Reliability and validity of the European Portuguese version of the EULAR Systemic Sclerosis Impact of Disease (ScleroID) questionnaire

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Abstract

Objective: The European Alliance of Associations for Rheumatology (EULAR) Systemic Sclerosis Impact of Disease questionnaire (ScleroID) is a new disease-specific and patient-derived outcome measure of systemic sclerosis (SSc) burden. This work aims to evaluate the feasibility, reliability and construct validity of the European Portuguese version of the EULAR ScleroID.

Methods: Participants were consecutively selected from all patients receiving care in the rheumatology department of a tertiary hospital who fulfilled ACR/EULAR classification criteria for SSc or EUSTAR criteria for Very Early Diagnosis of Systemic Sclerosis (VEDOSS). Feasibility was assessed by the proportion of missing ScleroID items. Reliability was assessed by internal consistency (Cronbach's alpha) and test-retest reliability (intraclass correlation coefficients [ICC]). Construct validity was evaluated by principal component analysis, by testing for ScleroID score differences between groups stratified by demographic data and disease subtypes, and by correlations between the ScleroID score and other measures of similar constructs (HAQ-DI, SHAQ, SF-36, EQ-5D, UCLA GIT 2.0 and ABILHAND-SSc). Floor and ceiling effects were measured.

Results: A total of 53 patients were enrolled, 12 of whom participated in a re-test. Two patients (3.8%) had missing data regarding at least one item of the ScleroID questionnaire. The ScleroID had a high level of internal consistency (Cronbach's alpha = 0.928) and moderate test-retest reliability (ICC 0.68, 95%IC 0.19-0.90). Principal component analysis revealed two components that were clinically meaningful, one mostly related to hand and musculoskeletal involvement, and the other to internal organ involvement. No floor/ceiling effects were identified for the total score. ScleroID was statistically significantly different between SSc subtypes, but there was no difference regarding sex, age or disease duration. Good correlations were found between the ScleroID and all other patient-reported outcomes, except for the SF-36 social role functioning, SHAQ Breathing VAS and SHAQ finger ulcer VAS scores (moderate correlation for all).

Conclusion: The European Portuguese version of the ScleroID score appears to be a feasible, reliable and valid measure of SSc disease burden. Further validation in other Portuguese cohorts is needed to ensure the generalizability of these findings.

Keywords: Validity; Scleroderma and related disorders; Outcome measures; Reliability; Patient reported experience measure.

Introduction

Patient-reported outcome measures (PROMs) capture the patient's perspective and experiences, serving as a vital part of the diverse set of outcome instruments available in clinical practice and clinical trials[1]. The development of PROMs in systemic sclerosis (SSc) has been a longstanding challenge, due to the heterogeneity and quantity of disease manifestations and constructs to measure. On the other hand, the difficulty of evaluating disease activity using objective measures such as laboratory investigations underscore the need to develop these PROMs.[1] Although few disease-specific instruments have been developed, most of the PROMs used in SSc are adapted from other diseases[2].

Recently, the European Alliance of Associations for Rheumatology Systemic Sclerosis Impact of Disease questionnaire (ScleroID) was developed, the first disease-specific and patient-derived PROM that aims to measure disease burden by assessing different dimensions of SSc. This questionnaire has been successfully validated in a large European clinical cohort using multiple translations[3]. It includes ten health dimensions, selected and weighted by patients, and reviewed by SSc experts: Raynaud's phenomenon, hand function, upper gastrointestinal (GI) symptoms, pain, fatigue, lower GI symptoms, life choices and activity limitation, body mobility, dyspnea, and digital ulcers. The final score is calculated as a weighted sum of all the items, each scored from 0 to 10. The ScleroID has been translated and cross-culturally adapted into European Portuguese following a forward-backward method, followed by a review by Portuguese SSc experts and a field test with cognitive debriefing by SSc patients.[4]

Face validity of the original ScleroID has been ensured by the involvement of patients and SSc experts in all steps of its development, including domain selection, question formulation and item weight. Similarly, the face validity of the European Portuguese translation was ensured by the involvement of SSc experts and patients.

The goal of this study was to assess the feasibility, reliability and construct validity of the European Portuguese version of the ScleroID.

Methods

Design, setting and study population

This study was conducted using a cross-sectional design. Participants were consenting adults consecutively selected from all patients receiving care in the rheumatology department of a tertiary hospital who fulfilled the 2013 American College of Rheumatology (ACR)/ European Alliance of Associations for Rheumatology (EULAR) criteria for the classification of SSc[5] or the Very Early Systemic Sclerosis (VEDOSS) criteria of the European Scleroderma Trials and Research Group (EUSTAR) [6]. Exclusion criteria were not being a native Portuguese speaker and reader and not being able to provide consent. Data was collected between January 2022 and June 2024.

