Universidade do Minho
Escola de Psicologia

Catarina da Silva Correia Pereira Leite

Psychosocial Characterization, Symptoms and Illness Perception in Scleroderma Patients: an international study

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Dissertação de Mestrado
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Professora Doutora Ângela da Costa Maia

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Psychosocial Characterization, Symptoms and Illness Perception in Scleroderma Patients: an international study

Abstract

Introduction: Scleroderma, also known as systemic sclerosis, is a rare, complex and autoimmune disease, with unknown origin and cause (Coelho, Oliveira, & Kroon, 2008). It affects the connective tissue and is characterized by vascular dysfunction, inflammatory and fibrotic of multiple organ systems (Nadashkevich, Davis, &Fritzer, 2004). The symptoms that suggest the existence of scleroderma include: Raynaud phenomenon, skin thickening, calcinosis and telangiectasia (Li, Sahhar, & Littlejohn, 2008). In addition to the swelling of the fingers and hands that may hinder mobility and become a very painful phenomenon (Li et al, 2008), the alterations in internal organs results in significant morbidity and mortality (Assassi, del Junco, Sutter, McNearney, Reveille, Karvanas, …,Gourh, 2009). Despite its severity, the disease remains unknown to many health professionals and may take several years before those affected are diagnosed. This disease is associated with pain, fatigue and progressive disability. Among the physical changes with more psychological impact are the changes in the skin (Malcarne, Hansdottir, Greenbergs, Clements & Weisman, 1999). Due to the disfigurement and physical changes that are typical of the disease, psychological morbidity is high and includes depression, body image problems.

Method: The participants in this study are 563 scleroderma patients from 12 European countries and Brazil. These participants are mostly women. The instruments used in this study were the “Canadian Scleroderma Patient Survey of Health Concerns and Research Priorities” and “The Brief Illness Perception Questionnaire (Brief IPQ)”.

Results: The most common symptoms and with more impact on the lives of the participants were joint pain, fatigue and Raynaud’s. Just under one fifth of the participants had a diagnosis of scleroderma with the first doctor consulted. Participants with more years of diagnosis revealed to be more satisfied with the medical care they receive. Participants reported a high degree of concern with scleroderma and 84% reported concerns with body image due scleroderma. Depression, anxiety and social phobia are common among scleroderma patients.

Conclusion: Due to gravity, lack of knowledge, unpredictability and disfigurement of scleroderma, the existence of psychological disorders in these patients is common.

Contents

Theoretical review

Introduction

Chronic diseases are the greatest cause of morbidity and mortality, and their diagnosis is, in most cases, a period of uncertainty, doubt and general crisis. Besides the physical well being, the patient also suffers psychological, social and functional changes which, as several studies show, present an increased possibility of anxiety and depression (Beutel & Schulz, 2011).

Scleroderma

Scleroderma is an autoimmune, connective tissue disease characterized by fibrosis, degenerative changes and vascular lesions in the skin, joints, and multiple internal organs (Nadashkevich et al, 2004). The disease may impair the proper functioning of respiratory, gastrointestinal, and cardiac systems and renal function, which leads to a significant incidence of morbidity and mortality (Li et al, 2008). Scleroderma is also characterized fibroblast activation, collagen overproduction, and pathological tissue fibrosis (Varga, 2008). The diagnosis, prognosis and treatment can be very difficult (Ruzek, Jha, Ledbetter, Richards & Garman, 2004). Scleroderma is a heterogeneous, but serious, disease of unknown cause (Coelho et al, 2008), where lung abnormalities are the leading cause of death (Gupta& Thabah, 2007). The two primary lung manifestations disease are interstitial lung disease and pulmonary vascular disease (Roth, Tseng, Clements, Furst, Tashkin, Goldin, … Elashoff, 2011). Patients with respiratory symptoms can have chronic aspiration, airway disease, neuromuscular weakness, extrinsic restriction secondary to a hide-bound chest, pleural effusions and lung cancer, and all of this cause clinically significant disease (Le Pavec, Launay, Mathai, Hassoun & Humbert, 2011). The organ system most frequently affected with scleroderma is the gastrointestinal tract (Plastiras, Tzivras & Vlachoyiannopoulos, 2007). The esophageal muscle weakness and incoordination can impair intake of solid aliments. With the malfunctioning of the lower esophageal sphincter esophageal reflux occurs what causes heartburn, a very common symptom in people with scleroderma. The commitment of the small intestine can be affected causing pain, hypomobility intestinal bloating, diarrhea, malabsorption and weight loss. The large intestine affectation can cause constipation (Christmann & Silver, 2010). Gastrointestinal involvement can be also debilitating and even life threatening (Plastiras et al, 2007). Cardiac involvement is common in scleroderma; the diagnosis is not always clear or may be associated with left ventricular diastolic dysfunction. The commitment of the heart may be due to cardiac fibrosis

and not just pulmonary arterial hypertension (Varga, 2008). The renal crisis in scleroderma may be caused by lesions in the arteries of medium size. These injuries can also causehyperreninemia with malignant hypertension, microangiopathic hemolytic anemia, and rapidly progressive renal failure (Varga, 2008). 15% of people suffer from scleroderma renal crisis; such crises develop mainly in the first 4 years of diagnosis (Varga, 2008).

The first symptoms of scleroderma are usually the appearance of Raynaud's phenomenon and skin changes typical of the disease (Vilella, Aznar & Julià, 2010). These usually occur on the hands and may also affect the arms, face, trunk, and legs.

Scleroderma means hard skin. With scleroderma skin loses elasticity and becomes harder. After initial hardening, the dermis is left with excess collagen and thus, beyond getting hard, it is also thicker (Kissin, Schiller, Gelbard, Anderson, Falanga, Simms, … Merkel, 2006). The greater involvement of the skin, the less is the chances of survival (Clements, Lachenbruch, Simmons, Sterz,& Furst, 1990). Thus, skin thickening is the hallmark of scleroderma, and its severity and extent is a way to differentiate the kind and severity of scleroderma (Steen & Medsger, 2001). The first skin changes are characterized by skin thickening and swelling. After this phase, the skin becomes firm and connected to the subcutaneous tissue, which may cause flexion contractures, limiting movement. Finally, the skin becomes thinner, which facilitates the appearance of ulcers and infections. In the face, lips become thinner; there is a reduction in the amplitude of mouth opening, loss of natural skin wrinkles and facial expression lines (Li et al, 2008). The reduction of mouth opening calls microstomia, this problem can reduce quality of life, because interferes with eating, brushing teeth and dental care (Pizzo, Scardina,& Messina, 2003).

Raynaud’s phenomenon may be described by digital artery vasospasm. These episodic clinical attacks occur mainly in the fingers (Brown, Middaugh, Haythornthwaite,& Bielory, 2001). The Raynaud's phenomenon is present in more than 95% of patients with scleroderma and usually precedes the appearance of scleroderma for several years (Merkel, Herlyn, Martin, Anderson, Mayes, Bell & Wigley, 2002). Thus Raynaud's phenomenon offers the best window for research of first steps in the pathogenesis of scleroderma (Kahaleh, 2004). This phenomenon is characterized by episodes of sudden, transient and recurrent changes in the fingers, caused by exposure to cold or emotional stress (Hinchcliff & Varga, 2008). Thus, the fingers turn white (vasospasm), then moving to a blue-violet hue (ischemia) and finally turn red (hyperemia) (Hinchcliff& Varga, 2008). Normally we can observe painful ulcers that can be very limiting for most of the movements and may be necessary to amputate part of fingers. Raynaud's phenomenon has a great impact in quality of life and functioning of patients with scleroderma, this impact varies with the frequency, duration and severity of attacks, with the presence and activity of digital ulcers, and with other problems

related with scleroderma such as hardening skin, pain, shortness of breath and functional disability (Merkel et al, 2002). Digital ulcers are necrotic lesions at the fingertips. 50% of people with diffuse or limited scleroderma, have digital ulcers (Chung& Fiorentino, 2006).

