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RUNNING HEAD: Scleroderma and close social relationships

Scleroderma Patient Perspectives on Social Support from Close Social Relationships RUNNING HEAD: Scleroderma and Close Social Relationships

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Abstract

Purpose: The purpose of this study was to deepen our understanding concerning the ways that social support from close relationships can impact a patients' ability to cope with scleroderma.

Method: Four focus groups were conducted with patients diagnosed with scleroderma (N = 19). A semi-structured interview guide was used. Discussions were recorded and transcribed, and a thematic analysis performed.

Results: Patients reported receiving three types of social support (i.e., emotional, informational, instrumental), with emotional support standing out as a priority. Patients also referred to three relational factors (i.e., communication style, active engagement, complementarity) that either enhanced or impeded the support received. More specifically, honest communication by the patients, patients' careful selection of sources of support, and close relationships motivated to learn and get involved enhanced social support. However, patients who avoided others or avoided speaking about scleroderma, people with a lack understanding, and people who do not get involved impeded support. **Conclusion:** Scleroderma patients might benefit from supportive interventions aimed at helping them cope better with the disease as a collective, rather than exclusively supporting patients to cope on their own. Findings from this study help better understand the unique experiences of scleroderma patients while receiving support in close relationships.

Keywords: scleroderma, systemic sclerosis, social support, close social relationships, coping, patient perspectives

Introduction

People diagnosed with rare chronic diseases can experience a wide variety of challenges related to disease symptoms, functional impairments, and emotional distress [1-3]. Thus, it is important for patients with rare diseases to develop coping skills that help them manage the physical and psychological impact of their disease. Yet, adopting effective coping skills can be a particular challenge for these patients because support services and knowledge about their illness are relatively scant. Further complicating this, coping is not just an individual process [4, 5]. Rather, according to Bandura's social cognitive theory, coping is a collective process that can be impacted by the support networks of patients, including their close social relationships and the community at large [4, 5].

Systemic sclerosis (SSc), also known as scleroderma, is a rare and chronic rheumatic disease that involves dysfunction of the immune system and excess formation of connective tissues [6]. This systemic disease includes two distinct sub-types (i.e., limited SSc and diffuse SSc) and is associated with a wide variety of physical symptoms, such as Raynaud's phenomenon, interstitial lung disease, ulcers, and joint contractures, among others [7-9]. The median survival time after receiving a diagnosis is only 11 years and there is no known cure at this time [6, 8].

Due to the elevated burden associated with SSc, which includes dealing with a variety of disease symptoms and an unpredictable disease progression, SSc patients tend to experience increased psychological distress [10-15]. Thus, learning strategies and behaviours to improve their ability to cope can greatly affect patient well-being [18, 19]. For one, understanding the social networks of SSc patients and how patients interact with

social support could be critical to understanding their coping needs and helping them better manage.

Social support has been associated with the prevention and improvement of patients' physical health outcomes, quality of life, and emotional well-being, as well as reduced morbidity and mortality [24-28]. Social support was originally described as information (e.g., tips to address needs) provided by a supportive network that would allow a person to feel cared for [29]. Currently, other types of social support have also been described, with the most common ones being instrumental support (e.g., assistance with tangible and concrete needs or daily tasks, such as providing patients helpful products or accessing resources for them) and emotional support (e.g., defined by more intangible qualities, like caring, sympathy, understanding, and comprehension) [24, 25].

According to research by Cantor, social support can further be classified into two main categories [30, 31]: formal and informal support. Formal support is frequently carried out in official, organizational settings and characterized by low levels of intimacy in the context of the relationship (e.g., health care professionals, nurses, physiotherapists) [31-33]. Informal support, on the other hand, involves close social relationships with people who are frequently in contact with the patient and with whom the patient feels more intimacy (e.g., family members and friends) [31-33]. Hence, while health care professionals will offer essential health information and care to patients as part of their occupational demands, close social relationships will often provide more emotional and tangible support for patients' daily needs with no financial benefit [34, 35]. Furthermore, while close relationships are typically defined by openness to self-disclosure contributing

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to more intimacy, the level of self-disclosure and intimacy within close relationships is highly variable and can change over time [36, 37].

Another significant feature of social support is that the provision and reception of support is not a linear phenomenon. Rather, it is a dynamic process influenced by many factors, including sociocultural (i.e., socio-demographic characteristics), situational or environmental (i.e., illness and illness-related stressors), temporal (i.e., timing of illness, disease stage and severity), and interpersonal or relational (i.e., the relational nature of the support system) [38, 39], and that may influence the type and extent of social support received. Most notably, this means that support from close social relationships can vary in terms of quality and quantity based on the patient's subjective perception of the received social support [28]. Perceptions of social support are what matters most because it is this perception that will influence the patient's openness to receive and benefit from the support provided [28, 40]. For instance, sociodemographic characteristics, like gender, are related to having different perceptions of support [22, 41-43]. In one review, men diagnosed with diabetes or heart failure perceived that they received more support from family members and were faced with fewer family barriers for engaging in self-care (e.g., being criticized about medical care) than women [22].

Although patients with SSc have identified social support as a priority for improving quality of life [12-14, 18, 19, 44-47], available evidence is limited, especially when comparing to research on more common chronic diseases. The purpose of the current study was therefore to develop a deeper understanding of the potential impact that support from close social relationships could have on the ability of SSc patients to actively cope with their disease. Notably, this exploratory investigation served as a follow-up to a previous focus group study looking at SSc patient and health care professional perspectives related to challenges in coping and different strategies aimed at enhancing coping with the disease [49]. The research questions guiding the current study were: (1) What types of social support do patients with SSc receive from close social relationships? (2) What are the different relational factors that impact a patients' perception of the social support received?