Data collection and variable definitions

One set of data was collected from each patient at one clinical visit, including age, sex, years of education, employment status, disease subtype (limited cutaneous SSc [lcSSc]; diffuse cutaneous SSc [dcSSc]; SSc sine scleroderma, VEDOSS), disease duration, clinical manifestations, and positivity for antinuclear antibodies (ANAs) and specific SSc antibodies.

Additionally, the following measurement instruments were collected, by self-completion of the European Portuguese versions of the questionnaires:

- ScleroID, a disease-specific patient-reported measure of SSc disease burden. The questionnaire is comprised of 10 items, scored from 0 (no impact) to 10 (extreme impact): Raynaud's phenomenon, hand function, upper GI symptoms, pain, fatigue, lower GI symptoms, life choices and activity limitation, body mobility, dyspnea, and digital ulcers. Each item is multiplied by a weight and the total score ranges from 0 to 10. The face validity, construct validity, internal consistency, test-retest reliability and sensitivity to change of the English version were previously tested and considered satisfactory[3]. The translation and cross-cultural adaptation of the original English version into European Portuguese was recently accomplished[4], but this version of the questionnaire is yet to be validated in Portuguese patients. The Portuguese version of the ScleroID questionnaire can be found in the supplementary materials (Appendix I).

- Short Form Health Survey (SF-36v2), a patient-reported measure of functional health and well-being comprised of 8 dimensions: physical functioning, bodily pain, role limitations due to physical health, general health perception, mental health, role

limitations due to emotional problems, vitality, and social functioning. The final score ranges from 0 (worst possible health) to 100 (best health status) and a score of 50 represents the mean for the general population. The questionnaire has been translated into European Portuguese, and the Portuguese population norms have been established[7].

- European Quality of Life 5 Dimensions 5 Levels (EQ-5D-5L), a self-reported measure of health-related quality of life for clinical and economic appraisal. It is composed of five dimensions, scored from 1 (best state) to 5 (worst state): mobility, self-care, usual activities, pain/discomfort, anxiety/depression. Each health state is assigned a unique score using a value set derived from specific populations. The value set for EQ-5D-5L has been determined for the Portuguese population, and the index ranges from -0.603 (worst state) to 1 (best state).[8] Additionally, a visual analogue scale (EQ-VAS) is scored, ranging from 0 (worst imaginable health) to 100 (best imaginable health).

- University of California Los Angeles Scleroderma Clinical Trials Consortium gastrointestinal tract 2.0 (UCLA-GIT 2.0), which captures the impact on quality of life of SSc-related GI tract involvement. The questionnaire has 7 scales - reflux, distention/bloating, diarrhoea, faecal soilage, constipation, emotional well-being, and social functioning[9]. All scales are scored from 0 (best state) to 3 (worst state), except the diarrhoea (0-2) and constipation (0-2.5) scales. The total score ranges from 0 (no GI symptoms) to 2.83. Recently, the European Portuguese version of the score has been validated[10].

- Scleroderma Health Assessment Questionnaire (SHAQ), comprised of the Health Assessment Questionnaire Disability Index (HAQ-DI) and six additional visual analogue scales (VASs) —pain, GI symptoms, breathing, Raynaud's phenomenon, finger ulcer, and overall disease severity. The HAQ-DI contains 20 items and measures eight domains: dressing and grooming, arising, eating, walking, hygiene, reach, grip, and activities[11]. Each item is scored from 0 (without difficulty) to 3 (unable to do). The highest-scored item in each domain determines the total score for that domain, except for the necessity of aids or devices, in which case the minimum score for the domain is 2. The total score is the average of the domains and ranges from 0 to 3. Each additional VAS has a 1-week recall period and ranges from 0 to 100mm. Recently, the European Portuguese version of the SHAQ was validated[12].

- The SSc-adapted ABILHAND questionnaire (ABILHAND-SSc), a measure of hand ability validated in SSc patients, comprised of 26 items, which are manual tasks rated by each patient according to their ability to perform them (impossible, difficult or easy). The final score is a measure of the patient's hand ability that ranges from 0 to 100%. The score has been subjected to a European Portuguese translation and cross-cultural adaptation,[13] but this version is yet to be validated. As far as the authors are aware, this is the only questionnaire directly measuring hand function in SSc patients that has been translated and culturally adapted into European Portuguese.

ScleroID validation

Feasibility and missing data

To evaluate the feasibility of the European Portuguese version of ScleroID, the proportion of missing values and their distribution among the 10 items was assessed. A proportion of missing values below 5% was considered ideal. Afterwards, for the remaining statistical analysis, missing values of the ScleroID score were imputed by the mean of the remaining cohort for the respective item, as recommended by the original authors[3].

