Reliability and validity of the Italian version of the UCLA Scleroderma Clinical Trial Consortium Gastrointestinal Tract Instrument in patients with systemic sclerosis

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ABSTRACT

Objective. *To test the acceptability, feasibility, reliability and validity of the Italian translated version of the UCLA Scleroderma Clinical Trial Consortium GIT (UCLA-SCTC GIT) 2.0. Gastrointestinal tract (GIT) involvement is frequent in systemic sclerosis (SSc). The UCLA-SCTC GIT 2.0 is a validated instrument for measuring the presence and impact of GIT symptoms in SSc patients.*

Methods. *Acceptability and feasibility of the questionnaire were evaluated based on the input from the patients. Internal consistency was evaluated by Cronbach's alpha. External consistency was measured by comparing with the Short Form (SF)-36 and EQ-5D by Spearman's rho, meaningful if ≥0.30.*

Results. *Sixty-two consecutive SSc patients (mean age 60.6) were recruited, 88.5% were female. The UCLA-SCTC GIT 2.0 was well accepted. Percentage of missing data in UCLA-SCTC GIT total score was 2 %. Internal consistency was acceptable (alpha ≥0.70) for all domains. Cronbach's alpha was ≥0.70 for all domains. UCLA-SCTC GIT 2.0 discriminated between patients with or without gastroesophageal reflux disease whether diagnosed clinically or by objective testing (p<0.01 for both). UCLA-SCTC GIT emotional well-being was correlated with the conceptually equivalent SF-36 mental health domains (correlation coefficient >0.35) and with the EQ-5D usual activities domain (0.38), thus reflecting the impact on everyday activities. The distention/bloating domain strongly correlated with the EQ-5D anxiety/depression domain (0.51) and reflux domain with role emotional of SF-36 (0.44).*

Conclusion. *This is the first validation study of the Italian version of UCLA-SCTC GIT 2.0. Our data support its feasibility, reliability, and validity in Italian SSc patients.*

Introduction

Systemic sclerosis (SSc) is a chronic systemic connective tissue disease characterised by diffuse organ fibrosis and vasculopathy. SSc may affect different organs and systems, and gastrointestinal tract (GIT) manifestations – both upper and lower – may be present in up to 90% of patients (1). GIT symptoms in patients with SSc may be associated with incremental reductions in health-related quality of life (HRQoL) (2-4).

Recently, our research team developed and revised a GIT instrument specific for SSc, called the University of California, Los Angeles Scleroderma Clinical Trial Consortium Gastrointestinal Tract Instrument (UCLA-SCTC GIT 2.0) (5, 6). UCLA-SCTC GIT 2.0 was developed to assess GIT symptoms severity and its impact on HRQoL and consists of 34 items with seven domains: reflux, distention/bloating, diarrhoea, faecal soilage, constipation, emotional well-being, and social functioning. The original English version of the instrument is feasible and showed acceptable reliability and validity (5). UCLA-SCTC GIT 2.0 has already proved useful in evaluating changes after treatment in a pilot study on the effectiveness of probiotics in ameliorating distention/bloating symptoms in SSc patients (7).

The aim of this study was to assess the acceptability, feasibility, reliability and construct validity of the Italian version of the UCLA-SCTC GIT 2.0.

Subjects and methods

Translation

The Italian version of the UCLA-SCTC GIT 2.0 instrument was produced by PharmaQuest Ltd., a specialist PRO translation and linguistic validation agency, using methodology conforming to the International Society for Pharma-

coeconomics and Outcomes Research (ISPOR) guidelines (8).

A concept elaboration document was produced to explore and clarify all concepts within the source English instrument. Two qualified medical translators then conducted independent forward translations of the instrument, using the concept elaboration document as guidance, and PharmaQuest's Italian in-country investigator reconciled them to create a single Italian version. This was translated back into English by two qualified medical translators and the project manager reviewed the back translations against the source text to identify any discrepancies. Any queries were discussed with the investigator to find the best possible translation solution, following which the translation report was reviewed and approved by the instrument developer. The translation then underwent independent proofreading, pilot testing (incorporating cognitive debriefing) with 5 native speakers of Italian with SSc, and a final proofreading stage. The results from the pilot testing confirmed that the Italian translation was culturally and linguistically suitable for use in Italy and conceptually equivalent to the source text.

