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Development of a new patient-reported outcome measure to assess activities and participation in people with systemic sclerosis: The Cochin 17-item Scleroderma Functional Scale.

Running head: Assessment of activities and participation in scleroderma.

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Bulleted statements.
What is already known about this subject?

- Patient-reported outcomes for people with systemic sclerosis rarely involved the target population in earliest stages of their construction.
- Instruments able to capture the specific needs of people with systemic sclerosis in terms of activities and participation are lacking.

What does this study add?

- The Cochin 17-item Scleroderma Functional Scale (CSF-17) is a new patient-reported outcome assessing global activities and participation specifically in people with systemic sclerosis.
- Its construction prioritized patients' perspectives at all stages.
- The CSF-17 could be used in clinical practice and research to assess the efficacy of complex multidisciplinary interventions targeting activity limitations and participation restriction in people with systemic sclerosis.

Abstract

Objectives. To develop a new patient-reported outcome measure assessing activities and participation in people with systemic sclerosis (SSc).

Methods. A provisional International Classification of Functioning, Disability and Health (ICF)-based 65-item questionnaire previously developed from interviews of people with SSc was sent by email to all patients followed in the internal medicine department of Cochin hospital (n=184) and enrolled in the Scleroderma Patient-centered Intervention Network Cohort. Items were reduced according to their metric properties. Dimensional structure of the questionnaire was assessed by principal component analysis, convergent and divergent validities by the Spearman correlation coefficient (ρ), internal consistency by the Cronbach α coefficient, and reliability by a test-retest method using intraclass correlation coefficient (ICC) and Bland and Altman analysis.

Results. Overall, 113/184 (61.4%) patients completed the provisional questionnaire. The item-reduction process resulted in a 17-item questionnaire, the Cochin 17-item Scleroderma Functional scale (CSF-17). Principal component analysis extracted 2 dimensions: 10 items related to mobility (CSF-17 section A) and 7 items related to general tasks (CSF-17 section B). We observed convergent validity of the CSF-17 total score with global activity limitation, pain, depression and aesthetic burden, and divergent validity with anxiety. The Cronbach α coefficient was 0.94 for section A and 0.95 for section B. ICC (n=25 patients) was 0.92 for CSF-17 total score. Bland and Altman analysis did not reveal a systematic trend for the test-retest.

Conclusions. The CSF-17 is a new patient-reported outcome assessing activities and participation specifically in people with SSc. Its content and construct validities are very good.

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Systemic sclerosis (SSc) is a rare autoimmune disease characterized by collagen deposition in skin and internal organs and vascular hyperreactivity. Two subsets of SSc are reported based on the extension of skin involvement and with different prognosis: limited cutaneous SSc (lcSSc) and diffuse cutaneous SSc (dcSSc) (1, 2). With improvement in patients' care, survival rates have increased. However, SSc remains a disabling condition and impairs patients' health-related quality of life (HRQoL) and functioning (3, 4), as defined by the WHO in the International Classification of Functioning, Disability and Health (ICF).

Guidelines regarding assessment of people with SSc have shifted from recommendations of quantifying organ damage to recommendations of measuring « functioning ». These recommendations rather refer to « physical functioning », than to functioning as defined by the WHO. Therefore, most of patient-reported outcome measures (PROM) (e.g. pain Numeric Rating Scale [NRS], Health Assessment Questionnaire [HAQ] (5) or Medical Outcome Study Short Form-12 items [MOS SF-12] (6-8)) (9) cover selected aspects of the whole individuals' experience of SSc and disregard others such as activities and participation. Some PROM have been designed to assess activities and participation in people with SSc (4, 10-12). However, the development of these instruments did not fully follow current guidelines (13-15). As reviewed by Pauling and colleagues, only 7/13 (53.8%) PROM aiming at assessing people with SSc involved the target population in the item and domain generation stage of the instrument construction (12, 16-23).

We aimed to develop a new PROM assessing activities and participation in people with SSc, using an original ICF-based patient-centered self-administered 65-item provisional questionnaire (24) and prioritizing people perspectives at all stages of the instrument construction.

Patients and methods

Study design overview. An original 65-item self-administered questionnaire was developed within the ICF conceptual framework and served as a provisional questionnaire (24). It was sent *via* email to participants enrolled in the Scleroderma Patient-centered Intervention Network (SPIN) Cohort (25), followed-up at the internal medicine department of Cochin hospital for item reduction and assessment of psychometric properties.

Provisional questionnaire. The development of the provisional ICF-based 65-item questionnaire had been reported (24). In a previous study (24), we developed a comprehensive ICF

core-set for SSc. Meaningful concepts were collected using data source triangulation from patients, experts and literature and linked to the best-matching ICF domains in accordance with prespecified standardized linking rules (26). To collect concepts from patients, we used a qualitative approach with focus group interviews of 18 patients followed-up in a French tertiary care center (Cochin Hospital, Paris, France), from October 2012 to June 2013. Inclusion criteria were: ≥18 years of age and a diagnosis of SSc according to the 2013 American College of Rheumatology (ACR) criteria and/or European League Against Rheumatism (EULAR) criteria **(Appendix 1)**. Patients generated 50 ICF categories belonging to the "activity and participation" domain. These categories were translated into understandable questions by a trained sociologist of the French ICF Research Branch, who double-checked with the reviewer who linked collected concepts to corresponding ICF categories, that ICF categories and derived questions were consistent. A total of 65 questions composed our provisional questionnaire **(Appendix 2)**. Participants in the present study were invited to report problems on the comprehensibility of the provisional questions in free text.

Participants. Participants were recruited from the SPIN Cohort (25). The SPIN Cohort is an online international cohort of patients with SSc that started enrollment in 2014. Eligibility criteria are: patients ≥18 years of age and be classified as having SSc according to the 2013 ACR criteria and/or EULAR criteria applied by a physician expert in SSc. For the present study, we included only French patients who were enrolled in SPIN through the internal medicine department of Cochin hospital, because our provisional questionnaire was developed in French. To improve the completion rates, one investigator contacted each patient by e-mail every 10 days. If the patient did not complete the questionnaire after 4 emails, she contacted the patient once by phone. If she failed to reach the patient or if the patient did not complete the questionnaire within 10 days after phone contact, the patient was considered a non-respondent **(Appendix 3)** (27).

Statistical analyses and sample size calculation. Statistical analyses were performed by using *ad hoc* routines implemented in R 3.3.1 software. Categorical data were summarized by numbers and percentages and continuous data by means and standard deviations. Quantitative variables were compared between two groups by the non-parametric Wilcoxon-Mann-Whitney test and qualitative variables by the Fisher's exact test. Correlations between two quantitative variables were evaluated using the non-parametric Spearman test. All tests were 2-sided. A p-

value <0.01 was considered as significant. In order to evaluate the structural validity of our questionnaire and its internal consistency and reproducibility, at least two times more patients than the total number of items was needed (28). Therefore, we sought to enroll 165 patients.

