Psychosocial aspects of scleroderma

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SYNOPSIS
Patients with systemic sclerosis (SSc; scleroderma) have to cope with not only the physical impacts of the disease, but also the emotional and social consequences of living with the condition. Because there is no cure for SSc, improving quality of life is a primary focus of treatment, and an important clinical challenge. The present review summarizes significant problems faced by patients with SSc, including depression, anxiety, fatigue, sleep disruption, pain, pruritus, body image dissatisfaction, and sexual dysfunction, and describes options to help patients cope with the consequences of the disease.

KEY POINTS BOX
- Symptoms of SSc, including fatigue, pain, pruritus, sleep problems, and sexual impairments negatively influence quality of life in many patients, and may lead to emotional consequences such as depression, anxiety, and body image distress due to appearance changes.
- Providing accessible information to SSc patients regarding problems common to people living with the disease, as well as information regarding useful resources and services to address these problems, can help SSc patients and can be easily implemented by health care professionals.
- In addition to referrals for professional health care interventions, low-intensity strategies such as self-management programs and support groups, may be helpful to some patients with SSc.
• Future research should focus on the development and testing of interventions designed specifically to meet the educational and psychosocial needs of patients with SSc.
INTRODUCTION

Systemic sclerosis (SSc, or scleroderma) has far-reaching consequences for physical health, as well as emotional and social well-being.\textsuperscript{1–4} Because there is no known cure for the disease, SSc treatment focuses on reducing symptoms and disability, and improving health-related quality of life (HRQL). The objectives of the present review are to summarize the impact of SSc on common patient-reported problems associated with HRQL and to describe potential interventions to support coping with the consequences of the disease.

Depression

Depression involves symptoms that may include sadness, loss of interest or pleasure, feelings of guilt or low self-esteem, poor concentration, and disturbed sleep or appetite. A study of 345 SSc patients enrolled in a Canadian registry reported the prevalence of major depressive disorder (MDD) was of 4% for the past 30-days, 11% for the past 12-months, and 23% for lifetime.\textsuperscript{5} A French study of 50 hospitalized SSc patients and 50 SSc patients who attended a patient organization meeting found that 19% had current MDD and 56% had lifetime MDD, and rates were higher in hospitalized (28% current) versus non-hospitalized (10% current) patients.\textsuperscript{6} Depression is substantially more common in SSc than in the general population and may be more prevalent than in other rheumatic diseases.\textsuperscript{5} Many patients with SSc and other chronic diseases who meet criteria for MDD at a given time point, however, may not meet criteria consistently. In the Canadian sample, for instance, only 3 of 12 patients with SSc who had a current major depressive episode at baseline met diagnostic criteria one month later.\textsuperscript{7} Some episodes may be time-limited and may resolve without targeted intervention or treatment. Others may reflect ongoing moderate symptoms that only variably meet criteria for formal diagnosis. Many SSc patients describe ongoing emotional distress from the burden of living with the disease, but
differentiate this qualitatively from what they would consider clinical depression.\textsuperscript{8} Cross-sectionally, factors associated with symptoms of depression in SSc include greater overall disease burden, which may involve degree of gastrointestinal involvement, breathing problems, skin involvement, and tender joints.\textsuperscript{9-11}

**Anxiety and Fear**

Anxiety can be a normal reaction to stress; however, it may also lead to mental health problems when experienced in excess. To date, only one study documented the prevalence of anxiety disorders among SSc patients. In that study, 49\% of 50 hospitalized patients and 50 patients who attended a patient meeting had at least one current anxiety disorder, and 64\% met criteria for at least one anxiety disorder in their lifetime. Social anxiety and generalized anxiety disorder were the most common.\textsuperscript{6} There was no difference in prevalence between hospitalized and non-hospitalized patients.

