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Nonpharmacological Treatment in Systemic Sclerosis

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Abstract

Systemic sclerosis (SSc) is a rare, severe connective tissue disease without available universal treatment. Organ involvement increases the mortality of this disease; nevertheless, skin involvement and the involvement of muscles and joints lead to loss of function, disability, and reduced quality of life.

Our experience and data from the literature suggest that physical activity interventions are an essential and integral part of treatment of patients with SSc. The limitation on most of the small number of existing studies aimed at nonpharmacological treatment for patients with SSc includes suboptimal methodological quality, low number of patients, lack of a control group or monitoring (follow-up), and short-term intervention.

The purpose of this chapter was to introduce the few studies that have examined this issue and to show that nonpharmacological therapy for diseases such as systemic sclerosis has been the focus of some research teams in recent years—the studies of which have demonstrated safety and benefits of these approaches.

Keywords: systemic sclerosis, function, disability, rehabilitation, physiotherapy, nonpharmacological interventions

1. Introduction

In the last few years, increased attention has been focused on nonpharmacological interventions in rare rheumatic diseases such as systemic sclerosis (SSc). However, there is still an unmet need to ensure medical awareness of the importance of physical therapy among patients, health professionals, rheumatologists, and other specialists to implement these nonpharmacological interventions into the daily care of patients with SSc.

Currently, physical therapy is considered a process by which patients try to restore and maintain their optimal physical, mental, social, occupational, and emotional state. It is therefore a comprehensive approach to the patient, which includes physical activity, as well as the principles of secondary prevention and healthy lifestyles, which are based on the patient's collaboration, activity, and

compliance. It is beyond doubt that physical inactivity is a major risk factor for both cardiovascular and respiratory systems, but also for functional limitations and reduced quality of life.

To acknowledge the importance of prevention and adherence to healthy lifestyle even before the onset of the disease, we can draw inspiration from the results of a recently published study on another, less rare, rheumatic disease, rheumatoid arthritis (RA): about the influence of environmental factors (e.g., the intensity and frequency of physical activity, of smoking, and different exposures) showing that patients with regular physical activity before the onset of RA suffer from a milder form of the disease and that obesity reduces the chance of achieving a good response to standard pharmacological approaches in RA [1, 2].

2. Nonpharmacological interventions in patients with systemic sclerosis

Tissue fibrosis is the dominant feature of SSc and affects both the skin and internal organs—especially the lungs, heart, and gastrointestinal tract [3]. While visceral involvement is responsible for increased mortality, musculoskeletal involvement (e.g., arthralgia/arthritis, flexion contractures, and muscle weakness) and skin involvement (skin thickening and tethering) pose a significant burden for the patient in terms of reduction of functionality, quality of life, and ability to work or perform activities of daily living [4].

Current recommendations for the treatment of SSc include symptomatic therapy of individual organ manifestations and complications of SSc. However, to date, we have no effective treatment of tissue fibrosis, for example, skin, lung, heart, and gastrointestinal tract. Nonpharmacological management of patients with SSc with the main focus on the musculoskeletal system and the affected skin may be useful to reduce disability and improve quality of life and contribute to reducing the burden of disease on the organism. Nevertheless, the major limitations of most of the small number of existing publications focused on physical interventions in patients with SSc include the diversity of interventions, small cohorts of patients, the absence of a control group, randomization and/or a follow-up, and a short duration of intervention [3].

The most frequently investigated nonpharmacological interventions include commonly used methods such as paraffin, manual lymphatic drainage, soft tissue techniques, stretching exercises for hands and face, massage, exercises to maintain range of motion in joints, mobilization, aerobic exercise, and strength training. Other studies focus on occupational therapy, physical therapy, sleep disturbances, sexual function, and depression.

The following paragraphs will provide an overview of the results of the available publications, which are divided according to the nature of the intervention.

2.1. Interventions using paraffin

One of the methods frequently used in physiotherapy of rheumatic diseases is paraffin treatment. Paraffin solidifies at a temperature of 52–62°C and yet gives latent heat. In order to utilize this heat, paraffin must be applied to the body in a fluid state and allowed to solidify in

the course of application. There are a few studies of paraffin in SSc patients: two randomized controlled studies (RCTs), one nonrandomized controlled trial and a series of three individual case studies.

The RCT by Sandqvist et al. evaluated the difference in intervention outcomes in 17 patients with SSc who underwent therapy with paraffin in combination with exercise on one hand chosen by random, and on the other hand received only exercise. Interventions continued daily for 1 month. Hand function was assessed at the beginning of the intervention and after the first month according to hand mobility and strength, the perception of pain, stiffness and elasticity of the skin. The authors demonstrated a significant improvement in flexion and extension of the fingers, thumb abduction, plantar flexion, perceived stiffness, and elasticity of the skin in patients who underwent paraffin intervention in combination with exercise compared to baseline levels. Furthermore, significant improvement in hand function (specifically in extension deficit, perception of stiffness, and elasticity of skin) was significantly greater in the hand that was treated with paraffin in combination with exercise than in the hand treated with exercise only [5].

In another RCT study [6], 16 patients with SSc underwent 12 times paraffin wrap therapy and then were randomly assigned to the intervention and control groups. The intervention group continued with paraffin therapy for 3 months, and the control group did not. Skin thickness decreased in all patients after 12 sessions of paraffin treatment. However, at the end of 3 months therapy in the intervention group, there was no significant difference between the intervention and control group [6].

One of the first studies, a nonrandomized controlled trial by Askew et al., recruited 10 patients with SSc and examined the effectiveness of once performed interventions including paraffin, friction massage, and exercise maintaining range of motion in the joints of the wrist and hand. Compared with seven patients with SSc, who did not attend any sessions, the interventions lead to significant improvements in the articular range of motion, skin elasticity, and overall function of the hand [7].

In a series of three individual case studies [8], respondents conducted active exercises with their hands after paraffin wrap 5 times per week for 8 weeks. The authors detected clinically significant improvements in both body function/structure measurements of hand function and in their ability to participate in activities in all participants [8].

