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Original article

Happiness, quality of life and their determinants among people with systemic sclerosis: a structural equation modelling approach

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Abstract

Background. Patients' objectives and experiences must be core to the study and management of chronic diseases, such as SSc. Although patient-reported outcomes are attracting increasing attention, evaluation of the impact of disease on the overall subjective well-being, equivalent to 'happiness', is remarkably lacking.
Objectives. To examine the determinants of happiness and quality of life in patients with SSc, with emphasis on disease features and personality traits.
Methods. Observational, cross-sectional multicentre study, including 142 patients, with complete data regarding disease activity, disease impact, personality, health-related quality of life (HR-QoL) and happiness. Structural equation modelling was used to evaluate the association between the variables.
Results. The results indicated an acceptable fit of the model to the data. Perceived disease impact had a significant negative direct relation with HR-QoL ($\beta = -0.79$, $P < 0.001$) and with happiness ($\beta = -0.52$, $P < 0.001$). Positive personality traits had a positive relation with happiness ($\beta = 0.36$, $P = 0.002$) and an important indirect association upon QoL ($\beta = 0.43$) and happiness ($\beta = 0.23$). Perceived disease impact is influenced by body image, fatigue and SSc-related disability to a higher degree ($\beta = 0.6$–0.7) than by disease activity ($\beta = 0.28$) or form ($\beta = 0.17$). Impact of disease had a much stronger relation with HR-QoL than with happiness.
Conclusions. The results suggest that treatment strategies targeting not only disease control but also the mitigation of relevant domains of disease impact (body image, fatigue, global disability) may be important to improve patients' experience of the disease. The reinforcement of resilience factors, such as positive psychological traits, may also play a contributory role towards better patient outcomes.

Key words: systemic sclerosis, happiness, personality traits, quality of life, outcome assessment (health care)

Rheumatology key messages

- To optimize quality of life and happiness, body image and fatigue should be addressed.
- 'Positive' personality traits influence perceived impact of the disease and are associated with greater levels of happiness.
- Strategies to improve bodily and subjective well-being should be considered in patient-centered care of SSc.

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CLINICAL SCIENCE

Introduction

SSc is a heterogeneous and challenging disease that can lead to a substantial decrease in quality of life (QoL) through physical, emotional and social impacts [1, 2]. SSc patients have to cope with an often progressive and disabling disease characterized by pain, fatigue, skin ulcers and the persistent threat of an unpredictable course punctuated by threatful complications such as shortness of breath, dysphagia, heart disease and premature death. Daily experience of the disease includes the burden imposed by body image changes upon self-esteem and social interactions [3–5]. Patients with SSc have poorer health-related QoL (HR-QoL) compared with the general population [6–8]. They report greater impairments in mental health and poorer perception of general health than patients with RA and SLE (after adjustments for age, gender, disease duration, comorbidities and disease activity) [8].

Thus, preserving and improving HR-QoL emerges as a crucial focus of management, but remains a difficult clinical challenge. This can only be achieved through considerate integration of patient-reported outcomes into clinical practice and research. However, so far the evaluation of HR-QoL has been limited to negative outcomes such as physical disability, functioning, depression, anxiety and distress [7–11].

Over the past decade, happiness has become a major topic not only in psychology, but also in public policy, economics and health [12]. The conceptualization of happiness is based mainly in three dimensions of well-being: affective (positive feelings and mood states such as happiness, joy and elation predominate over negative ones), eudaimonic (realization of personal potential and fulfilment of life goals) and subjective (global appraisal of how satisfied people are with their quality of life) well-being [12–15]. The potential associations between happiness and health are multiple and bidirectional. Good health is certainly expected to contribute to happiness, but this needs to be considered in the context of other factors, with emphasis on personality [12]. Research has demonstrated that personality traits have a major influence on how patients perceive their disease and their ability to cope with it, which in turn impacts the way they assess their health-related QoL [16–19].

Conversely, happiness has been argued to exert a positive impact upon physical health and longevity, through its influence on perceived disease impact, health-related behaviours and even biological processes [12, 20, 21]. Regarding the latter, consistent research has found an association between greater well-being and lower levels of inflammatory markers, such as CRP and IL-6 [22–24], while depression and anxiety have the opposite effect [25–27]. This link may be particularly relevant for SSc given the known involvement of CRP in disease activity and impaired lung function [12, 21]. Also, interventions targeted to increase well-being may have an impact upon inflammatory markers [28], but so far evidence does not allow firm conclusions regarding causal relationships between inflammation and well-being or happiness.

Immunological processes may have consequences upon happiness and depression [29], and these may in turn change the progress and therapeutic response of immune-mediated diseases [30, 31].