The course of SSc is highly unpredictable and patients may perceive the future as uncertain. For patients, worry about the future, including fear of disease progression, fear of becoming physically disabled, and fear of being dependent upon others are important sources of stress.\textsuperscript{12,13} Because SSc is unpredictable and associated with serious consequences, these concerns are realistic and in themselves do not represent anxiety disorders, for which irrational fear is typically a central component.\textsuperscript{13} Nonetheless, fear of progression can impact HRQL substantially. In a cross-sectional study of 215 SSc patients from the Netherlands, for instance, fear of progression was highly associated with symptoms of depression.\textsuperscript{13}

**Fatigue**
Fatigue from a chronic medical disease is characterized by persistent exhaustion that is disproportionate to exertion and not relieved by rest.\textsuperscript{14} Fatigue is the most commonly experienced symptom of SSc and has a substantial impact on HRQL, as well as the ability to carry out daily activities, including work.\textsuperscript{1,4,15-17} In one Canadian study, 89\% of 464 SSc patients reported fatigue at least some of the time, and 81\% of these patients indicated that fatigue had at least a moderate impact on their daily function.\textsuperscript{15} Levels of fatigue in SSc are similar to those experienced by patients with other rheumatic diseases and cancer patients undergoing active treatment.\textsuperscript{18} Cross-sectionally, greater fatigue in SSc is associated with increased medical comorbidities, current smoking, pain, breathing problems, and gastrointestinal symptoms.\textsuperscript{14} Longitudinally, fatigue severity has been associated with pain, severity of gastrointestinal involvement, and psychological variables, specifically ineffective coping skills.\textsuperscript{19}

**Sleep**

Significant sleep disruption is common in SSc and has broad implications for patients.\textsuperscript{15,20-22} A polysomnography study of 27 SSc patients found that, compared to age-adjusted norms, SSc patients had reduced sleep efficiency and rapid eye movement sleep, as well as increased arousal and slow wave sleep.\textsuperscript{20} Sleep disruption was associated with esophageal dyskinesia and dyspnea, which are common complications of SSc, as well as restless legs syndrome.\textsuperscript{20} Observational studies have linked dyspnea, pain, fatigue, pruritus, gastrointestinal symptoms, and depressive symptoms with self-reported poor sleep quality and sleep disruption in SSc.\textsuperscript{21,22}

**Pain**

Between 60 and 83\% of SSc patients report experiencing pain at any given time, and pain levels in SSc are similar to levels reported in chronic pain and rheumatic conditions.\textsuperscript{15,23-24} Pain in
SSc is associated with reduced HRQL, functional disability, work disability, sleep problems, and symptoms of depression.\textsuperscript{22-25} Patients with SSc describe their pain as both localized and generalized in quality,\textsuperscript{26} and sources can include pain from Raynaud’s phenomenon, gastrointestinal pain, joint and musculoskeletal pain, skin pain, and pain due to calcinosis and ulcers.\textsuperscript{1,23,26} Pain ratings are higher among patients with diffuse SSc compared to patients with limited disease, although this difference is generally small.\textsuperscript{23} Based on patient report, overall pain levels are associated with sleep problems, fatigue, and symptoms of depression, as well as physical function, reduced ability to carry out daily activities, work disability, and poorer HRQL.\textsuperscript{22,23,25,27} How patients with SSc describe their pain and the degree to which they believe they can manage it often reflects psychosocial factors, which should be considered in assessment and intervention, especially as related to pharmacologic treatment.\textsuperscript{28}

**Pruritus**

Pruritus, or itch, is common in SSc and is associated with HRQL, even after controlling for sociodemographic and other SSc symptom variables.\textsuperscript{29,30} Overall, 43\% of 959 SSc patients from a Canadian registry reported pruritus on most days in the last month.\textsuperscript{31} This rate was slightly higher, but not statistically significant, among patients with early SSc (< 5 years since onset of non-Raynaud’s symptoms; 46\%) versus those with longer disease duration (≥ 5 years; 41\%).\textsuperscript{31} The presence of pruritus is more common among patients with greater skin and gastrointestinal involvement.\textsuperscript{31}