Methods

Study Design

Focus groups were conducted to stimulate an open dialogue among participating SSc patients. This research strategy has often been used with minority populations [50] including rare diseases populations, like SSc [1, 2, 13-15, 44, 51]. Furthermore, focus groups can facilitate the discussion of sensitive topics [52] and improve the understanding of common experiences by promoting the exchange of differing viewpoints [52, 53]. For these reasons, the present study was framed within the boundaries of a social constructionist approach, with an emphasis on the importance of building shared meaning [53].

Participants

Recruiting individuals diagnosed with a rare disease can be challenging. As such, four focus groups were conducted during two major North American SSc conferences; two in Canada and two in the United States. The focus groups took place in September 2014 and July 2015. They were held in private meeting rooms that had been reserved in the same hotels as the conferences. Refreshments and/or meals were offered to patients with SSc as compensation for participating. We aimed to include 5 to 10 patients per focus group, with discussions lasting approximately 90 minutes.

Eligibility criteria for patients with SSc included: 1) having received a diagnosis of SSc, 2) being an English-speaker, and 3) being at least 18 years of age. Advertisements for study recruitment were emailed directly to registered attendees by conference organizers in the weeks leading up to the conferences.

Data Collection and Procedures

The study was approved by the research ethics committee of the Jewish General Hospital in Montreal, Canada. Before participating in a focus group, SSc patients signed an informed consent form and completed a socio-demographic questionnaire. Focus groups began with a standard statement about the purpose of the study, procedures to be followed by patients, as well as a basic introduction to the concept of social support. Next, a discussion took place that was centered on participant perspectives about social support while actively coping with and managing SSc. A semi-structured interview guide was used, which included specific questions and prompts (e.g., Describe your experiences getting support for coping with SSc. What do you need to feel supported? What type of support is most important to you?). However, the guide was also flexible enough to permit the expression of unique or new ideas related to the topic that were not present in the original interview guide, as well as foster discussions of ideas related to social support that might be uniquely important to SSc patients.

All focus groups were led by the same two moderators who were graduate-level trainees in psychology with research experience in SSc. The use of two moderators is consistent with standard methods for focus group research: one moderator primarily led

the discussion and the other mainly observed and took notes, but also contributed to the discussion whenever needed [54, 55]. The focus groups were audio and video recorded and transcribed verbatim by an external trained professional [55]. All transcripts were reviewed by the main moderator to ensure accuracy of content prior to coding.

Data Analysis

Focus group transcriptions were uploaded to the qualitative data analysis software *Atlas.ti* [56] to help with data storage, coding, and the organization and retrieval of codes. A thematic analysis was performed [55, 57, 58], which included two phases. In the first phase, a preliminary codebook with codes (i.e., selection of smaller, meaningful text segments) and themes (i.e., abstracted data that represent main findings) was developed [58]. Identified themes were based on pre-existing research concepts and the original ideas presented by the patients. In the second phase, the preliminary codebook was further refined through discussions between two coders who met regularly to revise the coding system. Any discrepancies in the coding system were resolved through either an in-depth discussion between coders until agreement was reached or by consultation with a third party, in this case, a member of the research team with a background in qualitative research. Reflexivity in the form of field and analytic memos was also conducted and used to enhance the thematic analysis [59].

For the current research report, only discussions that were related to the research questions were considered for analytical purposes. Consequently, participants' descriptions of social support received from health care professionals or other patients with SSc were excluded, as well as any comments not directly related to coping. To highlight the views of patients with SSc, we provided a summary of the focus group discussions and extracted representative quotes from the transcripts. Quotes were modified as needed to correct for grammatical errors or verbal fillers, while also taking care to maintain the original meaning expressed by patients [60]. In line with previous research endeavors [18, 49], and to preserve patients' anonymity, individual sociodemographic characteristics were not provided and pseudonyms were used when presenting selected quotes.

Results

Participant Characteristics

A total of 19 patients with SSc participated in the focus groups, of which 17 were women (1st focus group = 4 females, 2^{nd} = 4 females, 3^{rd} = 4 females, 4^{th} = 5 females, 2 males). The age of patients ranged from 31 to 74 years old with a mean age of 57.1 years old (Standard Deviation [SD] = 10.9). The mean number of years since receiving a SSc diagnosis was 18.2 years (SD = 13.2). While some patients had received their diagnosis only a year prior, others had been living with the diagnosis for up to 46 years. Seven (37%) patients reported a diagnosis of diffuse SSc; three (16%) reported limited SSc; nine (47%) reported CREST syndrome, which is a term used in the past to denote limited SSc and an acronym that stands for the main features of the disease, which are Calcinosis, Raynaud's phenomenon, Esophageal dysmotility, Sclerodactyly, and Telangiectasia. A total of 12 (63%) patients indicated being White and 15 (79%) had completed at least some college/university education. A more detailed summary of patient characteristics is provided in Table 1.

[Table 1 about here]

Coping with SSc with Social Support: Patient Perspectives

Patients shared their perspectives about how they actively coped with SSc while receiving support from close social relationships, including partners, children, siblings, extended family, and other people who were described as friends due to emotional intimacy or frequency of time spend together. Two main themes (and their correspondent sub-themes) were identified: 1) *types of perceived social support* (i.e., emotional support, informational support, and instrumental support) and 2) *relational factors that enhanced or impeded social support* according to patients' perception (i.e., communication style, active engagement, and complementarity). A diagram illustrating the relationships between the main themes and sub-themes related to coping while receiving support from close social ties are provided in Figure 1.

