Published in final edited form as:
Clin Exp Rheumatol. 2012 ; 30(2 0 71): S23–S29.

New directions for patient-centred care in scleroderma: the Scleroderma Patient-centred Intervention Network (SPIN)

Brett D. Thombs, PhD[1,2,3,7,8,9,10], **Lisa R. Jewett, MSc**[8,9], **Shervin Assassi, MD, MS**[11], **Murray Baron, MD**[3,8,10], **Susan J. Bartlett, PhD**[3], **Angela Costa Maia, PhD**[12], **Ghassan El-Baalbaki, PhD**[1,8,9], **Daniel E. Furst, MD**[13], **Karen Gottesman, BA**[14], **Jennifer A. Haythornthwaite, PhD**[15], **Marie Hudson, MD, MPH**[3,8,10], **PhD Ann Impens, MPH**[16], **Annett Korner, PhD**[5,6,8], **Catarina Leite, MSc**[12,17], **Maureen D. Mayes, MD, MPH**[11], **Vanessa L. Malcarne, PhD**[18], **Sarosh J. Motivala, PhD**[19], **Luc Mouthon, MD, PhD**[20,21], **Warren R. Nielson, PhD**[23], **Diane Plante**[24], **Serge Poiraudeau, MD, PhD**[20,22,25], **Janet L. Poole, PhD, OTR/L**[26], **Janet Pope, MD, MPH**[27], **Maureen Sauve, BA**[28,29], **Russell J. Steele, PhD**[4,8], **Maria E. Suarez-Almazor, MD, PhD**[30], **Suzanne Taillefer, PhD**[8,10], **Cornelia H. van den Ende, PhD**[31], **BSc Erin Arthurs**[8], **Marielle Bassel, BA**[8], **Vanessa Delisle, BSc**[1,8], **Katherine Milette, BSc**[5,8], **Allison Leavens, BSc**[8], **Ilya Razykov, BA**[1,8], and **Dinesh Khanna, MD, MS**[16]

[1]Department of Psychiatry, McGill University, Montréal, Québec, Canada [2]Department of Epidemiology, Biostatistics, and Occupational Health, McGill University, Montréal, Québec, Canada [3]Department of Medicine, McGill University, Montréal, Québec, Canada [4]Department of Mathematics and Statistics, McGill University, Montréal, Québec, Canada [5]Department of Educational and Counselling Psychology, McGill University, Montréal, Québec, Canada [6]Department of Oncology, McGill University, Montréal, Québec, Canada [7]School of Nursing, McGill University, Montréal, Québec, Canada [8]Lady Davis Institute for Medical Research, Jewish General Hospital, Montréal, Québec, Canada [9]Department of Psychiatry, Jewish General Hospital, Montréal, Québec, Canada [10]Division of Rheumatology, Jewish General Hospital, Montréal, Québec, Canada [11]University of Texas Health Science Center Houston, TX, USA [12]CIPSI – School of Psychology, University of Minho, Portugal [13]Division of Rheumatology, Geffen School of Medicine at the University of California in Los Angeles, USA [14]Scleroderma Foundation, Southern California Chapter, USA [15]Department of Psychiatry & Behavioral Sciences, Johns Hopkins University, Baltimore, MD [16]University of Michigan Scleroderma Program, Ann Arbor, MI [17]Federation of European Scleroderma Associations [18]Department of Psychology, San Diego State University, San Diego, California, USA [19]Cousins Center for Psychoneuroimmunology, UCLA Semel Institute, Los Angeles, California, USA [20]Université Paris Descartes, Paris, France; Assistance Publique-Hôpitaux de Paris (AP-HP) [21]Pôle de Médecine Interne, hôpital Cochin, Paris, France [22]Pôle Ostéoarticulaire, hôpital Cochin, Paris, France [23]Beryl & Richard Ivey Rheumatology Day Programs, St. Joseph's Health Care, London Ontario [24]Sclérodermie Québec, Montréal, Québec, Canada [25]IFR Handicap INSERM, Paris, France [26]Occupational Therapy Graduate Program, University of New Mexico, USA [27]University of Western Ontario, St. Joseph's Health Care, London, Ontario, Canada [28]Scleroderma Society of Canada, Ottawa, Ontario, Canada [29]Scleroderma Society of Ontario, Hamilton, Ontario, Canada [30]University of Texas MD Anderson Cancer Center, Houston, Texas, USA [31]Department of Rheumatology, Sint Maartenskliniek, The Netherlands

Abstract

Please address correspondence to: Brett D. Thombs, PhD, Jewish General Hospital, 4333 Cote Ste Catherine Road, Montreal, Quebec H3T 1E4, Canada. brett.thombs@mcgill.ca.

Systemic sclerosis (SSc), or scleroderma, is a chronic multisystem autoimmune disorder characterised by thickening and fibrosis of the skin and by the involvement of internal organs such as the lungs, kidneys, gastrointestinal tract, and heart. Because there is no cure, feasibly-implemented and easily accessible evidence-based interventions to improve health-related quality of life (HRQoL) are needed. Due to a lack of evidence, however, specific recommendations have not been made regarding non-pharmacological interventions (e.g. behavioural/psychological, educational, physical/occupational therapy) to improve HRQoL in SSc. The Scleroderma Patient-centred Intervention Network (SPIN) was recently organised to address this gap. SPIN is comprised of patient representatives, clinicians, and researchers from Canada, the USA, and Europe. The goal of SPIN, as described in this article, is to develop, test, and disseminate a set of accessible interventions designed to complement standard care in order to improve HRQoL outcomes in SSc.

