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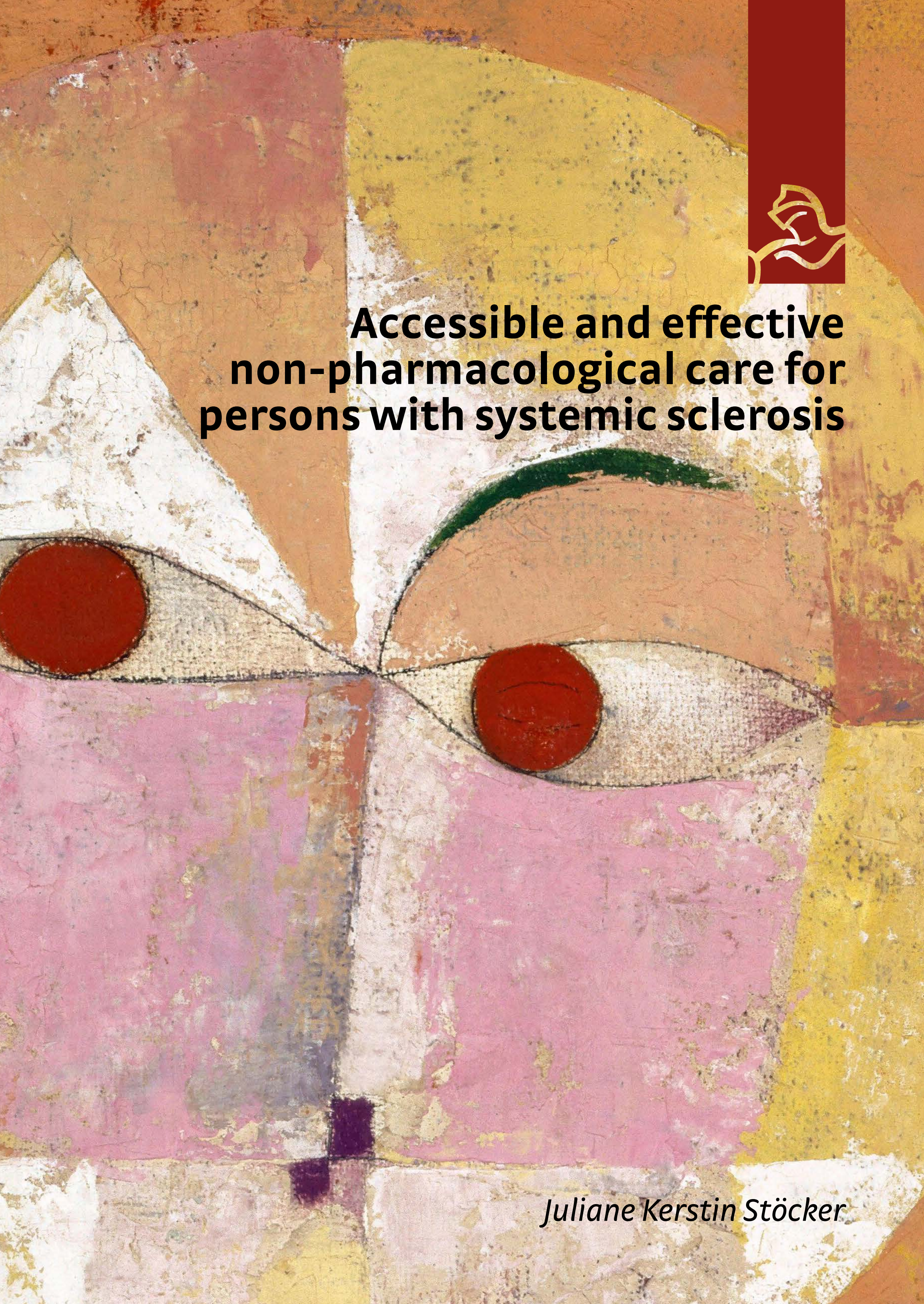

Accessible and effective non-pharmacological care for persons with systemic sclerosis

Juliane Kerstin Stöcker

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Artwork cover:
Senecio (1922) - Paul Klee
Paul Klee (1879-1940) is considered one of the most important visual artists of the 20th century. Klee joined the artist group Der Blaue Reiter, together with Wassily Kandinsky, Gabriele Münster and Franz Marc.

In addition to his artistic work, he taught at the Bauhaus in Weimar and Dessau before becoming a professor at the Düsseldorf Art Academy. In 1933 Klee was forced by the Nazis to leave the art academy because his work, like that of Marc Chagall, Wassily Kandinsky and Piet Mondrian, was considered 'entartete Kunst'. He died in 1940 as a result of a connective tissue disease that was still a mystery at the time, and which after his death was diagnosed as progressive systemic sclerosis. Paul Klee's death anniversary (June 29) marks World Scleroderma Day. A day to recognize the courage of those living with systemic sclerosis, and to demand equal treatment and equal care for persons with systemic sclerosis.

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For reasons of consistency, some terms have been standardized throughout the text.
As a consequence the text may differ from the published articles.

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Accessible and effective non-pharmacological care for persons with systemic sclerosis

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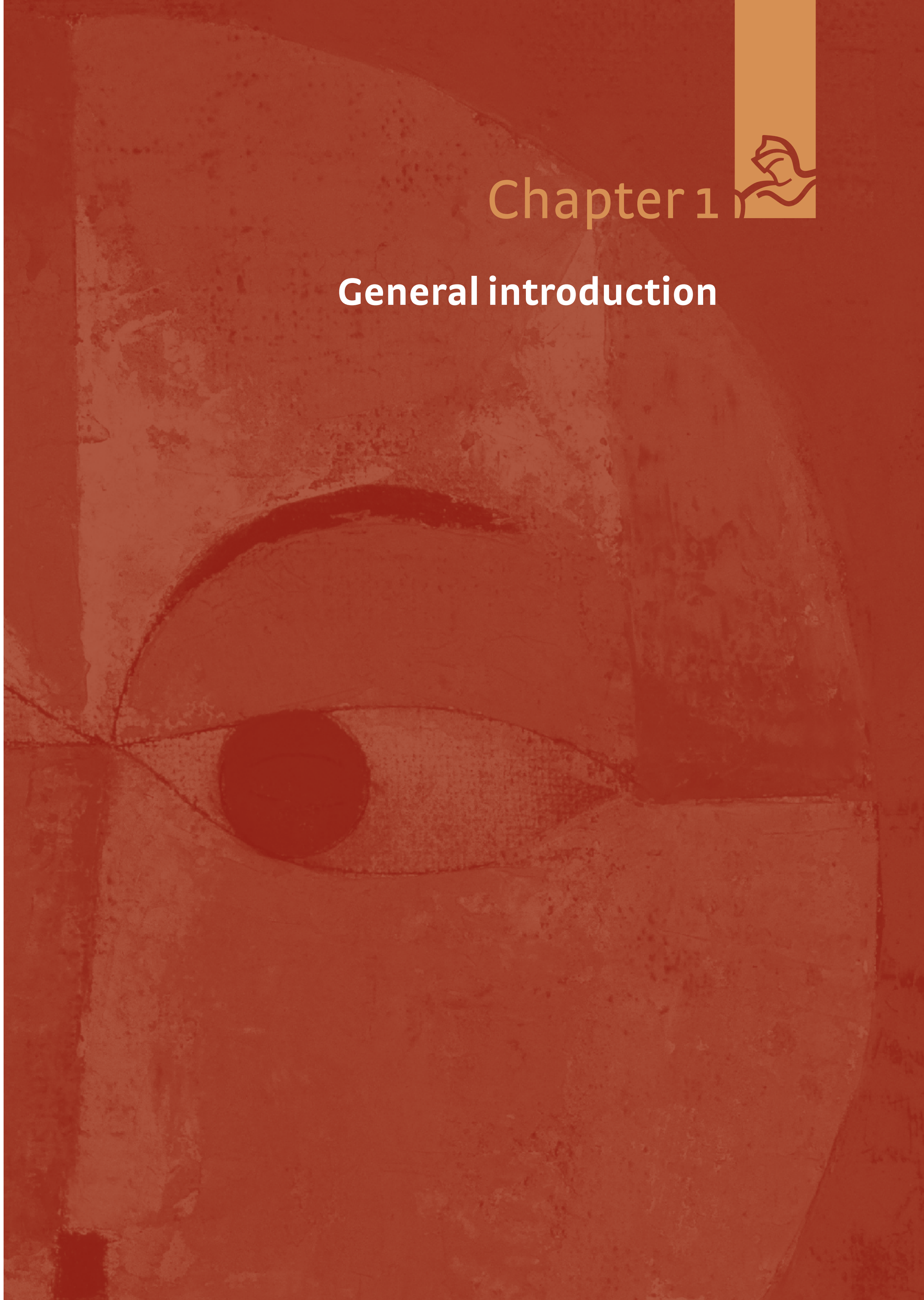
Chapter 1

General introduction



The management of systemic sclerosis (SSc) involves a combination of pharmacological and non-pharmacological care. Non-pharmacological care is delivered by health professionals such as physiotherapists, occupational therapists, psychologists, dieticians, dental hygienists, and social workers. They support persons with SSc in coping with the consequences of their disease and taking charge of the organization of their own care and the associated decision-making processes[1,2]. This thesis focusses on the coordination and quality of non-pharmacological care for persons with SSc, in particular the extent to which the care delivered by health professionals is aligned with their healthcare needs. It identifies possible targets for improving health professional care from the perspective of persons with SSc, health professionals, and rheumatologists, and provides evidence-based recommendations for a number of the most frequently described unmet care needs of persons with SSc.

Systemic sclerosis

The term scleroderma is derived from the Greek words scleros (hard), and derma (skin); hence the term "hard skin". The systemic form of scleroderma (systemic sclerosis, SSc) is a generalized connective tissue disorder characterized by thickening of the skin (scleroderma), microvascular and larger vascular lesions, fibrotic degenerative changes in muscles, joints and viscera mainly of the intestinal tract, heart, lungs, and kidneys, and disease specific autoantibodies[3]. SSc is a rare disease, with an estimated global annual incidence of less than 10 per 100,000 individuals. Estimates of prevalence vary widely depending on methods and diagnostic criteria used, and range from 7.2-44.3 per 100,000 individuals. The SSc diagnosis is usually made in persons aged between 34 and 60, and predominately affects women (ratio 3.8-15.1:1)[4]. The etiology of SSc is largely unknown, but it is believed that it involves both genetic and environmental factors[5]. Depending on the degree of skin involvement, SSc is divided into two main subtypes: limited cutaneous systemic sclerosis (lcSSc) and diffuse cutaneous systemic sclerosis (dcSSc). LcSSc is defined by skin thickening in areas distal to the elbows and knees, with or without facial effects, while in dSSc, skin thickening extends proximal to the elbows and knees, and/or trunk. Generally, dcSSc has a worse prognosis than lcSSc. Despite a better understanding of the underlying disease mechanisms, improved healthcare standards, and faster diagnosis, SSc is still associated with high mortality rates[6].

Consequences of systemic sclerosis

SSc is clinically characterized by a variety of symptoms, including Raynaud's phenomenon, skin thickening and skin fibrosis, digital ulcerations, pulmonary, renal or gastrointestinal involvement, and pulmonary arterial hypertension[7,8]. As a consequence, joint contractures and muscle weakness often lead to a progressive reduction of overall physical capacity and active range of motion in both upper and lower extremities[9,10]. This in turn can lead to severe hand function loss, impaired facial mimicry, incontinence, sexual dysfunction, and high levels of pain and fatigue[12-14]. As SSc progresses over time, additional psychological problems like anxiety, depressive symptoms, poor quality of sleep, and changes or impairment of self-efficacy and self-esteem can arise[15,16]. Fear of disease progression is a major concern, as SSc is a potentially life-threatening condition[6,17,18]. These physical and mental changes can lead to a severely reduced ability to perform activities of daily living (ADL), participation in work, leisure and social life, all consequently leading to a decreased health-related quality of life (HRQoL)[19-24]. In addition to SSc's direct impact, a lack of support from and inappropriate attitudes of family, friends or healthcare providers negatively impact HRQoL. Moreover, poorly coordinated healthcare services, systems and policies are cited as being responsible for disrupting day-to-day functioning of persons with SSc[21,25].

Current management of systemic sclerosis

Medical management

A variety of medical specialists such as rheumatologists, pulmonologists, gastroenterologists, cardiologists, dermatologists and nephrologists can be involved in the complex medical treatment of persons with SSc[26-28]. In general, the rheumatologist is the main coordinator of care. In the Netherlands, medical SSc management is not centralized; persons with SSc are treated in regional hospitals or centers of expertise, with a relatively small number in a shared care setting between both regional hospitals and centers of expertise[29]. Core elements of medical management are pharmacological treatment and tight monitoring of disease progression[30]. Despite several new promising therapies, including immunosuppressive therapy and new biological agents, there is as yet no evidence for the efficacy of a causal therapy[31].

Health professional management

Health professional care can play a significant role in SSc management as health professionals are well-equipped to address the variety of needs of persons with SSc[21]. In the Netherlands, health professionals from different professions predominantly active in primary care are involved in the treatment of persons with SSc. Persons with SSc can be referred to them, but they are also freely accessible. Health professional treatments consist of a careful and comprehensive assessment, including the setting of mutually agreed and clearly defined goals. Interventions offered by health professionals encompass a wide range of modalities, such as self-management support, exercise therapy, counselling, cognitive behavioral therapy, advice on splints, and the provision of orthotics and adaptive devices. Health professional treatments can be offered as individual actions, but can also be combined into comprehensive management strategies provided by one or more health professionals in a multidisciplinary setting.

In sum, living with SSc can have a large impact on persons' lives, and their physical and psychological health. Non-pharmacological treatment options relevant to improving HRQoL are target disease management, support with daily life coping mechanisms, and improving the relevant domains of disease impact (pain, fatigue, physical disfunction). However, access to and the quality of health care services available to persons with SSc delivered by health professionals are often substandard.

First, the use of health care delivered by health professionals is suboptimal, with persons with SSc reporting restricted access to health professional care[30]. The rheumatologist, as the main care coordinator, plays an important role in referrals to health professionals[32]. The referral process from rheumatologists to health professionals is not optimally aligned, and rheumatologists may reason from a different model of illness and health for this specific target group. Therefore, insights into the barriers impeding referrals as perceived by rheumatologists are needed.

Second, in alignment with advances in research and the global movement towards more person-centered health care, there is a tendency to develop treatment approaches based on self-management and shared decision-making[33]. New forms of collaboration between persons with SSc and healthcare providers have been established. Moreover, persons with SSc have been equipped with knowledge about self-management skills to be able to play a stronger role in the decision-making process. However, their expectations are not always met and there are still challenges to better match individual care expectations[34,35]. Insights in the experiences of persons with SSc about health professional care are needed to improve their access to care (e.g., by self-referral) and quality of care. This knowledge can then be used to better align the care delivered by health professionals with their healthcare needs.

Thirdly, many health professionals lack SSc-specific expertise because of the disease's rare occurrence and complexity. Health professionals have reported a great need for specialized training programs[36]. Furthermore, in recent decades, owing to changes in the Dutch health care system, health professional treatment has been increasingly transferred from hospital-based team care to a monodisciplinary primary care setting. As a result, the SSc-specific expertise of health professionals is fragmented in the Netherlands. It is likely that this lack of expertise negatively affects the content of care, therefore making an inventory of the extent health professionals are able to address the needs of persons with SSc is an important step towards improving educational offerings.

Finally, the lack of high quality evidence on the effectiveness of SSc-specific health professional treatments hampers establishing a set of specific recommendations on non-pharmacological treatment approaches[30]. The two largest systematic reviews to date focusing on the effectiveness and safety of non-pharmacological interventions only identify 33 studies that describe the effectiveness of a wide variety of interventions on an even wider variety of outcome measures[36,37]. Moreover, current SSc guidelines and recommendations from the European league against Rheumatism (EULAR) and the British Society of Rheumatology (BSR) do not or only superficially include recommendations regarding non-pharmacological care[30]. Therefore, recommendations for clinical practice need to be developed to support clinical decision making and to make treatment options transparent and accessible for all stakeholders. Creating recommendations for rare diseases like SSc presents specific challenges. The body of evidence is slim and inconsistent and published studies are often heterogeneous. A possible strategy for overcoming these barriers is the additional use of expert consensus and indirect evidence, i.e., evidence extrapolated from research involving patients with other (rheumatic) conditions with similar health consequences, a strategy advocated for the development of rare disease recommendations[38].

Roadmap of this thesis

The ***first*** aim of this thesis is to investigate possible targets for improving the indication for care delivered to persons with SSc by health professionals in persons with SSc from the perspective of essential stakeholders: persons with SSc, health professionals, and rheumatologists.

Chapter two provides insights into the rheumatologists' referral routine and identifies factors influencing rheumatologists' decisions about referral of persons with SSc to health professionals.

Chapter three adds the perspective of persons with SSc on care provided by health professionals with a focus on referral reasons, treatment goals, alignment with unmet care needs, and outcome satisfaction with health professional treatments.

Chapter four describes the spectrum of treatment options from the view of Dutch health professionals, including alignment of treatment goals with referral reasons, coverage of unmet care needs reported by persons with SSc, and quality of communication between health professionals and rheumatologists.

Based on these three perspectives, the ***second*** aim is to develop consensus and evidence-based recommendations for some of the most frequently described unmet care needs of persons with SSc, in order to facilitate accessible and effective non-pharmacological SSc care.

Chapter five describes the development of multidisciplinary consensus and evidence-based recommendations for non-pharmacological treatment of fatigue, hand function problems, and Raynaud's phenomenon/digital ulcers in persons with SSc.

Chapter six reflects on the research process, presenting a synthesis of the main findings of the first three studies. This chapter also introduces factors that can possibly contribute to more accessible and effective non-pharmacological SSc care, and presents methodological considerations.

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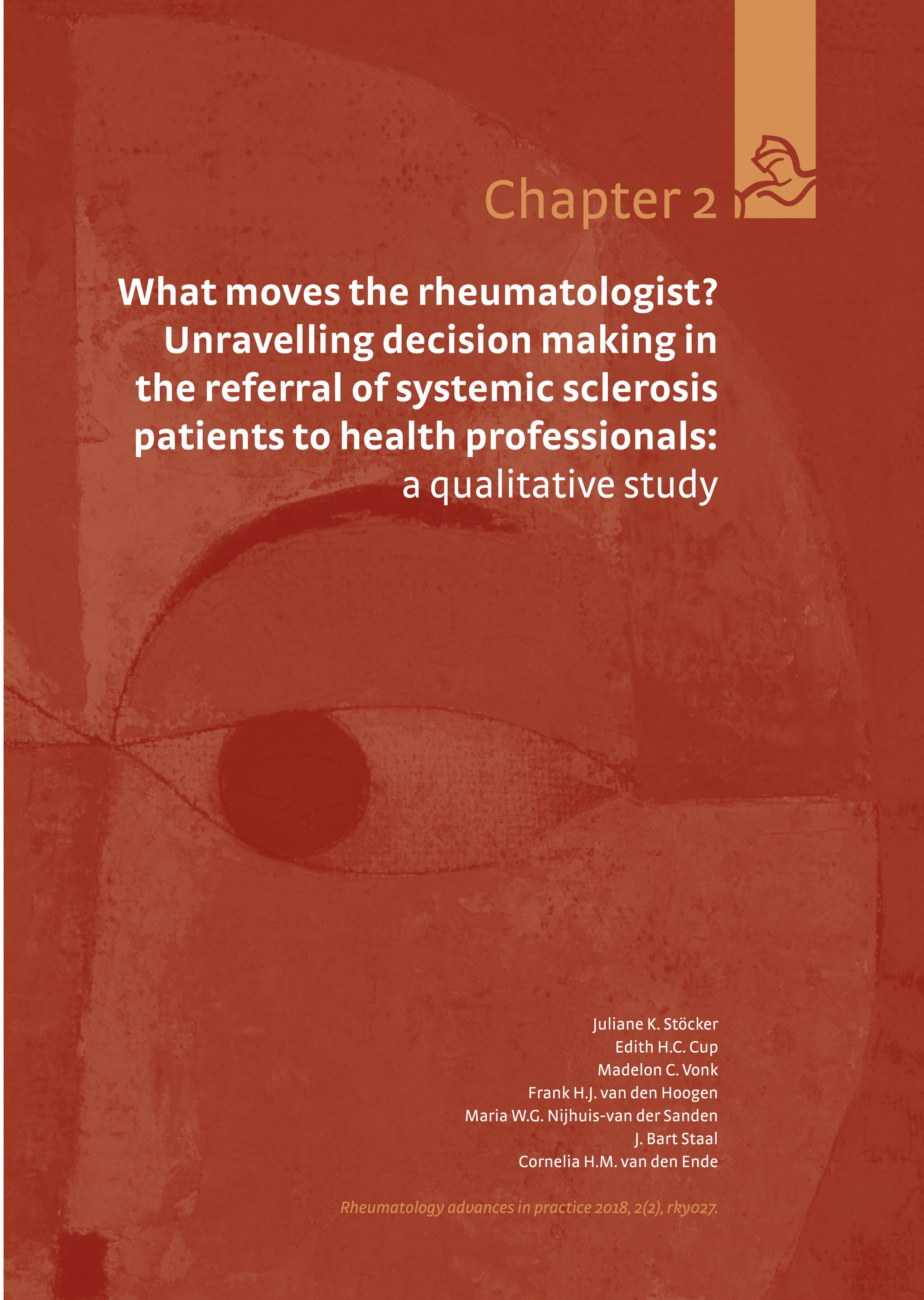

Chapter 2

What moves the rheumatologist? Unravelling decision making in the referral of systemic sclerosis patients to health professionals: a qualitative study

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Abstract

Objectives

Well-coordinated multidisciplinary non-pharmacological care is considered to be a cornerstone in the management of patients with systemic sclerosis. However, it has been discovered that unmet information and health care needs are common in patients with SSc. In addition, referrals by rheumatologists do not always correspond with potential treatment goals as identified by health professionals. The aim of this study was to gain insight into the current referral routine of rheumatologists in SSc patients and to identify and explore factors influencing rheumatologists' decisions about referral of SSc patients to health professionals.

Methods

Qualitative semi-structured interviews were held with 13 rheumatologists specializing in SSc management from different hospitals in The Netherlands.

Results

Our study identified rheumatologists' beliefs and local policy as influencing factors for referral to health professionals and a clear need for a better referral policy. Furthermore, a lack of knowledge about and low confidence in the competence of other disciplines were identified as barriers for referral to health professionals, which may possibly lead to undertreatment.

Conclusion

In the opinion of the majority of rheumatologists, adequate referral to health professionals requires an active role for the patient and increased visibility from health professionals.

Introduction

SSc is a complex and rare autoimmune disease with high morbidity and mortality[1,2]. Prevalence estimates vary around 20 per 100 000[3,4]. The main feature of SSc is skin fibrosis, but internal organs as well as muscles, joints and tendons can be affected as well. SSc has a significant impact on daily functioning, participation and quality of life[5,6]. A large proportion of patients experience a wide range of physical and psychological symptoms, such as chronic fatigue, pain, stiffness of joints, reduced hand function, reduced mouth opening, depression, body image distress and uncertainty about the future[7-10]. Pharmacological treatment of SSc has modest to moderate efficacy in terms of reducing morbidity and mortality, and disease-modifying medications are scarce, if not lacking completely[11].

In The Netherlands, non-pharmacological treatment is often provided as an adjunct to pharmacological treatment. Health professionals, such as nurses, social workers, psychologists, occupational therapists, physical therapists and podiatrists, play an important role supporting patients with SSc in coping with the consequences of their disease in daily life. The majority of patients have, on average, seven or more visits yearly to one or more health professionals[10]. However, research on the efficacy of non-pharmacological interventions for this specific patient group is sparse[12]. In her systematic review on the effectiveness of non-pharmacologic interventions, Willems *et al.* found 23 studies with wide variations in the content of interventions and outcome measures, but just 3 studies (randomized controlled trials) met the criteria for methodologically high quality[13]. As a result, non-pharmacological treatments in SSc vary widely among health professionals with respect to treatment goals and content of interventions[14,15]. Unmet information and health care needs are common among SSc patients[16]. This might be explained by the fragmented non-pharmacological care and lack of knowledge among physicians and patients about available treatment modalities for SSc. In addition, the majority of patients are dissatisfied with the coordination of care[10]. Furthermore, reasons for rheumatologists to refer SSc patients to health professionals, mainly focusing on functional impairment, do not correspond with the treatment goals of health professionals, which frequently focus on the patients' needs concerning daily activities and participation[17]. This implies that for SSc patients who receive care from multiple providers, attention should be given to the referral process, including communication among rheumatologists, SSc patients and health professionals[10].

This study is the first part of an umbrella project that aims to create transparency in the referral process of SSc patients from rheumatologists to health professionals using the view of all parties involved and to establish recommendations for improvement of the referral process. The objective of this study is to gain insight into the current referral routine of rheumatologists in patients with SSc and to identify and explore factors influencing rheumatologists' decisions about referral of SSc patients to health professionals.

Methods

We applied a qualitative study design using semi-structured interviews and inductive content analysis[18]. This methodology fits within an interpretive paradigm and enabled us to study the perspectives of specialized rheumatologists in terms of the SSc referral process and content. In order to report explicitly and comprehensively, the COnsolidated criteria for REporting Qualitative research checklist was used[19].

In The Netherlands, the majority of SSc patients are treated by rheumatologists with special expertise in SSc, appointed at several teaching and general hospitals across the country[10]. In this study we aimed to include rheumatologists with special expertise in the management and treatment of patients with SSc. Two scleroderma expert rheumatologists (M.V., F.H.) selected colleagues with special expertise in SSc from all rheumatologists registered in The Netherlands in August 2015 ($n = 361$). This resulted in a list of 24 rheumatologists. The minimum sample size for initial analysis was set at 40% ($n = 10$). We applied a stopping criterion of three, implying that data saturation is achieved after three new interviews without new ideas emerging[20]. Potential participants were invited by e-mail to participate in the study. Rheumatologists were included in the study after full oral informed consent, including quotations used in the published article, was obtained. The Institutional Review Board of the Radboud University Medical Centre, Nijmegen concluded that the Medical Research Involving Human Subjects Act did not apply to this study (protocol number RR-157-678). The interviews were carried out by the first investigator (J.K.S.) at each participant's home or workplace between September 2015 and May 2016.

Data were collected during semi-structured interviews. The use of an interview guide (for supplementary data see Appendix 1) ensured that the main issues were addressed. Interview questions were based on the evidence-based decision making model in order to take the view of the rheumatologist in their own context into account[21]. The semi-structured interview guide had an open-ended format. It focused on reasons for referring SSc patients to health professionals or not, factors important to rheumatologists when referring SSc patients and experiences with health professionals in daily practice.

In addition, self-reported demographic information was recorded. The interview started with open and explorative questions about factors influencing the rheumatologist's referral decisions, followed by in-depth and probing questions to extend the responses and help participants articulate their experiences. All interviews lasted between 45 and 60 min and were recorded digitally and transcribed verbatim[22].

