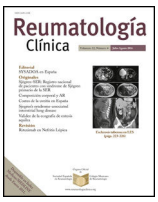
Reumatol Clin. 2019;15(5):282–288

Sociedad Española de Reumatología - Colegio Mexicano de Reumatología

Reumatología Clínica

www.reumatologiaclinica.org



Original Article

The Systemic Sclerosis Questionnaire (SySQ): Validation of the Translation of the Original German Version Into Spanish and Its Relationship to the Disease and to Quality of Life☆

Maria Pilar Cruz-Domínguez,[a,*] Moisés Casarrubias-Ramírez,[b] Victor Gasca-Martínez,[b] Cindy Maldonado García,[b] Rosa Angélica Carranza-Muleiro,[c] Gabriela Medina,[d] Grettel García-Collinot,[c] Daniel Hector Montes-Cortes[e]

[a] *División de Investigación en Salud, Hospital de Especialidades Centro Médico La Raza, Instituto Mexicano del Seguro Social, Mexico City, Mexico*
[b] *Departamento de Medicina Interna, Hospital de Especialidades Centro Médico La Raza, Instituto Mexicano del Seguro Social, Mexico City, Mexico*
[c] *Sección de Estudios de Posgrado e Investigación, Instituto Politécnico Nacional, Hospital de Especialidades Centro Médico La Raza, Escuela Superior de Medicina, Instituto Mexicano del Seguro Social, Mexico City, Mexico*
[d] *Unidad de Investigación en Epidemiología Clínica, Hospital de Especialidades Centro Médico La Raza, Instituto Mexicano del Seguro Social, Mexico City, Mexico*
[e] *División de Investigación en Salud, Unidad de Investigación en Epidemiología Clínica, Hospital Primero de Octubre ISSSTE, Hospital General Centro Médico La Raza, Instituto Mexicano del Seguro Social, Mexico City, Mexico*

ARTICLE INFO

Article history:
Received 25 January 2017
Accepted 1 September 2017
Available online 29 November 2018

Keywords:
Functionality
Quality of life
Systemic sclerosis

ABSTRACT

Objective: Translation, transculturation and validity of the self-administered questionnaire for functionality (Systemic Sclerosis Questionnaires [SySQ]) for use in Spanish patients with systemic sclerosis and its relationship to the severity of the disease and to quality of life.
Patients and methods: We conducted an observational analytical study to perform a cross-cultural validation of the self-administered questionnaire on functionality in scleroderma. The validity of the form and content was evaluated by an expert panel. The method included: (a) adaptation into Spanish of the construct for translation and back translation, and transculturation; (b) internal consistency with the SySQ (Cronbach's alpha), and (c) reproducibility was assessed taking into account all occasions in which the test was performed with Cohen's kappa. Additionally, we calculated the Spearman correlation coefficient with the Medsger severity scale, Health Assessment Questionnaire score and SF-36 score.
Results: We included 70 patients with systemic sclerosis: age 17–78 (51 ± 12) years, 65 (93%) were women, diffuse/limited subtype 64/36%, disease duration of .5–40 years. Optimal internal consistency for all categories of the final version of SySQ (Cronbach's α of .961) and intraobserver reliability in 2 tests over a 2-week interval (Cohen's kappa coefficient .618) and optimal interobserver reliability in 2 tests on the same day (Cohen's kappa coefficient .911). Moderate correlation between functionality by SySQ and by Health Assessment Questionnaire ($r = .573$, $P < .0001$). Inverse correlation between SySQ and quality of life mental health domain SF-36 ($r = -.435$, $P < .001$) and physical domain SF-36 ($r = -.638$, $P < .001$). Medsger severity scale (tendon, heart, lung, vascular) also showed significant correlation with SySQ.
Conclusions: SySQ in this validated Spanish version is a suitable instrument to measure functional status in patients with systemic sclerosis. Reduced functionality is related to greater tendon and peripheral vascular involvement and to a poorer quality of life.

© 2017 Elsevier España, S.L.U. and Sociedad Española de Reumatología y Colegio Mexicano de Reumatología. All rights reserved.

☆ Please cite this article as: Cruz-Domínguez MP, Casarrubias-Ramírez M, Gasca-Martínez V, Maldonado García C, Carranza-Muleiro RA, Medina G, et al. Cuestionario de funcionalidad para esclerosis sistémica (SySQ): validación en español del original en alemán y su relación con la enfermedad y la calidad de vida. Reumatol Clín. 2019;15:282–288.

* Corresponding author.
E-mail address: drapilarcd@live.com.mx (M.P. Cruz-Domínguez).

2173-5743/© 2017 Elsevier España, S.L.U. and Sociedad Española de Reumatología y Colegio Mexicano de Reumatología. All rights reserved.