2.2. Orofacial exercises

Orofacial region has many functions. Primarily, it ensures food intake and contributes to breathing. Coordinated interplay of activity of facial muscles allows for expression of verbal, articulate speech. In the orofacial region, we find more than fifty facial muscles that enable people to communicate nonverbally, to express their feelings. No other part of the human body possesses such a social function as the orofacial region. Through this region we communicate, we speak to others, express our feelings, and create new relationships. In patients with SSc, tightening of the skin and muscle atrophy in orofacial region lead to loss of facial expressions

and an appearance of a mask-like face. The affected orofacial region with skin thickening often contribute to inability of mechanical mouth opening, microstomia, which along with other symptoms, such as dry mouth, can lead to dental and mouth hygiene issues, and hampered alimentation.

In the RCT by Yuen Hon et al., the authors studied the effect of orofacial home exercise program to increase the orifice in adults with SSc. The study included 48 adult patients with SSc who were randomly divided into two groups and examined for oral aperture at baseline, 3-month, and 6-month intervals. The intervention group underwent a multifaceted oral health intervention including adaptive oral hygiene devices, instruction and demonstration on the use of the devices, brushing teeth for 2 min and flossing twice a day for 6 months. The control group received the usual dental care. Participants with orifices <40 mm in the intervention group underwent orofacial exercise program, which included manual mouth-stretching and oral augmentation exercises with a wooden stick, twice a day with a total of 6 minutes for 6 months. The results showed a significant increase in oral orifice in patients of the intervention group at 3 months, but not after 6 months evaluation. However, compliance with the program was low (48.9%), which could distort the results of this study [9].

An RCT by Maddali-Bongi et al. evaluated the effectiveness of an intervention program based on a combination of the Kabat's technique (proprioceptive neuromuscular facilitation, PNF), connective tissue massage, and kinesiotherapy specially developed for the face of SSc patients. Out of the 40 recruited SSc patients, 20 patients were randomly selected into the intervention group and underwent the intervention program for 9 weeks, 2 times a week for 1 h with a home exercise program. Twenty patients were randomly assigned into the control group, which underwent home exercise program only. Patients of both groups were evaluated at baseline, after 9 weeks and after 9 weeks of follow-up in terms of quality of life and health, stiffness of the facial area, opening of the mouth, and oral handicap. At the end of treatment, both groups showed improved orifice. However, in the follow-up period, improvement persisted only in the intervention group. The facial score in the intervention group improved both at the end of therapy and follow-up period, whereas in the control group, there were no changes. According to the results of quality of life and health status, no effect of therapy was detected in any group. A significant improvement was also found in oral handicap at the end of therapy in the intervention group, whereas in the control group, no changes were observed [10].

In an observational study by Poole et al., the authors investigated the improvement in oral hygiene in SSc patients who underwent a program involving structured oral hygiene and exercises for the face and hands. The authors recruited 17 patients with SSc who previously underwent basic dental checkup, including examination for decayed or missing teeth, calculus, sites that bleed upon probing, measures of oral aperture, and the patient hygiene performance index. The study also evaluated upper limb function involving strength, joint range and maneuverability. Participants underwent a structured exercise program including education on brushing and flossing techniques used, hand exercise of three series with five repetitions, once daily stretching, once daily facial exercise of five times stretching 3–5 seconds, individually adjusted dental equipment, and a 6-month supply of dental products. This program was

performed one or two times a day for 6 months. At the end of the 6-month intervention, there were a significant improvement in oral hygiene and a significant decline in the number of teeth with calculus and a reduction in bleeding gums. There were no differences in any of the upper extremity measures or oral aperture [11].

Another observational study by Pizzo et al. examined the effects of nonsurgical exercise program in 10 patients with severe SSc-associated microstomia (maximal mouth opening ≤ 30 mm). Patients were instructed to perform an exercise program, which included mouth stretching exercises for at least 15 minutes, twice a day, and oral augmentation exercise with a stick of soft wood daily during 18 weeks. After 18 weeks, the effect of exercise was evaluated measuring the maximal mouth opening. The exercise program improved mouth opening of all subjects. At the end of the period of 18 weeks, all patients noted that eating, speaking, and oral hygiene were easier [12].

In 2009, Maddali-Bongi et al. performed two other RCTs that examined the effect of specialized intervention programs and pointed out their safety and improvements in the hands and mouth (facial). These studies will be discussed in the following sections of this chapter.

At this point, it is worth to mention a study that can be useful for dentists treating this area in such a rare disease as SSc. Alantar et al. reviewed the literature and collected expert opinion for the design of preventive and curative treatment of oral and dental disability in patients with SSc. The authors point out that prevention of oral and dental complications is a major issue in patients with SSc. Dental treatment should be tailored to limitations in mouth opening, disease severity, and on-going treatments [13].

2.3. Comprehensive physical therapy for physical and/or psychological functioning

Physiotherapy is a vital part of nonpharmacological approaches, which deals with the prevention and treatment of disorders of the musculoskeletal system. Based on a comprehensive approach, physiotherapy pays special attention to the life situations, in which the person is. The aim of comprehensive physiotherapy is to find the best way to mitigate or eliminate health problems and to maintain the current effect in a long term. There are several studies available in this area of research.

In 2009, Maddali-Bongi et al. performed two RCTs that examined the effect of a specialized intervention program in the area of hands and mouth (face). The first study aimed to evaluate the effectiveness of the intervention program based on a combination of connective tissue massage and joint manipulation according to Mc Mennell specially designed for the hands of SSc patients. Of the 40 recruited SSc patients, 20 patients were randomly assigned to intervention and underwent the exercise program for 9 weeks, twice a week for one hour, and a home exercise program. Twenty patients were randomly assigned to the control group which underwent only the home exercise program. Patients of both groups were evaluated at baseline, after 9 weeks and after 9 weeks of follow-up in terms of quality of life, hand involvement, hand functional disability, and the measurements of range of motion. The intervention group demonstrated a significant improvement in all measured parameters, whereas the home

exercise program in the control group improved just closing the hand into a fist at the end of therapy [14].

