Patients' Perspectives and Experiences Living with Systemic Sclerosis: A Systematic Review and Thematic Synthesis of Qualitative Studies

Ayano Nakayama, David J. Tunnicliffe, Vivek Thakkar, Davinder Singh-Grewal, Sean O'Neill, Jonathan C. Craig, and Allison Tong

ABSTRACT. Objective. Systemic sclerosis (SSc) is a chronic, progressive autoimmune disease with major end-organ involvement. Much attention has been focused on the management of physical and clinical manifestations; however, the effect of the disease and treatment on the patient's identity, relationships, functioning, and mental well-being are less known. We aimed to describe the patients' perspectives and experiences of living with SSc.

Methods. Electronic databases were searched to October 2014. Thematic synthesis was used to analyze the findings.

Results. We included 26 studies involving 463 patients. Six key themes were identified: distressing appearance transformation (disturbing facial changes, stigmatizing sickness, unrecognizable self), palpable physical limitations (bodily restrictions, frustrating mind-body disconnect, pervasive fatigue, disabling pain), social impairment (breaking intimacy, struggling to fulfill family responsibilities, maintaining work, losing independence), navigating uncertainty (diagnostic ambiguity, medically fending for oneself, unpredictable course of illness), alone and misunderstood (fearful avoidance of fellow patients, invisible suffering), and gradual acceptance and relative optimism (adapting to change and accepting limitations, taking a positive spin, cautious hoping, empowering relationships, valuing medical support).

Conclusion. SSc is a rare and unpredictable illness that undermines patients' sense of certainty and control and impairs their self-image, identity, and daily functioning. Patient-centered care that encompasses strategies to promote self-esteem, resilience, and self-efficacy may help to improve treatment satisfaction and health and quality of life outcomes for patients with SSc. (First Release May 1 2016; J Rheumatol 2016;43:1363–75; doi:10.3899/jrheum.151309)

Key Indexing Terms:

SYSTEMIC SCLEROSIS	REVIEW	QUALITATIVE RESEARCH
QUALITY OF LIFE/PSYCHOLOGY	HEALTH BEHAVIOR	SOCIAL SUPPORT

Systemic sclerosis (SSc) is a connective tissue disorder characterized by progressive fibrosis and vasculopathy affecting the skin and internal organ systems. It mostly affects women and carries a mortality risk 4 times higher than age-matched individuals from the general population[1]. In the absence of a cure, patients face a lifetime of morbidity and treatment strategies that focus on limiting end-organ involvement[2]. As a result, the illness and treatment burden faced by patients can have detrimental consequences on their identity, relationships, functioning, and mental well-being.

Quantitative studies have shown that patients experience

From the Centre for Kidney Research, and Department of Rheumatology, The Children's Hospital at Westmead; Sydney School of Public Health, and Discipline of Paediatrics and Child Health, University of Sydney; School of Maternal and Child Health, University of New South Wales; Department of Rheumatology, Liverpool Hospital, Sydney; Department of Rheumatology, Canberra Hospital, Canberra; School of Medicine, University of Western Sydney, Campbelltown; South West Sydney Clinical School, University of New South Wales, Liverpool, Australia.

Supported by Scleroderma New South Wales, Australia. AT is supported by an Australian National Health and Medical Research Council Fellowship (ID 1037162).

A. Nakayama, MBBS, Centre for Kidney Research, The Children's Hospital at Westmead, and Department of Rheumatology, Canberra Hospital; D.J. Tunnicliffe, MIPH, Centre for Kidney Research, The Children's Hospital at Westmead, and Sydney School of Public Health, University of Sydney; V. Thakkar, MD, Department of Rheumatology, Liverpool Hospital, and School of Medicine, University of Western Sydney, and South West Sydney Clinical School, University of New South Wales; D. Singh-Grewal, PhD, Associate Professor, Department of Rheumatology, The Children's Hospital at Westmead, and Discipline of Paediatrics and Child Health, University of Sydney, and School of Maternal and Child Health, University of New South Wales, and Department of Rheumatology, Liverpool Hospital; S. O'Neill, PhD, Professor, Department of Rheumatology, Liverpool Hospital, and South West Sydney Clinical School, University of New South Wales; J.C. Craig, PhD, Associate Professor, Centre for Kidney Research, The Children's Hospital at Westmead, and Sydney School of Public Health, University of Sydney; A. Tong, PhD, Centre for Kidney Research, The Children's Hospital at Westmead, and Sydney School of Public Health, University of Sydney.

Address correspondence to Dr. A. Nakayama, Department of Rheumatology, Canberra Hospital, Yamba Drive, Garran ACT 2605, Australia. E-mail: Ayano.Nakayama@act.gov.au

Accepted for publication March 1, 2016.

high rates of depression, anxiety about disease progression, and low self-esteem[3,4]. Sexual impairment, difficulty with parenting, and impaired participation in life are also reported[5,6,7]. However, quantitative studies do not provide detailed explanations about patient attitudes and beliefs.

Qualitative research can offer rich narrative data to provide in-depth insight about patients' perspectives on living with SSc. Thematic synthesis of qualitative studies can generate a diversity of perspectives across different healthcare contexts, and involves high-level analytical abstraction of findings from individual studies. We aimed to describe the breadth of experiences and perspectives of patients to inform strategies to improve and direct care to patient-centered outcomes.

MATERIALS AND METHODS

We followed the Enhancing Transparency of Reporting the Synthesis of Qualitative research framework[8].

Selection criteria. Qualitative studies that reported the perspectives and experiences of adults (≥ 18 yrs of age) with SSc were eligible. This could include a patient's insight, attitudes, and beliefs about any topic relating to the illness and treatment of SSc. Qualitative studies that did not elicit perspectives or experiences from patients directly were excluded. Any type of qualitative studies were eligible (e.g., semi-structured interviews, focus groups, observations). Journal articles, conference abstracts, and Masters and PhD dissertations were included. Excluded were observational epidemiological studies, non-primary research articles (letters, commentaries, and reviews), and non-English articles.

Data sources and searches. We searched MEDLINE, Embase, PsycINFO, and CINAHL from database inception to October 13, 2014. We also searched Google Scholar, reference lists of relevant studies, British Library Electronic Digital Thesis Online Service, the Europe E-theses Portal, and ProQuest. We screened the abstracts and discarded those not meeting the inclusion criteria and then examined the full text of potentially relevant studies. One author (AN) screened abstracts, which was crosschecked by AT. There were no disagreements. The search strategy is provided in Appendix 1.

Comprehensiveness of reporting. We used a modified version of the consolidated criteria for reporting qualitative health research (COREQ) framework[9] to evaluate the completeness of reporting of each included study. Items specific to the research team, study methods, study setting, analysis, and interpretations were assessed. Two reviewers (AN and DT) independently assessed each study and resolved disagreements through a third reviewer (AT).

Synthesis of findings. We used thematic synthesis to analyze the data. We extracted all the participant quotations and text under the Results and/or Discussion section of each study and imported them into the HyperResearch software (ResearchWare Inc.; version 3.5.2). Preliminary concepts were inductively identified by AN without using a preexisting conceptual framework. The preliminary coding framework was discussed among authors AN, DT, VT, DSG, and AT. For each article, AN performed line-by-line coding into preliminary themes and subthemes with adjustments as new themes or subthemes emerged. AN, DT, VT, DSG, and AT identified conceptual links among themes to develop an analytical thematic schema. This research triangulation enhances the credibility of the findings and ensures that our analysis reflects the full breadth and depth of data.