**Body Image**

Acquired disfigurement from an injury or medical illness is often linked to problems with body image, including social avoidance.\textsuperscript{32} Appearance changes in highly visible areas of the body,
particularly the face and hands, are common in patients with SSc and contribute to body image distress, which in turn can be associated with symptoms of anxiety and depression.\textsuperscript{1,4,32-36} A number of cross-sectional studies have reported that appearance changes of the face, including changes to the mouth, as well as hand involvement, including skin thickening, have been consistently related to body image distress, including dissatisfaction with appearance, decreased appearance self-esteem, and symptoms of anxiety and depression.\textsuperscript{32-36} Other appearance changes, including telangiectasias, may also be associated with body image distress.\textsuperscript{36,37} Social discomfort due to changes in appearance is also related to age.\textsuperscript{36} Younger patients, for whom the importance of meeting new people and developing intimate relationships is more pronounced, may experience a greater negative impact of appearance changes on social relationships.\textsuperscript{36}

**Sexual Function**

Sexual dysfunction is a common problem among women with SSc.\textsuperscript{1,38-43} Compared to women in the general population, women with SSc are significantly less likely to be sexually active, and sexually active women with SSc are significantly more likely to be sexually impaired.\textsuperscript{39,40,42} Factors that are independently associated with being sexually active include younger age, fewer gastrointestinal symptoms, and less severe Raynaud’s phenomenon symptoms.\textsuperscript{41} Among women who are sexually active, sexual impairment is associated with older age, as well as more severe skin involvement and breathing problems. Vaginal pain is eight times as common among women with impairment compared to those without.\textsuperscript{41}

Among men with SSc, erectile dysfunction (ED) is common with onset typically occurring several years after the manifestation of the first non-Raynaud’s symptoms.\textsuperscript{44-46} While in the general population, ED is typically associated with atherosclerosis, in SSc, penile blood flow is impaired due to both myointimal proliferation of small arteries and corporal fibrosis.\textsuperscript{46} Men with
SSc who have ED are significantly more likely to be older than those without ED and tend to report non-SSc risk factors (e.g., alcohol consumption) at higher rates.\textsuperscript{45,46} SSc factors associated with ED include severe cutaneous, muscular, or renal involvement, diffuse disease, elevated pulmonary pressures, restrictive lung disease, endothelial dysfunction, and microvascular damage.\textsuperscript{45-47} Most men with SSc who have ED do not receive treatment.\textsuperscript{45} Among those who do, sildenafil appears to be commonly used, but its efficacy has not been established in SSc.\textsuperscript{44,45}

**CLINICAL MANAGEMENT**

In addition to the core medical treatment of SSc-related symptoms, providing services and interventions to help manage the psychological, behavioral, and social aspects of living with the disease is an important component of patient-centered care. There are challenges related to the development, testing, and delivery of such patient-centered interventions in SSc, however, including the small number of patients and limited resources available.\textsuperscript{2,3} Nonetheless, these types of psychosocial interventions have proven to be effective in reducing disability and improving HRQL in more common conditions, including rheumatic diseases,\textsuperscript{48,49} and can be reasonably implemented in SSc.

Stepped-care models for psychological, behavioral, and educational interventions involve matching interventions of differing intensities to patient needs. Generally, stepped-care starts off with the simplest, least intrusive intervention and proceeds to more intense treatment approaches as deemed necessary.\textsuperscript{1} Self-help can be a very useful first step towards addressing relatively mild problems associated with psychosocial functioning and HRQL. Providing accessible information to both patients and those who support them regarding issues common to people living with SSc, as well as information regarding useful resources and services to address such problems, is something that can be easily implemented by health care professionals.
should be aware of the important concerns that affect HRQL in order to help patients access appropriate resources and facilitate conversations that address concerns of individual patients. Additionally, links to other sources of information can be provided in clinics regarding self-help programs that are available as a first step in providing psychosocial support.

In the context of stepped care, low-intensity interventions are appropriate for people with less severe psychological concerns. However, referral to professional services is needed for patients with more complex or serious psychological problems such as severe depression, or when lower intensity methods do not work well. For these patients, a focused evaluation of their psychological symptoms and more intense intervention services provided by a mental health professional are often required.

