RHEUMATOLOGY

Rheumatology 2022;61:1476–1486
doi:10.1093/rheumatology/keab537
Advance Access publication 14 July 2021

Original article

Evidence and consensus-based recommendations for non-pharmacological treatment of fatigue, hand function loss, Raynaud's phenomenon and digital ulcers in patients with systemic sclerosis

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Abstract

Objective. SSc is a complex CTD affecting mental and physical health. Fatigue, hand function loss, and RP are the most prevalent disease-specific symptoms of systemic sclerosis. This study aimed to develop consensus and evidence-based recommendations for non-pharmacological treatment of these symptoms.

Methods. A multidisciplinary task force was installed comprising 20 Dutch experts. After agreeing on the method for formulating the recommendations, clinically relevant questions about patient education and treatments were inventoried. During a face-to-face task force meeting, draft recommendations were generated through a systematically structured discussion, following the nominal group technique. To support the recommendations, an extensive literature search was conducted in MEDLINE and six other databases until September 2020, and 20 key systematic reviews, randomized controlled trials, and published recommendations were selected. Moreover, 13 Dutch medical specialists were consulted on non-pharmacological advice regarding RP and digital ulcers. For each recommendation, the level of evidence and the level of agreement was determined.

Results. Forty-one evidence and consensus-based recommendations were developed, and 34, concerning treatments and patient education of fatigue, hand function loss, and RP/digital ulcers-related problems, were approved by the task force.

Conclusions. These 34 recommendations provide guidance on non-pharmacological treatment of three of the most frequently described symptoms in patients with systemic sclerosis. The proposed recommendations can guide referrals to health professionals, inform the content of non-pharmacological interventions, and can be used in the development of national and international postgraduate educational offerings.

Key words: SSc, fatigue, hand function loss, RP, digital ulcers

CLINICAL SCIENCE

Introduction

SSc is a complex, chronic and incurable CTD characterized by diffuse microangiopathy and immune dysregulation, ultimately leading to widespread skin and internal

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Submitted 15 April 2021; accepted 25 June 2021

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Rheumatology key messages

- We developed consensus and evidence-based recommendations for non-pharmacological treatment of systemic sclerosis-induced fatigue, hand function loss, and Raynaud's phenomenon/digital ulcers.
- Our recommendations provide direction for targeted referrals and treatments, tailored to the needs of the systemic sclerosis patient.

organ fibrosis [1]. Its prevalence is estimated to be 23 per 100 000 people [2]. The consequences of this complex disease significantly adversely affect both mental and physical health [3]. Fatigue, hand function loss and RP, which often lead to digital ulcers (DU), are the most prevalent disease-specific symptoms of limited (lcSSc) and diffuse cutaneous SSc (dcSSc) according to SSc patients [4–6]. All three symptoms interfere, to varying degrees, with the performance of everyday tasks and have a major impact on health-related quality of life (HRQoL) [7–10]. Fatigue significantly affects patients' ability to fulfil social roles, RP and DU are associated with significant pain and disability, and hand function loss worsens the ability to perform meaningful activities of daily living [10–15].

In the Netherlands, SSc care delivered by rheumatologists and nurses is offered in hospitals and rehabilitation centres; while care delivered by health professionals like physiotherapists, occupational therapists and psychologists is predominantly delivered in primary care settings. Depending on the nature of the patient's condition, his or her specific needs and the availability of caregivers at an institution or in the area, delivery of care includes, in addition to treatment by medical specialists, continuous or intermittent involvement of health professionals (HPRs) [16]. HPRs from different professions can be involved in the non-pharmacological treatment of patients with SSc-induced fatigue, hand function loss and RP/DU. In addition to rheumatologists and specialized nurses, there is a role for physiotherapists, occupational therapists, psychologists, dieticians and social workers. In clinical practice, the HPR treatment offer varies, as well as the content of the treatments, and there is little evidence available regarding HPR treatment for SSc-induced fatigue, hand function loss and RP/DU thus far [17–19]. Although HPRs offer numerous treatments to satisfy the unmet care needs of patients with SSc, and these patients are satisfied with the content and results of HPR treatments, fatigue, hand function loss and RP are uncommon reasons for referral to HPRs [18, 20, 21]. In a previous study, we found that rheumatologists are reluctant to refer their patients to HPRs due to a poor overview of HPR treatment options and a lack of published evidence [22]. Existing SSc guidelines and recommendations do not include recommendations regarding non-pharmacological care or only superficially include them. Specific recommendations on non-pharmacological treatment approaches for patients with SSc are not yet available [23].