Population

We recruited consecutive adult patients diagnosed as having SSc, referring to the Rheumatology Department of the Orthopedic Institute G. Pini (Milan) for clinical care from September 2012 to February 2014. Patients were included after obtaining written informed consent. The study was approved by the local ethics committee. Inclusion criteria were: adult patients fulfilling SSc ACR/EULAR 2013 criteria (9).

In the validation study we captured age, sex, cutaneous SSc subtype as defined by LeRoy (10), disease duration (from Raynaud's - RP and from the first non-RP symptom), serum C-reactive protein (CRP), presence of digital ulceration (active or previous) and medication use (proton pump inhibitors [PPI], prokinetics, oral low-dose corticosteroids, calcium channel blockers [CCBs], antibiotics, antidepressants, immunosuppressant therapy). Pulmonary fibrosis was assessed by high-resolution computed tomography (HRCT) scan and pulmonary function tests (forced vital capacity [FVC] and carbon monoxide diffusion lung capacity divided by alveolar volume [DLCO/VA]). All patients were tested for antinuclear antibodies (ANA) by indirect immunofluorescence (IIF). We also analysed anti-extractable nuclear antigen (ENA) autoantibodies specific for SSc via ELISA.

Health-related quality of life (HRQoL) instruments

Patients completed the UCLA-SCTC GIT 2.0, the Medical Outcomes Short Form (SF)-36 and the EQ-5D with three levels tool. UCLA-SCTC GIT 2.0 is a validated, patient-reported outcome measure to assess HRQoL and GIT symptoms presence and severity in SSc (5, 6). UCLA-SCTC GIT 2.0 has 34 items and seven domains: reflux, distention/bloating, diarrhoea, faecal soilage, constipation, emotional well-being, and social functioning. All domains are scored from 0 (higher HRQoL) to 3 (lower HRQoL) except the diarrhoea and constipation domains, which range from 0–2 and 0–2.5, respectively. The total GI score is the average of six of seven domains (excludes constipation) and is scored from 0 (better HRQo/L) to 2.83 (worse HRQOL). The English and other versions (including Italian) are available online at: http://uclascleroderma.researchcore.org/.

The SF-36 is a generic health status measure consisting of 36 items assessing eight domains (11). The SF-36 consists of four physical health domains (physical functioning [10 items], bodily pain [two items], role limitations due to physical health perceptions [four items], and general health perceptions [five items]), four mental health domains (mental health [five items], role limitations due to emotional problems [three items], vitality [four items] and social functioning [two items]), and a health transition domain [one item]. The four physical health domains are summarised into Physical Component Summary (PCS) score and the four mental health domains are summarised into Mental Component Summary (MCS) score.

The EQ-5D questionnaire with three levels is a generic measure of HRQoL developed and tested by the EuroQoL Group to provide a quantitative measure of health outcomes (12, 13) and was proved to be a valid tool in its Italian version in a first study on SSc patients (14). It consists of two main parts: the first part generates a health profile (EQ-5D profile), based on a descriptive system that defines health in terms of five dimensions, namely 'mobility', 'self-care', 'usual activities', 'pain or discomfort' and 'anxiety or depression'. Each dimension has three response categories corresponding to no problem/some problems/extreme problems. The second part of the questionnaire consists of a visual analogue domain (EQ-5D VAS), measuring overall HRQoL ranging from 0 (worst imaginable health state) to 100 (best imaginable health state).

We used a standard (4-wk) recall period.

Statistical methods

Acceptability and feasibility were assessed based on the complaints of the patients.

We calculated mean scores, standard deviations (SD), ranges and percentage of missing data. Floor and ceiling effects of UCLA-SCTC GIT 2.0 were calculated based on the percentage of respondents scoring the minimum (floor) and maximum (ceiling) possible scores.

Internal consistency of UCLA-SCTC GIT 2.0 reliability was measured by means of Cronbach's alpha (15). We assessed the construct convergent validity by exploring the association between the UCLA-SCTC GIT 2.0 and EQ-5D and SF-36 domains. Spearman's rho was used to assess correlations. We considered correlation coefficients ≤0.29 as small, between 0.30 and 0.49 as moderate, and ≥0.50 as large (16).