Low-intensity strategies that have been proven effective to improve coping with the consequences of more common chronic diseases, and that may be readily available to many patients with SSc, include self-management programs and support groups.48,49

**Self-management**

Across more prevalent diseases, supportive care programs, such as self-management programs, are increasingly included as core components of patient-centered care. Although there is currently no gold-standard definition, the term “self-management” has been defined as the ability of patients to manage the symptoms, treatment, physical, and psychological consequences, and lifestyle changes inherent in living with a chronic condition. Effective self-management involves the ability to monitor one’s condition and to have the cognitive, behavioral, and emotional responses necessary to maintain a satisfactory HRQL.50

In more prevalent diseases, such as arthritis, asthma, and diabetes, self-management interventions have been shown to provide benefits to participants in terms of knowledge,
performance of self-management behaviors, self-efficacy, and health status.\textsuperscript{50} A Cochrane systematic review (17 trials, \(N = 7,442\))\textsuperscript{49} found that low-cost self-management programs lead to small improvements in participants’ self-efficacy to manage their disease, which is the principal target of the programs, as well as improvements in self-rated health status (e.g., pain, fatigue, disability) and some health behaviors (e.g., exercise, cognitive symptom management). No randomized controlled trials (RCTs), however, have investigated the efficacy of self-management interventions for SSc. Two pre-post intervention studies of SSc self-management programs have been conducted.\textsuperscript{51,52} One described a mail-delivered self-management program provided to 49 patients with SSc,\textsuperscript{51} and the other was a pilot study of an internet-delivered self-management program with 16 SSc patients.\textsuperscript{52} The small sample sizes in these studies, however, limit the ability to draw conclusions about effectiveness. In addition, the Scleroderma Patient-centered Intervention Network (SPIN) is developing an online self-management program.\textsuperscript{2,3} However, there are currently no SSc-specific programs readily available to patients. An alternative is a general disease self-management program, such as the Chronic Disease Self-Management Program from Stanford University, which is available via the Internet. The Chronic Disease Self-Management Program, which is designed to teach self-care techniques useful to persons with many chronic diseases, improved self-efficacy for disease management and overall health status in an RCT that included patients with heart and lung disease, as well as diabetes.\textsuperscript{53}

**Support Groups**

A large number of patients with chronic medical illnesses, including those with SSc, join support groups in order to better cope with and manage their illness.\textsuperscript{54} Activities of support groups include giving and receiving emotional and practical support, as well as providing education and information to patients. The specific activities and focus, as well as facilitator training and
competence, may vary across support groups, which are typically organized locally. Patients may differ in the acceptability of the idea of attending a support group and the degree to which they may benefit from one. Due to their grass-roots nature, support groups can be configured in a variety of ways.\textsuperscript{55} For example, some support groups may meet face-to-face whereas others may “meet” online; some groups may be facilitated by a peer whereas others may be facilitated by a professional; and some may include structured educational activities whereas others may not.

Research on the effectiveness of support groups is scant, particularly with regard to lay-led groups. However, many people who do attend support groups describe feeling more empowered, more hopeful, and less alone following their group experience.\textsuperscript{54} In addition, some patients who attend these groups report feeling more in control of their life, as well as more knowledgeable about their illness, coping strategies, and developments in medical and self-help treatments.

Peer-led support groups have become increasingly popular in recent years.\textsuperscript{56} Consistent with this, most SSc support groups are peer-led rather than professionally-led. No studies, however, have examined the effectiveness of these groups on psychosocial or other coping-related outcomes. Given that they may be the sole source of SSc-specific support available to many patients and that they have been effective in other conditions, attending support groups may be beneficial for some patients with SSc. It is important to keep in mind, however, that support groups are meant to complement rather than supplement standard medical care. Medical professionals may want to discuss the possible benefits of attending a support group with their patients, as well as potential pitfalls. For patients who are interested in attending or joining a SSc support group, information can typically be found on local or national organization websites, such as the Scleroderma Society of Canada and the Scleroderma Foundation in the United States.\textsuperscript{57,58}
Attention and research regarding the psychological aspects of living with SSc have increased in recent years.\textsuperscript{1,4} Furthermore, as the low prevalence of SSc sometimes precludes high-quality research in this area, the substantial progress that has been made in establishing international research networks is promising and provides a solid foundation for future research endeavors.\textsuperscript{2,3} Future research should focus on gaining a better understanding of the psychosocial aspects of SSc, their interconnectedness, and the valid measurement of outcomes related to this, as well as on moving forward the development and testing of interventions that help patients with SSc cope with the everyday challenges related to living with their disease.

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