Qualitative data analysis followed the method of inductive content analysis adapted from Nayar and Stanley[18]. Analysis followed a six-step process of coding to create established meaningful themes:

- Step 1: Transcribed interviews were read through several times by the principal investigator to obtain a sense of the whole data set.
- Step 2: Initial coding: Two investigators (J.K.S., E.C.) independently coded the first three interviews by highlighting text fragments that appeared to capture key thoughts or concepts in relation to the research question to enhance. Subsequently J.K.S. and E.C. discussed the procedure and content of the analysis. J.K.S. continued to allocate codes to remaining transcripts.
- Step 3: Grouping codes into meaningful categories (J.K.S.).
- Step 4: The resulting categories were discussed with two members of the project group (E.C., C.H.M.E.). Minor adaptations were made by moving codes into other categories. For member checking, a short description of each category was sent to the participants and additionally to the panel of patient research partners (H.K., J.T.V., J.W.) for comment. This research triangulation enhanced the credibility of the findings[22].
- Step 5: The categories were grouped into meaningful themes. In defining the themes, the researcher paid attention to using the expressions of the participants in order not to lose the original meaning of the expression.
- Step 6: The resulting themes were discussed in the whole group of investigators until consensus was obtained. The discussion with the research team also enhanced the credibility.

Results

Study-wise data saturation was achieved at interview 13, as no new ideas had emerged after interview 10, thus the scheduling of interviews ended. None of the rheumatologists approached refused to participate or dropped out after giving informed consent. The 13 rheumatologists came from nine different centers in The Netherlands, with work experience within their specialty ranging from 3 to 30 years (Table 1).

Table 1: Characteristics of the interviewed rheumatologists ($n = 13$)

Characteristics	Values
Female, n (%)	7 (54)
Age, median (range), years	35 (33–61)
Years in practice, n (%)	
1–10	4 (31)
11–20	7 (54)
>21	2 (15)
Working in an academic hospital, n (%)	6 (46)
SSc patients in all treated patients, % (range)	35 (5–85)
Doctoral degree, n (%)	9 (69)
Involved in research, n (%)	6 (46)
Involved in SSc guidelines development (local or national), n (%)	6 (46)

Two major themes, beliefs and local policy and routines, were identified as influencing decision making with respect to referral of SSc patients to health professionals. We also found an additional theme reflecting the needs of the rheumatologists regarding professional multidisciplinary collaboration. The three themes, subthemes and associated categories are displayed in Table 2.

Table 2: Themes, subthemes and categories

Themes	Sub-themes	Categories
Beliefs	Beliefs about one's own professional role	A coordinator with a helicopter view
	Beliefs about the patient's role	Pro-active patients, rheumatologists in the lead and joint decision making
		Creative patients need less guidance
		Patients in charge
	Beliefs about the role of health professionals and rheumatology nurses	Role and competence of health professionals and effectiveness of non-pharmacological treatments
		Role of rheumatology nurse
Local policy, money and time		Local policy, money and time
		Referral to which health professional?
Needs		Active, visible health professionals
		Few large expert centers exchanging expertise with regional centers
		Regional expert networks

Theme 1: beliefs

Beliefs about one's own professional role

A coordinator with a helicopter view

The rheumatologist sees him/herself as a coordinator with a helicopter view, who gives the patient guidance and structure, especially at the beginning of the treatment.

P9: 'In principal, you are kind of the coordinating factor...you're almost a bit of the patient's GP... I do think, that as the rheumatologist you kind of need to keep a helicopter view of all the different aspects of what the patient is dealing with'.

They inform the SSc patient about pharmacological and non-pharmacological treatment options and in some cases about specific exercises. All rheumatologists express their intention to offer tailored care and are interested in seeing the patient from a holistic perspective. Their own continuous education and professional exchange are considered important factors for high-quality treatment, including evidence-based treatment options.

Beliefs about the patient's role

Proactive patients, rheumatologist in the lead and joint decision making

Expectations regarding the patient's role in the referral process vary among rheumatologists.

Some interviewees expect a very active role and feel that the patient has a responsibility to ask for a referral to care delivered by health professionals.

P1: 'The responsibility really does lie with the patient, they also need to make their own appointments, we don't do that for them'.

Several rheumatologists see themselves in the lead. They determine the policy for referral and propose this to the SSc patient. Other rheumatologists draw a picture of 'shared responsibility' based on the patient's complaint or request for assistance.

P6: 'But it's the case that I do feel it's my duty to inform that patient about everything that's available... You're sitting next to each other, you hear the story, you inform them, and together you make... I always have the feeling that we're making the decision together'.

All rheumatologists expect SSc patients to adhere to agreements made and to inform them about the progress of non-pharmacological treatments they receive. In addition, some rheumatologists mention behavior they find conducive to good cooperation, like the expression of the patient's own opinion, informing themselves prior to the consultation or preparing a list of points to discuss.

Rheumatologists state that SSc patients often focus on problems related to the complexity of the disease and their fear of potentially harmful medical examinations. As a result, they forget or do not have the time to discuss health professional treatment options.

P11: 'If the patient has a lot of medical problems, so you have to make medication changes... and the bit about multidisciplinary or possible referral to health professionals isn't discussed if you've only got a quarter of an hour'.

Creative patients need less guidance

In several interviews, a distinction was made between 'creative' and 'passive and uncertain' SSc patients. Creative patients are able to invent their own solutions to problems. It is easier for them to grasp new knowledge and make decisions about the referral process. They require less guidance and explanation. With passive and uncertain patients, the rheumatologist is more likely to propose solutions and to determine the policy for referral.

P2: 'You sometimes need to take the patients who demonstrate helpless behavior by the hand and actively show them the way. But in general, my impression is that the prognosis for a patient who's active, is better'.

Patients in charge

Several rheumatologists have a vision regarding the future role of the SSc patient as a partner and expect them to have an active role in the treatment process. They also have the same view with respect to their health professional and rheumatology nurse colleagues. They envisage a collaboration with four active parties.

P2: 'I think the patient should also make an active contribution to his care...So, I actually want there to be four active parties, and I prefer the responsibility to lie with the patient'.

Beliefs about the role of health professionals and rheumatology nurses

Beliefs about the role and competence of health professionals and the effectiveness of non-pharmacological treatments

Almost all rheumatologists stated that they have little or no knowledge about treatment options of health professionals. A few indicated they were familiar with the content of treatment offered by physiotherapists, occupational therapists, dieticians and hand therapists in their own center. Outcome expectations varied widely among the rheumatologists interviewed and were based on personal experience with health professionals. Rheumatologists with a clear structure of collaboration and regular exchange with health professionals within their own work setting expressed the added value of health professional treatments.

D10: 'An incredibly important role (health professionals). Not only in self-management, but disease perception, being able to support and steer where necessary...You will not cure the disease, but I think it has a huge potential in maintaining quality of life'.

Rheumatologists with little experience with health professional colleagues within their own institution felt uncertain about the effectiveness of non-pharmacological treatments.

D4: 'I realized myself, I do not know if it helps (health professional treatments) or whether it is coincidence or not'.

Rheumatologists with little or negative experiences with a specific health professional did not believe in the clinical reasoning skills of the health professionals, describing a lack of disease-specific knowledge and poor skills regarding reporting on treatment targets, content and outcome.

D6: '... and furthermore I do not refer to these health professionals because I think they cannot do anything at all, unless there is a very apparent reason for it'.

Beliefs about the role of the rheumatology nurse

In all interviews, the role of the rheumatology nurse was regarded more positively compared with other health professionals. Their qualifications and skills are also highly valued. All except one rheumatologist described a close cooperation with the rheumatology nurse, ranging from an advisory role to shared responsibility. The rheumatologist often focuses on the medical aspects and has full confidence in the rheumatology nurse's ability to address the non-pharmacological and multidisciplinary aspects, to identify problems not discussed and to give advice about health professional treatment. Rheumatology nurses are often seen as a key person for referrals.

In the case of new patients, the rheumatology nurse is often involved in the intake and, later on, is the representative of the other health professionals during multidisciplinary meetings. Four of the rheumatologists believe that, in the future, the rheumatology nurse could play a coordinating role between rheumatologists and other health professionals on a regular basis.

P9: 'In fact, I refer everybody who I diagnose with scleroderma to the rheumatology nurse. So that the nurse can give the patient more information about the clinical picture, can tell the patient about the challenges they'll face in everyday life. And I do use the nurse a bit as a guideline as to where I can further send the patient to'.

Theme 2: local policy and routines

Local policy, money and time

Local policy and, where present, care pathways have a significant impact on the intake and referral process. Some rheumatologists do not need to consider referral to health professionals, because intake and advice from all health care disciplines is part of the existing routine.

P11: 'Actually, I must say that here we're really connecting everything to the care path, so people come every year...So then in fact I don't need to refer them, because that happens automatically'.

Due to time constraints during follow-up consultations, referrals to health professionals initiated by the rheumatologist are mostly a result of the first consultation. Follow-up consultations are usually shorter and, due to the complexity of the disease, focus primarily on medical aspects. As a result, there is less attention given to non-pharmacological care.

P11: 'You know, if I'm really busy and my outpatient clinic runs over, that's also a factor that influences whether or not I remember to mention: "Oh yeah, maybe it might be wise for you to once go back to the occupational therapist sooner"'.

Financial aspects are taken into account when deciding about referral.

P3: 'Things that I come up against? I think mainly the payment structure. And in particular for physiotherapy. For example, systemic sclerosis isn't covered in its chronic form. So I always discuss with people, look at how much is covered in the insurance. Otherwise it's really expensive for people'.

Most rheumatologists express a clear preference for certain health professionals when referring their patients. This is often driven by costs, clinical pathways and internal policies with regard to referral to either hospital-based or primary care health professionals. Another reason for referral to a preferred therapist is trust and confidence in the expertise of colleagues they know personally.

All centers use a center-specific SSc intake list comprising medical and non-medical aspects that need to be addressed during consultation. Often there is a more comprehensive list for new patients, with additional questions about work and leisure activities as possible targets for health professional treatments.

In 11 of the 13 interviews, physical symptoms and functional limitations were the main reason for referring a patient to a health professional, while two rheumatologists base their decision on the actual or potential loss of the ability to perform everyday activities or to participate in society.

P12: 'As a rheumatologist you try to see if there are any physical limitations. If there are any, you quickly look at how you can… let's say, try to solve it with the help of occupational therapy or physiotherapy'.

Some rheumatologists consider non-pharmacological treatment options only after the failure of pharmacological treatment options.

Referral to which health professional?

Most rheumatologists regularly refer patients to physiotherapists and occupational therapists. Preferences for referral to either hospital-based or primary care health professionals differ. In general, physiotherapy in primary care is the first option, whereas in the case of occupational therapy, referral within the rheumatologist's own center is common. In many places, referral to dieticians is defined by care pathways. Referrals to hand therapists, dental hygienists, podiatrists, social workers and psychologists are made on a less regular basis. Travel distance to the therapist and associated costs and effort for the patient are also determining factors.

Theme 3: needs

The needs of the rheumatologist concern their working environment, collaboration with health professionals and their vision of collaboration in the future.

Need for active, visible health professionals

Generally rheumatologists express a need for sufficient visibility of health professionals and active communication about therapy goals and treatment content. All interviewees perceived a lack of published evidence on health professional interventions.

Few large expert centres exchanging expertise with regional centres

Due to the complexity of the disease, the rheumatologists prefer that all health care take place as close as possible to the patient's home environment. They believe in the importance of establishing a satellite system of a few specialized SSc centers facilitating smaller regional centers in knowledge exchange.

P5: 'Because SSc is so rare you can actually cluster the experience and that results in the people being treated better. Although… you mustn't specialize it that much so that people can only reach one center with their questions. There should at least always be a regional center'.

Need for regional expert networks

The rheumatologists wish more interaction with health professionals and a clear communication and collaboration structure. In addition, they prefer health professionals adopt a more active role in the SSc treatment process and provide more transparency about their area of expertise and treatment content. The latter could be achieved through the establishment of local and regional networks of health professionals with disease-specific knowledge and protocols. Finally, in daily practice rheumatologists value transparent communication, direct or by telephone or e-mail, and would appreciate more structured reports.

Discussion

This study focuses on daily routines and factors influencing Dutch rheumatologists' decision about the referral of patients with SSc to health professionals. We identified two major themes: beliefs and local policy and routines. The additional theme reflecting the needs of rheumatologists regarding professional multidisciplinary collaboration is not directly related to the research question, but rather reflects the rheumatologists' perspective on future challenges. As far as we know, this is the first qualitative study focusing on the experiences of rheumatologists on this specific topic.

Initially the goal of this study was to investigate factors that influence rheumatologists in their decision to refer SSc patients to health professionals, in order to bring transparency to their decision-making process. However, during the analysis phase and comparison with existing literature, it became clear that we were not dealing with a list of distinct factors, but rather with a complex reasoning structure underlying the rheumatologist's decision-making process.

Rheumatologists expressed the considerable value they attach to evidence-based practice, as well as for the credibility of the therapy content of other disciplines. A lack of evidence for non-pharmacological treatments and a correspondingly low confidence in their competence was often mentioned. There is indeed little evidence regarding treatments that specifically focus on SSc[13]. However, since non-pharmacological treatments often do not focus on a specific disease, but rather on limitations in activities, there is evidence for a large number of non-pharmacological treatments originally intended for other rheumatic conditions[23]. Therefore, in our opinion, a transfer of knowledge about the effectiveness of non-pharmacological treatment options could improve SSc care.

In analyzing rheumatologists' reasoning about decision making and referral we were able to distinguish the influence of strong local policy, financial aspects and time constraints. Our findings suggest that for referral to health professionals, in the absence of scientific evidence, rheumatologists predominantly make use of their personal experience, beliefs and local policy. This is in line with the rheumatologists' decision-making routine described by Ianello *et al.*[24]. These findings also underpin the findings of Gabbay and le May regarding 'collectively constructed mindlines' that are built up as a 'bank of personalized, flexible syntheses of all the different types of theoretical and experiential knowledge', and affect professionals' reasoning and decision making[24,25].

A common perception among the rheumatologists was a lack of confidence in the clinical reasoning competence of health professionals and their insufficient knowledge about options of non-pharmacological treatment. A strong relationship between these two factors has also been reported by Arena et al. and Suter *et al.*[26,27]. They found that a lack of knowledge about potential benefits is an important factor in the underutilization of treatments and rehabilitation and suggest that low perception of confidence in diagnostic and treating competence influences the decision of whether or not to refer a patient. On the same note, Larme and Pugh and Gallagher et al. found that a combination of lack of knowledge and a perception of low efficacy of treatment can negatively affect the patient's empowerment in their self-management[28,29].

One strength of this study is the involvement of a panel of patient research partners, who reflected upon and advised on the execution of the different steps of our research.

Due to the fact that the majority of SSc patients in The Netherlands are treated by specialized rheumatologists working in a teaching or general hospital setting, only those rheumatologists who, in the opinion of our two expert rheumatologists (F.H., M.V.), were considered to be experts were included in the study. As a result, we may have missed rheumatologists who see themselves as an expert. Moreover, we only interviewed rheumatologists about their opinion of the referral process. Therefore the perspective of patients and health professionals is missing. This means that the picture is not yet complete. Further research will be needed to identify the perspectives of all parties involved in the referral process.

Conclusions

Our study identified rheumatologists' beliefs and local policy as influencing factors for referral of SSc patients to health professionals and the clear need for a better referral policy. Furthermore, a lack of knowledge and low confidence in the competence of other disciplines were identified as barriers for referral to health professionals, which may possibly lead to undertreatment. The low confidence level is closely linked to beliefs regarding the rheumatologist's own role as well as that of the health professionals and SSc patients. Regular mutual contact between rheumatologists and health professionals seems to be a crucial factor in increasing confidence in non-pharmacological treatment options.

In the opinion of the majority of the rheumatologists interviewed, adequate referral to health professionals requires an active role on the part of the SSc patient, greater visibility of health professionals and a coordinating role of the specialized rheumatology nurse.

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Appendix 1: Interview guide

Interview guide

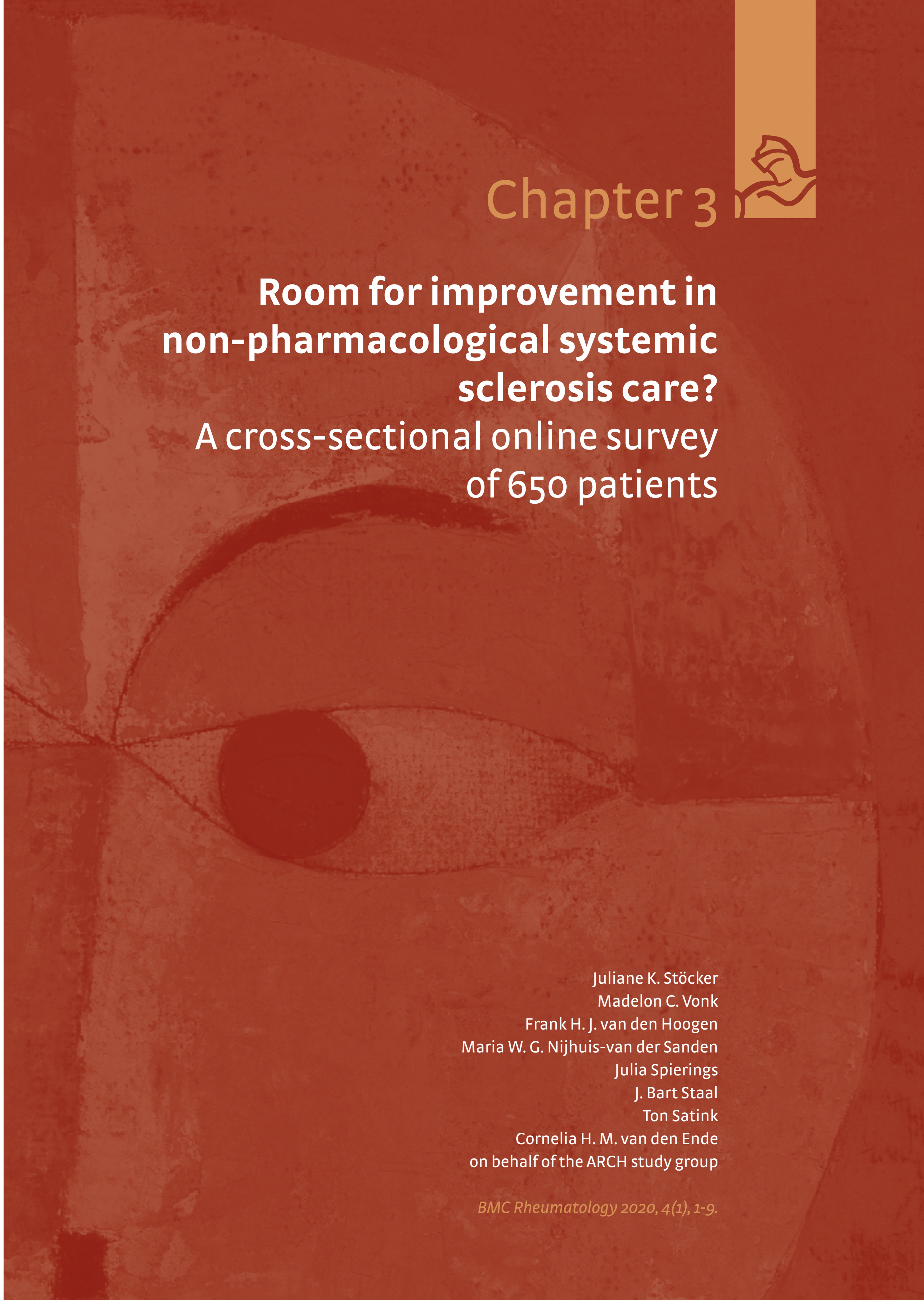
The moderator guide for the 45-60-minute interviews was divided into two modules

A -Participant information/ Introduction

- Explanation of interview purpose
- Description of audio recording and consent to continue
- Demographical questions (age, work experience with SSc)
- Case load of SSc patients
- Sources/ acquisition of (new) knowledge on SSc

B –Referral process to health professionals

- The role of the rheumatologist within the center
- Description of usual consultation with SSc patients/ Possible treatment patterns
- Process of decision making for referrals to health professionals
- Possible influences on decision making for referral
- Health professionals involved in patient management
- Knowledge about content of different health professional treatment options
- Personal evaluation on value and effectiveness of health professional treatment options in SSc care
- Point in time at which health professionals (possibly) become involved in patient care
- Role of the rheumatologist, the patient and health professionals on referral process
- To what extent are health professionals part of your daily work with SSc patients?
- Nature and extent of communication with health professionals about treatment goals and content of treatments



Chapter 3

Room for improvement in non-pharmacological systemic sclerosis care?

A cross-sectional online survey of 650 patients

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Abstract

Objective

To gain insight in the use of current systemic sclerosis (SSc) care provided by health professionals from the patient perspective. We focused on referral reasons, treatment goals, the alignment with unmet care needs, and outcome satisfaction.

Method

Dutch SSc patients from 13 participating rheumatology departments were invited to complete an online survey. Descriptive statistics were used to describe current use of non-pharmacological care and outcome satisfaction. Reasons for referral and treatment goals were encoded in International Classification of Function and Disability (ICF) terms.

Result

We included 650 patients (mean (standard deviation [SD]) age, 59.4 (11.4) years. 50% had contact with a health professional in the past year; 76.3% since disease onset. Physiotherapists were the most frequently visited in the past year (40.0%), followed by dental hygienists (11.4%) and podiatrists (9.2%). The three most common referral reasons were pain, joint mobility and cardiovascular functions. Fatigue, Raynaud's phenomenon, physical limitations, reduced hand function and joint problems were mentioned by more than 25% of all respondents as unmet needs. The proportion of patients treated in the past year by a health professional who were satisfied with knowledge and expertise of their health professionals was 74.4%; 73% reported improved daily activities and better coping with complaints. However, 48.9% perceived that the collaboration between rheumatologist and health professional was never or only sometimes sufficient.

Conclusion

Despite the high outcome satisfaction and good accessibility of health professionals, there are obstacles in the access to non-pharmacological care and communication barriers between health professionals and rheumatologists.

Introduction

Systemic sclerosis (SSc) is an orphan connective tissue disease characterized by progressive fibrosis and vasculopathy affecting the skin and multiple internal organs[1]. Despite a growing body of knowledge and new therapeutic approaches, SSc remains a potentially fatal disease with a high clinical burden[2,3]. SSc can affect the physical and psychological conditions, daily functioning, and participation in society. Pain, digital ulcers, fatigue, and joint contractures significantly contribute to impaired functional capacity and are associated with negative perceptions of illness severity[4-6]. Depression, distressing appearance transformation, social isolation, and Raynaud's phenomenon have high impact on health-related quality of life (HRQoL) in patients with SSc[7-9].

In recent years, an increased understanding of the disease and targeted research activities have led to an improved classification and a growing number of pharmacological treatment options for specific complications. Much effort has been made to identify the patients' perspective on their disease, quality of life and potential therapeutic targets[10-12]. Owing to the direct impact of the disease on daily functioning and psychosocial well-being of patients, non-pharmacological care is a key element of SSc care. So far, the evidence for non-pharmacological approaches in SSc is limited and specific guidelines are not available yet[13]. According to the updated European League Against Rheumatism (EULAR) recommendations, the evaluation of the efficacy of non-pharmacological treatments in SSc is on the research agenda for the next update[14].

Restricted access to trustworthy information, including knowledgeable health professionals, and lack of support in managing difficult social interactions and negative emotions are seen as unmet needs in SSc care[13]. A previous qualitative study among rheumatologists revealed barriers for referral to health professionals due to the lack of evidence for non-pharmacological treatments and a correspondingly low confidence of rheumatologists in health professional competences[15]. In the study of Willems et al. among European health professionals about the content of non-pharmacological care, discrepancies between physicians' reasons for referral and treatment targets as defined by health professionals were found. This also suggests a fragmented knowledge of physicians about the content of non-pharmacological care and a suboptimal communication between physicians and health professionals[16].

Today, patients have an important role in the organization of their own care[17]. Shared decision making contributes to optimal healthcare for SSc patients in terms of improvement of health outcomes, quality of care, and healthcare services. So far, it has not been investigated how SSc patients value non-pharmacological care, the coordination between rheumatologist and health professional, and to what extent this care fits the patients' needs. Therefore, it is important to involve the patients' perspective, as alignment in the communication between the different stakeholders is likely to lead to more effective personalized SSc care.

The purposes of this study were to provide insight in the use of the current SSc care provided by health professionals from the patient perspective. We focused on (1) the use of care (2) referral reasons and treatment goals, (3) their alignment with reported unmet care needs, and (4) outcome satisfaction with health professional.

Methods

Study design

A multicenter, cross-sectional, online survey was performed to explore health care utilization and perceptions of SSc patients in the Netherlands.

Participants

In the Netherlands, the Arthritis Research and Collaboration Hub (ARCH) was established as a nationwide effort to improve health care for patients with rare systemic autoimmune diseases, including SSc. The ARCH working group purposely selected the departments of rheumatology for the study, to ensure a representative patient population from both regional (*n* = 7) and university (*n* = 6) hospitals spread across the Netherlands. Patients with a registered diagnosis of SSc, treated in one of the 13 participating rheumatology departments, were selected from the patient administration system of the institution and invited to participate. Information about the survey was communicated to the patients by the treating rheumatologists. The invitation was accompanied by a written participant information letter and a reply card. After returning the reply card or sending a notification e-mail, a unique web link was distributed to enter the online survey. The inclusion criteria were as follows: being diagnosed with SSc, aged ≥18 years, and sufficient knowledge of the Dutch language. Data were processed anonymously. All participants provided informed consent when starting the web survey and before they were asked substantive questions.

Ethical approval was obtained by the Institutional Review Board of the Radboud university medical center, Nijmegen (protocol number: 2017–3621). The Strengthening the Reporting of Observational Studies in Epidemiology (STROBE) guidelines were followed[18].

Data collection

The online survey was hosted by Castor Electronic Data Capture (Castor EDC; Castor, Amsterdam, the Netherlands), a highly secured, cloud-based electronic data capture platform[19]. The survey questions were constructed based on the results of a literature review, three semi-structured multicenter focus group interviews with 23 patients, and interviews with 12 rheumatologists and five specialized nurses. Next, the survey was evaluated by the members of the ARCH SSc working group and a patient panel[20]. The questionnaire contained 67 multiple choice, multiple response, and open questions covering the following: [1] sociodemographic characteristics; [2] opinions on bottlenecks and areas for improvement; [3] perceived quality of care, and [4] non-pharmacological care. The survey was pilot tested in five SSc patients. To answer the research question of this study, we used data concerning non-pharmacological care and unmet needs in SSc care.

Description of the selected questions of the survey

Sociodemographic questions

Sociodemographic questions included sex, age, educational level, living situation, employment and disability status, and disease characteristics (disease subset, symptom onset and year of diagnose).

Unmet needs in SSc care

The question '*I would like more attention to be paid in my treatment to the following topics*' was assessed using a list of 27 yes/no questions on changed appearance, physical limitations, pain, fatigue, impaired walking and/or hand function, sleeping problems, psychological problems, sexual dysfunction, stomach and intestine problems, reduced mouth function, gynecological complaints, Raynaud's phenomenon, joint problems, loss of independence, loss of work / school, daily activities, and social life; insufficient support from social network, dealing with uncertainty, unpredictability of SSc, ambiguities about the diagnosis, feeling misunderstood, loneliness, loss of self-confidence, contact with other SSc patients, and the possibility to indicate other topics.