Reliability

Internal consistency was considered acceptable if Cronbach's alpha ≥0.7. Test-retest reliability was assessed using the intraclass correlation coefficient (ICC). Patients in a stable disease state (assessed by the attending physician) and receiving stable treatment were invited to retake the ScleroID within 15 to 30 days after their initial response and to send the filled questionnaire by mail. The authors considered this interval appropriate to minimize recall bias and ensure clinical stability. ICC estimates and 95% confidence intervals were calculated based on a single rating, absolute-agreement, 2-way mixed-effects model. ICCs were interpreted as follows: <0.5 – poor, 0.5 ≤ ICC ≤ 0.75 – moderate; 0.75 < ICC ≤ 0.90 – good; >0.9 – excellent.

Construct Validity

The dimensionality of ScleroID was assessed by principal component analysis (PCA). The suitability of PCA was assessed prior to analysis by testing two assumptions: linearity between all variables in the scale, and sampling adequacy. The first was tested by inspecting a correlation matrix, and linearity was assumed if all variables had at least one correlation coefficient ≥0.3. Sampling adequacy was tested through the Kaiser-Meyer-Olkin (KMO) measure and Bartlett's

test of sphericity. KMO measure was interpreted as follows: 0.7 ≤ KMO < 0.8 – acceptable; 0.8 ≤ KMO < 0.9 – good; KMO ≥ 9 – very good. Bartlett's test of sphericity was considered statistically significant if $p<0.05$. If all assumptions were met, PCA was performed. Components with Eigenvalues ≥1 were kept, and each item was considered to load into a component if the correlation was ≥0.6 (varimax orthogonal rotation). An item was considered to cross-load into multiple components if correlation coefficients were similar (Δ≤0.3) for those components.
Floor or ceiling effects were considered present if >15% of patients scored either the minimum or maximum value possible, respectively.

Hypothesis testing was used to evaluate ScleroID differences between groups stratified by sex, age, SSc subtype and disease duration. Convergent validity was assessed by testing correlations of the ScleroID score with other measures of similar constructs – HAQ-DI, SHAQ (disability), SF-36 (functional health and well-being), EQ-5D (quality of life), UCLA GIT 2.0 (impact of GI symptoms) and ABILHAND-SSc (hand function).

Continuous variables were described as means and standard deviations (SD) or medians and interquartile ranges (IQR), according to the normality of their distribution. Normality was assessed through the calculation of the z-scores for skewness and kurtosis (normal distribution assumed if the z-score was within ±2.58) if the sample was >50, and the Shapiro-Wilk test if the sample was <50. Categorical variables were described as proportions (%). For continuous variables, comparisons between groups were made using the t-test or Mann-Whitney U test (dichotomic independent variable) and the one-way ANOVA or Kruskal-Wallis H tests (categorical independent variable with >2 groups), according to the normality of their distribution and the presence of outliers. Associations between continuous variables were assessed through Pearson's or Spearman's correlation coefficients, depending on the normality of their distribution and the presence/absence of a linear relationship between the two variables. Strengths of correlation were classified as follows: low (≤3), moderate (0.3-0.49), good (0.5-0.79), very good (≥0.8). Statistical significance was set at $p<0.05$ for all tests. Statistical analysis was performed using IBM SPSS Statistics (v. 26.0.0.0).

Results

Sample

A total of 53 patients were enrolled in this study, 84.8% female, with mean age of 58.7 years and median disease duration of 10.9 years. Over half of the patients had lcSSc (58.5%), 28.3%

had dcSSc, 5 were classified as VEDOSS, and 2 had SSc sine scleroderma. Table I summarizes sociodemographic data, lifestyle and disease characteristics. Data regarding the measurement instruments EQ-5D-5L, SF-36v2, SHAQ, UCLA-GIT 2.0, and ABILHAND-SSc are presented in Table II. Mean total score of ScleroID was 4.60. The ranges, means and proportions of missing data for the total score and individual items are described in Table III.

Feasibility and missing data

Two patients (3.8%), both with lcSSc, had missing data regarding at least one item on the ScleroID questionnaire, and missing data was evenly distributed among the items (Table III). There was a low proportion of missing data for all individual items (<2%), in line with the EQ-5D-5L, SF-36v2, SHAQ and ABILHAND-SSc instruments. Missing items were imputed by the mean of the remaining cohort for that item, as described previously.

Reliability

The ScleroID had a high level of internal consistency, as determined by a Cronbach's alpha of 0.928, which remained high after a sensitivity analysis excluding VEDOSS and SSc sine scleroderma patients (Cronbach's alpha = 0.823). Although most patients were in a stable disease state, adherence to the retest was low, with twelve patients (22.3% of the whole cohort) mailing back the results, all of which had lcSSc (41.7%) or dcSSc (58.3%). ICC estimates and their 95% CIs are represented in Table IV. The items “Hand Function” and “Upper GI symptoms” had poor test-retest reliability; “Raynaud’s phenomenon”, “Pain”, “Lower GI symptoms”, “Daily activities”, “Mobility” and “Dyspnoea” had moderate reliability; “Fatigue” and “Digital ulcers” had good reliability. The total score had moderate test-retest reliability.