HPR recommendations not only could support HPRs in SSc treatment, but also could provide clinicians with guidance on timely referrals and access to adequate care for patients with SSc, fatigue, hand function loss and RP/DU. To address this need, this study aimed to develop HPR recommendations for the management and treatment of fatigue, hand function loss and RP/DU in patients with SSc. A multidisciplinary task force has been assembled to develop these recommendations based on evidence and consensus. These recommendations are targeted at all HPRs in the field of non-pharmacological SSc care and are potentially relevant to key stakeholders, namely SSc patients, as well as their patient organizations, rheumatologists and other (medical) care providers.

Methods

Design

We developed the aforementioned recommendations based on the standardized operating procedures for developing practice recommendations of the EULAR [24, 25]. Ethical approval for the face-to-face meeting was obtained by the Institutional Review Board of the Radboud University Medical Center, Nijmegen (approval number, 2019: 5868). The AGREE II-instrument was used to structure this manuscript [26].

Task force

A task force was convened to reach consensus on the recommendations based on clinical expertise, discussion and a literature review. It was led by two convenors, Cornelia (Els) van der Ende (E.E) . (researcher/physiotherapist) and J.K.S. (researcher/OT) and composed of seven Dutch SSc patient representatives, including representatives of the three Dutch patient organizations, who live in different disease stages. These representatives are experienced with fatigue, hand function loss or RP/DU, and underwent non-pharmacological treatments. Selection of patient representatives followed the EULAR recommendations for the inclusion of patient representatives in scientific projects [27]. Moreover, 13 experienced professionals from leading centres of expertise involved in SSc care in the Netherlands were included; among them, there were three rheumatologists, one internist/clinical immunologist, two physiotherapists, one occupational therapist, two psychologists, one dietician, one dental hygienist, one specialized

nurse and one social worker. In the selection of medical and HPR experts, attention was paid not only to their expertise in the treatment of patients with SSc and their work setting, but also to a good geographical distribution across the Netherlands. In addition, care was taken to achieve a reflection of the disciplines involved in the multidisciplinary treatment of patients with SSc [28]. Three mail rounds, two telephone meetings, and one face-to-face task force meeting took place between May 2019 and December 2020.

The development of the recommendations comprised four phases

Phase 1: Formulation of research questions for education and treatment of fatigue, hand function loss and RP/DU

During the first telephone meeting, the task force agreed on the method for formulating the recommendations based on the standardized operating procedures for developing practice recommendations of EULAR. Clinically relevant questions on patient education and non-pharmacological treatments were inventoried by email and summarized by a convenor (E.E .). Based on this inventory, draft research questions were developed by both convenors (E.E. and J.St.). In the second e-mail round, task force members provided feedback on the draft research questions. Through the discussion and refinement of concept research questions, definitive research questions were established by both convenors (E.E. and J.St.).

Phase 2: Development of statements for draft recommendations

During the face-to-face task force meeting, statements for draft recommendations were generated, collected and selected through a systematically structured discussion with the task force members, following the nominal group technique in two parallel groups with a balanced distribution of patient representatives and professionals. The nominal group technique was chosen as a formal consensus development method because it encourages idea generation and problem solving in a structured and balanced group process, and is known to support the development of clinical treatment guidelines for several diseases in a highly structured manner [29–31].