[Figure 1 about here]

Types of perceived social support

Emotional Support

Emotional support was characterized by the provision of support that is intangible and difficult to measure, such as an attitude or a gesture. Although patients reported receiving other types of support while interacting with people close to them, they often prioritized emotional support above all other forms. Patients expressed how being emotionally understood and encouraged by someone close to them served as an important source of comfort. For instance, it was helpful for patients to have someone close to them with a supportive attitude about the diagnosis and who knew them enough to know when and how to support them emotionally without causing more distress.

Having close social relationships with people who knew how to reduce the emotional strain associated with talking about the disease was also beneficial. Examples of this included people knowing when to talk (or not to talk) about the disease, using agreed-upon shorthand phrases that referred to topics the patients did not want to discuss explicitly (e.g., asking patient: "How many spoons do you have left?" instead of: "Are you tired?"), or consciously avoiding repeatedly naming the disease or discussing specific symptoms. These attitudes and gestures helped patients to cope better with their disease, and at the same time, avoid emotional exhaustion.

According to patients, they also felt emotionally supported by close social ties when the latter knew them so well that they were able to simply predict their needs. This minimized the necessity to talk about the disease and to repeatedly clarify their concerns. Jodie (63 years old, diffuse SSc, retired) shared:

If it wasn't for my husband, I don't know where I'd be because I'm independent, I don't like to ask for [things] and he reads my mind [...]; he knows [my needs] and he takes a lot of the load I have [...], but graciously and delicately.

Other times, patients felt emotionally supported by close social ties who were understanding and accepting of necessary lifestyle changes and adaptations they had to make, such as wearing gloves or sweaters in the summer to prevent circulation problems from being exacerbated by air conditioning. Aaron (74 years old, CREST syndrome, retired) explained: "If there's outdoor dining, I will ask if it's comfortable enough for them to eat outdoors. If not, I'll sit [indoors] with a jacket [to face the air conditioning]. But [often] they understand where I'm coming from."

Family members and friends also provided emotional support by motivating patients to continue coping actively (e.g., attending educational conferences, socializing) despite the many challenges they face. In so doing, these close relationships reduced patients' desire to engage in behavioral or cognitive avoidance, which patients reported as contributing to them being insufficiently informed and ill-prepared to cope with their disease. Karen (47 years old, CREST syndrome, on disability) explained that her mother had been especially motivating in terms of helping her overcome her hesitation about attending SSc conferences: "She says, 'You're going.' And so, she kind of just kicked me in my [butt] and [I decided], 'Okay. I'll do this just to keep my mother happy.'" Without motivation from an emotionally supportive close social tie, some SSc patients struggled to act on their knowledge or use positive coping skills because they felt too stressed. *Informational Support*

The patients also referred to the provision of advice, information, and expertise. Many patients reported that close social relationships offered them informational support, even prior to receiving a formal diagnosis of SSc. This information helped the patients discuss and gain knowledge about the likely causes of physical symptoms, as well as find ideas of what to discuss with their health care professionals, and prepare themselves for future challenges. Moreover, some patients discussed how family members who had previously received a SSc diagnosis (or another chronic disease diagnosis) shared helpful insights about managing symptoms, dealing with disease progression, and coping with daily disease-related challenges.

Family members also helped patients access information about SSc and disease management by sharing their own skills and expertise on how to access information, such as navigating the internet, accessing research evidence, and interpreting research findings. John (56 years old, diffuse SSc, employed full-time) shared his experience of the support received from his brother: "I can read [a research] paper [...]. [But] if I don't

understand something, my brother, [who] has a post doctorate degree [...] [and who] did research [in chronic diseases, will help]. I'll send it to him [...] and ask what he thinks [about it]." SSc patients in the current study also discussed the benefits of having close social ties that were knowledgeable about how to find and connect to available resources, such as support groups, educational conferences, and hospitals with specialized SSc services.

Likewise, family members who worked as health care professionals (e.g., nurses, doctors) were important sources of informational support for patients because they often had insider knowledge about how to navigate the health care system more efficiently (e.g., which specialist to see, how to access services and treatments, overcoming hospital bureaucracy). Stephanie (32 years old, limited SSc, on leave of absence) explained her experience:

My mother is a nurse [...and] I think that makes a huge difference [...compared] to SSc patients who don't have family in the medical field. It's [...] like a bonus. She knows where to go. Not about the disease, but where to [start in case of need].

Instrumental Support

Examples of instrumental support that patients shared mainly revolved around received support related to medical commitments, like offering them a lift to attend their appointments, accompanying them to medical appointments to improve recall of treatment recommendations, and identifying and connecting patients to medical specialists or doctors with expertise in SSc.

Patients appreciated as well the support provided by family members and friends that helped them manage daily life challenges, including their assistance with household

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tasks (e.g., redistribution of responsibilities in the household) or providing new adaptive resources or products (e.g., heating pads, extra warm clothing, adapted household tools like an electric can opener). Rachel (65 years old, limited SSc, retired) shared the following: "[My father-in-law is] just watching me, [...] realizing: 'That's not working for her. What can I do to make it work?" For instance, he noticed that, "I can't pull my oven [...] racks out. So he made me this little thing. It's almost like a back scratcher but it's more solid, and you reach in there and you pull your bottom rack out."