Keywords

scleroderma; psychosocial; health-related quality of life; patient-centred care

Introduction

More than 25 million people in North America and more than 30 million in Europe are affected by a rare disease, defined in the United States as a condition with fewer than 7 prevalent cases per 10,000 people and by the European Union as fewer than 5 prevalent cases per 10,000 (1). Patients with rare diseases face many challenges, perhaps foremost among these, the lack of rigorously tested interventions and evidence-based care standards to direct patient care (2, 3). Indeed, the complexities involved in ensuring that patients with rare diseases have access to effective drugs is well documented (1). Beyond drug therapies, organisations that represent patients with rare diseases often have a clear idea of what kind of disease-specific supportive care is needed and how it might be provided, but these ideas are rarely translated into accessible, evidence-based, patient-centred care programmes (2).

In order to develop an evidence-based patient-centred care programme for patients with a rare disease, an active partnership among researchers, health care providers, and patients is necessary (4, 5). Furthermore, effective research in rare diseases requires successful multicentre collaborations. The NIH, for instance, created the Rare Diseases Clinical Research Network to support research on rare diseases. In systemic sclerosis (SSc), or scleroderma, there are several examples of ongoing collaborative efforts. The international Scleroderma Clinical Trials Consortium conducts multicentre drug trials. There are national SSc registries in the UK, Germany, USA, Brazil, Australia, and Canada, and there is an international registry, established by the European League Against Rheumatism Scleroderma Trial and Research (EUSTAR), which consists of minimal essential data from over 8,000 patients across 92 centres (6). None of these, however, focuses on developing evidence-based patient-centred care interventions.

SSc is characterised clinically by thickening and fibrosis of the skin and by the involvement of internal organs, most commonly the lungs, gastrointestinal tract and heart (7, 8). Prevalence estimates vary from less than 0.1 to 4.9 cases per 10,000 (9) with the disease at least 4 times more prevalent in women than men (10). The pathogenesis and etiology of SSc are complex and not completely understood. However, vascular damage, immunological/inflammatory system activation, and excessive collagen production are known to be important in the development of the disease (11). Median survival time from diagnosis is approximately 11 years, and patients are 3.7 times more likely to die within 10 years of diagnosis (44.9% mortality) than age, sex, and race-matched individuals without SSc (12.0%

mortality) (12). The effects of SSc often result in significant disruptions to activities of daily living and can negatively affect quality of life, including physical and psychological well-being (13, 14). While treatments for some of the manifestations of the disease are available, there are no cures or true disease modifying agents for a majority of SSc complications (15). Given this, there is an urgent need for interventions to improve HRQoL.

Despite the marked physical, medical, and functional changes experienced by persons with SSc, HRQoL has received relatively little attention in SSc compared to other rheumatic diseases. Clinical trial data related to behavioural/psychological, educational, and physical/occupational therapy interventions are lacking, and the most recent management guidelines for SSc (15) were not able to make recommendations for or against these types of interventions. Nonetheless, a number of key problems have been identified that contribute to HRQoL for many people with SSc and are potentially amenable to interventions with non-pharmacological, adjunctive health care strategies.

Common problems in SSc potentially addressed by adjunctive health care interventions

SSc is notable for the many different problems faced by people living with the disease. A number of these problems have been reported by patients to significantly influence health and well-being, including limitations in physical mobility; pain; fatigue; sleep disturbance; psychological distress including depression and anxiety; sexual dysfunction; and body image distress (13, 14, 16–18).

Physical mobility/hand function

SSc results in a range of physical limitations, including difficulty with eating and speaking, reductions in exercise tolerance and muscle strength, and impaired hand function. Contractures and deformities of the hand, consisting of decreased flexion and limited extension as well as reduced thumb abduction, are common among people with SSc (19), and limited hand function is an important contributor to overall disability and reduced HRQoL (20, 21). A recent Canadian national survey (n=464) found that more than two-thirds of SSc patients reported experiencing hand stiffness, difficulty making a fist, or difficulty holding objects that impacted daily activities at least some of the time (16). Problems related to hand function were among the top in both frequency and impact, among the 69 symptoms/problems in the survey checklist. Not surprisingly, another recent study reported that 89% of the SSc patients sampled had problems with hand function, as skin and tendon involvement of the hands is nearly universal in SSc (22). There have been 3 randomised controlled trials (RCTs) of physical or occupational therapy interventions to improve hand function in SSc, but none has included more than 20 patients per treatment or control groups (23). Recently, Schouffoer *et al*. (24) reported that a 12-week multidisciplinary day patient treatment programme that included a physical therapy component yielded greater improvements in grip strength and on the Scleroderma Health Assessment Questionnaire (SHAQ) for 28 patients who received the intervention, compared to 25 patients who received usual care.

Pain

Pain levels in SSc are similar to those reported in other chronic pain and rheumatic conditions (25–27). Between 60–83% of SSc patients experience pain (25, 28), and it is a significant source of distress, reduced physical and social functioning, and disability for many people living with the disease (13, 25, 28). Pain in SSc may have numerous sources, including skin pain; pain associated with Raynaud's phenomenon; musculoskeletal pain;

pain in distal extremities due to tightness, calcinosis, and ulcers; gastrointestinal problems, as well as depressive symptoms (13, 14, 25, 29).

