Systemic sclerosis: rehabilitation as a tool to cope with disability

S. Maddali-Bongi, A. Del Rosso

Division of Rheumatology, Department of Clinical and Experimental Medicine, University of Florence, Florence, Italy.

Susanna Maddali Bongi, MD
Angela Del Rosso, MD, PhD

Both authors contributed equally to this work.

Please address correspondence to:
Angela Del Rosso, MD, PhD,
Division of Rheumatology,
Department of Clinical
and Experimental Medicine,
University of Florence,
viale Pieraccini 18,
50139 Firenze, Italy.
E-mail: angela.delrosso@fastwebnet.it

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ABSTRACT

In patients with systemic sclerosis (SSc), local disability of the hands and face, due to the involvement of skin, subcutaneous tissues and musculoskeletal system, is scarcely improved by pharmacological therapy, but may be treated efficaciously with rehabilitation, which can prevent and reduce local disability, thus ameliorating global disability and impaired Quality of Life, related to changes in the hands and face.

In SSc, in order to be efficacious, rehabilitation should: 1. include and use both local treatments of hands and face and global rehabilitation techniques; 2. be different according to the different SSc phases and subsets; 3. include different techniques to tailor treatment to the personal needs and abilities of the patients.

Systemic sclerosis (SSc) is a connective tissue disease characterised by microvascular changes, progressively leading to fibrosis of skin and internal organs and systems (lungs, heart, kidney, urogenital and gastrointestinal tract). Because of its complexity, SSc management should be carried out by a multidisciplinary team of physicians, coordinated by the rheumatologist.

Similarly, the impairment of skin, musculoskeletal system, and respiratory system requires a rehabilitation therapy, to be carried out by a multidisciplinary team (composed of physicians, physiotherapists, podiatrists, occupational therapists, etc.) (1). The rheumatologist has a central role in taking charge and in managing SSc patients, even in rehabilitation, as already recommended for Ankylosing Spondylitis by Assessment of SpondyloArthritis international Society and European League against Rheumatism (ASAS/EULAR) (2).

Patients affected by rheumatic chronic diseases such as SSc are difficult to treat because of systemic comorbidities and complications. Thus, according to the plan for rehabilitation drawn up by the Italian Ministry of Health in 2011, their rehabilitative treatment is necessary and complex (3).

In SSc, the use of physiotherapy and rehabilitation programmes is advised to prevent and to reduce disability derived from cutaneous and musculoskeletal involvement. However, few studies on the subject are published, and rehabilitative therapy for SSc patients is not widely known (4) and is not included in the EULAR recommendations for SSc management. Moreover, in daily practice, proper rehabilitation is seldom performed by physiotherapists and occupational therapists with specific skills in treating SSc patients (5).

This review describes hand and face changes in SSc patients, their correlation with Global Disability and Quality of Life, and rehabilitation therapy, that include both local treatments of hands and face and global rehabilitation techniques.

Hand and face changes and disability

Skin involvement is present in all SSc patients and has a centripetal course, affecting hands and face early and electively, with the potential involvement of all the other skin areas. In SSc, skin involvement evolves through oedematous, sclerotic and atrophic phases.

Hand disability develops since oedematous phase, in which oedema reduces fingers movement and hands functionality. Oedema is prominently related to the microangiopathy of the lymphatic vessels, contemporary to the microangiopathy of the blood microvessels, causing leakage of fluid and macromolecules in the interstitial tissues (6).

In the following sclerotic phase, the skin becomes fibrotic, increases its consistency and becomes inelastic, impossible to be lifted in folds, shiny, tight and adherent to the subcutaneous tissue. In the later atrophic stage, the skin is thinned, due to the reduction in thickness of the dermis.

In the hands, the progressive skin sclero-

sis leads, firstly, to flexion deformity of the fingers and, then, to the typical disabling claw hands, characterised by ankylosis in flexion of the proximal and distal interphalangeal (IP) joints, loss of flexion of the metacarpophalangeal (MCP) joints, loss of mobility of the thumb, blocked in adduction and flexion. The reduction of flexion-extension of the wrist is concomitant. In the atrophic stage, hand movements are further reduced, especially with regard to pronation-supination of the radio-ulnar joint.