Impact of Relational Factors on Social Support

Communication Style

One relational factor identified by SSc patients as either enhancing or impeding social support was communication style. For instance, many patients reported that when they made efforts to maintain an open and honest line of communication about the diagnosis and disease symptoms, functional limitations and abilities, social support needs, and emotional distress; then, the quality of social support received was often enhanced. For these patients, clarity in communication was crucial to resolving misunderstandings or incorrect assumptions about their illness and well-being. As such, when they clearly stated their needs, their close social relationships could respond and subsequently provide appropriate forms of support. Vicky (49 years old, diffuse SSc, retired) explained:

People aren't going to know what I need if I don't tell them. So I had to learn how to communicate more efficiently [...] and to tell people what I needed so that they

could be there for me. [...]. And so for me it [...was], "I need you to go to the grocery store for me," "I need you to pick up some meds," or "I need you to come over and wash my hair."

Patients also brought up two issues related to communication style that impeded their ability to cope better using social support. In some cases, patients withheld information about their diagnosis from close social ties out of fear of potential negative reactions. In turn, poor communication due to non-disclosure on the patients' side resulted in limited knowledge by family and friends, which then contributed to close relationships experiencing increased anxiety, and enacting more fearful and avoidant behaviors towards the patient. Vicky (49 years old, diffuse SSc, retired) explained: "It's hard for them to see me sometimes. They're always protective. And so sometimes [...], I don't share as much with them. [...] I tend to protect them."

Moreover, patients found that close social ties with insufficient understanding about the disease could sometimes hinder both communication and the patients' likelihood to receive appropriate support; a fact exacerbated by the patients' tendency to withhold information from friends and family members. Moreover, even close social ties that made conscious efforts to stay engaged and attuned to patient needs often lacked sufficient understanding of the disease in order to provide appropriate support. Thus, close social relationships that lacked an understanding of the disease often expressed unhelpful judgments, fears, or opinions that were unsupportive and led the patients to detrimental coping. Common examples were that people close to them didn't necessarily comprehend the patient's need to adapt their lifestyle, make adjustments to old hobbies, reduce the frequency of social activities, or minimize travel because of physical limitations and symptoms, including chronic fatigue or chronic pain.

Active Engagement

Patients with SSc shared that having close social ties that actively raised awareness about the disease in the community was helpful because it reduced barriers to coping that were related to a lack of disease knowledge (e.g., receiving negative judgment, misunderstandings while engaged in daily tasks). In addition, patients discussed the benefit of having close social ties that were motivated to learn about the disease and get involved in the patient's life, despite possible fears or personal discomfort. For instance, Rachel (65 years old, limited SSc, retired) shared: "My husband [goes] along [with me to SSc conferences]. And he's gotten very comfortable because he's seeing other people [with the disease]. [...] But he [wasn't always] comfortable [before]. So [...] education [was a] big thing [for him]." Increased exposure to SSc knowledge and other patients in the larger SSc community, may provide close social ties with opportunities to learn about the disease and reduce their distress about the unknown.

Close relationships also helped patients reduce their distress by developing a clear action plan for the patient, especially when they felt too overwhelmed to do so alone. Having immediate sources of support who were involved was especially beneficial for patients when facing scary times, for instance during extended hospital stays or while struggling with severe symptom flare-ups. Moreover, patients noted that the mere presence of close family and friends in these difficult, stressful, and uncertain times, was reassuring for them.

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Patients with SSc also discussed the negative impact of having close social ties that could not (or would not) get involved in disease management or coping efforts. For instance, having close relationships engage in fearful-avoidant behaviours (e.g., refusing to talk about SSc, avoiding spending time with patient) could interfere with patients' ability to receive support. Patients reported that many people close to them would react by keeping their distance because they were frightened of witnessing the impact of the disease or observing the patient's daily struggles. Amanda (56 years old, CREST syndrome, employed part-time) shared the following: "My niece and nephew [don't] want to be part of this because it's scary to [them]." Other times, patients reported that their close social ties were simply too focused on their own needs to provide reliable support. Patients described that regardless of the underlying motives of people close to them (e.g., anxiety, focused on their own lives), this lack of involvement contributed to increased difficulties coping with the disease, including more emotional distress and reliance on financial resources to compensate for the lack of support from close social ties.

Complementarity

This relational factor was understood as the congruence of coping styles between patients and their close social relationships. Patients discussed the importance of being selective and careful when choosing which close social ties to rely on for support, based on a cursory assessment. For instance, patients with SSc reported that they were more willing to interact with and receive support from close relationships when the support provided was consistent with their actual needs and preferences. Congruence of coping styles could either involve their unique way of listening to the patient's concerns, their approach to motivating them to cope, or the forms of practical assistance to carry out and facilitate patients' daily life. Jodie (63 years old, diffuse SSc, retired) explained that, for her, surrounding herself with people who could laugh and be positive was key:

I think it's [...] important [...] to surround yourself with people who are upbeat [and] have a good sense of humor [...]. I use [my] sense of humor to get me through [...] my day. But there are people out there, and they might have been friends at one time, but they [are] toxic in their way of being around you [because] they're negative. [I] don't want to be around people who are negative.

Patients also discussed the importance of ongoing congruity in terms of coping styles because disease symptoms and support needs can evolve over time due to disease progression. The ability of close social relationships to provide appropriate support may also improve over time, as these individuals may become more knowledgeable and comfortable with the illness and more attuned to the patients' needs.