Overall subjective well-being/happiness can, and probably should, be conceived as the ultimate target of medical care, together with prolongation of life. Many physicians and members of the community at large may even question the value of the second target in the absence of the first.

Recently, our research group highlighted that treatment strategies focussed solely on the control of disease activity can be expected to have only limited impact on QoL and happiness. Therefore, focus on improvement of the disease impact [RA impact of disease (RAID)] domains (i.e. pain, fatigue, emotional, functional, physical well-being, sleep and coping) should be given to optimize QoL and happiness in RA [19]. We have also shown that a 'positive' personality seems to play a pivotal role in these relations, supporting the need for an holistic assessment and approach, if 'happiness' is to be considered among the goals of care [13].

Studies on subjective well-being/happiness are remarkably lacking in SSc [1]. The underlying drivers need to be further explored, in order to serve the ethical imperative of putting the patients' needs and perspective at the core of the medical management strategy.

The aim of the present study was to evaluate the following hypotheses in patients with SSc:

H1: Disease activity and impact of disease are negatively associated with overall QoL and happiness;
H2: 'Positive' personality traits are related to happiness both directly and indirectly through perceived disease impact.

Methods

Participants and study design

This was a multicentre cross-sectional study carried out in six rheumatology departments in Portugal. Adult patients were included if they had (i) a diagnosis of SSc, according to the Le Roy and/or the ACR/EULAR classification criteria [32, 33], (ii) ability to understand and fill out the questionnaires, (iii) willingness to provide informed signed consent, and (iv) completed all the questionnaires required.

All patients were registered in the Rheumatic Diseases Portuguese Register (Reuma.pt). Patients' demographic, clinical and disease characteristics were collected.

All participants provided informed written consent before the start of study procedures, and the ethical approval was granted by Ethical Review Board at Centro Hospitalar e Universitário de Coimbra (CHUC-033-18).

Questionnaires/measures

Disability was measured as the mean of the five scleroderma-specific visual analogue scale (VAS) (0–10) included in the Scleroderma HAQ, assessing the impact of digestive symptoms, pulmonary symptoms, RP and digital ulcers upon daily activities, with an additional VAS for perceived overall disease severity [34]. This is designated hereafter as scleroderma-global VAS.

HR-QoL was accessed with the EuroQOL five dimensions (EQ-5D) questionnaire, which includes the dimensions mobility, self-care, usual activities, pain/discomfort and anxiety/depression [35]. Each dimension has three levels: no problems, some problems and severe problems. The combination of the five scores leads to an index score between −0.59 and 1.00 [35]. Higher scores indicate a perceived better health status and QoL. The EQ-5D questionnaire presents acceptable internal reliability for the current study ($\alpha = 0.75$) [36].

Fatigue was assessed using the Functional assessment of Chronic Illness Therapy (FACIT), which consists of 13 items assessing tiredness, weakness and difficulty conducting everyday activities due to fatigue in the past 7 days. Items are scored on a 5-point scale (0 = not at all, 4 = very much). All items, except items 7 and 8, are reverse-scored before being summed to obtain a total score (range 0–52). Higher scores reflect less fatigue [37]. The FACIT questionnaire presents excellent internal reliability for the current study ($\alpha = 0.94$) [36].

Satisfaction with body image was assessed using the Body Image Scale (BIS), which includes 10 items assessing affective (e.g. feeling self-conscious), behavioural (e.g. difficulty at looking at naked body) and cognitive (e.g. satisfaction with appearance) dimensions of body image, in the past 7 days [38]. A total score can be computed by summing all items (range 0–30). Higher scores indicate increasing distress or more body image concerns. The BIS questionnaire presents good internal reliability for the current study ($\alpha = 0.88$) [36].

Social support was assessed using the Satisfaction with Social Support Scale. This scale evaluates four dimensions: satisfaction with friendships, intimacy, family satisfaction and social activities. The total score ranges from 15 and 75. Higher scores correspond to greater satisfaction with social support [39]. The Satisfaction with Social Support Scale questionnaire presents good internal reliability for the current study ($\alpha = 0.88$) [36].

Personality was assessed by the Ten-Item Personality Inventory (TIPI), a brief measure of the Big-Five personality dimensions, each being scored as the mean of two items (7-point Likert scale) addressing extraversion, agreeableness, conscientiousness, emotional stability and openness to experience [40]. Higher scores indicate a stronger expression of the respective trait. We designated the latent higher order factor derived from TIPI as a 'positive' personality to represent the predominantly adaptive nature of the represented dimensions. We recognize that the term 'positive' is questionable, especially in the extremes of expression of certain traits, such as conscientiousness. The TIPI questionnaire presents good internal reliability for the current study ($\alpha = 0.81$) [36].