Cuestionario de funcionalidad para esclerosis sistémica (SySQ): validación en español del original en alemán y su relación con la enfermedad y la calidad de vida

Palabras clave:
Funcionalidad
Calidad de vida
Esclerosis sistémica

R E S U M E N

Objetivo: Traducción, transculturización y validez del cuestionario autoadministrado para funcionalidad (Systemic Sclerosis Questionnaire [SySQ]) en esclerosis sistémica al idioma español y su relación con la enfermedad y la calidad de vida.

Pacientes y métodos: Estudio observacional analítico. Validación realizada por panel de expertos, en apariencia y contenido. La metodología incluyó: a) adaptación al español del constructo por traducción, retrotraducción, y transculturización; b) consistencia interna para todas y cada una de las categorías del SySQ (alfa de Cronbach), y c) la reproducibilidad se evaluó tomando en cuenta todas las ocasiones en que se realizó la prueba con kappa de Cohen. Adicionalmente calculamos el coeficiente de correlación de Spearman con: 1) escala de severidad de Medsger; 2) Health Assessment Questionnaire, y 3) prueba SF-36.

Resultados: Se incluyeron 70 pacientes con esclerosis sistémica, edad 17-78 (51 ± 12) años, 65 (93%) mujeres, subtipo difuso/limitado 64/36%, evolución de la enfermedad 0,5-40 años. Observamos consistencia interna óptima de la versión final del SySQ (alfa de Cronbach 0,961) y buena reproducibilidad intraobservador entre pruebas con intervalo de 2 semanas (kappa de Cohen 0,618) y óptima interobservador el mismo día (kappa de Cohen 0,911). Moderada correlación entre los cuestionarios de funcionalidad SySQ y de discapacidad de Health Assessment Questionnaire ($r = 0{,}573$; $p < 0{,}0001$). Correlación inversa entre SySQ y calidad de vida en dominio mental del SF-36 ($r = -0{,}435$; $p < 0{,}001$) y en dominio físico del SF-36 ($r = -0{,}680$; $p < 0{,}001$). La escala de severidad de la enfermedad de Medsger (tendón, corazón, pulmón, vascular) también mostró correlación significativa con SySQ.

Conclusiones: Esta versión en español del cuestionario autoadministrado SySQ es un instrumento válido para evaluar el estado funcional de pacientes con esclerosis sistémica. La menor funcionalidad está relacionada con una mayor afección a tendón y vascular periférico y con una menor calidad de vida.

© 2017 Elsevier España, S.L.U. y Sociedad Española de Reumatología y Colegio Mexicano de Reumatología. Todos los derechos reservados.

Introduction

Systemic sclerosis (SS) or scleroderma is a rheumatologic disorder which mainly affects women, leading to vasculopathy, autoimmunity and fibrosis of the skin and visceral organs.[1] During its initial stage, progressive, vascular dysfunction predominates and involves organs such as the skin, lung, heart, muscles, tendons, digestive tract, kidney and general health status.[2] Many methods have been proposed for the measuring of SS activity, prognosis and assessment of therapeutic response, but few assess functionality. The speed of disease progression is highly varied for each individual and functionality is the main indicator of systemic improvement or impairment. The disability index of the Health Assessment Questionnaire (HAQ) is a tool which predicts survival. Although the HAQ is used in SS, it was designed for patients with rheumatoid arthritis. Its adaptation to SS (scleroderma Health Assessment Questionnaire) bears little correlation to disease severity.[3] In 1998, a functionality scale was designed in England, but the patients qualified their disability as considerably higher than their therapist and it was not used.[4] Another test refers solely to hand mobility in sclerodermia or the HAMIS Test.[5] In view of this situation Ruof et al.[6] designed a self-administered functionality questionnaire for SS (Systemic Sclerosis Questionnaire [SySQ]), which assesses the difficulty in carrying out everyday life activities and the intensity and frequency of symptoms with the highest functional impact in SS. This instrument was originally designed in German and has been validated into other languages, but not into Spanish. Scales were recently published for SS assessment but no functionality scale was specifically included for SS,[7] and it was therefore our objective to validate the SySQ functionality scale transculturally in Mexican patients with SS and assess its relationship with the HAQ disability scale, disease severity and quality of life.

Patients and Methods

We conducted an observational analytical study in the Internal Medicine Department of the Specialities Hospital of the La Raza Medical Centre, of the Mexican Social Security Institute. We included all patients of a cohort diagnosed with SS, according to the criteria of the American College of Rheumatology of 1980 and confirmed in their classification with the American College of Rheumatology and the European League Against Rheumatism of 2013,[8] who knew how to read and write and who signed the consent form. Excluded patients were those with syndromes overlapping clinical evaluation, mixed connective tissue disease, comorbidities affecting quality of life which were different from SS or those with impediments to questionnaire response. The protocol was approved by the local research and research ethics committee, registered with number R-2010-3501-40.