In the sclerotic phase, SSc patients have many characteristic changes of the face, such as microstomia, microcheilia, radial wrinkles around the mouth, nose sharpening, smoothing of wrinkles and amimia (meant as loss of facial movements reflecting emotions), defining the typical scleroderma face. These alterations cause disability in chewing food, slurred speech, difficulty in oral hygiene and dental treatments and aesthetic problems and, consequently, reduce both the oral health quality of life (OHQoL) and the overall quality of life (QoL) (7).

Anatomic and functional abnormalities of the hands and face are not only due to fibrosis and retraction of the skin, but they may also be caused and exacerbated by the impairment of musculoskeletal system and by microcirculation changes, represented by Raynaud's phenomenon and ulcers at fingertips, nose and ears, potentially evolving and leading to auto-amputation (8).

Bone resorption of temporomandibular joint (TMJ) may cause osteolysis of condyles, branches and angles of the mandible. The resulting TMJ impairment may lead to a severe painful dysfunction, causing impairment of the jaw movements and, therefore, a reduction of mouth opening.

OHQoL is further compromised by xerostomia, due to the frequent secondary Sjogren's syndrome, alterations of periodontal tissues, retraction of the palate and fibrosis of the lingual frenulum (9-11).

Relationship between hand and face changes, and disability and quality of life

The changes in the hands and face in the course of SSc, mainly due to the involvement of the skin, are severely disabling.

A longitudinal observational survey carried out on 745 Canadian patients shows that in SSc respiratory problems and the diffuse cutaneous SSc (dcSSc) subset are the major predictors of disability severity (12). In a series of 1250 patients, disability, assessed by Health Assessment Questionnaire (HAQ), is directly correlated with skin involvement, hand deformities, muscle strength and cardiac or renal impairment (13).

The changes in the skin of hands and face impair interpersonal relationships, self-esteem and psychosocial functions, so that skin modification in SSc may be considered among the factors leading to image dissatisfaction, which is higher than the image discomfort of patients suffering from extensive skin burns (14, 15).

SSc patients consider thin lips, mouth furrows, facies amimica and small mouth as the major causes of face dissatisfaction, and also refer face changes as leading to more concerns than hand changes (16).

Recently, using Oral Health Impact Profile (OHIP), a questionnaire that assesses the functional, social and psychological impact of the disease at the oral level, Baron *et al*. found that SSc patients had worse oral conditions and worse OHRQoL than healthy controls (17, 18).

A study of our group showed that in SSc patients local disability of the hands and face, measured by specific scales (Cochin Hand Function Scale disability [CHFDS], Hand Mobility in Scleroderma scale [HAMIS], and Mouth Handicap in SSc Scale [MHISS] (19, 20) is related to global disability (HAQ) and to QoL impairment (SF36) (21).

Regarding QoL, a more recent work carried out by our group investigated the independent determinants of physical and mental QoL (assessed by SPI and mental summary index [SMI] of SF36) in SSc patients. Functional Assessment of Chronic Illness Therapy-Fatigue (FACIT-F) and HAMIS (assessing fatigue and manual disability) were the independent determinants of SPI, while the independent determinants of SMI were FACIT-F, the Avoidance Strategies Subscale of Coping Orientation to Problems Experienced-New Italian Version (COPE-NIV), and the anxiety subscale of Hospital Anxiety Depression Scale (HADS-A). As HADS-A contributed significantly in the prediction of SMI of SF 36, its independent determinants (mouth disability, self-esteem and Social Support coping strategy (assessed by MHISS, Rosemberg self-esteem questionnaire [RSES] and COPE-NIV Social Support subscale) were included in the final model for SMI as factors indirectly affecting SMI (by increasing anxiety) (22).

In the work of Sandqvist *et al*. the difficulties in activities of daily living (ADL) were mainly related to the functional impairment of the hands, particularly to stiffness and reduction of grip strength and dexterity (23).

In another study, women affected with lcSSc presented a reduced work capacity of 50%, correlated with ADL impairment and well-being perception (24). In a subsequent investigation by the same authors, pain, fatigue and impairment of hand function were confirmed as causing a notable impact on work disability (25).

According to recent studies, the percentage of SSc patients who are not working is around 37% and global disability, health (26) and disease-related factors (27) are the parameters found as most associated to the work variable.

