHISS) scale *English*					
1. I have difficulties opening my mouth	0	1	2	3	4
2. I have to avoid certain drinks (sparkling, alcohol, acidic)	0	1	2	3	4
3. I have difficulties chewing	0	1	2	3	4
4. My dentist has difficulties taking care of my teeth	0	1	2	3	4
5. My dentition has become altered	0	1	2	3	4
6. My lips are retracted and/or my cheeks are sunken	0	1	2	3	4
7. My mouth is dry	0	1	2	3	4
8. I must drink often	0	1	2	3	4
9. My meals consist of what I can eat and not what I would like to eat	0	1	2	3	4
10. I have difficulties speaking clearly	0	1	2	3	4
11. The appearance of my face is modified	0	1	2	3	4
12. I have trouble with the way my face looks	0	1	2	3	4

	Mai	Raramente	Occasionalmente	Spesso	Sempre
Mouth Handicap in Systemic Sclerosis (MHISS) scale *Italian*					
1. Ho difficoltà ad aprire la bocca	0	1	2	3	4
2. Devo evitare alcuni tipi di bevande (frizzanti, alcoliche, acide)	0	1	2	3	4
3. Ho difficoltà a masticare	0	1	2	3	4
4. Il mio dentista ha difficoltà a prendersi cura dei miei denti	0	1	2	3	4
5. La mia dentatura si è alterata	0	1	2	3	4
6. Le mie labbra sono retratte e/o le mie guance sono infossate	0	1	2	3	4
7. La mia bocca è secca	0	1	2	3	4
8. Devo bere spesso	0	1	2	3	4
9. Devo mangiare le cose che posso e non le cose che vorrei	0	1	2	3	4
10. Ho difficoltà a parlare con chiarezza	0	1	2	3	4
11. L' aspetto della mia faccia si è modificato	0	1	2	3	4
12. L' aspetto della mia faccia mi crea problemi	0	1	2	3	4

Table 3 Mouth Handicap in Systemic Sclerosis (MHISS) scores in SSc Italian patients

	SSc	lSSc	dSSc	P (dSSc vs. lSSc)
MHISS subscale 1 (handicap related to reduced mouth opening)	6.60 ± 2.85	6.45 ± 2.98	7.28 ± 2.21	NS
MHISS subscale 2 (handicap related to sicca syndrome)	7.82 ± 2.59	7.45 ± 2.65	9.57 ± 1.27	0.0187
MHISS subscale 3 (handicap related to aesthetic concerns)	3.22 ± 1.14	3.15 ± 1.17	3.57 ± 0.97	NS
MHISS total	17.65 ± 5.20	17.06 ± 5.42	20.43 ± 2.88	0.0224

Association of MHISS with HAQ, SF36 and with clinical items

MHISS was not significantly related to HAQ neither to MSI and PSI of SF-36. The correlations of MHISS with age, disease duration and skin score were not significant, and MHISS scores were not significantly different between patients with interstitial lung disease, cardiac involvement, pulmonary hypertension, oesophageal involvement, muscle-skeletal involvement, sicca syndrome, telangiectasias and active ulcers and patients not presenting these clinical features.

Discussion

Our results support the validity and reliability of the Italian version of MHISS, as an instrument assessing the handicap and the impairment of OHRQoL, due to the involvement of mouth and face, in SSc Italian patients.

MHISS specifically measures face and mouth handicap and OHRQoL in SSc patients and evaluates health areas not assessed by HAQ and SF 36, such as impairment due to mouth opening reduction, mouth dryness and aesthetic concerns. In this context, the lack of correlation between MHISS and HAQ, not assessing mouth involvement, is not surprising and confirms the results presented in the validation study [13].

Also, the lack of correlation between SF36 mental and physical summary indexes, proposed in our work but not in the validation study, is in keeping with the previous finding. In fact, also SF36 questionnaire, a generic tool conceived to evaluate HRQoL in a wide range of different conditions and patients, does not assess mouth handicap in any of its items.

