

## STRATEGII DE RECUPERARE A PACIENȚILOR CU SCLEROZĂ SISTEMICĂ

### REHABILITATION STRATEGIES IN PATIENTS WITH SYSTEMIC SCLEROSIS

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**Cuvinte cheie:** sclerodermie, fibroză, tehnici de reabilitare, exerciții

#### Abstract

Systemic sclerosis (SSc), a multisystem disease involving microvascular system and the connective tissue is still surrounded by many mysteries and its treatment is often unsatisfying. Although skin fibrosis is the hallmark of this disease, musculoskeletal involvement is common. The thickening of the skin and joint involvement can cause serious musculoskeletal deficiencies and significant physical disabilities. Physical therapy is helpful to improve joint mobility, skin elasticity, muscle weakness and the ability to perform daily tasks. In this paper we present current rehabilitation techniques used for musculoskeletal impairments in persons with scleroderma. The most common rehabilitation techniques used for scleroderma patients are: connective tissue massage, hand and face stretching exercises, Kabat method, active exercises, joint manipulations and aerobic exercises.

#### Rezumat

Scleroza sistemică (SSc), o boala multisistemică care implică sistemul microvascular și țesutul conjunctiv este înconjurată încă de multe mistere, iar tratamentul ei este adesea nesatisfăcător. Deși afectarea cutanată reprezintă elementul central al acestei boli, mulți pacienți au acuze musculare și/sau articulare. Îngroșarea pielii și afectarea articulară pot provoca deficiențe musculo-scheletale și handicapuri fizice semnificative. Kinetoterapia este utilă pentru ameliorarea mobilității articulare, elasticității pielii, forței musculare și abilității de realizare a activităților zilnice. În lucrarea de față se prezintă tehnicile actuale de reabilitare musculo-scheletală utilizate la persoanele cu sclerodermie. Tehnicile cele mai utilizate în reabilitarea sclerodermiei sunt: masajul țesutului conjunctiv, exercițiile de întindere, metoda Kabat, exercițiile active, manipulările articulare și exercițiile de tip aerob.

#### Introduction

Systemic sclerosis (SSc) is a connective tissue disease of unknown etiology characterized by progressive fibrosis of skin and internal organs, including lung, heart, kidney and gastrointestinal tract [1,2]. Clinical manifestations of the disease are dominated by signs or symptoms of vascular dysfunction and excessive fibrosis with destruction of the normal architecture and organic tissue dysfunction.

Although skin lesion represents the primary element of this disease, patients may present muscular and/or joint damage [3,4,5].

The fibrotic process of SSc affects the tendons (causing tendon friction rubs), ligaments and joint capsules. The tendon friction rubs, common in patients with early-stage diffuse cutaneous SSc, are correlated with skin fibrosis and increased risk of scleroderma renal crisis.

They appear frequently at radiocarpal joint, ankles, but also in the subscapular bursa or at the thigh.

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The hallmark of SSc is induration and thickening of the skin. Hands are involved in most patients with SSc and they present contractures of the fingers and a claw-type deformity with metacarpophalangeal (MCP) extension, interphalangeal flexion and thumb adduction [4,5]. Moreover, the hand deformities and the limitations of finger movements have a significant impact on hand functionality.

On the other hand, tendinitis may cause severe pain and contributes to the development of tendon contractures. Swelling of the tendons and periarticular tissues in the wrist leads to medial nerve compression (carpal tunnel syndrome).

Arthralgia appears in the early stages of the diffuse form of SSc and may be confused with that of rheumatoid arthritis. Sometimes, the first sign of the disease is a symmetrical, seronegative polyarthritis, anodular and non-erosive [6]. Joint contractures associated with skin sclerosis contribute to reduced joint mobility.

In its evolution, resorption of the distal tufts of the digits (acro-osteolysis) is frequent and is due to inadequate vascular intake necessary for bone viability. Other sites of bone resorption include the mandible and the ribs.

Although vascular and fibrotic lesions of the organs are known to increase mortality and morbidity, musculoskeletal conditions also lead to significant physical disabilities [7,8].

Until now, there have been few clinical studies that evaluate the efficiency of rehabilitation techniques in SSc.

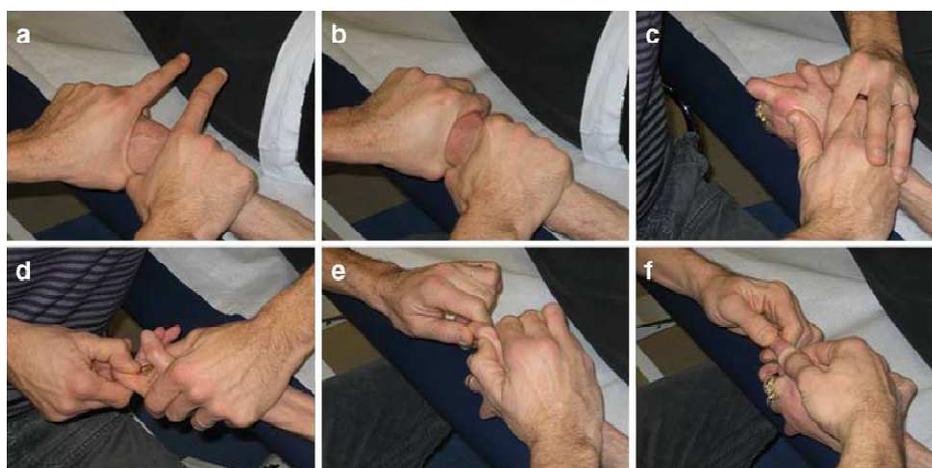
**The purpose** of this paper is to review existing evidence on the effectiveness of rehabilitation techniques in scleroderma patients.