Discussion

Social support provided by close social relationships is an important factor for patients coping with rare chronic diseases like SSc. Overall, patients in the current study found that receiving emotional, informational, and instrumental support from close social relationships allowed them to be better equipped to face the various challenges associated with the disease and to engage in more effective coping behaviours. Although all types of support were described as beneficial, receiving emotional support from close relationships was often prioritized because of the intimate knowledge these people had of the patient's needs and preferences (i.e., compared to health care professionals or community acquaintances). Additionally, different relational factors that may impact the ability of SSc patients to benefit from social support were identified. These relational factors included communication style, active engagement, and complementarity. Mainly, patients highlighted the benefits of clear and honest communication, interacting with people who were motivated to learn about SSc, and being selective when choosing their sources of social support. However, patients shared benefitting less from social support when they received judgment and criticism from them, when they avoided interacting with others or sharing details about the disease, or when family and friends refused to get involved in their care.

Even if the current research about the role of social support for SSc patients is limited to a few qualitative studies investigating SSc coping and quality of life [18, 19, 49], this study seems to corroborate previous findings. For instance, while patients with SSc reported benefitting from all types of social support, past research found that the key to navigating health care was relying on friends and family members who would not deny their limitations or necessary adaptations [19]. Past research also found that coping while dealing with the burden of the disease in daily life involved seeking out social support to cope with difficult emotions [18, 49], as well as to resolve challenges accessing information and resources [49]. Furthermore, some SSc studies found that disclosure/non-disclosure to others [13](i.e., communications style) and social isolation and social interactions [12](i.e., active engagement) can influence SSc patient quality of life.

The results of this study were also in line with findings from general chronic disease research [61-63]. These past studies found that both instrumental and emotional support received by family and friends improved coping behaviours [63] and disease

management outcomes for chronic disease patients [62]. Furthermore, although informational and instrumental support were identified as helpful at times, emotional support was deemed more effective at improving patient well-being and most important when received in the context of close relationships [61]. Past research also identified health care professionals as the most trustworthy source of information about the disease and medications [34], thus explaining why informational support from family and friends may be less critical for patients. Previous research investigating relational factors and coping in common chronic diseases seems consistent with the current findings as well [21, 64]. For instance, in one study, identifying and connecting to relevant social network resources (i.e., active engagement), negotiating within networks to help shape the relationship, expectations, and communication (i.e., communication style), and developing a shared perspective and shared objectives (i.e., complementarity) were identified as factors that could affect patient engagement with social support [64].

To our knowledge, this is the first study that has directly examined the role of social support in SSc, as previous studies have only inferred about social support through research on disease coping and quality of life [18, 19, 49]. Of note, social support was the focus of the current study, not social networks, as the goal was to explore the function of social network ties (e.g., social influence, companionship, types of support) not the structure of the social networks themselves (e.g., size, frequency of contact, intimacy) [24, 25, 28]. The current study therefore extended existing research knowledge by revealing a number of previously unknown priorities and needs for SSc patients while seeking social support from close relationships. For instance, patients with SSc discussed the benefits of receiving more concrete forms of support (i.e., instrumental and

informational support) from family and friends while navigating a wide variety of daily life situations. However, it was only when their close social ties had specialized knowledge or skills (e.g., enhanced understanding of healthcare system) that the patient came to rely on them for informational or instrumental support. Otherwise, receiving emotional support from family and friends was deemed more beneficial. Further, it was identified in the present study that while some patients with SSc did not seek support from others simply out of habit (e.g., self-sufficiency always encouraged as a family norm), many avoided requesting support out of fear that it would cause their close social ties to be excessively worried or anxious.

The current study brought to light the role of a few different relational factors in the context of SSc patients receiving social support from close social relationships. However, it is possible that other factors, not discussed in the current study, have a greater effect on SSc patient perceptions of social support. For patients with other chronic diseases, the socio-demographic characteristics of the patient, the family context, and the disease stage and severity [38, 39] can all have an influence on perceptions of social support. For instance, important gender differences have been found to be related to how chronic disease patients perceive and experience social support [22, 41-43, 65] In addition, because SSc involves many symptoms that are less visible (e.g., internal organ involvement) the close social ties of patients are often left with limited information about patient symptoms and well-being unless there is active communication. Thus, the nature of the disease might affect SSc patient perceptions of social support (i.e., lack of support for symptoms that are invisible to others) more so than relational factors that are often less disorder-specific. Finally, it is possible that there are yet unknown life context factors moderating the relationship between social support and coping for SSc patients [38, 39].

Study Limitations

Some important limitations should be considered. First, although efforts were made to increase the number of patients by running focus groups during SSc patient education conferences (i.e., to address SSc disease rarity and difficulties involved with bringing together a large number of patients) [1], the sample size remained small and did not allow for theoretical saturation [66, 67]. Second, because we sought to recruit patients at conferences, participants were sampled conveniently. The use of convenience sampling is a limitation because it might increase the likelihood that patients were people who were physically able to travel (e.g., experienced less severe disease involvement, had more financial means), motivated to learn about the disease, and thus had more experience coping, which may not be representative of all patients with SSc. On the other hand, enrollment of patients more active in the SSc community may have allowed for more fruitful focus group discussions. Third, assessing possible differences between or within sub-groupings of patients with SSc (e.g., men versus women, diffuse versus limited SSc sub-types) was not possible because the study was exploratory and not designed to assess group differences [52]. Finally, a fourth limitation involves the use of a semi-structured interview, which had the goal to better follow the flow of patients' discussions and to capture their unique interests [55]. Although this resulted in not being able to perform a detailed comparison of findings across focus groups, it was beneficial because it allowed for more flexibility to explore ideas that emerged directly from the patients [55].

