

## REVIEW ARTICLE

# Systemic Sclerosis (Scleroderma): An Integrated Challenge in Rehabilitation

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**ABSTRACT.** Casale R, Buonocore M, Matucci-Cerinic M. Systemic sclerosis (scleroderma): an integrated challenge in rehabilitation. *Arch Phys Med Rehabil* 1997;78:767-73.

Systemic sclerosis (SSc), a multisystem disease involving the microvascular system and the connective tissue, is considered one of the most difficult rheumatic diseases to treat. The natural history of the disease evolves from an edematous to a sclero-atrophic phase following two different temporal patterns: acute or chronic. The former leads to early death, and the latter evolves slowly toward severe disability that deserves rehabilitative intervention. Despite the poor prognosis, recent improvements in diagnosis and treatment have led to longer patient survival, thus increasing the need to intervene against the development of tissue fibrosis and contractures by using appropriate integrated rehabilitation programs. This article does not review the medico-pharmacological management of visceral manifestations of the disease. Rather, it is divided into six parts, which include analyses of the changes in skin, joints and tendons, and muscle induced by SSc; examination of the existing literature on rehabilitation strategies and treatments; discussions of the pain and peripheral sensory-motor system involvement that are present to a greater or lesser extent in almost all patients and influence not only the duration and outcome of rehabilitation but also the patient's family, social life, and working ability; and consideration of ergonomic and occupational interventions. No controlled studies have been done on the few rehabilitation guidelines and specific protocols identified, so it must be emphasized that this article is a summary of opinions expressed in the literature and the authors' own findings. Particularly lacking are studies on such aspects as ergonomics, work intervention, or the management of sexual dysfunction. Experience gained in the rehabilitation of skin burns and other rheumatic diseases forms the basis for a logical approach to SSc patients.

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**S**YSTEMIC SCLEROSIS (SSc) is a multisystem disorder affecting the microvascular system and the connective tissue, causing alterations in the skin (scleroderma) and in a variety of internal organs.<sup>1,2</sup> Vascular changes impair the function of heart, lung, kidney, and peripheral nervous system.<sup>1-3</sup>

The natural history of the disease evolves from an edematous to a sclero-atrophic phase.<sup>1</sup> The disease may progress following

two different temporal patterns, acute or chronic.<sup>2</sup> The former is characterized by diffuse skin involvement (extremities and trunk) and a poor prognosis because of the rapidity and severity of skin and internal organ involvement. In the latter, the disease, confined to the extremities (face, neck, hands, forearms, arms and, less frequently, feet and legs), evolves slowly toward severe disability deserving a rehabilitative intervention<sup>2</sup> (table 1).

Despite the poor prognosis, in recent years impressive improvements in diagnostic tools have been made and an early diagnosis, as well as early aggressive treatment, is now possible.<sup>1</sup> This has led to patients surviving longer, increasing the need to intervene against the development of tissue fibrosis and contractures. While heart, lung, and kidney involvement predominantly need a clinical-pharmacological approach, it is recognized that the major rehabilitative problems arise from skin induration and joint and muscle involvement.<sup>1</sup> These conditions are worsened by the frequently concomitant sensory-motor neuropathy. Although clinicians are interested in the importance of rehabilitative approaches to disabilities deriving from skin and joint modifications, only scanty specific literature on this subject is available. Furthermore, no interest has been devoted to the evaluation of the impairments, disability or the handicaps related to peripheral nervous system involvement and pain in SSc.

## PURPOSE

This article summarizes the current management of skin, joints and tendons, and muscles in SSc, and also reviews the management of pain, peripheral nervous system (PNS) involvement, and ergonomics and working interventions (outline in table 2).

MEDLINE and EMBASE (electronic version of printed *Excerpta Medica*) have been used as reference sources. Both sources, as well as an extended search of references identified from bibliographies of articles dating to the mid-1950s, showed a lack of controlled studies on rehabilitation in SSc. Information regarding current rehabilitation protocols in use in our institutions has also been used. This article, therefore, summarizes the existing literature on rehabilitation strategies and treatments published since the mid-1950s and reports some of our experience from the rehabilitation of SSc patients.