Non-pharmacological care

To assess the use of non-pharmacological care, patients were asked whether they consulted one or more health professionals because of SSc-related problems, since onset of the disease (yes/no) and during the last 12 months (yes/no). Patients who consulted one or more health professionals during the last 12 months were asked to identify the professional most frequently contacted. The list offered included the following health professionals: dietitians, occupational therapists, physiotherapists, hand therapists, speech- and language therapists, social workers, dental hygienists, exercise therapists, podiatrists, and psychologists. Moreover, patients could add other health professional disciplines to the list. Referral reasons and treatment goals were assessed by open-ended questions.

Two subscales, such as "coordination and alignment of care" (four questions) and "your health professional" (three questions) from the Consumer Quality Index (CQI) (rheumatoid arthritis, version 2.0), which has been found to be reliable to measure patients' experience with the quality of care in the field of rheumatology, were adapted for the current study[21]. Only questions of those two subscales focusing on communication, alignment, and outcome satisfaction with health professional treatments were selected. In addition, the wording "healthcare providers" was changed into "between rheumatologist and health professionals" in 4 questions of the subscale "coordination and alignment of care". Items were assessed on a four (five)-point Likert-scale [never, sometimes, most of the time, always, (I don't know)].

Data analysis

Descriptive statistics were used to describe demographic characteristics, unmet needs, current use of non-pharmacological care, and outcome satisfaction. Statistical analyses were conducted using Stata/IC 13.1 (StataCorp LP, College Station, TX). The free-text responses on the open-ended questions about reasons for referral to HPs and treatment goals were read and re-read to obtain an overview of the collected data. To examine the alignment of referral reasons to unmet needs, the concepts were compiled verbatim and subjected to an exploratory thematic analysis[22]. Coding discrepancies were resolved by discussion between two researchers (JS and CME) before refining the codes by summarizing and encoding in ICF terms (categories and subcategories) using the following:

- the updated ICF linking rules[23];
- the World Health Organization (WHO) ICF browser[24];
- the International Classification of Functioning, Disability and Health (ICF) Core Sets for rheumatoid arthritis[25]; and
- concepts of functioning and health as identified to be important to SSc patients[26].

The proportion of patients with unmet needs was calculated related to the number of patients that reported that need.

Results

A total of 2093 Dutch patients with SSc were invited to take part in the study from December 15th, 2017 to January 21st, 2018. Among the 2093 invited patients, 664 answered the survey. Data of 14 patients were excluded from the analysis, because of incompleteness. Thus, a total of 650 surveys were included in the analyses (Figure 1).

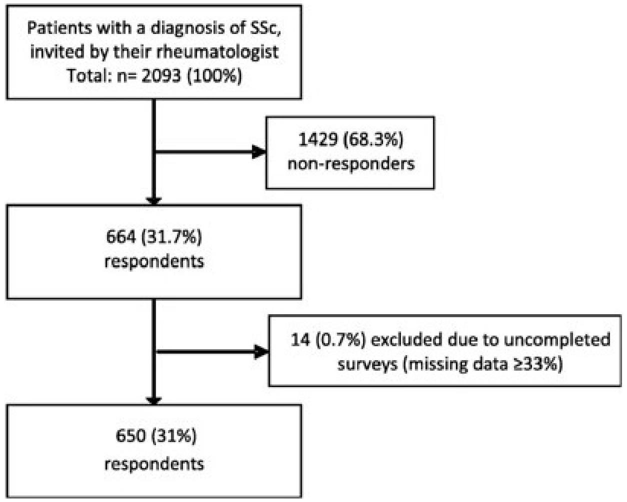

Figure 1: Flow diagram of patient selection procedure

Demographics and disease characteristics of the 650 respondents are displayed in Table 1. The majority of the responding patients were women (*N* = 486; 74.8%), with a mean age of 59.4 years (standard deviation [SD] = 11.4) and a mean time since onset of 8.2 years (SD = 8.0). About one third of the respondents received a higher education, and 82% (*N* = 533) were married or living together. Only 37.7% of the respondents were employed.

Table 1: Demographic and disease characteristics of 650 patients with SSc

Characteristics	Values
Female, N (%)	486 (74,8)
Age, years; mean (SD), range	59.4 (11.4), 18-87
Education level, N (%)	
0-12 years	443 (68.2)
> 12 years	207 (31.8)
Living alone, N (%)	117 (18.0)
Paid employment (%)	245 (37.7)
Disease subtype (%)	
Limited	227 (34.9)
Diffuse	132 (20.3)
Subtype unknown	291 (44.8)
Disease duration, years; N (SD), range*	8.2 (8.0), 0-51
Mean time between onset and diagnosis, years; N (SD)*	4.3 (6.9)
Women	4.8 (5.1)
Men	2.5 (7.3)

*Due to missing values, N=646

Use of care provided by health professionals

Since the onset of disease symptoms (mean time 8.2 years), 469 (76.3%) of the 650 participants had contacted one or more health professionals and half of them (324; 49.9%) had consulted at least one health professional in the last year. Approximately half of these patients (48.8%) were referred by a rheumatologist, a quarter of them (25.9%) contacted health professional themselves. The three most frequently visited health professionals were physiotherapist (40.0%), dental hygienist (11.4%), and podiatrist (9.2%) (Table 2). Approximately three quarters of all patients (76.3%) consulted at least one health professional since SSc onset for SSc-related problems and slightly more than half of these patients (56.6%) had contact with a physiotherapist.

Table 2: Health professional utilization by 650 patients with SSc

	Contacted in last 12 months N (%)	Contacted since onset SSc N(%)
Health professionals (all)	312 (41.1)	469 (76.3)
Physio therapist	260 (40.0)	367 (56.5)
Occupational therapist	58 (8.9)	155 (23.9)
Podiatrist	60 (9.2)	103 (15.9)
Hand therapist	18 (2.8)	58 (8.9)
Exercise therapist	17 (2.6)	28 (4.3)
Dietitian	51 (7.9)	108 (16.6)
Dental hygienist	74 (11.4)	95 (14.6)
Speech therapist	6 (0.9)	22 (3.4)
Psychologist	42 (6.5)	80 (12.3)
Social worker	15 (2.3)	64 (9.9)

Referral reasons and unmet needs in SSc care

Regarding the open-ended questions about referral reasons and treatment goals, we received a total of 697 encodable responses. We found that patients could not clearly distinguish between referral reasons (reflecting the rheumatologist's perspective) and treatment goals (reflecting the health professional's perspective) and consequently gave similar answers to both questions. Therefore, the responses of both questions were combined into one ("referral reasons") before initial coding. Within these responses, 143 different reasons for referral were identified and subsequently linked to 28 ICF-codes. The most common responses were related to the following ICF categories: pain in body part (38.9%), mobility of joint functions (28.7%), functions of the cardiovascular system (23.1%), functions of the skin and related structures (20.7%), and muscle functions (18.2%). The 15 most frequently mentioned referral reasons are shown in Table 3.

Table 3: Top 15 out of 27 different reasons for referral to non-pharmacological care (N=324)

Referral reason (ICF terms)	ICF code	N (%)
Body structures and functions		
Pain in body part	b2801	126 (38.9)
Mobility of joint functions	b710	93 (28.7)
Functions of the cardiovascular system, other specified and unspecified	b429	75 (23.1)
Functions of the skin and related structures, other specified	b898	67 (20.7)
Muscle functions, other specified and unspecified	b749	59 (18.2)
Neuromusculoskeletal and movement related functions	b7	30 (9.3)
Emotional functions	b152	27 (8.3)
Energy and drive functions	b130	16 (4.9)
Weight maintenance functions	b530	15 (4.6)
Respiration functions	b440	12 (3.7)
Blood vessel functions	b415	11 (3.4)
Activities and participation		
Self-care	d5	15 (4.6)
Hand and arm use	d445	34 (10.5)
Moving around in different locations	d460	12 (3.7)
Personal and environmental factors		
Assistive products and technology for personal use in daily living	e1251	11 (3.4)

Fatigue, Raynaud's phenomenon, physical limitations, reduced hand function, and joint problems were mentioned by more than 25% of all respondents as an unmet need in SSc care (Table 4). An analysis of potential associations of the number of unmet needs with disease duration, age, SSc subtype and education level revealed that participants with a lower level of education have on average 6.4% more unmet needs than participants in the higher educated group.

Table 4: Top 5 unmet needs compared to HP treatments aiming the specific unmet need

	More attention to … N (%)	Received non-pharmacological treatment in the last 12 months N (%)	Received treatment aiming at treatment goal related to unmet need N (%)
Fatigue	296 (45.6)	159 (24.5)	15 (5.1)
Raynaud's phenomenon	205 (31.6)	103 (15.9)	10 (4.9)
Physical limitations	193 (29.7)	119 (18,3)	93 (48.2)
Reduced hand function	177 (27.3)	100 (15.4)	23 (13.0)
Joint problems	163 (25.1)	82 (12.6)	81 (49.7)
No unmet needs	134 (20.7)	does not apply	does not apply

Alignment of reasons for referral and unmet needs

A relatively small percentage of the respondents (ranging between 4.9 and 13.0%) received non-pharmacological treatment addressing their specific unmet needs. Patients who had not reported any unmet need (20.7%) received a less frequently non-pharmacological treatment (Table 4).

Coordination and alignment of care

Nearly half of the 324 patients (*N* = 158, 48.9%) who received non-pharmacological treatment in 2017 perceived the collaboration between the rheumatologist and their health professional never or only sometimes as sufficient. Approximately two third of the patients (*N* = 214, 66.2%) reported insufficient agreements between the rheumatologist and the health professional, whereas more than half of the patients (*N* = 162, 50.2%) assumed that the advice given to the patient by the rheumatologist and health professional were never or rarely well-tuned (Table 5).

Table 5: Perceived quality of communication between patient, rheumatologist and HP, and outcome satisfaction with HP treatment (*N*=324)

	Always/ mostly	Never/ sometimes	I don't know
Perceived quality of communication			
Did your therapist, after your opinion, collaborate well with your rheumatologist?	56 (17.3)	158 (45.3)	109 (33.8)
How often did your rheumatologist and your therapist, in your opinion, make good agreements with each other?	27 (8.3)	214 (66.2)	82 (25.4)
How often did you think that the treatments and advices you received from your rheumatologist and your therapist were well tuned to each other?	60 (18.5)	162 (50.2)	101 (31.3)
How often was your rheumatologist aware of agreements you had with your therapist?	106 (32.9)	154 (47.7)	63 (19.5)
Outcome satisfaction			
Did your therapist have sufficient knowledge and expertise to treat you?*	240 (74.3)	36 (11.2)	47 (14.6)
Could you improve your daily activities through the treatment of your therapist?*	243 (75.2)	80 (24.8)	does not apply
Can you deal better with your complaints through the treatment of your therapist?*	243 (75.2)	80 (24.8)	does not apply

***Due to missing values, N=323**

Outcome satisfaction

A total of 240 (74.4%) out of the 324 respondents were satisfied with the knowledge and expertise of their health professionals regarding SSc treatment. The proportion of patients who could cope better with their complaints after the treatment and reported improvement in their daily activities was 73% (*N* = 156) (Table 5).

Discussion

The results of our study demonstrate that, from the patient's point of view, the reason for referral to health professionals was primarily the treatment of physical symptoms, such as mobility of joint functions and functions of the cardiovascular system. Reported unmet care needs as fatigue, Raynaud's phenomenon, and reduced hand function were not strongly covered by the referral reasons. Patients felt satisfied with health professional treatment content and outcomes. Despite this, communication and collaboration between rheumatologists and health professionals were rated rather low, and nearly one third of the patients was not able to judge the quality of communication between their rheumatologist and the health professional.

Our current study has shown that care for people with SSc is not yet optimal. We found three major areas that may be the causes of the different unmet needs for SSc care, which are as follows: underutilization of referrals to HP dealing with the psychosocial aspects of the disease, referrals that are not well aligned to the patients' unmet needs, and a suboptimal coordination and alignment of care.

Underutilization of non-pharmacological care services

Only approximately 50% of patients in our study used non-pharmacological care in the last year. Much of the reported referral reasons (reported by more than 30% of the patients) was related to treatment of physical symptoms. Referrals to occupational therapists, psychologists, and social workers, better equipped to address the psycho-social aspects of the disease, including emotional issues, impaired work, and decreased participation in social life, were much rarer[26]. This latter agrees with an earlier study of Willems and suggests that rheumatologists may be more likely to refer to physiotherapists and other HP disciplines who have a focus on the treatment of physical symptoms[16]. This strong focus on referrals to physical treatments possibly reflects obstacles from the following origins: rheumatologists, patients, and lack of evidence. Patients may not be aware enough of the possibilities of the non-pharmacological care. It is also possible that rheumatologists have a lack of knowledge of content and aims of non-pharmacological treatment options[15,16]. In addition, there is still a lack of strong evidence of the effectiveness of non-pharmacological treatment options10. However, since non-pharmacological treatments often do not focus on a specific disease, but rather on symptoms or limitations in activities, evidence for many non-pharmacological treatments originally intended for other rheumatic conditions could also be relevant in this patient group[27]. For instance the evidence for the effectiveness of treatments for commonly SSc specific problems such as fatigue, reduced hand function, and joint problems are already available in other rheumatological diseases[28-30]. HPs should take the opportunity and establish evidence-based recommendations for accessible and targeted non-pharmacological interventions.

Unmet needs

Along with the low number of referrals for psycho-social reasons, we found a limited alignment between unmet needs and reasons for (self)referral. Especially among patients who identified fatigue and Raynaud's phenomenon as an unmet care need, only a low percentage reported to actually be treated for this reason. Patients may hesitate to disclose certain topics during the consultation with the rheumatologist and therefore may not discuss their needs for information on non-pharmacological treatment options[14]. A recent study showed that patients with arthritis found it difficult to involve themselves in the decision making, often because they were unaware of having a choice[31]. This supports that the reported unmet care needs are not sufficiently addressed in daily SSc care and suggests that the use of care for SSc patients is still suboptimal. Psycho-social symptoms that are commonly experienced by SSc patients and have a major impact on daily activities and participation need to be considered as primary targets for interventions.

Coordination and alignment of care

In our study patients perceived the quality of communication and care coordination between rheumatologists and HPs as rather low. Well-coordinated and integrated care is considered as one of the eight important indicators of quality and safety, from the patient perspective30. It is not easy to offer SSc patients appropriate and well-coordinated care due to the complexity of the disease, the variability of the disease course, and the limited evidence-supported pharmacological and non-pharmacological treatment options[10,12,32].

However, poor communication and coordination, can create additional barriers to care access. SSc patients and their families are feel exposed to great barriers in access to and the quality of specialized and coordinated healthcare[16,33,34]. They describe themselves as being "passed around", have difficulties to find reliable information about their illness and treatment, and experience follow-up appointments logistically, physically and emotionally demanding. This in turn leads to emotional burden and frustration for the patients. This implies that, in daily practice, clinicians must invest even more in the quality of communication, particularly in the promotion of interdisciplinary communication. The use of patient decision aids leads to an increased communication and knowledge, more accurate risk perceptions, and a greater number of decisions consistent with SSc patients' values, and needs[35]. Our study underlines the importance to develop decision aids that support communication and may lead to decisions more consistent with the patients' needs.

Outcome satisfaction

In addition to the three areas of attention, we also found a supporting factor for the use of non-pharmacological care. In this study, patients perceived a high outcome satisfaction with non-pharmacological treatments, as well as high satisfaction with SSc specific knowledge and expertise of health professionals. They experienced improvement of daily activities and symptoms because of the non-pharmacological treatments. As far as we know, this is the first study describing the satisfaction with health professional treatment outcomes in SSc care from the patient perspective in such a large cohort. This underlines the added value of HPs in the treatment of problems that restrict SSc patients in daily activities, although there is not yet much evidence for non-pharmacological treatments.

Regarding our method, some limitations were found that may have influenced the described outcomes or their interpretation. Patients could not clearly distinguish between referral reasons and treatment goals. This might have led to a misinterpretation from the patients' perspective and made it impossible to distinguish between the rheumatologists' perspective as reflected in the referral reasons and the health professional treatment goals.

Another limitation of our study might be the relatively large percentage of respondents (58%) that were treated in hospitals specialized in SSc treatment. These patients may have different preferences than patients in small, local hospitals who did not participate.

Third, to recruit a large group of patients, we could only send one invitation without a reminder, which could explain the estimated response rate of 31%. However, the response rate will be slightly higher, as patients treated in shared care (39% of patients) could have received the invitation twice if both centers participated in the study. Compared to previous national and international SSc studies, the composition of our cohort is comparable in terms of demographic and disease specific characteristics. We found two minor differences that we believe do not affect the results of our study; namely large age range of the participants (18–87 years), which is often significantly narrower in comparable studies; and a relatively large percentage of participants, with an unknown SSc subtype (44.8%). However, this percentage is comparable with other surveys classifying patients in subtypes of SSc on the basis of self-report[10,36].

Conclusions

Reasons for referral, as well as communication and coordination of SSc care are not yet properly aligned between rheumatologists and health professionals and tuned to the patients' needs. Despite the high outcome satisfaction and the good accessibility of occupational therapists, psychologists, social workers, and hand therapists who are skilled to target unmet care needs such as psychological wellbeing, fatigue, daily functioning, and self-management, patients report relatively low utilization of health professional treatments. Our results suggest obstacles in the access to non-pharmacological care and barriers in communication between different (non-) pharmacological professionals. We recommend the development of easily accessible information and decision aids that give SSc patients and rheumatologists insights into the spectrum of non-pharmacological interventions and support the decision making for targeted referrals.

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Chapter 4

Opening the black box of non-pharmacological care in systemic sclerosis:
a cross-sectional online survey of Dutch health professionals

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Abstract

Purpose

To describe the spectrum of the health professional (HP) treatment approach for systemic sclerosis (SSc) from the perspective of Dutch HPs, including alignment of treatment goals set by HPs with self-reported referral reasons, coverage of patient-reported unmet care needs, and quality of communication between HPs and rheumatologists.

Method

Dutch HPs were invited through their patients with SSc to complete an anonymous online survey. The survey covered referral reasons, treatment goals, and interventions of the last patient treated, as well as the perceived quality of communication between HPs and rheumatologists. Referral reasons and treatment targets were linked to the International Classification of Functioning, Disability and Health following the refined ICF Linking Rules.

Results

Seventy-nine HPs from 8 professions (including 58 physiotherapists, 73%) completed the survey. One hundred and thirty-three different referral reasons were reported, yielding 58 different ICF codes, with 41 (70.7%) being linked to the ICF domain “body structures and functions.” The reported interventions focused on body functions/structures (27.9%), training of daily activities (25.6%), education and advice (26.3%), and psychosocial interventions (20.2%). The quality of communication between HPs and rheumatologists was perceived as low.

Conclusion

Our findings revealed numerous treatment options offered by Dutch HPs addressing the unmet care needs of patients with SSc. There is an overlap in the content of the various HP disciplines, and HP treatment goals are not sufficiently aligned with referrals of rheumatologists. HP treatment offer seemed inefficiently organized, possibly precluding rheumatologists from making targeted referrals. Communication between rheumatologists and HPs should be improved.

Introduction

Systemic sclerosis (SSc) is a rare and complex autoimmune disease with large differences in severity and extent. Its worldwide incidence is an estimated 13 people per million per year, and its prevalence is approximately 200 people per million[1]. SSc has a heterogeneous and often progressive nature that involves skin, vessels, joints, and internal organs, and it significantly impairs patients’ daily functioning and quality of life[2,3]. There is no effective treatment or cure for SSc yet, meaning that treatment is primarily aimed at controlling symptoms and maintaining quality of life[4]. As treatment options for life-threatening, organ-based complications improve, treatment approaches for nonfatal SSc complications require increased attention[5-7].

Due to the direct impact of SSc on daily functioning and psychosocial well-being of patients, non-pharmacological management and treatments are a key element of SSc care[8]. Health professionals in rheumatology (HPs), including occupational therapists, physiotherapists, psychologists, and social workers, play a vital role in the support of individuals with SSc manage their nonfatal SSc complications[9]. So far, no recommendations for the non-pharmacological care for SSc are formulated, but several high-quality randomized trials support the use of non-pharmacological treatment options to reduce the clinical burden of a variety of symptoms[6,10,11]. In addition, care by health professionals is also based on treatments proven to be effective in other rheumatic diseases. For instance, promising approaches to address fatigue in patients with RA and SLE are also applicable for patients with SSc[12-14].

In the past decades, owing to changes in the Dutch health care system, HP treatment has been transferred from hospital-based team care to a primary care setting. As a result, patients with SSc have more often been referred to HPs working in monodisciplinary primary care settings. Considering that, rheumatologists have more confidence in HP colleagues with whom they work on a daily basis in the same institution[15]. This development may have negatively affected rheumatologists’ knowledge of HP treatment options, adequate coordination of treatment, and the quality of communication between rheumatologists and HPs.

SSc patients consider non-pharmacological care as one of the five main issues affecting the quality of SSc care in need of improvement[9,16,17]. Spierings et al. identified the following the top five unmet care needs of patients with SSc: fatigue, Raynaud’s phenomenon, physical limitations, and impaired hand and joint function[17]. It remains unknown to what extent these five unmet care needs are addressed by HPs in the treatment of patients with SSc.

Therefore, the aim of our study was to examine the content and alignment of care delivered by Dutch HPs with patients’ most important needs.

Methods

Study design

A cross-sectional study using a web-based survey (SurveyMonkey®.com) was conducted to make an inventory of perceptions of Dutch HP treating patients with SSc. This study was reported according to the Strengthening the Reporting of Observational Studies in Epidemiology (STROBE) and using the Checklist for Reporting Results of Internet E-Surveys (CHERRIES)[18,19].

Survey

The survey questions were based on several preliminary investigations conducted by the Arthritis Research and Collaboration Hub (ARCH) working group: a literature review, three semi-structured multicenter focus group interviews, and individual interviews among patients, HPs, and rheumatologists[17].

The 23 survey questions were distributed over 14 webpages and divided into 4 domains: socio-demographic and work setting-related characteristics [12], referral to non-pharmacological care [2], treatment [5], and perceived quality of communication [4]. The survey included both open-ended questions, asking the participants to answer in their own words, and closed questions, providing multiple-choice and multiple-response questions.

Socio-demographic and work setting-related questions

The survey started with 12 socio-demographic and work setting-related questions: sex (woman, man); age (free text); educational level (bachelor, master, Ph.D., and others); work experience (free text); profession (dietitian, occupational therapist, physiotherapist, hand therapist, speech therapists, social worker, dental hygienist, exercise therapist, podiatrist, psychologist, and others); caseload of SSc patients in the past year (0–2, 3–6, 7 or more); SSc specialization (yes/no); working hours per week (32 or more, 20–31, 12–19, others); work domains of the past 5 years (patient care, research, education, management, and others); current work setting (academic hospital, regional hospital, health center, private practice, nursing home, rehabilitation center, and others); SSc specialization of work setting (yes, no); and participation in multidisciplinary SSc consultations (yes, no, and others).

Referral to non-pharmacological care

Types of referrer were assessed by means of a list of seven medical disciplines and an option to add new items. With the following answering format: never, sometimes, and always. Most common reasons for SSc referrals, as reported by HPs, were assessed by an open-ended question with three options for free text responses.

Treatment

Five questions assessed the HP treatments. HPs were asked to consider the last SSc patient treated to assess the following items: type of SSc (limited SSc, diffuse SSc, I do not know, others), main treatment goals (open-ended question with 3 options for free text responses), main interventions (multiresponse question divided into 4 domains, body structure and functions (20 items), activity and participation (9 items), education and advice (20 items), and psychosocial interventions (12 items). These multiresponse questions were used to prioritize items, participants could choose a maximum of three options, including an option to add a new item. Duration of HP treatment was assessed with two free text questions: duration in weeks and number of treatment contacts.

Perceived quality of communication

Perceived quality of communication was assessed by adapting four items of the Dutch version of the Consumer Quality Index Rheumatoid Arthritis (CQI-RA) (version 2.0), subscale ‘Communication’. The CQI-RA was found to be a reliable measure for patients’ experiences with the quality of rheumatic care20. For our study, we used the items ‘Parallel treatments were adjusted to one another’, ‘Various advises were integrated’, ‘Caregivers kept their appointments’ and ‘Caregivers were aware of other activities of caregivers’ and adapted them to measure the experiences of HP (see Table 4). The answering format of the items was: never, sometimes, usually, and always.

The survey was evaluated by members of the ARCH SSc working group and a patient panel of five patients. Only an individual code and Internet Protocol (IP) address was registered to guarantee the anonymity of the participants. Pilot testing of the questionnaire was undertaken in five HPs to ensure the relevance of the questions[17].

Sampling strategy

Sampling followed a targeted snowball sampling strategy[21]. Dutch HPs from different disciplines (including physiotherapists, occupational therapists, podiatrists, hand therapists, dietitians, dental hygienists, speech therapists, psychologists, and social workers) were eligible to participate in the study, if they were currently treating or had previously treated patients with SSc. There were no participation restrictions on the workplace, the case load, or the working environment. HPs were invited by their own patients with SSc who participated in a large-scale survey among 650 Dutch patients with SSc set up by the ARCH working group. Patients with SSc who participated in the study were asked to ‘snowball’ their treating HPs by providing them with an internet link we offered, or by writing down the name and address of the workplace of the HP, enabling us to invite the HP to take part in the study. An estimation of the sample size was not possible due to snowballing as sampling strategy and the unknown number of HPs working with SSc patients in the Netherlands. Eligible participants had 4 months to voluntarily complete the survey (December 2017 to March 2018). The survey link was open from the time the participants were first informed about the study. The cover letter, displayed on the first page of the survey, provided details about the background and purpose of the survey, along with the estimated duration of the survey (15 min). Informed consent was taken at the beginning of the survey.

Data handling and confidentiality

IP address checks have been performed to avoid duplicate answers from one respondent. The data processing was completely anonymous, with the IP addresses remaining with the first and corresponding author. A second author (CHME) had access to the individual codes and synthesized data without associated IDs. Only completed surveys were included in the analyses.

Ethical approval was obtained from the Institutional Review Board of the Radboudumc Nijmegen, the Netherlands, protocol (2017: 3621).

Data analysis

Statistical analysis

Socio-demographic and work setting-related data, HP interventions and perceived quality of communication were analyzed descriptively. Continuous variables, following a normal distribution, were reported as means and SD and categorical variables as absolute numbers and percentages. Statistical analyses were conducted using Stata/IC 13.1 (StataCorp LP, College Station, TX).