Fatigue

Levels of fatigue in SSc are similar to those reported by patients with other rheumatic diseases and cancer patients undergoing treatment, and higher than in the general population or among cancer patients in remission (30). Almost 90% of patients who responded to a Canadian survey (n=464) said that fatigue was present at least some of the time, and 72% said that fatigue had a moderate to severe impact on their ability to carry out daily activities (16). A Dutch study found that fatigue was present in 75% of 123 SSc patients and was reported as one of the most bothersome symptoms associated with the disease (31). Numerous studies have shown that fatigue in SSc is associated with the inability to carry out daily activities, work disability, and limited overall physical functioning, even after controlling for potential confounding variables such as education level, disease subtype, pain, sleep quality, and depressive symptoms (32–34). A number of medical comorbidities are associated with fatigue in SSc, including breathing and gastrointestinal problems, pain, and depression (35).

Sleep disturbance

Recent findings indicate that patients with SSc report marked sleep disturbance. One study found that compared to the US general population, patients with SSc reported significantly worse sleep quality and this poor sleep was associated with dyspnea, depressed mood, and severity of reflux symptoms (36). In another study, scores on a single-item sleep disruption scale were as high in Canadian SSc patients as those reported in rheumatoid arthritis patients, and higher than for the general population (37). Furthermore, pain was strongly associated with sleep disturbance. There is a need for more focused research on sleep problems in SSc.

Psychological distress/depression/anxiety

Depressive symptoms are another problem in SSc. One study of a French cohort found that 1 in 5 SSc patients met criteria for a current episode of Major Depressive Disorder (MDD) (38). The range of patients who report depressive symptoms or distress based on self-report questionnaires, which may or may not qualify as MDD, is understandably higher (ranging from 36–65% of patients above recommended cut-offs) (39). Cross-sectional studies have reported that sociodemographic factors (such as being unmarried and having lower education levels), disease variables (including overall disease severity, pain, more tender joints, breathing problems, and gastrointestinal functioning), and psychological factors (*e.g.* body image distress, poor social support), are associated with greater distress symptomatology (39–41).

Few studies have examined anxiety among patients with SSc, and all have involved small sample sizes and self-report questionnaires that assess levels of anxiety, but cannot determine the presence or absence of anxiety disorders (42–44). One Dutch study (n=123) reported that fear of the future, including the potential for uncontrolled and unpredictable disease progression, major loss of function, inability to work, dependency on others, and mortality were among the top concerns reported by SSc patients (31). More research is needed to determine the extent of these fears and their impact on people living with SSc, as well as the presence of anxiety disorders, more generally and the association between anxiety and other common HRQoL dimensions, such as pain, sleep disturbance, and body image distress (13).

Sexual dysfunction

The physical and psychological effects of SSc can lead to sexual dysfunction, including decreased desire and enjoyment, impaired arousal, and painful sex (45). For instance, skin tightening and discomfort, shrinking of the mouth, joint pain, Raynaud's phenomenon, gastrointestinal symptoms, vaginal tightness and dryness in women, and reduced penile blood flow in men are physical consequences of SSc that can impede sexual functioning (46–48). Emotional distress and depressive symptoms, as well as concerns about physical appearance, are psychological issues that may also impact sexual functioning among SSc patients. Although research in this area is limited, one study found that more than half of a sample of women with SSc reported impaired sexual function (47), and another reported that levels of sexual impairment in women with SSc are similar to or higher than levels for women with breast cancer, human immunodeficiency virus, or gynecologic cancer (49). Other factors associated with decreased sexual functioning in SSc include vaginal discomfort and pain, fatigue, disease duration, and overall marital dissatisfaction (50).

Body image distress

Acquired disfigurement from an injury or medical illness is often linked to problems with body image as well as social anxiety and avoidance (51). Appearance changes common in SSc often affect visible and socially relevant body parts, including the face and hands, and can potentially lead to challenges with social interactions. SSc patients have rated skin deformities as one of the most significant stressors associated with the disease (31). Existing studies have shown that more severe disease manifestations, such as significant skin changes in the hands, are associated with greater body image dissatisfaction, increased depressive symptoms, and reduced overall psychosocial functioning (31, 51, 52).

The Scleroderma Patient-centred Intervention Network (SPIN)

Given the well documented extent of problems related to HRQoL in SSc, we believe that SSc researchers are ready to embark on interventional studies to develop a patient-centred care programme for people with the disease. The first step towards this end occurred in the fall of 2008 when a panel of internationally-recognised experts in behavioural and psychological health in SSc, representatives from the Scleroderma Society of Canada, and Canadian Scleroderma Research Group researchers convened for a two-day meeting, funded by the Canadian Institutes of Health Research (CIHR). This meeting identified important areas of concern for people living with SSc related to HRQoL and developed a consensus research agenda, which was subsequently published (13). In the fall of 2010, SPIN was conceived, and its initial meeting was held in Montreal, also with funding from the CIHR. This meeting built upon the 2008 meeting and was organised with the purpose of laying the groundwork to develop an investigative infrastructure for implementing a patient-centred care programme and testing interventions targeting the HRQoL of people living with SSc. Meeting participants included researchers with expertise needed to develop and test patient-centred interventions focused on improving HRQoL in SSc; Canadian and international physician experts in SSc; and patient representatives from the Scleroderma Society of Canada, Scleroderma Society of Ontario, Sclérodermie Québec, the Scleroderma Foundation, and the Federation of European Scleroderma Associations (FESCA).