Phase 1. Translation and Transculturation

Translation and transculturation followed methodology used by other studies recommended by Falcão et al.[9] The tool was initially translated from German into Spanish by a native translator, creating the first Spanish version. The initial translation was reverted into the original language and later compared with the original tool. Any discrepancies found were jointly reviewed by an internist, a rheumatologist and the translator, adapting them and generating a second Spanish version. The semantic equivalence was analysed based on the vocabulary and corresponding grammar, idiomatic equivalence, translation of idiomatic expressions and conceptual equivalence, since the terms could be semantically equivalent but not conceptually so.

Following translation of the SySQ questionnaire a content assessment was made by a panel of 5 experts (2 rheumatologists,

2 clinical researchers, one internist). These included 3 domains concerning daily life activities, intensity and frequency of symptoms. Three domains were distributed into 4 categories: (1) general symptoms; (2) musculoskeletal symptoms; (3) cardiopulmonary symptoms, and (4) digestive tract symptoms, which are the main organs affected and represent the highest impact on functionality of patients with SS. This scale includes a section on the capacity to take an action (0 = no difficulty, 1 = a little difficulty, 2 = some difficulty, 3 = unable to do); another section describes the intensity of symptoms (0 = none, 1 = mild, 2 = moderate, 3 = very intense), and finally there is a section on symptom frequency (0 = no, 1 = sometimes, 2 = often, 3 = always). The final score of the scale is calculated by averaging the maximum value obtained in each domain (block of questions) divided between 3 (given that there are 3 domains), and this value ranges from 0 (normal functionality) to 3 (minimum functionality).

This version was later applied to a group of 10 patients, adding to each of the questions the option "not applicable" so as to identify the questions which were not understood or were not used regularly by our population and therefore were considered as culturally inappropriate. Questions with over 25% of "not applicable" were analysed by the committee and then replaced with others with the same concept. Patient opinions on the clarity and facility to respond to reactions were considered and appearance was corrected on 2 occasions and the content in 3 questions. These changes led to the new version which was re-applied to the patients, until "not applicable" was not obtained in over 15% of patients, generating the Mexican version to the Spanish version of the questionnaire.

Phase 2. Application of the Questionnaire and Assessment of the Psychometric Measurements

In phase 2 we included 70 consecutive patients in follow-up visits out of a cohort of 210 patients with SS. The SySQ functionality self-administered questionnaire (Fig. 1) was administered, and adjusted with corrective questions which are shown and explained in Fig. 2. The disability self-questionnaire (HAQ) was applied, as was SF-36 quality of life quesionnaire,[10] and the Medsger[11] severity index. The latter took into account laboratory and imaging studies which had been taken no more than 3 months after the appointment date.

The final version of the translated questionnaire was assessed by 3 interviews with each patient: initially by 2 different interviewers to assess interobserver reproducibility and 14 days later by the first interviewer to assess intraoberver reproducibility. The SySQ questionnaire responses from the first interview were taken for internal consistency analysis using Cronbach's alpha. Reproducibility was assessed by taking into account all the occasions when the Cohen kappa test was performed. To relate the SySQ self-administered questionnaire with scales which could affect functionality, we calculated Spearman's coefficient with the HAQ disability scale and with Medsger's disease severity scale as a good indicator of specific damage to organs.[12] We used the SF-36 quality of life questionnaire proposed as the appropriate one for this variable related to health in sclerodermia in a meta-analysis.[13,14]

Clinical and Paraclinical Data Collection

The clinical examination was performed the same day as the functional assessment for each patient included in the study. The paraclinical parameters were accepted within an interval of the last 3 months, routinely collected during follow-up. We included age, gender, SS subtype (diffuse or limited), duration of the disease from the first SS symptom and the antibody associated with sclerodermia by immunoenzymatic analysis. The severity of the skin and internal organ involvement was assessed using a scale modified by Rodnan and the Medsger scale, respectively.[11,15] Peripheral vascular involvements was assessed with the Medsger in gangrene or amputation, ulcers and scars. In the absence of these data the spontaneous active Raynaud phenomenon was assessed or placing the hands in water at 12 °C for 2–5 min to search for a change in colouring to paleness, hyperaemia or cianosis.[16]

Statistical Analysis

We calculated the sample size as 47 patients by considering the minimum correlation of .4, plus 10% of possible losses and with a statistical power of 80% we re-calculated the sample as 52 patients. Initially the simple was random but many people did not meet with the inclusion criteria and subjects were consecutively included from the whole cohort.

The results with normal distribution are presented as a mean ± standard deviation. Ordinal variables are expressed as percentage, median and minimum and maximum range.

We used Chronbach's alpha to assess the internal consistency of the questionnaire in its final version applied in the first interview, the kappa coefficient, to identify the proportion of concordance observed between the first and second response to the questionnaire in one same day (interobserver) and between the initial and another 2 weeks later (intraobserver), taking as values between 0 (no concordance) and 1 (maximum concordance).

The SySQ self-administered questionnaire was considered valid if a Crohbach alpha of >.8 was obtained and a Cohen kappa of >.6. We identified adequate or good correlation with the HAQ disability scales, quality of life (SF-36) and Medsger disease severity.[7,10–14] Statistical analysis was performed with SPSS v21 software.