### **The efficiency of rehabilitation techniques in sclerodermia**

There are a few randomized clinical studies that refer to the efficiency of rehabilitation techniques in systemic sclerosis. Additionally, these studies involved a small number of patients which have been followed up for a *short* period of *time* [9,10,11].

A recent study suggested that a combined rehabilitation program based on connective tissue massage, wrist joint manipulation (Mc Mennell) and home exercises was efficacious for hand involvement. Conjunctive tissue massage is a manual technique which improves local blood flow, elasticity of the connective tissue structures and allows its mobilization by stretching. After examination of the tissues in order to detect the presence of Raynaud phenomenon or digital ulcers, the treatment includes massage of forearm and hand, which lasts about 10 minutes/ limb [11,12].

The McMennell joint manipulation is a technique that allows regaining the “joint play”, which refers to normal movements between joint surfaces on multiple planes necessary for joint mobility [11] (Fig. 1). Treatment starts with wrist manipulation and continues with that of the metacarpophalangeal and interphalangeal joints (15 min per side).



**Fig. 1** Mc Mennell wrist manipulation (Bongi SM et. al)

S.M. Bongi et al. noticed that connective tissue massage associated with wrist manipulation of 40 scleroderma patients was effective for improving hand function and quality of life [11].

Also, the authors recommended a home daily exercise program consisting of active *range of motion exercises* for fingers, terminal and subterminal pinches, but also, thumb opposition movements. Wrist movements (flexion, extension, ulnar and radial deviation) and forearm pronation and supination were suggested [11].

Facial involvement in scleroderma patients causes not only functional impairments but also aesthetic modifications which patients find hard to accept. The fibrosis of facial soft tissues confers an inexpressive appearance (byzantine icon) with disappearance of folds and wrinkles, thin nose and a reduction mouth opening (microstomia). It is important to extend the face massage to the neck and adjacent regions.

In addition, the Kabat method has been recommended for the stimulation of the orbicularis oris, zygomaticus, nasalis, buccinator and lip levators muscles. A 9-week study conducted on 20 scleroderma patients analysed the efficiency of a combined rehabilitation program based on facial connective tissue massage, Kabat's technique and specific physical therapy exercises (active and passive exercises for temporo-mandibular joint). All patients underwent a daily home exercise program (opening and stretching of the mouth), mimic exercises for the facial muscles. At the end of the program, all patients recorded a significant improvement in the mouth opening, Rodnan skin score and the MHISS scale (Mouth Handicap in Systemic Sclerosis). This intervention did not record any improvement in the quality of life, respectively, in SF36 and HAQ scores [13,14]. Other studies demonstrated that aerobic training contributes to a increase in cardio-respiratory parameters in scleroderma patients. Training sessions consisted of moderate aerobic exercise on a treadmill, stationary bicycle, walking, swimming and aerobic dancing. However, few data are available in the literature regarding the efficacy of such interventions [15,16]. Antonioli et al. recommended an individualized program based on aerobic exercises, respiratory exercises and passive extensions of the hand. At the end of the observed period, patients recorded a better exercise tolerance, which was suggested by the reduction of the heart and dyspnoea rate. There has also been noted an improvement in hand mobility and in the quality of life [15].

Additionally, Oliviera et al. found in scleroderma patients a significant increase in VO<sub>2</sub> max after completing an 8-week aerobic training program. This improvement was not statistically significant [16].

### **Discussion**

Although reduced in number, most studies have underlined the importance of rehabilitation regimes in musculoskeletal involvement in SSc and have recommended connective tissue massage, passive extensions, active exercises and joint manipulations.

After reviewing the studies evaluated in this paper, we can notice that connective tissue massage, active and passive exercises associated with wrist manipulation are effective in increasing joint mobility, functional capacity and quality of life in SSc [11,12,13,14].

Aerobic training exercises are beneficial in improving cardiovascular function, endurance and quality of life; these being more associated with the improvement of the pulmonary function than with the muscular function [15,16].

### **Conclusions**

Conclusions must carefully be interpreted, because of the limited number of studies and small number of evaluated patients. Systemic sclerosis remains a challenging disease to treat, with the potential for severe morbidity and disability. Additional randomized clinical studies are required to further evaluate the validity of rehabilitation techniques.

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