Phase 3: Development of draft recommendations with level of evidence

Based on the collected task force meeting statements, draft recommendations were developed by both convenors (J.St.+E.E.). To determine the level of evidence for the draft recommendations, a literature search was performed. The PubMed/MEDLINE, Embase, CINAHL, PsycINFO, Cochrane Library and Web of Science databases were searched for key systematic reviews (SRs) and randomized controlled trials (RCTs) published after SRs that investigated the effectiveness of interventions targeting adults with SSc between January 1985 and September 2020. If no SRs or RCTs were available, international clinical practice guidelines or recommendations were consulted. According to the agreed method to answer research questions:

i. the literature search for ‘fatigue’ was expanded to include interventions for SLE and RA;
ii. the literature search for ‘joint protection’ (hand function loss) was expanded to also include interventions for RA and OA;
iii. the literature search for ‘RP’ was expanded to include interventions for primary RP; and
iv. because of the lack of evidence, 13 medical specialists were consulted about non-pharmacological advises regarding DU.

For every research question, the found publications were screened by J.St. and E.E. for eligibility through reading the title and abstract. Potentially relevant articles were identified, and full text articles were evaluated independently by both convenors (J.St. and E.E.) and discussed until an agreement was achieved. Methodological quality and risk of bias in individual studies were assessed according to study level using the adapted second version of A Measurement Tool to Assess systematic Reviews (AMSTAR 2), and the Joanne Briggs Institute critical appraisal checklist for RCTs was used to assess RCTs [32, 33]. Discrepancies in assessments between both convenors were discussed until consensus was reached. The Oxford Centre for Evidence-based Medicine levels of evidence were used to assign levels of evidence for each individual draft recommendation [34].

Phase 4: Determining the level of agreement regarding definitive recommendations

In the fourth and final phase, the level of agreement regarding each draft recommendation was determined by the task force and the 13 involved medical specialists using an individual anonymous voting procedure. A numeric rating scale from 1, which indicates total disagreement, to 10, which indicates total agreement, was used. The mean, SD., median, and range of the level of agreement for each recommendation were calculated. A recommendation was approved when $\geq$70% of the expert group indicated a score of $\geq$7 on the numeric rating scale.

Results

Three research questions were developed during phase 1: (i) Which non-pharmacological advices and interventions are meaningful to treat fatigue in patients with SSc?; (ii) Which non-pharmacological interventions and advices are meaningful to prevent hand function loss and improve hand function in patients with SSc?; and (iii) Which non-pharmacological interventions and advices are meaningful to prevent and/or cure RP and DU in patients with SSc? In the second phase, during the face-to-face taskforce meeting, 103 proposed statements for draft recommendations were collected in discussion of the two parallel groups. Thereafter, in the

third phase, the first author (J.St.) modified and reorganized individual statements according to research questions and removed duplicate statements. This process reduced the number of statements for draft recommendations to 41. Moreover, based on these statements, a gradation using a stepped care approach could be made. Generated draft recommendations subsequently formed the basis of the literature review. Supplementary Table 1 (available at *Rheumatology* online) shows the literature search strategy. A total of 20 articles were included; of these articles, there were ten SRs, seven RCTs, one study with a quasi-experimental design and two guidelines. Nine studies addressed fatigue, eight addressed hand function loss, two addressed RP/DU, and one addressed hand function loss and RP/DU. Supplementary Table S2 (available at *Rheumatology* online) summarizes the included articles with their corresponding quality and risk of bias scoring. Regarding the strength of draft recommendations, eight recommendations were graded as having a strength level I, which indicates the highest level of strength, six as having a strength level II, two as having a strength level III, seven as having a strength level IV, and 18 as having a strength level V, which indicates expert agreement. Supplementary Table S3 (available at *Rheumatology* online) summarizes the draft recommendations with their associated quality scoring and level of evidence.