Given that the clinical picture of SSc is wide and variable, HAQ and SF36, routinely used in evaluating SSc patients disability and HRQoL [2, 3], should be completed by a tool specifically assessing mouth disability. MHISS is potentially useful in daily clinical practice to evaluate handicap related to mouth and face because it is simple, easily understandable and not time-consuming, thus being of help in SSc routine assessment to evaluate and score the typical involvement of face and oral tissues, felt by SSc patients as a severe problem because the disease deeply changes aesthetic aspects and affects OHRQoL, leading to important handicap in daily activities, and impairing oral hygiene and dental treatments [9–12].

As in SSc an early and regular referral to an experienced dentist or dental specialist for preventive and regular dental and periodontal care is advocated, MHISS may be easily used also in the dentist setting in order to assess OHRQoL before evaluating and/or intervening on SSc patients and in their follow-up.

The lower values of MHISS total score and subscales, in our series in respect to the original one, may be explained by the higher number of lSSc in our cohort [13]. The different composition of our sample, in respect to the sample evaluated in the validation study, may reflect, however, the reality in everyday practice. In fact, we chose to evaluate consecutive patients attending our outpatient clinic and/or our Day Hospital, whilst Mouthon et al. evaluated members of a SSc patient association attending a national meeting. This sample, as underlined by the authors, might be more likely to suffer from a severe form of the disease than the general population of SSc patients. This can be

confirmed also by the lower HAQ-DI index values in our patients than in Mouthon series [13].

In our study, scores of subscale 1, assessing handicap related to reduced mouth opening and subscale 3 of MHISS, evaluating handicap related to aesthetic concerns, are not different between lSSc and dSSc. These results are in line with those reported in the validation study [13].

In our cohort of patients, differently from Mouthon [13], scores of subscale 2, evaluating handicap related to sicca syndrome, were higher in dSSc than in lSSc group. This is probably due to the higher proportion of patients affected from sicca syndrome in dSSc than in lSSc group.

We found a good correlation of MHISS with an item notably assessing orofacial disability in SSc patients, represented by mouth opening, also assessed in the validation work, used to test MHISS external consistency. This result is different from Mouthon's findings, which showed a fair correlation between these two items, but is in keeping with the results shown in a study of our group evaluating handicap and functionality of the hands in SSc patients [21]. We found that HAMIS scores were strongly correlated not only with a tool assessing hand function such as Cochin Hand Function Scale index but also with fist closure (evaluating hand disability) [21].

An important characteristic of a clinimetric scale is its sensitivity to change and the ability in following up the modifications over time of the items assessed. This was confirmed for tools evaluating hand functionality in SSc, such as Cochin Hand Function [5] and HAMIS scale [6], that were able to assess district-specific handicap and to follow-up disease evolution and treatments [7, 8]. In a longitudinal study following-up hand involvement and activities of daily living in early SSc patients, HAMIS was found to be the most sensitive test in assessing hand mobility changes over time [7]. A study from our group has showed that fist closure, HAMIS test and Cochin hand functional disability scale improved at the end of a 9-week rehabilitation protocol based on connective tissue massage and Mc Mennell joint manipulation and home exercises on SSc patients hands [8].

Similar results were shown for MHISS. In fact, in a recent study, we showed that in SSc patients with face involvement, a 9-week treatment with a combined rehabilitation program based on connective tissue massage, Kabat technique, facial physiotherapy and home exercises for mimic face muscles significantly improved mouth opening, facial skin score and MHISS. On the contrary, no improvement was shown in HAQ and SF36 scores [22].

These data suggest that MHISS might be introduced as a routine tool not only to assess SSc patients undergoing face and mouth rehabilitation, but also to evaluate the results after dental interventions.

In conclusion, validation in different languages versions of already validated questionnaires is of pivotal importance in standardizing the patient's assessment and follow-up over different countries. MHISS is a self-administered questionnaire specifically developed to measure mouth handicap in SSc, brief and easy to be filled in. Our results support its validity and reliability in the evaluation of mouth handicap in Italian SSc patients.

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