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Review

A review on quality of life in systemic sclerosis

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ABSTRACT

Systemic sclerosis (SSc) is a chronic multi-system autoimmune disease associated with disability and reduced quality of life. There is no effective treatment or cure to SSc, so it is important improve global health of these patients and reduce morbidity and mortality associated with SSc. It was made a literature review about quality of life in patients with SSc, regarding the several factors that should be considered and evaluated when attending to SSc patients. It was also considered the validated scales and questionnaires used to measure outcomes in patients with SSc. We concluded that it is important to have an interdisciplinary approach to SSc patients considering the patient's cognitive representations of the disease and what they value most like mobility and hand function, pain, fatigue, sleep, depression and body image.

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1. Introduction

Systemic sclerosis (SSc) is a chronic multi-system autoimmune disease with extremely heterogeneous manifestations, characterized by thickening and fibrosis of the skin, vasculopathy and involvement of various internal organs in different degrees. Patients with SSc report a number of problems associated with disability and reduced quality of life (QoL) [1–3], which in turn is an important factor for disease outcomes [4–6]. Also the presence of polyautoimmunity in SSc, a common condition affecting one quarter of the patients, could influence both disease phenotype and severity. These patients seem to have a milder disease [7].

There is no effective treatment or cure to SSc [8–10], so a primary goal of care is to reduce symptoms and disability and to improve health-related quality of life (HR-QoL), with both pharmacological and non-pharmacological interventions [8,11].

Patients with rare diseases and especially with SSc face unique challenges that are not covered by generic interventions or interventions developed for other chronic diseases [8,12], so to improve global health of these patients and reduce morbidity and mortality associated with SSc, it is important to measure outcomes with validated scales and questionnaires.

The authors decided to make a literature review about quality of life in patients with SSc.

2. Material and methods

A literature search was performed on October 1, 2014 using PubMed database with the keywords quality of life in conjunction with scleroderma or with systemic sclerosis. The results of the search included a total of 599 articles. After duplicates removal remained 337 articles selected. The abstracts of all original and review articles in English were read. Those thought to be relevant with full text available and pertinent secondary references were fully examined, and 117 articles were included.

3. Features related to quality of life in SSc patients

Knowledge about the frequency and perceived impact of the range of problems faced by individuals living with SSc, is limited [1,11]. It is important to know that there is a great difference between the assessment of the disease severity and HR-QoL [4,13]. The patient's cognitive representations of the disease are the most important determinants of physical and mental health. A recent study suggested that the fear of clinical consequences and the tendency to attribute each physical complaint to SSc are major contributors to the physical health, while the emotional responses to personal representation of the disease contribute to mental health [13,14]. The “Canadian Scleroderma Patient Survey of Health Concerns and Research Priorities” suggests that patients with SSc are more unsatisfied with healthcare than other chronically ill patients [2], because SSc involves more visible physical disfigurement that tends to worse over time, contributing to high psychological morbidity (more depressive symptoms and anxiety) [15], regular use of healthcare and related increased costs. Physicians may ignore or use unreliable measurements to evaluate psychological distress [16]. While physicians may prioritize objective indicators of disease status, patients may perceive other aspects of their disease experience as more debilitating or distressing [1,17,18], like limitations in mobility and hand function, pain, fatigue, sleep disturbance, depression, sexual dysfunction and body image distress from disfiguring changes in appearance (pigment changes, hand contractures and facial telangiectasias for example) [8].

Van Lankveld et al. [15] reported that fatigue, functional limitations, skin deformities, pain and disfigurement were the most annoying symptoms, whereas Suarez Almazor et al. [18] identified physical pain, coping skills, social aspects of living with the disease, physical

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