In the fourth and final phase, 29 of the 33 invited experts, which comprised the task force along with the consulted medical specialists, established the level of agreement for recommendations by voting. There were seven patient representatives, 10 HPRs and 12 medical specialists. Accordingly, 34 final recommendations were approved; 12 were on fatigue, eight were on hand function loss, and 14 were on RP/DU, and 90.4% of the expert group voted with a mean agreement of 8.3 [S.D. 0.6; and mean agreement of patient representatives, 8.5 (S.D. 0.5); HPR, 8.4 (S.D. 0.7); and medical specialists, 8.2 (S.D. 0.6)]. The average level of agreement for the final recommendations ranged from 7.2–9.4. Tables 1, 2 and 3 summarize the developed recommendations with references to the studies used, their level of evidence and their level of agreement.

Seven of the 41 draft recommendations did not meet the approval criteria of an agreement of $\geq$7 in $\geq$70% of the expert group, with an average of 54.7% of the expert group voting with a mean agreement of 6.7. The average level of agreement for disapproved draft recommendations ranged from 6.1–7.4. Table 4 gives an overview of the disapproved draft recommendations with references to the literature used, the level of evidence and the level of agreement.

Discussion

These are the first published recommendations on non-pharmacological interventions to treat the three most frequently reported symptoms of SSc, which are fatigue, hand function loss and RP/DU. These recommendations are based on the best available evidence, and the opinion and experience of patients with SSc in different disease states and experienced professionals from leading centres of expertise in the Netherlands. Thirty-four recommendations were developed; of these recommendations, 12 were on fatigue, eight were on hand function loss, 14 were on RP and DU and six were specifically on DU education.

Overall, 15 (51.7%) of the developed recommendations were based on expert opinions due to a lack of scientific evidence. Some topics addressed in the recommendations have not been investigated yet in previously published high-quality research. Such topics include treatments using assistive technology, the adjustment or alternation of environments, including the work environment, to restore energy in meaningful daily activities and the maintenance of the autonomy and independence of patients with SSc. Researchers should focus on further validating these recommendations, in order to provide SSc care with an even clearer substantiation using evidence-based practice.

We observed an overall slightly more positive view on the draft recommendations by the patient representative group compared with the rest of the expert group. However, a single draft recommendation on patient education for RP/DU, which clarifies the possible importance of a healthy diet with an adequate fat intake, was assessed noticeably more positively by patient representatives (mean agreement 9.0) than by the rest of the expert group (mean agreement of whole expert group, 6.7; HPRs, 6.6; and medical experts, 5.5). Consequently, this draft recommendation was not included in the final recommendations, although patient representatives showed a high acceptance. Further research should be conducted to verify this result. Moreover, six of the seven disapproved draft recommendations (*1 to *6) were excluded through expert agreement, although there is verifiable evidence from the literature indicating that these recommendations are valid. A possible explanation for this could be that the patient information and treatments in the aforementioned literature seemed unfamiliar to some experts because they are not often applied in the Netherlands.

The EULAR recommendations for patient education for people with inflammatory arthritis considered patient education as an integral part of standard care [54]. In this study, contrary to some existing recommendations, content for specific SSc-related patient education has been developed, and corresponding recommendations can contribute to the knowledge base related to multidisciplinary care and inform the content of self-management programs that focus on treating SSc and its consequences. Existing self-management programs for people with rheumatic diseases primarily aim at increasing knowledge, adhering to treatment, improving physical functioning and ensuring a healthy lifestyle. The

Table 1 Recommendations on patient education and treatments for systemic sclerosis patients with fatigue