Psychometric properties of the questionnaire. Patients completed the provisional questionnaire and other PROM:

- Five activity limitation scales: 1/ McMaster Toronto Arthritis Patient Preference Disability Questionnaire (MACTAR) ranging from 0 to 30 (29, 30), 2/ HAQ ranging from 0 to 3 (5), 3/ scleroderma HAQ (sHAQ) ranging from 0 to 3 (4), 4/ Cochin Hand Function Scale (CHFS) ranging from 0 to 90 (10, 11), and 5/ Mouth Handicap in Systemic Sclerosis scale (MHISS) ranging from 0 to 48 (12), with higher scores indicating higher activity limitation,
- One HRQoL scale: MOS SF-12, with its two components, the physical component score (PCS) ranging from 9.95 to 70.02 and the mental component score (MCS) ranging from 5.89 to 71.97 (6-8), with higher scores indicating better HRQoL, and
- Three impairment scales: 1/ Hospital Anxiety Depression Scale (HADS) subscales for anxiety (HADSa) and depression (HADSd) ranging from 0 to 21 (31), 2/ NRS for pain ranging from 0 to 10, and 3/ NRS for aesthetic burden ranging from 0 to 10, with higher scores indicating higher impairment.

We limited the number of PROM in order to reduce the burden of the survey and to avoid a too time-consuming participation for patients. The Patient Acceptable Symptom State (PASS) was assessed using a specific anchoring question: "Taking into account all you have to do during your daily life, your level of pain and your functional impairment, do you consider that your current state is acceptable?", with a "yes" or "no" answer (32). The PASS was defined as the 75th percentile of the score of a PROM in the group of patients who answered "yes" to the anchoring question (33) and its 95% confidence interval (95% CI) was assessed by bootstrap resampling. Wilcoxon test was used to compare measurements for qualitative variables among groups.

Reduction of items. To avoid nonresponse bias and irrelevant items (34), we used patients' perspectives to reduce the number of items. We removed items with a response rate <90% (35). To avoid floor and ceiling effects, we removed items for which more than 50% of the respondents had an extreme value (0 or 10) (36). To avoid redundancy, and after consensus between investigators (9), we removed items with an inter-item Spearman correlation coefficient >0.80 (37). This step was the only one of the CSF-17 development that did not involve patients.

Dimensional structure of the questionnaire. To assess the uni- or multidimensional character of the new questionnaire, we carried out a principal component analysis. Factors with an eigenvalue >1 were retained (38, 39). We further performed an exploratory factor analysis to determine which items belonged to which dimension using the Psych R package (40).

External validity. We hypothesized that our questionnaire would be highly correlated with questionnaires assessing activity limitation (HAQ, sHAQ, MACTAR, CHFS and MHISS) or impairments associated with activity limitation (MOS SF-12 PCS and pain NRS) and weakly to moderately correlated with questionnaires assessing symptoms of anxiety and depression (HADSa, HADSd and MOS-SF12 MCS) and aesthetic burden (aesthetic burden NRS) (10, 12). The Spearman correlation coefficient (ρ) was calculated. Correlation was considered weak if $\rho<0.35$, moderate if $0.35\leq\rho<0.50$, and high if $\rho\geq0.50$ (41). We did not prespecify hypotheses concerning correlations with mean disease duration, because based on literature and our own experience, these are inconsistent.

Internal consistency. To verify that all items of the new questionnaire assessed the same concept, we calculated the Cronbach α coefficient (42). Its 95% CI was assessed by a bootstrap resampling. A Cronbach α coefficient was considered acceptable if >0.70 (43).

Test-retest reliability and agreement. An independent sample of 75 French patients with SSc of the SPIN Cohort, who were "non-respondents" in the first step or recently included, were invited to complete the new questionnaire twice with at least a 1-week interval. To assess reliability, we used ICC (44) and Bland and Altman analysis (45). ICC was considered acceptable if >0.7 (46). Even though we did not assess whether participants were stable in the interim period, given the chronic nature of SSc, we assumed it was unlikely that participants' health status and/or treatments would change within the interim period.

Ethical consideration. Our study protocol was approved by our Institutional Review Board (*CPP Île-de-France I*). Patients from the SPIN Cohort gave their written informed consent to participate in the online cohort study and received an electronic note to inform them about the specific additional research question addressed in the present study.

Patient and public involvement. Patients and public were not involved in the co-production of the present research.

Results

Participants. From February to March 2018, 113/184 (61.4%) patients completed at least 1 questionnaire: 109/113 (96.4%) the provisional questionnaire and 85/113 (75.2%) at least another questionnaire. Among respondents, 102/113 (90.3%) were women, 41/113 (37.3%) had dcSSc and 68/113 (61.8%) lcSSc. Mean age was 56.0 (14.6) years and mean disease duration 9.9 (7.4) years **(Table 1)**. Participants did not report any problems with the comprehensibility of the provisional questions.

Reduction of items. 24/65 (36.9%) items had a completion rate <90%, 27/65 (41.5%) a floor effect and 0/65 (0.0%) a ceiling effect **(Appendix 4)**, leaving 27/65 items in the questionnaire. Of these 27 items, 10/27 (37.0%) items were redundant with an inter-item Spearman correlation coefficient ≥0.80 and were removed **(Appendix 5)**.

Dimensional structure of the questionnaire. Exploratory analysis extracted 2 factors with eigenvalues of 10.6 and 1.9 explaining 59% and 11% of the variance, respectively **(Fig. 1)**. This result was confirmed by explanatory factor analysis **(Appendix 6)**.

Cochin 17-item Scleroderma Functional scale. The final questionnaire was named the Cochin 17-item Scleroderma Functional scale (CSF-17). It included 17 items distributed in 2 sections: section A (10 items, factor 1) assessing mobility, and section B (7 items, factor 2) assessing general tasks and demands. The French original version of the questionnaire is presented in **Appendix 7** and a provisional English version in **Table 2**. Full validation and transcultural adaptation are currently ongoing. Sections were named after the ICF domains to which their items mostly belonged**.** Because our scale assessed activities and limitations, we chose that the minimal score for an item (i.e. 0) would correspond to the absence of activity limitations and/or participation restriction and the maximal score (i.e. 10) to maximal activity limitations and/or participation restriction. The score of each section is the sum of the score for each item: section A score ranges from 0 to 100 and section B score ranges from 0 to 70. The total score for the CSF-17 was defined as the sum of the score for each section and ranges from 0 (no limitation) to 170 (maximal limitation).

External validity. External validity was calculated for each dimension. Sections A and B of the CSF-17 showed high correlation with the HAQ (ρ=0.84 and ρ=0.63), sHAQ (ρ=0.85 and

ρ=0.70), MACTAR (ρ=0.50 and ρ=0.54), CHFS (ρ=0.74 and ρ=0.58), MIHSS (ρ=0.67 and ρ=0.55), MOS-SF12 physical component score (ρ=-0.83 and ρ=-0.58) and pain NRS (ρ=0.66 and ρ=0.60), and weak to moderate correlation with the HADSa subscale (ρ=0.27 and ρ=0.49) and MOS-SF12 mental component score (ρ=-0.04 and ρ=-0.38), respectively. Sections A and B of the CSF-17 also showed high correlation with the HADSd subscale (ρ=0.50 and ρ=0.75) and aesthetic burden NRS (ρ=0.53 and ρ=0.54), respectively. Sections A and B of the CSF-17 showed moderate to high correlation with all 8 domains of the HAQ **(Table 3)**.