The aim of the second study was to evaluate the efficacy of a specific and comprehensive rehabilitation program tailored for patients with SSc. Hand involvement was treated with a combination of connective tissue massage and Mc Mennell joint manipulation. Manual lymph drainage was applied if patients had edematous hands. Face involvement was treated with a combination of Kabat's method, connective tissue massage, and kinesiotherapy. Other techniques used in the study included hydrokinesiotherapy (for patients without ulcers), land-based rehabilitation (for patients with ulcers), and respiratory rehabilitation exercises. The authors recruited twenty patients with SSc, who were randomly divided into two groups and evaluated at baseline and after 9 weeks in terms of quality of life, health, hand function, range of motion, water volumetric test, orifice, Duruoz scale, and purpose-built-questionnaire for hand and face Involvement. The intervention group was evaluated also after 9 weeks of follow-up. The intervention group 10 patients underwent a specific rehabilitation program, one or two times per week for 9 weeks. The control group underwent only educational advice and medical information for patients with SSc. At the end of treatment, patients in the intervention group improved in all measured parameters. In follow-up, orofacial function and mobility as well as general health condition were partially lost, whereas only the mobility and function of the hand were preserved. No changes in the examined parameters were observed in the control group [15].

In a controlled clinical study by Antonioli et al., the authors evaluated the effect of a specialized individual rehabilitation program that consisted of warm-up and cool-down exercises, training of motor functions, diaphragmatic breathing, controlled coughing exercises, treadmill, freewalking, finger stretching, and occupational therapy for 2 weeks of daily 30-minute sessions (10 sessions in total). Physical therapy was also prescribed to 13 patients with joint-related problems. At-home exercise program was prescribed on days when the intervention program was terminated. Patients were reevaluated after 2 and 4 months. The control group consisted of 17 patients who did not receive any form of rehabilitation. The authors evaluated aerobic capacity, hand function, limitations in activities of daily living, quality of life, and skin-related problems. At the end of the 4-month period, the examiners observed an improvement in the perception of quality of life, better exercise tolerance, and better mobility of hand in a significant number of patients with SSc [16].

Since patients with SSc reported great need to receive support and education regarding their disease, Kwakkenbos et al. developed a short, group-based psychoeducational program, and evaluated it in an observational study using pre- and posttest design. Participants filled out a questionnaire assessing their physical and mental condition. Subsequently, patients were asked to evaluate the program content using a questionnaire. Completely filled questionnaires were available for data analysis from 41 patients. The authors detected a high satisfaction of patients with the program, smaller helplessness after the intervention, and a higher acceptance of patients' limitations. However, no differences were observed in depressed mood or in physical function. Thus, this psychoeducational program contributed to meeting the reported needs of patients and to improving the care of patients with SSc [17].

In an RCT by Schouffoer et al., the authors compared the effectiveness of a 12-week multidisciplinary nursing program with usual outpatient care for SSc patients. The multidisciplinary nursing program included individual treatment goals once a week for 12 weeks, general exercises, hand/mouth exercises, educational sessions under supervision of a local physical therapist, and a home-based exercise program at least 6 days a week for 12 weeks. Twenty-eight patients with SSc were randomly selected into the intervention group (twenty-five of them completed the treatment), and twenty-five patients were randomly assigned to the control group. The examiners evaluated the hand function, grip strength, maximal mouth opening, aerobic capacity, gait, quality of life, and health status at baseline, at the end of treatment (12 weeks), and at the end of follow up (after 24 weeks). Results showed that the 12-week daily multidisciplinary care program was more effective than regular outpatient care with regard to significant improvement in grip strength, mouth opening, gait, and quality of life after 12 weeks, whereas the results in other followed parameters showed no difference between the intervention and control group. At the end of the follow-up period, after 24 weeks, only improvement in grip strength was maintained [18].

Our own preliminary results (unpublished data) from an ongoing single-center controlled study further support the role of intensive physiotherapy in patients with SSc. Our project aimed to address some limitations of existing studies and to assess the efficacy of a long-term (24-week intervention, 24-week follow-up), intensive (1 h physiotherapy + 0.5 h occupational therapy twice weekly, and home-exercise for 0.5 h five times a week), tailored physiotherapy program on function and impairment of the hands and face, and quality of life and disability in patients with SSc. We recruited 27 SSc patients into the intervention group, which underwent the 24-week intervention, and 29 SSc patients into the control group, which received an education and textbook on home exercise at baseline only. The intervention program unit comprised several physiotherapy/occupational therapy techniques, such as warm-up with infrared lamp, manual lymphatic drainage, skin wrinkling, fascial techniques, postisometric muscle relaxation, joint mobilization, passive/active muscle stretching and exaggerated facial mimicry training, soft ball facilitation techniques, sensoric stimulation with terabeans and therapeutic plasticine. Our program not only prevented the natural course of progressive deterioration of function of hands/mouth (observed in the control group) but also led to a significant improvement in monitored parameters (e.g., delta finger to palm, hand/finger grip strength, HAMIS-Hand Mobility in Scleroderma, interincisor/lip distance), which was clinically meaningful in a substantial proportion of patients.

2.4. Stretching interventions

Stretching includes special exercises leading to increased mobility of the body that can address a variety of health-related issues. It is a method, which can gently prepare your muscles to increased physical stress, but also to very ordinary functioning of our body during daily life activities. Stretching increases performance, maintains the elasticity of muscles and tendons, prevents muscle imbalance, improves joint mobility, and helps to maintain correct posture, proper breathing, and efficient and economical movement. Furthermore, stretching reduces muscle tone and brings overall relaxation, provides prevention from muscle- and joint-related injuries, improves response and readiness, and increases resistance to fatigue. To sum up the

benefits of stretching, this technique maintains overall good physical and mental condition. For patients, most of all, it is a method/technique that maintains the elasticity of tendons, muscles, and improves joint mobility. Therefore, not only in healthy individuals but also in patients with SSc, it is the only way to recover, or aim to restore physiologic length of shortened muscles.

Mugii et al., in their observational study, compared the effect of autostretching of individual fingers in patients with SSc. Forty-five patients with SSc were instructed how to autostretch their fingers. The individual fingers were held with the other hand in the stretched position for 10 s, and this was repeated 3–10 times a day. To evaluate the effect of autostretching intervention, passive range of motion of the fingers was evaluated using a goniometer during the first visit, after 1 month and 1 year of the autostretching program. Quality of life was also assessed at the first visit and after a year using the Health Assessment Questionnaire (HAQ). The authors demonstrated a significant improvement in the overall range of passive joint motion of each finger after 1 month of autostretching, which was further improved or maintained even after 1 year. Although the range of motion of the fingers was lower in patients with diffuse cutaneous (dc) SSc than in patients with limited cutaneous (lc) SSc during the first visit, the examiners detected a significant improvement regardless of the disease duration or severity of skin sclerosis. The study team noted that stretching your fingers can improve the function of fingers, because HAQ score assessing the hand function activities, such as eating and grasping, decreased significantly [19].