Results

We included 70 patients with SS, of 51.32 ± 11.95 (17–78) years of age, 65 (93%) women, 45 (64.3%) of whom presented with the diffuse cutaneous variety, and 25 (35.7%) with limited cutaneous type, 5(7%) of them without dermal involvement or "sine sclerodermia".[17,18] Their evolution was 7 (.5–40) years (Table 1). The organs most frequently involved were skin (93%), vascular (77%) and gastrointestinal (77%) system; the least affected was the kidney (8.5%) (Table 1).

Each of the 3 domains of SySQ were affected to some extent in around 70% of cases. Functionality (SySQ) was most frequently affected in the capacity for pressure or in reaching (82.9%) and less regularly when walking (57.1%); the total score was 1.66 (.6–3.0) (Table 2). Frequency of symptoms was high in 10% of patients and in 58.5% to a lesser extent, and for this reason was the domain with the highest score (2.1–3). Although ability to undertake daily life activities affected 70% of the patients, this was to a very low degree, and obtained the lowest score (1.0–3) (Table 2). The internal consistency was as good as the original tool, since Cronbach's alpha was .96 for the total of the tool and in the original article Cronbach's alpha was .93. We observed good reproducibility of the baseline test in duplicate on the first day and assessed by different researchers (Cohen kappa .911) and a second test with a 2-week interval by the same researcher (Cohen kappa of .618).

Seventy percent (70%) of the patients included manifested affected functionality, mainly classified as "with some difficulty", and the symptoms which limited their functionality were mostly "of moderate intensity" and "often" in frequency (Table 2).

The HAQ disability questionnaire uses a total score range from 0 to 3, similar to SySQ. With HAQ we obtained a total score of a little less than that obtained with SySQ (1.0–2.8). The 8 domains of the HAQ scale were affected in 58%–65% of the patients, and with a mild disability median (Table 3). The abilities domain of the SySQ