Analysis of open-ended questions

The qualitative data analysis of the answers to open-ended questions followed an adapted form of “meaning condensation”[22]. First, all answers to the open questions about referral reasons and treatment goals were read through by the principal investigator (JS) to obtain an overview of the collected data. Second, all data were divided in ‘meaning units’, defined as specific text units, either a few words or a part of a sentence with a common meaning. Third, concepts within each meaning unit were identified. Sometimes one meaning unit could contain several concepts. For instance, the meaning unit, “Staying fit so that my client can keep walking > 5 km.” contains the concepts ‘maintaining physical fitness’ and ‘walking longer distances’. All resulting concepts were linked to the most appropriate ICF category according to established linking rules[23,24]. The purpose of the matching process was to translate the concepts from the HPs’ answers into the most appropriate ICF categories. The ICF classification uses a hierarchical structure organized in chapters, or ‘first level’ categories, which subdivide the four separate concepts of body functions, Body structures, activities and participation and environmental factors. Each chapter contains numerous categories (second, third, and fourth levels), which form the classification unit. The specificity increases from the first to the fourth level. As an example, the concept ‘walking longer distances’ was linked to d450 Walking. ‘Maintaining physical fitness’ was linked to d5701 Managing diet and fitness.

In accordance with the linking rules, interactive discussions were held to resolve coding discrepancies (JS and CHME). Finally, all assigned ICF codes were re-read repeatedly by the main coder (JS) to ensure that the linked ICF codes reflected the meaningfulness of the concept. Through this process, the large number of answers to the open questions on referral reasons and treatment goals were reduced to a smaller amount of clearly defined ICF terms. These were used to compare treatment goals with reasons for referral and unmet care needs.

Results

Participants, origin, and content of referrals

We obtained 81 completed surveys. One duplicate response set was identified and excluded from the analysis, and another set was excluded for not meeting the inclusion criteria; the person was a medical doctor. Thus, data from 79 surveys taken by eight HP professions were analyzed. Table 1 presents the HPs’ socio-demographic and work-related characteristics.

The larger proportion of participants was female (*n* = 52; 67%). Physiotherapists were the largest group represented (*n* = 58; 73%), followed by dietitians (*n* = 6; 8%) and occupational therapists (*n* = 5; 6%). Nineteen (24%) of the respondents reported to have treated 3 or more patients with SSc in the past year. Most HPs (*n* = 60, 69%) worked in private practices. In all, 21 (26.6%) HPs felt specialized in SSc care, and 11 (13.9%) found that their workplace was specialized in SSc. Only six HPs (5.6%) regularly participated in multidisciplinary SSc meetings. HPs reported that rheumatologists were the most frequent referrers (*n* = 56, 73.7%). Nearly one-third (*n* = 22, 29.0%) of the reported referrals were patient self-referrals. All other referrals were distributed among general practitioners (*n* = 14, 18.4%), dermatologists (*n* = 4, 5.3%), other medical specialists (*n* = 14, 18.5%) and other HPs (*n* = 4, 5.3%).

Table 1: Characteristics of 79 health professionals working with patients with SSc and frequency of referrals from different sources

Characteristics			
Female, N (%)			52 (65.8)
Age, years; mean (SD), range			41.2 (13.6), 22–82
Education level, N (%)			
Bachelor diploma			53 (67.1)
Master diploma			22 (27.9)
PhD			4 (5,1)
Patients with SSc per year, N (%)			
0–2 patients			60 (76,0)
3–6 patients			14 (17,7)
≥7 patients			5 (6.3)
Specialized in SSc treatment N (%)			21 (26,6)
Institution/practice specialized in SSc treatment N (%)			11 (13.9)
Regular participation in multidisciplinary consultation of patients with SSc N (%)			6 (5.6)
Profession N (%)			
Physiotherapist			58 (73.5)
Dietitian			6 (7.6)
Occupational therapist			5 (6.3)
Podiatrist			4 (5,1)
Skin therapist			3 (3.8)
Speech- and language therapist			1 (1.3)
Dental hygienist			1 (1.3)
Psychologist			1 (1.3)
Practice setting N (%)*			
Private practice			60 (69.0)
Hospital or treatment center			27 (31.1)
School/university			2 (2.3)
Other			7 (8.1)
Category of work during the last 5 years N (%)*			
Clinical patient care/rehabilitation			83 (73.6)
Education			13 (11.6)
Management			12 (10,7)
Research			8 (7,1)
Years worked in clinical practice as a health professional, years; mean (SD), range			16.9 (12.2), 0.5–42.0
Frequency of HP referrals from different sources N (%)*,**			
	Never	**Some**	**Most/all**
General practitioner	62 (81.6)	13 (17.1)	1 (1.3)
Rheumatologist	20 (26.3)	41 (54.0)	15 (19.7)
Dermatologist	72 (94.7)	3 (4.0)	1 (1.3)
Other medical specialist	62 (81.6)	11 (14.5)	3 (4.0)
Other health professional	72 (94.7)	4 (5.3)	0 (0.0)
Self-referral	54 (71.1)	17 (22.4)	5 (6.6)

* multiple answers possible; **n=76

The 129 concepts on referral reasons, collected from open-ended questions, could be linked to 47 unique ICF codes and included 31 ICF codes on Body structures and functions (89 concepts), 13 on Activities and participation (36 concepts), and 3 on Environmental factors (4 concepts). Table 2 presents the ten most frequently mentioned referral reasons together with the reporting disciplines. Seven of the ten most frequently cited referral reasons were aimed at Body structures and functions. In addition, up to four HP disciplines received referrals with identical referral reasons.

Table 2: Ten most frequently mentioned referral reasons and reporting disciplines

	ICF code		N	N per HP discipline* reporting the referral reason
1	Aerobic capacity	b4551	11	11 PT
2	Sensation of pain	b280	8	8 PT
3	Mobility of joint functions	b710	8	6 PT, 1 ST, 1 P
4	Carrying out daily routine	d230	8	6 PT, 2 OT
5	Respiration functions	b440	7	7 PT
6	Managing one's own activity level	d2303	7	4 PT, 3 OT
7	Energy level	b1300	6	3 PT, 1 OT, 1 D, 1 P
8	Weight maintenance functions	b530	6	6 D
9	Muscle power functions	b730	5	5 PT
10	Hand and arm use	d445	5	1 PT, 1 HT, 3 ET

*PT=Physio therapist, ST=Skin therapist, P=Podiatrist, OT=Occupational therapist, D=Dietitian

Treatment goals and interventions

Analysis of the reported treatment goals revealed 209 concepts that could be coded into 66 unique ICF codes. Most of the treatment goals were aimed at Body structures and functions (*n* = 35 ICF codes, consisting of 119 concepts), a smaller part focused on Activities and participation (*n* = 27 ICF codes, consisting of 86 concepts) and only a small amount of the treatment goals aimed at Environmental factors (*n* = 4 ICF codes, consisting of four concepts). Nine participants did not report any treatment goals. Table 3 shows the ten most frequently mentioned treatment goals, together with the number of disciplines that reported the respective treatment goal.

Table 3: Ten most frequently mentioned treatment goals and reporting disciplines

	ICF code		N	N per HP discipline* reporting the referral reason
1	Aerobic capacity	b4551	25	25 PT
2	Managing daily routine	d2301	15	13 PT, 2 OT
3	Managing one's own activity level	d2303	15	12 PT, 3 OT
4	Mobility of several joints	b7101	12	11 PT, 1 P
5	Muscle power functions	b730	12	12 PT
6	Sensation of pain	b280	11	9 PT, 2 P
7	Managing diet and fitness	d5701	9	4 PT, 5 D
8	Other functions of the skin	b830	7	6 PT, 1 ST
9	Moving around	d455	6	6 PT
10	Hand and arm use	d445	6	6 PT

*PT=Physiotherapist, ST=Skin therapist, P=Podiatrist, OT=Occupational therapist, D=Dietitian

A total of 605 interventions (8.8 average per participant) were reported, with the treatment focus more or less evenly distributed across the following 4 components: Bodily functions/structures (27.9%), Training of activities (25.6%), Education/advice/instruction (26.3%), and Psychosocial interventions (20.2%). The most frequently mentioned interventions (top four per topic) are presented in Table 4. Within these most frequently mentioned interventions, we found five interventions or strategies that are applied by up to six different HP disciplines: walking/cycling (4), exercise activities/sport (6), household (5), self-management/self-monitoring (4), and motivational interviewing (5) (Table 4).

Table 4: Interventions applied by the 79 HPs, top 4 per topic

Intervention	N (%)	Number of HP per discipline* focusing on the intervention (disciplines N)
Body functions and/or structures		
Physical activity promotion	38 (48.1)	33 PT, 1 OT, 4 D
Training of body functions (e.g., muscular strength, range of motion)	34 (43.0)	34 PT
Aerobic capacity training	25 (31.7)	25 PT
Balance/coordination training	14 (17.7)	13 PT, 1 P
Activities		
Walking/biking	45 (57.0)	39 PT, 3 D, 2 P, 1 S (4)
Movement activities/sports	45 (57.0)	37 PT, 3 D, 2 HT, 1 S, 1 P, 1 OT (6)
Leisure activities	19 (24.1)	17 PT, 1 ET, 1 ST
Household	16 (20.3)	11 PT, 2 OT, 1 HT, 1 D, 1 P (5)
Education/advice/instruction		
Graded activity	44 (55.7)	38 PT, 4 OT, 1 HT, 1 P (4)
Physical activity	42 (53.2)	39 PT, 1 D, 2 HT
Lifestyle (e.g., smoking, cold, silver gloves)	14 (17.7)	12 PT, 2 OT
Energy conservation	12 (15.2)	10 PT, 1 OT, 1 D
Psychosocial interventions		
Self–management/self–monitoring	53 (67.1)	42 PT, 4 OT, 3 D, 2P, 2 KT (5)
Relaxation strategies/stress management/biofeedback therapy	23 (29.1)	20 PT, 2 OT, 1 D
Motivational Interviewing	13 (16.5)	6 PT, 3 OT, 2 D, 1 DH, 1 P (5)
Problem–solving training	10 (12.7)	9 PT, 1 S

*PT=Physiotherapist, P=Podiatrist, OT=Occupational therapist, D=Dietitian, HT=Hand therapist, DH=Dental hygienist, S=Speech therapist, ST=Skin therapist

Alignment of referral reasons and HP treatment goals

In all, 17 of 129 (13.2%) referral reason concepts matched with one of the treatment goal concepts at the patient level. In 10 cases, referral reasons fully matched with treatment goals. The ICF codes d230/2303 (Carrying out daily routine/Managing one's own activity level) corresponded in four cases, whereas b4551 (Aerobic capacity), and b280 (Sensation of pain) corresponded in two cases. The other corresponding codes were: b4550/b4551 (General physical endurance/Aerobic capacity), b710/b7101 (Mobility of joint functions/Mobility of several joints), b730 (Muscle power functions), d445 (Hand and arm use), s320/s3200 (Structure of mouth/Teeth), and s7502 (Structure of ankle and foot).

Correspondence between treatment goals and unmet care needs

The examined unmet care needs *fatigue, Raynaud's phenomenon, joint problems, physical function, and hand function* were covered by 108 out of the total of 209 ICF codes extracted from reported HP treatment goals. Since the unmet care needs described relate to physical symptoms and not to the transcending health information such as situations and daily activities, almost exclusively ICF codes from chapters b (Body functions) and s (Body structures) could be assigned.

In 57 (81.4%) of the 70 cases in which participants provided information about treatment goals, we found ICF codes directly associated with 1 or more of the 5 unmet care needs. In half of all cases, we found agreement with the unmet care need *fatigue* (*n* = 16, 22.9%), *Raynaud's phenomenon* (*n* = 12, 17.1%), *joint problems* (*n* = 18, 25.7%), *physical function* (*n* = 35, 50.0%), and *hand function* with nine associable cases (12.9%). In 13 cases, we did not find a direct connection with 1 of the 5 unmet care needs; 7 of them concerned the participating dieticians and the 1 of them the only participating oral hygienist.

Quality of communication between HP and rheumatologists

Figure 1 illustrates the percentage of participants' perceptions on the quality of communication. Nearly one-third (29%) of those questioned could not make any statements about the cooperation and/or the quality of communication. Overall, slightly above 40% of the participants had a positive view about the quality of communication. One-quarter of the HPs reported that they are mostly satisfied with the agreements they have with the rheumatologists. Almost 40% of HPs rarely or never inform the rheumatologist about the goals, progress, and outcomes of their treatment.

Figure 1: Quality of communication between HPs and rheumatologists, %

Discussion

This cross-sectional survey study revealed that HPs use a broad spectrum of treatment goals (unique ICF codes, *n* = 66) and interventions (*n* = 51). At the HP group level, the ten most common referral reasons and treatment goals were considerably similar. However, analysis at an individual level indicated discrepancies between the self-reported referral reasons provided by the HPs and the reported treatment goals, suggesting insufficient alignment between referral reasons and treatment goals. On the other hand, we demonstrated that HP treatment goals indeed match the most important unmet care needs of SSc patients reported in the literature. Another critical finding was that relatively few HPs communicated with the rheumatologists and only some HPs reported to have agreements with rheumatologists, implying a poor quality of communication between HPs and rheumatologists.

Missing coherence between referral reasons and HP approach

We found discrepancies between rheumatologists' referral reasons and the reported HP treatment goals and interventions. HPs report interventions that are not mentioned in the referral reasons such as education, psychosocial interventions, and interventions aimed at social or environmental factors. A possible explanation for this may be insufficiently targeted referrals by rheumatologists. One study among Dutch rheumatologists specialized in SSc, the rheumatologists indicated to be insufficiently aware of the non-pharmacological treatment options15. In the absence of available evidence-based guidelines, practice-based evidence recommendations based on consensus could be a good option to share information about existing HP treatment options with referrers and patients. In addition, practice-based or consensus-based non-pharmacological recommendations could also be a good adjunct to the next update of the European League Against Rheumatism (EULAR) recommendations for the treatment of SSc of Kowal-Bielecka et al.[25].

Good correspondence between treatment goals and unmet care needs and SSc-ICF core set

Our results establish that the reported treatment goals covered the five most important unmet care needs of patients with SSc: fatigue, Raynaud's phenomenon, joint problems, physical function, and hand function described by Spierings et al. (2019). Our findings complement those of a European study by Willems et al. on the content of HP SSc care identifying fatigue, Raynaud's phenomenon, and hand function as the most important treatment goals[26]. These results suggest that HPs are indeed able to identify the care needs of patients with SSc. Despite these promising results, written consensus- and evidence-based recommendations need to be established to make the possibilities of HP care more visible for patients with SSc and rheumatologists.

Large overlap in interventions

Our results reveal that in some cases up to six disciplines indicate that they focus on the same areas of intervention. Due to the quantitative nature of our study, it is unclear whether they actually offer the same interventions or whether they are working with a different focus and intervention strategy. This overlap of the intervention offer could make it difficult for referrers to refer patients with SSc targeted to the best matching HP discipline because the spectrum of interventions offered is large but without clear distinctions. Studies with a qualitative approach could help to further specify the content of the interventions offered and allow referrers to make more targeted referrals to the most appropriate HP disciplines[27].

Quality communication between HPs and rheumatologists

Our results suggest a suboptimal communication of the HPs with the rheumatologists. Due to transitions of the health care system in the Netherlands, the work setting of HPs delivering care for patients with rheumatic diseases moved from larger hospitals to primary care setting. As a result, possibilities for specialization and multidisciplinary collaboration for HPs in the Netherlands have thinned, thereby reducing direct interaction of HPs with their medical and other HP colleagues. Due to the broad alignment with different target groups in primary care, there is a decrease in specialized HPs for the treatment of rare disorders. This new situation requires new models of care because the complex situation of people with SSc requires specialized care. A digital network, such as ParkinsonNet[28], could be a possible component of such a new care model. ParkinsonNet is a network of more than 3400 specialized health care providers with national coverage in the Netherlands. The model of ParkinsonNet has also been adopted in other countries[29]. Such a network could connect patients and the various health care providers in a targeted manner and thereby increase communication, the quality of multidisciplinary collaboration, and thus the quality of SSc care.

This study had a number of limitations. One limitation was the rather low response rate. We expected that by approaching the HPs through the 650 SSc patients who had participated in the previous survey study, a larger number of HPs would be reached. A possible explanation for the relatively small number of participating HPs is that they, although invited by their patients, subsequently did not participate because they felt insufficiently specialized in SSc. This explanation is supported by the low number of HPs (around 25%) that reported to feel specialized in SSc treatments. The second limitation is the limited use of validated questionnaires, for instance to examine the heterogeneity of interventions. Another limitation might be that our results on referral reasons are based on self-report by HPs, which could lead to recall bias. A content analysis of referral letters would be an option to obtain more reliable information.

Conclusion

We found a broad spectrum of treatment options offered by Dutch HPs that address the unmet care needs of patients with SSc. An overlap in the content of the care delivered by the various HP disciplines was noted, and the referrals of rheumatologists were not sufficiently aligned with HP treatment goals. The HP offer seems to be inefficiently organized, which may prevent rheumatologists from making targeted referrals. Strategies for better communication between rheumatologists and HPs should be developed and implemented.

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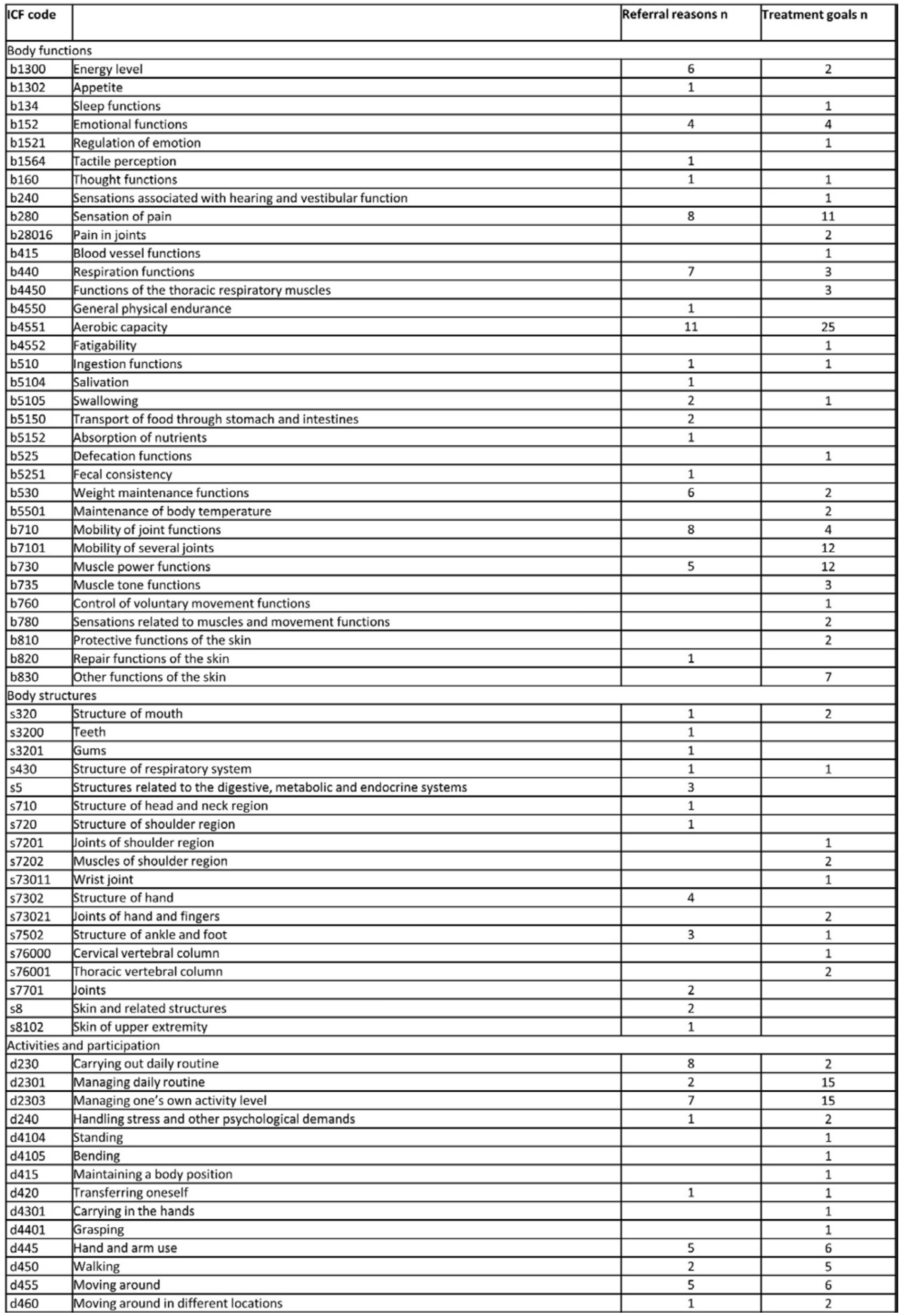
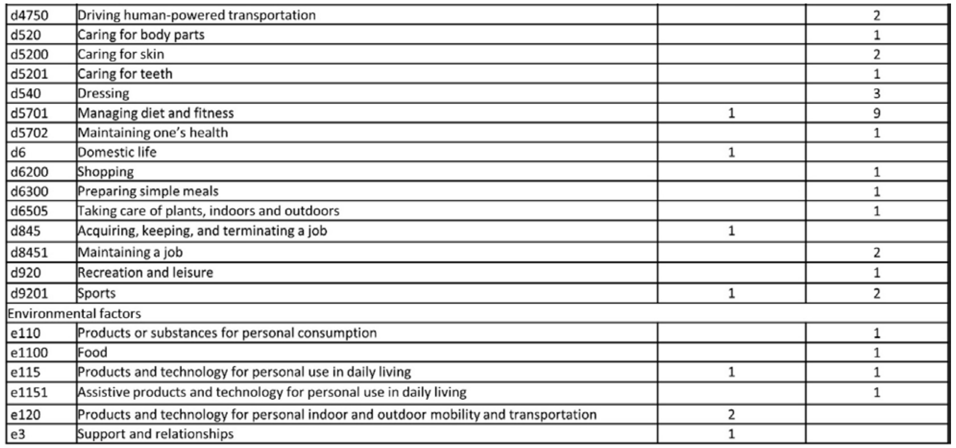
Appendix 1:

Reasons for referral and treatment goals as reported by 79 HPs, expressed in ICF codes





ICF code		Referral reasons n	Treatment goals n
Body functions			
b1300	Energy level	6	2
b1302	Appetite	1	
b134	Sleep functions		1
b152	Emotional functions	4	4
b1521	Regulation of emotion		1
b1564	Tactile perception	1	
b160	Thought functions	1	1
b240	Sensations associated with hearing and vestibular function		1
b280	Sensation of pain	8	11
b28016	Pain in joints		2
b415	Blood vessel functions		1
b440	Respiration functions	7	3
b4450	Functions of the thoracic respiratory muscles		3
b4550	General physical endurance	1	
b4551	Aerobic capacity	11	25
b4552	Fatigability		1
b510	Ingestion functions	1	1
b5104	Salivation	1	
b5105	Swallowing	2	1
b5150	Transport of food through stomach and intestines	2	
b5152	Absorption of nutrients	1	
b525	Defecation functions		1
b5251	Fecal consistency	1	
b530	Weight maintenance functions	6	2
b5501	Maintenance of body temperature		2
b710	Mobility of joint functions	8	4
b7101	Mobility of several joints		12
b730	Muscle power functions	5	12
b735	Muscle tone functions		3
b760	Control of voluntary movement functions		1
b780	Sensations related to muscles and movement functions		2
b810	Protective functions of the skin		2
b820	Repair functions of the skin	1	
b830	Other functions of the skin		7
Body structures			
s320	Structure of mouth	1	2
s3200	Teeth	1	
s3201	Gums	1	
s430	Structure of respiratory system	1	1
s5	Structures related to the digestive, metabolic and endocrine systems	3	
s710	Structure of head and neck region	1	
s720	Structure of shoulder region	1	
s7201	Joints of shoulder region		1
s7202	Muscles of shoulder region		2
s73011	Wrist joint		1
s7302	Structure of hand	4	
s73021	Joints of hand and fingers		2
s7502	Structure of ankle and foot	3	1
s76000	Cervical vertebral column		1
s76001	Thoracic vertebral column		2
s7701	Joints	2	
s8	Skin and related structures	2	
s8102	Skin of upper extremity	1	
Activities and participation			
d230	Carrying out daily routine	8	2
d2301	Managing daily routine	2	15
d2303	Managing one’s own activity level	7	15
d240	Handling stress and other psychological demands	1	2
d4104	Standing		1
d4105	Bending		1
d415	Maintaining a body position		1
d420	Transferring oneself	1	1
d4301	Carrying in the hands		1
d4401	Grasping		1
d445	Hand and arm use	5	6
d450	Walking	2	5
d455	Moving around	5	6
d460	Moving around in different locations	1	2
d4750	Driving human-powered transportation		2
d520	Caring for body parts		1
d5200	Caring for skin		2
d5201	Caring for teeth		1
d540	Dressing		3
d5701	Managing diet and fitness	1	9
d5702	Maintaining one’s health		1
d6	Domestic life	1	
d6200	Shopping		1
d6300	Preparing simple meals		1
d6505	Taking care of plants, indoors and outdoors		1
d845	Acquiring, keeping, and terminating a job	1	
d8451	Maintaining a job		2
d920	Recreation and leisure		1
d9201	Sports	1	2
Environmental factors			
e110	Products or substances for personal consumption		1
e1100	Food		1
e115	Products and technology for personal use in daily living	1	1
e1151	Assistive products and technology for personal use in daily living		1
e120	Products and technology for personal indoor and outdoor mobility and transportation	2	
e3	Support and relationships	1	

Chapter 5

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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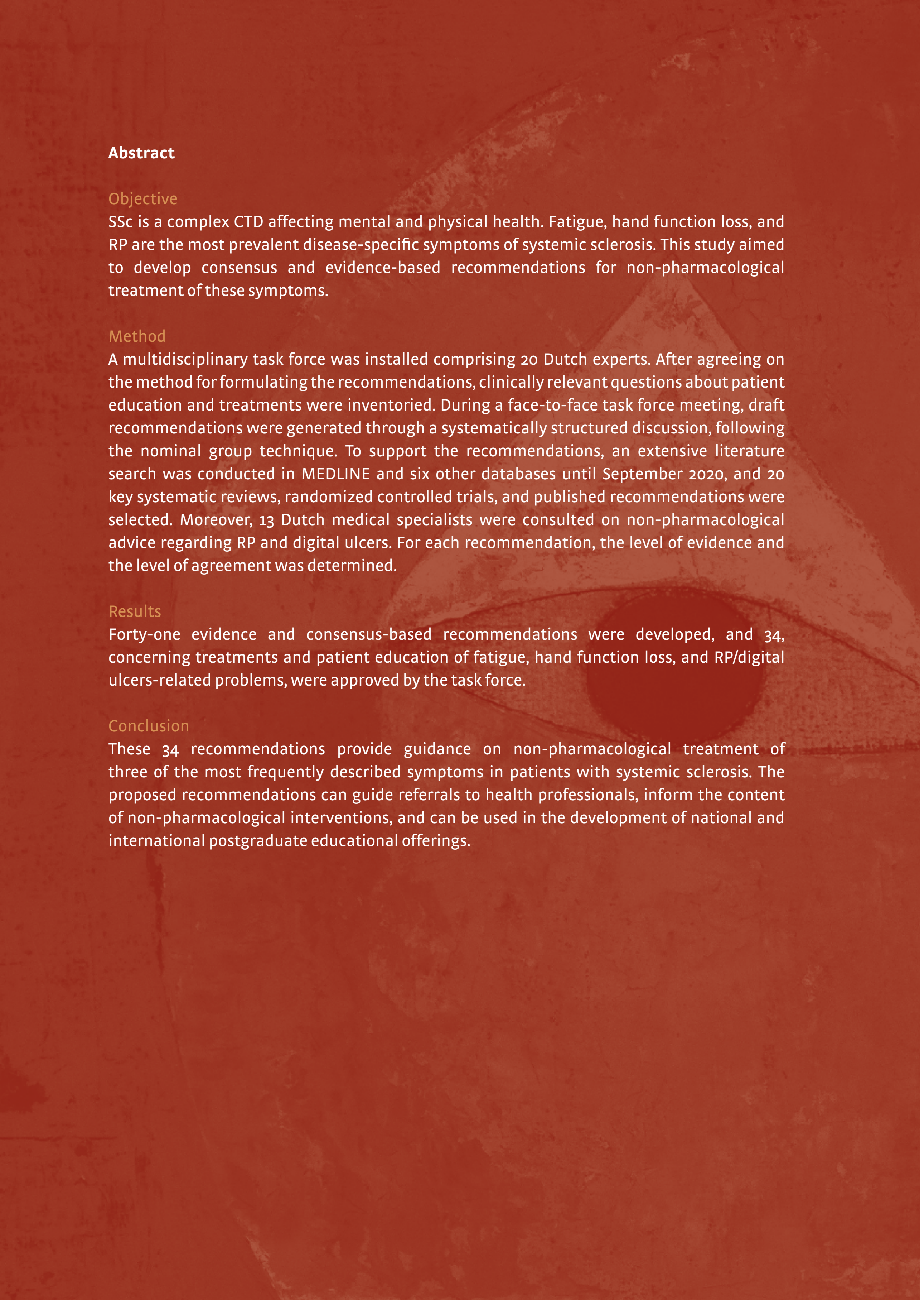


Abstract

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.