Happiness was assessed through the Subjective Happiness Scale (SHS) a 4-item measure (7-point Likert scale) [41]. A higher mean score indicates more intense perception of a 'happy life'. The SHS questionnaire presents good internal reliability for the current study in its 3-item version ($\alpha_{3\ items} = 0.83$ *vs* $\alpha_{4\ items} = 0.54$) [36].

Disease activity was assessed by the European Scleroderma Trials and Research group (EUSTAR) score, which is a weighted 10-point activity score composed by: Δ-skin = 1.5 (Δ = patient assessed worsening during the previous month), modified Rodnan skin score (mRss) >18 = 1.5, digital ulcers = 1.5, tendon friction rubs = 2.25, CRP >1 mg/dl = 2.25 and diffusing capacity of the lung for CO (DLCO) % predicted <70% = 1.0 [42]. A score ≥2.5 indicates an active disease.

Subset disease form was classified into limited and diffuse forms, defined according to Le Roy classification [32].

Data analysis

The software SPSS, v.23 (IBM, Armonk, NY, USA) was used to perform descriptive and correlational analyses. The assumptions of normality were not fulfilled [43]. Spearman correlation analyses were conducted to examine the associations between disease activity, disease impact, personality, HR-QoL and happiness, and interpreted according to Cohen's benchmark values as small (<0.30), medium (0.30–0.50) or large (>0.50) [44].

Structural equation modelling (latent variable structural model) was used to estimate the association between these variables and performed with STATA 15.0. Given that the assumptions of normality were not fulfilled [45], we used a maximum likelihood estimation with Satorra–Bentler correction. Variance inflation factor values were <5 for all variables included in the model, excluding multicollinearity as a major issue. A set of goodness of fit indices were used to test the plausibility of the model: (i) the χ^2 (χ^2) and normed χ^2 (χ^2/df), (ii) the comparative fit index (CFI), (iii) the Tucker–Lewis Index (TLI) and (iv) the root mean square error of approximation (RMSEA). A good fit of the models was assumed when: the ratio of χ^2 to its degrees of freedom was <3.0; CFI and TLI were >0.90; RMSEA values <0.06 were considered ideal, and values between 0.08 and 0.10 were considered acceptable [46].

In the initial tested models, several steps were taken that led to the final proposed model:

i. Initially a theoretical model was proposed based on clinical plausibility and available literature (impact disease→happiness and QoL; positive personality→happiness and impact disease; EUSTAR activity score and disease form→impact disease; and, lastly, QoL→happiness).
ii. Other paths with theoretical and clinical plausibility were tested (such as disease form→EUSTAR activity

score). Through the modification indexes we realized that these variables were correlated but not directly related.

iii. Following the analysis of the modification indices, three covariances were entered in the measurement model due to clinical plausibility and theoretical (conceptual/semantic overlap) justification (body image–social support; agreeableness–conscientiousness; item 1 and item 2 of the SHS).

iv. mRss was excluded from the model due to its collinearity with body image and because it ultimately had an overall negative effect on the fit of the model.

v. The fourth question of SHS showed a totally discordant profile vis-à-vis the other three, and was therefore excluded from the happiness construct, as technically recommended [46]. This issue has been reported in a previous study [19].

After we applied these steps, the initially proposed model was readjusted accordingly. Statistically significant differences were assumed for direct, indirect and total effects with $P < 0.05$.

Results

Patient characteristics

A total of 142 patients were included with a median (interquartile range) age of 60.6 (17.4) years, 92% women and disease duration since diagnosis of 6.9 (9.1) years. Approximately 75% had the limited cutaneous form of disease. Eighteen patients (12.7%) had an active disease (i.e. EUSTAR activity score ≥2.5). Baseline sociodemographic, clinical characteristics and patient-reported outcomes are detailed in Table 1.

Correlation coefficients

Correlation coefficients are presented in Table 2. HR-QoL was found to be strongly and inversely correlated with overall disease severity VAS, fatigue and body image, and presented medium correlations with scleroderma-global VAS. The personality traits extraversion, emotional stability and openness to experience presented small to medium positive correlations with HR-QoL.

Happiness (3-items score) presented medium positive correlations with HR-QoL, extraversion and emotional stability, and a small positive correlation with openness to experience.

Finally, DAS showed small to medium positive association with perceived disease impact—namely with scleroderma-global VAS, pulmonary VAS, overall disease severity VAS and body image. In addition, the DAS showed a small negative correlation with QoL and no significant correlation with any personality trait.

Structural equation modelling

Five models were run separately for each of the personality traits as preliminary analyses to the final model. All traits were significantly associated with impact of disease and happiness, except for trait consciousness.