Future Directions for Research

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Based on the current study findings, there are many additional topics that would be beneficial to research in the future. For instance, some studies have found that different sources of support, including family and close friends, health care professionals, community acquaintances, and even other chronic disease patients, might provide different kinds of support and have different effects on patient coping [38]. Some studies have even found differences in social support received from family members versus close friends [21]. In time, quantitative research assessing the relationship between perceived social support and SSc patient health outcomes, like coping behaviours or medication adherence, should also be conducted [68]. Finally, research into chronic diseases has found that patients' experiences of social support and their tendencies towards helpseeking (or not) can vary depending on different socio-demographic characteristics, such as race/ethnicity [69] and gender [65]. Because of the small proportion of men diagnosed with SSc, as well as the small proportion of patients who identify as part of a racial or ethnic minority group, the perspectives and experiences of these individuals are often under-represented in research. Thus, conducting research into racial, ethnic, and gender differences in coping and social support would be important for understanding ways to support the entire SSc community.

Practical Implications

Helping SSc patients engage with social support while coping with the disease is one possible way of improving patient quality of life. Therefore, in addition to helping patients access community-based support tailored to their unique needs, which is an initiative currently pursued by the Scleroderma Patient-centered Intervention Network [48, 70], programs tailored to helping family members and friends of patients could also be beneficial. Research has investigated, for example, the benefits of family education programs, family support programs, and peer support programs for helping the social network of patients with common chronic diseases [20-22]. However, it is still unclear which strategies and behaviours community programs for family members should target (e.g., learning about supportive communication, helping patients with concrete goalsetting) [22]. Overall, having a better understanding of the influence of social support received by family and friends of SSc patients might provide an increased understanding of how to develop and tailor family-centered interventions to help patients cope.

Conclusions

Receiving social support while coping with a chronic disease like SSc is important for improving the quality of life and emotional well-being of patients. Patients with SSc experience numerous challenges related to their disease, and they often cope better when supported by close social relationships. However, lack of or non-adaptive social support can also serve as an impediment to coping depending on the individuals involved and the nature of their relationship. This study, affords a first glimpse into possible ways that social support can improve, as well as hinder, disease management and coping for patients with SSc. A deeper understanding of the role of close social relationships while coping is also important because coping does not happen in isolation. Community-based support programs intended to help patients learn tools to manage disease symptoms, navigate medical care, reduce emotional distress, and improve communication about the disease and personal needs should also include interventions aimed at fostering supportive relationships with close ones. The entire community of patients with SSc could benefit from new knowledge and skills to better navigate life post-diagnosis. A stronger social support network might prevent patients from having to manage their numerous disease-related challenges in isolation.

Declaration of Interest Statement

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References

- Institut National d'Excellence en Santé et Services Sociaux (INESSS). Prise en charge des maladies rares : Expériences étrangères. ETMIS. 2011;7(6):1-63.
- Kole A, Faurisson F. Rare disease social epidemiology: Analysis of inequalities. In: de la Paz MP, Groft SC, editors. Rare diseases epidemiology. New York (NY): Springer Science & Business Media; 2010; p. 223-50.
- [3] Canadian Organization for Rare Disorders. Now is the time: A strategy for rare diseases is a strategy for all canadians. Toronto (ON): CORD; 2015.
- [4] Bandura A. Health promotion from the perspective of social cognitive theory. Psychol Health 1998;13(4):623-49.
- [5] Bandura A. Exercise of human agency through collective efficacy. Curr Dir Psychol. Sci 2000;9(3):75-8.
- [6] Mayes M. Systemic sclerosis: A. clinical features. In: Klippel JH, Stone JH, Crofford LJ, White PH, editors. Primer on the rheumatic diseases. 13th ed. New York (NY): Springer and Arthritis Foundation; 2008; p. 343-50.
- [7] What is scleroderma? [Internet]. Danvers (MA): Scleroderma Foundation; c2017
 [cited 2017 Oct 19]. Available from: http://www.scleroderma.org/site/PageServer?pagename=patients_whatis#.VfB6dLQ 77G4.
- [8] Mayes M. Scleroderma epidemiology. Rheum Dis Clin North Am. 2003;29(2):239-54.

- [9] Domsic RT, Medsger Jr TA. Disease subsets in clinical practice. In: Varga J, Denton C, Wigley F, Allanore Y, Kuwana M, editors. Scleroderma: From pathogenesis to comprehensive management. 2nd ed. New York (NY): Springer; 2017; p. 39-48.
- [10] Sandqvist G, Åkesson A, Eklund M. Daily occupations and well-being in women with limited cutaneous systemic sclerosis. Am J Occup Ther. 2005;59(4):390-7.
- [11] Oksel E, Gündüzoğlu NÇ. Investigation of life experiences of women with scleroderma. Sex Disab. 2014;32(1):15-21.
- [12] Cinar FI, Unver V, Yilmaz S, et al. Living with scleroderma: Patients' perspectives, a phenomenological study. Rheumatol Int. 2012;32(11):3573-9.
- [13] Joachim G, Acorn S. Life with a rare chronic disease: The scleroderma experience. J Adv Nurs. 2003;42(6):598-606.
- [14] Suarez-Almazor ME, Kallen MA, Roundtree AK, et al. Disease and symptom burden in systemic sclerosis: A patient perspective. J Rheumatol. 2007;34(8):1718-26.
- [15] Mendelson C, Poole JL. Become your own advocate: Advice from women living with scleroderma. Disabil Rehabil. 2007;29(19):1492-501.
- [16] Buch M, Seibold J. Systemic sclerosis: C. treatment and assessment. In: Klippel JH, Stone JH, Crofford LJ, White PH, editors. Primer on the rheumatic diseases. 13th ed. New York (NY): Springer and Arthritis Foundation; 2008; p. 359-62.
- [17] Kowal-Bielecka O, Landewe 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(5):620-8.