Rehabilitation

The changes in hands and face of SSc patients, despite their high frequency and the prominent disability that they cause, are undervalued and not thoroughly investigated and treated, probably because they are overshadowed by the concurrent and often severe systemic symptoms of the disease (5).

Hand and face disabilities are frequent, different according to SSc phases and also to subsets, as demonstrated by a study of our group, in which patients with dcSSc present higher HAQ, lower Summary Physical Index (SPI) of SF36, major disability in hands and face than patients with the limited cutaneous subset of SSc (lcSSc) (28). Local disability is also prominently related to global disability and QoL. For

all these reasons, SSc patients should be treated attentively and with different techniques, in order to tailor the treatment to the personal needs and abilities of the patients.

Hand and face problems related to SSc are scarcely improved by the pharmacological therapy usually used to treat the disease, while rehabilitation has a role in coping with the progressive involvement of the skin and periarticular tissues that may lead to severe deformities, causing disability and impairing QoL.

However, although rehabilitation is an effective tool to reduce disability in SSc, there is no prove yet that it could prevent the disease evolution.

1. Hand rehabilitation

In patients with SSc, hand rehabilitation aims to improve hand movement, functionality and strength, as well as to increase participation in the ADL, such as self-care, home management, work and leisure activities and, ultimately, to ameliorate QoL.

A Japanese open study showed in 45 SSc patients that a programme of stretching exercises of the fingers, self-administered by the patients for 1 month, improved the function of the fingers, with the results maintained for 1 year (29).

Some studies used finger mobilisation exercises preceded by the application of paraffin. The randomised controlled trial (RCT) carried out by Sanqvist *et al*., over 1 month, included 17 SSc patients, in which one hand was treated daily with paraffin bath in combination with hand exercises and the other hand, treated only with exercises, was considered a control. The finger exercises and paraffin significantly improved the mobility of the hands, stiffness and skin elasticity, while the hands treated just with exercises only had the benefit of an improvement in mobility (30). The work of Mancuso and Poole included just 3 women treated with paraffin bath and mobility exercises for 8 weeks. The patients, evaluated according to the parameters of the ICF (International Classification of Functioning, Disability and Health), reported a significant improvement in joint motion, skin thickness, grip and pinch strength and dexterity (31).

On the whole, these studies indicate that exercises of finger stretching and mobility, especially if preceded by paraffin baths, favour vasodilation and joint flexibility and reduce skin stiffness. In our opinion, they should be performed regularly by SSc patients in order to maintain and improve hand function and skin flexibility.

Dynamic splints, applying a constant force of stretching on the joints, might be effective in maintaining or improving the extension of the proximal IP joints (PIP) (32). However, the only work evaluating the use of dynamic splints in SSc showed a significant improvement in PIP mobility in only 1 out of 19 patients enrolled in the study. As the use of splints is controversial because they may create friction on the skin and may contribute to the development of skin ulcers, there is no evidence to recommend their use in SSc.

Our group performed two RCTs on the rehabilitation of SSc hands using different methods according to the stage of the disease.

The efficacy of manual lymphatic drainage (MLD), carried out for 5 weeks according to the Vodder method, was evaluated on the oedematous hands of 20 patients in the early stage of SSc and compared to a control observational group (15 patients). In the treatment group, MLD significantly reduced hand volume and improved their function (assessed by HAMIS), pain and the perception of oedema and their interference in ADL (assessed by visual analogue scale [VAS]). Also, global disability (HAQ) and QoL (SPI and SMI of SF36) were significantly improved, with all the results (except SMI) maintained at a 9-week follow-up (33).

To treat hands in the later sclerotic phase we used a 9-week rehabilitative programme, specifically designed for SSc and tailored to the individual patients, combining connective tissue massage, Mc Mennell manipulation and daily home exercises, compared to home exercises only, performed by the control group. The 20 patients treated with the combined protocol, with respect to the control group (20 patients), significantly improved in fist closure, hand function (evaluated by HAMIS and CHFDS), global disability (HAQ) and QoL (SPI and SMI of SF36). All the results, except SPI and SMI, were maintained at a 9-week follow-up. In the control group, the home exercise programme improved only fist closing (34).

These studies indicate that rehabilitation techniques should be integrated, chosen and tailored to SSc patients, differently according to the phase of their disease.