Internal consistency. Cronbach α coefficient was 0.94 (95% CI 0.92 to 0.96) for section A and 0.95 (95% CI 0.93 to 0.96) for section B. Correlation between each item and the CSF-17 score was good for all items and ranged from 0.72 to 0.87 for section A and from 0.76 to 0.88 for section B (**Appendix 8**).

Test-retest reliability. From May to June 2018, 34/75 (45.3%) patients completed the CSF-17, and 25 of these 34 (73.5%) patients also completed the retest (mean interval [SD]: 14.2 [6.8] days) **(Appendix 9)**. ICC (n=25) was 0.92 (95% CI 0.83 to 0.96) for the CSF-17 total score, 0.90 (95% CI 0.79 to 0.95) for section A and 0.94 (95% CI 0.85 to 0.97) for section B (**Appendix 10**). The Bland and Altman analysis showed no systematic trend. Mean difference was -4.2 (95% CI -34.1 to 25.7) for the CSF-17 total score, -2.6 (95% CI -22.8 to 17.1) for section A and -1.7 (95% CI -13.8 to 10.6) for section B (**Fig. 2**).

CSF-17 score and patient acceptable symptom state. In all patients, mean (SD) was 42.3 (38.4) for the CSF-17 total score, 25.5 (24.0) for section A and 16.8 (17.1) for section B. Section A score was lower in patients with lcSSc compared to dcSSc. Section B score did not differ between patients with lcSSc and dcSSc (**Table 4**). There were no significant correlations between mean disease duration and CSF-17 section A, section B and total score (ρ=0.05, ρ=0.10 and ρ=0.08, respectively). Overall, 40/106 (37.7%) patients considered their state as acceptable according to the anchor. PASS (95% CI) was 31.0 (18.5-37.0) for the CSF-17 total score, 21.3 (14.3-24.0) for section A and 9.3 (5.0-12.0) for section B (**Table 4**).

Discussion

The CSF-17 is a new self-administered questionnaire assessing activities and participation in people with SSc. The CSF-17 has 2 dimensions. The first dimension includes 10 items related to mobility: 5/10 items related to upper limb mobility (1, 4, 5, 6, 10), 3/10 to general mobility (2, 3, 9) and 2/10 to lower limb mobility (7, 8). The second dimension includes 7 items related to cognitive functions and complex tasks, which we assume were related to the chronic aspects of the disease. While literature and experts usually focus on hand- and mouth-specific activity limitation, general and lower limb mobilities were also limited in patients with SSc. This result is consistent with our previous finding that general mobility and walking were the most frequent patient-perceived activity limitation and reported in 34.3% (29) and 54.6% (30) of people with SSc, respectively.

We confirmed our hypotheses regarding external validity of the CSF-17, except for a convergent correlation with symptoms of depression and aesthetic burden. The convergent correlation with symptoms of depression might be explained by items related to psychological demands and socio-professional interactions of the CSF-17. Mobility and aesthetic burden might be correlated with depression (47). Therefore, the correlation between aesthetic burden and mobility could be biased. Because the ICF conceptual framework does not contain items related to aesthetic burden, this might explain its resurgence through other dimensions. Even though we confirmed the convergent correlation of the CSF-17 and the MACTAR, it was the lowest correlation coefficient of all instruments for both dimensions of the CSF-17. This result is consistent with our previous finding that the MACTAR weakly correlates with other PROM and add non-redundant and relevant information to other scores (29, 30).

Overall reliability of the CSF-17 was promising: Cronbach α coefficients were excellent indicating that all items within each dimension referred to the same concept and ICCs were very good ≥ 0.75. Further, the Bland and Altman analysis did not reveal a systematic trend for the test-retest suggesting that the CSF-17 is a repeatable measure.

The CSF-17 total score was higher in patients with dcSSc compared to patients with lcSSc, suggesting the ability of the CSF-17 to capture higher levels of global activity limitation in patients with a more severe disease. Because evaluating the clinical significance of a PROM can be challenging (9, 48), we also estimated the PASS thresholds of the CSF-17 total, section A and B scores. Our estimates could be useful in interpreting the clinical relevance of the CSF-17 in clinical practice and trials.

Our study has limitations. We enrolled French participants only and our sample size was

small. However, there is only little guidance on sample size calculations for questionnaire development (49). Some patients found our internet survey burdensome and invasive. Among those who provided an explanation for not participating in the survey: six expressed tiredness of online surveys, two could not use a computer, two preferred to answer to the survey by phone, one during a face-to-face meeting and one refused to answer because "questionnaires never reflect what she experiences in daily life". This highlights limitations of internet-based surveys including selection bias of individuals sufficiently motivated and computer-skilled and a partial transfer of the research burden to participants (50, 51). All participants enrolled in the SPIN Cohort were followed-up in tertiary care centers and might not be representative of the French population with SSc. Comparing antinuclear antibody-based disease subsets for the CSF-17 score could have added relevant information. However, we did not collect antinuclear antibody status. We did not assess whether participants were able to distinguish between 11 response options. We did not have indications they were not, and did not find a systematic trend of response pattern for any of the 17 questions and 2 domains **(Appendix 11)**. Finally, we collected data during the winter of 2018, which could have led to an overestimation of the CSF-17 score.

In sum, the CSF-17 is a new self-administered questionnaire designed to assess SSc-specific global activity limitation and participation restriction. Its content validity and psychometrics properties are very good. We have planned to use the international SPIN cohort as a platform for additional external validation.

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Figure legends.

Figure 1. Principal component analysis of the Cochin 17-item Scleroderma Functional Scale (CSF-17).

Figure 2. Bland-Altman plots (n=25); (A) Cochin 17-item Scleroderma Functional Scale (CSF-17) total score; (B) CSF-17 section A score; (C) CSF-17 section B score.

Table 1. Demographical and clinical data of respondents.