RESULTS

Literature search and study descriptions. Twenty-six articles (12 journal articles, 7 abstracts, and 7 dissertations) involving 463 participants were included (Figure 1). The study characteristics are provided in Table 1[10–19,20–29,30,31,32,33,34,35]. There were 343 women (84%), and the age of participants ranged from 22 to 86 years. Subtypes of SSc were predominantly patient-reported and specified in 9 studies (diffuse cutaneous SSc, n = 77; limited cutaneous SSc, n = 139). The disease duration/time since diagnosis ranged from 0 to 48 years. Data were collected using interviews (14 studies), focus groups (10 studies), phone interviews (3 studies), and an open-ended questionnaire (1 study).

Comprehensiveness of reporting. The comprehensiveness of reporting was variable, with studies reporting 2 to 23 out of the 26 possible items included in the modified COREQ framework (Table 2[10–19,20,21,22,24,25,26,27,28,30,31,32,33,34,35]). There were 14 (54%) studies that stated the participant selection strategy and 23 (88%) that described the participant characteristics. A topic/question guide was provided in 20 studies (77%), and audio or visual recording was reported in 18 (69%). There were 17 studies (65%) that specified the use of researcher triangulation in data analysis, and 20 (77%) provided raw data including quotations.

Key synthesis. We identified 6 themes: distressing appearance transformation, palpable physical limitations, social impairment, navigating uncertainty, alone and misunderstood, and gradual acceptance and relative optimism. The themes are described in the following section, illustrative quotations are provided in Table 3[10–19,20,21,23,24,25,26,27,28,35], and conceptual links among themes are presented in Figure 2.

Distressing appearance transformation. Radical facial changes were the most devastating aspect of SSc for some patients. Patients felt they looked terrifying, unattractive, and undesirable, using terms such as "freak," "Dracula," "monster," or "ugly duckling" to describe themselves[28]. This caused some to become emotionally distressed to the point of contemplating suicide. Patients were painfully aware of their facial changes and desperately tried to hide their appearance by wearing their hair over their face and using sunglasses, hats, or makeup.

Telangiectasia gave the appearance of "chicken pox" and patients felt ostracized and often needed to explain to others that they were not contagious[12]. Patients felt particularly exposed to prejudice, which they attributed to their bodily deformities. For example, 1 patient was asked to sit out of a meeting so "nobody would have to look at [their] hands"[15].

Patients felt a deep sense of grief over their lost identity because they no longer recognized themselves as a result of their dramatically changed appearance. They were ashamed of the person they had transformed into, desperately wanting others to know "this isn't me"[28]. Patients were reminded of their mortifying transformation when old friends had "no clue" of who they were or mistook old photos to be someone else.

Palpable physical limitations. For some, SSc was physically limiting as their skin hardened to become "like a

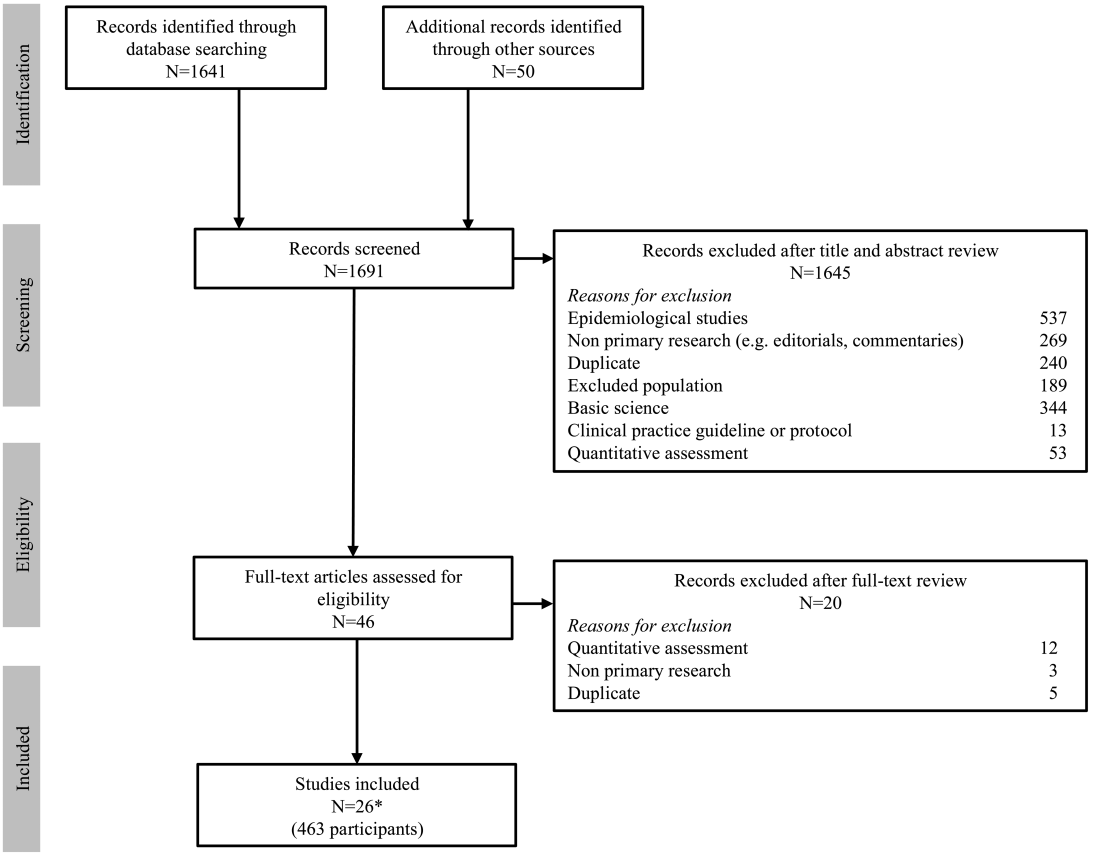

Figure 1. Search results.

mannequin"[15]. Patients felt they had to abandon previous leisure and work activities, and blamed themselves for "ruining everybody's time"[21].

Some patients articulated an exasperating mismatch between their mental capacity and physical capability. Although patients continued to be mentally astute, they felt forced to pursue less physically demanding jobs[16].

Patients found their lack of energy "overwhelming"[13]. It impeded their ability to work, participate in leisure activities, and look after themselves and their family. Basic self-management tasks such as applying ointment several times a day became "too onerous"[14]. Weekends were spent resting, and hobbies were replaced by "lying in bed"[13].

Raynaud phenomenon, calcinosis, and digital ulcers were described as being intensely painful and debilitating by some patients. This was emotionally distressing and also limited patients' ability to work, go outdoors, or even walk.

Social impairment. Women felt that they were undesirable, unattractive, and had "less to offer"[28]. Those who were single were afraid of remaining unwanted. Some had the painful experience of being told they were "no longer sexually attractive" by their husbands, or treated as if it was an embarrassment to be seen in public together. Lower libido, decreased secretions, and physical pain were barriers to sexual activity and made some women feel guilty.

Patients struggled to fulfill roles as parents, grandparents, and children. In particular, women felt inadequate because they could not care for their children or perform household duties and men regretted being unable to be physically active with their children. One patient felt guilty about being unable to reciprocate the help provided by parents.

Diminishing work capacity made patients feel pressured and disappointed with themselves. They reduced their work hours because of exhaustion, or sought help with fine motor tasks such as stapling, typing, and filing because of hand deformities. Some experienced discrimination by being unfairly demoted or treated as if they were "disabled"[24]. Others were well supported and were given flexible working hours, permission to work from home, or had modified work environments such as the use of personal heaters and changes in door handles.