The core mission of SPIN is to develop and evaluate effective, feasible, easily accessible, cost-efficient non-pharmacological interventions for patients with SSc. Thus, SPIN's patient-centred research programme will capitalise on technological approaches; and interventions will be designed to be accessible via telecommunication such as instructional/downloadable videos, e-mail, internet or telephone, but will not require face-to-face contact with care providers, such as sessions with psychotherapists or customising splints and assistive

devices by physical or occupational therapists. SPIN will pilot each of the proposed interventions. Following this groundwork, the group will recruit patients through SPIN investigative centre sites and SPIN-affiliated patient organisations for participation in RCTs.

There are many areas where SSc patients would potentially benefit from accessible supportive care. As a first step, SPIN members prioritised areas that have been identified by patients as having a high impact on HRQoL and where there is enough basic research to confidently develop and test an intervention in SSc. An additional consideration involved selecting intervention areas where there is a SSc-specific component that would not likely be addressed by interventions that are generally applied across disease groups. Thus, current SPIN priorities include the development and testing of interventions to improve hand function, reduce symptoms of psychological distress, address body image concerns related to significant disfigurement, as well as to implement a general programme for self-management geared to the needs of SSc patients.

A. Overview of SPIN approach and research priorities

Innovations being proposed by SPIN that will facilitate the conduct of rigorous testing and subsequent delivery of prioritised supportive care interventions include (1) the elaboration of an international collaborative network of major SSc treatment and research centres and existing networks to conduct supportive care intervention trials; (2) the use of the cutting-edge *cohort multiple randomised controlled trial* (cmRCT) design (53), which was developed specifically for pragmatic trials conducted to inform clinical practice in (i) open trials where treatment as usual is compared with the offer of treatment, (ii) where rigorous blinding is not realistic, and (iii) where it is important to be able to conduct multiple trials; (3) capitalising on technological approaches including downloadable videos, e-mail, internet and telephone; and (4) partnering with patient organisations who, in collaboration with SPIN, will provide supportive care services to patients across a wide geographic range on an ongoing basis, following the initial development and testing stage.

The cmRCT design (53) was developed to address some of the shortcomings of pragmatic trial designs, which are highly relevant to SPIN, including recruitment difficulty inherent in the randomisation process for traditional RCTs; in nonpharmaceutical trials, the difficulty blinding patients and the “disappointment bias” that can influence outcome reporting among patients who are not assigned to the intervention; and the ability to conduct only a single randomised trial in a group of recruited patients. In the cmRCT design, a large observational cohort is recruited and a set of core outcomes is regularly measured in these patients. Patients consent upon enrollment in the cohort to provide data that can be used to evaluate the benefit of treatments for their condition. For a given RCT conducted with the cohort, eligible patients are identified, and a random selection of eligible patients is offered the trial intervention. Outcomes among patients who receive the intervention are then compared to outcomes among eligible patients who did not receive the intervention. Only patients who are offered the intervention are notified and consented for the trial. An important feature of the cmRCT design is that enrollees can participate in more than one study (though not at the same time) being conducted in the cohort, which is important given the difficulty recruiting patients with a rare disease, like SSc.

The first priority of SPIN will be to develop a set of supportive care interventions. As this is occurring, we will begin to recruit the SPIN cohort, from which patients will be recruited for specific SPIN intervention trials. An important part of the SPIN program will be its focus on outcome measures. The SPIN Measurement Core will rely upon the OMERACT (Outcome Measures in Rheumatology) framework (54, 55) to select the most appropriate outcome measures for each intervention, prioritising measures that have met or are close to meeting OMERACT criteria. An important task of the SPIN Measurement Core will be to use the

cohort and included interventions to conduct further validation work on important patient-oriented outcome measures. The following sections describe interventions that have been prioritised for initial development by SPIN.

B. Physical/occupational therapy to improve hand function

Several small RCTs have found that physical/occupational therapy interventions improve hand function in SSc (19). While the methods and outcome measures are reasonably well established, there remains a major gap in the testing and implementation of interventions that improve hand function, as well as appropriate care delivery. A Canadian Scleroderma Research Group needs survey identified that only 10% of Canadian patients with hand problems receive physical/occupational therapy (Bassel *et al*., under review). Patients often report that they cannot find therapists who are experienced in working with SSc or that they have had negative experiences with therapists unfamiliar with SSc. In order to address this, SPIN has prioritised a plan to develop and test a brief DVD/downloadable instructional video to educate physical/occupational therapists on exercises unique to SSc to improve hand function for patients. Patients in need of a physical/occupational therapist (PT/OT) will be able to share the instructional material with a PT/OT, which would support active SSc patient management and help address a major need. A PT/OT, after a brief, 10-minute DVD/online training session, will be able to deliver more effective therapy. Planned outcomes will include improved overall hand function, as assessed by Cochin Hand Function Scale, as well as increased joint motion, grip strength, and grip pinch (19). Additionally, the McMaster-Toronto Arthritis Patient Preference Disability Questionnaire (MACTAR) (56), a functional scale that includes patient priorities and has been used successfully in SSc (57), will be included as an outcome measure of the degree of difficulty experienced by patients when performing key activities of daily living.