	All n=113	dcSSc n=41 (36.3%)	lcSSc n=68 (60.2%)	Unspecified subset n=4 (3.5%)	p-value*
Women, n/N (%)	102/113 (90.3)	35/41 (85.4)	63/68 (92.6)	4/4 (100.0)	-
Age (years), Age (years), mean (SD)	56.0 (14.6)	53.4 (12.5)	57.6 (15.7)	56.8 (16.2)	-
Duration of the disease (years), mean (SD)	9.9 (7.4)	9.3 (6.3)	10.3 (8.0)	6.5 (3.1)	-
Body mass index (kg/m²), mean (SD)	23.2 (4.8)[a]	23.1 (5.2)	23.2 (4.7)	NA	-
Sclerodactyly, n/N (%)	84/109 (77.1)	37/41 (90.2)	47/67 (70.1)	3/4 (75.0)	-
Digital ulcer, n/N (%)	45/110 (40.9)	22/41 (53.7)	23/68 (33.8)	0/4 (0.0)	-
Telangectasias, n/N (%)	69/109 (63.3)	20/40 (50.0)	48/68 (70.6)	3/4 (75.0)	-
Stiffness of small joints (finger, wrist), n/N (%)	35/107 (32.7)	18/40 (45.0)	16/66 (24.2)	1/4 (25.0)	-
Stiffness of large joints (elbow, hip, knee, ankle), n/N (%)	22/106 (20.8)	13/40 (32.5)	9/65 (13.8)	0/4 (0.0)	-
Gastrointestinal tract distal involvement, n/N (%)	33/109 (30.3)	9/41 (22.0)	24/67 (35.8)	0/4 (0.0)	-
Pulmonary fibrosis, n/N (%)	47/110 (42.7)	25/41 (61.0)	22/68 (32.4)	0/4 (0.0)	-
Pulmonary arterial hypertension, n/N (%)	7/110 (6.4)	4/41 (9.8)	3/68 (4.4)	0/4 (0.0)	-

Scleroderma renal crisis, n/N (%)	6/110 (5.5)	4/41 (9.8)	2/68 (2.9)	0/4 (0.0)	-
HAQ (0-3) mean (SD)	1.1 (0.8)[b]	1.4 (0.8)[i]	0.8 (0.6)[n]	1.8 (0.4)[v]	0.001
sHAQ (0-3) mean (SD)	1.00 (0.7)[b]	1.1 (0.8)[i]	0.9 (0.6)[n]	2.0 (0.5)[v]	0.135
MACTAR (0-30) mean (SD)	16.8 (8.9)[c]	19.3 (7.2)[j]	15.7 (9.5)[o]	7.0 (-)[w]	0.148
CHFS (0-90) mean (SD)	18.0 (18.7)[d]	26.5 (10.2)[i]	12.0 (13.8)[p]	46.0 (43.8)[v]	0.001
MHISS (0-48) mean (SD)	19.0 (12.5)[e]	24.7 (12.7)[k]	15.2 (11.1)[q]	21.5 (10.6)[v]	0.004
MOS SF 12 PCS (9.95-70.02) mean (SD)	37.3 (10.5)[f]	33.2 (9.5)[l]	40.7 (9.9)[r]	23.0 (2.3)[v]	0.004
MOS SF 12 MCS (5.89-71.97) mean (SD)	43.3 (12.0)[f]	44.6 (12.1)[l]	42.4 (12.0)[r]	43.2 (18.4)[v]	0.574
HADS anxiety subscale (0-21) mean (SD)	7.9 (4.4)[g]	7.8 (4.7)[l]	8.0 (4.3)[s]	9.5 (2.1)[v]	0.903
HADS depression subscale (0-21) mean (SD)	6.2 (4.4)[g]	7.4 (4.9)[l]	5.4 (3.9)[s]	7.5 (5.0)[v]	0.032
Pain NRS (0-10) mean (SD)	4.5 (3.0)[h]	5.0 (2.6)[m]	4.1 (3.2)[t]	8.5 (2.1)[v]	0.493
Aesthetic burden NRS (0-10) mean (SD)	4.2 (3.3)[e]	5.0 (3.2)[m]	3.7 (3.1)	5.0 (7.1)[v]	0.393

CHFS: Cochin Hand Function Scale; dcSSc: diffuse cutaneous systemic sclerosis; HADS: Hospital Anxiety Depression Scale; HAQ: Health Assessment Questionnaire; lcSSc: limited cutaneous SSc; MACTAR: McMaster Toronto Arthritis patient preference questionnaire; MHISS: Mouth Handicap In Systemic Sclerosis; MOS SF-12 PCS: 12-Item Short Form Health Survey Physical Component Score; MOS SF-12 MCS: 12-Item Short Form Health Survey Mental Component Score; NA : not available; NRS: Numeric Rating Scale; sHAQ: scleroderma Health Assessment Questionnaire.

*Comparisons between dcSSc and lcSSc groups at baseline were not prespecified but were performed for self-administered questionnaires upon reviewer's request using t-test. A p-value < 0.01 was considered significant.

[a]n=110, [b]n=79, [c]n=65, [d]n=89, [e]n=80, [f]n=77, [g]n=71, [h]n=82, [i]n=33, [j]n=28, [k]n=31, [l]n=30, [m]n=32, [n]n=44, [o]n=38, [p]n=54, [q]n=47, [r]n=45

[s]n=40, [t]n=48, [u]n=46, [v]n=2, [w]n=1.

Table 2. Provisional English translation of the Cochin 17-item Scleroderma Functional scale (CSF-17).

Because of my systemic sclerosis, I feel limited in the following daily activities:	
Section A. Mobility	
1	Writing with a pen or a pencil
2	Changing my body position
3	Standing up alone
4	Lifting and carrying objects in my hands even when moving
5	Manipulating small objects using my fingers and hands
6	Moving arms (raise, flex, extend)
7	Walking
8	Running
9	Using public transportation (bus, metro, tramway)
10	Thinkering, gardening, feeding and taking care of my domestic animals
Section B. General tasks and demands	
11	Learning new things
12	Focusing my attention
13	Solving problems of daily life
14	Undertaking a complex task requiring several steps
15	Managing my own activity level
16	Handling stress and other psychological demands
17	Handling responsibilities in my personal and professional life

NOTE. The items were originally formulated in French.

Table 3. CSF-17 convergent and divergent validities.

	Section A		Section B		n=
	ρ	p-value	ρ	p-value	
Convergent validity: | ρ | > 0.50					
sHAQ	0.85	< 0.0001	0.70	< 0.0001	76
HAQ	0.84	< 0.0001	0.63	< 0.0001	76
• **HAQ Domain 1**	0.59	< 0.0001	0.48	< 0.0001	76
• **HAQ Domain 2**	0.59	< 0.0001	0.46	< 0.0001	76
• **HAQ Domain 3**	0.60	< 0.0001	0.52	< 0.0001	76
• **HAQ Domain 4**	0.71	< 0.0001	0.48	< 0.0001	76
• **HAQ Domain 5**	0.64	< 0.0001	0.39	< 0.001	76
• **HAQ Domain 6**	0.80	< 0.0001	0.60	< 0.0001	76
• **HAQ Domain 7**	0.41	< 0.001	0.42	< 0.001	76
• **HAQ Domain 8**	0.82	< 0.0001	0.54	< 0.0001	76
MACTAR	0.50	< 0.0001	0.54	< 0.0001	63
CHFS	0.74	< 0.0001	0.58	< 0.0001	70
MHISS	0.67	< 0.0001	0.55	< 0.0001	70
MOS SF-12 PCS	-0.83	< 0.0001	-0.58	< 0.0001	64
Pain NRS	0.66	< 0.0001	0.60	< 0.0001	75
HADS depression subscale	0.50*	< 0.0001	0.75*	< 0.0001	62
Aesthetic burden NRS	0.53*	< 0.0001	0.54*	< 0.0001	74
Divergent validity: | ρ | < 0.50					
HADS anxiety subscale	0.27	< 0.05	0.49	< 0.0001	64
Disease duration	0.21	0.04	0.18	0.09	94
MOS SF-12 MCS	-0.04	0.77	-0.38	< 0.01	64

CHFS: Cochin Hand Function Scale; HADS: Hospital Anxiety Depression Scale; HAQ: Health Assessment Questionnaire; MACTAR: McMaster Toronto Arthritis patient preference questionnaire; MHISS: Mouth Handicap In Systemic Sclerosis; MOS SF-12 PCS: 12-Item Short Form Health Survey Physical Component Score; Study MOS SF-12 MCS: 12-Item Short Form Health Survey Mental Component Score; NRS: Numeric Rating Scale; sHAQ: scleroderma Health Assessment Questionnaire.
*unexpected correlation.