Regarding the final model, the results obtained in the hypothesized model indicated an acceptable fit to the data [$\chi^2_{(82)} = 133.74$, $\chi^2/df = 1.63$, $P < 0.005$; CFI = 0.91; TLI = 0.90; RMSEA = 0.06, $P = 0.05$, 90% CI = 0.04–0.08] and supported all driving hypotheses. The direct path

Table 1 Baseline characteristics of the 142 patients with SSc included in the study

Socio-demographic	
Age, years	60.6 (17.4)
Women, *n* (%)	131 (92.3)
Marital status, %	
Married/single/divorced	73.4/19.5/7.1
Education, years	9 (8)
Employment status, *n* (%)	
Retired, employed, unemployed	49.2/44.3/6.5
Disease characteristics	
SSc duration since first symptoms (years)	10.9 (10.1)
SSc duration since diagnosis (years)	6.9 (9.1)
Limited/diffuse, *n* (%)	107 (75.4)/35 (24.6)
mRSS (0–51)	4.0 (9.0)
RP, *n* (%)	133 (93.7)
Skin thickening, *n* (%)	112 (78.9)
Telangiectasia, *n* (%)	68 (47.9)
History of digital ulcers, *n* (%)	57 (40.1)
Gastrointestinal involvement,[a] *n* (%)	51 (35.9)
Pulmonary involvement (CT or X-ray),[b] *n* (%)	41 (28.9)
Artralgias/arthritis, *n* (%)	45 (31.7)
Calcinosis, *n* (%)	18 (12.7)
Myalgias/myositis, *n* (%)	5 (3.5)
ACA positive, *n* (%)	85 (59.9)
Anti-Scl70 positive, *n* (%)	56 (39.0)
Current treatment with DMARDs, *n* (%)	41 (28.9)
Patient-reported outcomes	
Scleroderma-global VAS (0–10)	3.4 (3.8)
Digestive VAS	2.0 (5.0)
Pulmonary VAS	1.0 (5.0)
Raynaud VAS	5.0 (7.0)
Digital ulcers VAS	0.0 (7.0)
Overall disease severity VAS	5.3 (5.0)
EQ-5D (−0.59–1.0)	0.49 (0.38)
FACIT fatigue score (0–52)	34.0 (18.3)
Social perception scale (15–75)	52.0 (17.0)
Subjective happiness scale (1–7)	4.5 (1.5)
Body Image Scale (0–30)	6.0 (13.0)
Ten-Item Personality Inventory (1–7)	
Extraversion	4.0 (3.0)
Agreeableness	6.5 (1.5)
Conscientiousness	6.0 (2.0)
Emotional stability	4 (1.5)
Openness to experience	4.5 (2)
EUSTAR DAS (0–10)	0 (1.5)
Inactive (<2.5) *vs* active (≥2.5), *n* (%)	124 (87.3) *vs* 18 (12.7)

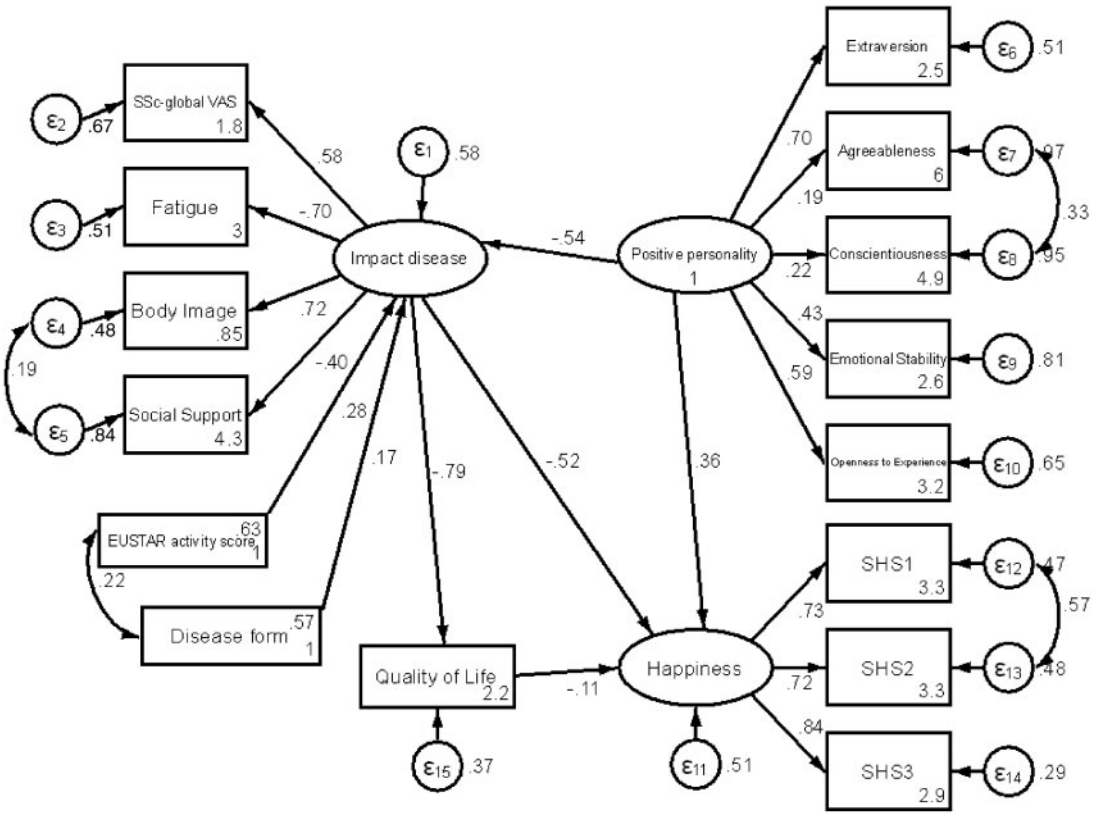
Data are shown as median (interquartile range) unless otherwise stated. [a]Dysphagia and/or heartburn and/or bloating and/or vomiting and/or diarrhoea and/or constipation; [b]Either ground glass or interstitial fibrosis as detected at lung high-resolution CT or X-ray. VAS: visual analogue scale; mRss: modified Rodnan skin score; EQ-5D: EuroQOL five dimensions; FACIT: Functional Assessment of Chronic Illness Therapy-Fatigue; EUSTAR: European Scleroderma Trials and Research group.