Construct validity

Inspection of the correlation matrix showed that all variables had at least one correlation coefficient ≥ 0.3. The sampling adequacy was good (KMO = 0.880) and Bartlett’s test of sphericity was statistically significant ($p<0.0001$). The PCA revealed two components with Eigenvalues ≥ 1 that explained 72.4% of variance: component one with Eigenvalue=6.24 (variance explained = 62.4%) and component two with Eigenvalue=1.01 (variance explained = 10.1%). The loading of each item onto the two components (rotated coefficients) can be found in the Appendix II of the supplementary materials.

All measures significantly loaded onto one of the two components. Component one was made of the following items: “Raynaud’s phenomenon”, “Hand Function”, “Pain”, “Fatigue”, “Daily

Activities", "Mobility", and "Digital ulcers". Component two was made of the following items: "Upper GI symptoms", "Lower GI symptoms" and "Dyspnoea". Fatigue had similar loading in both components (Δ≤0.3), as the item likely cross-loads them both.

Regarding floor/ceiling effects, no patient had the lowest or highest possible total ScleroID score. However, a floor effect was observed for the individual items "Upper GI symptoms", "Lower GI symptoms", "Mobility", "Dyspnoea" and "Digital ulcers", while no ceiling effects were identified (Table III).

ScleroID differences between groups stratified by sex, age and selected clinical variables are presented in Table V. The ScleroID score was statistically significantly different between SSc subtypes, but *post-hoc* pairwise comparisons showed no difference between any two groups. There was no difference regarding sex, age group or disease duration. ScleroID did not correlate with the continuous variables age (r=0.049, p=0.726) and disease duration (ρ=0.000, p=0.999).

The correlation coefficients between total ScleroID score and other patient-reported outcomes are presented in Table VI. A good correlation with all scores was found, except for the SF-36 social role functioning, SHAQ Breathing VAS and SHAQ finger ulcer VAS scores (moderate correlation for all). The direction of the correlation was also aligned with the intended interpretation of each score.

To evaluate the impact of including VEDOSS and SSc sine scleroderma patients in the assessment of construct validity, a sensitivity analysis was performed in which only dcSSc and lcSSc patients were included. The results of the PCA, of comparisons between groups stratified by clinical and sociodemographic variables and of the correlations with age, disease duration and other scores were similar to the main analysis, and their overall conclusion the same (Appendix III, IV and V of the supplementary materials).

Discussion

PROMs are a crucial aspect of assessing disease activity and burden in rheumatic inflammatory diseases. The EULAR ScleroID questionnaire is a comprehensive PROM, recently developed with strong input from patients, with the purpose of capturing disease impact in its various health domains[3].

We have shown that the European Portuguese version of the ScleroID is a feasible, valid and reliable measure of disease burden in Portuguese SSc patients. Its simple design (10 items rated from 0 to 10) contributes to its feasibility, demonstrated by the low proportion of missing data.

The final score, ranging from 0 to 10 (lowest to highest burden) is easily calculated and interpreted in clinical and research settings.

The questionnaire had a high level of internal consistency, and the total score had moderate test-retest reliability. The individual items “Hand Function”, “Upper GI symptoms” and “Dyspnoea” had particularly poor test-retest reliability and the confidence intervals for the ICC estimates were generally wide. While this could be explained by real variability in symptom severity or measurement error, we believe the most likely explanation is the small sample size of the test-retest analysis (22.3% of the cohort), leading to a lack of precision. Also of note, the test-retest reliability in VEDOSS or SSc sine scleroderma patients is unclear, as none of these patients participated in the retest.

Construct validity was first assessed by principal component analysis, which revealed two components. The first included the items “Raynaud’s phenomenon”, “Digital ulcers”, “Hand Function”, “Pain”, “Daily Activities”, “Fatigue” and “Mobility”. The second was composed of “Upper GI symptoms”, “Lower GI symptoms” and “Dyspnoea”. These components are clinically meaningful - the first is mostly related to hand and musculoskeletal involvement, and the second to internal organ involvement (GI tract and lung). The former explained a higher proportion of the variance of the ScleroID score. Also of note, the “Fatigue” item had similar loading in both components and thus cross-load is likely.

The ScleroID score demonstrated no ceiling or floor effects, as no single patient achieved the minimum possible of 0 or the maximum of 10. As such, the whole score seems able to capture a wide range of SSc disease impact. However, it should be noted that floor effects were found for the items “Upper GI symptoms”, “Lower GI symptoms”, “Mobility”, “Dyspnoea” and “Digital ulcers”. This could imply that these individual items are not sensitive enough to detect small differences in their respective domains, particularly in patients with mild symptoms.