C. Telephone-based peer support to address psychological distress

Peer support has been shown to be an effective first step to reduce symptoms of depression and prevent MDD in a number of patient groups (58, 59). An RCT of telephone-based peer support (59) among postpartum women (n=701) reduced symptoms of depression by almost two-fold. In that study, trained community volunteers who had previously experienced and recovered from postpartum depression provided telephone-based support. A similar approach would be well suited to SSc. Based on our preliminary work, it is clear that many people with SSc feel socially isolated and that they would benefit from talking openly with somebody who has experienced the problems they encounter. Although face-to-face support groups for people with SSc who live in metropolitan areas exist, they are generally poorly attended and are largely inaccessible to patients in suburban or rural areas. A telephone-or internet-based support intervention would reduce barriers to attendance, such as difficulty travelling due to disability from SSc and would be available to patients who live far from urban centres. In addition, telephone-based support is flexible, private, and non-stigmatising. The planned primary outcome will be symptoms of depression, as measured with the Patient Health Questionnaire-9 (PHQ-9), which has been validated for use in SSc (40).

D. Internet-based group support to reduce body image distress

Social skills training programmes (*e.g.* 60, 61) and cognitive-behavioural techniques (62) have been recommended as strategies to reduce social avoidance resulting from disfigurement and appearance-related concerns among groups with acquired disfigurement. Changing Faces, a non-profit organisation in the United Kingdom, has been a leader in developing workshop-based interventions for persons with visible disfigurement, with a focus on social skills training and cognitive behavioural therapy for social anxiety. Rigorous evidence of the effectiveness of these workshop-based interventions is lacking. However,

there is some documentation of positive outcomes in terms of reduction of social avoidance, anxiety, and distress among disfigured individuals at post-workshop follow-up (60).

The content of these social skills training workshops are potentially applicable to the appearance and body image concerns of people with SSc. The disfigurement experienced by many patients can be quite salient and is often found in visible and socially relevant body parts.

As a result, anxiety about the social reactions of others and avoidance of social interactions may be an important issue for many SSc patients. Given this, the content and strategies incorporated in the social skills training workshops will serve as the foundation for the development of a workshop-based intervention that we plan to adapt for easy delivery to SSc patients with significant body image concerns, via either telephone or internet. The method of delivery of these workshops will depend largely on the comfort levels of the patients, as video-based interactions may not be appealing to individuals with concerns about their physical appearance. The Brief-Satisfaction with Appearance Scale, which has been adapted and validated for SSc, (63) and which measures appraisal of appearance and comfort in social interactions, will be used as the primary outcome measure.

E. Internet-based self-management for scleroderma

Effective management of chronic illness places extensive day-to-day demands on patients and their caregivers. Patients must adhere to medication and other medical recommendations, make significant lifestyle and behavioural changes, and learn to cope with psychological and social issues inherent to living with a chronic illness (64, 65). As a result, and because of recognition of the benefits that are accrued when patients increase personal control in health matters, patient self-management programmes have been increasingly emphasised. The Chronic Disease Self-Management Programme (CDSMP), which was developed by Kate Lorig and modeled on her Arthritis Self-Management Programme, is designed to teach self-care techniques useful to persons with various chronic diseases (66). The CDSMP has been widely adapted, and an internet-based version of the programme improved self-efficacy and overall health status in a RCT that included patients with heart disease, lung disease, and type II diabetes (66). Although the program is designed for general use by patients with chronic diseases, in focus groups conducted by the Canadian Scleroderma Research Group, patients with SSc indicated that they would be more likely to find the programme beneficial if topics were geared towards managing the unique demands of living with SSc (*e.g.* skin care strategies, strategies to overcome eating difficulties and temperature changes) and if other participants in the workshops also had SSc. Preliminary work has been conducted to determine how best to adapt the CDSMP to meet the needs of patients with SSc, as well as how best to adapt the technology platform for a SSc-focused programme. Trials of CDSMP typically measure self-efficacy, as well as more standard outcomes (*e.g.* pain, fatigue) and the best outcomes to measure in a group of SSc patients with a diverse set of problems will be an important part of the project development stage.

Conclusions

Numerous sources of evidence highlight the negative effects of SSc on HRQoL; however, no rigorous trials have been conducted to examine treatment strategies and no recommendations have been made regarding intervention approaches. One of the primary reasons that SPIN has been created is to provide a network of experts to fill the gap in SSc psychosocial and allied health research and clinical practice, by developing and testing patient-centred interventions with the goal of broad dissemination of evidence-based interventions for use by a spectrum of people with the disease as well as providers that care for these patients. Ideally, in the long-term, interventions developed and tested by SPIN will

be incorporated into a menu of options to address the diverse problems experienced by people living with SSc, which can be made available through outlets, such as the websites of patient organisations across the globe.

Acknowledgments

Funding and competing interests The initial organisational meeting for SPIN was funded by a Canadian Institutes of Health Research (CIHR) Meetings, Planning, and Dissemination grant to B.D. Thombs (KPE-109130), Sclérodermie Québec, and the Lady Davis Institute for Medical Research of the Jewish General Hospital, Montréal, Québec. SPIN receives funding support from the Scleroderma Society of Ontario, the Scleroderma Society of Canada, and Sclérodermie Québec. B.D. Thombs and M. Hudson are supported by New Investigator awards from the CIHR, and Établissement de Jeunes Chercheurs awards from the Fonds de la Recherche en Santé Québec (FRSQ). M. Baron is the director of the Canadian Scleroderma Research Group, which receives grant funding from the CIHR, the Scleroderma Society of Canada and its provincial chapters, Scleroderma Society of Ontario, Sclérodermie Québec, and the Ontario Arthritis Society, and educational grants from Actelion Pharmaceuticals and Pfizer. M.D. Mayes and S. Assassi are supported by the NIH/NIAMS Scleroderma Center of Research Translation grant no. P50-AR054144. S.J. Motivala is supported by an NIH career development grant (K23 AG027860) and the UCLA Cousins Center for Psychoneuroimmunology. D. Khanna is supported by a NIH/ NIAMS K23 AR053858-04) and NIH/ NIAMS U01 AR057936A, the National Institutes of Health through the NIH Roadmap for Medical Research Grant (AR052177), and has served as a consultant or on speakers bureau for Actelion, BMS, Gilead, Pfizer, and United Therapeutics.