Erectile dysfunction in men with scleroderma is common. Several studies suggest that erectile dysfunction associated with scleroderma is due to the damage of blood vessels and fibrotic changes (Ostojic& Damjanov, 2007). In turn, women with scleroderma are more likely to suffer from dyspareunia, vaginal dryness and vaginal ulcers compared with women with other rheumatic diseases (Knafo, Jewett, Bassel & Thombs, 2010). Despite these physical changes, according Knafo et al (2010) is necessary to determine which psychosocial characteristics of the disease can contribute to sexual dysfunction.

In addition to other symptoms, patients often suffer from a wide range of musculo-skeletal disorders such as joint pain, arthritis, flexion contractures of the joints and muscle weakness (Ostojic, Zivojinovic, Reza & Damjanov, 2010). In some patients with scleroderma the skeletal mucles alterations may range from mild to severe, few patients develop a debilitating or even life-threatening course (Olsen, King & Park, 1996).

Types of Scleroderma

Scleroderma is divided into three types: limited, diffuse and systemic sclerosis *sine scleroderma*.The most common types of Scleroderma are limited scleroderma and diffuse scleroderma (Hinchcliff& Varga, 2008). These types vary in the extension of damaged skin, and in relation to the organs affected (Li et al, 2008).

The most common is limited scleroderma, which affects approximately 60% of people with scleroderma. CREST syndrome is a variant of this type of scleroderma(Hinchcliff& Varga, 2008). The meaning of the acronym CREST: calcinosis, Raynaud's phenomenon, esophageal dysmotility, sclerodactyly (tension in the skin of the fingers, which can hinder or even prevent the mobility of the fingers and create contractures) and telangiectasia (dilation of blood vessels in the outer layer of skin) (Li et al, 2008).

The diffuse scleroderma affects about 35% of people with scleroderma (Hinchcliff& Varga, 2008). People with this type are more likely to develop pulmonary interstitial disease and heart or kidney disease in an early stage of the disease (Li et al, 2008). Finally, systemic sclerosis sine scleroderma is the least common, affecting only 5% of people with this disease and manifests itself unaccompanied by changes in the skin, including skin thickening (Hinchcliff& Varga, 2008). It is considered that a patient has systemic sclerosis sine scleroderma if he or she has all of these assumptions: Raynaud’s, positive ANA (anti-nuclear

antibody) and one of the following: distal esophageal hypomotility, small bowel hypomotility, pulmonary interstitial fibrosis, primary pulmonary arterial hypertension, cardiac involvement or renal crisis (Poormoghim, Lucas, Fertig & Medsger, 2000).

The localized forms of the disease are morphea and linear scleroderma. Limited scleroderma or morphea is characterized by a thickening of the skin with increased collagen deposition (Zulian, Athreya, Laxer, Nelson, de Oliveira, Punaro, … Harper, 2006). Linear scleroderma is characterized by scleroatrophic lesions that affects asymmetrically arms and legs (Grosso, Fioravante, Biasi, Conversano, Marcolongo, Morgese & Balestri, 2003).Linear scleroderma is the most common type of localized scleroderma in children and adolescents and is characterized by the appearance of one or more strips, usually on the upper or lower limbs, and may be associated with plaques of morphea. When one of these linear lesions affect the face or scalp is called scleroderma *en coup de sabre*, a subtype of linear scleroderma (Kasapçopur, Ozkan & Tuysuz, 2003). The different form of localized scleroderma affects mainly children and is not associated with Raynaud's phenomenon or significant visceral manifestations (Hinchcliff& Varga, 2008).

Prevalence

The prevalence of this disease is 30 per 100,000 people (Chifflot, Fautrel, Sordet, Chatelus & Sibilia, 2008), with a proportion of four women for every man affected, especially over 40 years old. In Portugal, there is no actual data about the number of people diagnosed with scleroderma, but it is estimated that there are over 3,000 people, attending the studies of scleroderma’s prevalence.

Impact of Scleroderma

Scleroderma has a physical, psychological and socialimpact.

Physical impact

Fatigue is considered one of the symptoms of greater impact among patients (Taillefer, Bernstein, Schieir, Buzza, Hudson, Scleroderma Society of Canada, … Thombs, 2010), independently of the scleroderma type (Morell-Dubois, Condette-Wojtasik, Clerson, Berezné, Launay, Lambert, ... Hachulla, 2011).In terms of physical impact, fatigue is often considered by people with chronic diseases such as the symptom that most affect their quality of life (Swain, 2000), being a symptom that includes complex interactions between biological, psychosocial and behavioral processes. According to Swain (2000), persistent fatigue in chronic disease involves continuing tiredness disproportionate to the effort made and that is not relieved by rest.It is a common and incapacitating symptom related with precarious sleeping quality, greater pain, depressive symptoms and a deficit in physical and social functioning (Sandusky, McGuire, Smith, Wigley & Haythornthwaite, 2009). Fatigue is defined as the experience of feeling weak, tired and lacking energy. In scleroderma, this is similar to that of patients who are receiving treatment for cancer (Thombs, Fuss, Hudson, Schieir, Taillefer, Fogel, … Canadian Scleroderma Research Group, 2008).

Experiencing difficulties when trying to sleep is also one of the symptoms that affect patients more severely (Taillefer et al, 2010). Patients' sleep can be interrupted by gastroesophageal reflux disease, pulmonary fibrosis with dyspnea and the restless legs symptom (Abad, Sarinas & Guilleminault, 2008), which reduces sleep efficiency.

Pain is common among patients with scleroderma (Benrud-Larson, Haythornthwaite, Heinberg, Boling, Reed, White & Wigley, 2002). Painful symptoms are associated with the frequency of Raynaud's phenomenon, digital ulcers, joint pain and gastrointestinal symptoms (Morell-Dubois et al, 2011). Digital Ulcers are extremely painful and cause significant functional disability (Chung& Fiorentino, 2006). The pain is crucial in physical functioning and social adaptation, both relevant factors in quality of life (Benrud-Larson et al, 2002).

Psychological impact

Psychological factors with confirmed relevance to scleroderma include pain, depression, and distress about disfigurement, and physical and social function (Haythornthwaite, Heinberg & McGuire, 2003).

Scleroderma affects body image and is associated with depressive and anxiety symptoms and sexual dysfunction. Many rheumatic diseases (scleroderma is part of this group) involves visible physical changes associated with disease progression (Malcarne et

al, 1999). However, while in other types of diseases the changes remains, in scleroderma these alters over time and tend to worsen (Taillefer et al, 2010), existing facial disfigurement and usually hands disfigurement.

These physical disfiguring changes are in visible and important interpersonal contact body parts. From the beginning, scleroderma causes significant physical changes, such as swelling of fingers, loss of natural skin wrinkles, the change on the skin for brighter, hypo-or hyper-pigmentation of the skin, facial changes at as the loss of flexibility in the lips, loss of ability to fully open the mouth, difficulty in completely close the lips and changes in the appearance of the eyes and nose (Malcarne et al, 1999). Age and disability are associated with dissatisfaction with body image, and younger patients with severe symptoms of the disease have an increased risk of developing body image concerns (Benrud-Larson, Heinberg, Boling, Reed, White, Wigley & Haythornthwaite, 2003). The results of the "Canadian Scleroderma Patient Survey of Health Concerns and Research Priorities" show that 77% of participants reveal concern about their physical appearance due to scleroderma, related to the hands and / or face (Taillefer et al, 2010).