The total score was statistically significantly different across SSc subtypes, although pairwise comparisons showed no difference between any two groups. This was likely due to the low sample size in the VEDOSS and SSc sine scleroderma subgroups, which had numerically lower ScleroID scores when compared to both dcSSc and lcSSc. This group difference is clinically expected and contributes to the score’s construct validity. On the other hand, no difference regarding sex, age or disease duration was noted. The ScleroID score had good correlations with measures of disability (SHAQ), functional health and well-being (SF-36), quality of life (EQ-5D),

GI symptoms impact (UCLA GIT 2.0) and hand function (ABILHAND-SSc), showing good convergent validity.

This study had a few weaknesses. One limitation already outlined is the low sample size in the test-retest reliability analysis, in addition to the assumption that the disease remains stable in the 15 to 30 days between tests. Additionally, all included patients were recruited from a single tertiary centre, which limits the generalizability of our findings. Another clear weakness is the absence of a sensitivity to change analysis. In the original development cohort, the ScleroID had better sensitivity to change than all other comparator PROMs.[3] As for the strengths, this study involved patients with varying educational backgrounds and employment status. Patients with different SSc subtypes were also included, although the number of SSc sine scleroderma and VEDOSS patients was low. Sensitivity analyses excluding these patients yielded results and overarching conclusions regarding internal consistency and construct validity that were consistent with those of the whole cohort.

The results of this study apply to the Portuguese population and not to other Portuguese-speaking populations, for which specific cross-cultural adaptations and validation analysis should be performed due to the linguistic and cultural differences. Additionally, further validation efforts in different centres should be pursued to ensure generalizability to the Portuguese population.

Conclusion

The European Portuguese version of the ScleroID appears to be feasible, reliable and valid for use in clinical and research settings to capture the disease burden of SSc. These results should be further validated in different cohorts.

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Tables and Figures

Table I – Patient's characteristics

		Missingness (%)
Age (years) – mean ± SD	58.7 ± 12.6	0
Female sex – n (%)	45 (84.9)	0
SSc subtype		0
lcSSc – n (%)	31 (58.5)	
dcSSc – n (%)	15 (28.3)	
SSc sine scleroderma – n (%)	2 (3.8)	
VEDOSS – n (%)	5 (9.4)	
Disease duration – median (IQR)	10.9 (12.3)	0
Clinical manifestations		
Skin thickening proximal to MCPs – n (%)	22 (41.5)	0
Puffy fingers – n (%)	17 (32.1)	0
Sclerodactyly – n (%)	32 (60.4)	0
Digital ulcers – n (%)	22 (41.5)	0
Pitting scars – n (%)	18 (34.0)	0
Telangiectasia – n (%)	35 (66.0)	0
Capillaroscopy abnormalities – n (%)	35 (66.0)	0
Pulmonary arterial hypertension – n (%)	4 (7.5)	0
Interstitial lung disease – n (%)	11 (20.8)	0
Raynaud's phenomenon – n (%)	51 (96.2)	0
Arthralgia – n (%)	28 (52.8)	0
Myositis – n (%)	4 (7.5)	0
Upper GI involvement – n (%)	33 (62.3)	0
Lower GI involvement – n (%)	7 (13.2)	0
Renal involvement – n (%)	1 (1.9)	0
Immunological profile		
ANAs – n (%)	52 (98.1)	0
Anti-centromere – n (%)	28 (52.8)	0
Anti-topoisomerase I – n (%)	15 (28.3)	0
Employment status		1.9
Employed – n (%)	18 (34.6)	
Unemployed – n (%)	4 (7.6)	
Retired – n (%)	30 (57.7)	
Education		1.9
No formal education – n (%)	1 (1.9)	
4 years – n (%)	14 (26.9)	
5-12 years – n (%)	24 (46.2)	
>12 years – n (%)	13 (25.0)	

ANAs – antinuclear antibodies; dcSSc – diffuse cutaneous systemic sclerosis; GI – gastrointestinal; lcSSc – limited cutaneous systemic sclerosis; MCPs – metacarpophalangeal joints; SD – standard deviation; VEDOSS – very early diagnosis of systemic sclerosis