2. Face rehabilitation

In SSc, face rehabilitation aims to improve mouth functions (mouth opening, chewing, swallowing) and disability, mime facial expression, movements and alignment of the cervical spine, often impaired by tissue retraction.

Stretching home exercises were used to improve mouth opening in SSc patients. The exercises included the exaggeration of the usual facial movements, and oral-augmentation exercises (manual stretching of the mouth, executed by thumbs and by tongue depressors). Two studies conducted on small cohorts of patients showed a significant impact of these exercises on mouth opening (35, 36).

However, a more recent RCT, lasting 6 months, evaluating daily manual mouth-stretching and oral-augmentation exercises in 13 SSc patients *versus* a control group (15 patients), showed that exercises significantly improved mouth opening at 3 months, but not at the 6-month evaluation (37). These results may be due to the low adherence rate to the programme and to insufficient frequencies, repetitions and durations of the exercises: 3 minutes twice a day *versus* 5 minutes, 3 times/day, as programmed in the previously published studies (35, 36).

On the whole, these studies indicate that home facial exercises may be helpful in improving mouth opening. According to our experience, these self-managed exercises (easy to do and not time consuming), should be done regularly by SSc patients as, by improving oral function, they may improve the patient's ability to eat, to talk, to perform dental hygiene and, therefore, may ameliorate OHRQoL.

In 20 SSc patients in the sclerotic phase, a RCT of our group examined

the efficacy of a 9-week programme combining connective tissue massage, Kabat's technique (method of proprioceptive neuromuscular facilitation), physiotherapy (executed twice a week) and daily home exercises compared to 20 patients performing home exercises only (control group). At the end of the treatment, the combined programme significantly ameliorated mouth opening, facial skin score and oral disability (measured by MHISS). The improvement in mouth opening and in skin score was maintained after 9 weeks of follow up. The control group reported only a significant improvement of mouth opening, lost at follow-up (38).

In SSc patients with severe oral problems, however, face rehabilitation should be accompanied by an evaluation and treatment of TMJ, whose dysfunction does not allow the total recovery of oral disabilities. A randomised controlled trial of our group lasting 12 weeks on SSc patients with TMJ dysfunction, compared the effects of a rehabilitation programme including intra-and extra-oral manipulation of TMJ, connective tissue massage, Kabat's technique and stretching and mobilisation of the cranio-cervical region and home exercises *versus* only home facial exercises. According to preliminary results obtained in 26 patients, Helkimo scale (assessing TMJ dysfunction) improved in all its components (anamnestic, dysfunction and occlusal indexes) only in the group executing the combined programme (13 patients), while the group executing exercises (13 patients) ameliorated only in the occlusal index. Both protocols improved cervical spine mobility and reduced facial skin score, with the results confirmed at follow-up. Only the combined protocol improved MHISS. Generally, both protocols were efficacious on TMJ and cervical mobilisation, TMJ disability, and skin score. However, the combined protocol seems to ameliorate more items and to be more efficacious than home exercises (39).

According to these data, protocols integrating different rehabilitation techniques tailored to SSc patients are more efficacious than facial exercises.

3. Global rehabilitation

In SSc patients, as well as in patients affected by other chronic systemic rheumatic diseases, rehabilitation, should not only treat the areas most affected by the disease, but should include a global rehabilitation programme, in order to improve posture, breathing, muscle strength and overall QoL. The musculoskeletal system should be treated in its entirety in order to prevent or to reduce the impairments in all the other areas, and to prevent damage due to postural adjustments. A careful clinical and clinimetric assessment, including both local and global evaluation allows to draw up a comprehensive rehabilitation programme, that should be tailored and centred on the physical, psychological and social situation of the patient (40).

Aerobic exercises at progressively increasing intensity (41), also associated with muscle-strengthening exercises (42), improved aerobic capacity and muscle strength in SSc patients with no pulmonary involvement. These results should be considered with attention, as the results come not from RCTs but from studies conducted on small groups of patients with disease in remission. In SSc, due to the involvement of internal organs and musculoskeletal system, aerobic activity and strengthening exercises should be performed with caution, respecting the threshold of pain and the onset of fatigue. Thus, the level and the intensity of the exercises should be adapted and tailored to each patient (41). Till now, only one pilot study has evaluated a small group of SSc patients with pulmonary impairment an intensive aerobic exercise and muscle endurance training programme, that improved muscular endurance and aerobic capacity (43).