Table 4. CSF-17 scores and patient acceptable symptom state estimates.

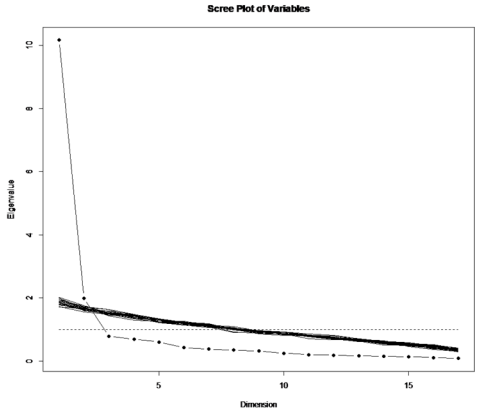
	CSF-17 scores			
	All	**dcSSc**	**lcSSc**	***p-value****
	n=106	**n=41**	**n=65**	
CSF-17 total, mean (SD)	47.3 (38.5)	60.3 (39.7)[a]	37.9 (34.9)[b]	< 0.01
CSF-17 section A, mean (SD)	29.8 (24.9)	39.3 (25.8)[d]	23.1 (22.1)[e]	< 0.01
CSF-17 section B, mean (SD)	17.9 (17.3)	21.8 (18.7)[f]	15.3 (15.8)[g]	< 0.10
	CSF-17 patient acceptable symptom state estimates			
	All	**dcSSc**	**lcSSc**	
	n=40**	**n=13**	**n=26**	
PASS total, 75th percentile (IC 95%)	31.0 (18.5-37.0)	24 (20.0-62.0)	31.8 (14.0-45.0)	
PASS section A, 75th percentile (IC 95%)	21.3 (14.3-24.0)	22 (14.9-29.0)	20.3 (10.0-34.0)	
PASS section B, 75th percentile (IC 95%)	9.3 (5.0-12.0)	11 (3.0-30.0)	9 (4.3-12.0)	

CSF-17: Cochin 17-item Scleroderma Functional scale; dcSSc: diffuse cutaneous systemic sclerosis; lcSSc: limited cutaneous SSc; PASS: Patient Acceptable Symptom State.

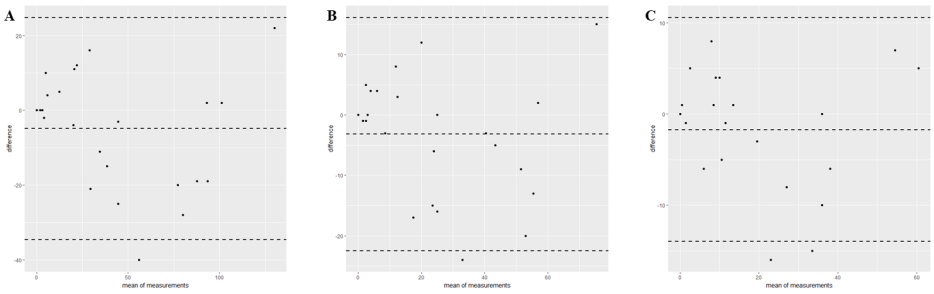
[a]n=37; [b]n= 51; [d]n=37; [e]n=53; [f]n=38; [g]n=56

*Non-parametric Wilcoxon-Mann-Whitney Test.

**Including 1 patient with no information about the disease subset.



bjd_18922_f1.tif



bjd_18922_f2.tif

Appendix 1. Demographical and clinical data of patients included in the focus groups.

	All patients n=18
Women, n/N (%)	10/18 (55.6)
Age (years), mean (SD)	56.5 (16.2)[a]
Disease subset, n/N (%)	
Diffuse cutaneous	8/16 (50.0)
Limited cutaneous	8/16 (50.0)
Disease duration (years), mean (SD)	10.8 (9.9)[b]
Disease considered severe by the treating physician, n/N (%)	11/16 (68.8)
Scleroderma renal crisis	0/16 (0.0)
Pulmonary arterial hypertension*	4/16 (25.0)
Pulmonary fibrosis	9/16 (56.3)
Cutaneous involvement extended to the trunk	2/16 (12.5)
Gastrointestinal tract involvement	6/16 (37.5)
Modified Rodnan skin score (0-51), mean (SD)	14.3 (9.6)[c]
Microstomia (mouth opening < 40 mm), n/N (%)	6/12 (50.0)
Mouth opening (mm), mean (SD)	36.8 (7.1)[d]
Sclerodactylia, n/N (%)	12/12 (100.0)

*confirmed by right heart catheterization; [a]n=16; [b]n=17; [c]n=10; [d]n=13.

Appendix 2. ICF-based 65-item self-administered provisional questionnaire.

Learning and applying knowledge
<u>Because of my systemic sclerosis</u>, I feel limited in the following daily activities:
Q1. Learning new things?
Q2. Focusing my attention?
Q3. Reading?
Q4. Writing with a pen or a pencil?
General tasks and demands
<u>Because of my systemic sclerosis</u>, I feel limited in the following daily activities:
Q5. Solving problems of daily life?
Q6. Making decisions?
Q7. Undertaking a complex task requiring several steps?
Q8. Undertaking multiple simultaneous or successive tasks?
Q9. Carrying out daily routine (planning, carrying out, undertaking tasks and demands of daily life)?
Q10. Managing my own activity level?
Q11. Handling stress and other psychological demands?
Q12. Handling responsibilities in my personal and professional life?
Mobility
<u>Because of my systemic sclerosis</u>, I feel limited in the following daily activities:
Q13. Changing my body position?
Q14. Kneeling down, squatting alone?
Q15. Standing up alone?
Q16. Bending forward?
Q17. Maintaining a lying position?
Q18. Maintaining a standing position?
Q19. Lifting and carrying objects in my hands even when moving?
Q20. Manipulating small objects using my fingers and hands?
Q21. Moving arms (raise, flex, extend)?
Q22. Pulling or pushing an object?
Q23. Walking?
Q24. Moving downwards (a step, a slope, a ladder)?
Q25. Moving upwards or downwards (a step, a stool, a slope, a ladder)?

Q26. Running?

Q27. Going somewhere (inside and outside the home)?

Q28. Taking a plane or a train?

Q29. Using public transportation (bus, metro, tramway)?

Q30. Driving (a car, a motocycle, a bicycle)?