Table II – Patient reported outcome measures

	Measure	Min-Max	Mean±SD	Median (IQR)	Missingness (%)
EQ-5D-5L	Index	0.19-1.00	0.71±0.21	—	0
	VAS	4-90	60.5±20.4	—	1.9
SF-36v2	Physical functioning	5-100	52.5±24.0	—	0
	Physical role functioning	0-100	43.2±25.6	—	1.9
	Bodily pain	0-100	43.7±22.1	—	0
	General health perceptions	0-75	33.4±16.2	—	0
	Vitality	0-87.5	36.2±20.7	—	0
	Social role functioning	0-100	61.1±24.7	—	0
	Emotional role functioning	0-100	51.2±25.1	—	1.9
	Mental health	10-100	55.0±24.4	—	0
SHAQ	HAQ-DI	0-2.22	0.99±0.63	—	0
	Pain VAS	1-86	42.4±24.5	—	1.9
	GI symptoms VAS	0-100	27.7±30.1	—	1.9
	Breathing VAS	0-86	25.1±28.7	—	1.9
	Raynaud's phenomenon VAS	0-88	41.6±29.7	—	1.9
	Finger ulcer VAS	0-92	25.2±30.2	—	1.9
	Overall disease severity VAS	0-90	47.0±27.5	—	1.9
UCLA-GIT 2.0	Reflux	0-2.13	0.61±0.57	—	5.7
	Distension/bloating	0-3	1.01±0.85	—	1.9
	Faecal soilage	0-3	—	0.00 (0.00)	1.9
	Diarrhoea	0-2	—	0.50 (1.00)	1.9
	Social functioning	0-2.67	—	0.00 (0.54)	5.67
	Emotional wellbeing	0-2.89	—	0.22 (0.86)	9.4
	Constipation	0-2.50	—	0.50 (0.75)	7.6
	Total score	0-2.02	—	0.36 (0.60)	13.2
ABILHAND-SSc	Measure	33.7-100	37.2±11.6	—	0

IQR – interquartile range; Max – maximum; Min – minimum; SD – standard deviation; VAS – Visual analogue scale

Table III – ScleroID questionnaire

Item	Mean±SD	Min-Max	Lowest possible value N (%)	Highest possible value N (%)	Missingness N (%)
Raynaud's phenomenon	5.02±2.87	0-10	4 (7.5)	2 (3.8)	1 (1.9)
Hand Function	5.85±2.82	0-10	4 (7.5)	2 (3.8)	0 (0)
Upper GI symptoms	3.72±3.11	0-9	11 (20.8)	4 (7.5)	0 (0)
Pain	5.38±3.01	0-10	4 (7.5)	3 (5.7)	0 (0)
Fatigue	5.79±2.92	0-10	2 (3.8)	4 (7.5)	0 (0)
Lower GI symptoms	4.29±3.24	0-9	11 (20.8)	0 (0)	1 (1.9)
Daily activities	4.92±2.80	0-10	5 (9.4)	2 (3.8)	1 (1.9)
Mobility	4.75±3.26	0-10	8 (15.1)	2 (3.8)	0 (0)
Dyspnoea	2.67±2.90	0-9	21 (39.6)	0 (0)	0 (0)
Digital ulcers	2.68±3.26	0-9	26 (49.1)	0 (0)	0 (0)
Total Score	4.60±2.37	0.33-8.40	0 (0)	0 (0)	0 (0)

GI – gastrointestinal; Max – maximum; Min – minimum; SD – standard deviation

Table IV – Test-retest reliability of the ScleroID

Item	ICC	95% CI
Raynaud's phenomenon	0.660	0.147-0.890
Hand Function	0.457	-0.125-0.806
Upper GI symptoms	0.409	-0.169-0.783
Pain	0.723	0.296-0.911
Fatigue	0.845	0.548-0.953
Lower GI symptoms	0.600	0.073-0.865
Daily activities	0.731	0.283-0.915
Mobility	0.731	0.296-0.915
Dyspnoea	0.500	-0.016-0.820
Digital ulcers	0.754	0.349-0.922
Total Score	0.676	0.190-0.895

CI – confidence interval; GI – gastrointestinal; ICC - interclass correlation coefficient

Table V – ScleroID score stratified by sociodemographic and clinical variables

		N	Median (IQR)	Statistic	p-value
Sex*	Female	45	4.98 (4.08)	U=159.0, z = 0.52	0.617
	Male	8	3.31 (3.89)		
SSc subtype*#	Diffuse	15	5.97 (3.10)	H(3)=7.89	**0.048**
	Limited	31	4.79 (3.94)		
	VEDOSS	5	1.22 (3.38)		
	Sine scleroderma	2	2.17 (—)		
Age+	<50 years	12	4.35 (5.18)	H(2)=1.38	0.505
	[50-70[years	30	4.96 (3.91)		
	≥70 years	11	3.58 (3.88)		
Disease duration*	<5 years	13	5.01 (4.10)	U= 221.5, z = -0.80	0.433
	≥5 years	40	4.57 (3.92)		

IQR – interquartile range; H - Kruskal-Wallis H; U - Mann-Whitney U, z = standardized test statistic

*Distributions of ScleroID scores were not similar between groups (assessed by visual inspection of a boxplot). As such, statistical analysis was carried by evaluation of mean ranks. Medians and IQR are reported for easier interpretation.

#Pairwise comparisons were performed using Dunn's (1964) procedure with a Bonferroni correction for multiple comparisons. This post hoc analysis revealed no statistically significant differences in ScleroID score between any two groups.

+Age categories were calculated by identifying quartiles and rounding to the nearest multiple of 5.