Patients were annoyed and frustrated that "everything takes longer to do"[13], and some struggled to complete simple tasks such as drinking water or opening doors. Patients loathed the idea of becoming dependent on others and were

The Journal of Rheumatology 2016; 43:7; doi:10.3899/jrheum.151309

Table 1. Characteristics of the included studies.

Study ID	Country	n	Age, Yrs	Sex, M:F	Disease Duration/ Time since Diagnosis, Yrs	Scleroderma Type			Data Collection	Conceptual Methodological Framework	Analysis	Main Topic
						dcSSc	lcSSc	NS				
Peer-reviewed journal articles												
Brown, *et al*[10]	UK	5	40–75	0:5	NS	NS	NS	5	Face-to-face, semi-structured interviews	Qualitative study	Content and thematic analysis interviews	Group education experiences
Cinar, *et al*[11]	Turkey	16	Mean 48 (SD 12)	0:16	Mean 13 (SD 8)	NS	NS	16	Face-to-face, semi-structured interviews	Phenomenology	Colaizzi's phenomenological data analysis	Illness experiences
Ennis, *et al*[12]	UK	113	Median 63 (IQR 14)	17:96	Median 12 (IQR 12)	NS	81	32	Open-ended questionnaire	Qualitative study	Thematic analysis	Telangiectasias and body image dissatisfaction
Joachim and Acorn[13]	Canada	13	46–79	0:13	2–17	NS	NS	13	Focus groups	Qualitative study	Thematic analysis	Illness experiences
Kocher, *et al*[14]	Germany, Switzerland	8	44–77	3:5	0–11	4	3	1	Face-to-face interviews	Qualitative study	Content and thematic analysis	Skin and mucosa care, general illness experiences
Mendelson and Poole[15]	USA	11	43–84	0:11	Mean 16 (SD 4)	7	1	3	Focus groups	Qualitative study	Immersion and crystallization method of thematic analysis	Illness experience, self-management
Mendelson, *et al*[16]	USA	32	Mean 47 (SD 8)	6:26	Mean 10 (SD 9)	18	NS	14	Structured phone interviews	Qualitative study	Thematic analysis	Work experiences
Oksel and Gündüzoğlu[17]	Turkey	20	Mean 51 (SD 10)	0:20	Mean 9 (SD 8)	NS	NS	20	Face-to-face, semi-structured interviews	Phenomenology	Colaizzi's phenomenological data analysis	Illness experiences
Sandqvist, *et al*[18]	Sweden	17	38–62	1:16	2–22	2	15		Focus groups	Qualitative study	Thematic analysis	Work experiences
Stamm, *et al*[19†]	Austria, Romania, Sweden, Switzerland	63	30–85	10:53	0.5–48	25	38		Focus groups	Qualitative study	"Meaning condensation" thematic analysis	Illness experiences
Stamm, *et al*[20†]	Romania, Norway, Netherlands, Germany, Sweden, Austria, UK, Switzerland	63	30–85	10:53	0.5–48	25	38		Focus groups	Qualitative study	Content analysis	Functioning in daily life
Suarez-Almazor, *et al*[21]	USA	19	Median 49	6:13	Mean 8	18	1		Face-to-face, semi-structured individual interviews and focus groups	Grounded theory	Thematic analysis	Illness experiences
PhD and Masters dissertations												
Ananian[22]	USA	NS	NS	NS	NS	NS	NS	NS	Focus groups	Grounded theory	Social ecological model	Barriers to exercise/ experiences of management of arthritis
Lees[23]	UK	5	35–86	0:5	Mean 14	NS	NS	5	Face-to-face, open-ended interviews	Qualitative study	Thematic analysis	Arthritis self-management program experiences

Table 1. Continued.

Study ID	Country	n	Age, Yrs	Sex, M:F	Disease Duration/ Time since Diagnosis, Yrs	Scleroderma Type			Data Collection	Conceptual Methodological Framework	Analysis	Main Topic
						dcSSc	lcSSc	NS				
Johnson[24]	UK	6	40s, 50s, 60s	1:5	NS	NS	NS	5	Face-to-face, semi-structured interviews	Biographical methods**	Thematic analysis	Work experiences
Mills[25]	Canada	9	NS	NS	NS	NS	NS	9	Face-to-face, semi-structured interviews	Grounded theory	Thematic analysis	Living well with chronic disease
Newton[26]*	Canada	16	28–79	0:16	NS	NS	NS	16	Face-to-face, semi-structured interviews	Qualitative study	Thematic analysis	Emotional distress
Thorne[27]	USA	4	NS	NS	NS	NS	NS	4	Face-to-face, semi-structured interviews	Naturalistic inquiry, phenomenology, grounded theory	Phenomenological and grounded theory analysis	Relationships with health professionals
Wood[28]	USA	12	34–62	0:12	5–43	3	NS	9	Face-to-face, semi-structured interviews	Grounded theory	Thematic analysis	Facial changes
Conference abstracts												
Decuman, *et al*[29]	Belgium	14	NS	NS	NS	NS	NS	14	Semi-structured interviews	Qualitative study	NS	Work experiences
Mendelson, *et al*[30]	USA	25	Mean 54	2:23	Mean 12	NS	NS	25	Semi-structured phone interviews	Qualitative study	Thematic synthesis	Leisure activities experiences
Mittoo 1, *et al*[31]‡	USA, Canada	NS	NS	NS	NS	NS	NS	NS	Focus groups	Qualitative study	Thematic analysis	Interstitial lung disease experiences
Mittoo 2, *et al*[32]‡	Canada	5	NS	NS	NS	NS	NS	5	Focus groups	Qualitative study	Thematic analysis	Interstitial lung disease experiences
Mouthon, *et al*[33]	France	25	NS	NS	NS	NS	NS	25	Semi-structured interviews, life story interviews, focus groups, patient observations	Qualitative study	NS	Illness and disease management experiences
Poole, *et al*[34]	USA	10	Mean 45	10:0	4	NS	NS	NS	Phone interviews	Qualitative study	Content analysis	Experience of being a father
Stamm, *et al*[35]	Austria	15	Mean 46	7:8	NS	NS	NS	15	Biographic narrative interviews	Qualitative study	Narrative biographic method	Illness experience

* Same study published as a journal article in 2012. † Same scleroderma subjects in both journal articles. ‡ Mittoo 2, *et al* may be a study that forms a part of Mittoo 1, *et al*. ** Biographical methods use life histories as a method of social research. dcSSc: diffuse cutaneous systemic sclerosis; lcSSc: limited cutaneous systemic sclerosis; NS: not stated; IQR: interquartile range.

Table 2. Comprehensiveness of reporting in included studies (consolidated criteria for reporting qualitative health research).