Self-care

Because of my systemic sclerosis, I feel limited in the following daily activities:

Q31. Washing myself?

Q32. Caring for my physical appearance (combing, shaving, removing hair, brushing teeth, caring for skin, hands, feet, making up, choosing my clothes)?

Q33. Toileting?

Q34. Putting on clothes, taking off clothes, putting on footwear, taking off footwear?

Q35. Eating?

Q36. Looking after my health?

Domestic life

Because of my systemic sclerosis, I feel limited in the following daily activities:

Q37. Shopping?

Q38. Preparing meals?

Q39. Doing housework (washing dishes, washing clothes, housekeeping, ironing, cleaning)?

Q40. Thinkering, gardening, feeding and taking care of my domestic animals?

Q41. Assisting others (familly members, neighbours, relatives) according to their needs?

Communication

Because of my systemic sclerosis, I feel limited in the following daily activities:

Q42. Expressing myself and making myself understood in oral language?

Q43. Starting a conversation or conversing with one person or many people?

Q44. Using a landline or a mobile phone?

Q45. Using a computer (reading the computer screen and/or writing using a keyboard)?

Interpersonal interactions and relationships

Because of my systemic sclerosis, I feel limited in the following daily activities:

Q46. Interacting with someone in a contextually and socially appropriate manner?

Q47. Accepting bodily contact (allowing physical contact, hugging)?

Q48. Forming and terminating relationships?

Q49. Regulating emotions, verbal aggression and physical aggression in interactions with others?

Q50. Engaging in contacts with strangers for specific purposes (asking for directions, making a purchase, reporting a

problem)?

Q51. Having and maintaining relationships with friends?

Q52. Having and maintaining relationships with the members of my family?

Q53. Creating and maintaining close or romantic relationships with someone?

Q54. Having a satisfying sexual life?

Major life areas

Because of my systemic sclerosis, I feel restricted in participating in the following daily activities:

Q55. Taking an exam?

Q56. Engaging in an educational program (being present, being diligent)?

Q57. To seek, to change, to find or to keep a job?

Q58. Doing all the required tasks and activities of my job?

Q59. Working full-time?

Community, social and civic life

Because of my systemic sclerosis, I feel restricted in participating in the following daily activities:

Q60. Travelling in France or overseas?

Q61. To do sport?

Q62. Going to cultural events (shows, museums, exhibitions)?

Q63. Doing handicrafts (sewing, collections, craftwork)?

Q64. Having and developing my spiritual life?

Q65. Participating in local and political life as a citizen (vote, local debate, unionism)?

NOTE. The items were originally in the French language.

Appendix 3. Checklist for Reporting Results of Internet E-surveys (CHERRIES).

Item Category	Checklist Item	Explanation
Design	Describe survey design	It was a closed-survey. The target population and sample frame were French patients of the e-cohort SPIN, described in Appendix 3, **see p. 38**
Institutional Review Board (IRB) approval and informed consent process	IRB approval	Approved by our IRB (*Comité de Protection des Personnes Île-de-France I*), **see p.12**
	Informed consent	Patients from the SPIN e-cohort gave their written informed consent to participate in the e-cohort. Along with the link to the online questionnaires, patients received an electronic note to inform them about the specific research question addressed in the present study, **see p.12**
	Data protection	For the provisional questionnaire, no personal information was collected in addition to the questionnaires answers, and answers were stored on a secured server at the clinical research unit. For the test-retest, name, gender and answer to the questionnaire were collected and stored on a secured Google drive.
Development and pre-testing	Development and testing	Online version of the provisional questionnaire was developed and its functionality was tested in July 2017. The online version of the final questionnaire for the test-retest was developed and tested in May 2018.
Recruitment process and description of the sample having access to the questionnaire	Open Survey vs closed survey	Closed survey, a secured link and personalized password were send to each participant.
	Contact mode	Initial contact was made by mail.
	Advertising the survey	No advertising was made.
Survey administration	Web/E-mail	The questionnaires were stored on a website, with automatic method for capturing responses in the database.
	Context	The survey was on the website of the clinical research unit of Paris Descartes: http://www.rechercheclinique pariscentre.fr. Only participants contacted by mail received the link to the secured online platform.
	Mandatory/voluntary	It was a voluntary survey.
	Incentives	No incentives were used.
	Time/Date	For the provisional questionnaire : from February 6, 2018 to March 31, 2018, **see p.13** For the test-retest : from May 16, 2018 to June 20, 2018, **see p.22**
	Randomization of items or questionnaires	No randomization planned or needed for the purpose of this study.
	Adaptative questioning	No randomization planned or needed for the purpose of this study.
	Number of items	For the provisional questionnaire: 155 items, distributed as follows: - Provisional scale: 65 items - HAQ : 20 items - sHAQ: 5 items - MACTAR: 7 items - CHFS: 18 items - MHISS: 12 items - HADS: 14 items - SF-12: 12 items - NRS pain: 1 item - NRS aesthetic burden: 1 item

		For the test-retest : 17 items
	Number of screens (pages)	For the provisional questionnaire : 26 pages For the test-retest : 1 page
	Completeness check	For the provisional questionnaire, completeness was checked after the questionnaire has been submitted. No item was mandatory. For the test-retest, completeness was checked before the questionnaire has been submitted, all the items were mandatory.
	Review step	Patients were able to change their answers with a review step, and they had the possibility to have several accesses to complete or modify their answers.
Response rates	Unique site visitor	Each participant had a unique identifying code and a personalized access-link and password. Patients' answers were saved under their identifying codes.
	View rate (ratio unique site visitors/unique survey visitors)	N/A; only patients of the survey could access to the internet platform.
	Participation rate (ratio unique survey page visitors/agreed to participate)	For the provisional questionnaires: 113/184 invited participants answered (61.4% of answer rate), **see p.13** For the test-retest: 34/75 (45.3%) and 24/34 (70.6%) invited participants answered
	Completion rate (ratio agreed to participate/finished survey)	For the provisional questionnaire: 109/113 (96.4%) completed the provisional questionnaire (subject to item ratio of 1.7) and 85/113 (75.2%) at least another questionnaire in addition to the provisional questionnaire. For the test-retest, the completion rate was 100% (34/34 for the test and 24/24 for the retest).
Preventing multiple entries from the same individual	Cookies used	No cookies were used.
	IP check	IP addresses were not checked.
	Log file analysis	N/A
	Registration	Patient's answers were registered under his identifying code, given by the personalized access-link and password. Multiples entries were allowed, patients could change their answers and the last answer was kept for analysis.
Analysis	Handling of incomplete questionnaires	For the provisional scale an item without answer was assumed irrelevant. For the other scales, in case of a missing item, the questionnaire was not examined, excepted for the SF-12 and MACTAR where imputations performed. No incomplete questionnaire was allowed for the test-retest.
	Questionnaires submitted with atypical timestamp	The time needed to fill in a questionnaire was not used to exclude questionnaires.
	Statistical correction	N/A

Appendix 4. Characteristics of the ICF-based 65-item self-administered questionnaire (109 patients with complete dataset).