Table VI – Correlations between ScleroID score and other patient-reported outcomes

		Coefficient	p-value
EQ-5D-5L	Index	r=-0.746	p<0.0001
	EQ-VAS	r=-0.572	p<0.0001
SF-36v2	Physical functioning	r=-0.609	p<0.0001
	Physical role functioning	r=-0.598	p<0.0001
	Bodily pain	ρ=-0.658	p<0.0001
	General health perceptions	r=-0.591	p<0.0001
	Vitality	r=-0.631	p<0.0001
	Social role functioning	r=-0.450	p<0.001
	Emotional role functioning	r=-0.573	p<0.0001
	Mental health	r=-0.529	p<0.0001
SHAQ	HAQ-DI	r=0.644	p<0.0001
	Pain VAS	r=0.644	p<0.0001
	GI Symptoms VAS	ρ=0.531	p<0.0001
	Breathing VAS	ρ=0.433	p<0.001
	Raynaud's phenomenon VAS	ρ=0.666	p<0.001
	Finger ulcer VAS	ρ=0.464	p<0.001
	Overall disease severity VAS	ρ=0.667	p<0.0001
UCLA-GIT 2.0	Total Score	ρ=0.631	p<0.0001
ABILHAND-SSc	Measure	ρ=-0.698	p<0.0001

r – Pearson's coefficient; VAS – visual analogue scale; ρ – Spearman's coefficient

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Supplementary Material

Appendix I – European Portuguese Version of the ScleroID questionnaire

Impacto da Doença - Esclerose Sistémica
Questionário EULAR ScleroID

Tendo em conta as diferentes dimensões da esclerose sistémica, indique o quanto é que elas o/a afetaram, durante os últimos 7 dias. Por favor, responda usando a escala, e escolhendo o número que melhor quantifica cada uma das seguintes dimensões:

1- Fenómeno de Raynaud:
Faça um círculo à volta do número que melhor descreve a gravidade do seu fenómeno de Raynaud, durante os últimos 7 dias.

Nenhuma	0	1	2	3	4	5	6	7	8	9	10	Extremamente grave

2- Avaliação da função das mãos:
Faça um círculo à volta do número que melhor descreve as limitações que sente na função das mãos, devido à sua esclerose sistémica, durante os últimos 7 dias.

Nenhuma	0	1	2	3	4	5	6	7	8	9	10	Limitação grave

3- Sintomas do sistema digestivo superior (por exemplo: dificuldade em engolir, refluxo, vómitos):
Faça um círculo à volta do número que melhor descreve a gravidade dos sintomas do sistema digestivo superior, devido à esclerose sistémica, durante os últimos 7 dias.

Nenhuma	0	1	2	3	4	5	6	7	8	9	10	Extremamente grave

4- Dor:
Faça um círculo à volta do número que melhor descreve a intensidade da dor que sentiu, devido à esclerose sistémica, durante os últimos 7 dias.

Nenhuma	0	1	2	3	4	5	6	7	8	9	10	Extremamente grave

5- Cansaço/Fadiga:

Faça um círculo à volta do número que melhor descreve a fadiga/cansaço geral que sentiu, devido à esclerose sistémica, durante os últimos 7 dias.

Nenhum	0	1	2	3	4	5	6	7	8	9	10	Extremamente grave

6- Sintomas do sistema digestivo inferior (por exemplo: sensação de "barriga inchada", diarreia, obstipação, incontinência fecal)

Faça um círculo à volta do número que melhor descreve a gravidade dos sintomas do sistema digestivo inferior, devido à esclerose sistémica durante os últimos 7 dias.

Nenhuma	0	1	2	3	4	5	6	7	8	9	10	Extremamente grave

7- Limitações nas atividades diárias e opções de vida (por exemplo: cuidados pessoais, atividades da vida social, trabalho)

Faça um círculo à volta do número que melhor descreve a gravidade das limitações nas atividades de vida diária e opções de vida, devido à esclerose sistémica durante os últimos 7 dias.

Nenhuma	0	1	2	3	4	5	6	7	8	9	10	Extremamente grave

8- Mobilidade corporal

Faça um círculo à volta do número que melhor descreve, o quanto a mobilidade do seu corpo foi afetada pela esclerose sistémica, durante os últimos 7 dias.

Nada	0	1	2	3	4	5	6	7	8	9	10	Extremamente grave

9- Falta de ar

Faça um círculo à volta do número que melhor descreve a gravidade da falta de ar que sentiu, devido à esclerose sistémica, durante os últimos 7 dias.

Nenhuma	0	1	2	3	4	5	6	7	8	9	10	Extremamente grave

10-Úlceras (Feridas) nos dedos

Faça um círculo à volta do número que melhor descreve o quanto as suas úlceras digitais o afetaram, durante os últimos 7 dias.