References

1. WASTFELT M, FADEEL B, HENTER JI. A journey of hope: lessons learned from studies on rare diseases and orphan drugs. J Intern Med. 2006; 260:1–10. [PubMed: 16789973]
2. REIMANN A, BEND J, DEMBSKI B. (Patient-centered care in rare diseases. A patient organisations' perspective). Bundesgesundheitsblatt Gesundheitsforschung Gesundheitsschutz. 2007; 50:1484–93. [PubMed: 18026887]
3. OLSON AR. The need to know. JAMA. 2007; 297:787–8. [PubMed: 17327512]
4. CANADIAN MEDICAL ASSOCIATION. Health care transformation in Canada. 2010.
5. LEWIS S. Patient-centered care: An introduction to what it is and how to achieve it. A discussion paper for the Saskatchewan Ministry of Health. 2009
6. GALLUCCIO F, WALKER UA, NIHTYANOVA S, et al. Registries in systemic sclerosis: a worldwide experience. Rheumatology. 2011; 50:60–8. [PubMed: 21148153]
7. SEIBOLD, J. Scleroderma. In: HARRIS, ED.; BUDD, RC.; FIRESTEIN, GS.; GENOVESE, MC.; SERGENT, JS.; RUDDY, S.; SLEDGE, CB., et al., editors. Kelley's Textbook of Rheumatology. Elsevier; Philadelphia: 2005. p. 1279-308.
8. WIGLEY, FM.; HUMMERS, LK. Clinical features of systemic sclerosis. In: HOCHBERG, MC.; SILMAN, AJ.; SMOLEN, JS.; WEINBLATT, ME.; WEISMANN, WH., editors. Rheumatology. Mosby; Philadelphia: 2003. p. 1463-80.
9. CHIFFLOT H, FAUTREL B, SORDET C, CHATELUS E, SIBILIA J. Incidence and prevalence of systemic sclerosis: a systematic literature review. Semin Arthritis Rheum. 2008; 37:223–35. [PubMed: 17692364]
10. BERNATSKY S, JOSEPH L, PINEAU CA, BELISLE P, HUDSON M, CLARKE AE. Scleroderma prevalence: demographic variations in a population-based sample. Arthritis Rheum. 2009; 61:400–4. [PubMed: 19248123]
11. DENTON CP, BLACK CM, ABRAHAM DJ. Mechanisms and consequences of fibrosis in systemic sclerosis. Nat Clin Pract Rheumatol. 2006; 2:134–44. [PubMed: 16932673]
12. MAYES MD, LACEY JV JR, BEEBE-DIMMER J, et al. Prevalence, incidence, survival, and disease characteristics of systemic sclerosis in a large US population. Arthritis Rheum. 2003; 48:2246–55. [PubMed: 12905479]
13. THOMBS BD, VAN LANKVELD W, BASSEL M, et al. Psychological health and well-being in systemic sclerosis: State of the science and consensus research agenda. Arthritis Care Res. 2010; 62:1181–9.