Items	Missing answer	Floor effect	Ceiling effect	50 percentiles	Decision rule
1	6.4	45.9	0.9	1.00	yes
2	6.4	43.1	0	1.00	yes
3	6.4	57.8*	0.9	0.00*	no
4	6.4	36.7	0.0	2.00	yes
5	6.4	36.7	0.9	2.00	yes
6	6.4	49.5	1.8	0.00*	no
7	6.4	38.5	1.8	2.00	yes
8	7.3	37.6	0.9	2.00	yes
9	7.3	38.5	0.9	2.00	yes
10	6.4	25.7	0.9	3.00	yes
11	6.4	25.7	1.8	3.00	yes
12	8.3	40.4	1.8	1.00	yes
13	4.6	44	0.9	1.00	yes
14	4.6	30.3	4.6	2.00	yes
15	4.6	30.3	0.9	2.00	yes
16	4.6	42.2	0.9	1.00	yes
17	4.6	59.6*	0.0	0.00*	no
18	5.5	37.6	0.9	1.00	yes
19	5.5	22.9	1.8	3.00	yes
20	4.6	18.3	1.8	4.00	yes
21	4.6	43.1	0.9	1.00	yes
22	4.6	25.7	0.9	2.50	yes
23	4.6	41.3	0.9	1.00	yes
24	6.4	39.4	0.9	1.00	yes
25	6.4	39.4	0.9	2.00	yes
26	5.5	22	16.5	4.00	yes
27	6.4	51.4*	0.9	0.00*	no
28	8.3	48.6*	0.0	0.00*	no
29	6.4	45	1.8	1.00	yes
30	5.5	56*	3.7	0.00*	no
31	8.3	55*	0.0	0.00*	no
32	8.3	46.8	0.0	0.00*	no
33	7.3	60.6*	0.9	0.00*	no
34	8.3	47.7	0.0	0.00*	no
35	7.3	58.7*	0.9	0.00*	no
36	8.3	52.3*	0.0	0.00*	no

37	9.2	36.7	2.8	1.00	yes
38	10.1*	39.4	1.8	1.00	no
39	11*	29.4	1.8	2.00	no
40	9.2	30.3	1.8	2.00	yes
41	9.2	36.7	3.7	2.00	yes
42	10.1*	56*	0.0	0.00*	no
43	9.2	56.9*	0.0	0.00*	no
44	9.2	60.6*	0.0	0.00*	no
45	11*	58.7*	0.0	0.00*	no
46	12.8*	57.8*	0.0	0.00*	no
47	12.8*	56.9*	1.8	0.00*	no
48	11.9*	42.2	0.9	1.00	no
49	13.8*	60.6*	0.9	0.00*	no
50	11.9*	57.8*	0.9	0.00*	no
51	12.8*	56*	0.0	0.00*	no
52	11.9*	48.6	3.7	0.00*	no
53	12.8*	47.7	1.8	0.00*	no
54	12.8*	37.6	10.1	2.00	no
55	19.3*	41.3	8.3	0.00*	no
56	17.4*	46.8	8.3	0.00*	no
57	18.3*	45	11.9	0.00*	no
58	21.1*	33	10.1	1.00	no
59	20.2*	30.3	16.5	3.00	no
60	11.9*	44	4.6	0.50	no
61	11.9*	18.3	11.0	4.00	no
62	11.9*	42.2	3.7	1.00	no
63	11.9*	23.9	8.3	3.50	no
64	11.9*	57.8*	1.8	0.00*	no
65	11.9*	54.1*	4.6	0.00*	no

*reason(s) why the item was rule-out.

Appendix 5. Redundancy between the 27 remaining items (84 patients with complete dataset) (**indicates items with redundancy***).**

	1	**2**	**4**	**5**	**7**	**8**	**9**	**10**	**11**	**12**	**13**	**14**	**15**	**16**	**18**	**19**	**20**	**21**	**22**	**23**	**24**	**25**	**26**	**29**	**37**	**40**	**41**
1	1	0.78	0.45	0.72	0.68	0.65	0.66	0.65	0.68	0.64	0.48	0.42	0.41	0.47	0.41	0.43	0.47	0.51	0.47	0.39	0.44	0.33	0.37	0.40	0.49	0.35	0.52
2		1	0.42	0.67	0.62	0.56	0.58	0.62	0.66	0.58	0.40	0.31	0.20	0.35	0.47	0.37	0.40	0.33	0.43	0.41	0.37	0.36	0.30	0.38	0.47	0.39	0.45
4			1	0.47	0.51	0.55	0.52	0.53	0.38	0.29	0.57	0.56	0.54	0.61	0.60	0.68	0.68	0.57	0.65	0.54	0.51	0.54	0.52	0.47	0.64	0.57	0.53
5				1	0.77	0.74	0.81*	0.76	0.65	0.66	0.56	0.47	0.42	0.52	0.53	0.54	0.50	0.51	0.56	0.55	0.55	0.49	0.45	0.55	0.62	0.60	0.68
7					1	0.90*	0.90*	0.76	0.74	0.75	0.65	0.47	0.49	0.57	0.52	0.57	0.55	0.51	0.59	0.57	0.58	0.54	0.47	0.59	0.65	0.60	0.70
8						1	0.89*	0.76	0.68	0.66	0.64	0.45	0.53	0.55	0.56	0.61	0.53	0.47	0.58	0.64	0.59	0.56	0.51	0.61	0.68	0.61	0.73
9							1	0.78	0.71	0.69	0.61	0.48	0.52	0.58	0.57	0.61	0.56	0.55	0.60	0.61	0.62	0.56	0.49	0.60	0.64	0.59	0.69
10								1	0.68	0.65	0.54	0.43	0.36	0.47	0.46	0.60	0.57	0.47	0.63	0.53	0.53	0.53	0.56	0.51	0.64	0.64	0.69
11									1	0.73	0.57	0.29	0.34	0.40	0.42	0.47	0.39	0.38	0.50	0.37	0.44	0.37	0.36	0.41	0.52	0.49	0.56
12										1	0.48	0.34	0.32	0.46	0.44	0.49	0.39	0.42	0.49	0.42	0.41	0.40	0.31	0.45	0.59	0.51	0.58
13											1	0.61	0.66	0.71	0.55	0.56	0.56	0.57	0.62	0.55	0.67	0.54	0.54	0.58	0.61	0.59	0.64
14												1	0.81*	0.76	0.63	0.69	0.66	0.74	0.68	0.58	0.65	0.64	0.72	0.59	0.59	0.55	0.52
15													1	0.80*	0.62	0.72	0.62	0.63	0.65	0.68	0.76	0.66	0.66	0.67	0.56	0.57	0.61
16														1	0.67	0.65	0.65	0.73	0.65	0.63	0.69	0.60	0.56	0.58	0.58	0.57	0.54
18															1	0.76	0.57	0.62	0.69	0.82*	0.70	0.71	0.55	0.67	0.63	0.62	0.60
19																1	0.71	0.62	0.87*	0.70	0.72	0.77	0.70	0.68	0.80*	0.75	0.71
20																	1	0.65	0.76	0.57	0.62	0.59	0.57	0.52	0.67	0.63	0.58
21																		1	0.70	0.61	0.62	0.59	0.59	0.52	0.56	0.46	0.48
22																			1	0.67	0.72	0.76	0.71	0.65	0.78	0.72	0.72
23																				1	0.85*	0.84*	0.67	0.79	0.62	0.61	0.66
24																					1	0.88*	0.70	0.76	0.65	0.60	0.70
25																						1	0.73	0.80*	0.68	0.62	0.68
26																							1	0.73	0.63	0.65	0.68
29																								1	0.67	0.71	0.82*
37																									1	0.74	0.76
40																										1	0.81*
41																											1

Appendix 6. Factor analysis.