## SKIN

There are several methods of evaluating the degree of skin involvement<sup>4-10</sup> (table 3). In the advanced phase of SSc the skin is tight, pale, and waxy; the patient has a progressive impairment of finger flexion, evolving into finger retraction, which gives the hand a "clawlike" aspect.<sup>11</sup> In the more severe forms, skin induration also affects oropharyngeal tissues (microstomia) and sexual organs, generating problems in food supply and sexual functions; these extreme expressions of the disease are terminal end points.<sup>1</sup>

The purpose of a rehabilitative approach to skin involvement, however, is to prevent or delay this kind of evolution, starting with a rational program in the early phase of the disease. There are currently no rehabilitative guidelines for skin problems of

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Table 1: Clinical and Pathogenic Classification of SSc

<b>Diffuse Cutaneous Subset</b>
Onset of Raynaud's phenomenon within 1 year of onset of skin changes (puffy or hidebound), truncal and acral skin involvement.
Presence of tendon friction rubs.
Early and significant incidence of interstitial lung disease, oliguric renal failure, diffuse gastrointestinal disease, and myocardial involvement.
Absence of anticentromere antibodies and 30% positivity of antitopoisomerase I (Scl-70).
Nail-fold, capillary dilatation, and capillary destruction.
<b>Limited Cutaneous Subset</b>
Raynaud's for years or decades.
Skin involvement limited to hands, face, feet, and forearms (acral) or absent.
Significant late incidence of pulmonary hypertension with or without interstitial lung disease, trigeminal neuralgia, skin calcifications, telangiectasia.
High incidence of anticentromere antibodies (70% to 80%).
Dilated nail-fold, capillary loops, usually without capillary dropout.

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patients with SSc, but the experience gained in the rehabilitation of skin burns<sup>12</sup> may form the basis of a logical approach to rehabilitation in patients with SSc.

### Motion Exercises

Range of motion (ROM) exercises may prevent skin retraction and help skin vascularization. For example, exercises from Bell's palsy rehabilitation may be used for the face.<sup>13</sup> Our experience is that exercises that may be useful include stretching the skin and oral mucosa by activation of the orbicularis oculi and perioral muscles and those for mouth excursion (ie, yawning with open mouth).<sup>14</sup> Our program includes not only active but also passive exercises for the trunk and the limbs, depending on the clinical phase of SSc. The literature does not differentiate between the rehabilitation of edematous or atrophic skin; only nonspecific motion exercises, regardless of the clinical phase, have been used.<sup>15</sup> Our approach to the rehabilitative treatment of skin problems in SSc relies on the differentiation of programs for atrophic and edematous skin. For instance, in the edematous phase motion exercises are performed carefully to avoid provoking skin stretching that may result in pain and worsening of stiffness. When skin induration is present, passive and active movements are used with progressive loads.

### Massage

Patients with SSc have a dramatic decrease in cutaneous oxygenation due to vessel obliteration.<sup>3</sup> Massages may complement ROM in stimulating vascular function. Massage may mobilize skin fluids, reduce edema, and increase skin temperature

by dilating microcirculation through a local or reflex action<sup>16</sup>; it may also have a local analgesic effect by increasing the pain threshold.<sup>17</sup> Basic maneuvers are gliding, stroking, kneading, and compression of soft tissues.<sup>18</sup> Among various kinds of massage, "Bindegewebsmassage" or connective tissue (reflexogenic) massage<sup>19</sup> and Soft Tissue Mobilization (STM)<sup>20</sup> have been usefully employed in our patients. Although no comparative data are available, these types of massage place fascia and muscle in an elongated position and may be helpful when tissue fibrosis is not dominant.