- [18] Gumuchian ST, Peláez S, Delisle VC, et al. Understanding coping strategies among people living with scleroderma: A focus group study. Disabil Rehabil. 2017 [Oct 20];[1-10]. DOI:10.1080/09638288.2017.1365954
- [19] Mouthon L, Alami S, Boisard A-S, et al. Patients' views and needs about systemic sclerosis and its management: A qualitative interview study. BMC Musculoskelet Disord. 2017 [May 30];[1-10]. DOI:10.1186/s12891-017-1603-4.
- [20] Fisher EB, Coufal MM, Parada H, et al. Peer support in health care and prevention: Cultural, organizational, and dissemination issues. Annu Rev Public Health. 2014;35:363-83.
- [21] Gallant MP, Spitze GD, Prohaska TR. Help or hindrance? how family and friends influence chronic illness self-management among older adults. Res Aging. 2007;29(5):375-409.
- [22] Rosland AM, Piette JD. Emerging models for mobilizing family support for chronic disease management: A structured review. Chronic Illn. 2010;6(1):7-21.
- [23] Holt-Lunstad J, Smith TB, Layton JB. Social relationships and mortality risk: A meta-analytic review. PLoS Med. 2010;7(7):e1000316.
- [24] Berkman LF, Glass T. Social integration, social networks, social support, and health.In: Berkman LF, Kawachi I, editors. Social epidemiology. New York (NY): Oxford University Press; 2000; p. 137-73.
- [25] Smith KP, Christakis NA. Social networks and health. Annu Rev Sociol. 2008;34:405-29.
- [26] Uchino BN. Social support and physical health: Understanding the health consequences of relationships. New Haven (CT): Yale University Press; 2004.

- [27] Cohen S, Gottlieb BH, Underwood LG. Social relationships and health. In: Cohen S, Underwood LG, Gottlieb BH, editors. Social support measurement and intervention: A guide for healh and social scientists. New York (NY): Oxford University Press; 2000; p. 3-28.
- [28] Heaney CA, Israel BA. Social networks and social support. In: Glanz K, Rimer BK, Viswanath K, editors. Health behavior and health education: Theory, research, and practice. 4th ed. San Francisco (CA): Jossey-Bass; 2008; p. 189-210.
- [29] Cobb S. Social support as a moderator of life stress. Psychosom Med. 1976;38(5):300-14.
- [30] Cantor MH, Mayer MJ. Factors in differential utilization of services by urban elderly. J Gerontol Soc Work. 1979;1(1):47-61.
- [31] Cantor MH. Neighbors and friends: An overlooked resource in the informal support system. Res Aging. 1979;1(4):434-63.
- [32] Chappell N, Blandford A. Informal and formal care: Exploring the complementarity. Ageing Soc. 1991;11(3):299-317.
- [33] Pinquart M, Sörensen S. Older adults' preferences for informal, formal, and mixed support for future care needs: A comparison of Germany and the United States. Int J Aging Hum Dev. 2002;54(4):291-314.

[34] Hesse BW, Nelson DE, Kreps GL, et al. Trust and sources of health information: The impact of the internet and its implications for health care providers: Findings from the first health information national trends survey. Arch Intern Med. 2005;165(22):2618-24.

- [35] Hall JA, Roter DL, Katz NR. Meta-analysis of correlates of provider behavior in medical encounters. Med Care. 1988;27(7):657-75.
- [36] Green K, Derlega VJ, Mathews A. Self-disclosure in personal relationships. In:
 Vangelisti AL, Perlman D, editors. Cambridge handbook of personal relationships.
 1st ed. Cambridge (UK): Cambridge University Press; 2006; p. 409-28.
- [37] Laurenceau JP, Kleinman BM. Intimacy in personal relationships. In: Vangelisti AL, Perlman D, editors. Cambridge handbook of personal relationships. 1st ed. Cambridge (UK): Cambridge University Press; 2006; p. 637-56.
- [38] Ell K. Social networks, social support and coping with serious illness: The family connection. Soc Sci Med. 1996;42(2):173-83.
- [39] Revenson TA. Social support and marital coping with chronic illness. Ann Behav Med. 1994;16(2):122-30.
- [40] Lakey B, Orehek E. Relational regulation theory: A new approach to explain the link between perceived social support and mental health. Psychol Rev. 2011;118(3):482.
- [41] Dunbar SB, Clark PC, Quinn C, et al. Family influences on heart failure self-care and outcomes. J Cardiovasc Nurs. 2008;23(3):258-65.
- [42] Manne SL, Zautra AJ. Coping with arthritis: Current status and critique. Arthritis Rheum. 1992;35(11):1273-80.
- [43] Cornwell B. Independence through social networks: Bridging potential among older women and men. J Gerontol B Psychol Sci Soc Sci. 2011;66(6):782-94.
- [44] Stamm TA, Mattsson M, Mihai C, 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(6):1074-9.