Items	Studies Reporting Each Item	No. Studies
Personal characteristics		
Interviewer/facilitator identified	10, 14, 15, 18, 24–28	9
Occupation of the interview facilitator	10, 24, 25, 27	4
Experience or training in qualitative research	20, 22, 27, 35	4
Relationship with participants		
Relationship established prior to study commencement	14	1
Participant selection		
Selection strategy, e.g., snowball, purposive, convenience, comprehensive	10, 13–16, 20–28	14
Method of approach or recruitment	12–16, 19, 21–28	14
Sample size	10–30, 33–35	24
Number and/or reasons for nonparticipation	12, 13, 18, 21, 24, 26, 28	7
Setting		
Venue of data collection	10–16, 22, 24–28, 30	14
Presence of nonparticipants, e.g., clinical staff	18, 19	2
Description of the sample	10–28, 30, 32, 34, 35	23
Data collection		
Questions, prompts, or topic guide	10–22, 24–26, 28, 31, 32, 34	20
Repeat interviews/observations	13, 35	2
Audio/visual recording	10, 11, 13–16, 18, 19, 21–28, 34, 35	18
Field notes	10, 11, 13, 15, 19, 22, 24, 25, 28, 35	10
Duration of data collection	10, 13-15, 21–26, 28	11
Translation and interpretation, NA if English-speaking	11, 14, 19, 20	4
Protocol for data preparation and transcription	10, 13, 15–22, 24–28, 34, 35	17
Data, or theoretical, saturation	19, 21, 22, 24	4
Data analysis		
Researcher/expert triangulation, multiple researchers involved in coding and analysis	10–14, 18–22, 24, 25, 27, 30–32, 35	17
Translation, specifies language in which analysis was done, NA if English	14, 19	2
Derivation of themes or findings, e.g., inductive, constant comparison	10–12, 14–28, 30–32, 34, 35	23
Use of software, e.g., NVivo	15, 16, 21, 22, 24, 25, 28	7
Participant feedback on findings	11, 17, 24, 27, 28, 31	6
Reporting		
Participant quotations or raw data provided, e.g., picture, diary entries	10–28, 35	20
Range and depth of insight into participant perspectives of SSc, thick description provided	11–16, 18, 21, 24–28	13

NA: not applicable; SSc: systemic sclerosis.

determined to remain independent. Some women felt frustrated by their husbands who would "smother" and "wait on (them)"[15]. In contrast, others were saddened by the lack of support they received.

Navigating uncertainty. Some patients were frightened and exasperated when their symptoms progressed and remained unexplained despite seeking medical support. Patients had their symptoms dismissed or were misdiagnosed as having depression, cancer, or other illnesses. When a diagnosis was finally made, this brought great relief and a sense of closure to some. One patient stated, "I was so thrilled… It didn't matter that it is a potentially fatal disease"[27].

Some patients felt confused and misinformed by poor communication about their illness. Patients "felt obliged to do their own research"[14], but became disturbed by images of facial changes or statistics on reduced life expectancy. They mistrusted some health professionals who were perceived as being uneducated and suspected that some medications were unnecessary. Another problem was that the timing and the characteristics of disease manifestations were unpredictable. Symptoms could "jump from place to place"[13]. Patients could feel well and then "something else pops up"[13]. Patients were terrified of developing breathing difficulties, facial or hand changes, and the possibility of impending death. Some felt they had no control over what could happen, and were unable to understand why symptoms worsened.

Alone and misunderstood. Although support groups offered a rare opportunity to meet other patients, some found the experience horrifying. Seeing other patients with physical deformities, in wheelchairs, or taking supplemental oxygen was a frightening glimpse into their potential future. Accordingly, 1 patient remarked that it was common for patients to "avoid interaction with one another"[11]. Some patients were concerned about scaring others who were less affected. In contrast, others found support groups a valuable way to share their experiences and felt hope when they met others who were living well with SSc.

Patients without visible features of SSc were angered by family members who told them they "looked good" or were "doing fine"[23]. Although they had "pain, fatigue, and constant

Table 3. Illustrative quotations from study participants.

Themes	Quotations	Contributing Studies
Distressing appearance transformation		
Disturbing facial changes	Ninety-nine percent (of my self-esteem) has been affected because of my face. I mean, if my face wasn't disfigured like this, I call it disfigured, okay, or changed, and if I just had the scleroderma that didn't do anything to your face, yes, I could handle that. It wouldn't be as dramatic or drastic. It's been such a bad change. But like I said, yes, this scleroderma to my face has just really, really pulled me down. I mean, just no self-esteem[28]. Because I think it is scarier; maybe because there is so much wrapped up in your face; it reflects your emotions, it reflects your feelings, it reflects your attractiveness. People are going to perceive you as stupid or intelligent... The face is much more significant in some way[28].	11–14, 17, 21, 25, 26, 28
Stigmatizing sickness	A number of times people thought I had measles or chicken pox. I try to explain it's not contagious[12]. I was teaching in a men's school, and we had a large meeting, and the principal asked me to sit in the hallway during this meeting, that way, nobody would have to look at my hands. I said, "So do you even want me coming to the meeting?" "Well you have to talk." "But you don't want me in the room?" "Right."[15]	11–15, 20, 21, 25, 28
Unrecognizable self	Another participant described her worries saying, "Dying doesn't worry me, but dying totally ugly or handicapped when I don't recognize myself does." She added, "I see differences in my facial structure already. I'd rather die before I don't recognize myself anymore."[13] It bothers me because people who see me for the first time, I want to say, "This isn't me. Let me show you what I really look like." And the first thing I want to do is show them pictures [of myself before the facial changes]. "That's what I look like." Rather than saying, "Yeah, what you see, this is it."[28]	12, 13, 15, 17, 20, 21, 25, 28
Palpable physical limitations		
Bodily restrictions	In describing the onset of scleroderma, one woman said, "It started with shortness of breath." Upon bronchoscopy, she found she had only 25% lung capacity. She now has difficulty breathing almost all the time and is waiting for a lung transplant. Prior to having these symptoms, she described herself as leading an active life and spending much of her time outside working in her garden[13]. I don't feel free, which naturally restricts my movements and breathing[17].	10, 11, 13–17, 19–21, 23, 25, 26, 28
Frustrating mind-body disconnect	One participant summed up the challenge of coping with this disease when she said, "My biggest struggle has been that my mind has not figured out yet that my body doesn't keep up any more. My mind thinks it should still be able to do what we used to do and you can't".[15]	15, 16
Pervasive fatigue	My leisure activity is lying in bed. I feel that I never get enough sleep[13]. You don't feel like going out. You don't feel like doing anything. I've become a couch potato[21].	10, 13, 14, 17, 18, 21, 23, 25, 26, 35
Disabling pain	'Cause I'm scared something will touch it...; With the gangrene...the pain that you felt in your fingers as they were dying was so excruciating that you almost begged to say please cut it off[21]. The calcinosis hurts...in my toes, I can't walk, I can't wear sandals[21].	13, 14, 21, 23, 25, 26, 28
Social impairment		
Breaking intimacy	My husband deserves more. And I can't fake it and I'm too dry to do anything about it[21]. I gave my [husband] a kiss. And he's like, "No, give me a good kiss." It was like, well, this is as good as it gets! (Laugh) I can't pucker my lips. So just in that instant, he really, really hurt my feelings[28]. Eventually [my husband] left me. And it was devastating. Really devastating. Now, I didn't want to be single. Because I had all these problems [including facial changes] and who was going to want me[28].	12, 17, 19, 21, 28
Struggling to fulfill family responsibilities	I am ill. Why me? I could not stand up, walk, care for my children, cannot comb my daughter's hair, cooking is completely difficult for me. But I should do all these[11]. My parents have always helped me; I cannot help them with anything[18].	11, 13–18, 20, 21, 26
Maintaining work	Oh, it's totally subsumed [home life]. I mean, that is a challenge because I got home at 6:00 tonight. Normally, this is about when I get home, and I can wash my face and go to bed right now. I mean, I could totally check out. I think my life is very skewed toward work because I'm required to be there, and home can suffer. But I try to work through it[16]. People like myself could cope with going back to work if you've got an employer that understands[24].	16–18, 21, 24–26, 28
Losing independence	It is hard for me to open the tap. I told my husband several times that I could not open it since it was difficult for me. He ignored my words. So I have not said anything about it. He has not helped me[11]. I mean I hate to think of somebody having to dress me, or feed me. I mean I think that if and when that time comes, I think I will be probably the most depressed person on the face of the earth[17].	11, 13–15, 17, 18, 21, 23, 25, 26
Navigating uncertainty		
Diagnostic ambiguity	After being sick, or knowing there was something wrong, for nearly 17 years, I got a diagnosis of progressive systemic sclerosis, which I had never heard of. But I was so excited, I was so thrilled for this man to be telling me I had this disease! It was stupid, you know — to have an answer. It didn't matter that it is a potentially fatal disease. None of that stuff mattered[27]. At first, I was scared out of my wits. I didn't know what to do about it. Why was it all of a sudden spreading? What was going on? Is it cancer? I was told I had cancer, actually. And thank God, I didn't. But scared, absolutely terrified, absolutely terrified. What was going on? ...And the [doctors] keep telling you, "There's nothing wrong. It's all in your mind." That just made it worse. You think, "Gee, I must be nuts. I must be out of my mind."[28]	13–15, 25–28

Table 3. Continued.