	Level of evidence	Reference	Level of Agreement (0–10) Mean (s.d.); Median (range)
PATIENT EDUCATION			
Persistent fatigue can have far-reaching consequences for activities of daily living and social participation. It is therefore essential that all patients with SSc who report symptoms of fatigue are properly informed about measures that support self-management skills.			
All SSc patients who report fatigue should receive patient information about the following aspects.			
1. Maintaining good physical condition and regular exercise	I	[35–37]	9.3 (0.8) 9 (8–10)
2. Principles of energy conservation and good sleep hygiene	I	[36, 38]	8.8 (1.0) 9 (7–10)
3. Relaxation exercises	I	[38]	7.5 (1.7) 7 (4–10)
4. A healthy diet	V	n/a	7.9 (1.8) 8 (2–10)
5. The possible link between fatigue and drug side effects	V	n/a	7.2 (2.1) 8 (2–10)
TREATMENTS			
a) Treatments for SSc patients with persistent fatigue and related restrictions in whose activities of daily living and who have an imbalance between mental load and mental resilience			
6. Psychoeducational interventions (individually or in a group) aimed at principles of goal setting, energy conservation, dealing with the social environment and relaxation should be offered to SSc patients with fatigue. These interventions can be performed by a skilled health professional, e.g. a nurse, social worker or occupational therapist.	I	[36, 38, 39]	8.7 (1.1) 9 (7–10)
7. CBT under the supervision of a psychologist should be offered to SSc patients with fatigue, if there are severe impediments to activities of daily living.	I	[38–40]	8.2 (1.2) 8.0 (6–10)
8. Participation in available online and face-to-face courses with fellow patients, provided by trained patient representatives: e.g. ReumaUitgedaagd! (self-management training for people with rheumatism) should be offered to patients with SSc with fatigue.	V	n/a	8 (1.3) 8 (5–10)
9. In order to maintain the ability to work, SSc patients with fatigue should be guided in adapting the work environment or switching to different work by a skilled health professional, e.g. an occupational therapist or social worker.	V	n/a	8.3 (1.2) 8.0 (5–10)
b) Treatments for SSc patients with persistent fatigue where reduced physical resilience plays a role			
10. SSc patients with fatigue should receive support to improve exercise capacity and incorporate more physical activity into daily life with the guidance of health professionals such as physical therapists.	I	[35–38, 41–43]	9.1 (1.0) 9 (7–10)
11. Advice about a healthy diet and preventing malnutrition offered by e.g. a dietician should be offered to SSc patients with fatigue.	V	n/a	7.9 (1.2) 8 (6–10)
c) Multidisciplinary treatments			
12. A multidisciplinary rehabilitation program should be offered to SSc patients with severe fatigue symptoms that lead to problems in several domains of activities of daily living.	V	n/a	8.3 (1.6) 9 (3–10)

Level of evidence (according to the standards of the Oxford Centre for Evidence Based Medicine), Level of Agreement for the recommendations, Numeric Rating Scale from 0 (total disagreement) to 10 (total agreement) reported as mean (range), *n/a* not applicable. CBT: cognitive behavioural therapy.

Table 2 Recommendations on patient education and treatments for systemic sclerosis patients with hand function loss

		Level of evidence	Reference	Level of Agreement (0–10) Mean (s.d.); Median (range)
PATIENT EDUCATION				
Provide patient education and advice to all SSc patients with hand function loss to enhance their knowledge about treatment options and promote self-management. The patient education and advice should be tailored to the individual patient but should contain the following aspects.				
13.	Independently and regularly doing hand exercises to maintain hand mobility and strength.	II	[19, 44–47]	9.1 (0.9) 9.0 (7–10)
14.	A continuous use of hands in activities of daily living to maintain hand functionality.	V	n/a	8.7 (1.3) 9 (5–10)
15.	Avoiding cold and keeping the hands warm.	IV	[48]	9.0 (0.8) 9 (8–10)
16.	Good hand care, for example by moisturizing the skin (especially with lanolin-based products) and wearing protective gloves.	IV	[48]	8.3 (1.2) 8 (5–10)
TREATMENTS				
a) Treatments for SSc patients whose activities of daily living are restricted due to limitations in hand function				
17.	Passive and active hand function exercises to promote hand mobility, functionality and strength, under the guidance of a skilled health professional (e.g. a hand therapist), should be offered to SSc patients who experience restrictions in the performance of daily activities due to hand function loss.	II	[19, 44–47]	8.5 (0.9) 8.0 (7–10)
18.	Learning ergonomic measures under the guidance of a health professional such as an occupational therapist should be offered to SSc patients who experience restrictions in the performance of daily activities due to hand function loss.	I	[45, 49]	8.4 (1.1) 8 (7–10)
19.	The adaption of hobbies and work (including volunteer work) to enable participation in meaningful activities of daily living, under the guidance of a health professional, e.g. a social worker or occupational therapist, should be offered to SSc patients who experience hand function loss.	II	[47]	7.9 (1.2) 8 (5–10)
b) Multidisciplinary treatments:				
20.	A multidisciplinary rehabilitation should be offered to SSc patients with hand disabilities that lead to problems in multiple domains of activities of daily living.	II	[19, 47]	8.0 (1.4) 8 (3–10)