Items	Factor 1 (Section A)	Factor 2 (Section B)
1	0.30	0.77
2	0.17	0.77
4	0.63	0.39
5	0.47	0.72
7	0.43	0.82
10	0.42	0.75
11	0.26	0.79
12	0.24	0.79
13	0.60	0.54
15	0.83	0.15
19	0.84	0.30
20	0.70	0.33
21	0.65	0.36
23	0.78	0.31
26	0.74	0.20
29	0.77	0.23
40	0.74	0.36

	Factor 1 (Section A)	Factor 2 (Section B)
SS loadings	6.21	5.32
Proportion of variance explain by each factor	0.37	0.31
Root mean square of the residuals	0.05	
Tucker Lewis Index of factoring reliability	0.87	
Root mean square error of approximation (95% CI)	0.123 (0.10 to 0.14)	

Appendix 7. Original French version of the Cochin 17-item Scleroderma Functional scale (CSF-17).

À cause de ma sclérodermie systémique, je suis gêné(e) pour réaliser les activités quotidiennes suivantes :	
Partie A. Mobilité	
1	Écrire à l'aide d'un stylo ou d'un crayon
2	Changer la position de mon corps
3	Me relever, me mettre debout seul(e)
4	Soulever et porter des objets ou des charges y compris en me déplaçant
5	Manipuler de petits objets ou des objets fins
6	Bouger les bras (lever, plier, tendre)
7	Marcher
8	Courir
9	Utiliser les transports en commun (bus, métro, tramway)
10	Bricoler, jardiner, nourrir et prendre soin de mes animaux de compagnie
Partie B. Tâches et demandes générales	
11	Apprendre de nouvelles choses
12	Fixer mon attention
13	Résoudre les problèmes du quotidien
14	Entreprendre une tâche complexe nécessitant plusieurs étapes
15	Adapter mes activités quotidiennes à mon niveau d'énergie
16	Gérer la pression et le stress
17	Assumer mes responsabilités dans ma vie personnelle et professionnelle

Appendix 8. Internal consistency.

Items		Correlation
	Section A, Cronbach α = 0.94 IC95% [0.92-0.96]	**ρ**
1	Writing with a pen or a pencil	0.72
2	Changing my body position	0.73
3	Standing up alone	0.78
4	Lifting and carrying objects in my hands even when moving	0.87
5	Manipulating small objects using my fingers and hands	0.77
6	Moving arms (raise, flex, extend)	0.73
7	Walking	0.81
8	Running	0.73
9	Using public transportation (bus, metro, tramway)	0.79
10	Thinkering, gardening, feeding and taking care of my domestic animals	0.80
	Section B, Cronbach α = 0.95 IC95% [0.93-0.96]	**ρ**
11	Learning new things	0.82
12	Focusing my attention	0.76
13	Solving problems of daily life	0.81
14	Undertaking a complex task requiring several steps	0.88
15	Managing my own activity level	0.83
16	Handling stress and other psychological demands	0.79
17	Handling responsibilities in my personal and professional life	0.79

Correlation of each item with its dimension score (analysis with the item removed from of the score).

ρ=correlation within each item and its subpart score, calculated without the item.

Appendix 9. Demographical and clinical data of respondents for the test-retest.

	All **n=25**
Interval between test and retest (days), mean (SD)	14.2 (6.8)
Women, n/N (%)	18/25 (72.0)
Age (years), mean (SD)	52.5 (12.8)[a]
lcSSc, n/N (%)	18/22 (81.8)
dcSSc, n/N (%)	7/22 (31.8)
Duration of the disease (years), mean (SD)	9.7 (5.6)[a]
Body mass index (kg/m²), mean (SD)	23.2 (4.1)[a]
Sclerodactyly, n/N (%)	14/22 (63.3)
Digital ulcer, n/N (%)	8/22 (36.4)
Telangectasias, n/N (%)	12/22 (54.5)
Stiffness of small joints (finger, wrist), n/N (%)	4/22 (18.2)
Stiffness of large joints (elbow, hip, knee, ankle), n/N (%)	3/22 (13.6)
Gastrointestinal tract distal involvement, n/N (%)	2/22 (9.1)
Pulmonary fibrosis, n/N (%)	9/22 (40.9)
Pulmonary arterial hypertension, n/N (%)	1/22 (4.5)
Scleroderma renal crisis, n/N (%)	1/22 (4.5)

dcSSc: diffuse cutaneous systemic sclerosis; lcSSc: limited cutaneous SSc.

[a]n=22 (4 patients recently included with no available medical information).

Appendix 10. Stability of the CSF-17 at 1-week interval.

Items		ICC	95% CI
	CSF-17 section A	**0.90**	**[0.79 - 0.95]**
1	Writing with a pen or a pencil	0.85	[0.66 - 0.96]
2	Changing my body position	0.69	[0.41 - 0.86]
3	Standing up alone	0.64	[0.10 - 0.86]
4	Lifting and carrying objects in my hands even when moving	0.74	[0.42 - 0.91]
5	Manipulating small objects using my fingers and hands	0.90	[0.73 - 0.97]
6	Moving arms (raise, flex, extend)	0.87	[0.72 - 0.96]
7	Walking	0.87	[0.66 - 0.97]
8	Running	0.89	[0.79 - 0.95]
9	Using public transportation (bus, metro, tramway)	0.51	[0.03 - 0.85]
10	Thinkering, gardening, feeding and taking care of my domestic animals	0.86	[0.68 - 0.94]
	CSF-17 section B	**0.94**	**[0.85 - 0.97]**
11	Learning new things	0.77	[0.14 - 0.95]
12	Focusing my attention	0.86	[0.70 - 0.95]
13	Solving problems of daily life	0.92	[0.81 - 0.97]
14	Undertaking a complex task requiring several steps	0.87	[0.68 - 0.96]
15	Managing my own activity level	0.85	[0.57 - 0.95]
16	Handling stress and other psychological demands	0.81	[0.51 - 0.94]
17	Handling responsibilities in my personal and professional life	0.86	[0.62 - 0.96]
	CSF-17 Total	**0.92**	**[0.83 - 0.96]**

95% CI: 95% confidence interval; CSF-17: Cochin 17-item Scleroderma Functional scale; ICC: intraclass correlation coefficient.

Appendix 11. Response pattern for the 17 questions and 2 domains of the CSF-17. Y-axis represents frequency of the responses and X-axis represents the response options.