Method

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.

Conclusion

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.

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

Study 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 centers 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, S.D., median, and range of the level of agreement for each recommendation were calculated. A recommendation was approved when ≥70% of the expert group indicated a score of ≥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. Appendix 1 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. Appendix 2 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. Appendix 3 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.

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

		Level of Evidence	Reference	Level of Agreement (0-10) Mean (SD); 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.	Cognitive behavioural therapy (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 (SD); 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.	A 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. CBT: cognitive behavioural therapy.

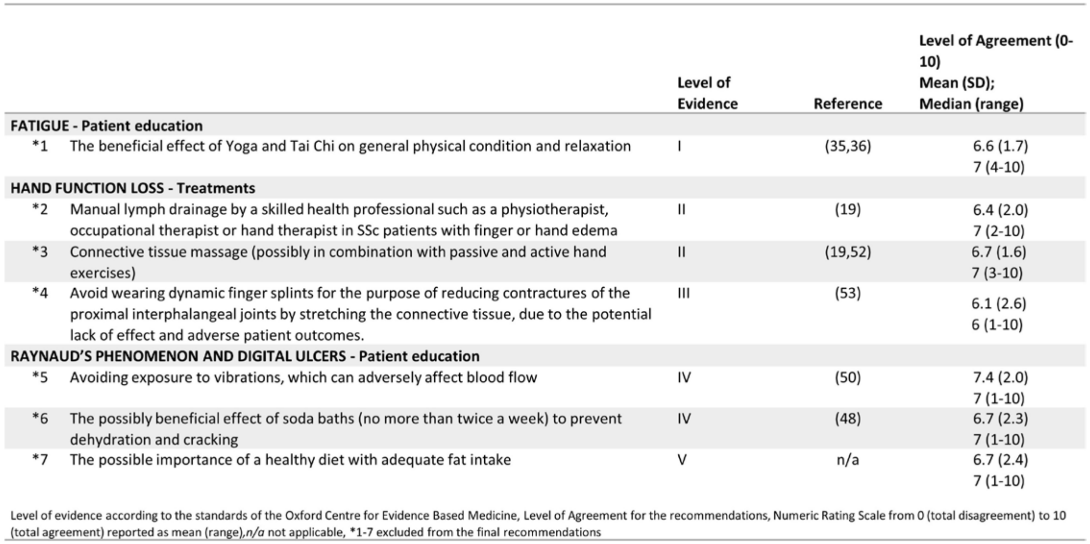
Table 3: Recommendations on patient education and treatments for SSc patients with Raynaud's phenomenon and/or digital ulcers

		Level of Evidence	Reference	Level of Agreement (0-10) Mean (SD); 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. 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. CBT: cognitive behavioural therapy.

Seven of the 41 draft recommendations did not meet the approval criteria of an agreement of ≥7 in ≥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.

Table 4: Draft recommendations which were excluded from the final recommendations in phase 4 through the determination of the level of agreement bij the task force members and medical experts



		Level of Evidence	Reference	Level of Agreement (0-10) Mean (SD); 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 edema	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)
RAYNAUD'S PHENOMENON 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

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 centers 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 care54. 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 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 focusing 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 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.

Conclusions

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.

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Appendix 1:
Search strategy

1	**systemic sclerosis[Mesh]**
2	diffuse scleroderma[Mesh]
3	limited scleroderma[Mesh]
4	syndrome CREST[Mesh]
5	localized scleroderma[Mesh]
6	systemic sclerosis
7	scleroderma
8	SSc
9	Skin
10	1 or 2 or 3 or 4 or 5 or 6 or 7 or 8 or 9
11	“Connective tissue disease*” or CTD
12	“Systemic lupus erythematosus” or SLE
13	“Rheumatoid arthritis” or RA
14	Osteoarthritis or OA
15	“Raynaud’s phenomenon” or “Raynaud’s syndrome” or RP
16	11 or 12 or 13 or 14 or 15
17	"non pharmacological intervention*"
18	"non pharmacological treatment*"
19	rehabilitation[Majr] or rehabilitation[subheading] or rehabilitation
20	"multidisciplinary treatment*"
21	("multicomponent treatment*" or "interdisciplinary treatment*" or "multimodal treatment”)
22	("multidisciplinary intervention*" or "multicomponent intervention*" or "interdisciplinary intervention*" or "multimodal intervention”)
23	17 or 18 or 19 or 20 or 21 or 22
24	physical therapy modalities[Mesh] or "physical modalities"
25	exercise therapy[Mesh] or exercise*
26	Intervention*
27	hydrotherapy[Mesh] or hydrotherapy
28	balneology[Mesh] or balneotherapy
29	range of motion, articular[Mesh] or "range of motion"
30	"physical therapy" or "physical therapies" or "physiotherapy*" or “physio therapy”
31	massage or "connective tissue massage"
32	mobilization or "skin mobilization"
33	training
34	thermotherapy
35	stretching
36	"paraffin wax" or paraffin
37	24 or 25 or 26 or 27 or 28 or 29 or 30 or 31 or 32 or 33 or 34 or 35 or 36
38	cognitive therapy[Mesh]
39	behavior therapy[Mesh]
40	psychotherapy[Mesh]
41	counseling[Mesh] or counseling or counselling
42	vocational rehabilitation[Mesh] or "vocational rehabilitation"
43	health education[Mesh]
44	“patient education”
45	Self care[Mesh] or “self-care”
46	"cognitive behaviour therapy" or "cognitive behavior therapy" or "cognitive behaviour therapies" or "cognitive behaviour therapies" or CBT
47	patient education
48	"self management" or “self-management”
49	38 or 39 or 40 or 41 or 42 or 43 or 44 or 45 or 46 or 47 or 48
50	occupational therapy[Mesh] or "occupational therapy" or "occupational therapies"
51	“hand therapy” or “hand therapist”
52	splints[Mesh]
53	leisure activities[Mesh] or "leisure activities"
54	activities of daily living[Mesh] or "activities of daily living" or “ADL” or “iADL”
55	self-help devices[Mesh] or "self-help device*" or “adaptive devices”
56	orthotics or orthotic* or brace*
57	"joint protection"
58	"activity pacing"
59	50 or 51 or 52 or 53 or 54 or 55 or 56 or 57 or 58
60	Rehabilitation nursing[Mesh] or "rehabilitation nursing" or nurse or nursing or “rheumatology nurse”
61	60
62	nutrition therapy[Mesh]
63	nutritional support[Mesh]
64	nutrition or diet*
65	dietitian*
66	62 or 63 or 64 or 65
67	podiatry[Mesh] or podiatry or podiatrist
68	"foot care"
69	67 or 68
70	oral hygiene[Mesh] or "oral hygiene"
71	dental hygiene[Mesh] or "dental hygiene"
72	70 or 71
73	Fatigue or fatigued
74	Tiredness or tired

75	Exhaustion or exhaust*
76	„Mental exertion“
77	„Physical exertion
78	Energy
79	"energy conservation"
80	rest
81	73 or 74 or 75 or 76 or 77 or 78 or 79 or 80
82	23 or 37 or 49 or 59 or 61 or 66 or 69 or 72 or 81
83	10 and 82
84	23 and 82
85	Limits: Humans, English, German, Dutch, systematic review, randomized controlled trial, date 1985-current

Appendix 2:
Included articles with their associated quality and risk of bias scoring

Ref	**Author (year)**	**Aim**	**Number of studies included (for SR)/ Total no. of participants**	**Interventions included in each review (SR)/ Intervention (RCT)**	**Outcome**	**Between group differences/ power**
Systemic sclerosis						
19	Willems et al. (2015) (SSc) Systematic review	Effectiveness of non-pharmacological care	23 studies, of which 9 RCTs/ n= n/a	education, oral therapy, comprehensive rehabilitation program for physical and/or psychological functioning, self-management, comprehensive rehabilitation for the hands and/or face, aerobic training, stretching, paraffin treatment, carbon dioxide baths, manual lymph drainage, ultrasound, extracorporeal shock wave therapy, biofeedback, deep oscillation, transcutaneous electrical nerve stimulation, and home total parenteral nutrition	Manual lymph drainage reduced hand volume, edema, and pain, and improved hand function and perceived QOL, multidisciplinary team-care program improves hand grip strength	n/a
49	Liem et al. (2019) (SSc) Systematic review	Effect and safety of exercise therapy	9 studies (4 RCTs, 4 ODs, 1 Single subject experimental design), n= n/a	Hand exercises, Orofacial exercises, Aerobic and muscle-strengthening exercises	Improvement of hand mobility and hand function, significant improvement in grip strength	n/a
50	Stefanantoni et al. (2016) (SSc) RCT	Effect of occupational therapy intervention, integrated with a self-administered stretching program on the hands	n= 31, 16 intervention, 15 in control group	Instruction of an occupational therapist in performing hand exercises + tailor-made instructions + weekly telephone contact for 1 month	Hand function significantly improved T1 (at one month) and T2 (at three months)	Significant difference concerning COPM performance, with greater improvements in the OT group respect to the control one at T 1 (p<0.02); power n/a

57	Horvath et al. (2017) (SSc) RCT	Efficacy of intensive hand physical therapy	n= 53, 31 intervention, 22 control group	stretching, occupational therapy in combination with mud baths, soft tissue massage, during 3 weeks, daily; Control group: large joints only, not hands	Significant improvements in hand function and finger to palm, improvements in grip strength	Significant difference in HAQ and DASH scores in intervention group at T2 (p<0.05); power 0.74
51	Landim et al. (2019) (SSc) Quasi experimental study, pre- and post-assessment	Development and evaluation of a home-based program for hand function	n= 22	Home based hand exercises over 8 weeks, assessment at baseline, 4 and 8 weeks	Significant improvements in hand pain, overall hand function, hand mobility, delta finger-to-palm, grip strength, tip pinch, and key pinch strength	n/a
52	Rannou et al. (2017) (SSc) RCT	Evaluation of the impact of a supervised standardized and individualized physical therapy program followed by daily home sessions at 1 year on disability	n= 218, 110 intervention, 108 control group	Personalized program, depending on problems and their localization. Intervention focused on increasing joint mobility, muscle strength, daily activities, mouth, skin. Given by an occupational and physiotherapist. Four weeks, 3 times a week + home exercises	At T1 (one month) improved global hand mobility, and reduced hand disability and pain, at T2 (at three months) improved hand mobility, At T3 (at 12 months) no between-group differences	Significant difference in HAQ-DI (-0.14, p<0.01), and hand mobility and function (-3.65, p<0.01) at T1, significant difference in HAQ-DI (-0.12, p=0.054) at T2; power 80%
53	Denton et al. (2016) (SSc) Guideline	BSR and BHPR guideline for the treatment of systemic sclerosis	n/a	n/a	Physical therapy, massage therapy and other programs to improve exercise capacity, Raynaud's phenomenon: Patient education (cold avoidance, staying warm, smoking cessation), Skin involvement: - Moisturization (particularly with lanolin-based products), avoidance of frequent bathing with harsh detergents, - Treatment of telangiectasia with skin camouflage and laser therapy	n/a
58	Seeger et al. (1987) (SSc) RCT	To determine whether or not the proximal interphalangeal joint contractures in patients with progressive systemic sclerosis can be reduced, modified, or reversed through a regimen of dynamic splinting designed to stretch connective tissue	n= 38 hands, 19 intervention, 19 control group	Splinting 8 hours per day for the 2-month study period, unsplinted hand served as control	Neither the splinted nor the control hands of the 8 subjects, changed significantly in active PIP extension at 1 or 2 months. No significant difference between splinted and unsplinted hands in range of motion. No statistically significant evidence to indicate that the splints had served to maintain the PIP extension when compared to the unsplinted hand as neither group worsened significantly. The splint and the protocol employed did not improve nor maintain PIP extension for this group of scleroderma subjects. 11 dropouts: 7 due to intervention, 4 due to reasons not related to the disease.	n/a
56	Mitropoulos et al. (2018) (SSc) RCT	Effects of a supervised and individually-tailored exercise programme based on arm cranking (ACE) and cycle ergometry (CE) on microvascular reactivity, aerobic capacity, exercise tolerance and enjoyment levels	n= 34, 11 arm crank ergometer, 11 cycle ergometer, 12 control group	12-week exercise programme, twice-weekly supervised exercise sessions, 5 min warm-up, followed by 30 s at 100% and 30 s passive recovery for a total of 30 min, and a 5 min cool-down period. the control group did not perform any type of supervised physical activity	Aerobic exercise, involving the upper limbs may improve the microvascular reactivity through an enhancement of the endothelial-dependent function.	No significant differences between groups, significant reduced discomfort due to Raynaud's pain in all groups (p<0.05) at T1; power n/a

Systemic lupus erythematosus							
47	Yuen & Cunningham (2014) (SLE) Systematic review – narrative synthesis	Mixed – any interventions for SLE included	26 studies, of which 9 RCTs / n= 2789)	Exercise (7 studies, of which 4 RCTs) Primarily aerobic, 3x per week, 30-50 minutes, 8 to 12 weeks. Psychosocial interventions (7 studies of, which 5 RCTs) CBT, psychoeducation, counselling, psychotherapy and biofeedback	Exercise All but one study (3 of the 4 RCTs and all 3 non-RCTs) showed significantly reduced fatigue Psychosocial interventions 3 of the 7 studies demonstrated decrease in fatigue	n/a	AMSTAR medium
43	del Pino-Sedeño et al. (2016) (SLE) Systematic review – narrative synthesis	Non-pharmacological interventions for SLE	12 studies, of which 7 RCTs/ n= 549	Exercise Aerobic in all, include strengthening in one. 8weeks – 8 months, typically 3x per week, 30-60 minutes. Behavioral and psychological approaches, CBT, psycho-education, relaxation, self-management and counselling.	All studies report significant fatigue reduction on at least one measure (effect sizes ranged from 0.24 – 0.84). All exercise studies report significantly reduced fatigue (effect size ranged from 0.30 – 0.89). All behavioral and psychological approach studies found statistically significant reductions in fatigue, with moderate to high effect sizes.	n/a	AMSTAR high
48	O'Dwyer et al. (2017) (SLE) Systematic review - meta-analysis	Exercise and physical activity in SLE	11 studies, of which 6 RCTs and 5 quasi-RCTs/ n=469, 7 studies included in meta-analyses	Exercise and physical activity	The two studies assessing fatigue showed significantly improved fatigue in the exercise intervention groups compared to controls.	n/a	AMSTAR high
42	Poole et al. (2019) (SLE) Systematic review	Occupational therapy interventions in SLE	20 studies, 10 RCTs/ n=893	10 physical activity interventions , 10 psychoeducational interventions	Moderate evidence supports physical activity to improve depression, fatigue, exercise tolerance, and function without exacerbation of disease symptoms.	n/a	AMSTAR high
Rheumatoid arthritis (Osteoarthritis)							
40	Cramp et al. (2013) (RA) Systematic review – meta-analysis	Non-pharmacological interventions for fatigue in rheumatoid arthritis	13 RCTs / n= 1556	Physical activity including pool based-therapy, yoga, dynamic strength training, aerobics, Tai Chi. Psychosocial interventions – expressive writing, CBT, mindfulness, lifestyle management, energy conservation, self-management and group education.	Pooled effects Physical activity interventions significantly reduced fatigue (SMD= -0.36, 95%CI -0.62 to -0.10). Psychosocial interventions) significantly reduced fatigue (SMD= -0.24, 95% CI -0.40 to -0.07).	n/a	AMSTAR high
41	Siegel et al. (2017) (RA) Systematic review - meta-analysis	Occupational therapy interventions in RA	51 studies, of which 5 systematic reviews, meta-analyses, or RCTs, 19 articles on physical activity, 32 on psychoeducational interventions/ n= n/a	Physical activity interventions home exercise and coaching, dynamic exercise, aquatic exercise, aerobic exercise, resistive exercise, and Tai Chi and yoga Psychoeducational interventions patient education and self-management, cognitive–behavioral therapy, multidisciplinary approaches, joint protection, assistive devices, emotional disclosure, and comprehensive occupational therapy.	Physical activity interventions exercise appeared to improve function and fatigue; yoga resulted in improvements in vitality maintained at a 2-mo follow-up assessment Psychoeducational interventions Patient education and self-management interventions showed small benefit in managing fatigue	n/a	AMSTAR medium

45	Shen et al. (2020) (RA) Systematic review - meta-analysis	Cognitive behavioral interventions in RA	6 RCTs/ n=430	Effects of CBT on anxiety, depression and fatigue levels	Significant reduction in all three outcome areas: anxiety, depression and fatigue symptoms	n/a	AMSTAR medium
44	Hewlett et al. (2019) (RA) RCT	Reduction of arthritis fatigue impact	n= 333; 175 intervention, 158 control group	CBT training vs. usual care, seven-session group CBA course for RA fatigue self-management delivered by rheumatology nurses and occupational therapists	Reduced fatigue impact beyond usual care both at 6 months and 2 years. RAFT had high patient attendance and satisfaction. Improvements were also seen in emotional fatigue, living with fatigue, coping with fatigue and self-efficacy.	Significant difference in fatigue impact after 26 weeks (-0.59, p=0.03); power n/a	JBI high
46	Azeez et al. (2020) (RA) RCT	Exercise programme on body composition, aerobic capacity, muscle strength and cognition	n= 66, 33 intervention, 33 control group, after three months n= 24/21	3-month personalized exercise programme plus three sessions with PT during study period, standard care: advice on benefits of exercise and recommendations	GFI was significantly reduced (compared with control group) after 3 months.	Significant difference in fatigue scores (p=0.025); power n/a	JBI high
54	Bobos et al. (2019) (RA and OA) Systematic review - meta-analysis	Effectiveness of joint-protection programs on pain, hand function, and grip strength	17 studies (all RCTs), n=2120	Instruction on exercise or JP education, and was administered individually or to a group and was delivered by an occupational therapist, Outcomes: pain levels, Self-report hand function, and grip strength	JP reduced pain for people with RA at midterm and long-term follow-ups. At midterm, JP programs were superior to usual care/ control in terms of grip strength levels. Short term, midterm, long-term effects on function levels.	n/a	AMSTAR high

Raynaud's phenomenon							
55	Belch et al. (2017) (Raynaud's phenomenon) Guideline	ESVM guidelines–the diagnosis and management of Raynaud's phenomenon.	n/a	Diagnosis and management of RP: diagnostics, lifestyle management, pharmacological and non-pharmacological care.	Education and avoidance of triggers are key. It is important to advise patients about protecting themselves from the cold. Lifestyle measures such as wearing gloves when handling frozen food should be adopted. Simple suggestions such as keeping the trunk warm and providing occupational therapy aids such as key holders to use when the fingers are numb can all help. Education is important and an occupational therapist can provide useful advice, as can patient self-help groups. It is essential that patients with RP stop smoking and, where applicable, avoid vibration exposure. Recommendation 10: Lifestyle change is an effective means of controlling RP attacks and should include dressing warmly, ceasing smoking, avoiding triggers such as cold, and an occupational therapy assessment for aids if difficulties are reported (Grade IIa – Level C).	n/a	Recommendations Grade IIa – Level C
SSc, Systemic sclerosis; SLE, Systemic lupus erythematosus; RA, Rheumatic arthritis; OA, Osteoarthritis; RP, Raynaud's phenomenon; QOL, quality of life; JP, joint protection; SR, systematic review; RCT, Randomized controlled trial; COPM, Canadian occupational performance measure; OT, occupational therapy; GFI, Global fatigue index; RAFT, reducing arthritis fatigue ;HAQ, Health Assessment Questionnaire ; HAQ-DI, Health Assessment Questionnaire-Disability Index; DASH, Disabilities of the Arm, Shoulder and Hand questionnaire; CBT, cognitive behavioral treatment							

Appendix 3:
Draft recommendations with their associated quality scoring and level of evidence

Recommendation		Supportive evidence	Quality score of Systematic reviews	Quality score of RCTs	Additional studies/ Recommendations	Oxford level of evidence
FATIGUE **Patient education**						
1	Maintaining good physical condition and regular exercise	(38-40) RA,RA, SLE	medium: n=1[5] high: n=2[1,6]			I
2	Principles of energy conservation and good sleep hygiene	(39,41) SLE, RA	medium: n=1[5] high: n=1[3]			I
3	Relaxation exercises	(41) SLE	high: n=1[3]			I
*	The beneficial effect of Yoga and Tai Chi on general physical condition and relaxation	(38, 39) RA, RA	high: n=2[1,5]			I
4	A healthy diet	n/a				V
5	The possible link between fatigue and drug side effects	n/a				V
FATIGUE **Treatments**						
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 nurse, social worker or occupational therapist.	(39, 41, 42) SLE, RA, RA	medium: n=1[5] high: n=1[3]	high: n=1[8]		I
7	Cognitive behavioural therapy (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.	(41-43) SLE, RA, RA	medium: n=1[7] high: n=1[3]	high: n=1[8]		I
8	Participation in available online and face-to-face courses with fellow sufferers, provided by trained patient representatives: e.g. ReumaUitgedaagd! (self-management training for people with rheumatism) should be offered to patients with SSc with fatigue.	n/a				V
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 an occupational therapist or social worker.	n/a				V
10	SSc patients with fatigue should receive support to improve exercise capacity and incorporate more physical activity into daily life with the guidance of a physical therapist.	(38-41, 44-46) Ra, SLE, SLE, SLE, RA, SLE, SSc	medium: n=2[2,5] high: n=4[1,3,4,6]	high: n=1[1,9]		I
11	Advice about a healthy diet and preventing malnutrition offered by a dietician should be offered to SSc patients with fatigue.	n/a				V
12	A multidisciplinary rehabilitation program should be offered for SSc patients with severe fatigue symptoms that lead to problems in several domains of activities of daily living.	n/a				V
HAND FUNCTION PROBLEMS **Patient education**						
	Independently and regularly doing hand exercises to maintain hand mobility and strength	(47-51)	medium: n=1[10,11]	high: n=2[13,16]	Quasi experimental high: n=1[15]	II
14	A continuous use of hands in activities of daily living to maintain hand functionality	n/a				V
15	Avoiding cold and keeping the hands warm	(52)			Recommendations: Grade III – Level C	IV
16	A good hand care, for example by moisturizing the skin (especially with lanolin-based products) and wearing protective gloves	(52)			Recommendations: Grade III – Level C	IV
HAND FUNCTION PROBLEMS **Treatments**						
17	Passive and active hand function exercises to promote hand mobility, functionality and strength, under the guidance of a hand therapist, should be offered to SSc patients who experience restrictions in the performance of daily activities due to hand function problems	(47-51)	medium: n=1[10,11]	high: n=2[13,16]	Quasi experimental high: n=1[15]	II
18	Learning ergonomic measures under the guidance of an occupational therapist should be offered to SSc patients who experience restrictions in the performance of daily activities due to hand function problems	(49,53) RA/OA, SSc	high: n=1[12]	high: n=1[13]		I
*	Manual lymph drainage by a physiotherapist, occupational therapist or hand therapist in SSc patients with finger or hand edema	(47)	medium: n=1[10]			II
*	Connective tissue massage (possibly in combination with passive and active hand exercises)	(47,56)	medium: n=1[10]	medium: n=1[14]		II
19	The adaption of hobbies and work (including volunteer work) to enable participation in meaningful activities of daily living, under the guidance of a social worker or occupational therapist, should be offered to SSc patients who experience hand function problems	(51)		high: n=1[16]		II
*	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.	(57)		medium: n=1[18]		III
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	(47,51)	medium: n=1[10]	high: n=1[16]		II
RAYNAUD'S PHENOMENON AND DIGITAL ULCERS **Patient education**						
21	Quitting smoking	(54) Raynaud			Recommendations: Grade IIa – Level C	IV
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	(54) Raynaud			Recommendations: Grade IIa – Level C	IV

	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 hand washing iii. avoiding contact with cold objects (wearing gloves to remove items from the fridge/freezer, using a heated keyboard and mouse)	(54) Raynaud			Recommendations: Grade IIa – Level C	IV
24	Wearing fingertip protection (e.g., thermoplastic material or neoprene) to prevent pain when performing actions that cannot be avoided	n/a				V
*****	Avoiding exposure to vibrations, which can adversely affect blood flow	(54) Raynaud			Recommendations: Grade IIa – Level C	IV
25	Preventing infections and wounds through good hygiene, avoiding bruising and hazardous work, and using gloves for protection	n/a				V
26	Promoting good blood circulation through the use of a stress ball and having sufficient exercise throughout the day	n/a				V
27	Avoiding prolonged, static postures	n/a				V
28	Hydration of the skin around the ulcers with products based on lanolin, petroleum jelly or cetomacrogol	(52)			Recommendations: Grade III – Level C	IV
*****	The possibly beneficial effect of soda baths (no more than twice a week) to prevent dehydration and cracking	(52)			Recommendations: Grade III – Level C	IV
	Avoiding frequent exposure to water with aggressive cleaning agents	(52)			Recommendations: Grade III – Level C	IV
30	The importance of avoiding finger punctures	n/a				V
31	Avoiding manipulation of ulcers (e.g., by squeezing out calcium deposits or cutting away hard skin)	n/a				V
*****	The possible importance of a healthy diet with adequate fat intake	n/a				V
FENOMEEN VAN RAYNAUD EN DIGITALE ULCERA **Treatments**						
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 physiotherapist, should be offered to SSc patients with Raynaud's phenomenon/ digital ulcers	(55)		medium: n=1[19]		III
33	In case of vasculopathy of the feet, advice about suitable, non-restrictive footwear (for indoor and outdoor use), by a podiatrist, should be offered to SSc patients	n/a				V
34	Advice on the protection of the fingertips with special gloves or by using adaptive devices, provided by an occupational therapist, should be offered to SSc patients whose activities of daily living are restricted due to Raynaud's phenomenon/ digital ulcers	n/a				V

* Refers to draft recommendations which were excluded from the final recommendations in phase 4 through the determination of the level of agreement bij the task force members and medical experts

Chapter 6

General discussion

Introduction

The overarching aim of my thesis was to investigate possible target points for improving the indication for non-pharmacological SSc care and to facilitate accessible and effective non-pharmacological care for persons with SSc. To achieve these aims, I conducted several studies: a qualitative interview study and two cross-sectional survey studies, followed by the development of multidisciplinary recommendations based on the scientific literature and expert consensus.