All analyses were performed using SPSS software version 21 (College Station, Texas) and $p<0.05$ was considered statistically significant.

Results

Sixty-two consecutive patients with SSc were recruited. Patients had a

Table I. Demographic and clinical characteristics of the study group.

Number of patients	62
Mean age (SD)	60.6 (14.4)
Mean disease duration (SD)	10.8 (9.4)
Mean duration from RP onset (SD)	14.3 (12.1)
Female	88.5%
lcSSc	70.5%
dcSSc	21.3%
veSSc	8.2%
ANA +	27.9%
ACA +	41.0%
ATA +	27.9%
Mean FVC (SD)	102% (20%)
Mean DLCO (SD)	71.4% (20.3%)
Mean DLCO/VA (SD)	78.5% (16.8%)
HRCT documented fibrosis (n/a)	49.2% (1.6%)
Mean left ventricular ejection fraction (SD)	61.8% (4.6%)
PAP mmHg (SD)	28.0 (6.9)
CRP (SD)	0.33 (0.79)
Digital ulcers	
Never	57.4%
Previous	16.7%
Active	24.6%
Positive barium x-ray (n/a)	50.8% (14.8%)
Positive EGD (n/a)	24.6% (11.6%)
GERD clinical diagnosis (n/a)	41.0% (29.5%)
Alternating bowel	1.6%
Diverticulosis	3.3%
Malabsorption	1.6%
Ongoing PPI	70.5%
Prokinetics	41.0%
Low dose CS	27.9%
Antibiotics	8.2%
Antidepressants	26.2%
Immunosuppressants	29.5%
Calcium channel blockers	37.7%

ACA: anti-centromere antibodies; ANA: anti-nuclear antibodies; ATA: anti-topoisomerase antibodies; CRP: C reactive protein; CS: corticosteroids; dcSSc: diffuse cutaneous systemic sclerosis; DLCO: diffusion lung carbon monoxide; EGD: esophagogastroduodenoscopy; FVC: forced vital capacity; GERD: gastroesophageal reflux disease; HRCT: high resolution computed tomography; lcSSc: limited cutaneous systemic sclerosis; n/a: not available; PAP: Pulmonary arterial pressure; PPI: proton pump inhibitors; RP: Raynaud's phenomenon; SSc: systemic sclerosis; VA: Alveolar volumn; veSSc: very early systemic sclerosis.

mean (SD) age of 60.6 (14.4) years, were predominantly female (88.5%) and had limited cutaneous SSc (lcSSc; 70.5%), whereas 8.2% were classified as having very early SSc (veSSc; Table I). The mean (SD) disease duration from RP onset was 10.8 (9.4) years. Mean (SD) score of the UCLA-SCTC GIT 2.0 ranged from 0.80 (0.82) for distention/bloating to 0.15 (0.39) for the diarrhoea domain (Table II). Cronbach's alpha was ≥0.70 for all domains. Percentage of missing data in UCLA-SCTC GIT score was 2% *versus* 5% and 9% for EQ-5D and SF-36. No comments or complaints were reported after completion.

Table II. Descriptive statistics and internal consistency reliability of the UCLA-SCTC GIT 2.0 instrument.

	Mean (SD) score	Minimum score	Maximum score	Ceiling/floor effect	Cronbach's alpha	Test-retest stability (Spearman's rho)
Reflux	0.44 (0.51)	0.000	1.625	33.3/0.0	0.79	0.96
Distention/bloating	0.80 (0.82)	0.000	3.000	23.8/0.3	0.75	0.55
Faecal soilage	0.23 (0.59)	0.000	3.000	81.0/0.2	n/a	0.10
Diarrhoea	0.15 (0.39)	0.000	2.000	81.0/0.0	0.73	0.58
Social functioning	0.22 (0.35)	0.000	1.660	52.4/0.0	0.71	0.54
Emotional wellbeing	0.22 (0.41)	0.000	2.220	57.1/0.0	0.88	0.79
Constipation	0.46 (0.55)	0.000	2.500	41.3/0.0	0.81	0.75
Total GIT score	0.34 (0.37)	0.000	1.584	11.1/0.0	0.74	0.38

GIT: gastro-intestinal tract; n/a: not applicable.
All domains and Total GIT score are scored from 0 (better HRQoL) to 3 (worse HRQoL) except diarrhoea and constipation domains that ranges from 0–2 and 0–2.5, respectively.
Floor/ceiling: percentage of patients reporting worst possible score (floor effect)/best possible score (ceiling effect).