There are few studies assessing the utility of global and multidisciplinary rehabilitation programmes in SSc. Antonioli *et al*., in 16 SSc patients, evaluated the efficacy of a 2-week programme of supervised exercises (including breathing and stretching exercises) movements, added to physical therapy and followed by home exercises, compared to a control group (17 patients in a waiting list). After 4 months, SSc patients treated with the multidisciplinary programme improved in QoL, mobility of the hands and tolerance to exercise (44).

In a cohort of SSc patients we evaluated, in a randomised controlled trial, the efficacy of a tailored rehabilitation programme integrating face and hand rehabilitation and global tailored rehabilitation, compared to a control group (following only an educational programme). The face was treated with connective tissue massage, Kabat method and physiotherapy, and the hands were treated with connective tissue massage and Mc Mennell manipulations (in the sclerotic phase) and MLD (in the oedematous phase).

The global techniques consisted of hydrokinesitherapy including breathing, strengthening and stretching exercises, for patients without skin ulcers and not suffering from fear of water, or land-based individual exercises including breathing, stretching and body awareness exercises. At the end of the treatment (performed 2/week for 9 weeks) the patients in the experimental group significantly improved in the parameters of general health (SF36 SPI and SMI, HAQ) as well as in hands (HAMIS, CHFDS, hand closing) and face parameters (mouth opening and face disability, assessed by a specific questionnaire). At a 2-month follow-up, the improvement obtained in HAMIS and mouth opening were maintained. No change was observed in the control group (45).

The results of this study highlight the usefulness in SSc of a customisation and a differentiation of rehabilitative programmes, tailored to the different disease phases and involved areas. Thus, a multidisciplinary approach, integrating both local and global methods, is fundamental in the rehabilitation of SSc patients.

Interestingly, all the global exercises performed in our study (45), including breathing exercises, were based on body awareness, and executed under the guide of physiotherapists with an expertise in Mind Body Therapies (MBT) such as Yoga and Rességuier Method (RM) (46). MBT could be useful in SSc, as they are in other chronic rheumatic diseases. In fact, these techniques, integrating physical, mental and emotional

Table I. Rehabilitation in SSc patients.

Author	Study design	intervention group	Duration of intevention	Control group	Number of patients	Outcome measure	Results
Hand rehabilitation in SSc patients							
Mugii (2006)	Pretest-post test	Home program self-administered exercises for finger stretching	1 month	N/A	TG: 45 SSc	Hand function (finger passive ROM test, hand skin score) Limitations in activities (HAQ) Skin score	Significant Improvement of: fingers passive ROM (maintained at 12 months follow up); hand function (HAQ)
andqvist, (2004)	RCT	Randomly selected hand treated with paraffin bath once a day. after the paraffin bath, hand exercises performed daily	1 month	Other hand, treated with hand exercises	TG: 17 SSc CG: 17 SSc	Hand function (ROM, HAMIS, grip force) Pain (VAS pain) Skin concerns (VAS stiffness, VAS skin elasticity)	TG: finger ROM increased, stiffness and skin elasticity significantly improved; CG: ROM significantly improved. TG improved significantly more than CG for finger ROM, skin stiffness, and elasticity
Mancuso (2009)	Series of single patients	Paraffin bath, ROM exercises 5 days/week	8 weeks	N/A	TG: 3 SSc	Joint motion Pain Grip and pinch strength Dexterity Hand function Skin thickness	Significant Improvement in joint motion skin thickness, grip and pinch strength, dexterity
Seeger (1987)	RCT	Randomly selected hand treated with dynamic splints	2 months	Other hand	TG: 19 SSc CG: SSc 19	Joint motion	1/8 patients who finished the study: significant improvement in ROM of proximal interhalangeal joints
Maddali Bongi (2011)	RCT	Manual lymph drainage of the hands 1/week	5 weeks	Waiting list	TG: 20 SSc CG: 15 SSc	Hand function (hand volumetry, HAMIS, VAS hand oedema, VAS hand pain, VAS interference oedema, VAS interference pain) global disability (HAQ) Quality of life (SF36)	TG: significant reduction of hand volume, improvement of HAMIS, VAS pain, VAS oedema, VAS interference oedema, VAS interference pain; significant improvement of HAQ and SF36
Maddali Bongi (2009)	RCT	Connective tissue massage plus McMennell joint manipulation (2/week) plus home ROM daily exercises	9 weeks	home ROM daily exercises	TG: 20 SSc CG: 20 Ssc	Hand function (HAMIS, Cochin scale, hand opening, fist closure) global disability (HAQ) Quality of life (SF36)	TG group improved significantly for fist closure, hand function (HAMIS, Cochin Scale, global disability (HAQ), and quality of life (SF36) CG improved significantly in fist closure
Face rehabilitation in SSc patients							
Naylor (1984)	RCT	Home daily mouth stretching exercises plus Oral augmentation exercises	3 months	Control: Home daily facial grimacing exercises	TG:5 SSc CG:4 SSc	mouth opening	3 months: TG: had a higher increase in mouth opening than CG (not significantly different) 6 months: When CG received the intervention (months 3-6) mouth opening improved; and original IG (who continues intervention) further improved
Pizzo (2003)	Pretest-post test	Home daily mouth stretching exercises	18 weeks	N/A	TG: 10 SSc	mouth opening	Significant Increase in mouth opening; Improvements in eating, speaking and ability to do oral hygiene
Yuen (2012)	RCT	Home daily mouth stretching exercises plus Oral augmentation exercises	6 months	usual dental care	TG:13 SSc CG:15 SSc	mouth opening	In TG significantly increase in mouth opening compared to CG at 3 months but not at 6 months.