This final chapter of my thesis starts with a reflection on the process of my PhD research, followed by a synthesis of the main findings of the first three studies (**chapters 2 to 4**). Based on these findings and the literature, we then introduce and interpret factors that we believe form the important elements of our roadmap to accessible and effective non-pharmacological SSc care. The chapter closes with some methodological considerations.

Reflection

Developing a research topic is often a personal process that unfolds over time. As an occupational therapist in rheumatology rehabilitation, I have always been interested in how multidisciplinary team care contributes to the quality of individual care. Early in my career, I became acquainted with scleroderma patients and the problems they experienced in their daily lives. I quickly understood that I was working with ‘persons with a disease’, including their daily activities, fears, dreams and social environment. I began to understand the value of giving them an individual perspective on their treatment, rather than using the more collective term "patients" (in the original sense of the term: suffering or enduring). This experience has broadened my understanding of the complexity of multidisciplinary SSc care and the importance of the collaboration for both care recipients and care providers.

While working on my master's thesis, I learned that the number of persons with SSc being referred for health professional care had been steadily declining, although the number of referrals had never been particularly high. Monthly, only about 10 persons with SSc were enrolled in health professional treatments, while about 800 were being treated at the university hospital where I worked. This inspired me to investigate the problem to determine what the process and content of the indication for non-pharmacological SSc treatment is.

My initial hypothesis was that the problem of not referring persons with SSc to health professionals was due to blips in the process. I thought that, if we were able to map the perspectives of important stakeholders (those with SSc, health professionals, and rheumatologists), we could use their opinions to systematically develop a strategy or tool for optimizing the indication for non-pharmacological care of persons with SSc. In the first study among specialized Dutch rheumatologists and their motivations for referral or not, role division in decision-making and individual experiences with non-pharmacological care (**chapter 2**), revealed more complex considerations, beliefs and structural barriers than I had expected. Fortunately, I had the opportunity to join the Arthritis Research and Collaboration Hub (ARCH) project. ARCH is a Dutch initiative, established as a nationwide effort to improve health care for persons with rare systemic autoimmune diseases, including SSc. More specifically, the cooperation with ARCH and its large network of experts and researchers gave me the opportunity to extend my previous work by combining different expert perspectives in two comprehensive studies with a nation-wide representative sample of persons with SSc and their treating health professionals (**chapters 3 and 4**).

The results of these first three studies together with the results of a multidisciplinary working conference hosted by ARCH helped us to identify and prioritize widely accepted target points for improving clinical practice and future research goals. In addition, it provided distinct directions to the content of the research questions on the basis of which we developed the multidisciplinary recommendations for health professional treatment (**chapter 5**).

Barriers to accessible and effective non-pharmacological SSc care

In the first three studies (**chapters 2-4**) I explored the barriers to accessible and effective high quality care for persons with SSc[1,2]. According to Donabedian, aspects of quality of care can be divided into three categories: structure, process and outcome[3]. *Structure* refers to the organizational factors under which care is delivered and can directly influence care processes and healthcare outcomes. *Process* is the actual delivery and receipt of care in clinical practice that involves interactions between users and the healthcare structure. *Outcomes* are consequences that can be influenced both by structures and by processes[3]. I used these three categories to structure our results on the perspectives of rheumatologists, persons with SSc, and health professionals, in order to visualize target points for improving indication for health professional SSc care. Table 1 illustrates the multitude of factors that emerged.

Table 1: Barriers to accessible and targeted non-pharmacological SSc care

Structure	Process	Outcomes
• RH experience restrictive institutional policy[1]	• Low quality of communication between HP and RH[1, 2, 3]	• Few referrals of persons with SSc to HP[3]
• Lack of HP evidence[1]	• Insufficient alignment of HP treatment goals with RH referral reasons[3]	• Persons with SSc experience unmet care needs[2]
• 70% of HP work in private practices[3]		
• Broad HP treatment offer-not visible for persons with SSc[3]	• Overlap in treatment domains between HP disciplines[3]	• HP treatment outcomes not visible to RH[1]
• RH lack of trust in HP competences[1]		• Weak role of persons with SSc in treatment process[1]
• RH lack of knowledge about HP treatment options[1, 2]		• Weak role of HP in treatment process[1]

Our findings presented in **chapters two to four** indicate that there are structural and procedural gaps in the organization of SSc care that not only impede provision of accessible and effective non-pharmacological care, but also negatively influence the subjective treatment outcomes of persons with SSc, reflected in the unmet care needs experienced by persons with SSc (**chapter 3**). Our findings corroborate those of previous work from the perspective of persons with SSc, indicating that unmet information and care needs are common[4-6]. These unmet care needs relate to the coordination of care and limited access to trustworthy information, including knowledgeable health professionals. Accordingly, persons with SSc consider non-pharmacological care as one of the most important improvement aspects affecting their quality of care[7-11]. Consistent with the literature, in my opinion, the findings of the three studies (**chapters 2 to 4**) cannot be viewed in isolation, as structure, process and healthcare outcomes intervene with each other.

In terms of accessibility and effectiveness of care, we searched for factors that may contribute to persons with SSc having reduced access to the health structures and care processes they need, and what might contribute to the care they receive being considered less effective. First, non-pharmacological treatment options appear to be underutilized due to an insufficient number of referrals, as reflected in the unmet care needs of persons with SSc. The low quality of communication between rheumatologists and health professionals, experienced by all stakeholders, the lack of strong evidence for the effectiveness of non-pharmacological treatments (**chapter 2**), and the overlap in treatment domains of several health professional disciplines (**chapter 4**), may be the reason for rheumatologists' lack of awareness regarding treatment options of other health professional disciplines[12]. This information gap together with the institutional barriers rheumatologists experience may in turn lead to referrals of persons with SSc to health professionals working in monodisciplinary primary care settings, without connection to interdisciplinary networks (**chapter 4**). This then could lead to rheumatologists referring less frequently to health professionals, as they have more confidence in the expertise of colleagues they know personally and work with on a daily basis (**chapter 2**). Secondly, the treatment offer of health professionals is insufficiently visible, amongst others this is a result of the transition of recent Dutch healthcare system changes which forced hospital-based team care to relocate in a primary care setting[13]. This physical distance between rheumatologists working in hospitals and health professionals working in private practices could be a reason why the treatment offer and outcomes of health professionals are invisible to rheumatologists, and that rheumatologists' referral reasons often do not correspond to the actual treatment goals of the health professionals. This lack of structural cooperation, together with the lack of evidence for the effectiveness of non-pharmacological treatments, is also a possible reason for the weak role of health professionals in the treatment process, as perceived by rheumatologists. The perceived unmet care needs of persons with SSc and their weak role in the treatment process as perceived by rheumatologists indicate a need for another treatment approach (**chapters 2 and 3**). An approach based on self-management support can provide health professionals and rheumatologists with the necessary tools to view a person with SSc more holistically, to be able to give structure to the type of interventions, and thus to pay more attention to person-centered care.

Exploring possibilities to improve quality of non-pharmacological care in SSc

Facilitating self-management

To empower persons with SSc to take an active role in their treatment process, greater efforts need to be made to support them in gaining a good understanding of their disease, and to build their ability to effectively deal with its practical, physical and psychological consequences. Self-management interventions go beyond the pharmacological treatment of disease and its symptoms, and actively investigate and address the impact of disease on a person's priorities and quality of life[14]. Persons with SSc themselves, but also health professionals and rheumatologists, have an important role to play in developing adequate self-management skills.

Non-pharmacological interventions can support persons with SSc in developing both disease and daily-life self-management skills as they are targeted at the physical, psychological and social domain and thereby can improve quality of life[15,16]. As an integral part of supported self-management, patient education content developed following our recommendations can be used throughout the course of the disease to help persons with SSc achieve and maintain independence[17]. In addition, the recently published 2021 EULAR recommendations for the implementation of self-management strategies in patients with inflammatory arthritis could be deployed, as the formulated overarching principles and recommendations align seamlessly with SSc outcome domains that may be influenced by self-management, including pain, fatigue, sleep, emotional and physical well-being, disability, quality of life, and self-efficacy[14].

By regularly examining how persons with SSc feel and how they perceive their day-to-day functioning, the rheumatologist could identify additional preferences with regard to bio-psycho-social treatments and thereby promote referral to health professionals who offer appropriate self-management support. In SSc care, shared decision making (SDM) is viewed as an important approach to both promote person centered care, and to involve persons with SSc in decision-making about all aspects of their care, in order to achieve informed preferences[18]. However, studies on clinical management and the use of SDM in SSc and other rheumatic diseases suggest that discussions initiated by rheumatologists are often limited to asking patients about their pain, disability and disease activity and emphasis is still on medical management[7,19-24]. A conscious use of patient-reported outcome measures (PROMs) to support the shared decision making approach could support rheumatologists in mapping the whole spectrum of bio-psychosocial aspects in persons with SSc[25]. PROMs such as the Cochin 17-item Scleroderma Functional scale (CSF-17), the EULAR Systemic Sclerosis Impact of Disease (ScleroID) questionnaire, or the Self-Efficacy for Managing Chronic Disease (SEMCD) scale are validated and reliable in persons with SSc, and they incorporate items on management of daily activities, psychological demands, responsibilities in personal and professional life, concepts of self-management, and frequently reported unmet care needs[26-28]. The use of these or similar PROMs prior to a consultation could provide both the person with SSc and the rheumatologist with an understanding bio-psychosocial aspects that preoccupy the person, thereby strengthening the role of persons with SSc in the treatment process, and promote targeted referral to an appropriate health professional[23].

Self-management:
- *Health professionals can support the development of self-management skills through education and interventions targeting the bio-psychosocial priorities of persons with SSc.*
- *Rheumatologists can promote targeted referral to appropriate health professionals by the use of patient reported outcome measures to identify preferences with regard to bio-psychosocial treatments.*

Strengthening the role of health professionals

In the Netherlands, the current trend is to implement an SSc shared care approach between highly specialized (tertiary) centers and regional hospitals[11]. However, although it has been determined that a range of health professionals should be part of this infrastructure, widely accepted criteria for SSc-specific expertise of health professionals available at these centers have not yet been defined[29]. The following elements could form a basis for establishing a set of criteria for SSc-specific expertise and competences of health professionals working in these centers:
- detected unmet care needs of persons with SSc
- proven effective health professional treatments
- established recommendations on health professional treatment options in SSc
- the 2019 EULAR recommendations for the generic core competences of health professionals in rheumatology[16]

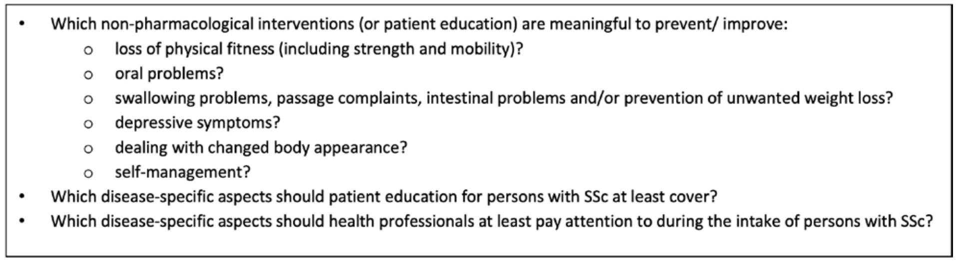
The 2014 Update of the EULAR standardized operating procedures for EULAR-endorsed recommendations could be followed to establish the criteria systematically and to ensure key stakeholder participation (persons with SSc, a diverse group of health professionals, and rheumatologists)[30]. Once established, these criteria can be used to determine which health professionals in centers of expertise are needed to guarantee this expertise. Subsequently, all persons with SSc treated in the expertise centers should have access to these health professionals to ensure that no healthcare needs are overlooked. In addition, an important task of these health professional experts will be to facilitate the training of colleagues at other centers. Establishing criteria for expertise and thus a minimal group of health professional experts in SSc expertise centers can strengthen the role of SSc health professionals and lead to a better alignment between treatment domains of the different health professional disciplines. This in turn can lead to the provision of more accessible non-pharmacological treatment for persons with SSc.

To strengthen the role of health professionals:
- *criteria for SSc-specific expertise and competences of health professionals should be developed*
- *expert health professionals need to be part of the multidisciplinary team of SSc expertise centers*

Extension of recommendations

Our recommendations focus on non-pharmacological treatment and education for the three most frequently described unmet care needs of persons with SSc: fatigue, hand function loss, and Raynaud's phenomenon/ digital ulcers. It will be important to extend these recommendations to address all bio-psychosocial aspects faced by persons with SSc in the context of their daily lives. Established care questions and unmet care needs of persons with SSc could be used for this purpose. Moreover, the as yet undeveloped recommendations from the research questions established by our task force in the initial phase (**chapter 5**) could form valuable input (Table 2).

Table 2: Additional research questions not yet developed into SSc health professional recommendations.



- Which non-pharmacological interventions (or patient education) are meaningful to prevent/ improve:
  - loss of physical fitness (including strength and mobility)?
  - oral problems?
  - swallowing problems, passage complaints, intestinal problems and/or prevention of unwanted weight loss?
  - depressive symptoms?
  - dealing with changed body appearance?
  - self-management?
- Which disease-specific aspects should patient education for persons with SSc at least cover?
- Which disease-specific aspects should health professionals at least pay attention to during the intake of persons with SSc?

A further extension of our recommendations based on the available evidence-based treatment offer of health professionals could also be considered, as this treatment offer may cover additional blind spots in SSc care that have arisen due to the way unmet health care needs have been mapped. In this respect, that persons with SSc must be involved in all phases of recommendation development.

Further work is needed to extend the presented recommendations at an international level in order to create transparency on non-pharmacological treatment options and to promote targeted non-pharmacological care. In addition, the translation of existing evidence-based treatment options for other chronic (rheumatic) conditions for use in persons with SSc should be further investigated. For example, behavioral techniques, such as energy conservation and activity stimulation, have been beneficial in several chronic conditions[31].

Future extensions of recommendations on non-pharmacological treatment in SSc
- *should address all bio-psycho-social aspects faced by persons with SSc*
- *could be structured by unmet care needs of persons with SSc, and established research questions*
- *should explore the translation of existing evidence-based health professional treatment options from other chronic (rheumatic) conditions*

eHealth possibilities

Web-based or so called eHealth technologies are becoming increasingly important for disease- and self-management as they provide the opportunity to connect care recipients and care providers, regardless of the distance between them, and provide viable options for accessible, cost-effective and timely dissemination of information[32,33]. An important positive aspect of eHealth-supported care is the support of self-management behaviors for persons with SSc[25,34-37]. Instructions for self-management as well as advice on a healthy lifestyle have feasible eHealth potential[15]. To date, only one internet-based self-management program has been found satisfactory for persons with SSc. It showed that their knowledge, skills and confidence in managing health improved, and that their fatigue and depressive symptoms reduced[34,35,38].

To take the lead in their self-management (daily life, disease, psychological and social aspects) persons with SSc can also make use of appropriate SSc mobile health (mHealth) technologies that support self-management behaviors[39]. For the development, evaluation and implementation of mHealth applications to support self-management in persons with rheumatic and musculoskeletal diseases, the EULAR has published a list of points to be considered when developing such applications[40]. In addition, existing applications such as the 'remote self-assessment tool for digital ulcers in patients with SSc' or the Activiteitenweger App (activity calculator app) could be considered[41,42]. However, the development, implementation and use of e/mHealth technology is still impeded by a lack of legal, financial, and organizational support, and their applicability is limited if developed without end-user participation[43,44]. In addition, levels of readiness to use of eHealth information varies widely among persons with SSc and health professionals, with a reported lack of experience with eHealth support (e.g., apps, online forums, self-help groups) and difficulties in assessing the quality of eHealth information[37]. To successfully design eHealth technology that fits the needs and skills of persons with SSc and their treating health professionals, their early and active involvement in the development process, using co-creation solutions, is advised[37,45]. As greater eHealth literacy is associated with better access to healthcare, more proactive self-management and improved health-related outcomes, guidelines for the use of eHealth services are needed[46,47]. Hybrid forms of non-pharmacological eHealth interventions and direct, face-to-face care, with pre-consultation intake applications (e.g., PROMs) and self-management applications may be applicable and cost-effective future SSc care strategies[48-50].

e-Education

Due to long distances and complex healthcare structures, persons with SSc often lack access to coordinated, specialized non-pharmacological care with health professionals expert in SSc[43,51]. To bundle information and build up shared expertise, accessible (online) information material on treatment options of health professionals, and lay translations of professional literature and existing guidelines could be provided for persons with SSc, health professionals, and rheumatologists. In addition, this can improve the expertise and alignment between treatment domains of health professionals. A comparable offer has been developed in the Netherlands for persons with Parkinson's disease. ParkinsonNet is a national network with standardized and integrated delivery of evidence-based care for persons with Parkinson's disease provided by health professionals with specialist expertise[52]. The online platform (www.parkinsonnet.nl) gives persons with Parkinson's disease access to information about treatment options from various health professional disciplines, existing treatment guidelines, and experienced health professionals in their region[53].

Virtual online platform

To reduce structural and procedural barriers and implement the proposed improvements arising from this thesis, I advise the development of a virtual online platform. The ARCH digital environment (www.arch.nl/archdigitaal) could qualify for this purpose, as it aims to reduce the scarcity of organized expertise in the field of SSc, and it is a pre-existing platform. To date, the platform supports the shared care approach for persons with SSc by facilitating collaboration between medical specialists in regional meetings. It consists of an app for healthcare providers and a personal web environment for care recipients. An online 'care finder' (zorgzoeker) for finding health professionals close to home could also be integrated on the platform.

e-Collaboration

Recently, education, counseling, coaching and (the promotion of) exercise and physical activity have been shown to be appropriate non-pharmacological interventions for SSc eHealth-enhanced care[15]. Therefore, online consultations with experienced health professionals may be an appropriate option for persons with SSc with special care needs, or those who are prevented by their illness from traveling long distances to see a specialized health professional. In addition, integrating all health professionals in the online multidisciplinary team meetings would improve the frequency and quality of communication between stakeholders and improve SSc care quality[54].

eHealth

- *eHealth can support self-management behaviors in persons with SSc*
- *providing online educational resources can facilitate uniform access to trustworthy information, such as patient education, health professional treatment options, and professional literature*
- *a virtual online platform can support networking between persons with SSc and healthcare providers and improve the latter's frequency and quality of communication*
- *online consultations with expert health professionals and eHealth enhanced SSc care may be an appropriate option for persons with SSc with special care needs or those who cannot easily travel.*

Methodological considerations

I would like to highlight some methodological strengths and limitations that have emerged from these studies. A first strength is the mix of qualitative and quantitative data collection methods in the first three studies. While the qualitative interviews with the rheumatologists (**chapter 2**) gave us profound insights into their thoughts and beliefs, the surveys conducted with persons with SSc and health professionals (**chapters 3 and 4**) focused on a broad overview of the current use of systemic sclerosis care. This promoted data synthesis and thereby identified target points for improving indications for non-pharmacological care. Secondly, our recommendations for non-pharmacological treatment of fatigue, hand function loss, Raynaud's phenomenon, and digital ulcers in SSc (**chapter 5**) are based on recent scientific evidence as well as expert opinion, and were formulated according to standardized procedures for developing practice recommendations[30,55]. A third strength is that our recommendations were developed in close collaboration with a multidisciplinary SSc task force, from the initial draft of research questions to agreement on the final recommendations. This unique project is an example of good communication and collaboration between persons with SSc and healthcare providers from different disciplines who jointly develop person-centered information in an evidence-based way, in order to promote accessible and effective non-pharmacological care. Our recommendations form potential next steps to improving the quality of non-pharmacological SSc care and are a valuable addition to the existing EULAR and BSR/BHPR medical/pharmacological recommendations for the treatment of systemic sclerosis[56,57].

A limitation in the development of the recommendations (**chapter 5**) is the lack of an implementation plan. The value of professional recommendations to improve the quality of care is widely recognized. However, adherence with guidelines and recommendations was found to be sub-optimal in the absence of active implementation strategies[58,59]. Implementation of the recommendations is therefore needed to integrate the recommended actions into daily clinical practice and to improve non-pharmacological care for the individual[30,60]. One strategy for implementing recommendations is through improved professional education[61]. A second strategy is to inform persons with SSc and their patient organizations about the existence of the recommendations and their relevance to managing their own situation. Careful assessment and exploration of potential barriers for the target audience is a critical component of any knowledge translation project involving rare diseases[62]. To enhance adoption of our recommendations, strategies using implementation science and methods such as the use of provider education, audit procedures, and critical evaluations of implementation are indicated[63,64]. Another limitation of the recommendations is their focus on the Dutch environment. An example of this is the exclusion of some of the draft recommendations, some with a high level of evidence, due to the low level of agreement and low acceptance by the task force. In our opinion, the task force gave a good reflection of the most important stakeholders in the current situation in the Netherlands. However, they cannot be considered internationally representative, so the excluded recommendations may well be accepted in other countries.

In summary, the studies in this thesis describe possible targets for improving the indication for non-pharmacological SSc care. These can contribute to enabling more accessible and effective non-pharmacological care and may ultimately result in enhanced quality of life of persons with SSc. As a first response to the identified barriers, we developed multidisciplinary recommendations for education and treatment of frequently reported unmet care needs in persons with SSc. These recommendations can contribute to a strengthened role of health professionals in the multidisciplinary treatment, as well as more targeted referrals of persons with SSc to non-pharmacological care. We expect them to contribute to a better quality of communication and improved organization of SSc-related non-pharmacological care.

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Summary

Systemic sclerosis (SSc) is a complex and rare autoimmune disease. Due to the serious physical and psychological consequences of the disease, persons with SSc experience limitations in daily life and a reduced quality of life. Up to now there is no effective treatment or cure for SSc, meaning that treatment is primarily aimed at controlling symptoms and maintaining quality of life. Providing optimal care for persons with SSc is challenging as it requires experienced healthcare providers, a multidisciplinary approach, and accessible and effective treatment options. Rheumatology health professionals like occupational therapists, physiotherapists and psychologists play an important role in the multidisciplinary care of persons with SSc. Non-pharmacological treatments offered by health professionals provide support with self-management, and are relevant to improve the quality of life of persons with SSc. But although the importance of accessible and effective care delivered by health professionals is broadly recognized, not all persons with SSc receive the same standard of non-pharmacological care. The aim of this thesis is to contribute to the improvement of care for persons with SSc. It focuses on accessible and effective care delivered by health professionals, by investigating possible target points for improving the indication for non-pharmacological care (**chapter 2, 3, and 4**), and facilitating accessible and effective non-pharmacological care for persons with SSc (**chapter 5**).

In **chapter 2** we explored current referral routines of Dutch rheumatologists, and the factors that influence their decisions about referral of persons with SSc to health professionals. Qualitative semi-structured interviews with Dutch rheumatologists with special expertise in the management of persons with SSc were conducted. A qualitative inductive content analysis, following a six-step process of coding was performed to establish meaningful themes. Thirteen rheumatologists with a broad range of work experience within their specialty, from nine different centres in the Netherlands were interviewed. Two major themes were identified as influencing rheumatologists' decision making with respect to referral of persons with SSc to health professionals: 'beliefs' (e.g., own professional role, role of persons with SSc, and role of health professionals) and 'local policy and routines' (e.g., costs, clinical pathways, internal policies, and preferences regarding certain health professionals). In addition, a third additional theme reflecting the needs of the rheumatologists regarding professional multidisciplinary collaboration emerged (e.g., active, visible health professionals, few large expert centres exchanging expertise with regional centres). The results of our research provide a national perspective on factors influencing rheumatologists' decision for referral of persons with SSc to health professionals. A lack of knowledge about health professional treatment options and a low confidence in the competence of other disciplines were identified as barriers for referral to health professionals, which may possibly lead to undertreatment.

Chapter 3 describes a study in which we gained insight into the use of current systemic sclerosis care delivered by health professionals from the perspective of persons with SSc. We focused on referral reasons, treatment goals, the alignment with unmet care needs, and outcome satisfaction with health professional treatments. A total of 650 Dutch persons with SSc from 13 participating rheumatology departments completed an online survey. Descriptive statistics revealed that half of all participants had contact with a health professional in the past year and three quarters since the onset of their disease. Most common referral reasons from the perspective of persons with SSc were pain, limited joint mobility and cardiovascular functions. Persons with SSc reported fatigue (46%, n = 295), Raynaud's phenomenon (31%, n = 204), physical limitations (30%, n = 192), hand function loss (27%, n = 177), and joint problems

(25%, n = 163) as important unmet care needs. Satisfaction with the knowledge and expertise of their health professionals was high (74% of all participants), as well as the improvement of daily activities and coping strategies (73% of all participants). However, more than 50% perceived that the collaboration between their rheumatologist and health professional was insufficient.

Chapter 4 describes a cross-sectional study examining the knowledge and experiences of Dutch health professionals regarding the content of care delivered and the extent to which it is tailored to the unmet care needs of persons with SSc. A web survey among health professionals was used for this purpose, consisting of both closed and open questions. We assessed self-reported referral reasons, treatment goals, and interventions of the last treated person with SSc, as well as the perceived quality of communication between health professionals and rheumatologists. Based on the responses from the seventy-nine health professionals, we identified one hundred and thirty-three different unique reasons for referral, of which 70% were related to ICF-domain 'body structures and functions'. The broad spectrum of reported interventions on the other hand focused on body functions and structures (28%), training daily activities (26%), education and counselling (26%) and psychosocial interventions (20%). A comparison of the reported treatment options of health professionals with unmet health care needs expressed by persons with SSc (**chapter 2**) showed that, among numerous others, the three most frequently mentioned unmet health care needs are covered. Additionally, we found a considerable overlap in the content of the various health professional disciplines. Furthermore, treatment goals were found to be insufficiently aligned with referral reasons of rheumatologists. The quality of communication between health professionals and rheumatologists was perceived as low.

In **chapter 5** multidisciplinary recommendations for non-pharmacological treatment of fatigue, hand function loss, Raynaud's phenomenon, and digital ulcers in persons with SSc were developed. The recommendations were based on research evidence and consensus among experts, following the standardized operating procedures of the EULAR (European Alliance of Associations for Rheumatology). The task force was composed of seven persons with SSc and 15 experienced professionals (rheumatologists, internist/clinical immunologist, physiotherapists, occupational therapist, psychologists, dietician, dental hygienist, specialized nurse, social worker, and researchers) in the Netherlands. In 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, and 20 key systematic reviews, RCTs, and published recommendations were selected. Moreover, 13 Dutch medical specialists were consulted on non-pharmacological advice regarding Raynaud's phenomenon and digital ulcers. For each recommendation the level of evidence and the level of agreement was determined. In total, 34 recommendations, concerning treatments and patient education for fatigue, hand function loss, and Raynaud's phenomenon/ digital ulcers were developed and approved by the task force.