Level of evidence (according to the standards of the Oxford Centre for Evidence Based Medicine), Level of Agreement for the recommendations, Numeric Rating Scale from 0 (total disagreement) to 10 (total agreement) reported as mean (range), *n/a* not applicable.

approaches used were found to be mainly didactic and were mostly instructional, counselling and practical exercises [54]. A deeper understanding of factors that influence self-management may improve self-management outcomes among patients with SSc and may inform treatment options tailored to meet individuals' needs and improve health outcomes and consequently the HRQoL of SSc patients.

Similar to the updated 2017 EULAR recommendations for treating systemic sclerosis, we used supportive evidence extrapolated from studies involving patients with other (rheumatic) conditions, including SLE, RA/OA and primary RP for developing these recommendations [55]. This particularly applies to recommendations focussing on fatigue treatment. This could be seen as a limitation of our study. On the other hand, it is likely that in SSc, non-specific factors contribute to fatigue. Non-specific psycho-social aspects include coping skills, depression, lifestyle considerations, such as physical activity, diet or smoking, and also other contributors, such as comorbid conditions, simultaneous pain or sleep disorders. These non-disease-specific factors are also described in other chronic rheumatologic conditions, such as RA and SLE [56, 57]. Therefore, we assumed that apparently effective non-pharmacological interventions in such diseases should also be considered for SSc patients. For example, behavioural techniques, such as energy conservation and activity stimulation, have shown benefits in several

Table 3 Recommendations on patient education and treatments for systemic sclerosis patients with RP and/or digital ulcers

		Level of Evidence	Reference	Level of Agreement (0–10) Mean (s.d.); Median (range)
PATIENT EDUCATION				
All SSc patients with problems due to Raynaud's phenomenon and/or digital ulcers should receive patient education about the following aspects.				
21.	Quitting smoking.	IV	[50]	9.4 (0.9) 10 (7–10)
22.	Avoiding triggers that can elicit an attack of Raynaud's, such as sudden changes in temperature, drinking large amounts of coffee or energy drinks, and stress.	V	[50]	9.0 (1.2) 9.0 (5–10)
23.	Practical advice for protection against cold and avoiding temperature differences: i. use of special clothing, silver gloves, heated gloves, or heating pads; ii. drying the skin thoroughly after showering or washing hands; iii. (iii) avoiding contact with cold objects (wearing gloves to remove items from the fridge/freezer, using a heated keyboard and mouse).	IV	[50]	8.9 (1.1) 9 (6–10)
24.	Wearing fingertip protection (e.g. thermoplastic material or neoprene) to prevent pain when performing actions that cannot be avoided.	V	n/a	7.8 (1.9) 8 (3–10)
25.	Preventing infections and wounds through good hygiene, avoiding bruising and hazardous work, and using gloves for protection.	V	n/a	8.7 (1.7) 9 (3–10)
26.	Promoting good blood circulation through the use of a stress ball and having sufficient exercise throughout the day.	V	n/a	7.4 (2.3) 8 (1–10)
27.	Avoiding prolonged, static postures.	V	n/a	7.3 (2.2) 8 (1–10)
In SSc patients with digital ulcers, attention should also be paid to the following aspects.				
28.	Hydration of the skin around the ulcers with products based on lanolin, petroleum jelly or cetomacrogol.	IV	[48]	8.0 (1.6) 8 (4–10)
29.	Avoiding frequent exposure to water with aggressive cleaning agents.	IV	[48]	8.1 (1.7) 8 (3–10)
30.	Avoiding finger punctures.	V	n/a	8.0 (1.8) 8 (4–10)
31.	Avoiding manipulation of ulcers (e.g. by squeezing out calcium deposits or cutting away hard skin).	V	n/a	8.3 (1.9) 9 (4–10)
TREATMENTS				
Treatments for SSc patients whose activities of daily living are restricted due to Raynaud's phenomenon and/or digital ulcers.				
32.	Exercise therapy (with an arm bicycle) to promote general blood circulation and support the integration of exercise activities in daily life, guided by a health professional (e.g. a physiotherapist), should be offered to SSc patients with Raynaud's phenomenon/digital ulcers.	III	[51]	7.4 (1.7) 8 (2–10)
33.	In case of vasculopathy of the feet, advice about suitable, non-restrictive footwear (for indoor and outdoor use), by a health professional such as, for example, a podiatrist, should be offered to SSc patients.	V	n/a	7.8 (1.9) 8 (1–10)
34.	Advice on the protection of the fingertips with special gloves or by using adaptive devices, provided by a health professional (e.g. an occupational therapist), should be offered to SSc patients whose activities of daily living are restricted due to Raynaud's phenomenon/digital ulcers.	V	n/a	8.2 (1.7) 9.0 (3–10)