The use of heavy petrolatum or lanolin lubricants to protect the skin during massages has been proposed in SSc patients.<sup>15</sup> Lubricants have also been recommended for sexual intercourse because vaginal dryness occurs in a high percentage of SSc patients (71%) leading to ulcerations and dyspareunia.<sup>21</sup> Sexual problems may also occur in men with SSc because of skin induration leading to a loss of penile length and erectile function.<sup>22</sup> Penile implantation surgery has been proposed,<sup>23</sup> but no extensive follow-up data are available.

### Heat

Superficial moist heat applied for 3 minutes increases soft tissue temperature about 3°C down to a depth of 1 cm, producing a beneficial sensation of warm skin and increasing skin flexibility.<sup>14</sup> Because the skin in SSc patients is poorly perfused and hypoxic,<sup>10</sup> hypoperfused skin may easily overheat, resulting in tissue damage. Paraffin baths of hands and feet<sup>15,25</sup> have been suggested in SSc patients. Ultrasound has also been reported to have a favorable effect in SSc.<sup>26</sup> We found that in SSc patients spas have some benefit, possibly because the emollient effect of water immersion and the application of mud generates increased skin temperature. Spa therapy may help the patient's compliance by promoting social interaction.

CO<sub>2</sub> laser has been used for the treatment of skin induration and vascular insufficiency. Our experience with this technique in 25 patients with SSc suggests that it reduces the number of attacks of Raynaud's phenomenon and fingertip pain, and increases reepithelization of finger lesions and digital blood flow, with a satisfactory subjective improvement in hand function.

Nonpharmacological management of attacks of Raynaud's

Table 2: Outline of the Contents

<b>Skin</b>
Motion Exercises
Massages
Heat
Acupuncture and TENS
<b>Joints, Tendons, and Muscle</b>
Rest
Active and Passive Exercises
Heat
Stretching
Splinting and Prosthesis
<b>Pain</b>
TENS and Acupuncture
Laser
Heat and Cold
Topical Treatments
<b>Peripheral Nervous System</b>
Prevention of Secondary Damage
Electrical Stimulation
Passive Range of Motion
Reeducation
<b>Ergonomic and Working Intervention</b>

Table 3: Methods of Evaluating the Degree of Skin Involvement

Skin score	Black <sup>4</sup>
Ultrasound	Uhn and colleagues <sup>5</sup>
Durometer	Falanga and Bucalo <sup>6</sup>
Elastomer	Ballou and colleagues <sup>7</sup>
Slip test	Bennis and colleagues <sup>8</sup>
Magnetic resonance imaging	Richard and colleagues <sup>9</sup>
Skin O <sub>2</sub>	Silverstein and colleagues <sup>10</sup>

phenomenon relies mainly on prevention. Improvement in the acral circulation in SSc patients by prevention may include hypnosis and autogenic training,<sup>27</sup> education about hand protection in the cold, the use of warm mittens, and temperature biofeedback<sup>28</sup> to increase skin temperature of the fingers.

### Acupuncture and Transcutaneous Electrical Nerve Stimulation (TENS)

Manual or electric acupuncture as well as transcutaneous electrical nerve stimulation (TENS) have been reported to improve skin vascularization; acupuncture increases skin blood flow, possibly through the activation of vasoactive neuropeptides,<sup>29,30</sup> and TENS increases skin temperature as an effect of reflex vasodilatation.<sup>31</sup>

Manual acupuncture has been successfully used to improve blood flow in patients with xerostomia.<sup>32</sup> In one SSc case with painful ulcers and Raynaud's phenomenon, Kaada,<sup>33</sup> using acupuncture-like low-frequency TENS, reported a significant improvement in the clinical picture and reduction of pain and an improvement of calcinosis and dysphagia. Although some success in Raynaud's phenomenon control may be achieved with physical treatments, the major therapeutic intervention relies on education about how to avoid thermal stress, protecting hands from the cold, and increasing skin temperature.<sup>27,28</sup>

Acupuncture has been shown to have some effects on inflammatory processes.<sup>34</sup> In one study 34 SSc patients were treated with low-frequency electrical acupuncture. In these patients, data showed improvement in the inflammatory reaction but no changes in the immune reactions or the clinical picture.<sup>35</sup>

### JOINTS, TENDONS, AND MUSCLE

Patients with SSc may develop a symmetric painful arthropathy<sup>36</sup> involving the wrists, fingers, knees, and ankles. In many patients the major complaint is reduction of temporomandibular joint range and subsequent difficulty in nutritional support. In the majority of SSc patients, the articular involvement is caused by skin thickening and retraction over and around the joint leading to joint disuse.<sup>37</sup> Mild weakness and progressive muscular atrophy may be present and accompanied by the elevation of muscular enzymes.