In terms of depressive symptoms, the same Canadian study showed that about half of the participants report feeling "down, depressed or hopeless" at least several days in the previous two weeks.

The manifestation of depressive symptoms is favored in people with rheumatic diseases (Kobayashi-Gutiérrez,Martinez-Bonilla, Bernard-Medina, Troyo-Sanroman, González-Díaz, Castro-Contreras, ... Torres-Mendoza, 2009), and a frequent complication in scleroderma (Benrud-Larson et al, 2002). The symptoms of scleroderma can be painful, such as digital ulcers, Raynaud's phenomenon, skin changes, joint contractures and gastroesophageal reflux (Benrud-Larson et al, 2002), and severity of symptoms of pain is associated with depression (Kobayashi-Gutiérrez et al, 2009). In addition to pain, feelings of shame, disgust of themselves and self-deprecation of the body, are factors contributing to the adjustment to disease (Richards, Herrick, Griffin, Gwilliam & Fortune, 2004) and provide depressive symptoms (Roca, Wigley & White, 1996). On the other hand lowsocial support is one factor that predisposes to depression in these patients (Legendre, Allanore, Ferrand & Kahan, 2005). These symptoms of depression are in turn associated with an impairment of vocational, social and sexual adjustment (Benrud-Larson et al, 2002), directly affecting the quality of life (Danieli, Airò, Bettoni, Cinquini, Antonioli, Cavazzana, … Cattaneo, 2005).

The few studies conducted with these patients suggest that the quality of life is significantly diminished, and that half of patients with scleroderma are depressed. Depression symptoms have a considerable negative impact on clinical outcomes. Studies on women with scleroderma (females are more affected by the disease) suggest that these

patients show a high degree of dissatisfaction with body image, with depression being a moderating variable between dissatisfaction with body image and psychosocial function (Benrud-Larson et al, 2003).

In addition to depression, anxiety is also common in patients with scleroderma (Legendre et al, 2005). Anxiety symptoms are associated with clinical pain and changes in quality of life (Baubet, Ranque, Taïeb, Bérezné, Bricou, Mehallel, … Mouton, 2011) these symptoms are predicted by cognitive factors in relation to fear of negative evaluation (Richards et al, 2004).

The physical and psychological symptoms, including concerns about body image and depressive symptoms may have an impact on sexual functioning of patients with scleroderma (Thombs, Jewett, Assassi, Baron, Maia, El-Baalbaki, … Khanna, in press), of which women with scleroderma have greater commitment and sexual distress (Knafo, 2010).

Social Impact

Due to the high physical and psychological morbidity, social impact of this disease can be observed in the social costs associated with it. In addition to the costs associated with high use of health care with long periods of morbidity (Hansdottir, Malcarne, Furst, Weissman & Clements, 2004), the "Canadian Scleroderma Patient Survey of Health Concerns and Research Priorities" shows that half of the participants who were working indicated that the scleroderma affect their ability to perform their functions at work and one quarter of the participants also noted that scleroderma have an economic impact in their lives(Taillefer et al, 2010). This study also suggests that patients with scleroderma are more dissatisfied with health care than other chronically ill patients.

In the same study, almost half of participants reported that scleroderma affects very much or extremely, their capacity to work. More than half of participants reported that there was little to no understanding by their workplace of how their medical situation affected their capacity to work. Work disability constitutes a serious problem to scleroderma patients and this problem is associated with pain, fatigue and impaired hand function (Sandqvist, Scheja & Hesselstrand, 2010).

Illness perception

Patients with chronic illnesses often have an array of difficulties that may vary, depending of their concept about their disease, especially perceived severity and the type of development and control the person perceives. The illness representation that the patient creates is based on the information that he may have received from his doctors; and information from family and friends (Richards, Herrick, Griffin, Gwilliam, Loukes & Fortune, 2003). The literature reviewed allowed to locate only one study (carried out with only 49 participants) on the perception of the disease in scleroderma, showing that illness perception is related to emotional disturbance (Richards et al, 2003).This study also found that about half of the people with scleroderma do not understand the disease, making adjustment a difficult task, since people who have a good understanding are more likely to distance themselves emotionally, giving them a greater sense of control.

Intervention

Once scleroderma is a heterogeneous disease, treatment should be adapted to the needs of each patient (Hinchcliff& Varga, 2008). In addition to the medical treatment of symptoms of the disease, and considering all the implications of physical, psychological and social causes of the scleroderma, the psychological support is essential.

At present it is acknowledged that the treatment offered to these patients is not sufficient and an international team of researchers, health professionals and patients(Thombs et al., in press), are developing an intervention program for patients with scleroderma (The Scleroderma Patient-Centered Intervention Network - SPIN). This group is composed of representatives of patient organizations in Canada, the United States and Europe, doctors and researchers with expertise in this pathology, and researchers with expertise in health services research, health economics, health technology assessment, health policy and utilization of technology to provide health services to patients. The aim of the group, based on priority needs identified for these patients, respond to major problems: improving the functioning of the hands, reducing depressive symptoms, treat concerns about body image, and implement a general self-management oriented to the needs of patients (Thombs et al, in press). As the authors affirm, the creation of this group and putting into practice of his proposals will help to fill the gap in psychosocial research of scleroderma and also in clinical practice, providing the development and testing of intervention models, accessible to a large number of people with scleroderma.

It is estimated that there are more than 3,000 patients in Portugal. However, so far there has been no attempt to do a symptomatic characterization, or a study on the illness perception, the psychological and social impact of the disorder. The results of Canadian study showed several disturbing signs that reveal the needs for action in this area. On the European scene, there are few studies and, due to the absence of a psychosocial characterization, it is not possible to define the priority areas of intervention.This study aims to fill this gap, andallow patients with scleroderma expose the symptoms that most affect their life, how they feel about the disease and the perception of threat they feel about it.

Empirical study

The main objectives of this study were:

(1) To characterize the main symptoms of the disease and their impact on the quality of life among scleroderma patients;
(2) To assess the way that patients evaluate their satisfaction with medical care in terms of general satisfaction, technical quality, interpersonal manner, communication, financial aspects, time spent with doctor and accessibility and convenience.
(3) To describe the psychological symptoms, specially depressive symptoms;
(4) To assess the impact of physical appearance on social functioning;
(5) To understand the illness perception in terms of consequences, timeline, personal control, identity, coherence, emotional representation andillness concern.
(6) To evaluate the impact of the symptoms of the disease and illness perception on physical appearance

With this research we want to promote research on scleroderma calling attention to the various realities of the disease and raise awareness among health professionals and the general public to this problem. More specifically, to inform health professionals so that they can provide health care, more aware of the psychological reality of scleroderma.

Method

Participants

This study has 563 participants belonging to various European countries and Brazil. The European countries involved in the study are:France, United Kingdom, Cyprus, Hungary, Poland, Ireland, Finland, Denmark, Norway, Portugal, Spain and Italy. Data was collected between December 2010 and July 2011.

The criterion for participation in this study was that the participants need to have scleroderma. Participation was voluntary.

The mean age of participants were 47 years old (SD=14.1) and they were mostly female (N=490, 89%). Most of the participants were white (N= 511, 93%) and married or living as married (N= 373, 68%). Only 12% (N= 65) of the participants live alone and most participants have a college degree (N= 180, 37%). 34% (184) of participants reported that

were disable, on sick leave or unemployed, of those 184 participants, 108 (59%) have also reported that scleroderma was the reason.