Table 2 Spearman correlation coefficients among variables

		1	2	3	4	5	6	7	8	9	10	11	12	13	14	15	16	17	18	19	20	21	22
Impact of disease	SSc global VAS (1)	1.00																					
	Digestive VAS (2)	0.63**	1.00																				
	Pulmonary VAS (3)	0.68**	0.52**	1.00																			
	Raynaud VAS (4)	0.77**	0.34**	0.41**	1.00																		
	Digital ulcers VAS (5)	0.76**	0.32**	0.39**	0.54**	1.00																	
	Overall disease severity VAS (6)	0.68**	0.23**	0.34**	0.50**	0.40**	1.00																
	Fatigue (7)	−0.43**	−0.37**	−0.38**	−0.17*	−0.28**	−0.31**	1.00															
	Body image (8)	0.45**	−0.37**	−0.36**	0.19**	0.31**	0.40**	−0.56**	1.00														
	Social (9)	−0.15	−0.08	−0.12	−0.15	0.04	−0.15	0.22**	−0.17*	1.00													
Disease form (10)		0.29**	0.25**	0.31**	0.22*	0.09	0.22**	−0.09	0.26**	−0.04	1.00												
Quality of life (11)		−0.42**	−0.28**	−0.23**	−0.30**	−0.20**	−0.50**	−0.58**	−0.55**	0.30	−0.11	1.00											
Positive personality	Extraversion (12)	−0.15	−0.01	−0.06	−0.09	−0.12	−0.14	0.21*	−0.25**	0.43**	0.006	0.28**	1.00										
	Agreeableness (13)	−0.08	0.09	−0.08	−0.06	0.08	−0.05	0.15	−0.10	0.10	0.04	−0.03	0.15	1.00									
	Conscientiousness (14)	−0.01	−0.01	−0.05	−0.07	−0.04	0.16	0.15	−0.09*	0.21*	0.01	0.02	0.13	0.36**	1.00								
	Emotional stability (15)	−0.13	−0.16	−0.07	−0.08	0.01	−0.12	0.31**	−0.19*	0.26**	0.03	0.28**	0.25**	0.10	0.10	1.00							
	Openness to experience (16)	−0.09	−0.02	−0.01	−0.08	−0.13	−0.10	0.24**	−0.15	0.18*	0.04	0.22**	0.48**	0.09	0.21*	0.15	1.00						
Happiness	SHS1 (17)	−0.19**	−0.06	−0.07	−0.23**	−0.08	−0.36**	−0.23**	−0.32**	0.43**	−0.15	0.38**	0.25**	0.03	0.001	0.31**	0.18*	1.00					
	SHS2 (18)	−0.19**	−0.06	−0.11	−0.15	−0.08	−0.31**	0.24**	−0.31**	0.36**	−0.11	0.39**	0.22**	−0.02	0.04	0.28**	0.25**	0.79**	1.00				
	SHS3 (19)	−0.28**	−0.18**	−0.10	−0.21	−0.15	−0.30**	0.35**	−0.32**	0.37**	−0.12	0.35**	0.38**	0.15	0.07	0.34**	0.27**	0.61**	0.61**	1.00			
	SHS4 (20)	0.16	0.12	0.09	0.08	0.04	0.17*	−0.21*	0.14	−0.27**	0.03	−0.21*	−0.22**	−0.12	−0.15	−0.23**	−0.05	−0.17*	−0.18*	−0.22**	1.0		
	SHS 3 items score (21)	−0.25	−0.12	−0.11	−0.22**	−0.11	−0.36**	0.31**	−0.36**	0.44**	−0.15	0.42**	0.33**	0.06	0.04	0.35**	0.27**	0.91**	0.90**	0.85**	−0.22**	1.00	
EUSTAR score (22)		0.33**	0.20*	0.31**	0.18*	0.22**	0.31**	−0.19*	0.29**	0.07	0.22**	−0.26**	−0.09	−0.01	0.11	0.10	0.01	−0.08	−0.11	−0.10	0.008	−0.09	1.00