Muscles can be affected in three ways in SSc. The most common finding is diffuse disuse atrophy. Nonprogressive, usually mild, proximal myositis with increased creatine phosphokinase (CPK) may be present when a rheumatoid-like synovitis is present. Progressive myositis, leading to different degrees of diffuse atrophy and fibrotic retraction of the muscle, occurs in some cases when the inflammatory reaction is severe. The functional assessment of joints is based on their ROM and joint pain, while muscle impairment is assessed by clinical and electrophysiological evaluations of strength and localized muscle fatigue.

#### Rest

We prescribe rest for short periods of time and only in selected cases of SSc in which the myositic reaction is severe. Similarly, when synovitis is prominent, rest may be prescribed. Splints may be applied locally for short periods of time,<sup>38</sup> but systemic rest, highly recommended in arthritis, is not suggested for SSc.

#### Active and Passive Exercises

Skin and joint involvement leads to a progressive reduction in ROM. Joint structure may become deformed and the muscles shorten during prolonged immobilization, even in the absence of any joint and/or muscle inflammatory process.<sup>1</sup> Although

Chazen and colleagues<sup>39</sup> reported the usefulness of an exercise program in the maintenance of ROM of joints in 19 children affected by focal scleroderma, no specific exercises were proposed by the authors. Before prescribing either passive or active exercises in SSc patients, the degree of joint limitation and inflammation, the periarticular skin involvement, and the condition of the muscles should be evaluated. The exercise program should be slow and progressive, periodically reevaluated and adjusted according to the progression of the disease.

Although passive exercises may benefit patients with joint retraction and severe muscle weakness, they may worsen joint inflammation in patients with severe arthropathy. Active muscular contractions (isometric or isotonic) are important to maintain and/or try to restore full ROM. Isometric exercises require little time, are usually easy to perform, and are best suited for patients with arthropathy because no joint movement occurs. Isotonic exercises may provoke muscle soreness and are not suited to SSc patients with biomechanically impaired joints. High- and low-repetition rates of isokinetic exercises may be used to avoid articular overload. In our SSc patients, using high-repetition rates, we recorded little soreness after this kind of exercise. When mild myositis was present only high-repetition rates were employed. Physical capacity may be maintained by means of a continuous training program involving walking, light running, swimming, cycling, and cranking activities on a daily basis.<sup>40</sup> Active and passive exercises include facial muscle rehabilitation and treatment of reduction in temporomandibular joint articulation range.<sup>41</sup> Perioral muscle exercises are important in the management of dysphagia and may also include motor exercises to improve tongue placement.<sup>42</sup>

#### Heat

Exposing the hands to a temperature of 40°C to 45°C may improve metacarpophalangeal joint stiffness by 20%<sup>43</sup> and an increase of 5°C to 7°C significantly increases collagen extensibility,<sup>44</sup> suggesting a beneficial use of heat in improving ROM in SSc patients. Relatively high temperatures should be used with caution because of the risk of skin damage. Hand flexion-extension improved in two cases of SSc treated with ultrasound.<sup>26</sup> Recently, ultrasound has also been proposed as a way to improve temporomandibular joint function in SSc patients.<sup>42</sup> Radon spas in association with physical therapy have been reported to improve ROM in SSc patients, although no controlled studies are available.<sup>45</sup>