14. HAYTHORNTHWAITE JA, HEINBERG LJ, MCGUIRE L. Psychologic factors in scleroderma. Rheum Dis Clin North Am. 2003; 29:427–39. [PubMed: 12841303]
15. KOWAL-BIELECKA O, LANDEWÉ R, AVOUAC J, et al. EULAR recommendations for the treatment of systemic sclerosis: a report from the EULAR Scleroderma Trials and Research group (EUSTAR). Ann Rheum Dis. 2009; 68:620–8. [PubMed: 19147617]
16. BASSEL M, HUDSON M, TAILLEFER SS, SCHIEIR O, BARON M, THOMBS BD. Frequency and impact of symptoms experienced by patients with systemic sclerosis: results from a Canadian National Survey. Rheumatology. 2011; 50:762–7. [PubMed: 21149249]
17. JEWETT, LR.; HAYTHORNTHWAITE, JA.; THOMBS, BD. Psychosocial issues and care for patients with systemic sclerosis. In: VARGA, J.; DENTON, CP.; WIGLEY, FM., editors. Scleroderma: From pathogenesis to comprehensive management. Springer; In press
18. JEWETT, LR.; HUDSON, M.; THOMBS, BD. A 38-year-old woman with elevated muscle enzymes, Raynaud's phenomenon and positive anti-topoisomerase I antibody: Is she depressed?. In: SILVER, RM.; DENTON, CP., editors. Case Studies in Systemic Sclerosis. Springer; In press
19. POOLE JL. Musculoskeletal rehabilitation in the person with scleroderma. Curr Opin Rheumatol. 2010; 22:205–12. [PubMed: 20019617]
20. MANCUSO T, POOLE JL. The effect of paraffin and exercise on hand function in persons with scleroderma: a series of single case studies. J Hand Ther. 2009; 22:71–7. [PubMed: 18950987]
21. RANNOU F, POIRAUDEAU S, BEREZNE A, et al. Assessing disability and quality of life in systemic sclerosis: Construct validities of the Cochin Hand Function Scale, Health Assessment Questionnaire (HAQ), Systemic Sclerosis HAQ, and Medical Outcomes Study 36-Item Short Form Health Survey. Arthritis Rheum. 2007; 57:94–102. [PubMed: 17266096]
22. KALLEN MA, MAYES MD, KRISEMAN YL, De ACHAVAL SB, COX VL, SUAREZ-ALMAZOR ME. The Symptom Burden Index: Development and initial findings from use with patients with systemic sclerosis. J Rheumatol. 2010; 37:1692. [PubMed: 20516027]
23. SCHOUFFOER AA, NINABER MK, BEAARTVAN DE, VOORDE LJ, et al. Randomized comparison of a multidisciplinary team care program with usual care in patients with systemic sclerosis. Arthritis Care Res. 2011; 63:909–17.
24. BENRUD-LARSON LM, HAYTHORNTHWAITE JA, HEINBERG LJ, et al. The impact of pain and symptoms of depression in scleroderma. Pain. 2002; 95:267–75. [PubMed: 11839426]
25. DANIELI E, AIRO P, BETTONI L, et al. Health-related quality of life measured by the Short Form 36 (SF-36) in systemic sclerosis: correlations with indexes of disease activity and severity, disability, and depressive symptoms. Clin Rheumatol. 2005; 24:48–54. [PubMed: 15300468]
26. JOHNSON SR, GLAMAN DD, SCHENTAG CT, LEE P. Quality of life and functional status in systemic sclerosis compared to other rheumatic diseases. J Rheumatol. 2006; 33:1117–22. [PubMed: 16622903]
27. SCHIEIR O, THOMBS BD, HUDSON M, et al. Prevalence, severity, and clinical correlates of pain in patients with systemic sclerosis. Arthritis Care Res. 2010; 62:409–17.
28. HANSDOTTIR I, MALCARNE VL, FURST DE, WEISMAN MH, CLEMENTS PJ. Relationships of positive and negative affect to coping and functional outcomes in systemic sclerosis. Cognitive Ther Res. 2004; 28:593–610.
29. THOMBS BD, BASSEL M, McGUIRE L, SMITH MT, HUDSON M, HAYTHORNTHWAITE JA. A systematic comparison of fatigue levels in systemic sclerosis with general population, cancer and rheumatic disease samples. Rheumatology. 2008
30. Van LANKVELD WG, VONK MC, TEUNISSEN H, Van Den HOOGEN FH. Appearance self-esteem in systemic sclerosis--subjective experience of skin deformity and its relationship with physician-assessed skin involvement, disease status and psychological variables. Rheumatology. 2007; 46:872–6. [PubMed: 17308314]
31. HUDSON M, STEELE R, LU Y, THOMBS BD, CANADIAN SCLERODERMA RESEARCH GROUP, BARON M. Work disability in systemic sclerosis. J Rheumatol. 2009; 36:2481–6. [PubMed: 19797513]
32. SANDQVIST G, EKLUND M. Daily occupations--performance, satisfaction and time use, and relations with well-being in women with limited systemic sclerosis. Disabil Rehabil. 2008; 30:27–35. [PubMed: 17852220]