SSc global VAS: scleroderma global visual analogue scale; SHS: Subjective Happiness Scale; EUSTAR score: DAS assessed by European Scleroderma Trials and Research group. *$P < 0.05$; **$P < 0.001$.

FIG. 1 Estimated standardized direct effects for the proposed model



All effects are statistically significant ($P < 0.05$) except for quality of life→happiness. Circles represent latent factors. Squares represent measured variables. Arrows show a hypothesized direct relationship between the two variables. Curved lines with an arrow represent a covariance. Circles with the letter 'E' written in it represent the associated error. SSc-global VAS: scleroderma global Visual Analogue Scale; EUSTAR: European Scleroderma Trials and Research group; SHS: Subjective Happiness Scale.

coefficients for the model are shown in Table 2 and Fig. 1.

H_1—Disease activity and perceived disease impact are negatively associated with overall QoL and happiness.

Disease activity (EUSTAR activity score) and disease form showed a significant positive direct relation with disease impact ($\beta = 0.28$, $P < 0.001$; $\beta = 0.17$, $P = 0.02$, respectively), which in turn showed a significant negative direct relation with HR-QoL ($\beta = -0.79$, $P < 0.001$) (Table 3, Fig. 1).

Several other indirect effects were observed between disease activity, disease form and happiness ($\beta = -0.12$, $P = 0.004$; $\beta = -0.08$, $P = 0.06$, respectively) and HR-QoL ($\beta = -0.22$, $P < 0.001$; $\beta = -0.14$, $P = 0.02$, respectively).

Disease impact had a much stronger negative relation with HR-QoL than with happiness ($\beta = -0.52$, $P = 0.001$). HR-QoL and happiness had no statistically significant relationship.

H_2—'Positive' personality traits are related to happiness both directly and indirectly through perceived disease impact.

'Positive' personality traits had a total effect of 0.59 on happiness, through a direct effect of $\beta = 0.36$ ($P = 0.002$) and indirect effect of $\beta = 0.23$ ($P = 0.006$) through the disease impact. The model also shows a direct negative relation between 'positive' personality and disease impact ($\beta = -0.54$, $P < 0.001$).

Importantly, disease impact had a total effect of -0.44 on happiness, of which $\beta = 0.08$ ($P = 0.05$) was an indirect effect through HR-QoL (Table 3, Fig. 1).

In a model where an independent variable has an effect on a dependent variable, but part of the effect is assumed to occur through a third variable, the 'indirect effect' represents the portion of the 'total effect' of the independent variable on the dependent variable that is explained by this third variable. The remainder of the total effect that is unexplained by the third variable is referred to as the 'direct effect', representing the effect of the independent variable on the dependent variable controlling for the third variable.

Therefore, the 'total effect' is equal to the sum of the direct and indirect (in linear systems) [47, 48].