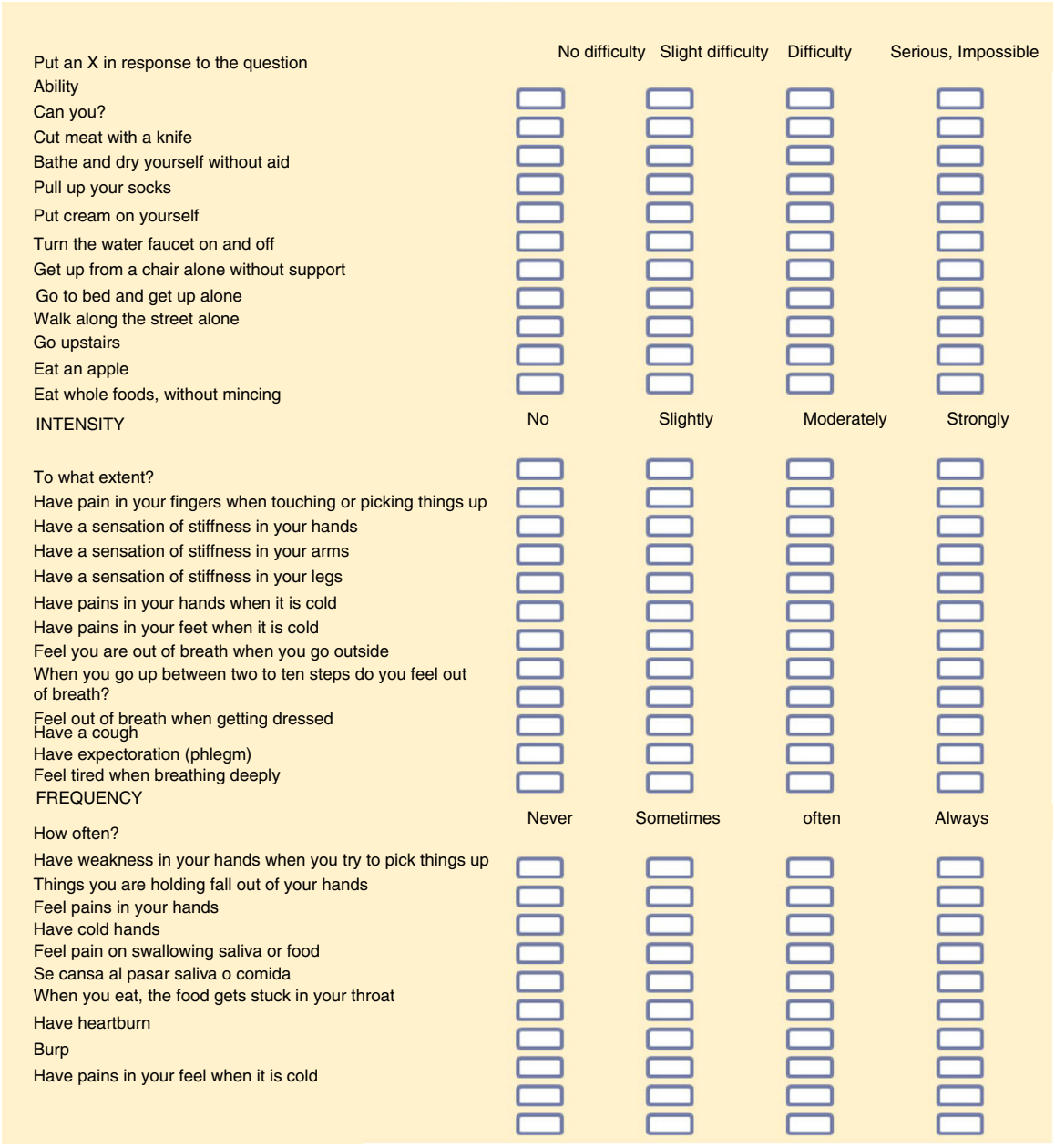


Fig. 1. Mexican version in Spanish of the self-administered functionality scale of the Systemic Sclerosis Questionnaire.
Score. *First.* 32 scores were obtained in 3 domains. We chose the highest score for each domain (11 responses to ability, 12 to intensity and 9 to frequency). Initial scores were therefore reduced to 3. *Second.* To avoid confusions in low scores (0 and 1) a requirement was made to look at the corrective questions, as described in the section on Material and method. *Third.* The mean of the 3 values of the domains was calculated: (a) abilities, (b) intensity, and (c) frequency. This will be the final score of the SySQ questionnaire. Interpretation: 0 = no difficulty or no symptoms; 1 = slight difficulty, presences of mild symptoms and/or on some occasions; 2 = serious difficulty, moderate symptoms and/or symptoms present frequently, and 3 = incapacity, strong or intense symptoms which always present.

scale also had a median in the range described as slight difficulty on carrying out everyday activities (Tables 2 and 3). Spearman's correlation coefficient between the HAQ and SySQ scales was moderate ($r = .573$, $P = .0001$).

We use the SF-36 (best 0-worst 100) questionnaire to analyse quality of life. In the physical domain we found 35.9 (2.8–82.5). The domain which most affected patients was body pain (98.4%; 45.0–100), followed by physical role (45.7%; .0–100) (Table 4). The mental domain affected 45% (9.3–90.8), vitality and mental health 100% of patients (50.10–100 and 48.10–96, respectively), and the emotional role affected 75.7% (3.3, 0–100) (Table 4).

The Spearman correlation was inverse between HAQ and the mental domain of the SF-36 ($r = -.390$, $P = .001$) and the physical domain of the SF-36 ($r = -.620$, $P = .0001$). The relationship was also inverse between the functionality SySQ scale and the mental domain of the SF-36 ($r = -.435$, $P = .0001$) and the physical domain of the SF-36 ($r = -.680$, $P = .0001$) (Table 5)

The SySQ correlation was moderate with the severity of the Medsger disease in the tendon ($r = .458$; $P < .0001$), and adequate for the lung, heart, vascular and muscular system (Table 5).

The degree of limitation of everyday life SySQ activities with the overall Medsger severity scale had a moderate to good correlation ($r = .563$, $P < .0001$), which shows that the degree of dysfunction in everyday activities has a direct correlation with the degree of involvement to organs in SS. Analysis by domains with the best correlation was the tendon and the intensity of the Raynaud phenomenon ($r = .401$, $P < .003$) (Table 5).

Corrective questions. When the patient obtains scores 0 and 1 in a section. The questions of the SySQ items are rephrased with the use of corrective questions. If 2 or 3 is obtained as a score this step is ommitted.

Mark which daily life activities you have difficulty with and require assistance.

___ Cut meat with a knife
___ Bathe and dry yourself without aid
___ Put your socks on
___ Put cream on yourself
___ Turn the water faucet on and off
___ Get up from a chair alone without suppor
___ Go to bed and get up from bed alone
___ Walk along a flat street
___ Go upstairs
___ Eat an Apple

___ Eat whole foods without cutting them up into small pieces

Mark whether these symptoms are so intense that they prevent you carrying out your daily activities:

___ Pain in your fingers when touching or holding things
___ A sensation of stiffness in your hands
___ A sensation of stiffness in your arms
___ A sensation of stiffness in your legs
___ Pain in your hands when it is cold
___ Pain in your feet when it is cold
___ Feel you are out of breath when you go outside

___ When you go up betweentwo to ten steps do you feel out of breath?
___ Feel out of breath when getting dressed
___ Have a cough
___ Have expectoration (phlegm)
___ Feel tired when breathing deeply

Mark whether these symptoms are so frequent that they prevent you carrying out your daily activities:

___ Weakness in your hands when you are holding things
___ Things fall from your hands when you are holding them
___ Feel pains in your hands
___ Have cold hands
___ Feel pain on swallowing saliva or food
___ gets tired when passing saliva or food
___ When you eat, the food gets stuck in your throat

___ Heartburn
___Burping

Example: if in the ability to carry out activities section the patient answered the following:
Can you….