Total GIT score correlated with disease duration since RP and first non-RP symptom (0.38 and 0.33 respectively). An objective diagnosis of gastroesophageal reflux disease (GERD), was moderately correlated with the reflux and distension/bloating domains (correlation coefficient 0.44 and 0.31 respectively) and with the total GIT score (correlation coefficient 0.37). An ongoing PPI therapy was, as expected, correlated with reflux domain (correlation coefficient 0.50). Modified Rodnan's skin score was moderately correlated with reflux domain, distention/bloating, emotional well-being and total GIT score (correlation coefficient 0.59, 0.50, 0.50 and 0.54 respectively). A moderate correlation was also observed for anti-depressive therapy with reflux domain and total GIT domain (correlation coefficient 0.35 and 0.32 respectively).

UCLA-SCTC GIT 2.0 was able to discriminate between patients without or with GERD both clinically (n=25) –mean score (SD) 0.21 (0.22) *versus* 0.69 (0.56), $p<0.01$ – and by Barium x-ray or oesophagogastroduodenoscopy (EGDS): mean score (SD) 0.28 (0.32) *versus* 0.64 (0.58), $p<0.01$. In one patient malabsorption was diagnosed and UCLA-SCTC 2.0 score was significantly different for the diarrhoea domain – score of 1.25 *versus* a mean score (SD) of 0.32 (0.39) – and in one patient alternating bowel was present and UCLA-SCTC 2.0 score was significantly different for the faecal soilage domain -2.00 *versus* a mean score (SD) of 0.05 (0.21).

Test-retest was evaluated in 25 patients after 4 weeks (Table II). We found correlation coefficients >0.50 for all the domains except for faecal soilage, but in this case two patients who had not symptoms at the time of completion of the first questionnaire complained about new-onset faecal soilage, so they were not stable. We found particularly strong stability for reflux, emotional wellbeing, constipation (correlation coefficient 0.95, 0.79, 0.75 respectively). Total GIT score had a correlation coefficient of 0.38 for test-retest stability. Ceiling effect ranged from 11.1% for the total GIT score to 81.0% for the faecal soilage and diarrhoea domains (Table II).

We found moderate correlation between the emotional well-being domain of the UCLA-SCTC GIT 2.0 and the role-emotional, mental health domains and MCS of the SF-36 (correlation coefficient 0.45, 0.37 and 0.41 respectively; Table III); reflux domain of the UCLA-SCTC GIT 2.0 was moderately correlated with role-emotional domain of SF-36 (correlation coefficient 0.44) and distention/bloating domain was moderately correlated with social functioning domain of SF-36 (correlation coefficient 0.37). Social functioning domain of the UCLA-SCTC GIT 2.0 and the social functioning and role-emotional domains of SF-36 had a correlation coefficient of 0.37 and 0.35, respectively.

Table III. Spearman's correlation coefficients of UCLA-SCTC GIT 2.0 and EQ-5D and SF-36.

	Reflux	Distention/ Bloating	Faecal Soilage	Diarrhoea	Social Functioning	Emotional Wellbeing	Constipation	Total GIT score
EQ-5D Mobility	ns	ns	ns	ns	ns	ns	ns	ns
EQ-5D Self care	ns	ns	ns	ns	ns	ns	ns	ns
EQ-5D Usual activities	+0.27*	+0.26*	+0.30 ns	ns	ns	+0.38**	ns	+0.28*
EQ-5D Pain/discomfort	ns	ns	ns	ns	ns	ns	ns	ns
EQ-5D Anxiety/depression	+0.26*	+0.51**	ns	ns	+0.31*	ns	ns	+0.43**
EQ-5D VAS	0.24 ns	ns	ns	ns	ns	ns	ns	ns
SF-36 PF	0.21 ns	ns	ns	ns	ns	ns	0.33*	ns
SF-36 RP	ns	0.25 ns	ns	ns	ns	0.34*	0.34 ns	ns
SF-36 BP	ns	-0.24 ns	ns	ns	ns	ns	ns	ns
SF-36 GH	0.22 ns	0.23 ns	ns	ns	ns	0.24 ns	ns	0.23
SF-36 VT	ns	ns	ns	ns	ns	ns	ns	ns
SF-36 SF	0.28 ns	0.37 ns	0.27 ns	ns	0.37*	0.34*	ns	0.35*
SF-36 RE	0.44*	0.23 ns	ns	ns	0.35*	0.45**	ns	ns
SF-36 MH	ns	0.29*	ns	ns	ns	0.37**	0.25 ns	0.27 ns
PCS	ns	0.23 ns	0.25 ns	ns	ns	0.29 ns	0.34*	0.22 ns
MCS	ns	ns	ns	ns	ns	0.41**	ns	ns