Table 3 Direct, indirect and total effects between parameters

	Unstandardized effects	Standardized effects	Standard Error	Significance level
Direct effects				
Impact of disease←positive personality	−0.71	−0.54	0.08	<0.001
Impact of disease←EUSTAR activity score	0.34	0.28	0.07	<0.001
Impact of disease←disease form	0.64	0.17	0.08	0.02
Happiness←impact of disease	−0.37	−0.52	0.20	0.001
Happiness←positive personality	0.33	0.36	0.12	0.002
Happiness←quality of life	−0.47	−0.11	0.15	0.49
Extraversion←positive personality	1.00	0.70	0.08	<0.001
Agreeableness←positive personality	0.16	0.19	0.08	0.03
Conscientiousness←positive personality	0.22	0.22	0.10	0.03
Emotional stability←positive personality	0.50	0.43	0.09	<0.001
Openness to experience←positive personality	0.74	0.59	0.08	<0.001
Scleroderma global VAS←impact of disease	1.00	0.58	0.07	<0.001
Fatigue←impact of disease	−5.04	−0.70	0.05	<0.001
Body image←impact of disease	3.29	0.72	0.04	<0.001
Social←impact of disease	−3.12	−0.40	0.08	<0.001
Quality of life←impact of disease	−0.13	−0.79	0.04	<0.001
SHS1←happiness	1.00	0.73	0.07	<0.001
SHS2←happiness	0.99	0.72	0.07	<0.001
SHS3←happiness	1.30	0.84	0.07	<0.001
Indirect effects				
Happiness←impact of disease	0.06	0.08	0.09	0.50
Happiness←EUSTAR activity score	−0.10	−0.12	0.04	0.004
Happiness←disease form	−0.19	−0.08	0.11	0.06
Happiness←positive personality	0.22	0.23	0.08	0.006
Quality of life←EUSTAR activity score	−0.04	−0.22	0.01	<0.001
Quality of life←disease form	−0.08	−0.14	0.04	0.02
Quality of life←positive personality	0.09	0.43	0.02	<0.001
Total effects[a]				
Happiness←impact of disease	−0.31	−0.44	0.09	0.001
Happiness←positive personality	0.54	0.59	0.14	<0.001

Unstandardized direct effects come directly out of the estimation procedure. Due to the metric differences of the instruments. in this case. standardized direct effects should be preferred to indicate the strength of the associations (magnitude between −1.0 and +1.0). Higher absolute values indicate a stronger (positive or negative) association. [a]The remaining total effects are isolated direct or indirect effects and are therefore not repeated. VAS: visual analogue scale; EUSTAR: European Scleroderma Trials and Research group; SHS: Subjective Happiness Scale.

Discussion

This multicentre study provides an innovative model elucidating the relations between disease form, activity and impact, personality traits, HR-QoL and happiness in people with SSc.

Overall, the results indicate that disease impact and positive personality traits are related to QoL and happiness in patients with SSc. Our model also shows HR-QoL is essentially related to disease impact. Happiness has a negative relationship with perceived impact of disease that is strongly buffered by indirect and direct effects of 'positive' personality.

Surprisingly, happiness is almost unrelated to HR-QoL. These two concepts, which we expected to be related, seem to represent distinct dimensions in patients' perspectives, probably with HR-QoL related to the ability to perform activities, and happiness with the satisfaction of doing things, i.e. enjoyment of life as a whole.

Disease impact is predominantly related with body image, fatigue and disability, with smaller contributions of satisfaction with social support and, to a lower extent, disease activity and disease form.

The importance of body image as a factor of disease impact certainly deserves to be highlighted. The association between these two constructs has already been underlined by previous studies and attributed not only to the debilitating and stigmatizing physical changes, but also to a marked dissatisfaction with self-image and a fractured identity [49–51]. SSc patients have higher levels of body image dissatisfaction, and fear of negative evaluation as well as lower self-rated attractiveness [50]. The disfigurement and stiffness of the skin may lead to difficulty in social and sexual interactions [52], social anxiety and avoidance of social situations [50], and contribute to depression [50, 53].

Previous research also showed a significant association between higher mRSS and reduced physical and mental component scores of the 36-item Short Form (SF-36) [8]. The observations highlight the importance of body image in SSc and emphasize the need for studies to better understand the underlying mechanisms and

potential therapeutic interventions. Conversely, skin improvement is paralleled by increases in QoL (SF-36 Physical Component Summary score) and function in patients with early diffuse SSc patients [54].

The important impact of fatigue upon HR-QoL is in agreement with previous reports [55, 56]. Fatigue has been more strongly and regularly associated with psychosocial disorders, with emphasis on depression, than with organ involvement [50]. In fact, data relating fatigue and disease activity are very contradictory [57, 58].

The scleroderma-global VAS directly captures the impact of several organ manifestations upon daily activities, as perceived by the patient [59]. It is interesting to note that this load is higher than that associated with the measure of disease activity by the EUSTAR score. These findings underline the differences between the patients' and the physician/researcher's perspective, and highlight the need to understand the views and needs of patients to optimize communication and share decision-making about treatment strategies.

Positive personality traits are the fourth most influential factor on perceived disease impact in our model, preceding social support, and disease activity and form. The importance of psychological factors in this context has been scarcely investigated except for depression [9, 53]. Depression has been associated with mRSS, oesophageal involvement, digital ulcers and self-rated disability (i.e. HAQ). Anti-depressants have also shown to improve functional outcome [9, 53]. Positive personality traits are a powerful protection against depression and anxiety (which may add to the already burdening impact of SSc), and this may be one of the multiple pathways by which personality influences the perceived impact of disease [13, 18].