1. Cut meat with a knife (0) (no difficulty)

2.Bathe and dry yourself without aid (1) (slight difficulty)

3. Put your socks on (2) (serious difficulty)

4. Put cream on yourself (3) (impossible)
The score will be (3). i.e. the highest value of the items which compose the domain a) ability to undertake activities. In all domains where a score of (2) or (3) was obtained it is not necessary to look at corrective questions.

Second. Look at the corrective questions. Many people get confused at this point. The task is made easier if they understand the meaning of the corrective questions. Their purpose is to avoid overly low scores which occur if the patient responds that they carry out their activities without difficulty (0) or with slight difficulty (1), but they realise they need help from someone else, some type of utensil, technical aid to carry out these same activities or that the symptoms are so intense or frequent that they affect their everyday activiites. In this case the initial score of the domain of (0) or (1) becomes (2) but never (3). If one domain obtains a score of (2) or (3) correcctive questions are omitted.

Example:

If in the intensity of symptoms domain the patient answered:
How frequent are your symptoms….

1. Weakness in your hands when holding things (0) never

2. Things you are holding in your hands fall

(1) sometimes
But if they later marked that a symptom limits their everyday activities then the score is two instead of one.

Third: Calculate the mean. Find the mean of the 3 values corresponding to the 3 domains described. A) Abilities b) Intensity, c) Frequency. This will be the final score of the SySQ questionnaire. The score of the SySQ will range between 0 (no difficulty) to 3 (incapacity), 0 (no symptoms) to 3 (strong and intense symptoms) and 0 (no symptoms) to 3 (always). If the patient does not answer an item it will be assigned the highest vlaue of the remaining ones which make up that domain. If a complete domain had no answers, it is invalid.

Fig. 2. Corrective questions for the version into Spanish of the Systemic Sclerosis Questionnaire.

Discussion

The Spanish version of the functionality questionnaire of systemic sclerosis (SySQ) proved to be valid and reliable for Mexican patients with SS, using a methodology for transition and transculturation which is reproducible and reflects to some extent the severity of organ involvement.

In contrast to other rheumatic diseases, few questionnaires have been designed for patients with SS: As a result, functional limitation in these patients has been measured in clinical trials by the HAQ-DI disability index, a tool developed to measure disability in patients with rheumatoid arthritis. Other tools have been developed and validated, the majority focusing on specific organs involved in the SS.[10,11] This tool was previously published by our group as a summary presented in a congress,[19] and later translated and validated into Portuguese and Italian.[20,21] In Mexico there are no specific tools in the Spanish language to assess functionality in these patients.

Ruof et al.[6] developed the questionnaire in German, covering general symptoms, in addition to gastrointestinal, cardiopulmonary and musculoskeletal ones. We assessed the SySQ, HAQ and SF-36 scales in order to find the differences and similarities on classifying the ability of patients to undertake everyday activities and the relationship with the damage to different organs. We found there was a good relationship between functionality and the severity of the Raynaud phenomenon, because as this is one of the initial signs of the disease it represents an impact from the beginning of SS. Also with tendon involvement, which usually appears

Table 1
Patient Characteristics.

Demographics		
Age (years), mean ± standard deviation		51 ± 12
Gender, n (%)		
Female		65 (93)
Male		5 (7)
Evolution (years), median (min-max)		7 (.–40)
Clinical presentation. Subtype of systemic sclerodermia, n (%)		
Diffuse cutaneous		45 (64)
Limited cutaneous		25 (36)
Antibodies, n (%)[a]		
Antinuclear antibodies		67 (88.2)
Nucleolar		24 (35.3)
Anti-Scl70		16 (23.5)
Anticentromere		14 (20.6)
Anti-RNP		7 (10.3)
Organ or system	*Median (min-max)*	*Affected patients, n (%)*
General state of health	0 (0–3)	18 (26)
Peripheral vascular	1 (0–4)	54 (77)
Skin	1 (0–4)	65 (93)
Tendon	1 (0–4)	41 (59)
Muscular	1 (0–3)	46 (66)
Gastrointestinal tract	1 (0–2)	54 (77)
Lung	1 (0–4)	38 (54)
Heart	0 (0–4)	15 (21)
Kidney	0 (0–2)	6 (9)

Anti-RNP: anti-ribonucleoprotein antibodies; max: maximum; min: minimum.
[a] Indirect immunofluorescence or enzyme-linked immunosorbent assay (ELISA).

Table 2
Systemic Sclerosis Questionnaire Functionality Scale.

Domain	Median (min–max)	Affected patients, *n* (%)
Limitation of activities	1 (0–3)	48 (69)
Intensity of symptoms	2 (0–3)	49 (70)
Frequency of symptoms	2 (1–3)	50 (71)

max: maximum; min: minimum.

Table 3
Health Assessment Questionnaire Disability Scale.