Level of evidence (according to the standards of the Oxford Centre for Evidence Based Medicine), Level of Agreement for the recommendations, Numeric Rating Scale from 0 (total disagreement) to 10 (total agreement) reported as mean (range), *n/a* not applicable.

TABLE 4 Draft recommendations excluded from the final recommendations through determination of the level of agreement

	Level of Evidence	Reference	Level of Agreement (0–10) Mean (S.D.); Median (range)
FATIGUE—Patient education			
*1 The beneficial effect of yoga and tai chi on general physical condition and relaxation.	I	[35, 36]	6.6 (1.7) 7 (4–10)
HAND FUNCTION LOSS—Treatments			
*2 Manual lymph drainage by a skilled health professional such as a physiotherapist, occupational therapist or hand therapist in SSc patients with finger or hand oedema.	II	[19]	6.4 (2.0) 7 (2–10)
*3 Connective tissue massage (possibly in combination with passive and active hand exercises).	II	[19, 52]	6.7 (1.6) 7 (3–10)
*4 Avoid wearing dynamic finger splints for the purpose of reducing contractures of the proximal interphalangeal joints by stretching the connective tissue, due to the potential lack of effect and adverse patient outcomes.	III	[53]	6.1 (2.6) 6 (1–10)
RP AND DIGITAL ULCERS—Patient education			
*5 Avoiding exposure to vibrations, which can adversely affect blood flow.	IV	[50]	7.4 (2.0) 7 (1–10)
*6 The possibly beneficial effect of soda baths (no more than twice a week) to prevent dehydration and cracking.	IV	[48]	6.7 (2.3) 7 (1–10)
*7 The possible importance of a healthy diet with adequate fat intake.	V	n/a	6.7 (2.4) 7 (1–10)

Level of evidence according to the standards of the Oxford Centre for Evidence Based Medicine, Level of Agreement for the recommendations, Numeric Rating Scale from 0 (total disagreement) to 10 (total agreement) reported as mean (range), *n/a* not applicable, *1–7 excluded from the final recommendations.

chronic conditions [57]. Moreover, low-impact aerobic exercises that gradually increase in intensity, duration and frequency may be effective for reducing fatigue as such exercises have demonstrated beneficial effects on RA, SLE and initial positive results in patients with SSc [58]. The possibility of conducting adequately powered, high-quality RCTs involving only patients with SSc is limited due to the rarity and clinical heterogeneity of SSc. As SSc-specific evidence on non-pharmacological interventions is limited, in our opinion the way we developed these recommendations is a valid, second-best and efficient method.