33. SANDQVIST G, SCHEJA A, EKLUND M. Working ability in relation to disease severity, everyday occupations and well-being in women with limited systemic sclerosis. Rheumatology. 2008; 47:1708–11. [PubMed: 18815157]
34. THOMBS BD, HUDSON M, BASSEL M, TAILLEFER SS, BARON M, CANADIAN SCLERODERMA RESEARCH GROUP. Sociodemo-graphic, disease, and symptom correlates of fatigue in systemic sclerosis: evidence from a sample of 659 Canadian Scleroderma Research Group Registry patients. Arthritis Rheum. 2009; 61:966–73. [PubMed: 19565539]
35. FRECH T, HAYS RD, MARANIAN P, CLEMENTS PJ, FURST DE, KHANNA D. Prevalence and correlates of sleep disturbance in systemic sclerosis - results from the UCLA scleroderma quality of life study. Rheumatology. 2011
36. MILETTE K, RAZYKOV I, POPE J, et al. Clinical correlates of sleep problems in systemic sclerosis: the prominent role of pain. Rheumatology. 2011; 50:921–5. [PubMed: 21169344]
37. BAUBET T, RANQUE B, TAIEB O, et al. Mood and anxiety disorders in systemic sclerosis patients. Presse Med. 2011; 40:e111–9. [PubMed: 21055901]
38. THOMBS BD, TAILLEFER SS, HUDSON M, BARON M. Depression in patients with systemic sclerosis: A systematic review of the evidence. Arthritis Rheum. 2007; 57:1089–97. [PubMed: 17665491]
39. MILETTE K, HUDSON M, BARON M, THOMBS BD, CANADIAN SCLERODERMA RESEARCH GROUP. Comparison of the PHQ-9 and CESD depression scales in systemic sclerosis: internal consistency reliability, convergent validity and clinical correlates. Rheumatology. 2010; 49:789–96. [PubMed: 20100794]
40. THOMBS BD, HUDSON M, TAILLEFER SS, BARON M, CANADIAN SCLERODERMA RESEARCH GROUP. Prevalence and clinical correlates of symptoms of depression in patients with systemic sclerosis. Arthritis Rheum. 2008; 59:504–9. [PubMed: 18383422]
41. ANGELOPOULOS NV, DROSOS AA, MOUTSOPOULOS HM. Psychiatric symptoms associated with scleroderma. Psychother Psychosom. 2001; 70:145–50. [PubMed: 11340416]
42. LEGENDRE C, ALLANORE Y, FERRAND I, KAHAN A. Evaluation of depression and anxiety in patients with systemic sclerosis. Joint Bone Spine. 2005; 72:408–11. [PubMed: 16214073]
43. MOZZETTA A, ANTINONE V, ALFANI S, et al. Mental health in patients with systemic sclerosis: a controlled investigation. J *Eur Acad Dermatol Venereol*. 2008; 22:336–40. [PubMed: 18269602]
44. BANCROFT, J. Human Sexuality and its Problems. 3rd ed.. Churchill Livingstone; Edinburgh, UK: 2009.
45. SCHOVER, LR.; JENSEN, SR. Sexuality and chronic illness: A comprehensive approach. Guilford; New York: 1988.
46. SAAD SC, PIETRZYKOWSKI JE, LEWIS SS, et al. Vaginal lubrication in women with scleroderma and Sjögren's syndrome. Sex Disabil. 1999; 17:103–13.
47. SAAD SC, BEHREND AE. Scleroderma and sexuality. J Sex Res. 1996; 33:15–20.
48. KNAFO R, THOMBS BD, JEWETT L, HUDSON M, WIGLEY F, HAYTHORNTHWAITE JA. (Not) talking about sex: a systematic comparison of sexual impairment in women with systemic sclerosis and other chronic disease samples. Rheumatology. 2009; 48:1300–3. [PubMed: 19692458]
49. SCHOUFFOER AA, Van Der MAREL J, Ter KUILE MM, et al. Impaired sexual function in women with systemic sclerosis: a cross-sectional study. Arthritis Rheum. 2009; 61:1601–8. [PubMed: 19877108]
50. MALCARNE VL, HANSDOTTIR I, GREEN-BERGS HL, CLEMENTS PJ, WEISMAN MH. Appearance self-esteem in systemic sclerosis. Cognitive Therapy and Research. 1999; 23:197–208.
51. BENRUD-LARSON LM, HEINBERG LJ, BOLING C, et al. Body image dissatisfaction among women with scleroderma: Extent and relationship to psychosocial function. Health Psychology. 2003; 22:130–9. [PubMed: 12683733]
52. RELTON C, TORGERSON D, O'CATHAIN A, NICHOLL J. Rethinking pragmatic randomised controlled trials: introducing the “cohort multiple randomised controlled trial” design. BMJ. 2010; 340:c1066. [PubMed: 20304934]

53. MERKEL PA, CLEMENTS PJ, REVEILLE JD, et al. Current status of outcome measure development for clinical trials in systemic sclerosis. Report from OMERACT 6. J Rheumatol. 2003; 30:1630–47. [PubMed: 12858472]
54. FURST D, KHANNA D, MATUCCI-CERINIC M, et al. Systemic sclerosis – continuing progress in developing clinical measures of response. J Rheumatol. 2007; 34:1194–200. [PubMed: 17477486]
55. TUGWELL P, BOMBARDIER C, BUCHANAN WW, GOLDSMITH CH, GRACE E, HANNA B. The MACTAR Patient Preference Disability Questionnaire – an individualized functional priority approach for assessing improvement in physical disability in clinical trials in rheumatoid arthritis. J Rheumatol. 1987; 14:446–51. [PubMed: 3305931]
56. MOUTHON L, RANNOU F, BEREZNE A, et al. Patient preference disability questionnaire in systemic sclerosis: a cross-sectional survey. Arthritis Rheum. 2008; 59:968–73. [PubMed: 18576298]
57. MEAD N, LESTER H, CHEW-GRAHAM C, GASK L, BOWER P. Effects of befriending on depressive symptoms and distress: systematic review and meta-analysis. Br J Psychiatry. 2010; 196:96–101. [PubMed: 20118451]
58. DENNIS CL, HODNETT E, KENTON L, et al. Effect of peer support on prevention of postnatal depression among high risk women: multisite randomised controlled trial. BMJ. 2009; 338:a3064. [PubMed: 19147637]
59. ROBINSON E, RUMSEY N, PARTRIDGE J. An evaluation of the impact of social interaction skills training for facially disfigured people. Br J Plast Surg. 1996; 49:281–9. [PubMed: 8774241]
60. KAPP-SIMON KA, SIMON DJ, KRISTOVICH S. Self-perception, social skills, adjustment, and inhibition in young adolescents with craniofacial anomalies. Cleft Palate Craniofac J. 1992; 29:352–6. [PubMed: 1643066]
61. NEWELL RJ. Altered body image: A fear-avoidance model of psycho-social difficulties following disfigurement. J Adv Nurs. 1999; 30:1230–8. [PubMed: 10564423]
62. JEWETT LR, HUDSON M, HAYTHORNTHWAITE JA, et al. Development and validation of the Brief-Satisfaction with Appearance Scale for systemic sclerosis. Arthritis Care Res. 2010; 62:1779–86.
63. NEWMAN S, STEED L, MULLIGAN K. Self-management interventions for chronic illness. Lancet. 2004; 364:1523–37. [PubMed: 15500899]
64. MENDELSON C, POOLE JL. Become your own advocate: advice from women living with scleroderma. Disabil Rehabil. 2007; 29:1492–501. [PubMed: 17852224]
65. LORIG KR, RITTER PL, LAURENT DD, PLANT K. Internet-based chronic disease self-management: a randomized trial. Med Care. 2006; 44:964–71. [PubMed: 17063127]