Themes	Quotations	Contributing Studies
Medically fending for oneself	However, even after receiving a name for these "mysterious" symptoms, some of them still felt uninformed. They stated that they did not understand the medical terms used by physicians and had to enquire several times. Some participants felt obliged to do their own research on the Internet. In many cases they had received disturbing information, as in the case of one woman who read of a life expectancy of five years[14].	10, 11, 13–15, 19, 21, 25, 27, 28
Unpredictable course of illness	You get one thing under control and then something else pops up[13].	11, 13, 15, 17, 20, 21, 26, 28
	It was very hard, you don't know what's going on and the symptoms are developing, you know, you're not stabilized. So you know, it starts with the Raynaud, then the acid reflux, then the skin, then it spreads. Then you get darker, then you know you're getting them all. So you're waiting for that day when you can't breathe anymore or — but it hasn't happened so that's good[26].	
	It's always lurking in the corner. What's going to come next?[21]	
Alone and misunderstood		
Fearful avoidance of fellow patients	When I met with other patients and listened to them, I feel myself bad. They give me negative feelings. Some of them asked me to meet. I did not accept their proposal. I told them that you make me depressed and I could not cope with it. I do not want to meet with them so often[11].	10, 11, 13, 15, 17, 21, 25, 26, 28
	When I was attending the support group meetings, there was a lot of patients that their symptoms were pretty far advanced, and then every once in a while, we'd go to a meeting and find out that nice lady down the road had already passed on. It was getting to where I almost hated to go because I hated to see people that I was getting to know getting worse, even though the support group was good cause everyone shared experiences, but on the other hand, it was frightening[15].	
Invisible suffering	...scleroderma and Raynaud disease is something that nobody can see unless you are all crippled up. But with me, having Raynaud and scleroderma, people cannot see it. People say that you are looking good. Like with your family, if your family was interested in it or was into it by going to some of the meeting, then they would realize that you do have something bad. And it would help them [family] a lot to understand what is going on with me. ...you can talk to them and they think that you are doing fine when you are in fact not...People think, oh well, but they really don't have a clue about what it is all about. Sure it is good to do things at this age, but it is really hard to do things in so much pain?[23]	23, 26–28
Gradual acceptance and relative optimism		
Adapting to change and accepting limitations	For instance, if it is hard for me to wash dishes, I find a proper sponge for my hand. There are many small or big sponges that I can hold. I could not prefer those with two levels. Because they are so thick and I could not wash the glasses with them. Firstly, I had difficulty in doing it but later I overcame this diffculty. Now I know that I should wash the dishes and I find the easiest way to do this. I always ask myself "how can I do this?", "how can I cope with it?"[11]	10–16, 18, 21, 23, 25, 26, 28
	After a family dinner on Sunday, at 5 o'clock, I am so tired. If I sit for 20 minutes, I'll regroup. After having scleroderma for some time, I know how to handle this[13].	
Taking a positive spin	I can't use my hands the way I used to...I can tell my lungs have gotten weaker...[compared] to the next person...I feel healthy...I'm the lucky one[21].	12, 13, 15, 21, 23, 28
	Having this, being so ill and near death made me a better person. I appreciate life. It's precious. I work harder. I achieve more[13].	
Cautious hoping	I had no idea what SSc was. I had no idea if I was going to die, be in a wheelchair, I had no idea… I saw a lady, I was 31 at the time, but there was a girl, probably in her 20s, that was in a wheelchair, and it looked like she was on her deathbed. She was so thin; she had oxygen, a wheelchair, and I thought, oh my gosh, that's going to happen to me. But I kept on going to these support group meetings, and at one of the meetings, I met a lady who was probably about my age now and said she had had SSc for 25 years, and she was full of life and just going on, and I thought okay, I'm going to be like her[15].	13, 15, 16, 21, 25, 26, 28
	I have faith in God…[who] has all power and if God wants this thing cured. He usually can do it like that[21].	
Empowering relationships	My husband tells me after 30 years of marriage, we are the same people inside, we just have different wrappers[12].	10–16, 18, 21, 23, 25–28
	[My children] know that there's a change in the way I look. But to them, it's not a big deal because I'm still who I am on the inside[28].	
Valuing medical support	He was very concerned. He took my hand and told me what happens, and he was sorry that there's nothing that he can do. He was very concerned. He had very good bedside manners. He was very good about it. Really explained everything, and he was very good. He was very comforting, really. He's a very good doctor[28].	15, 21, 27, 28

SSc: systemic sclerosis.

worry"[26], if they brought these up, they felt they were being perceived as "complainers"[23].

Gradual acceptance and relative optimism. Over time, some patients became confident and determined that they knew "how to handle"[13] their disease. For example, patients were able to apply their own eyeliner, wash dishes by using a smaller sponge, or open cans by using a towel or knife. They created their "own therapy mix" for finger ulcers and