An RCT by Vannajak et al. recruited 28 patients with SSc who were divided into two groups of 14 patients. Both groups received the same daily home treatment with one difference: one group with gloves and the other group without gloves. The 2-week daily home intervention included a Thai massage, stretching exercises and heating. The authors assessed the function of the hand using Hand Mobility in Scleroderma (HAMIS). Both groups showed a significant improvement in the function of the hand. Wearing gloves, however, lead to greater thumb mobility [20].

2.5. Aerobic training and combined endurance/resistance training

The rehabilitation of patients with impaired cardiovascular system is based on endurance, aerobic training, that is, a prolonged dynamic loading at or below the anaerobic threshold. Regular aerobic exercise improves the efficiency of the cardiovascular system (lungs, heart, and circulatory system), and regular endurance and strength training induce characteristic changes that lead to improved physical condition. These changes represent the training effect and help to achieve greater physical load with lower heart-rate responses.

Pinto et al. in their observational study focused on the effects of 12 weeks of combined resistance and aerobic training program (concurrent training) in 11 SSc patients. Supervised concurrent training session consisted of treadmill warm-up (5 min) followed by resistance training (30 min), treadmill aerobic training (20 min), and stretching exercises (5 min). Resistance training included five exercises for the main muscle groups: bench press, leg press, latissimus pull down, leg extension, and seated row. The concurrent training program was performed in one hour sessions, twice per week for 12 weeks. The study showed that

concurrent 12-week training program was safe and significantly improved muscle strength, function and aerobic capacity in SSc patients [21].

The aim of a pilot study by Alexanderson et al. was to determine the effect of 8-week intense aerobic and endurance training program in four patients (three women and one man) with SSc with 50–100% of forced vital capacity (FVC). The authors detected significantly improved muscular endurance in three participants and significantly or clinically significantly improved aerobic capacity in two participants [22].

Oliveira et al., in their prospective nonrandomized controlled trial, examined whether patients with SSc have reduced exercise capacity compared to healthy individuals, and whether aerobic exercise is safe for patients with SSc and improves their aerobic capacity. In this study, 7 patients with SSc (without interstitial lung disease) and 7 healthy controls underwent an 8-week aerobic exercise program of moderate intensity. Participants were evaluated by cardiopulmonary stress test before and after 8 weeks of training, and examined for blood lactate concentration and oxygen saturation at each visit and at rest, and quality of life. Patients with SSc and healthy controls underwent 40-min training twice weekly. The first five minutes of the exercise program were reserved for the warm-up, when the speed was gradually increased until a target heart rate. The last five minutes of exercise were represented by the cool-down with slowing down the speed to a standstill. In the first session of the program, the participants underwent 15 min of aerobic exercise at their respective target heart rate. In the second session, aerobic training was increased to 20 min, in the third session to 25 min, and in all subsequent sessions to 30 min. Both groups showed a significant improvement in peak VO₂ but with no significant difference between the study groups. Furthermore, both groups improved in exercise intensity which was documented by significantly increased peak lactate concentration in blood. No significant changes in SSc skin score or quality of life were detected [23].

2.6. Interventions on lower extremities

In SSc commonly affected area of the body is the lower limbs, especially their muscular strength and function, the loss of which has an impact on quality of life of patients with SSc. Therefore, Lima et al. in their cross-sectional study assessed peripheral and respiratory muscle strength in patients with SSc and examined their correlation with the 6-minute walk test and quality of life. Secondary aim of this study was to characterize the nutritional status, lung function, functional ability and quality of life of SSc patients compared with healthy controls. The study included 20 patients with SSc and 20 healthy control subjects. Results showed that patients with SSc have significantly reduced respiratory muscle strength, decreased strength in m. quadriceps femoris, and increased fatigability compared to healthy controls [24].

2.7. Self-management programs

Poole et al., in 2013 and 2014, assessed their two self-management programs and their impact on self-management for patients with SSc. The first program (2013) consisted of a workbook and a DVD (sent by postmail) that provided information on medical aspects of the disease, dysphagia, fatigue management, advocacy, activities of daily living, oral hygiene, skin and

wound care, psychosocial changes, exercises, and other features of the condition. Forty-nine participants evaluated the effect of the program providing feedback through series of questions, filling out the protocol on health condition, and through their participation in the evaluation interview. The results of the study showed a decrease in depression, fatigue and pain, and an improvement in the hand function, self-efficacy for controlling pain, and self-efficacy "other." However, the only statistically significant change was in self-sufficient pain control. Moreover, the study showed that the program was easy to use and should help individuals with SSc in self-sufficiency [25].

In another pilot study (2014), Poole et al. assessed the effectiveness of internet-based self-management program for patients with SSc. Participants logged on to a password-protected web site and completed the program, which consisted of 10 modules (e.g., coping and body image/appearance, fatigue and energy conservation, self-advocacy), an exercise video, learning activities, worksheets, and resources. Participants proceeded through the modules at their own pace over 10 weeks. Participants were encouraged to log on to the discussion board, participate in an interactive component of the web site, and respond to questions posted for each module. Participants completed pre- and postintervention questionnaires on perceived self-efficacy, health efficacy, ability to manage care, functional disability, depression, pain, and fatigue. They also completed an eight-question evaluation form regarding satisfaction with the web site, program content, discussion boards, and learning activities. Sixteen participants completed the study and postintervention measures. The authors demonstrated significant improvements in mean scores for ability to manage health care and efficacy, and significant decreases in fatigue and depression. Self-efficacy was improved as well, but not significantly [26].

2.8. Splinting interventions

Functional impairment of the hand in SSc patients is one of the major problems encountered in rehabilitation practice. Research in this area also includes splinting interventions of the hand and wrist, which aim to facilitate restoration of the lost function. In a 2-month study by Seeger et al., the authors investigated splinting effect on the secondary contractures in 19 patients with SSc. Specifically, whether dynamic splinting can improve flexion contractures in the proximal interphalangeal joints (PIP). Of the eight patients who completed the study, only one had significant improvement in range of motion as a result of PIP joint splinting, which shows that the study failed to demonstrate the use of splints as a suitable method to maintain PIP extension compared with the control hand [27].