Domain	Median (min–max)	Affected patients, *n* (%)
Get dressed	1 (0–3)	41 (58)
Get up	1 (0–3)	41 (58)
Eat	1 (0–3)	45 (64)
Walk	1 (0–3)	43 (62)
Hygiene	1 (0–3)	39 (56)
reach	1 (0–3)	45 (64)
Pressure	1 (0–3)	45 (64)
Others	1 (0–3)	46 (65)

max: maximum; min minimum.

Table 4
SF-36 Quality of Life Questionnaire.

Domain	Median (min-max)	Cronbach's alpha
Physical control		
Physical function	47.5 (0–100)	.938
Physical role	45.7 (0–100)	.874
Body pain	45 (0–100)	.944
General health status	27.5 (0–85)	.867
Mental control		
Emotional role	33.3 (0–100)	.662
Social function	50 (0–100)	.87
Vitality	50 (10–100)	.878
Mental health	48 (10–96)	.806

max: maximum; min minimum.

Table 5
Spearman's Correlation of Systemic Sclerosis Questionnaire With Health Assessment Questionnaire, SF-36 and Medsger.

	Correlation coefficient	*P*
HAQ	.573[a]	<.001
SF-36	–.564[a]	<.001
Mental control	–.435[a]	<.001
Physical control	–.638[a]	<.001
Medsger	.563	<.0001
General	–.044	.716
Peripheral vascular	.401[a]	.003
Skin	.202	.093
Tendon	.458[a]	<.0001
Muscular	.260[b]	.029
Gastrointestinal	.194	.107
Pulmonary	.360[b]	.002
Cardiac	.336[b]	.004
Renal	.116	.339

HAQ: Health Assessment Questionnaire.
[a] Correlation moderate to good. Spearman correlation coefficient between the analysed tools.
[b] Weak correlation.

during the progressive course of the disease. Other domains which were statistically significant in the relationship between functional status and severity of the disease were muscular, cardiac and pulmonary signs as a direct link between functionality and organicity assessable through the SySQ questionnaire.

In our study, the questionnaire translated into Spanish proved to be reproducible, and usable in the Mexican population. The validity and transcultural adaptation of the original tool was achieved for application to the Mexican population.

In our study we found a similar spectrum of patients to those found in previous studies where functionality indexes for SS were tried, although the tools used were not specific to this disease.[5–7]

The importance of our study is based on the objective data provided by the SS clinic of our hospital for a comprehensive assessment of the patients and this afforded us the necessary elements for correct application of the severity indexes.

It is important to consider that the questionnaire does not directly assess the Raynaud phenomenon or renal involvement but the presence of pain is an indirect detail demonstrating the existence of this vascular phenomenon and renal involvements is infrequent in the Mexican population.[22] Furthermore, the psychological sphere of the patient has an effect on the self-appreciation of their abilities to carry out daily life, frequency and intensity of symptoms, with consequent greater or lesser appreciation of the same. The psychological area is not considered in the SySQ, and could therefore be complemented with other tools which are able to identify this concern.

Conclusions

The Spanish version of the SySQ questionnaire is valid, reliable, reproducible and accurate for the assessment of functionality in the Mexican population with SS. Our Spanish version of the self-administered SySQ functionality questionnaire had a coherent and inverse relationship with the HAQ disability scale, and with the physical and mental sections of the SF-36 quality of life scale. It also proved to be more closely related to the severity of the involvement of the tendon and peripheral vascular system in SS: The construction of new and better tools are required to allow us to assess not just the severity of symptoms but also the activity and chronicity of SS. The assessment of functionality in SS many also be an indirect indicator of treatment response.

Ethical Liabilities

Protection of people and animals. The authors declare that the procedures followed complied with ethical guidelines of the committee responsible for human experimentation and comply with the World Medical Association and the Declaration of Helsinki.

Data confidentiality. The authors declare that they have adhered to their centre of work on the publication of patient data.

Right to privacy and informed consent. The authors declare that they have obtained the informed consent from the patients and/or subjects referred to in this article. This document is held by the corresponding author.

Conflict of Interests

The authors have no conflicts of interests to declare

Acknowledgements

The authors would like to thank the teacher Hannelore Rayle Staib for translation of the SySQ scale from German into Spanish.