- [45] Cinar FI, Unver V, Cinar M, et al. Coping strategies for activities of daily living in women whose hands affected by systemic sclerosis. J Clin Nurs. 2014;23(11):1630-8.
- [46] Mendelson C, Poole JL, Allaire S. Experiencing work as a daily challenge: The case of scleroderma. Work. 2013;44(4):405-13.
- [47] Newton EG, Thombs BD, Groleau D. The experience of emotional distress among women with scleroderma. Qual Health Res. 2012;22(9):1195-206.
- [48] Thombs BD, Jewett LR, Assassi S, et al. New directions for patient-centred care in scleroderma: The scleroderma patient-centred intervention network (SPIN). Clin Exp Rheumatol. 2012;30(2 Suppl 71):S23-9.
- [49] Milette K, Thombs BD, Maiorino K, et al. Challenges and strategies for coping with scleroderma: Implications for a scleroderma-specific self-management program.Disabil Rehabil. 2018:1(35). Located at: https://mc.manuscriptcentral.com/dandr.
- [50] Barbour R. Focus groups. In: Bourgeault I, Dingwall R, de Vries R, editors. The SAGE handbook for qualitative methods in health research. Thousand Oaks (CA): Sage; 2010; p. 327-52.
- [51] Gumuchian ST, Peláez S, Delisle VC, et al. Exploring sources of emotional distress among people living with scleroderma: A focus group study. PLoS One. 2016;11(3):e0152419.
- [52] Barbour R. Doing focus groups. London (UK): Sage; 2008.
- [53] Gergen KJ. Relational being: Beyond self and community. New York (NY): Oxford University Press; 2009.
- [54] Barbour RS. Making sense of focus groups. Med Educ. 2005;39(7):742-50.

- [55] Guest G, MacQueen KM, Namey EE. Applied thematic analysis. Thousand Oaks (CA): Sage; 2012.
- [56] Friese S. Qualitative data analysis with atlas TI. 2nd ed. Thousand Oaks (CA): Sage;2014.
- [57] Braun V, Clarke V. Using thematic analysis in psychology. Qual Res Psychol. 2006;3(2):77-101.
- [58] Vaismoradi M, Jones J, Turunen H, et al. Theme development in qualitative content analysis and thematic analysis. J Nurs Educ Pract. 2016;6(5):100-10.
- [59] Morrow SL. Quality and trustworthiness in qualitative research in counseling psychology. J Couns Psychol. 2005;52(2):250-60.
- [60] Poland BD. Transcription quality. In: Gubrium JF, Holstein JA, editors. Handbook of interview research: Context and method. Thousand Oaks (CA): Sage; 2002; p. 628-49.
- [61] Thoits PA. Mechanisms linking social ties and support to physical and mental health. J Health Soc Behav. 2011;52(2):145-61.
- [62] Rosland A, Kieffer E, Israel B, et al. When is social support important? the association of family support and professional support with specific diabetes selfmanagement behaviors. J Gen Intern Med. 2008;23(12):1992-9.
- [63] Gallant MP. The influence of social support on chronic illness self-management: A review and directions for research. Health Educ Behav. 2003;30(2):170-95.
- [64] Vassilev I, Rogers A, Kennedy A, et al. The influence of social networks on selfmanagement support: A metasynthesis. BMC Public Health. 2014;14(1):719.

- [65] Kristofferzon M, Löfmark R, Carlsson M. Myocardial infarction: Gender differences in coping and social support. J Adv Nurs. 2003;44(4):360-74.
- [66] Kitzinger J. Qualitative research. introducing focus groups. BMJ. 1995;311(7000):299-302.
- [67] Carlsen B, Glenton C. What about N? A methodological study of sample-size reporting in focus group studies. BMC Med Res Methodol. 2011;11(1):26.
- [68] Uchino BN. Understanding the links between social support and physical health: A life-span perspective with emphasis on the separability of perceived and received support. Perspect Psychol Sci. 2009;4(3):236-55.
- [69] Rees CA, Karter AJ, Young BA. Race/ethnicity, social support, and associations with diabetes self-care and clinical outcomes in NHANES. Diabetes Educ. 2010;36(3):435-45.
- [70] Kwakkenbos L, Jewett LR, Baron M, 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(8):e003563.

Figure 1. Participant perspectives on types of social support received from close social relationships and relational factors impacting support

SCLERODERMA AND CLOSE SOCIAL RELATIONSHIPS

Variables	
Age, mean (standard deviation)	57.1 (10.9)
Female gender, <i>n</i> (%)	17 (89.5)
Race/ethnicity, n (%)	
White	12 (63)
African American or Black	2 (10.5)
Asian	2 (10.5)
Aboriginal	2 (10.5)
Not specified	1 (5)
Level of Education, n (%)	
High school graduate	4 (21)
Some college/university	6 (32)
College/university degree	6 (32)
Postgraduate degree	3 (16)
Occupational status*, n (%)	
Homemaker	1 (5)
Retired	8 (42)
On disability	9 (47)
On leave of absence	1 (5)
Full-time employed	3 (16)
Part-time employed	1 (5)
Scleroderma subtype, n (%)	

Table 1. Socio-demographic characteristics of patients with scleroderma (N=19)

5)

*Total n (%) is greater than 19 (100%)

because some patients reported more than one occupational status