2.9. Physical therapy

In addition to the abovementioned list of interventions, there are few studies that evaluate the effectiveness of some other nonpharmacological approaches in SSc including the use of physical therapy, which is the practical application of physical treatments procedures and methods on living organism.

Manual lymphatic drainage (MLD) stimulates the lymphatic system and reduces swelling. It is one of the main components of a combined physical therapy, which is used for the treatment of

lymphedema [28]. It is a technique that applies a force to the interstitial fluid and proteins within the initial lymphatics, thereby shifting toward collaterals and/or normally functioning lymphatics [29]. This technique not only directly softens the tissues, but perhaps also removes excessive local fluid. Therefore, it is speculated that the MLD technique should modify the mechanical properties, such as strain, of the skin and subcutaneous tissue. Strain is defined as deformation of a solid due to stress and is considered to be one of the measures representing tissue hardness [30].

The effect of manual lymphatic drainage to reduce swelling, improve the functionality of the hand and the perceived quality of life (QoL) in patients with SSc in edematous stage of disease was assessed in an RCT by Bongi et al. The study recruited 35 patients with SSc who were in the edematous disease stage (swelling of the hands), 20 of which were randomly assigned to the intervention group, and underwent MLD 1h once a week for 5 weeks according to the Vodder procedures, and 15 of which served as a control group. The study demonstrated a reduction in the hand volume, improved hand function, and perceived swelling and pain. Reduction in the volume and improvement in hand function were also maintained at follow-up. Furthermore, the authors detected an improvement in overall disability and quality of life in patients who received therapy. In the control group, no improvement was observed. The authors concluded that the application of MLD is effective in the treatment of the hand in edematous SSc by reducing hand volume, edema, and pain, and improving hand function and perceived quality of life. A study on a larger cohort of patients and with a longer follow-up is needed to assess the effect of MLD on different disease subsets as well, and to verify whether the technique could be of help in preventing the transition from hand edema to fibrosis [31].

Beyond examining the effects of MLD, researchers also investigated the effects of other types of physical therapy in the rehabilitation of patients with SSc.

Sallam et al. evaluated the impact of 14-day transcutaneous electrical nerve stimulation (TENS) of gastrointestinal (GI) acupoints on GI symptoms and quality of life in 17 SSc patients. This treatment significantly increased sympathetic and vagal activity in comparison with the basal value, lead to normalization of sympathovagal balance, and increased physical function score, which is closely related to the change in the sympathovagal balance. This provides the potential to treat the upper GI tract symptoms; however, further studies are needed to support these results [32].

In a pilot study by Tinazzi et al., the authors investigated the effect of extracorporeal shock wave therapy on the skin of 30 patients with SSc. This study was based on a hypothesis that the treatment effect of extracorporeal shock wave may decrease damage to the endothelial cells and skin fibrosis in patients with SSc. The authors demonstrated rapid and sustained reduction in Rodnan skin scores and a reduction in visual analog scale. Furthermore, an improvement in skin structure and vascularization was recorded 90 days after treatment. The number of endothelial progenitor cells and circulating endothelial cells increased 60 and 90 days after treatment, whereas serological biomarkers showed no differences before and after treatment [33].

Results of a prospective RCT by Sporbeck et al. point out the beneficial impact of biofeedback and deep oscillation (three times a week for 4 weeks) on Raynaud's phenomenon in patients with SSc [34].

In an observational study by Uhlemann et al., the authors evaluated the effect of ultrasound applied to the hands with an intensity of 0.6 W/cm^2 , for 6 minutes, 3 times a day for 6 days on the hand function and strength. Pain decreased in 18 of 24 patients. At the end of therapy, no increase in pain was observed, and the hand strength significantly improved in all patients [35].

Milačić et al. examined the change in volume of orifice before and after combined hyperbaric oxygen (HBO) therapy with facial physical therapy. Patients received HBO therapy ten times (2.0 ATA) and infrared phototherapy using Solux lamp 15 min before kinesiotherapy in front of mirrors daily for 10 days. This treatment targeted facial muscles. The results showed a statistically highly significant difference in improvement before and after treatment, and that use of HBO in the treatment of these patients has a significant role [36].

3. Conclusion

Chronic rheumatic diseases have a significant impact on the function, quality of life, and ability to work and represent a significant clinical and economic burden on healthcare systems. It is generally known that adequate physical activity is considered one of the adequate means to maintain or improve the quality of life, both physically and mentally. Nonpharmacological therapy plays a key role in the treatment of most rheumatic diseases. However, its effect for some of them has not been adequately studied or been little researched. A limitation of most of the small number of existing works aimed at nonpharmacological treatment for patients with SSc is low methodological quality, low number of patients, lack of a control group or monitoring (follow-up), and short-term intervention. The purpose of this chapter was to introduce the few studies that have examined this issue (**Table 1**) and to show that nonpharmacological therapies for diseases such as systemic sclerosis have been the focus of some research teams in recent years, the studies of which have demonstrated safety and benefits of these approaches.

Nonpharmacological therapy in patients with SSc includes, in particular, methods commonly used, for example, paraffin, manual lymphatic drainage, massage, stretching exercises to maintain range of motion in joints, mobilization, aerobic exercises, specially developed programs for the face and hands, self-management programs, splinting, and physical therapy. Other studies also investigated occupational therapy, sexual dysfunction, education, psychosocial status, and nutrition in patients with SSc.