#### Stretching

Passive and active stretching help the patient to maintain joint ROM and to elongate tendons and muscles, especially when fibrotic retractions are present. It has been demonstrated that collagen and connective tissues exhibit a quality of plastic elongation when undergoing constant and prolonged tension.<sup>46</sup> Careful and slow sustained stretching, performed manually with weights, traction, and serial casting may also be helpful for skin retraction. Manual elongation of the joints may also be helpful because of its analgesic effect.<sup>47</sup>

#### Splinting and Prosthesis

Characteristic hand deformities occurring in SSc include loss of flexion of the metacarpophalangeal joints, extension of the proximal interphalangeal joints, and thumb abduction, opposition, and flexion, resulting in a decreased web space. Although dynamic splinting is appropriate for contractures, few studies address the efficacy of splinting in SSc.<sup>48,49</sup> A study of 19 SSc patients who used individually constructed dorsal thermoplastic hand splints that maintained the wrist in approximately 15°

extension and the metacarpophalangeal joints at 0° and that had a dynamic proximal interphalangeal joints extension outrigger failed to demonstrate that the splints maintained the proximal interphalangeal extension when compared with the unsplinted hand. Furthermore 21% of the patients had splint-exacerbated Raynaud's phenomenon.<sup>50</sup> Because of this problem we advise against the use of dynamic splinting to correct proximal interphalangeal joint contractures or maintain proximal interphalangeal joint extension in SSc patients.

Microstomia progresses in SSc patients and is always a severe complication in the atrophic phase. For prevention and, when possible, its partial correction, tissue-borne or tooth-borne microstomia appliances might be considered.<sup>51</sup> Another important problem affecting SSc patients is vaginal tightness and constricted introitus. In a recent study, 5 of 60 patients complained of vaginal involvement.<sup>21</sup> No solution to this problem has been proposed.

Ulcerations and autoamputation of fingers and toes are seen as a result of severe acral anoxia due to Raynaud's phenomenon.<sup>52</sup> Surgical toe amputations are rarely performed in SSc, and amputation of the lower extremities at or proximal to the midtarsal level has been previously reported in only eight cases.<sup>37,53</sup> Although rarely performed, when amputations occur, they heal well, the skin tolerates wearing a prosthesis, and patients may become successful prosthesis users.<sup>54</sup> The systemic manifestations of SSc are often so severe that the patient may die before he or she can benefit from prosthetic training.

### PAIN

In some patients joint pain becomes one of the main symptoms. Although analgesic medications may be effective, anti-inflammatory and analgesic treatments may elicit additional problems in these patients in whom esophageal reflux and gastrointestinal involvement are very frequent. We recommend physical therapy and topically applied treatments as a safe approach to chronic pain control. Some of the treatments already mentioned in the section devoted to skin induration may be used for pain control.

### TENS and Acupuncture

The efficacy of TENS<sup>55,56</sup> and acupuncture<sup>57,58</sup> has been described in the management of pain in osteoarthritis and rheumatoid arthritis. We suggest the use of TENS in SSc in two particular conditions: (1) presence of polyarticular spontaneous pain where high intensity, low-frequency acupuncturelike TENS is more effective activating a diffuse endorphinergic pain control system<sup>59</sup>; (2) joint pain provoked by passive and active mobilization of a particular limb segment where high-frequency, low-intensity TENS applied to the painful joint and/or to the related nervous trunk supplying it may relieve pain by segmentary inhibition at spinal level.<sup>60</sup> Some of our patients reported that they were beneficially treated with acupuncture. Acupuncture acts through the activation of different antinociceptive systems: an endorphinergic naloxone dependent system<sup>61</sup> and a naloxone independent nonopiatergic system, the latter probably mediated by serotonin at spinal level,<sup>62</sup> and therefore may be useful in controlling spontaneous and provoked joint pain in these subjects. In our opinion the use of acupuncture in these patients is to be avoided because of their skin involvement.

### Laser

Laser light has been reported to control rheumatoid, myofascial, and neuropathic pain.<sup>63-65</sup> We used CO<sub>2</sub> laser therapy in 25 SSc patients with positive results, although this was an uncontrolled study.