Table 1
Demographics of participants

	Frequency	Percent
Gender:		
Female	490	88.9
Male	61	11.1
Racial/Ethnic Background:		
White	511	93.2
Black	16	2.9
Other	20	3.6
Current Marital Status		
Single	89	16.3
Married or living as married	373	68.3
Separated	25	4.6
Divorced	48	8.8
Widowed	11	2
Current family dwelling composition		
Alone	65	12.4
Living with spouse/partner	210	40.2
Living with spouse/partner and children	151	28.9
Living with children, no spouse/partner	29	5.5
Living with friends	5	1
Other	4	.8
Highest level of education		
Primary school	45	9.2
Secondary school	71	14.4
Higher education	157	32
University	180	36.7
Master	24	4.9
PhD	13	2.6
Current employment status		
Working full-time	145	26.7
Working part-time	54	9.6
Homemaker	37	6.8
Student	26	4.8
Retired	98	18
Disabled, sick leave or unemployed	184	33.8
If Yes --» because of scleroderma	108	58.7

Instruments

Canadian Scleroderma Patient Survey of Health Concerns and Research Priorities (Scleroderma Society of Canada and Canadian Scleroderma Research Group) was given to us by the authors with the proper authorization to translate. The authors created this questionnaire in order to access the most important aspects of living with scleroderma.To prepare for this questionnaire the team studied the existing literature and questionnaires about scleroderma and other rheumatic diseases.

The questionnaire has 11 Sections (Demographics; Diagnosis and Disease; Healthcare Services Utilization; Healthcare Services Reimbursement; Healthcare Services Reimbursement Needs; Medical Care; Symptoms; Employment; Sensations; Physical Appearance; Commentaries).

Section A (Demographics) is based on “Arthritis Treatment and Care in Canada Survey”.The Section B (Diagnosis and Disease)aims to describe the medical status of participants, such as determining the type of scleroderma and other health problems. Section C (Healthcare Services Utilization)aims to make a list of health services most often used by patients with scleroderma and also approach issues concerning mental health. Section D (Healthcare Services Reimbursement) andSection E (Healthcare Services Reimbursement Needs) will not be included in this study because health systems are different in the various countries that constitute this study. Section F (Medical Care) aims to assess what people with scleroderma feel about the medical care provided to them.This section uses an existing survey:*The Patient Satisfaction Questionnaire Short-Form*(Marshall & Hays, 1994). Section G (Symptoms) aims to list the frequency and impact of 69 symptoms. This section is based on *The Scleroderma Assessment Questionnaire (SAQ)*(Ostojic& Damjanov, 2006) and several articles. Section H (Employment) aims concerns about employment. Section I (Sensations) assesses depression, anxiety and social phobia. Depression is assessed with the instrument *The PHQ-2 a new depression diagnostic and severity measure*(Kroenke & Spitzer, 2002). The cut-off of this two items instrument is three, higher than or equal to three means sensitivity to severe depression. The first item assesses anhedonia and the second item assesses dysphoria. The sum of two items is a maximum of 6. Anxiety is assessed with the instrument *Generalized Anxiety Disorder (GAD)*(Kroenke, Spitzer, Williams, Monahan,& Lowe, 2007) and includes two items. The cut off this two items instrument is three, higher than or equal to three means generalized anxiety disorder. The sum of two items is a maximum of 6.Finally, social phobia is evaluated with the instrument*Mini SPIN: a brief screening assessment for generalized social anxiety disorder* (Connor, Kobak, Churchill, Katzelnick,& Davidson, 2001). The cut off the items of this instrument is six. Higher than or

equal to six means general social anxiety disorder. The sum of items varies between zero and a maximum of twelve. Section J (Physical Appearance) aims to assess concerns about body image.The three questions in this section are from the instrument*The Body Image Disturbance Questionnaire* (Cash, Phillips, Santos,& Hrabosky, 2004).Finally, the last section called "Commentary" is an open question that allows participants to write about something they think is relevant and has not been covered in the questionnaire.

The Brief Illness Perception Questionnaire (Brief IPQ)(Broadbent, Petrie, Main & Weinman, 2006), in existing versions in several languages (see"http://www.uib.no/ipq/"). This Questionnaire has nine items and assesses nine dimensions of illness perceptions, one per item. The dimensions assessed are: consequences, timeline, personal control, treatment control, identity, coherence, emotional representation, concern and causal attributions. The dimensions are evaluated quantitatively. A high score on the dimensions "consequences" and "timeline" means a more negative and more chronic illness perception, respectively. In the dimensions "personal control" and "treatment control", the higher the score represents more control. A high result in the dimension "identity" means that the participant associates a high number of symptoms to the illness.In turn, a high result in the dimension "coherence" reflects a poor understanding about the illness.In the "emotional representation" a high score means that the participant has negative emotions regarding the illness.In the "concern" the result reflect the level of concern about the illness; the higher result is equivalent to greater concern.The compute of the first eight items also allow evaluating the perception of threat in relation to illness. Highest score reflect greater perceived threat.

Procedure

After the contact with the Canadian team, we contacted the Federation of European Scleroderma Associations (FESCA) and all the European associations of scleroderma and all accepted to collaborate with the study.

The various associations have issued the study in several ways: sharing the details on their webpage, writing an advertisement in their newsletters and sending information to the email addresses of patients.

In Portugal, besides the support of the Portuguese Association of Patients with Scleroderma (APDE), we contacted all 50 hospitals treating patients with this condition.After the first contact we have submitted the project to the respective ethics committees. We obtained positive responses from all major national hospitals who are dealing with this

pathology.We got in contact with several doctors who treat patients with scleroderma and asked collaboration to disseminate the study to their patients. At the Hospital of S. João (Porto) it has been possible to collect data directly with patients. In the remainder, some had the collaboration of doctors who gave the questionnaires and helped to fill, in other cases, doctorsshared the questionnaire's link with the patients so that they would go online and fill it out.

For Brazil, we contacted the Association of Brazilian Patients with Scleroderma (ABRAPES).

Two methods were used in order to reach as many patients as we can and to get them to fill out the survey:

(1) We used Survey Monkey to create online versions of the survey. For each country we have created a different link.

(2) We also printed out on paper to provide the survey to patients who had no Internet access.

Confidentiality and anonymity were guaranteed either the online version or paper version of the questionnaire. In both versions were not asked the participants' names, date of birth, telephone number, and anything that would allowthe participants' identification.

To perform this study we had to translate the questionnaire to several languages. The Canadian team has provided us the version of the questionnaire in English and French. To cover a larger number of countries was necessary to translate the questionnaire into other languages. The languages chosen were: Danish, Portuguese, Spanish, Italian and German.The translation into Danish and German were made by the associations of scleroderma patients from Denmark and Belgium, respectively. Regardless of the language, some questions of the questionnaires (e.g. educational level) were adapted for each country.

Procedure for Data Analysis

The data analysis, descriptive and inferential, was performed using the statistical software package SPSS 18 (for Windows).

We did the exploratory data analysis to verify if the normality and homogeneity of the variables was guaranteed, which did not happen. However, we chose to do both tests, parametric and nonparametric, the results were equivalent, so we chose to present the results of parametric tests, since they are more robust (cf. Martins, 2011, p. 240).

We use descriptive statistics to describe the data. To make inferences from the data to general conditions, we used inferential statistics.In analyzing the results the differences are considered significant if p <.05.

Results

Diagnosis and Disease

In this section we summarize diagnosis and disease information. The Table 2 refers diagnosis information. Most of participants reported having been diagnosed with Diffuse Scleroderma or CREST. Rheumatology is the medical specialty most reported regarding the diagnosis of scleroderma.