References

1. Allanore Y, Simms R, Distler O, Trojanowska M, Pope J, Denton CP, et al. Systemic sclerosis. Nat Rev Dis Primers. 2015;1:15002.
2. Becker MO, Riemekasten G. Risk factors for severity and manifestations in systemic sclerosis and prediction of disease course. Expert Rev Clin Immunol. 2016;12:115–35.
3. Medsger TA Jr. Natural history of systemic sclerosis and the assessment of disease activity, severity, functional status, and psychologic well-being. Rheum Dis Clin North Am. 2003;29:255–73.
4. Silman A, Akesson A, Newman J, Henriksson H, Sandquist G, Nihill M, et al. Assessment of functional ability in patients with scleroderma: a proposed new disability assessment instrument. J Rheumatol. 1998;25:79–83.
5. Sandqvist G, Nilsson JÅ, Wuttge DM, Hesselstrand R. Development of a modified hand mobility in scleroderma (HAMIS) test and its potential as an outcome measure in systemic sclerosis. J Rheumatol. 2014;41:2186–92.
6. Ruof J, Brühlmann P, Michel BA, Stucki G. Development and validation of a self-administered systemic sclerosis questionnaire (SySQ). Rheumatology (Oxford). 1999;38:535–42.
7. Pope J. Measures of systemic sclerosis (scleroderma): Health Assessment Questionnaire (HAQ) and Scleroderma HAQ (SHAQ), physician- and patient-rated global assessments, Symptom Burden Index (SBI), University of California, Los Angeles, Scleroderma Clinical Trials Consortium Gastrointestinal Scale (UCLA SCTC GIT) 2.0. Baseline Dyspnea Index (BDI) and Transition Dyspnea Index (TDI) (Mahler's Index), Cambridge Pulmonary Hypertension Outcome Review (CAMPHOR), and Raynaud's Condition Score (RCS). Arthritis Care Res (Hoboken). 2011;63 Suppl 11:S98–111.
8. Van den Hoogen F, Khanna D, Fransen J, Johnson SR, Baron M, Tyndall A, et al. 2013 classification criteria for systemic sclerosis: an American College of Rheumatology/European League against Rheumatism collaborative initiative. Arthritis Rheum. 2013;65:2737–47.
9. Falcão DM, Ciconelli RM, Ferraz MB. Translation and cultural adaptation of quality of life questionnaires: an evaluation of methodology. J Rheumatol. 2003;30:379–85.
10. Khanna D, Furst DE, Clements PJ, Park GS, Hays RD, Yoon J, et al. Responsiveness of the SF-36 and the Health Assessment Questionnaire Disability Index in a systemic sclerosis clinical trial. J Rheumatol. 2005;32:832–40.
11. Medsger T, Silman A, Steen V, Black C, Akesson A, Bacon P, et al. A disease severity scale for systemic sclerosis: development and testing. J Rheumatol. 1999;26:2159–67.
12. Khanna D, Merkel PA. Outcome measures in systemic sclerosis: an update on instruments and current research. Curr Rheumatol Rep. 2007;9:151–7.
13. Hudson M, Thombs BD, Steele R, Panopalis P, Newton E, Baron M. Health-related quality of life in systemic sclerosis: a systematic review. Arthritis Rheum. 2009;61:1112–20.
14. Vilagut G, Ferrer M, Rajmil L, Rebollo P, Permanyer-Miralda G, Quintana JM, et al. [The Spanish version of the Short Form 36 Health Survey: a decade of experience and new developments]. Gac Sanit. 2005;19:135–50 [in Spanish].
15. Poormoghim H, Lucas M, Fertig N, Medsger TA. Systemic sclerosis sine scleroderma. Arthritis Rheum. 2000;43:444–51.
16. Vera-Lastra O, Sauceda-Casas CA, Cruz Domínguez MD, Mendoza Alvarez SA, Sepulceda-Delgado J. Systemic sclerosis sine scleroderma in Mexican patients. Case reports. Reumatol Clin. 2017, pii: S1699-258X(16)30159-00.
17. Clements PJ, Lachenbruch PA, Seibold JR, Zee B, Steen VD, Brennan P, et al. Skin thickness score in systemic sclerosis: an assessment of interobserver variability in 3 independent studies. J Rheumatol. 1993;20:1892–6.
18. Lau CS, Khan F, Brown R, McCallum P, Belch JJ. Digital blood flow response to body warming, cooling, and rewarming in patients with Raynaud's phenomenon. Angiology. 1995;46:1–10.
19. Cruz-Dominguez MP, Casarrubias-Ramirez M, Gasca MV, Vera-Lastra OL, Jara-Quezada LJ, Montes-Cortes DH. Self-administered systemic sclerosis questionnaire Validation of a Spanish version (SYSQ) in Mexicans. Arthritis Rheum. 2011;63:665 [abstract].
20. Lacerda RI, Medeiros L, Medeiros EA. Translation, cultural adaptation and validation into Portuguese (Brazil) in Systemic Sclerosis Questionnaire (SySQ). Rev Bras Reumatol. 2014;54:95–101.
21. Uras C, Giannantoni P, Tabolli S, Di Giulio P, Peghetti A, Cianchini G, et al. Measuring disability of women with systemic sclerosis: validation of the Italian version of the systemic sclerosis questionnaire. G Ital Dermatol Venereol. 2016;151:332–9.
22. Rojas-Serrano J, Codina-Velásquez H, Medrano-Ramírez G, Abraham SJ, Vera-Lastra O, Vázquez-Mellado J. [Lower incidence of severe damage to target organs in Mexican patients with systemic sclerosis and diffuse skin affection]. Reumatol Clin. 2008;4:3–7 [in Spanish].