Quality of treatment depends on the cooperation of a multidisciplinary team. A physician should properly choose the right medical treatment. A physiotherapist must select an adequate therapy to be individually tailored for a specific patient. An exercise program should be feasible even in the home environment, and patient's family or partner should get involved. Modifications to the home and work environments should be consulted with an occupational therapist. A psychologist or a psychosomatic medicine specialist can help not only the patient

Name of study	Author, year	Study design	Type of intervention	Description of intervention in intervention group	Description of intervention in control group	Number of individuals in intervention group	Number of individuals in control group	Duration of the intervention	Postintervention assessment	Results
Evaluation of paraffin bath treatment in patients with systemic sclerosis [5]	SANDQVIST, 2004	RCT	Paraffin	Paraffin in combination with exercise	Only exercise	17 SSc patients	17 SSc patients	1 month daily	After 1 month	The results showed that hand exercise in combination with paraffin bath seemed to be a suitable form of treatment in the effort to improve mobility and perceived hand stiffness
Paraffin hand bath for scleroderma [6]	PILS, 1991	RCT	Paraffin	12 times paraffin wrap therapy which continued for next 3 months	12 times paraffin wrap therapy	8 SSc patients	8 SSc patients	3 months	After 3 months	Skin thickness decreased in all patients after 12 sessions of paraffin treatment. However, at the end of 3 months therapy in the intervention group, there was no significant difference between the intervention and control group
Objective evaluation of hand function in scleroderma patients to assess effectiveness of physical therapy [7]	ASKEW, 1983	CCT	Paraffin	Paraffin, friction massage and exercise maintaining ROM in the joints of the wrist and hand	-	10 SSc patients	7 SSc patients	Once performed interventions	After 2 hours	Significant improvement in the articular ROM, skin elasticity and overall function of the hand
The effect of paraffin and exercise on hand function in persons with scleroderma: a	MANCUSO and POOLE, 2009	3x single case study	Paraffin	Active exercises with hands after paraffin wrap	-	3 SSc patients	-	5 times per week for 8 weeks	1 month and at 2 months after intervention	The results of this study suggest clinical efficacy of paraffin and exercises to

Name of study	Author, year	Study design	Type of intervention	Description of intervention in intervention group	Description of intervention in control group	Number of individuals in intervention group	Number of individuals in control group	Duration of the intervention	Postintervention assessment	Results
series of single case studies [8]										improve activity/ participation involving the use of the hands
Effect of orofacial exercises on oral aperture in adults with systemic sclerosis [9]	YUEN, 2012	RCT	Orofacial treatment	Multifaceted oral health intervention	Usual dental care	26 SSc patients	22 SSc patients	Twice a day with a total of 6 minutes for 6 months	After 3 months, and 6 months	A significantly larger increase in oral aperture for participants receiving the orofacial exercise program was found when compared to those in the usual care at 3 months, but not at 6 months evaluation. Participants' adherence rate to the exercise program was low (48.9%)
The rehabilitation of facial involvement in systemic sclerosis: efficacy of the combination of connective tissue massage, Kabat's technique and kinesitherapy [10]	MADDALI-BONGI, 2011	RCT	Orofacial treatment	Combination of the Kabat's technique, connective tissue massage, and kinesiotherapy specially developed for the face of SSc patients with a home exercise program	Only home exercise program	20 SSc patients	20 SSc patients	9 weeks, 2 times a week for 1 hour	After 9 weeks and after 9 weeks of follow-up	The combination of connective tissue massage, Kabat's technique, kinesiotherapy, and home-based exercises was more effective than a home exercise program alone in the rehabilitative treatment of SSc facial involvement
Oral hygiene in scleroderma: the effectiveness of a	POOLE, 2010	OD	Orofacial treatment	Structured oral hygiene and exercises for the face and hands	–	17 SSc patients	–	6 months daily	After 6 month intervention	There was a significant improvement in

Name of study	Author, year	Study design	Type of intervention	Description of intervention in intervention group	Description of intervention in control group	Number of individuals in intervention group	Number of individuals in control group	Duration of the intervention	Postintervention assessment	Results
multidisciplinary intervention program [11]										oral hygiene and a significant decrease in the number of teeth that bled on probing and in subgingival calculus
Effects of a nonsurgical exercise program on the decreased mouth opening in patients with systemic sclerosis [12]	PIZZO, 2003	OD	Orofacial treatment	Mouth stretching exercises for at least 15 minutes, twice a day, and oral augmentation exercise with a stick of soft wood daily	–	10 SSc patients	–	18 weeks	After 18 weeks	The exercise program improved the mouth opening of all subjects. At the end of the 18 week period, all patients commented that eating, speaking and oral hygiene measures were easier
Efficacy of connective tissue massage and Mc Mennell joint manipulation in the rehabilitative treatment of the hands in systemic sclerosis [14]	BONGI, 2009	RCT	Comprehensive physical therapy for physical and/or psychological functioning	Combination of connective tissue massage and joint manipulation according to Mc Mennell specially designed for the hands, and a home exercise program	Only home exercise program	20 SSc patients	20 SSc patients	9 weeks twice a week, 1 h per session	After 9 weeks, and after 9 weeks of follow-up	The combination of connective tissue massage, Mc Mennell joint manipulation and home exercise program was effective in the rehabilitative treatment of SSc hands. This combined treatment may lead to an improvement in hand function and QoL
Efficacy of a tailored rehabilitation	BONGI, 2009	RCT	Comprehensive physical therapy for	Combination of connective tissue massage and Mc	Educational advices and	10 SSc patients	10 SSc patients	One or two times per week for 9 weeks	After 9 weeks and after 9 weeks of follow-up	The association of disease-specific and global

Name of study	Author, year	Study design	Type of intervention	Description of intervention in intervention group	Description of intervention in control group	Number of individuals in intervention group	Number of individuals in control group	Duration of the intervention	Postintervention assessment	Results
program for systemic sclerosis [15]			physical and/or psychological functioning	Mennell joint manipulation (MLD if patients had edematous hands. Face involvement was treated with a combination of Kabat's method, connective tissue massage, and kinesiotherapy. Hydrokinesiotherapy (for patients without ulcers) or land-based rehabilitation (for patients with ulcers), and respiratory rehabilitation exercises)	medical information					rehabilitative techniques designed and tailored for SSc patients improved disability, HRQoL, hand and face disability and functionality, with its effects partially maintained at the follow-up
An individualized rehabilitation program in patients with systemic sclerosis may improve quality of life and hand mobility [16]	ANTONIOLI, 2009	CCT	Comprehensive physical therapy for physical and/or psychological functioning	Rehabilitation program that consisted of warm-up and cool-down exercises, training of motor functions, diaphragmatic breathing, controlled coughing exercises, treadmill, freewalking, finger stretching, and occupational therapy (physical therapy was also prescribed to 13 patients with articular problems) and home exercise program	–	16 SSc patients	17 SSc patients	2 weeks daily 30-minute sessions (10 sessions in total)	After 2 and 4 months	This study suggests that a significant proportion of patients with SSc experience an improvement in their perception of QoL, a better exercise tolerance, and a better hand mobility after a rehabilitation program consisting of a 2-week period of daily individual 30-min sessions of outpatient care, followed by at-home exercise program