The participants were diagnosed between 1 and 56 years ago (mean =8.45 ((*SD* = 7.21)). Only 19% (*N*=94) of participants got the diagnosis with the first doctor. However, 17% (*N*=86) were obliged to consult more than five physicians for diagnosis.

The information about Raynaud's indicates that 455 participants (91%) reported having Raynaud's.

Table 2
Diagnosis Information

	Frequency	Percent
Type of scleroderma:		
Limited	80	16.1
Diffuse	145	29.1
CREST	104	20.9
SINE	8	1.6
Linear	17	3.4
Morphea	22	4.4
Systemic	71	14.3
Unknown	49	9.8
Who diagnose		
Rheumatologist	265	53.6
Family doctor	45	9.1
Dermatologist	89	18
Internist	59	11.9
Pneumologist	7	1.4
Other	29	5.9

Table 3 lists all other medical conditions reported. Most of participants reported at least one other medical conditions (73%). The most conditions reported are esophageal dysfunction, pain, lung disease, osteoarthritis and depression.

Table 3
Other Medical Conditions

	Frequency	Percent
Esophageal dysfunction	199	35.3
Pain	123	21.8
Lung disease	115	20.4
Osteoarthritis	110	19.5
Depression	97	17.2
Ulcer or Stomach disease	76	13.5
High blood pressure	61	10.8
Rheumatoid Arthritis	56	9.9
Fibromyalgia	48	8.5
Heart disease	19	3.4
Diabetes	19	3.4
Hearing problems	35	6.2
Anemia	13	2.3
Kidney disease	11	2
Lupus	10	1.8
Cancer	6	1.1
Other	162	28.8

Healthcare Services Utilization

More than a half percent of participants (53%) were hospitalized at least once in the past because of scleroderma symptoms and 38% of the participants reported having access to adequate mental health care. However 65% never participated in support groups and the reason most often mentioned (48%) is that none is easily available.

Healthcare Services Reimbursement Needs

Table 4 contains the information about the health care services that the participants think considered necessary but were not receiving because of the cost. No more than one third of the participants reported that there were healthcare services they needed that they were not receiving because of the cost. The most often health services that participants reported they needed but were not receiving due to this reason were physiotherapy, vitamins and/or supplements, massage therapy, exercise therapy, and prescribed medicaments.

Table 4
Need for health care

	Frequency	Percent
None	178	32
Osteopathy	19	3
Post-surgical homecare	1	0
Personal care attendant services	4	1
Prescription medicaments	73	13
Non-prescription medicaments	47	8
Exercise therapy	93	17
Chiropractic therapy	9	2
Occupational therapy	6	1
Physiotherapy	116	21
Massage therapy	94	17
Acupuncture	70	12
Naturopathy	9	2
Vitamins and/or supplements	98	17
Assistive devices	6	1
Meal preparation aid services	34	6
Transportation	14	3
Personal care attendant services	3	1
House cleaning services	8	1
Bandages, ointments, and creams	60	11
Other	20	4

Medical Care

Table 5 summarizes the level of satisfaction of the participants with the medical care they are receiving with respect to seven domains: general satisfaction, technical quality of care, interpersonal manner, communication, financial aspects, time spent with physician, accessibility and convenience of care. Response options ranged from 1 (agree strongly) to 5 (strongly disagree). Some questions wore recoded so that for all items, higher scores represented higher satisfaction with medical care. In general, participants reflect to have positive attitudes regarding the medical care provided to them.The area where they are less satisfied is with the time spent with the doctor, and they show greater satisfaction with the interpersonal manner.

Table 5
Mean and standard deviation of Satisfaction with medical care

	Satisfaction with Medical Care
General Satisfaction	**3.2 (1.03)**
Medical care about perfect	3.25 (1.07)
Dissatisfaction with some things about medical care	3.16 (1.3)
Technical Quality of Care	**3.35 (0.86)**
Doctor's office has everything to provide complete care	3.2 (1.21)
Doubts about doctor diagnosis	2.89 (1.23)
Doctor are careful to check everything in the treatment	3.45 (1.1)
Doubts about the ability of doctors	3.34 (1.24)
Interpersonal Manner	**3.62 (0.94)**
Doctors act too businesslike and impersonal toward	3.3 (1.21)
Doctors treat in a very friendly and courteous manner	3.99 (0.98)
Communication	**3.2 (0.98)**
Doctors are good explaining the reason for medical tests	3.57 (1.09)
Doctors sometimes act ignoring	2.88 (1.19)
Financial Aspects	**3.18 (1.08)**
Confidence in being able to obtain medical care without financial loss	3.23 (1.19)
Prices of medical care is above the economic possibilities	3.21 (1.28)
Time Spent with Physician	**3.14 (0.85)**
Treatments are done in a hurry	2.82 (1.06)

Doctors spend plenty time in attendance	3.49 (1.1)
Accessibility and Convenience	**3.19 (0.8)**
Easy access to medical specialists required	3.54 (1.16)
People have to wait too long for emergency treatment	2.97 (1.13)
Idea that it is difficult to quickly get an appointment for medical care	2.99 (1.21)
Easy access to doctor when necessary	3.31 (1.09)

Note: Standard Deviation are in ().

Symptoms

The frequency of each of the 69 different symptoms in the past year is summarizedin Table 6, as well as its impact on the participant's abilities to carry out their everyday activities for those who reported experiencing the symptom. As we can see, the symptoms most commonly reported by patients are joint pain, fatigue, Raynaud's, muscle pain, hardening/tightening of skin, difficulty sleeping, difficulty remember things and heartburn. Fatigue is the symptom reported as having more negative impact on the daily lives of patients.

Table 6
Presence, frequency and impact ofsymptoms

	Symptom reported	Symptom reported most of the time/always	Symptom impact reported moderate/ severe/ extremely severe
Hardening/Tightening of Skin	422 (89%)	244 (51%)	307 (68%)
Itchy Skin	494 (88%)	259 (46%)	218 (48.2)
Changes in Skin Color	465 (83%)	268 (48%)	181 (41%)
Dilated Blood Vessels on the Hands	427 (76%)	259 (46%)	211 (53%)
Dilated Blood Vessels on the Face	377 (67%)	214 (38%)	146 (38%)
Skin Rashes	382 (68%)	182 (32%)	136 (35%)
Hypersensitivity to the Sun	413 (73%)	232 (41%)	177 (43%)
Skin Infections due to Open Sores	341 (61%)	179 (32%)	180 (48%)
Calcium Deposits in the Skin	329 (58%)	204 (36%)	163 (49%)
Skin Pain	437 (78%)	264 (47%)	241 (59%)