**Table 4: Central, Peripheral, and Autonomic Abnormalities Found in SSc**

Central Nervous System (CNS)
Psychiatric disorders
Modification of EEG pattern
Bulbar palsy
Peripheral Nervous System (PNS)
Optic neuritis
Trigeminal neuropathy
7th, 8th, 9th, 12th cranial nerve neuropathy
Median nerve compression
Mononeuritis multiplex
Polyneuropathy
Autonomic Nervous System (ANS)
Sympathetic dysfunction
Parasympathetic dysfunction
Sympatho-vagal mixed involvement

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### Heat and Cold

Although both cold and heat have been used in the management of osteoarthritis pain, in SSc, cold is not recommended for SSc because of the presence of vasomotor instability (Raynaud's phenomenon). Moreover, since severe fingertip anoxia, as seen during Raynaud's attacks induced in SSc by cold exposure, may lead to ulcerations, we suggest education on hand protection in the cold and the use of temperature biofeedback.<sup>27,28</sup> Heat, in its different forms of application (paraffin baths, ultrasound, and short wave diathermy) is used in degenerative joint diseases,<sup>66</sup> but should be used with caution in SSc patients because their skin is poorly perfused and easily overheated.<sup>67</sup>

### Topical Treatments

Topical treatments such as rubbing blistering fluids onto the skin have been widely used as home remedies for joint pain. They produce "hyperstimulation analgesia."<sup>68</sup> Recently, capsaicin, a neurotoxin derived from chili pepper, has been proposed as an effective topical treatment for the control of joint pain in osteoarthritis.<sup>69</sup> In SSc, the use of capsaicin may be considered in the future not only as a pain controller but also as a vasodilating drug as a consequence of its action on vasomotor thin unmyelinated C-fibers.<sup>69</sup> Poor skin perfusion together with the presence of sensory disturbances may lower the benefit/risk ratio in using hyperstimulation techniques.

### THE PERIPHERAL NERVOUS SYSTEM

Some cases of central nervous system involvement have been recorded in SSc patients, although it is not possible to specify whether this is a primary event or the consequence of internal organ alterations<sup>70</sup> (table 4). Peripheral nervous system (PNS) involvement is a much more common event in SSc patients featuring a peripheral sensory-motor neuropathy, often associated with autonomic involvement. The onset and evolution of PNS involvement is unpredictable; early signs of sensory-motor neuropathies have been reported in cranial as well as acral nerves. More subtle and not easily detectable are the early signs of autonomic impairment that in some cases may even predate sensory-motor involvement.<sup>70,71</sup> Peripheral nerve involvement in SSc rarely leads to severe or complete paralysis or sensory loss.<sup>70</sup> In our experience, carpal tunnel syndrome is the most common abnormal finding.<sup>72</sup> In SSc, the weakness of the prime mover, for instance in the hand muscles, may contribute to the development of metacarpophalangeal joint deformity. Failure to recognize a weakness of the prime movers may result in the underestimation of the underlying neuropathy and inappropriate

rehabilitation. Peripheral nerve lesions in SSc may be predominantly sensory.<sup>71</sup> Unrecognized sensory loss may explain some hand function inefficiency in the presence of adequate muscle strength. This may happen in patients with recurrent joint inflammation with compromised proprioception. We tailor the rehabilitation program to the different peripheral motor, sensory, and autonomic impairments.

### Prevention of Secondary Damage

In SSc, the main rehabilitation strategy for peripheral neuropathy is based on prevention of secondary damage and relapses, monitoring of functional and lesional recovery, and sensory-motor reeducation. Careful positioning of the joint and muscles may prevent articular deformities and muscle shortening, and helps to avoid compression pathologies of the nerve. Splinting may be useful, taking into account the limitations and contraindications reported in our earlier discussion of splinting and prosthesis. Atrophic-degenerative muscle changes may reduce the potential for full rehabilitation even if regeneration occurs.<sup>73</sup>