Name of study	Author, year	Study design	Type of intervention	Description of intervention in intervention group	Description of intervention in control group	Number of individuals in intervention group	Number of individuals in control group	Duration of the intervention	Postintervention assessment	Results
Addressing patient health care demands in systemic sclerosis: pre- and postassessment of a psychoeducational group programme [17]	KWAKKENBOS, 2011	OD	Comprehensive physical therapy for physical and/or psychological functioning	A short, group-based psychoeducational program	-	41 SSc patients	-	The intervention was scheduled over 3 weekends	6 weeks after the intervention and 6 months postintervention	Patients reported less helplessness after the intervention, and higher acceptance of their limitations. However, no difference in depressed mood and physical functioning was observed. Patients reported high satisfaction with the content of the program
Randomized comparison of a multidisciplinary team care program with usual care in patients with systemic sclerosis [18]	SCHOUFFOER, 2011	RCT	Comprehensive physical therapy for physical and/or psychological functioning	Multidisciplinary team care program (1 day per week; individual treatments, group exercises, and group education)	Regular outpatient care	25 SSc patients	25 SSc patients	12 weeks	0, 12, and 24 weeks	In patients with SSc, a 12-week multidisciplinary day patient treatment program was more effective than regular outpatient care
The efficacy of self-administered stretching for finger joint motion in Japanese patients with systemic sclerosis [19]	MUGII, 2006	OD	Stretching	Autostretching (individual fingers were maintained in a stretched position using the opposite hand for 10 seconds and this was repeated 3–10 times)	-	45 SSc patients	-	1 year	After 1 month and 1 year	The total passive ROM was significantly improved in each finger after 1 month of finger stretching and 1 year after the first visit. Eating and gripping was also significantly improved
The short-term effect of gloving in	VANNAJAK, 2014	RCT	Stretching	The home program combined traditional	Same program	14 SSc patients	14 SSc patients	2 weeks daily	After intervention	Both groups showed a

Name of study	Author, year	Study design	Type of intervention	Description of intervention in intervention group	Description of intervention in control group	Number of individuals in intervention group	Number of individuals in control group	Duration of the intervention	Postintervention assessment	Results
combination with traditional Thai massage, heat, and stretching exercise to improve hand mobility in scleroderma patients [20]				Thai massage with stretching exercises and heat with gloves	without gloves					significant improvement in hand mobility after 2 weeks of daily home exercise program. Wearing the glove, however, resulted in better thumb mobility
Efficacy and safety of concurrent training in systemic sclerosis [21]	PINTO, 2011	OD	Aerobic training and combined endurance/resistance training	Combined resistance and aerobic training program (concurrent training)	–	11 SSc patients	–	12 weeks	After intervention	This study demonstrates that a 12-week concurrent training program is safe and substantially improves muscle strength, function, and aerobic capacity in SSc patients
Intensive aerobic and muscle endurance exercise in patients with systemic sclerosis: a pilot study [22]	ALEXANDERSON, 2014	Pilot study	Aerobic training and combined endurance/resistance training	Intense aerobic and endurance training program	–	4 SSc patients	–	6 weeks (noninterventional baseline period) and 8 weeks (exercise intervention period 3× a week)	Every other week throughout the 14-week study	Three participants improved significantly in muscular endurance, and two participants improved significantly or clinically relevantly in aerobic capacity. All other variables remained unchanged, except for a trend toward reduced fatigue
Aerobic exercise is safe and effective in	OLIVEIRA, 2009	CCT	Aerobic training and combined endurance/	Aerobic exercise program of moderate intensity	Aerobic exercise program of	7 SSc patients	7 healthy volunteers	8 weeks	Pre- and posttest	Both groups showed a significant

Name of study	Author, year	Study design	Type of intervention	Description of intervention in intervention group	Description of intervention in control group	Number of individuals in intervention group	Number of individuals in control group	Duration of the intervention	Postintervention assessment	Results
systemic sclerosis [23]			resistance training		moderate intensity					improvement in peak VO ₂ but with no significant difference between the study groups. Furthermore, both groups improved in exercise intensity which was documented by significantly increased peak lactate concentration in blood. No significant changes in SSc skin score or QoL were detected
Lower limb muscle strength is associated with functional performance and quality of life in patients with systemic sclerosis [24]	LIMA, 2015	Cross-sectional study	Lower extremities treatment	Study assesses the peripheral and respiratory muscle strength in individuals with SSc and investigates their correlation with the 6MWD and QoL measurements		20 SSc patients	20 healthy volunteers	April 2013 and January 2014		Patients with SSc exhibited reduced respiratory muscle and quadriceps strength and an increase in its fatigability. In these individuals, there was a relationship between quadriceps strength, functional capacity, and QoL
Evaluation of a mail-delivered, print-format, self-management program for persons with systemic sclerosis [25]	POOLE, 2013	Pilot study	Self-management program	The program consisted of a workbook and exercise DVD that provided information on medical aspects of the disease, dysphagia, fatigue	–	49 SSc patients	–	4–6 weeks	–	Participants consistently reported that the program was easy to use. Depression, fatigue, and pain decreased, and hand function, self-