Joint Pain	448 (94%)	297 (63%)	375 (82%)
Swollen Joints	473 (84%)	282 (50%)	317 (73%)
Muscle Pain	422 (89%)	274 (58%)	351 (79%)
Tender Joints	476 (85%)	282 (50%)	299 (71%)
Difficulty Making a Fist	448 (80%)	311 (55%)	270 (67%)
Difficulty Fully Opening the Hand	418 (74%)	268 (48%)	223 (56%)
Difficulty Holding Objects	468 (83%)	264 (47%)	281 (67%)
Stiffness in the Hands	495 (88%)	343 (61%)	448 (80%)
Difficulty Turning on a Faucet	442 (79%)	235 (42%)	393 (70%)
Difficulty Self-Washing	378 (67%)	189 (34%)	351 (62%)
Difficulty Dressing	416 (74%)	194 (35%)	353 (63%)
Difficulty Walking	463 (82%)	265 (47%)	415 (74%)
Difficulty Climbing Stairs	467 (83%)	317 (56%)	422 (75%)
Difficulty Getting In/Out of a Car	428 (76%)	236 (42%)	368 (65%)
Dry Eyes	437 (78%)	284 (50%)	411 (73%)
Burning Sensation in the Eyes	401 (71%)	209 (37%)	379 (67%)
Gritty Sensation in the Eyes	415 (74%)	200 (36%)	347 (62%)
Dry Mouth	466 (83%)	311 (55%)	372 (76%)
Difficulty Chewing	389 (69%)	217 (39%)	373 (66%)
Difficulty Swallowing	442 (79%)	229 (41%)	364 (65%)
Painful Swallowing	345 (61%)	162 (29%)	344 (61%)
Choking While Eating Dry Foods	383 (68%)	188 (33%)	352 (63%)
Bad Taste in the Mouth at Night	415 (74%)	232 (41%)	303 (54%)
Heartburn	395 (87%)	201 (44%)	256 (67%)
Dental Caries or Decay	386 (69%)	205 (36%)	358 (64%)
Shortness of Breath	388 (86%)	170 (38%)	267 (69%)
Persistent Coughing	442 (79%)	239 (43%)	399 (71%)
Chest Pain	403 (72%)	203 (36%)	388 (69%)
Kidney Failure	292 (52%)	238 (42%)	415 (74%)
Rapid Heart Rate	407 (72%)	220 (39%)	352 (63%)
Irregular Heart Rate	396 (70%)	212 (38%)	359 (64%)
Fainting or Near Fainting	299 (53%)	145 (26%)	390 (69%)
Inflammation of the Liver	200 (36%)	139 (25%)	418 (74%)
Carpal Tunnel Syndrome	381 (68%)	221 (39%)	420 (75%)
Finger Ulcers	342 (61%)	216 (38%)	436 (77%)
Mouth Ulcers	281 (50%)	144 (26%)	377 (67%)

Nose Ulcers	225 (40%)	141 (25%)	375 (67%)
Migraine Headaches	400 (71%)	174 (31%)	391 (69%)
Diarrhea	476 (85%)	206 (37%)	382 (68%)
Constipation	432 (77%)	224 (40%)	384 (68%)
Stool Incontinence	303 (54%)	176 (31%)	415 (74%)
Nausea	422 (75%)	178 (32%)	352 (63%)
Unwanted Weight Loss	303 (54%)	172 (31%)	386 (69%)
Vaginal Dryness	455 (81%)	349 (62%)	436 (77%)
Erectile Dysfunction	51 (73%)	25 (43%)	34 (97%)
Gland Swelling	270 (48%)	167 (30%)	364 (65%)
Thyroid Problems	239 (43%)	198 (35%)	398 (71%)
Blood Clots	184 (33%)	149 (27%)	402 (71%)
Recurrent Fever	271 (48%)	158 (28%)	386 (69%)
Fatigue	419 (93%)	290 (64%)	346 (84%)
Difficulty Sleeping	402 (88%)	202 (44%)	291 (72%)
Difficulty Concentrating	382 (85%)	130 (29%)	251 (63%)
Difficulty Remembering Things	394 (88%)	119 (26%)	224 (55%)
Medication Side Effects	422 (75%)	203 (36%)	370 (66%)
Raynaud's	407 (90%)	289 (64%)	327 (80%)
Food Getting Stuck in the Mouth	361 (64%)	192 (34%)	373 (66%)
Difficulty Opening the Mouth	385 (68%)	230 (41%)	384 (68%)
Difficulty Brushing the Teeth	311 (55%)	198 (35%)	378 (67%)
Numbness in Feet or Lower Legs	463 (82%)	227 (40%)	390 (69%)

Note: Percentage related to the subjects that answer the item is in ().

Employment

Table 7 describes the impact of scleroderma in the participants' employment.Almost half the participants reported that scleroderma affects their ability to work somewhat, very much or extremely. A little more than half of the participants reported that scleroderma is not been accommodated by the workplace neither understood in terms of the ability to perform job-related tasks.

Table 7
Frequency of the impact in employment

	Employment' impact	
	Not at all/ a little bit	Somewhat/ very much/ extremely
Affect the ability to work	194 (35%)	278 (49%)
Affect the ability to meet financial obligations	323 (57%)	135 (24%)
Been accommodated by the workplace	293 (52%)	159 (28%)
Been understood by the workplace in terms of the ability to perform job-related tasks	292 (52%)	164 (29%)

Note: Percentage related to the subjects that answer the item is in ().

Depression and anxiety

The assembly of two items that constitute the PHQ-2 assessed depression severity measuring anhedonia and dysphoria, respectively. The results are in Table 8 and show that 38% of participants show sensitivity to severe depression.

Table 8
Frequency of PHQ-2 items

	Not at all	Several days	More than half the days	Nearly every day
Little interest or pleasure in doing things	114 (26%)	173 (39%)	68 (16%)	85 (19%)
Feeling down, depressed, or hopeless	128 (29%)	177 (40%)	55 (13%)	80 (18%)

Note. Percentage related to the subjects that answer the item is in ().

The assessment of anxiety symptoms is calculated using GAD-2. The results of these items are in Table 9 and the application of cut-off criteria show that 32% of participants reported symptoms of generalized anxiety symptoms.

Table 9
Frequency of GAD-2 items

	Not at all	Several days	More than half the days	Nearly every day
Feeling nervous, anxious, or on edge	132 (30%)	204 (46%)	61 (14%)	45 (19%)
Not being able to stop or control worrying	174 (39%)	175 (40%)	51 (12%)	42 (10%)

Note. Percentage related to the subjects that answer the item are in ().

The assessment of the general social anxiety symptoms (social phobia) was made using three items of Mini-SPIN. The results of these items are in Table 10. Using the cut –off of the instrument 30% of participants reported social phobia symptoms.

Table 10
Frequency of tree Mini-SPIN items

	Not at all	A little bit	Somewhat	Very much	Extremely
Fear of embarrassment causes avoid to do things or speak to people	205 (47%)	111 (25%)	46 (11%)	48 (11%)	30 (7%)
Avoid activities in which is the centre of attention	164 (37%)	88 (20%)	63 (14%)	85 (19%)	42 (10%)
Being embarrassed or looking stupid are among the worse fears	191 (43%)	96 (22%)	53 (12%)	65 (15%)	38 (9%)

Note: Percentage related to the subjects that answer the item are in ().

Body Image

Assessing body image, 34% of participants show body image disturbance. Table 11 describes the participants' concerns about changes in the body image, due to scleroderma.

Table 11
Frequency of Body Image Concerns

	Never or a little	Moderate, a lot or extremely
Preoccupation with parts of the body considered not attractive	190 (44%)	247 (57%)
Impairment in social, occupational or other important areas of functioning, due to changes caused by scleroderma	243 (56%)	195 (45%)
Avoidance of things due to the disfigurement by scleroderma	290 (66%)	147 (34%)

Note: Percentage related to the subjects that answer the item are in ().

Illness Perception

Table 12 summarizes the results of the means obtained in the dimensions of the Brief IPQ. Results show that participants are aware of the chronicity of the disease.Regarding the illness concern, participants report a high degree of concern with scleroderma. Participants are considerably emotionally affected by the illness. Regarding illness identity and

consequences, participants reported that scleroderma is associated with several symptoms, which considerably affects their lives.In general the participants believe considerably in the effectiveness of treatments and in the fact that they have some illness personal control. The subscale with the lowest score was coherence, showing that participants do not understand scleroderma.