All negative values unless marked (+); * indicate $p<0.05$, ** $p<0.01$. PCS: physical health component summary; MCS: mental health component summary; GH: general health perceptions; PF: physical functioning; RP: physical role functioning; BP: bodily pain; VT: vitality; SF: social role functioning; RE: emotional role functioning; MH: mental health.

Table IV. Mean (SD) UCLA-SCTC GIT 2.0 scores and SD for the different SSc subtypes.

Mean (SD) score	Reflux	Distention/ Bloating	Diarrhoea	Constipation	Faecal Soilage	Emotional well-being	Social functioning	Total GIT score
veSSc	0.02 (0.05)	0.21 (0.29)	0.00 (0.00)	0.00 (0.00)	0.05 (0.13)	0.04 (0.06)	0.17 (0.30)	0.05 (0.08)
lcSSc	0.45 (0.53)	0.84 (0.83)	0.17 (0.44)	0.21 (0.46)	0.27 (0.39)	0.24 (0.48)	0.48 (0.55)	0.36 (0.39)
dcSSc	0.54 (0.47)	0.77 (0.81)	0.46 (0.97)	0.00 (0.00)	0.08 (0.14)	0.19 (0.22)	0.40 (0.52)	0.34 (0.31)

dcSSc: diffuse cutaneous systemic sclerosis; lcSSc: limited cutaneous systemic sclerosis; veSSc: very early systemic sclerosis.

It is noteworthy that the anxiety/depression domain of the EQ-5D correlated significantly with the distention/bloating domain (0.51 $p<0.001$) and the total GIT score (0.43 $p<0.001$), as shown in Table III.

We also distinguished results based on the different disease subtypes (Table IV) and found that the higher mean scores (SD) in veSSc were related to distention/bloating 0.208 (0.29) and constipation 0.167 (0.30); in lcSSc higher scores were reported for distention/bloating 0.84 (0.83), constipation 0.48 (0.55) and reflux 0.45 (0.53); for diffuse cutaneous SSc (dcSSc) distention / bloating 0.78 (0.81); reflux 0.54 (0.47); faecal soilage 0.46 (0.97) and constipation 0.40 (0.52).

Discussion

Involvement of GIT is present in 90% of all patients with SSc and is associated with a decline in HRQoL (17). Both upper and lower GIT can be involved in SSc with a consequent high burden in terms of morbidity, mortality and HRQoL (17, 18). To date, the management of gastrointestinal complications in SSc is still based on limited data and different approaches which often vary amongst clinicians (19). With this as background, a feasible and reliable instrument able to detect and quantify GIT symptom severity and its impact on HRQoL was lacking until the UCLA-SCTC GIT questionnaire was developed and the 2.0 version optimised (20).

The original English version of the UCLA-GIT 2.0 had satisfactory reliability and validity (5).

We translated the UCLA-SCTC GIT 2.0 instrument into Italian and found good reliability as well (as assessed by Cronbach's alpha ≥0.70 in all its domains). Also, test-retest stability was acceptable (0.38 for the total GIT score) and particularly reliable for the reflux, emotional wellbeing and constipation domains (0.95, 0.79, 0.75 respectively correlation coefficient). These findings support the use of the UCLA-SCTC GIT 2.0 as a tool in clinical trials aimed at evaluating GIT treatment.