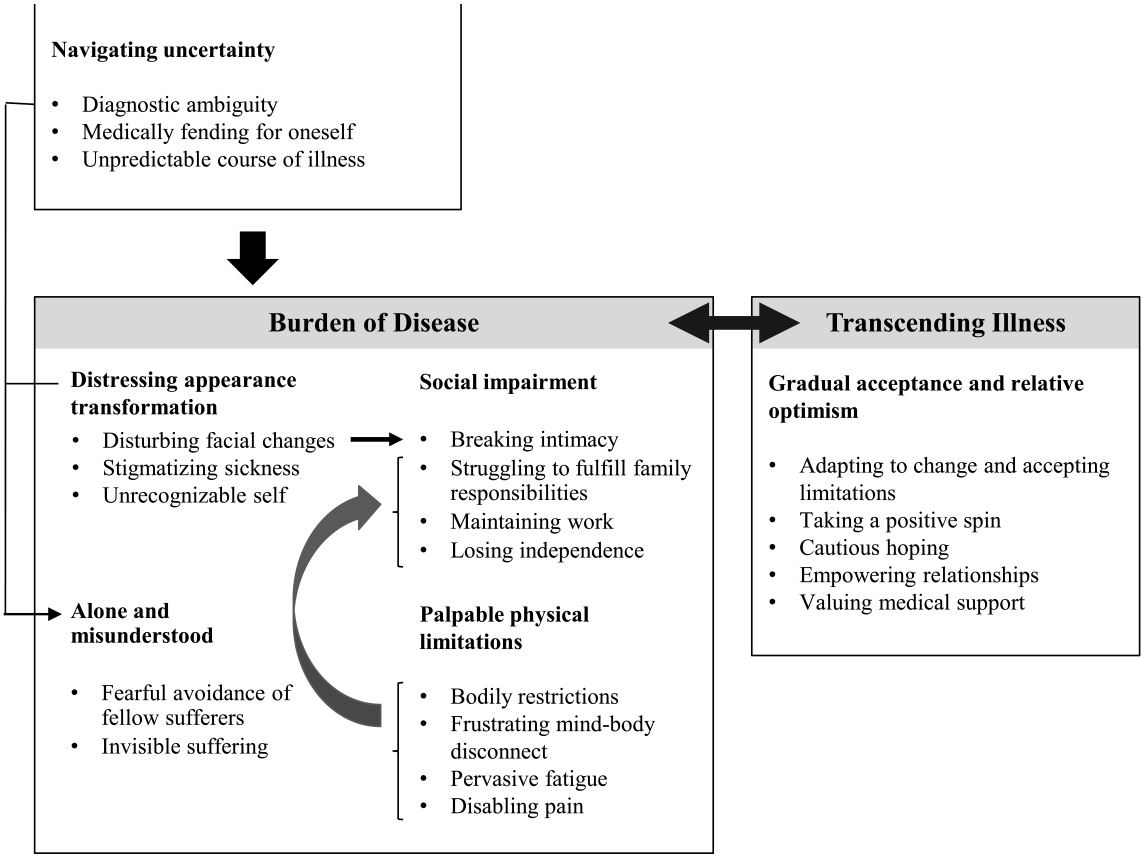

Figure 2. Thematic schema representing experiences and perspectives of adults living with scleroderma.

pain[14]. Patients accepted new restrictions such as not being able to ski because of Raynaud phenomenon or having to take breaks or accept help to cope with home and work life.

Some patients were grateful for the experience of having SSc because it helped them appreciate life, work harder, and achieve more. One patient described the illness as a "path of personal growth and spiritual transformation"[28]. Some attributed this attitude to being an older person or having a positive character. Other patients, including those with life-threatening disease, transitioned from devastation and despair to relative optimism and peace.

Patients held faith in their religion or advances in medicine to discover a cure for SSc. Some patients were hopeful that they would remain stable in their disease course and not become affected like others with advanced SSc.

Being treated in the same way by family and friends despite physical changes of SSc was particularly important for patients. One patient was touched by her husband who said, "After 30 years of marriage, we are the same people inside, we just have different wrappers"[12]. Patients found enjoyment in their relationships with family and friends in the place of physical leisure activities. Women appreciated husbands who would help them with household chores and looking after their children.

Patients valued honest and empathetic communication by physicians, which helped to counter the devastation of being told of frightening disease manifestations. Another source of great relief was to be diagnosed early in the course of the illness. In contrast to the frustration felt by patients when encountering doctors who were uneducated about SSc, 1 patient was grateful and impressed by the enthusiasm of a health professional to learn more about SSc and to teach others.

DISCUSSION

Patients with SSc contend with an uncertain prognosis and feel isolated by the rarity of the disease. Fatigue, pain, and hand deformities created frustration and guilt because patients were unable to fulfill their previous work and family roles. Also, the visible physical changes affecting their face and hands marred their self-perception such that they described their appearance as unsightly and repulsive. For this reason, some withdrew within themselves, which consequently placed strain on relationships and intimacy with their partner. Some learn to cope and adapt and felt their experience of being severely ill or having facial deformities led to personal enlightenment and emotional growth. While some patients were well supported in their workplace, others felt aggrieved by perceived discrimination.

There were some apparent differences across populations

using qualitative comparisons. Older patients with established disease appeared to cope better with physical deformities because over time they had learned to adapt and accept their limitations. Younger patients at earlier stages of their disease were more emotionally affected by facial and hand deformities and found it difficult to cope with work and looking after families.

Some of the findings in our study are similar to the experiences of patients with other chronic conditions. The uncertain course of the illness, fatigue, pain, role reversals, and dependence on others have been identified in patients with systemic lupus erythematosus and rheumatoid arthritis (RA)[36,37]. Patients with psoriasis also experience issues with self-esteem, stigmatization, and embarrassment[38]. The effect of facial changes was particularly striking in our review and highlights the importance of this aspect of the disease in patients with SSc. Some avoid meeting others with the same condition because the physical changes in others are so painful to observe.

Our study provides a range of insights about the concerns, attitudes, and experiences of patients with SSc. We coded the data using software to facilitate a systematic and auditable process. Researcher triangulation was conducted to ensure that all data were reflected in the analysis, and independent assessment of transparency of reporting was performed. However, our study has some potential limitations. Studies that did not report participant quotations contributed to a lesser extent to the findings. The majority of studies were conducted in English-speaking, high-income countries, and although many of these countries are multicultural, the transferability of our findings to other cultures is uncertain. We were unable to describe the differences between sexes because the majority of data from the included studies were from women. The differences between types of SSc (limited or diffuse) and specific organ involvement could also not be included because this was largely unreported.

Patients with SSc appreciate doctors who communicate empathetically and openly to them about their disease. Comprehensive education about the diagnosis, disease manifestations, treatment options, and likely prognosis balanced with empathy, support, and fostering hope for better outcomes and advances in medicine can help to mitigate fear and confusion. Online and written patient educational material from patient support organizations such as the Scleroderma Australia and New South Wales (Australia), Scleroderma Foundation (United States), Scleroderma Society of Canada, and the Federation of European Scleroderma Associations (Europe) can be useful adjuncts in patient education. A study of 49 patients with SSc found that a mail-delivered, self-management program including a workbook and exercise DVD was effective in decreasing depression, fatigue, and pain, and improved self-efficacy in managing pain[39]. This program included information on medical aspects of the disease, dysphagia, fatigue management, advocacy, activities of daily living, oral hygiene, skin and wound care, psychosocial changes, and exercises. Another study evaluated online information and support in a survey of 429 patients with SSc[40]. The study found that many patients with SSc use online resources and concluded that they need more information on physical, psychological, and social consequences of their disease. An interactive health communication application that includes e-consults and home access to electronic medical records and provides information about disease and treatments, and online peer-support forums, were perceived to be potentially useful by these patients.

The psychosocial effect of SSc is complex and requires a multidisciplinary approach. Women may feel unattractive and lose self-esteem, isolating themselves and withdrawing from society. Social workers and psychologists as well as patient-support forums may help provide counseling for patients. Use of cosmetics and skin care, advice from other patients, online resources, dermatologists, or beauticians may help improve appearance and self-esteem. Cognitive behavioral therapy can improve stress, depression, and disease activity in patients with RA[41]. In patients with body dysmorphic disorder, cognitive behavioral therapy has been used to successfully modify intrusive thoughts of body dissatisfaction, overvalued beliefs about physical appearance, exposure to avoided body image situations, and elimination of body checking[42].

Patients can struggle to maintain work and experience discrimination and lack of support in the workplace. Research has highlighted the importance of multidisciplinary care in improving patient outcomes in rheumatic diseases. Occupational therapy interventions in patients with RA and osteoarthritis (OA) have shown improved functional and work outcomes including work satisfaction and work performance, and decreased pain, helplessness, and disease activity[43,44].