Conclusions

The studies in this thesis show target points for the improvement of the indication for non-pharmacological SSc care. These target points can contribute to enabling more accessible and effective non-pharmacological care and may ultimately result in enhanced quality of life of persons with SSc. The multidisciplinary recommendations for education and treatment of frequently reported unmet care needs in persons with SSc can contribute to a strengthened role of health professionals in the multidisciplinary treatment and more targeted referrals of persons with SSc to non-pharmacological care. They may also contribute to a better quality of communication and improved organization of SSc-related non-pharmacological care.

Nederlandse samenvatting

Systemische sclerose (SSc) is een complexe en zeldzame auto-immuunziekte. Door de ernstige lichamelijke en psychische gevolgen van de ziekte ervaren personen met SSc zowel beperkingen in het dagelijks leven als ook en een verminderde kwaliteit van leven. Tot nu toe is er geen effectieve behandeling om personen met SSc te genezen, wat betekent dat de behandeling primair gericht is op het beheersen van symptomen en het behouden van kwaliteit van leven. Het bieden van optimale zorg voor personen met SSc is een uitdaging omdat het ervaren zorgverleners, een multidisciplinaire aanpak en toegankelijke en effectieve behandelingsopties vereist. Reumatologen zoals ergotherapeuten, fysiotherapeuten en psychologen spelen een belangrijke rol in de multidisciplinaire zorg voor personen met SSc. Niet-medicamenteuze behandelingen aangeboden door paramedici bieden ondersteuning bij zelfmanagement en zijn relevant om de kwaliteit van leven van personen met SSc te verbeteren. Maar hoewel het belang van toegankelijke en effectieve zorg door paramedici algemeen wordt erkend, ontvangen niet iedereen met SSc dezelfde standaard van niet-farmacologische zorg.

Het doel van dit proefschrift is een bijdrage te leveren aan de verbetering van de zorg voor personen met SSc. Het richt zich op toegankelijke en effectieve zorg geleverd door paramedici. Dit door mogelijke knelpunten in de indicatie voor niet-farmacologische zorg te onderzoeken (**hoofdstuk 2, 3 en 4**) en daarnaast toegankelijke en effectieve niet-farmacologische zorg voor personen met SSc te faciliteren (**hoofdstuk 5**).

In **hoofdstuk 2** hebben we de huidige verwijzingsroutines van Nederlandse reumatologen onderzocht en factoren die van invloed zijn op hun beslissingen over verwijzing van personen met SSc naar paramedici. Hiervoor zijn kwalitatieve semigestructureerde interviews afgenomen bij Nederlandse reumatologen met speciale expertise in de behandeling van personen met SSc. Er is en kwalitatieve inductieve inhoudsanalyse in zes stappen uitgevoerd om tot betekenisvolle thema’s te komen. Dertien reumatologen met een brede werkervaring binnen hun specialisme, afkomstig uit negen verschillende medische centra in Nederland, zijn geïnterviewd. Twee belangrijke thema's werden geïdentificeerd die van invloed zijn op de besluitvorming van reumatologen met betrekking tot verwijzing van personen met SSc naar paramedici: 'overtuigingen' (bijv. eigen professionele rol, rol van personen met SSc en rol van gezondheidswerkers) en 'lokaal beleid en routines' (bijv. kosten, klinische trajecten, intern beleid en voorkeuren met betrekking tot bepaalde paramedici). Daarnaast kwam een aanvullend derde thema naar voren dat de behoeften van de reumatologen met betrekking tot professionele multidisciplinaire samenwerking weergeeft (bijv. actieve, zichtbare paramedici, weinig grote expertisecentra die expertise uitwisselen met regionale centra). De resultaten van ons onderzoek bieden een nationaal perspectief op factoren die van invloed zijn op de beslissing van reumatologen om personen met SSc wel of niet door te verwijzen naar paramedici. Een gebrek aan kennis over de behandelmogelijkheden van paramedici en een laag vertrouwen in de competentie van andere disciplines werden geïdentificeerd als barrières voor verwijzing naar paramedici, wat mogelijk kan leiden tot te weinig paramedische behandelingen voor personen met SSc.

Hoofdstuk 3 beschrijft een onderzoek waarin we inzicht verkregen hebben in het gebruik van de huidige niet-farmacologische SSc zorg door vanuit het perspectief van personen met SSc. We hebben ons gericht op verwijzingsredenen, behandeldoelen, afstemming op onvervulde zorgbehoeften en tevredenheid over de uitkomst van paramedische behandelingen. In totaal hebben 650 Nederlanders met SSc van 13 deelnemende reumatologieafdelingen een online enquête ingevuld. Uit de beschrijvende statistiek bleek dat de helft van alle deelnemers in het afgelopen jaar, en driekwart sinds het begin van hun ziekte, contact had met één of meerdere paramedici. De meest voorkomende verwijzingsredenen vanuit het perspectief van personen met SSc waren pijn, beperkte gewrichtsmobiliteit en cardiovasculaire functies. Personen met SSc rapporteerden vermoeidheid (46%, n = 295), het fenomeen van Raynaud (31%, n = 204), fysieke beperkingen (30%, n = 192), handfunctieverlies (27%, n = 177) en gewrichtsproblemen (25%, n = 163) als belangrijke onvervulde zorgbehoeften. De tevredenheid over de kennis en expertise van hun paramedici was hoog (74% van alle deelnemers), evenals de verbetering van dagelijkse activiteiten en coping strategieën (73% van alle deelnemers). Meer dan 50% vond echter dat de samenwerking tussen hun reumatoloog en paramedicus onvoldoende was.

Hoofdstuk 4 beschrijft een cross-sectioneel onderzoek naar de kennis en ervaringen van Nederlandse paramedici met betrekking tot de inhoud van de geleverde zorg en de mate waarin deze is afgestemd op de onvervulde zorgbehoeften van personen met SSc. Hiervoor is gebruik gemaakt van een online enquête onder zorgprofessionals, bestaande uit zowel gesloten als open vragen. Zelf gerapporteerde verwijzingsredenen, behandeldoelen en interventies van de laatst behandelde persoon met SSc, evenals de waargenomen kwaliteit van communicatie tussen paramedici en reumatologen zijn hiervoor geëvalueerd. Op basis van de reacties van de negenenzeventig paramedici identificeerden we honderddrieëndertig verschillende unieke redenen voor verwijzing, waarvan 70% gerelateerd was aan het ICF-domein 'lichaamsstructuren en -functies'. Het brede spectrum van gerapporteerde interventies was daarentegen gericht op lichaamsfuncties en -structuren (28%), training van dagelijkse activiteiten (26%), educatie en counseling (26%) en psychosociale interventies (20%). Een vergelijking van de gerapporteerde paramedische behandelopties met door personen met SSc gerapporteerde onvervulde zorgbehoeften (**hoofdstuk 2**) toonde aan dat, naast andere, de drie meest genoemde onvervulde zorgbehoeften worden gedekt. Daarnaast vonden we een grote inhoudelijke overlap tussen de verschillende paramedische discipline. Verder bleken behandeldoelen onvoldoende aan te sluiten bij verwijsredenen van reumatologen. De kwaliteit van de communicatie tussen paramedici en reumatologen werd als laag ervaren.

Hoofdstuk 5 geeft de ontwikkeling van multidisciplinaire aanbevelingen voor de niet-farmacologische behandeling van vermoeidheid, verlies van handfunctie, het fenomeen van Raynaud en digitale ulcera bij personen met SSc weer. De aanbevelingen zijn gebaseerd op onderzoeksgegevens en consensus onder experts, volgens de gestandaardiseerde werkprocedures van de EULAR (European Alliance of Associations for Rheumatology). De taskforce bestond uit zeven personen met SSc en 15 ervaren professionals (reumatologen, internist/klinisch immunoloog, fysiotherapeuten, ergotherapeut, psychologen, diëtist, mondhygiënist, gespecialiseerd verpleegkundige, maatschappelijk werker en onderzoekers). In een bijeenkomst van de taskforce zijn conceptaanbevelingen gegenereerd door middel van een systematisch gestructureerde discussie, waarbij de nominale groepstechniek gevolgd is. Om de aanbevelingen te onderbouwen, is een uitgebreide literatuurstudie uitgevoerd en zijn 20 belangrijke systematische reviews, RCTs en gepubliceerde aanbevelingen geselecteerd. Bovendien werden 13 Nederlandse medisch specialisten geraadpleegd voor niet-farmacologische adviezen over het fenomeen van Raynaud en digitale ulcera. Voor elke aanbeveling is het evidentie niveau en het niveau van overeenstemming bepaald. In totaal zijn 34 aanbevelingen met betrekking tot behandelingen en voorlichting van personen met SSc voor vermoeidheid, verlies van handfunctie en het fenomeen van Raynaud/digitale ulcera ontwikkeld en goedgekeurd door de taskforce.

Conclusies

De in dit proefschrift beschreven studies tonen aangrijpingspunten voor de verbetering van de indicatie voor niet-farmacologische SSc-zorg. Deze aangrijpingspunten kunnen bijdragen aan het mogelijk maken van meer toegankelijke en effectievere niet-farmacologische zorg en kunnen uiteindelijk leiden tot een betere kwaliteit van leven van personen met SSc. De multidisciplinaire aanbevelingen voor educatie en behandeling van vaak gerapporteerde onvervulde zorgbehoeften van personen met SSc kunnen bijdragen aan een krachtigere rol van paramedici in de multidisciplinaire behandeling en meer gerichte verwijzingen van personen met SSc naar niet-farmacologische zorg. Ook kunnen ze bijdragen aan een betere kwaliteit van communicatie en een betere organisatie van SSc-gerelateerde niet-farmacologische zorg.

Deutsche Zusammenfassung

Systemische Sklerose (Sklerodermie, SSc) ist eine komplexe und seltene Autoimmunerkrankung. Aufgrund der schwerwiegenden körperlichen und psychischen Folgen der Erkrankung erfahren Betroffene Einschränkungen im täglichen Leben und eine reduzierte Lebensqualität. Bisher existiert keine wirksame Behandlung oder Heilung für SSc, was bedeutet, dass die Behandlung in erster Linie darauf abzielt, die Symptome zu kontrollieren und die Lebensqualität zu erhalten. Die Bereitstellung einer optimalen Gesundheitsversorgung für Menschen mit SSc ist eine Herausforderung, da sie erfahrene Gesundheitsdienstleister, einen interdisziplinären Ansatz sowie gut zugängliche und wirksame Behandlungsoptionen erfordert. Gesundheitsfachkräfte wie Ergotherapeuten, Physiotherapeuten und Psychologen spielen eine elementare Rolle bei der interdisziplinären rheumatologischen Behandlung von Meschen mit SSc. Behandlungen, die von Therapeuten dieser Berufsgruppen angeboten werden, unterstützen das Selbstmanagement und sind relevant, um die Lebensqualität von Menschen mit SSc zu verbessern. Doch obwohl die Bedeutung einer zugänglichen und wirksamen Gesundheitsversorgung durch Gesundheitsfachkräfte in der Rheumatologie allgemein anerkannt ist, erhält nicht jeder mit der Diagnose SSc den gleichen Standard an therapeutischer Gesundheitsversorgung.

Ziel dieser Doktorarbeit ist es, einen Beitrag zur Verbesserung der Gesundheitsversorgung von Menschen mit SSc zu leisten. Sie richtet sich auf eine zugängliche und wirksame Behandlung durch Gesundheitsfachkräfte, indem mögliche Ansatzpunkte zur Verbesserung der therapeutischen Gesundheitsversorgung untersucht werden (**Kapitel 2, 3 und 4**) und eine zugängliche und wirksame therapeutische Gesundheitsversorgung für Personen mit SSc mit konkreten Empfehlungen unterstützt werden (**Kapitel 5**).

In **Kapitel 2** untersuchten wir die aktuellen Überweisungsroutinen niederländischer Rheumatologen und Faktoren, die ihre Entscheidungen über die Überweisung von Personen mit SSc an Gesundheitsfachkräfte beeinflussen. Qualitative semi-strukturierte Interviews mit niederländischen Rheumatologen mit besonderer Expertise in der Behandlung von Personen mit SSc wurden durchgeführt. Eine qualitative induktive Inhaltsanalyse nach einem sechsstufigen Codierungsprozess wurde durchgeführt, um behandlungsrelevante Themen zu ermitteln. Dreizehn Rheumatologen mit einem breiten Spektrum an Berufserfahrung in ihrem Fachgebiet, aus neun verschiedenen spezialisierten Krankenhäusern in den Niederlanden wurden befragt. Es wurden zwei Hauptthemen identifiziert, die die Entscheidungsfindung von Rheumatologen in Bezug auf die Überweisung von Betroffenen an Gesundheitsfachkräfte beeinflussen: „Überzeugungen“ (z. B. eigene berufliche Rolle, Rolle von Personen mit SSc und Rolle von Gesundheitsfachkräften) und „lokale Politik und Routinen“ (z. B. Kosten, Behandlungsrichtlinien und Präferenzen in Bezug auf bestimmte Gesundheitsfachkräfte). Darüber hinaus entstand ein drittes zusätzliches Thema, das die Bedürfnisse der Rheumatologen in Bezug auf eine professionelle multidisziplinäre Zusammenarbeit widerspiegelt (z. B. aktive, sichtbare Gesundheitsfachkräfte, wenige große Expertenzentren, die Fachwissen mit regionalen Zentren austauschen). Die Ergebnisse unserer Forschung bieten eine nationale Perspektive auf Faktoren, die die Entscheidung von Rheumatologen bei Überweisung von Personen mit SSc an Gesundheitsfachkräfte beeinflussen. Als Hindernisse für die Überweisung an Gesundheitsfachkräfte, die möglicherweise zu einer unzureichenden Behandlung führen können, wurden fehlendes Wissen über die Behandlungsoptionen von Gesundheitsfachkräften und ein geringes Vertrauen in die Kompetenz anderer Fachdisziplinen identifiziert.

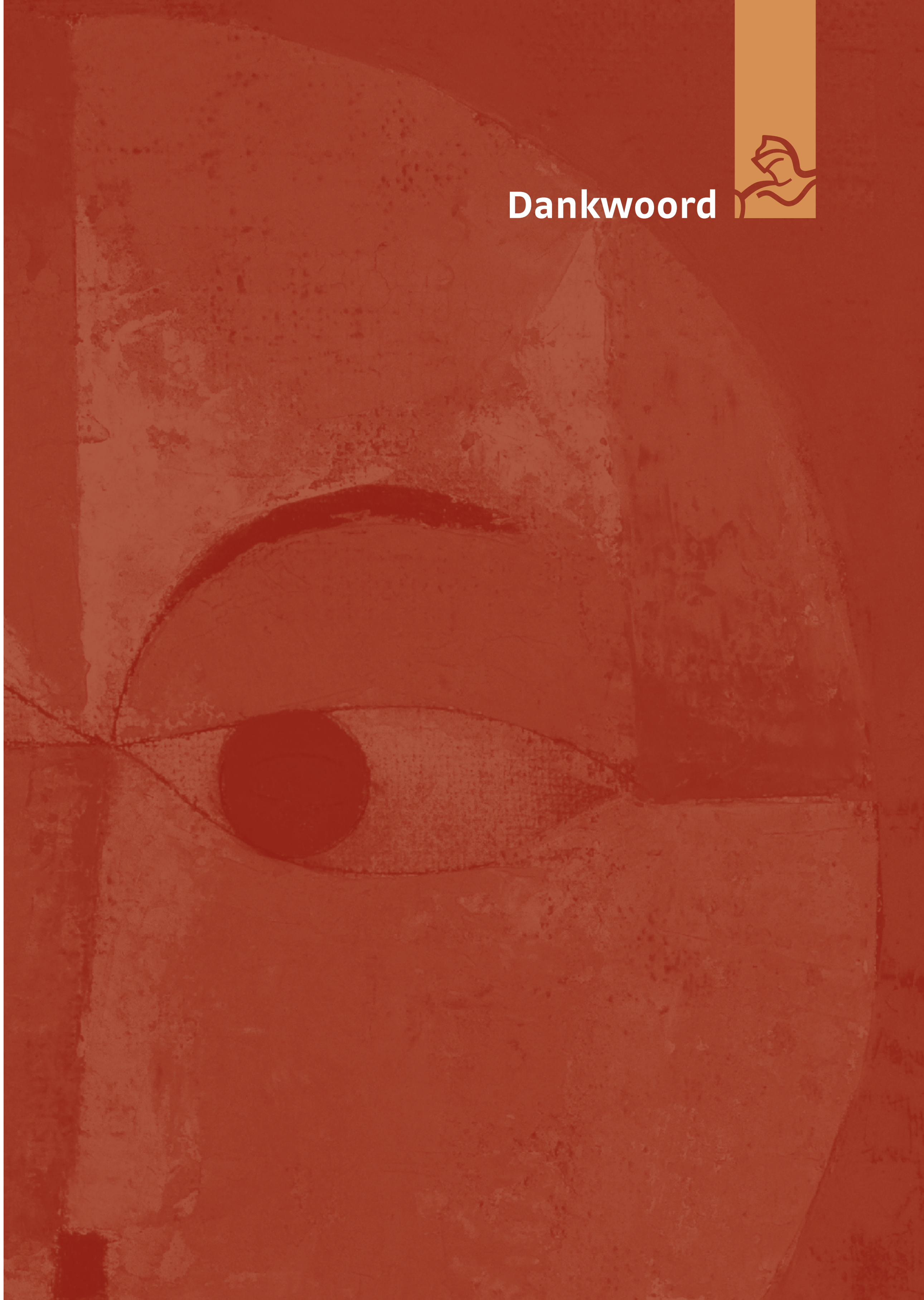
Kapitel 3 beschreibt eine Studie, in der wir aus der Perspektive von Personen mit SSc einen Einblick in die Nutzung der aktuellen therapeutischen Versorgung durch Gesundheitsfachkräfte gewonnen haben. Hierbei lag unser Fokus bei Überweisungsgründen, Behandlungszielen, die Ausrichtung auf unerfüllte Bedürfnisse in der therapeutischen Versorgung und die Ergebniszufriedenheit mit Behandlungen durch therapeutische Fachdisziplinen. Insgesamt haben 650 niederländische Personen mit SSc aus 13 teilnehmenden Rheumatologie-Abteilungen an unserer Online-Umfrage teilgenommen. Die deskriptive Statistik ergab, dass die Hälfte aller Teilnehmer im vergangenen Jahr und drei Viertel seit Beginn ihrer Erkrankung Kontakt zu einer Gesundheitsfachkraft hatten. Die häufigsten Überweisungsgründe aus Sicht der Be waren Schmerzen, eingeschränkte Beweglichkeit der Gelenke und kardiovaskuläre Funktionen. Personen mit SSc berichteten über Müdigkeit (46%, n = 295), Raynaud-Phänomen (31%, n = 204), funktionelle Einschränkungen (30%, n = 192), Handfunktionsverlust (27%, n = 177) und Gelenk-Probleme (25%, n = 163) als wichtige unerfüllte Bedürfnisse in der therapeutischen Versorgung. Die Zufriedenheit mit dem Wissen und der Expertise der Therapeuten war hoch (74% aller Teilnehmer), ebenso wie Verbesserungen der Alltagskompetenz und dem Erlernen von Copingstrategien (73% aller Teilnehmer). Allerdings empfanden mehr als 50% aller Teilnehmer die Zusammenarbeit zwischen ihrem Rheumatologen und dem Therapeuten als unzureichend.

Kapitel 4 beschreibt eine Querschnittsstudie, die das Wissen und die Erfahrungen niederländischer Gesundheitsfachkräfte in Bezug auf den Inhalt der erbrachten therapeutischen Versorgung und das Ausmaß, in dem Therapien auf unerfüllte Bedürfnisse in der therapeutischen Versorgung von Personen mit SSc zugeschnitten sind, untersucht. Zu diesem Zweck wurde eine Online-Umfrage unter Gesundheitsfachkräften durchgeführt, die sowohl aus geschlossenen als auch aus offenen Fragen bestand. Ausgewertet wurden selbstberichtete Überweisungsgründe, Behandlungsziele und therapeutische Interventionen der zuletzt behandelten Person mit SSc sowie die wahrgenommene Qualität der Kommunikation zwischen Gesundheitsfachkräften und Rheumatologen. Basierend auf den Antworten der 79 Therapeuten identifizierten wir 133 verschiedene Gründe für die Überweisung, von denen 70% mit der ICF-Komponente „Körperstrukturen und -funktionen" in Zusammenhang standen. Das breite Spektrum der berichteten Interventionen konzentrierte sich hingegen auf Körperfunktionen und -strukturen (28%), Training der täglichen Aktivitäten (26%), Patientenaufklärung und Beratung (26%) und psychosoziale Interventionen (20%). Ein Vergleich der berichteten Behandlungsoptionen von Gesundheitsfachkräften mit unerfüllten Bedürfnissen in der therapeutischen Versorgung, die von Personen mit SSc berichtet wurden (**Kapitel 2**), zeigte, dass neben zahlreichen anderen die drei am häufigsten genannten unerfüllten Bedürfnisse in der therapeutischen Versorgung abgebildet werden. Darüber hinaus fanden wir eine erhebliche inhaltliche Überschneidung der verschiedenen therapeutischen Disziplinen. Außerdem zeigte sich, dass therapeutische Behandlungsziele ungenügend mit den Überweisungsgründen der Rheumatologen übereinstimmen. Die Qualität der Kommunikation zwischen Gesundheitsfachkräften und Rheumatologen wurde als gering empfunden.

In **Kapitel 5** wurden multidisziplinäre Therapie-Empfehlungen zur nicht-pharmakologischen Behandlung von Fatigue, Handfunktionsverlust, Raynaud-Phänomen und digitalen Ulzera bei Personen mit SSc entwickelt. Die Empfehlungen wurden mit Hilfe von Forschungsergebnissen und Konsens unter Experten erstellt, basierend auf den standardisierten Arbeitsanweisungen der EULAR (European Alliance of Associations for Rheumatology). Die Task Force bestand aus sieben Personen mit SSc und 15 erfahrenen Behandlern und Forschern (Rheumatologen, Internisten/klinische Immunologen, Physiotherapeuten, Ergotherapeuten, Psychologen, Ernährungsberater, Dentalhygieniker, spezialisierte Krankenschwester, Sozialarbeiter und Forscher) in den Niederlanden. In einem Face-to-Face-Task-Force-Meeting wurden Empfehlungsentwürfe durch eine systematisch strukturierte Diskussion basierend auf der nominellen Gruppentechnik erstellt. Um die Empfehlungen zu untermauern, wurde eine umfangreiche Literaturrecherche durchgeführt und 20 wichtige systematische Reviews, RCTs und veröffentlichte Empfehlungen und Richtlinien ausgewählt. Darüber hinaus wurden 13 niederländische Fachärzte zu therapeutischen Empfehlungen zum Raynaud-Phänomen und digitalen Ulzera konsultiert. Für jede Empfehlung wurde der Grad der Evidenz und der Grad der Zustimmung bestimmt. Insgesamt wurden 34 Empfehlungen zu Behandlungen und Patientenaufklärung bei Fatigue, Handfunktionsverlust und Raynaud-Phänomen/digitalen Ulzera entwickelt und von der Task Force angenommen.

Schlussfolgerungen

Die Studien dieser Arbeit zeigen Ansätze auf, wie eine verbesserte nichtmedikamentöse Behandlung in der therapeutischen SSc-Versorgung gelingen kann. Diese Ansatzpunkte können dazu beitragen, eine zugänglichere und effektivere therapeutische Gesundheitsversorgung zu ermöglichen, und können letztendlich zu einer verbesserten Lebensqualität von Personen mit SSc führen. Die multidisziplinären Empfehlungen zur Patientenaufklärung und Behandlung von häufig berichteten unerfüllten Bedürfnissen in der therapeutischen Versorgung Betroffenen können zu einer gestärkten Rolle von Gesundheitsfachkräften bei der multidisziplinären Behandlung und gezielteren Überweisungen von Betroffenen an Gesundheitsfachkräfte beitragen. Sie können auch zu einer besseren Qualität der Kommunikation und einer verbesserten Organisation der SSc-bezogenen therapeutischen Gesundheitsversorgung beitragen.



Dankwoord

Met dit proefschrift gaat mijn promotietraject ten einde. Onderzoek doe je nooit alleen! Graag maak ik van de gelegenheid gebruik om iedereen die hieraan heeft bijgedragen te bedanken. Allereerst wil ik alle ervaringsdeskundigen met systemische sclerose en zorgprofessionals voor hun deelname aan de onderzoeken in dit proefschrift bedanken.

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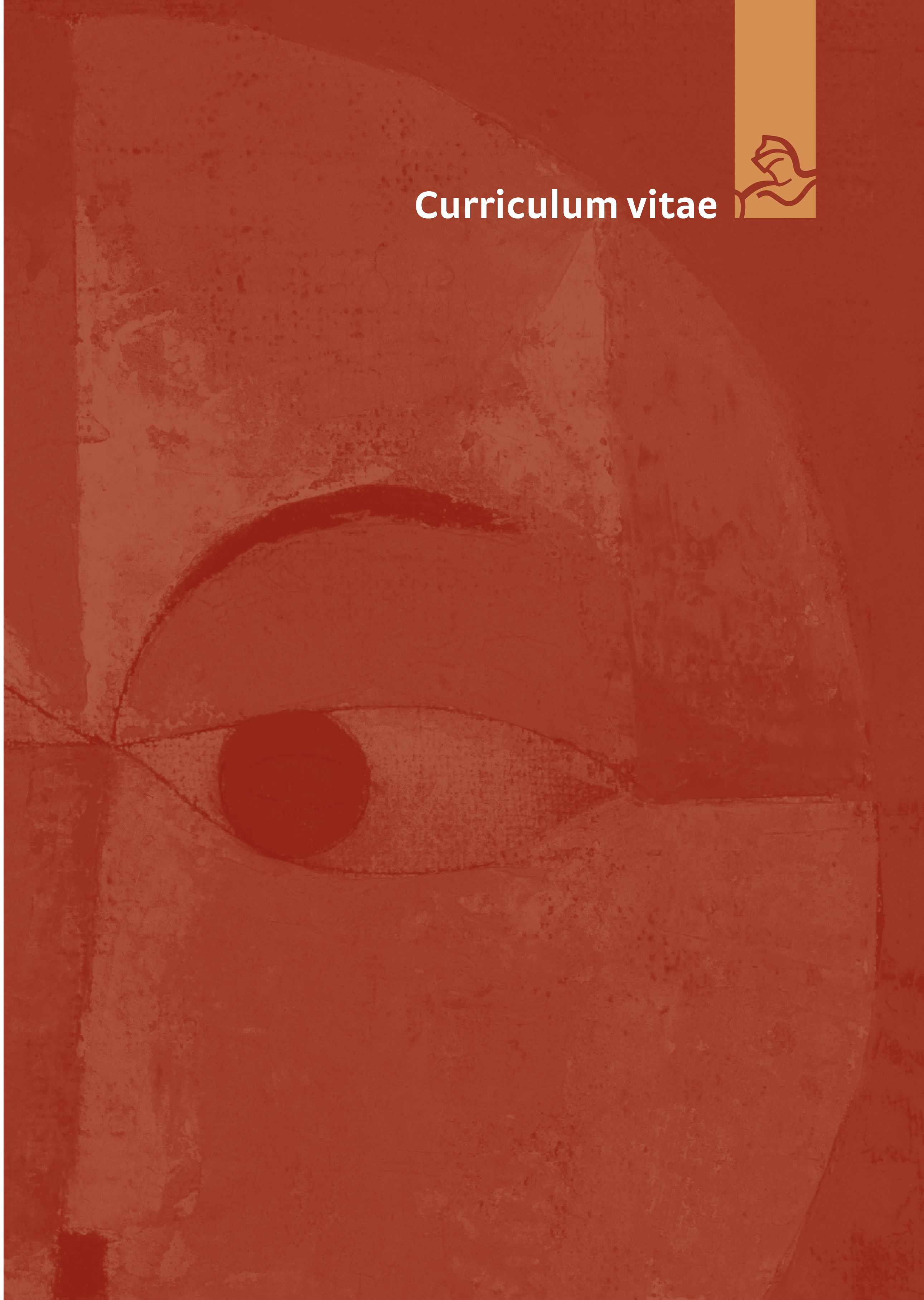
Juliane

Curriculum vitae



Juliane Kerstin Stöcker werd geboren op 15 oktober 1981 in Kreuztal, Duitsland. Na het afronden van de middelbare school in 1999, startte zij in datzelfde jaar met de opleiding tot ergotherapeut welke zij in 2002 afrondde. In 2003 begon haar carrière als ergotherapeut in de Baumrainklinik, een Revalidatiecentrum voor conservatieve orthopedie, traumatologie, gehoorstoornissen, tinnitus en interne geneeskunde in Bad Berleburg, Duitsland. Tijdens haar werk in de Baumrainklinik raakte ze geïnteresseerd in het werken met mensen met systemische sclerose.