Table 12
Means of Illness Perception

Subscales	*Mean*
Consequences	6.22 (2.74)
Timeline	9.33 (1.8)
Personal Control	5.79 (2.44)
Treatment Control	5.03 (2.41)
Identity	6.17 (2.4)
Illness concern	7.19 (2.53)
Coherence	3.97 (2.54)
Emotional Representation	6.58 (2.74)

Note: Standard Deviation are in ().

Inferential Statistics

The correlation among the variables age, years with diagnosis, frequency of the symptoms, impact of the symptoms, body image, depression, anxiety, social phobia, illness perception and satisfaction with medical care is presented in table 13.

Table13
Spearman correlations test between variables

Variable	1	2	3	4	5	6	7	8	9
1. Age									
2. Years with diagnosis	2.11**								
3. Frequency of Symptoms	.15**	.09*							
4. Impact of Symptoms	-.90*	0	.10*						
5. Body Image	-.13**	.07	.04	.38**					
6. Depression	-.09*	-.06	-.28**	.31**	.13**				
7. Anxiety	-.07	.04	0	.21**	.34**	.08			
8. Social Phobia	-.01	.01	.03	.25**	.53**	.10 *	.43**		
9. Illness Perception	.01	.07	.22**	.26**	.37**	-.01	.32**	.36**	
10. Satisfaction with medical care	-.07	.15**	.01	0	-.03	-.06	-.03	-.01	.10

Note: *p<.05; **p<.01

The years with diagnosis is positively correlated with the age (r= 2.11, p<.01). Older participants have more years with diagnosis.

The frequency of the symptoms is positively correlated with the age (r= .15, p<.01) and with the years with diagnosis (r= .09, p<.05). Older participants and with more years with diagnosis reported higherfrequency of the symptoms of scleroderma.

The impact of symptoms is negatively correlated with age (r= -.9, p<.05) and positively correlated with the frequency of the symptoms (r= .1, p<.05). Younger participants

reported more impact of symptoms. Participants who reported more frequency of symptoms, also reported more impact of the symptoms.

Body image is negatively correlated with age (r= -.13, p<.01) and positively correlated with impact of symptoms (r= .38, p<.01). Younger participant reported worse body image. Participants who reported more impact of the symptoms also reported worse body image.

Depression is negatively correlated with age (r= -.09, p<.05) and with frequency of symptoms (r= -.28, p<.01), and is positively correlated with the impact of symptoms (r= .31, p<.01) and with body image (r= .13, p<.01). Younger participants and participants with less frequency of symptoms reported more symptoms of depression. Participants with more impact of symptoms and participants with worse body image reported more symptoms of depression.

Anxiety is positively correlated with the impact of symptoms (r= .21, p<.01) and with body image (r= .34, p<.01). Participants with more impact of symptoms and participants with worse body image reported more symptoms of anxiety.

Social phobia is positively correlated with the impact of symptoms (r= .25, p<.01), with body image (r= .53, p<.01), with depression (r= .1, p<.05) and with anxiety (r_p= .43, p<.01). Participants who reported more impact of the symptoms, participants with worse body image, participants with depression and participants with anxiety, reported more symptoms of social phobia.

Illness perception is positively correlated with frequency of symptoms (r= .22, p<.01), with impact of symptoms (r= .26, p<.01), with body image (r= .37, p<.01), with anxiety (r= .32, p<.01) and with social phobia (r= .36, p<.01). Participants who reported more frequency of symptoms, more impact of symptoms, with worse body image, with anxiety and with social phobia, perceive illness as more threatening.

Satisfaction with medical care is positively correlated with years with diagnosis (r= .15, p<.01). Participants with more years with diagnosis are more satisfied with the medical care.

Hypotheses testing

Hypothesis 1:

Participants with depressive symptoms have a worse body image, report more symptoms of pain, more impact of pain, more symptoms of fatigue, more symptoms associated with the disease, more impact of these symptoms, have a more negative illness perception and are more dissatisfied with medical care they receive.

Table 14
Results of *t test for independent samples comparing participants with and without depression*

	With Depression	Without Depression	*t* (*df*)
	Mean (SD)	*Mean (SD)*	
Body image	8.87 (3.35)	6.51 (2.73)	-7.91 $_{(422)}$ ***
Frequency of symptoms of pain	7.18 (2.27)	6.71 (2.2)	-2.07 $_{(392)}$ *
Impact of symptoms of pain	7.3 (2.35)	6.46 (2.43)	-3.59 $_{(438)}$ ***
Fatigue	2.99 (1.18)	2.54 (1.17)	-3.68 $_{(399)}$ ***
Frequency of symptoms	39.41 (20.15)	38.99 (17.48)	-.23 $_{(438),}$ *n.s.*
Impact of symptoms	41.79 (11.67)	36.7 (12.97)	-4.15 $_{(438)}$ ***
Illness perception	52.02 (9.74)	44.77 (11.48)	-6.71 $_{(422)}$ ***
Medical care	62.9 (108.43)	50.51 (91.28)	-1.21 $_{(389)}$, *n.s.*

Note: * $p<.05$; *** $p<.001$

As we can see, participants with depression report worse body image, more symptoms and impact of pain, more fatigue, more impact of symptoms of the disease and perceive illness as more threatening. There were no differences between group on frequency of symptoms and medical care

Hypothesis 2:

Participants with anxiety symptoms have a worse body image, report more symptoms of pain, more impact of pain, more symptoms of fatigue, more symptoms associated with the disease, more impact of these symptoms, have a more negative illness perception and are more dissatisfied with medical care they receive.

Table 15
Results of *ttest for independent samples comparing participants with and without anxiety*

	With Anxiety *Mean (SD)*	Without Anxiety *Mean (SD)*	*t*
Body Image	8.83 (3.6)	6.81 (2.83)	-6.06 $_{(424)}$ ***
Frequency of symptoms of pain	7.1 (2.33)	6.86 (2.19)	-.93 $_{(393)}$, *n.s.*
Impact of pain symptoms	7.46 (2.24)	6.52 (2.44)	-3.65 $_{(439)}$ ***
Fatigue	2.85 (1.25)	2.67 (1.3)	-1.3 $_{(400)}$, *n.s.*
Frequency of symptoms	38.93 (19.89)	39.44 (17.93)	-.07 $_{(439)}$, *n.s.*
Impact of symptoms	38.93 (19.89)	39.44 (17.93)	-.26 $_{(439)}$, *n.s.*
Illness perception	52.75 (10.49)	45.83 (11)	-5.82 $_{(423)}$ ***
Medical care	48.5 (87.8)	56.16 (99.57)	-.7 $_{(391)}$, *n.s.*

Note: *** p<.001

Participants with anxiety have a worse body image, reported more impact of symptoms of pain and perceive illness as more threatening, but are not different from the ones without anxiety in the other variables.

Hypothesis 3:

Participants with social phobia symptoms have a worse body image, report more symptoms and impact of pain, more symptoms of fatigue, more symptoms associated with the disease, more impact of these symptoms, have a more negative illness perception and are more dissatisfied with the medical care they receive.

Table 16
Results of t test for independent samples between participants with and without social phobia and other variables

	With Social Phobia *Mean (SD)*	Without Social Phobia *Mean (SD)*	*t*
Body image	9.34 (3.48)	6.54 (2.64)	-9.05 $_{(421)}$***
Frequency of symptoms of pain	7.29 (2.32)	6.73 (2.17)	-2.29 $_{(393)}$ *
Impact of symptoms of pain	7.26 (2.4)	6.57 (2.41)	-2.73 $_{(437)}$ **
Fatigue	2.96 (1.2)	2.61 (1.16)	-2.71 $_{(397)}$ **
Frequency of symptoms	39.27 (21.27)	39.21 (17.12)	-.03 $_{(437)}$, *n.s.*
Impact of symptoms	41.98 (12.15)	37.20 (12.72)	-3.67 $_{(437)}$ ***
Illness perception	52.39 (10.18)	45.48 (11.04)	-6.04 $_{(421)}$ ***
Medical care	56.2 (96.1)	54.9 (99.38)	-.12 $_{(389)}$, *n.s.*

Note:*p<.05; ** p<.01; *** p<.001

Participants with social phobia have significantly worse body image, reported more symptoms of pain, more impact of symptoms of pain, more fatigue, more impact of symptoms of the disease and perceive illness as more threatening.