### Electrical Stimulation

Electrical stimulation in denervated muscle of rabbit seemed to open a new frontier for the treatment of peripheral nerve lesions<sup>74</sup> but in a recent review of the use of electrical stimulation of denervated muscle, Herbison et al<sup>75</sup> concluded that the use of electrical currents in improving reinnervation is questionable and remains a matter of debate.<sup>76,77</sup> Electrical currents have been reported to increase ROM, together with hot packs, splinting, and gentle stretching, in a case of linear scleroderma with hemiatrophy and a severe knee flexion contracture.<sup>78</sup>

### Passive ROM

Passive mobilization has been shown to positively influence reinnervation and the sprouting process after nerve crush injuries in animals.<sup>79</sup> It influences collagen fibers, which are more easily elongated at temperatures of 40°C to 45°C.<sup>39,40</sup> This evidence suggests that heating collagen fibers by means of ultrasound might be used in SSc patients before stretching as well as passive ROM exercises. Passive or assisted movements are used when a severe motor impairment is present.

### Reeducation

Reinnervation may occur even after a long period. The maintenance of adequate sensory stimulation may assist sensory recovery. Disuse provokes receptor degeneration, whereas continuous receptor stimulation induces the formation of new receptors.<sup>80</sup>

### ERGONOMIC AND WORKING INTERVENTIONS

Scleroderma has a peak incidence during the highly productive decade of a patient's working life (35 to 44 years of age).<sup>81</sup> It is more common and starts earlier in black people compared with Caucasians and in women compared with men, with the probability of surviving for 7 years being 87% for limited sclerosis and 72% for diffuse disease,<sup>81</sup> with some patients surviving as long as 15 years.<sup>82</sup> Ergonomics and working interventions must be individualized and match practical and economic factors with the evaluation of the patient to enhance his or her compliance with the treatment of the disease.

Occupational and environmental influences, such as exposure to cold, have been reported to be clearly linked with SSc (Raynaud's phenomenon),<sup>83</sup> while others such as silica dust, benzene, urea formaldehyde, appetite suppressants, other drugs, and silicone breast implants have been reported to play a possible complementary role in influencing susceptibility to SSc.<sup>84</sup> When

reintroducing the patient to his or her former working place, cold-induced vasospastic attacks as well as other occupational and environmental influences should be also taken into account and avoided with proper educational approaches.

Ninety percent of SSc patients complain of loss of hand grasp ability because of skin retraction and musculoskeletal problems.<sup>42</sup> Factors such as puffy fingers, calcium deposits, and wrist extension reduction have been identified as risk factors for late development of a high level of hand disability.<sup>85</sup> Treatments started during the early phase of the disease may help to prevent the extreme disability seen in untreated patients. Flexion contractures of the proximal interphalangeal joints may limit tip-to-tip prehension or complete grasp and lead to weak lateral pinch, resulting in decreased dexterity. As a result, patients cannot perform manual activities that require grasping or pinching, and many daily living activities and work capacity are frequently impaired.

Because of morphologic changes, a sclerodermic hand may not show grasp pattern comparable to the normal prehension pattern.<sup>86</sup> A qualitative examination should be performed. It may also be helpful to use a quantitative classification of hand usage specific for SSc patients because of the variable time course of the hand involvement through the clinical phases (edematous/atrophic) of the disease. Little data exist in the literature on this subject. The assessment of general physical disability<sup>87</sup> and grasp pattern<sup>86</sup> has been proposed in SSc patients. However, no data on ergonomic and working interventions are available in the literature on SSc, nor have such strategies been validated; thus, SSc patients are usually treated with protocols proposed for hand stiffening or originally tailored for burns and osteoarthritis.<sup>88</sup>

Rehabilitation cannot stop the pathogenic progression of SSc. Nevertheless, exercise performed on a regular basis may combat contractures. Rehabilitation programs also may reduce social handicaps in the family and the workplace, and improve general lifestyle and activities of daily living. Because psychological adjustments to the illness can be influenced by factors such as functional ability, social support, and attitude toward the severity of the disease progression,<sup>90,91</sup> more attention should be drawn to these problems, to develop an integrated approach to the rehabilitative treatment of SSc.

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