In 2004 verhuisde zij naar Zwitserland waar zij tot 2011 als ergotherapeut voor het Institut für Physikalische Medizin aan het Universitair Ziekenhuis in Zürich werkte. Hier werkte zij in de reumatologiezorg en chronische pijn revalidatie, daarnaast initieerde zij het multidisciplinaire sclerodermie assessment.

Vanaf 2008 combineerde zij deze baan met de studie tot European Master of Science in Occupational Therapy die zij in 2010 met haar thesis getiteld ‘Systemic sclerosis: The patient perspective on daily occupations’ afrondde.

In 2011 verhuisde Juliane naar Nederland waar zij als docent bij de bachelor opleiding ergotherapie aan de Hogeschool van Arnhem en Nijmegen (HAN) aan de slag ging. In 2015 ontving ze een lerarenbeurs van de Nederlandse Organisatie voor Wetenschappelijk Onderzoek (NWO) voor haar promotie en startte ze een deeltijdonderzoeksfunctie bij de Sint Maartenskliniek in samenwerking met de HAN en het Radboudumc.

Het resultaat van haar onderzoek is beschreven in dit proefschrift, is gepubliceerd in peer-reviewd internationale tijdschriften en heeft geleid tot verschillende presentaties op (inter) nationale congressen. Juliane is lid van de Nederlandse health professionals in de reumatologie (NHPR) en was lid van het European League Against Rheumatism (EULAR) orphan disease programme Grants Advisory Council (GAC).

List of publications

Stöcker JK, Schouffoer AA, Spierings J, Schriemer MR, Potjewijd J, Pundert L, van den Hoogen FHJ, Nijhuis-van der Sanden MWG, Staal JB, Satink T, Vonk MC, van den Ende CHM & the Arthritis Research and Collaboration Hub study group. 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. Rheumatology 2022, 61(4), 1476-1486.

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Stöcker JK, Cup EHC, Vonk MC, vanden Hoogen FHJ, Nijhuis-van der Sanden MWG, Staal JB, van den Ende CHM. What moves the rheumatologist? Unravelling decision making in the referral of systemic sclerosis patients to health professionals: a qualitative study. Rheumatology Advances in Practice 2018, 2(2), rky027.

Mattsson M, Boström C, Mihai C, **Stöcker JK**, Geyh S, Stummvoll G, Gard G, Möller B, Hesselstrand R, Sandqvist G, Draghicescu O, Gherghe AM, Voicu M, Distler O, Smolen JS, Stamm TA. Personal factors in systemic sclerosis and their coverage by patient-reported outcome measures: A multicentre European qualitative study and literature review. European Journal Of Physical And Rehabilitation Medicine 2015, 51(4), 405-421.

Stamm TA, Mattsson M, Mihai C, **Stöcker JK**, Binder A, Bauernfeind B, Stummvoll G, Gard G, Hesselstrand R, Sandqvist G, Draghicescu O, Gherghe AM, Voicu M, Machold KP, Distler O, Smolen J S, Boström C. Concepts of functioning and health important to people with systemic sclerosis: a qualitative study in four European countries. Annals of Rheumatic Diseases 2011; 70:1074-1079.

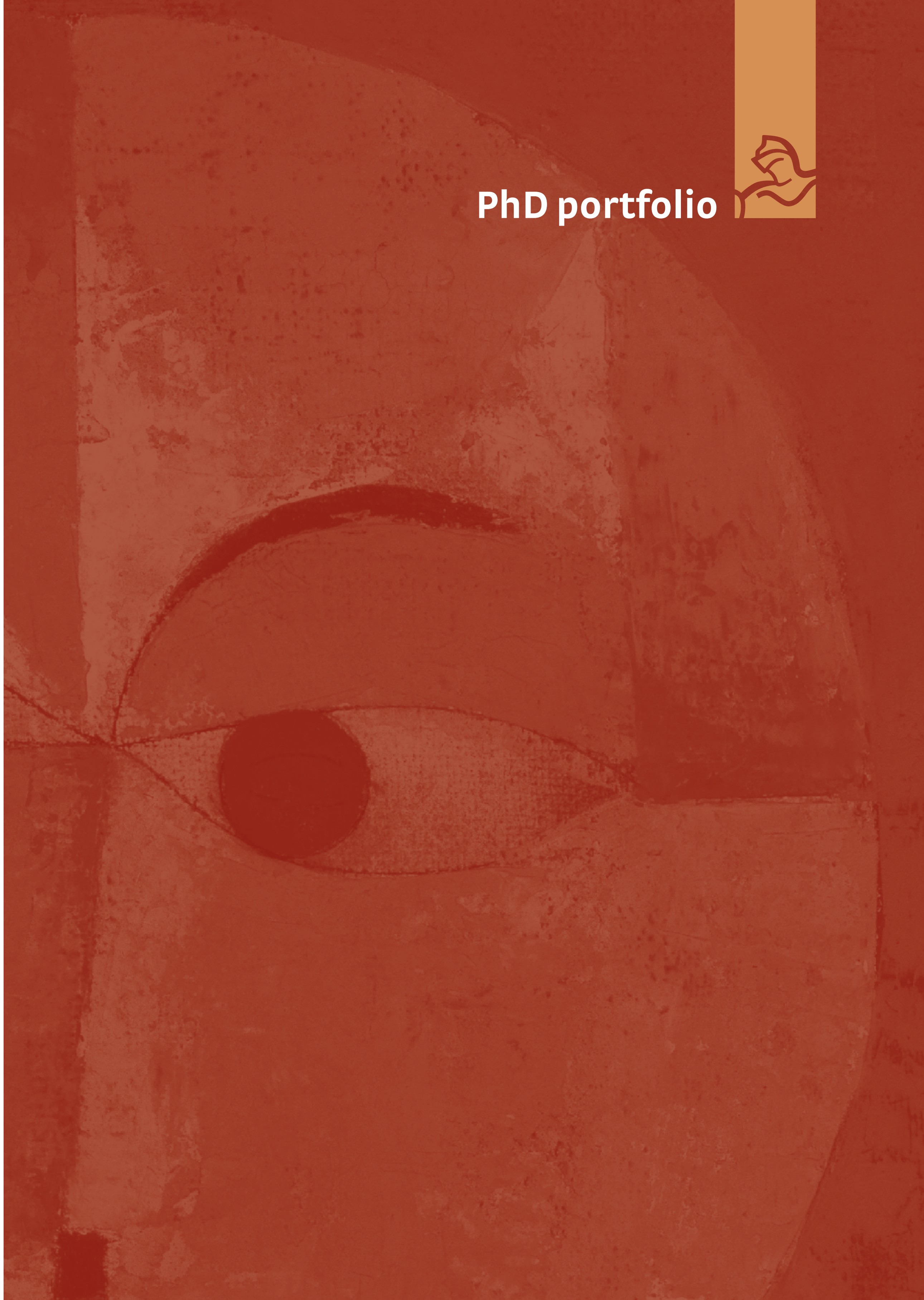
Stöcker JK, Koppermann I, Distler O. Optimierte interdisziplinäre Untersuchungen bei der systemischen Sklerose (SSc) – Das SSc- Assessment. Rheuma Suisse 2010; 1(2):9-12.

Research data management

All studies presented in this thesis are part of the scholarship research programme for teachers which is financed by the Dutch Research Council (NWO). They were conducted in accordance with the principles of the declaration of Helsinki. For all studies, the review board of the Arnhem-Nijmegen Region Medical Research Ethical Committee provided a waiver as they did not fall within the remit of the law of the Medical Research Involving Human Subjects Act.

All participants were well informed about the content, potential impact and publication of the data prior to the studies. Written informed consent was obtained by all participants prior to any study procedures. The privacy of the participants of all studies of this thesis is warranted by the use of study codes. Documents linking the study codes to personally identifiable information have been stored and protected digitally. Only researchers who were directly involved in the studies have access to these documents.

All data raw and processed data are stored on the Sint Maartenskliniek department server at V:/_reuma_research_studies and V:/_reuma_research_archief. The data will be stored for 15 years after termination of the study concerned. The use of these data for future research is only possible after renewed permission by the participants as recorded in their informed consents.



PhD portfolio

PhD portfolio of Juliane Kerstin Stöcker

Department: Reumatologie
PhD period: 01/01/2016 – 09/03/2023
PhD Supervisor(s): Prof. dr. F.H.J. van den Hoogen, Prof. dr. M.W.G. Nijhuis – van der Sanden
PhD Co-supervisor(s): Dr. C.H.M. van den Ende, Dr. J.B. Staal

Training activities	Hours
Courses	
RIHS - Introduction course for PhD candidates (2016)	15.00
Radboudumc - Scientific integrity (2020)	20.00
BROK course (2018)	10.00
RIHS – Introduction course for PhD Candidates (2017)	8.00
Scientific integrity course (2020)	16.00
Principles of Research in Medicine and Epidemiology (2016)	40.00
Thesis writing week (2020)	20.00
Seminars	
Journal clubs (2016-2022)	36.00
Research meetings (2016-2022)	36.00
Conferences	
SSc symposium (SMK) (2016)	5.00
EULAR London congress poster presentation (2016)	14.00
EULAR Madrid congress poster presentation (2017)	14.00
EULAR Amsterdam congress guided poster tour (2018)	14.00
EULAR Madrid congress oral presentation and chair (2019)	20.00
NVR Najaarsdagen Arnhem (2016-2019)	32.00
NVR Najaarsdagen oral presentation (2022)	14.00
Other	
Member of the Occupational Therapy Research Network Nijmegen (NEON (2016-2023)	40.00
Member of Nederlandse HealthProfessionals in de Reumatologie (NHPR) (2016-2023)	40.00
Reviewing scientific publications (2019)	8.00
Member of the Werkzame Factoren in Fysiotherapie en Paramedisch Handelen research groep, HAN University of Applied Sciences (2016-2023)	20.00
Teaching activities	
Lecturing	
Teaching in OT Bachelor of Science education 3 days a week (2016-2023)	4000.00
Lectures on qualitative and design thinking research methodology (2016-2023)	120.00
Supervision of internships / other	
Supervision of OT Bachelor students in research projects (2016-2023)	448.00
Supervision of interprofessional Bachelor students in research projects (2022-2023)	80.00
Supervision Bachelor project 'Systemische sclerose: Ergotherapeutische beleving, expertise en ervaring' (2016)	20.00
Total	**5090.00**

Theses Sint Maartenskliniek

Theses Sint Maartenskliniek

Mulder, M. (2022). *Going off-road. Exploring and mapping psoriatic arthritis.* Radboud University Nijmegen, Nijmegen. The Netherlands.

Huiskes, V. (2022). *The synergistic role of patients and healthcare providers in reducing drug-related problems.* Radboud University Nijmegen, Nijmegen. The Netherlands.

Marsman, D. (2022). *Polymyalgia rheumatica. Clinical characteristics and new treatment opportunities.* Radboud University Nijmegen, Nijmegen. The Netherlands.

Alingh, J. (2021). *Effect of robotic gait training on the post-stroke gait pattern. Evaluation of LOPES II.* Radboud University Nijmegen, Nijmegen. The Netherlands.

Van Dijsseldonk, R. (2021). *Step into the future: mobility after spinal cord injury*. Radboud University Nijmegen, Nijmegen, The Netherlands.

Pelle, T. (2021). *Beating osteoarthritis by e-self management in knee or hip osteoarthritis.* Radboud University Nijmegen, Nijmegen. The Netherlands.

Van Heuckelum, M (2020). *Novel approaches to improve medication adherence in rheumatoid arthritis.* Radboud University Nijmegen, Nijmegen. The Netherlands.

Mathijssen, E. (2020). *The voice of patients with rheumatoid arthritis.* Radboud University Nijmegen, Nijmegen. The Netherlands.

Bakker, S. (2019). *Regional anesthesia and total knee arthroplasty. Anesthetic and pharmacological considerations.* Radboud University Nijmegen, Nijmegen. The Netherlands.

Claassen, A. (2019). *Strategies for patient education in rheumatic diseases.* Radboud University Nijmegen, Nijmegen. The Netherlands.

Fenten, M. (2019). *Optimizing locoregional anesthesia in fast track orthopaedic surgery.* Radboud University Nijmegen, Nijmegen. The Netherlands.

Minten, M. (2019). *On the role of inflammation and the value of low dose radiation therapy in osteoarthritis.* Radboud University Nijmegen, Nijmegen. The Netherlands.

Verhoef, L. (2019). *Effective and efficient use of bDMARDs in rheumatoid arthritis.* Radboud University Nijmegen, Nijmegen. The Netherlands.

Bekker, C. (2018). *Sustainable use of medication. Medication waste and feasibility of redispensing.* Utrecht University, Utrecht. The Netherlands.

Bikker, I.(2018). *Organizing timely treatment in multi-disciplinary care.* University of Twente, The Netherlands.

Bouman, C. (2018). *Dose optimisation of biologic DMARDs in rheumatoid arthritis: long-term effects and possible predictors.* Radboud University Nijmegen, The Netherlands.

Mahler, E. (2018). *Contributors to the management of osteoarthritis.* Utrecht University, The Netherlands.

Tweehuysen, L. (2018). *Optimising biological treatment in inflammatory rheumatic diseases. Predicting, tapering and transitioning.* Radboud University Nijmegen, Nijmegen, The Netherlands.

Geerdink, Y. (2017). *Getting a grip on hand use in unilateral cerebral palsy.* Radboud University, Nijmegen, The Netherlands.

Remijn, L. (2017). *Mastication in children with cerebral palsy.* Radboud University, Nijmegen, The Netherlands.

Selten, E. (2017). *Beliefs underlying treatment choices in osteoarthritis.* Radboud University, Nijmegen, The Netherlands.

Van Hooff, M. (2017). *Towards a paradigm shift in chronic low back pain? Identification of patient profiles to guide treatment.* VU University Amsterdam, Amsterdam, The Netherlands.

Lesuis, N. (2016). *Quality of care in rheumatology. Translating evidence into practice.* Radboud University, Nijmegen, The Netherlands.

Luites, J. (2016). *Innovations in femoral tunnel positioning for anatomical ACL reconstruction.* Radboud University, Nijmegen, The Netherlands.

Pakvis, D. (2016). *Survival, primary stability and bone remodeling assessment of cementless sockets. An appraisal of Wolff's law in the acetabulum*. Radboud University, Nijmegen, The Netherlands.

Schoenmakers, K. (2016). *Prolongation of regional anesthesia. Determinants of peripheral nerve block duration*. Radboud University, Nijmegen, The Netherlands.

Altmann, V. (2015). *Impact of trunk impairment on activity limitation with a focus on wheelchair rugby.* Leuven University, Leuven, Belgium.

Bevers, K. (2015). *Pathophysiologic and prognostic value of ultrasonography in knee osteoarthritis.* Utrecht University, Utrecht, The Netherlands.

Cuperus, N. (2015). *Strategies to improve non-pharmacological care in generalized osteoarthritis.* Radboud University, Nijmegen, The Netherlands.

Kilkens, A. (2015). *De ontwikkeling en evaluatie van het Communicatie Assessment & Interventie Systeem (CAIS) voor het aanleren van (proto-)imperatief gedrag aan kinderen met complexe ontwikkelingsproblemen.* Radboud University, Nijmegen, The Netherlands.

Penning, L. (2015). *The effectiveness of injections in cuffdisorders and improvement of diagnostics.* Maastricht University, Maastricht, The Netherlands.

Stegeman, M. (2015). *Fusion of the tarsal joints: outcome, diagnostics and management of patient expectations.* Utrecht University, Utrecht, The Netherlands.

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Wiegant, K. (2015). *Uitstel kunstknie door kniedistractie.* Utrecht University, Utrecht, The Netherlands.

Willems, L. (2015). *Non-pharmacological care for patients with systemic sclerosis.* Radboud University, Nijmegen, The Netherlands.

Witteveen, A. (2015). *The conservative treatment of ankle osteoarthritis.* University of Amsterdam, Amsterdam, The Netherlands.

Zwikker, H. (2015). *All about beliefs. Exploring and intervening on beliefs about medication to improve adherence in patients with rheumatoid arthritis.* Radboud University, Nijmegen, The Netherlands.

Koenraadt, K. (2014). *Shedding light on cortical control of movement.* Radboud University, Nijmegen, The Netherlands.

Smink, A. (2014). *Beating Osteoarthritis. Implementation of a stepped care strategy to manage hip or knee osteoarthritis in clinical practice.* VU University Amsterdam, Amsterdam, The Netherlands.

Stolwijk, N. (2014). *Feet 4 feet. Plantar pressure and kinematics of the healthy and painful foot.* Radboud University, Nijmegen, The Netherlands.

Van Kessel, M. (2014). *Nothing left? How to keep on the right track. Spatial and non-spatial attention processes in neglect after stroke.* Radboud University, Nijmegen, The Netherlands.

Brinkman, M. (2013). *Fixation stability and new surgical concepts of osteotomies around the knee.* Utrecht University, Utrecht, The Netherlands.

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Severens, M. (2013). *Towards clinical BCI applications: assistive technology and gait rehabilitation.* Radboud University, Nijmegen, The Netherlands.

Stukstette, M. (2013). *Understanding and treating hand osteoarthritis: a challenge.* Utrecht University, Utrecht, The Netherlands.

Van der Maas, A. (2013). *Dose reduction of TNF blockers in Rheumatoid Arthritis: clinical and pharmacological aspects.* Radboud University, Nijmegen, The Netherlands.

Zedlitz, A. (2013). *Brittle brain power. Post-stroke fatigue, explorations into assessment and treatment.* Radboud University, Nijmegen, The Netherlands.

Beijer, L. (2012). *E-learning based speech therapy (EST). Exploring the potentials of E-health for dysarthric speakers.* Radboud University, Nijmegen, The Netherlands.

Hoogeboom, T. (2012). *Tailoring conservative care in osteoarthritis.* Maastricht University, Maastricht, The Netherlands.

Boelen, D. (2011). *Order out of chaos? Assessment and treatment of executive disorders in brain-injured patients.* Radboud University, Nijmegen, The Netherlands.

Heesterbeek, P. (2011). *Mind the gaps! Clinical and technical aspects of PCL-retaining total knee replacement with the balanced gap technique.* Radboud University, Nijmegen, The Netherlands.

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Smulders, E. (2011). *Falls in rheumatic diseases. Risk factors and preventive strategies in osteoporosis and rheumatoid arthritis.* Radboud University, Nijmegen, The Netherlands.

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Vriezekolk, J. (2011). *Targeting distress in rheumatic diseases.* Utrecht University, Utrecht, The Netherlands.

Willems, P. (2011). *Decision making in surgical treatment of chronic low back pain. The performance of prognostic tests to select patients for lumbar spinal fusion.* Maastricht University, Maastricht, The Netherlands.

Aarts, P. (2010). *Modified constraint-induced movement therapy for children with unilateral spastic cerebral palsy: the Pirate group intervention.* Radboud University, Nijmegen, The Netherlands.

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Ruiter, M. (2008). *Speaking in ellipses. The effect of a compensatory style of speech on functional communication in chronic agrammatism.* Radboud University, Nijmegen, The Netherlands.

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De Haart, M. (2005). *Recovery of standing balance in patients with a supratentorial stroke.* Radboud University, Nijmegen, The Netherlands.

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Jongerius, P. (2004). *Botulinum toxin type-A to treat drooling. A study in children with cerebral palsy.* Radboud University, Nijmegen, The Netherlands.

Van de Crommert, H. (2004). *Sensory control of gait and its relation to locomotion after a spinal cord injury.* Radboud University, Nijmegen, The Netherlands.

Van der Linde, H. (2004). *Prosthetic prescription in lower limb amputation. Development of a clinical guideline in the Netherlands.* Groningen University, Groningen, The Netherlands.

Hendricks, H. (2003). *Motor evoked potentials in predicting motor and functional outcome after stroke.* University of Nijmegen, Nijmegen, The Netherlands.

Hosman, A. J. F. (2003). *Idiopathic thoracic spinal deformities and compensatory mechanisms.* University of Nijmegen, Nijmegen, The Netherlands.

Donker, S. (2002). F*lexibility of human walking: a study on interlimb coordination.* Groningen University, Groningen, The Netherlands.

Hochstenbach, J. (1999). *The cognitive, emotional, and behavioural consequences of stroke.* University of Nijmegen, The Netherlands.

De Kleuver, M. (1998). *Triple osteotomy of the pelvis. An anatomical, biomechanical and clinical study.* University of Nijmegen, Nijmegen, The Netherlands.

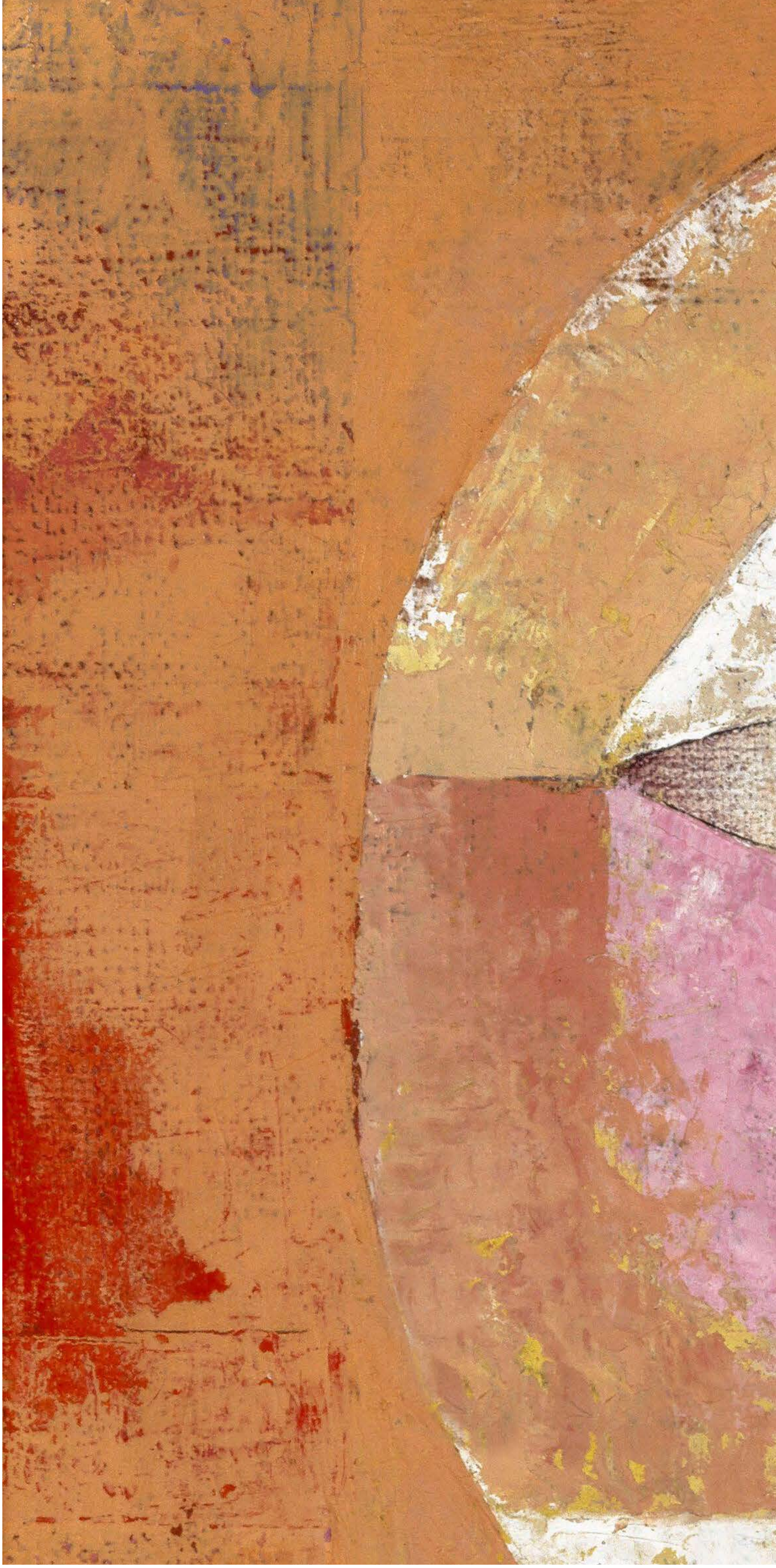
Van Balen, H. (1997). *A disability-oriented approach to long-term sequelae following traumatic brain injury. Neuropsychological assessment for post-acute rehabilitation*. University of Nijmegen, Nijmegen, The Netherlands.

Tromp, E. (1995). *Neglect in action: a neuropsychological exploration of some behavioural aspects of neglect.* University of Nijmegen, Nijmegen, The Netherlands.

Van Lankveld, W. (1993). *Coping with chronic stressors of rheumatoid arthritis.* University of Nijmegen, Nijmegen, The Netherlands.

Geurts, A. (1992). *Central adaptation of postural organization to peripheral sensorimotor impairments.* University of Nijmegen, Nijmegen, The Netherlands.

De Rooij, D. (1988). *Clinical and serological studies in the connective tissue diseases.* University of Nijmegen, Nijmegen, The Netherlands.



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