Discussion

This study aims mainly to characterize the most common symptoms of scleroderma, the impact felt by patients, assess satisfaction with medical care, psychological symptoms and illness perception. Another objective is to understand the correlation between the variables: age, years with diagnosis, frequency of the symptoms, impact of the symptoms, body image, depression, anxiety, social phobia, illness perception and satisfaction with medical care. And finally the study aims to evaluate if there are differences in the dimensions: body image, frequency of symptoms of pain, impact of symptoms of pain, fatigue, frequency of symptoms related to scleroderma, impact of this symptoms, illness perception and medical care,when comparingpatients with andwithout depressive symptoms; patients with andwithout symptoms of anxiety and patients with andwithout symptoms of social phobia.

The five most common symptoms reported by patients were: joint pain (94%), fatigue (93%), Raynaud's (90%), muscle pain (89%) and hardening / tightening skin (89%). These results were approximated to the Canadian study (Taillefer et al, 2010). Regarding the impact of symptoms, the five symptoms with the greatest impact were: fatigue (84%), joint pain (82%), Raynaud's (80%), Stiffness in the hands (80%) and muscle pain (79%). Also in this case the results are similar to those of Canada (Tailleffer et al., 2010).

Only 19% of patients were able to get the diagnosis of scleroderma with the first doctor consulted. This could be explained by the difficulty of diagnosis of scleroderma. Often the symptoms of scleroderma tend to be confused with symptoms of other diseases for a long time, moreover, as already stated, this disease remains unknown to many health professionals.In general, participants showed satisfaction with the medical care they receive and this result is similar to that obtained by Taillefer et al. (2010).Participants with more years of diagnosis revealed to be more satisfied with the medical care they receive.This result can be explained because there is less uncertainty about the diagnosis when compared with those diagnosed more recently.There were no differences in satisfaction with medical care among people with and without depression, or with and without anxiety or with and without social phobia. We also found no significant correlations between satisfaction with medical care and age, or frequency or impact of symptoms.Further studies must be made to determine which variables are related to satisfaction with medical care.

Almost a third of participants reported that they do not have access to all medical care due to its cost. Scleroderma involves many treatments and, depending on the health system of each country, not all treatments are free or reimbursed to patients. This data can be even

more relevant with the fact that almost half the participants showed that scleroderma affects the ability to work.

Regarding depression, the results suggest that 38% of participants have sensitivity for severe depression. This finding is consistent with the fact that depression is quite common in scleroderma (Benrud-Larson et al, 2002).The results suggest that younger participants reported more symptoms of depression andthis finding supports the conclusion of Roca (1996) that younger patients have more depressive symptoms.An interesting relation to depression, is that participants who reported less frequent symptoms have fewer symptoms of depression, however there are no differences between participants with and without depression (a measure that uses a cut-off value) in the frequency of symptoms reported. Those who have more impact report more symptoms of depression and there are differences between participants with and without depression in the account of the impact of symptoms. The interpretation of these data suggests that the impact perceived by the participants is important in the existence of depressive symptoms and not the frequency of symptoms, or it can be that subjects with more depressive humor report higher impact of the symptoms.

About one third of participants (34%) have body image disturbance. In particular, 84% of participants report experiencing preoccupation with body image due to scleroderma. This result is approximate to that obtained by Taillefer et al. (2010) which was 77%. Participants who reported worse body image reported more symptoms of depression, and there are differences in participants with and without depression in body image, since those who have depression have poorer body image.This finding is not surprising since scleroderma involves many physical changes visible in parts of the body, and depression has been reported as a common problem in people affected with various types of deformation (Thompson, 2001).

The results indicate that in relation to anxiety, 32% of participants reported symptoms of generalized anxiety. This finding is consistent with Thompson (2001) which says that anxiety is very prevalent in people with some type of physical disfigurement; in particular, anxiety is common in scleroderma patients (Legendre et al., 2005). Participants with worse body image reported more symptoms of anxiety, and there are differences between participants with and without anxiety, participants with anxiety have a worse body image. We found no relationship between the frequency of symptoms of scleroderma and anxiety. This finding is consistent with (Baubet, 2011) and can be explained by the fact that the emotional response is more related to the meaning given to the disease thanto disease severity.

Regarding the impact of the symptoms of pain, there are differences between participants with and without anxiety, with the first report more symptoms of pain.Baubet (2011) also associated symptoms of pain, a characteristic of scleroderma, with anxiety.

The symptoms of social phobia are reported by30% of the participants. More than half of participants reported that they avoid activities which they are the center of attention; this finding is consistent with Taillefer (2010). Participants with worse body image reported more symptoms of social phobia and there are differences between participants with and without social phobia: those with social phobia reported worse body image.According to Thompson (2001) people with disfigurement tend to avoid social situations.People with social phobia reported more frequency and impact of symptoms of pain and more impact of scleroderma symptoms than those without social phobia.

Regarding the illness perception, participants report a high degree of concern with scleroderma. This can be explained by the severity of the symptoms of scleroderma and also by its unpredictability and unfamiliarity.In fact, participants reported that they do not understand the disease, consistent with Richards et al. (2003).This idea can be supported by other results of this study that indicate that there is a correlation between the frequency and impact of symptoms of scleroderma with the perception of the disease.According to Richards et al. (2003) Illness perception is related to emotional disturbance. In this study, illness perception is positive correlated with anxiety and social phobia, however there is no significant correlation with depression and this is a puzzling result.We found that people which have perceived greater threat of disease, reported worse body image. This can be explained by another result, in which the participants reveal that the symptoms are seriously affecting their life.

Finally, participants who reported more symptoms, also reported higher impact of symptoms. Perhaps because of the fact that people who experience a large number of symptoms, may perceive the impact of symptoms as more severe and more affecting their quality of life.

Limitations and Future Studies

There were several limitations identified during the implementation of this study.

The time available to develop the project was too short, given the extent of it. It took a long time to coordinate contacts with all institutions concerned, particularly Portuguese hospitals and international associations. Thus, we could not include in this study all data collected since the collection still running. Despite this limitation and the extension of the survey, the number of participants exceeded our initial expectations.

Once that had to be made several translations of survey, it was not available online simultaneously for all countries, due to this factor and also because the number of patients is not the same in all countries (the number of the population is very different in the countries involved), the number of participants from various countries was not proportional.

It would have been interesting to have included the social support variable in this study.

The aim of this study was not to evaluate predictors of psychological adjustment, but in the future it will be important to evaluate the contribution of some clinical variables for the depression and anxiety symptoms. Another alternative would be to evaluate the way depressive symptoms influence the report of symptoms.

Analysis should consider the different types of scleroderma, because there are many differences in symptoms and severity of for instance a limited type and diffuse type. Are there differences in adaptation to the disease between different types of scleroderma? This is a question which remains for future studies.

We intend to continue research in this area so far largely neglected.The next step is to enter the remaining data in the study, including the following countries: Germany, Belgium and Switzerland.

We maintain contact with the Canadian team and also with a Dutch team that did the same study of Dutch patients and we as a project to establish a cooperation plan to compare the data.

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