The strengths of this project are the broad participation of patient representatives and professionals and its systematic approach that is based on the standardized operating procedures to combine practice and evidence-based knowledge of EULAR. Therefore, the resulting recommendations can be used for all stakeholders: support HPRs in the treatment of SSc patients, guidance for rheumatologists, and other medical or non-medical care providers on timely referrals, and thus better access to information regarding HPR treatment for SSc patients and their patient organizations. Another strength is the division of recommendations into a stepped care approach including patient education, single or multiple HPR treatments and a multidisciplinary approach. Both the consensus meeting and literature suggested that different disease manifestations require different treatment approaches. All patients diagnosed with SSc need patient education regarding clinical manifestations and possible disease consequences to manage SSc. Moreover, patients with single, non-lethal disease consequences can often receive help through specific, individualized treatments. However, when patients report limitations that cause restrictions in multiple areas of activities of daily living, multidisciplinary treatment with appropriate specialists should be considered.

A possible limitation in this study is that we used a pragmatic literature research approach to answer research questions, and that we refrained from statistical pooling of data of findings of individual RCTs due to the heterogeneity of interventions and outcome measures. As, a consequence, we did not provide information about the magnitude of effects (and thus the clinical relevance of findings). Draft recommendations formulated by the task force in the face-to-face meeting were the main factors deciding whether recommendations were approved. However, in disease settings in which evidence is limited by a small patient sample and the rapid development of the disease, this approach can help to inform the content of HPR interventions and can also be used in the development and/or optimization of

research studies and national postgraduate educational offerings. By performing a thorough literature search on systematic reviews and recently published RCTs we got insight into the (lack of) evidence basis of each individual recommendation. To ensure the high quality of statements, all articles found were assessed for their quality, risk of bias and subsequently the level of evidence. Another potential study limitation might be that, while the literature used originates from the international field of expertise, the expertise of experts involved is probably mainly based on the Dutch health care system. As those roles may vary per country, local adaptations may be needed if the recommendations stated in this study are used in other countries.

Conclusion

The 34 recommendations stated in this study provide guidance on the non-pharmacological management of three of the most frequently described symptoms of SSc. The proposed recommendations can inform the content of non-pharmacological interventions in the Netherlands and can also be used in the development and optimization of national and international postgraduate educational offerings. More research, particularly regarding assistive technology, the adaptation of the patients' (work) environment to restore energy, and self-management strategies to support meaningful daily activities, is needed to enhance the autonomy and independence of patients with SSc.

Acknowledgements

The authors would like to thank J. Auwema, S. Dittmar, J.J.K.C. Welling, T. van Helmond, A Vedder, B. Oosterveld, L. Kwakkenbos, B. van der Swaaij and J. Alberts-Pijnenborg for attending the task force meetings and the face-to-face consensus meeting. We would also like to thank A.E. van der Bijl, F. Bonte-Mineur, H.K.A. Knaapen-Hans, and A.E. Voskuyl for their valuable contribution to the content of recommendations regarding digital ulcers and their participation in the anonymous voting procedure to establish the level of agreement. The authors are grateful to the Dutch Society for Lupus, APS, Scleroderma, and MCTD, Stichting Sclerodermie Nederland, and Scleroderma framed for their collaboration. The authors also acknowledge the Dutch Arthritis Foundation (ReumaNederland) for funding the ARCH initiative.

Funding: This work was supported by the Netherlands Organization for Scientific Research (NWO) PhD program for teachers (grant number 023.006.043). The funding organization played no role in the design of this study, collection, analysis and interpretation of data, or any role in writing the manuscript.

Disclosure statement: The authors have declared no conflicts of interest.

Data availability statement

The datasets used and/or analysed during this study are available from the corresponding author upon reasonable request.

Supplementary data

Supplementary data are available at *Rheumatology* online.

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