Name of study	Author, year	Study design	Type of intervention	Description of intervention in intervention group	Description of intervention in control group	Number of individuals in intervention group	Number of individuals in control group	Duration of the intervention	Postintervention assessment	Results
Taking charge of systemic sclerosis: a pilot study to assess the effectiveness of an internet self-management program [26]	POOLE, 2014	Pilot study	Self-management program	management, advocacy, activities of daily living, oral hygiene, skin and wound care, psychosocial changes, exercises, and other features of the condition The program consisted of 10 modules (e.g. coping and body image/appearance, fatigue and energy conservation, self-advocacy), an exercise video, learning activities, worksheets, and resources	–	16 SSc patients	–	Over 10 weeks	–	efficacy for controlling pain, and self-efficacy “other” improved; however, the only statistically significant change was in self-efficacy for pain There were significant improvements in mean scores for ability to manage care and health, and significant decreases in fatigue and depression. Self-efficacy improved, but not significantly
Effects of splinting in the treatment of hand contractures in progressive systemic sclerosis [27]	SEEGER, 1987	Pilot study	Splinting interventions	Dynamic splinting of the hand daily for 8 hours (could decrease proximal interphalangeal (PIP) flexion contractures?)	The nonsplinted hand	8 SSc patients	8 SSc patients	2 months	After 1 and 2 months	Just one of eight patients experienced a statistically significant improvement in PIP range of motion as a result of the splinting. There was no evidence that the use of the splints served to maintain PIP extension when compared with the control hand

Name of study	Author, year	Study design	Type of intervention	Description of intervention in intervention group	Description of intervention in control group	Number of individuals in intervention group	Number of individuals in control group	Duration of the intervention	Postintervention assessment	Results
Manual lymph drainage improving upper extremity edema and hand function in patients with systemic sclerosis in edematous phase [31]	BONGI, 2011	RCT	Physical therapy	MLD 1 session a week – (lasting 1 hour)		20 SSc patients	15 SSc patients	14 weeks: 5 weeks of rehabilitation and 9 weeks of follow-up	At the end of the treatment, and after a follow-up of 9 weeks	The application of MLD is effective in the treatment of the hand in edematous SSc by reducing hand volume, edema, and pain, and improving hand function and perceived QoL
Transcutaneous electrical nerve stimulation (TENS) improves upper GI symptoms and balances the sympathovagal activity in scleroderma patients [32]	SALLAM, 2007	CCT	Physical therapy	Home use TENS application at two GI acupoints on GI symptoms and QoL		17 SSc patients	9 healthy controls	14 days	The electrocardiogram was recorded for two intervals: baseline and TENS application	Prolonged TENS application improved GI symptoms and restored the sympathovagal balance, with an impressive correlation with improved physical functioning scores
Effects of shock wave therapy in the skin of patients with progressive systemic sclerosis: a pilot study [33]	TINAZZI, 2011	CCT	Physical therapy	ESWT of the arm and the hand of one arm	Second arm was without intervention	30 SSc patients	30 SSc patients	Three sittings	Before and immediately after ESWT and at 7, 30, 60, and 90 days after the treatment	The results of this study suggest that ESWT is a novel and efficacious treatment that can be added to the pharmacological therapy in order to decrease endothelial cell damage and skin fibrosis in patients with SSc. This treatment is well tolerated and can be repeated without side effects; in the

Name of study	Author, year	Study design	Type of intervention	Description of intervention in intervention group	Description of intervention in control group	Number of individuals in intervention group	Number of individuals in control group	Duration of the intervention	Postintervention assessment	Results
Effect of biofeedback and deep oscillation on Raynaud's phenomenon secondary to systemic sclerosis: results of a controlled prospective randomized clinical trial [34]	SPORBECK, 2012	RCT	Physical therapy	The effect of deep oscillation and biofeedback on RP secondary to SSc	No intervention or therapies with an expected effect	8 SSc patients biofeedback and 10 SSc patients—deep oscillation	10 SSc patients	3 times per week for 4 weeks	After 4 and 12 weeks	majority of cases it determines a rapid improvement in skin elasticity and skin wellness, even if the effects tend to reduce during the time Biofeedback resulted in an improvement in RP as determined by score reduction of visual analogue scale compared with patients of the control group, whereas deep oscillation revealed a tendency for improvement. The study underlines the beneficial role of physiotherapy for the treatment of SSc-related RP
Multiple daily ultrasound treatment of patients with progressive systemic scleroderma [35]	UHLEMANN, 1990	OD	Physical therapy	Ultrasound therapy of the hands with an intensity of 0.6W/cm ² , 6 minutes per region	–	24 SSc patients	–	3 times a day during 6 days	After 6 days	Pain decreased in 18 of 24 patients. At the end of therapy, no increase in pain was observed, and the hand strength significantly improved in all patients

Name of study	Author, year	Study design	Type of intervention	Description of intervention in intervention group	Description of intervention in control group	Number of individuals in intervention group	Number of individuals in control group	Duration of the intervention	Postintervention assessment	Results
Mouth opening scope in the patients with systemic sclerosis: Simultaneous application of a hyperbaric oxygenation and physical therapy [36]	MILAČIĆ, 2014	Prospective study	Physical therapy	Combined HBO therapy with facial physical therapy	–	13 SSc patients	–	Daily 10 days	After intervention	The therapy showed a statistically significant difference and proved that the combination of HBO therapy and physical therapy has its place in treating these patients

Acronyms: SSc, systemic sclerosis; RCT, randomized controlled trial; CCT, controlled clinical trial; OD, observational design; QoL, quality of life; HRQoL, health-related quality of life; ROM, range of motion; VO₂, oxygen uptake; MLD, manual lymphatic drainage; 6MWD, 6-min walk distance; TENS, transcutaneous electrical nerve stimulation; GI, gastrointestinal; ESW, extracorporeal shock wave; ESWT, extracorporeal shock wave therapy; RP, Raynaud's phenomenon; HBO, hyperbaric oxygenation.

Table 1. Overview of available studies on nonpharmacological interventions in patients with systemic sclerosis.

but also the family or a partner to solve the psychological problems that could be associated with the establishment of the diagnosis.

It is clear from the systematic literature search of available studies that nonpharmacological treatment for these patients has its positive results and has been investigated worldwide. However, since it is a rare disease, the strength of evidence of efficacy of nonpharmacological treatment is limited. The aforementioned limitations suggest that there is an unmet need for international multicentric cooperation, teamwork, and unified projects with a solid design in order to explore this area to arrive at definite conclusions and treatment recommendations. Thus, even nonpharmacological approaches could offer specific techniques that could be implemented and would effectively contribute to greater self-sufficiency, and easier self-management of patients with SSc.

It is also important to carry out further studies to assess other neglected areas such as psychosocial status, depression or sexual dysfunction in order to provide a complex therapy by a team of specialist offering a versatile assistance to patients with SSc.

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