Conducting adequately powered trials of interventions to improve health-related quality of life is difficult in patients with rare diseases such as SSc. The Scleroderma Patient-centered Intervention Network is an international collaboration of patient organizations, clinicians, and researchers that aims to develop a research infrastructure to test accessible, low-cost, self-guided online interventions to reduce disability and improve quality of life for people with SSc[45]. Our review suggests that interventions focusing on changes in appearance and improving self-esteem, practical tips on how to cope with disability, fatigue, and pain, improving workplace support and understanding, and promoting self-efficacy and resilience are needed. A review of 34 controlled psycho-educational trials in arthritis reported that although comparisons with the varied interventions are difficult, overall there is improved pain, depressive symptoms, self-efficacy, coping abilities, and self-management behaviors with these interventions. Most patients in these trials had either RA or OA and evaluated either cognitive-behavioral therapy or self-management pro-

grams[46]. Trials aimed at improving quality of life outcomes in patients with SSc are lacking.

The Health Assessment Questionnaire-Disability Index (HAQ-DI) and the Medical Outcomes Study Short Form-36 (SF-36) are validated outcome measures of function and quality of life in SSc that have met the Outcome Measures in Rheumatology Clinical Trials filter[47]. The Scleroderma HAQ is a modified version of HAQ-DI with additional SSc-specific items[48]. All of these measures, however, do not assess the effect of changes in physical appearance or self-esteem, which we have found are important to patients in our study. Other quality-of-life tools addressing this, such as satisfaction with appearance scales[49], have been used in patients with SSc and could be incorporated into existing tools. The European League Against Rheumatism Scleroderma Trials and Research group initiative is currently developing an International Classification of Functioning (ICF) core set for SSc[50,51]. Our systematic review could help guide initial data collection for this ICF core set. For example, the ICF is used to quantify the effect and burden on functioning of health conditions, but does not identify all the domains (e.g., personal identity).

Our systematic review has identified several knowledge gaps that require further study. This includes patients' perspectives on medication taking and fears of genetic transmission onto future children. Understanding of how patients learn to cope well with their illness including practical tips for other patients would be useful for designing future educational programs and interventions. Finally, the male SSc experience and the differences between subtypes of SSc are other undetermined areas for further research.

SSc is a rare and unpredictable illness that can be perceived as disfiguring. It undermines patients' sense of certainty and control, and impairs their self-image and daily functioning. A multidisciplinary approach to patient-centered care that encompasses strategies to promote self-esteem, self-efficacy, and open communication may help to improve treatment satisfaction, health, and quality of life outcomes for patients with SSc.

ACKNOWLEDGMENT

We acknowledge Associate Professor Kathleen Tymms for providing input on the interpretation of the data and manuscript revision.

REFERENCES

1. Elhai M, Meune C, Avouac J, Kahan A, Allanore Y. Trends in mortality in patients with systemic sclerosis over 40 years: a systematic review and meta-analysis of cohort studies. Rheumatology 2012;51:1017-26.
2. Kowal-Bielecka O, Landewé R, Avouac J, Chwiesko S, Miniati I, Czirjak L, et al; EUSTAR Co-Authors. 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.
3. Mills SD, Fox RS, Merz EL, Clements PJ, Kafaja S, Malcarne VL, et al. Evaluation of the Satisfaction with Appearance Scale and its short form in systemic sclerosis: analysis from the UCLA scleroderma quality of life study. J Rheumatol 2015;42:1624-30.
4. Kwakkenbos L, van Lankveld WG, Vonk MC, Becker ES, van den Hoogen FH, van den Ende CH. Disease-related and psychosocial factors associated with depressive symptoms in patients with systemic sclerosis, including fear of progression and appearance self-esteem. J Psychosom Res 2012;72:199-204.
5. Levis B, Burri A, Hudson M, Baron M, Thombs BD; Canadian Scleroderma Research Group (CSRG). Sexual activity and impairment in women with systemic sclerosis compared to women from a general population sample. PLoS One 2012;7:e52129.
6. Poole JL, Willer K, Mendelson C, Sanders M, Skipper B. Perceived parenting ability and systemic sclerosis. Musculoskeletal Care 2011;9:32-40.
7. Poole JL, Chandrasekaran A, Hildebrand K, Skipper B. Participation in life situations by persons with systemic sclerosis. Disabil Rehabil 2015;37:842-5.
8. Tong A, Flemming K, McInnes E, Oliver S, Craig J. Enhancing transparency in reporting the synthesis of qualitative research: ENTREQ. BMC Med Res Methodol 2012;12:181.
9. Tong A, Sainsbury P, Craig J. Consolidated criteria for reporting qualitative research (COREQ): a 32-item checklist for interviews and focus groups. Int J Qual Health Care 2007;19:349-57.
10. Brown SJ, Somerset ME, McCabe CS, McHugh NJ. The impact of group education on participants' management of their disease in lupus and scleroderma. Musculoskeletal Care 2004;2:207-17.
11. Cinar FI, Unver V, Yilmaz S, Cinar M, Yilmaz F, Simsek I, et al. Living with scleroderma: patients' perspectives, a phenomenological study. Rheumatol Int 2012;32:3573-9.
12. Ennis H, Herrick AL, Cassidy C, Griffiths CE, Richards HL. A pilot study of body image dissatisfaction and the psychological impact of systemic sclerosis-related telangiectases. Clin Exp Rheumatol 2013;31 Suppl 76:12-7.
13. Joachim G, Acorn S. Life with a rare chronic disease: the scleroderma experience. J Adv Nurs 2003;42:598-606.
14. Kocher A, Adler S, Spichiger E. Skin and mucosa care in systemic sclerosis—patients' and family caregivers' experiences and expectations of a specific education programme: a qualitative study. Musculoskeletal Care 2013;11:168-78.
15. Mendelson C, Poole JL. Become your own advocate: advice from women living with scleroderma. Disabil Rehabil 2007;29:1492-501.
16. Mendelson C, Poole J, Allaire S. Experiencing work as a daily challenge: the case of scleroderma. Work 2013;44:405-13.
17. Oksel E, Gündüzoğlu NC. Investigation of life experiences of women with scleroderma. Sex Disabil 2014;32:15-21.
18. Sandqvist G, Hesselstrand R, Scheja A, Hakansson C. Managing work life with systemic sclerosis. Rheumatology 2012;51:319-23.
19. Stamm TA, Mattsson M, Mihai C, Stöcker J, Binder A, Bauernfeind B, et al. Concepts of functioning and health important to people with systemic sclerosis: a qualitative study in four European countries. Ann Rheum Dis 2011;70:1074-9.
20. Stamm T, Hieblinger R, Bostrom C, Mihai C, Birrell F, Thorstensson C, et al. Similar problem in the activities of daily living but different experience: a qualitative analysis in six rheumatic conditions and eight European countries. Musculoskeletal Care 2014;12:22-33.
21. Suarez-Almazor ME, Kallen MA, Roundtree AK, Mayes M. Disease and symptom burden in systemic sclerosis: a patient perspective. J Rheumatol 2007;34:1718-26.
22. Ananian CA. Factors that influence exercise in non-exercisers, insufficiently active individuals and exercisers with arthritis: a quantitative and qualitative approach [dissertation]. Columbia: University of South Carolina; 2005.
23. Lees RJ. The effectiveness of an arthritis self-management program on a population of persons with scleroderma [dissertation]. Vancouver: University of British Columbia; 1990.