The potential beneficial effects of 'positive' personality traits in our study are encouraging. They seem to mitigate the perceived impact of disease, thus potentially reducing the resultant suffering. This endows these traits with an indirect positive effect upon QoL and happiness, which is reinforced by significant direct effects in the case of happiness. The most relevant personality traits are extraversion and openness to experience.

Although personality traits are considered stable in adult life [60, 61], there is evidence that disease conditions can modulate the expression of naturally occurring traits [13]. Interestingly, 'positive' personality traits can influence the adoption of positive coping strategies [62] and the levels of subjective well-being [13]. More importantly, several psychological techniques, with emphasis in positive psychology interventions, mindfulness [63], savouring positive experiences [64], social activity and support groups [65], can demonstrably reinforce the expression of positive, adaptive traits and thus foster their potential benefits upon the patients' experience of the disease and their subjective well-being [66, 67]. Psychotherapy might also mitigate the decisive negative influence of body image in this context [68]. This may be particulary important for patients faced with profound losses in terms of their identity, family and social roles [49]. This underlines the need to consider psychological assessment and intervention in the holistic management of patients with SSc.

Our findings were, overall, similar to those found in a previous similar model in people with RA [19]. The effect of perceived impact of disease on QoL was similar in both conditions (SSc: $\beta = -0.79$, $P < 0.001$ *vs* RA: $\beta = -0.70$, $P < 0.001$). However, the effect of disease impact upon happiness was higher in SSc ($\beta = -0.52$, $P = 0.001$) than in RA ($\beta = -0.17$, $P = 0.02$). We believe that many specific features of SSc probably contribute to this, including the unpredictable disease course, the lack of effective therapies, and the impact on body image, self-esteem and social interaction, among others [9, 49, 64]. These factors certainly deserve further investigation as a means to try to minimize their impact upon happiness.

While interpreting our results, some limitations should be considered. The sample size and the imbalance in the number of patients with limited and diffuse forms impose statistical limitations and make it impossible to carry out group comparisons. The assessment of happiness was based on a unique and brief questionnaire, leaving out subcomponent analysis. This could be tackled in future studies by including additional relevant measures of positive affect and eudaimonic well-being, for example. The observational and cross-sectional nature of the study limits any interpretation or conclusion regarding the presence and direction of causal links among variables. Thus, future studies should replicate these findings using longitudinal designs that allow for the establishment of causality. Also, and despite the clinical importance of including subjective assessment and perspective of the patient, the use of self-report measures implies some limitations and potential biases that should be considered when interpreting the findings. We have not considered comorbidities and other factors (genetic, cultural, education, work, etc.) that may influence subjective well-being [12]. It is also true that our conclusions do not necessarily apply to other cultures. Regarding the measurement of personality, our decision to have all traits represented in a single 'positive and adaptive' dimension is questionable in the light of research on personality, which currently debates the validity of superordinate personality factors [69]. Although we cannot completely rule out that our 'positive personality' factor does not reflect a tendency towards positive evaluation and responding rather than a 'predominantly adaptive nature of the represented dimensions' [69], the results of the preliminary analyses using each trait separately showed that the effect is not general. Not all traits contribute to the measures of interest, as evidenced by the lack of association between consciousness and impact of disease and happiness. Preliminary analyses also showed that the effect of the general factor is not simply and especially linked to a particular trait, as for example neuroticism.

Conversely, this study adopted a sound and comprehensive methodology in many aspects: our construct of

perceived disease impact combines a wide scope of domains of interest for people with SSc, such as body image [49, 51], fatigue [55], scleroderma-global VAS [59] and social support [65]. Many of these domains, as well as personality traits, have seldomly been addressed in SSc. Finally, it represents a novel effort to widen the assessment of disease impact and a relevant contribution towards the ethical imperative of promoting person-centered care.

In summary, our results indicate that body image and fatigue are major domains for perceived disease impact and that a 'positive personality' has a considerable influence on how patients perceive their disease and manage their overall subjective well-being.

These findings highlight that treatment strategies in SSc should not only target disease control but also consider distinct interventions to mitigate all domains of perceived disease impact. We need to gain a better understanding of fatigue in SSc and how to treat it, but also need to use available resources, such as self-management and physical exercise programs [70, 71]. We must pay more considerate and committed attention to the impact of body image, and develop better and novel ways of improving it [72–74]. Psychological interventions hold great promise as a mean to enhance patients' resilience, reduce the impact of disease and optimize their ability to live full and fulfilling lives. We cannot imagine higher or more noble objectives for the care of patients with SSc.

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Data availability statement

Available upon request.

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