Nada	0	1	2	3	4	5	6	7	8	9	10	Extremamente grave

ScleroID scoring

ScleroID final value = (**Raynaud** value (range 0-10) x 0.117) + (**fatigue** value (range 0-10) x 0.114) + (**Hand function** value (range 0-10) x 0.109) + (**pain** value (range 0- 10) x 0.104) + (**life choices** value (range 0-10) x 0.098) + (**upper GI symptoms** value (range 0-10) x 0.096) + (**body mobility** value (range 0-10) x 0.096) + (**lower GI symptoms** (range 0-10) x 0.093) + (**dyspnea** value (range 0-10) x 0.091) + (**digital ulcers** value (range 0-10) x 0.083)

Thus, the range of the final SCLEROID value is 0-10 where higher figures indicate worse status.

Appendix II – Principal component analysis (item loading) of the whole cohort

Loading of items onto each component

	Rotated* component coefficients	
	Component 1	**Component 2**
Raynaud	**0.805**	0.221
Hand Function	**0.846**	0.350
Upper GI symptoms	0.149	**0.875**
Pain	**0.835**	0.357
Fatigue	**0.644**	0.599
Lower GI symptoms	0.457	**0.768**
Daily activities	**0.779**	0.400
Mobility	**0.779**	0.400
Dyspnoea	0.245	**0.700**
Digital ulcers	**0.645**	0.093

*Varimax rotation; Major loadings (correlations >0.6) in bold; GI – gastrointestinal

Appendix III – Principal component analysis (VEDOSS and sine scleroderma patients excluded)

The sampling adequacy was good (KMO = 0,853) and Bartlett's test of sphericity was statistically significant (p<0.0001). The PCA revealed two components with Eigenvalues ≥ 1 that explained 69.85% of variance; component one with Eigenvalue=5.91 (variance explained = 59,1%) and component two with Eigenvalue=1.08 (variance explained = 10,8%).

Loading of items onto each component

	Rotated* component coefficients	
	Component 1	**Component 2**
Raynaud	**0.801**	0.191
Hand Function	**0.836**	0.330
Upper GI symptoms	0.146	**0.852**
Pain	**0.829**	0.362
Fatigue	**0.624**	0.595
Lower GI symptoms	0.491	**0.742**
Daily activities	**0.789**	0.382
Mobility	**0.776**	0.362
Dyspnoea	0.143	**0.711**
Digital ulcers	**0.607**	0.011

*Varimax rotation; Major loadings (correlations >0.6) in bold; GI – gastrointestinal

Appendix IV – ScleroID differences by sociodemographic and clinical variables (VEDOSS and sine scleroderma patients excluded)

		N	Mean (SD)	Statistic	p-value
Sex	Female	38	5.10 (2.22)	t(44)=1.055	0.297
	Male	8	4.19 (2.25)		
Age[+]	<50 years	8	4.81 (2.30)	F(2,43)=0.171	0.844
	[50-70[years	29	5.08 (2.19)		
	≥70 years	9	4.60 (2.51)		
Disease duration	<5 years	13	5.08 (4.10)	t(44) = 0.250	0.804
	≥5 years	33	4.89 (2.26)		

F - ANOVA F-test; SD – standard deviation; t - Student's t-test

[+]Age categories were calculated by identifying quartiles and rounding to the nearest multiple of 5.

ScleroID did not correlate with the continuous variables age (ρ=0.075, p=0.619) or disease duration (ρ=0.092, p=0.542)

Appendix V – Correlations between ScleroID score and other patient-reported outcomes (VEDOSS and sine scleroderma patients excluded)

		Coefficient	p-value
EQ-5D-5L	Index	r=-0.720	p<0.0001
	EQ-VAS	ρ=-0.486	p<0.001
SF-36v2	Physical functioning	ρ=-0.550	p<0.0001
	Physical role functioning	ρ=-0.514	p<0.0001
	Bodily pain	ρ=-0.596	p<0.0001
	General health perceptions	r=-0.631	p<0.0001
	Vitality	ρ=-0.563	p<0.0001
	Social role functioning	ρ=-0.373	p=0.011
	Emotional role functioning	ρ=-0.497	p<0.0001
	Mental health	ρ=-0.518	p<0.0001
SHAQ	HAQ-DI	r=0.636	p<0.0001
	Pain VAS	r=0.538	p<0.0001
	GI Symptoms VAS	ρ=0.514	p<0.0001
	Breathing VAS	ρ=0.390	p=0.008
	Raynaud's phenomenon VAS	ρ=0.654	p<0.001
	Finger ulcer VAS	ρ=0.415	p=0.005
	Overall disease severity VAS	ρ=0.640	p<0.0001
UCLA-GIT 2.0	Total Score	ρ=0.621	p<0.0001
ABILHAND-SSc	Measure	ρ=-0.649	p<0.0001

r – Pearson's coefficient; VAS – visual analogue scale; ρ – Spearman's coefficient