24. Johnson A. Work experiences of people with scleroderma, an exploratory study [dissertation]. Bristol: University of the West of England; 2006.
25. Mills SL. Living well with chronic illness: aligning tensions, attitudes, strategies, and meanings [dissertation]. Vancouver: University of British Columbia; 2006.
26. Newton E. The experience of emotional distress in women with scleroderma: a qualitative study [dissertation]. Montreal: McGill University; 2010.
27. Thorne SE. Navigating troubled waters: chronic illness experience in a health care crisis [dissertation]. Ohio: The Union Institute of Advanced Studies; 1991.
28. Wood SL. Losing face: The effects of scleroderma-related facial changes on the relationship with self and others [dissertation]. San Francisco: California Institute of Integral Studies; 2000.
29. Decuman S, Smith V, Grypdonck M, De Keyser F, Verhaeghe S. Factors influencing the occupational trajectory of patients with systemic sclerosis: a qualitative study. Clin Exp Rheumatol 2015;33 Suppl 91:S26-30.
30. Mendelson CF, Greaves J, Poole JL. Changes in leisure participation in persons with systemic sclerosis [abstract]. Arthritis Rheum 2012;64:S1112.
31. Mittoo S, Frankel S, LeSage D, Castelino FV, Christopher-Stine L, Danoff S, et al. Patient perspective informs core sets, constructs of metrics and communication tools for patients with connective tissue disease related interstitial lung disease [abstract]. Arthritis Rheum 2012;64:S630-1.
32. Mittoo S, Saketkoo L, Swigris J, LeSage D, Fischer A, Frankel S. Living with connective tissue related interstitial lung disease: patient experiences of the disease over time [abstract]. Arthritis Rheum 2011;63 Suppl 10:1470.
33. Mouthon L, Alami S, Desjeux D, Hachulla E, Poiraudeau S. Patients', relatives' and practitioners' views concerning SSc and its management: a qualitative interview study [abstract]. Rheumatology 2012;51:ii75.
34. Poole JL, Haygood D, Mendelson CF. The challenges of being a father with systemic sclerosis [abstract]. Arthritis Rheum 2011;63 Suppl 10:2412.
35. Stamm T, Dür M, Sadlonova M, Stoffer M, Haider S, Smolen J. Life stories, gender and chronic autoimmune diseases: results of a qualitative study. Ann Rheum Dis 2013;71 Suppl 3:755.
36. Sutanto B, Singh-Grewal D, McNeil HP, O'Neill S, Craig JC, Jones J, et al. Experiences and perspectives of adults living with systemic lupus erythematosus: thematic synthesis of qualitative studies. Arthritis Care Res 2013;65:1752-65.
37. Lempp H, Scott D, Kingsley G. The personal impact of rheumatoid arthritis on patients' identity: a qualitative study. Chronic Illn 2006;2:109-20.
38. Uttjek M, Nygren L, Stenberg B, Dufåker M. Marked by visibility of psoriasis in everyday life. Qual Health Res 2007;17:364-72.
39. Poole JL, Skipper B, Mendelson C. Evaluation of a mail-delivered, print-format, self-management program for persons with systemic sclerosis. Clin Rheumatol 2013;32:1393-8.
40. van der Vaart R, Repping-Wuts H, Drossaert CH, Taal E, Knaapen-Hans HK, van de Laar MA. Need for online information and support of patients with systemic sclerosis. Arthritis Care Res 2013;65:594-600.
41. Sharpe L, Sensky T, Timberlake N, Ryan B, Brewin CR, Allard S. A blind, randomized, controlled trial of cognitive-behavioural intervention for patients with recent onset rheumatoid arthritis: preventing psychological and physical morbidity. Pain 2001; 89:275-83.
42. Rosen JC, Reiter J, Orosan P. Cognitive-behavioral body image therapy for body dysmorphic disorder. J Consult Clin Psychol 1995;63:263-9.
43. Macedo AM, Oakley SP, Panayi GS, Kirkham BW. Functional and work outcomes improve in patients with rheumatoid arthritis who receive targeted, comprehensive occupational therapy. Arthritis Rheum 2009;61:1522-30.
44. Baldwin D, Johnstone B, Ge B, Hewett J, Smith M, Sharp G. Randomized prospective study of a work place ergonomic intervention for individuals with rheumatoid arthritis and osteoarthritis. Arthritis Care Res 2012;64:1527-35.
45. Kwakkenbos L, Jewett LR, Baron M, Bartlett SJ, Furst D, Gottesman K, et al. The Scleroderma Patient-centered Intervention Network (SPIN) Cohort: protocol for a cohort multiple randomised controlled trial (cmRCT) design to support trials of psychosocial and rehabilitation interventions in a rare disease context. BMJ Open 2013;3.
46. Hawley D. Psycho-educational interventions in the treatment of arthritis. Baillieres Clin Rheumatol 1995;9:803-23.
47. Furst D, Khanna D, Matucci-Cerinic M, Clements P, Steen V, Pope J, et al. Systemic sclerosis - continuing progress in developing clinical measures of response. J Rheumatol 2007;34:1194-200.
48. Steen VD, Medsger TJ Jr. The value of the Health Assessment Questionnaire and special patient-generated scales to demonstrate change in systemic sclerosis patients over time. Arthritis Rheum 1997;40:1984-91.
49. Heinberg LJ, Kudel I, White B, Kwan A, Medley K, Wigley F, et al. Assessing body image in patients with systemic sclerosis (scleroderma): validation of the adapted satisfaction with appearance scale. Body Image 2007;4:79-86.
50. World Health Organization. International classification of functioning, disability and health (ICF). [Internet. Accessed March 22, 2016.] Available from: www.who.int/classifications/icf/en
51. Saketkoo LA, Escorpizo R, Keen KJ, Fligelstone K, Distler O; EUSTAR. International Classification of Functioning, Disability and Health Core Set construction in systemic sclerosis and other rheumatic diseases: a EUSTAR initiative. Rheumatology 2012;51:2170-6.

APPENDIX 1. Search strategy

Database, Search Range	Date of Search	Search Terms
Ovid MEDLINE, 1946 to 13/10/2014	13/10/2014	[exp Scleroderma, Systemic/ OR (systemic adj3 sclero$).tw. OR systemic sclerosis.tw. OR systemic scleroderma.tw. OR (diffuse adj2 sclero$).tw OR (limited adj2 sclero$).tw.] AND [exp qualitative research/ OR qualitative.tw. OR interview$.tw. OR focus group$.tw. OR (thematic$ or theme$).tw. OR grounded theory.tw. OR phenomenol$.tw. OR content analysis.tw. OR ethnograph$.tw. OR exp decision making/ OR exp Illness Behavior/ OR exp Knowledge/ OR Health Knowledge, Attitudes, Practice/ OR observation$.tw.]
Embase, 1974 to 13/10/2014	13/10/2014	[exp systemic sclerosis/ OR (systemic adj3 sclero$).tw. OR systemic sclerosis.tw. OR systemic scleroderma.tw. OR (diffuse adj2 sclero$).tw OR (limited adj2 sclero$).tw.] AND [exp qualitative.tw. OR interview$.tw. OR focus group$.tw. OR (thematic$ or theme$).tw. OR grounded theory.tw. OR phenomenol$.tw. OR content analysis.tw. OR ethnograph$.tw. OR exp decision making/ OR exp Illness Behavior/ OR observation$.tw. OR exp health belief/ OR exp social belief/ OR exp Knowledge/ OR Health Knowledge, Attitudes, Practice/ OR exp social psychology/]
PsycINFO, 1806 to 13/10/2014	13/10/2014	systemic sclerosis.tw. OR systemic scleroderma.tw. OR (systemic adj3 sclero$).tw OR (diffuse adj2 sclero$).tw. OR (limited adj2 sclero$).tw.
CINAHL, to 13/10/2014	13/10/2014	(MH "Scleroderma, Systemic+") Limiters - Clinical Queries: Qualitative - Best Balance