As far as construct validity is concerned, we found a moderate correlation (correlation coefficient >0.35) between conceptually equivalent mental health domains of the UCLA-SCTC GIT 2.0 and SF-36 such as the emotional well-being domain of the UCLA-GIT 2.0 and the role-emotional domain, mental health domain and MCS of the SF-36; the social functioning domains between the two instruments had a moderate correlation as well (correlation coefficient ≥0.35).

We did not find any significant correlation between physical function of SF-36 and symptom domains of UCLA-SCTC GIT 2.0 apart from constipation and role physical of SF-36. More notably, we also found more significant associations (correlation coefficient

≥0.30) between the reflux domain of the UCLA-SCTC GIT 2.0 and the mental health domains of the SF-36 such as role emotional and the SF-36 MCS compared to the physical health domains. Indeed, the UCLA-SCTC GIT 2.0 reflux domain was moderately correlated with SF-36 role limitation due to emotional problems, a domain which is part of the mental more than the physical health. Furthermore, the anxiety/depression domain of the EQ-5D was strongly associated with the distention/bloating domain and moderately with the total GIT score. Although small, an association was observed even between the distention/bloating domain and the SF-36 mental health. Finally, the UCLA-SCTC GIT 2.0 emotional wellbeing domain significantly correlated with EQ-5D usual activities and, although to a lesser extent, with the role limitation physical problems. Our group already found an association with symptom severity as reflected by the reflux and constipation domains and depressed mood (2). Data from the current study further support these data and suggest that GIT involvement has greater impact on mental compared to physical health.

Although UCLA-SCTC GIT 2.0 domain scores did not significantly correlate with physical functioning, as already found by the French group who translated and validated the questionnaire for French SSc patients (21), we further demonstrated that GIT symptoms may interfere with accomplishing daily activities related to physical activities, as shown by the presence of a small correlation between the total GIT score and EQ-5D usual activities (0.28 $p<0.05$).

In comparison to our original study of the English version, several baseline differences in the study population were noted (5). The Italian version was assessed on a small population (n=62 *vs*. 152); our patients were older (mean age = 60.6 *vs*. 50.9 years); and this study included fewer patients with dcSSc (21.3% *vs*. 55.3%). The HRQoL, as captured by the SF-36 showed mean SF-36 PCS and MCS scores of 40.8 and 45.6, respectively, compared to 36.7 and 47.1 in the original study. The Italian version of our instrument showed a higher mean score only in the constipation domain (0.48 *vs*. 0.43). Conversely, the other domains including the total GIT score had lower mean scores: reflux (0.44 *vs*. 0.69), distention / bloating (0.80 *vs*. 1.07), faecal soilage (0.23 *vs*. 0.30), diarrhoea (0.15 *vs*. 0.56), social functioning (0.22 *vs*. 0.26), emotional well-being (0.22 *vs*. 0.49) and total GIT score (0.34 vs. 0.66), suggesting somewhat milder GIT involvement in the sample of the current study. Both studies showed a high ceiling effect, ranging from 11.1% in total GIT score to 81.0% in faecal soilage and diarrhoea domains in the current study. Test-retest stability was performed in 25 patients out of 62. Not all patients were stable and some of them complained about the onset of new symptoms regarding faecal soilage. This is the reason of the low score in this domain and, likely of the low GIT total score we found in our study.

Of note, we even distinguished the population of the study into three different groups based on SSc subtypes (veSSc, lcSSc, dcSSc) and found that higher scores in veSSc were those regarding distention/bloating and constipation, whereas those in lcSSc and dcSSc were distention/bloating, constipation and reflux domains. Although no statistical significant difference was found in the three groups, these observations may suggest a slightly different involvement in the three SSc subtypes, which prompt a more thorough and possibly longitudinal study to better assess this aspect.

Strengths include comprehensive development and evaluation of the UCLA SCTC GIT 2.0. Then, we validated the reflux scale vs. physician diagnosed GERD. One limitation of our study was the relatively small number of patients (n=62) from a single Italian clinical centre. Further longitudinal studies are required to assess the predictive validity in clinical practice and its suitability in clinical trials.

In conclusion, we found UCLA-SCTC GIT 2.0 to be an acceptable, reliable and valid tool, useful to assess GIT symptoms in Italian speaking patients with SSc.

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