Maddali Bongi (2011)	RCT	connective tissue massage plus Kabat's method plus kinesi therapy (2/week for 9 weeks) daily mouth stretching and Oral augmentation exercises (for 18 weeks)	9 weeks	mouth stretching and Oral augmentation exercises for 18 weeks	TG:13 SSc CG:15 SSc	Mouth function (MHISS, mouth opening) facial Skin score global disability (HAQ) Quality of life (SF36)	in TG: significant improvement of mouth opening, facial skin score and MHISS. In CG: significant improvement of mouth opening,
Maddali Bongi (2014)	RCT	intra-and extra-oral manipulation of temporomandibular joint, connective tissue massage, Kabat's technique, and stretching and mobilisation of the cranio-cervical region plus daily home mouth stretching and Oral augmentation exercises		mouth stretching and Oral augmentation exercises	TG:13 SSc CG:13 SSc	TMJ dysfunction (Helkimo scale : anamnestic, dysfunction and occlusal indexes), face disability (MHISS); facial skin score; mobility of cervical spine	TG: improvement of the 3 indexes of Helkimo scale; MHISS; skin score, mobility of cervical spine CG: improvement of occlusal indexes of Helkimo scale, skin score, mobility of cervical spine
Global rehabilitation in SSc patients							
Oliveira (2009)	Pretest posttest,	Aerobic exercises 2/week	8 weeks		TG:7 SSc CG:5 healthy subjects	Treadmill stress test VO2, VCO2, VE Blood lactate concentration Oxygen saturation	Both groups improved significantly in VO2 (no difference between groups) and in exercise (significant increase in lactate concentration)
Pinto (2011)	Pretest posttest,	Aerobic, resistance and stretching exercises, 2/week	12 weeks	N/A	TG: 11 SSc	Aerobic capacity (VO2 peak, AT, RCP, heart rate rest, heart rate peak exercise); Lower and upper limb dynamic strengths (1 repetition of a leg press and bench press,), isometric strength (back pull and handgrip tests), balance and mobility (timed up-and-go test), muscle function (timed-stands test),	Significant improvement of leg and bench press, back pull, handgrip strength and Muscle function; Significant increase of time-to-exhaustion, significant reduction of heart rate at rest; Increase of workload and time of exercise at ventilatory thresholds and peak of exercise.
Alexanderson, (2014)	Pretest posttest,	Aerobic, resistance and stretching exercises, 3/week	6 weeks	N/A	TG: 4 SSc with lung impairment	Physical capacity (six-minute walk test), aerobic capacity (submaximal treadmill test) and muscle endurance (Functional Index 2); global disability (HAQ) Quality of life (SF36) Raynaud, Fatigue and Global Health (VAS)	3 patients improved significantly in muscular endurance; 2 patients improved significantly in aerobic capacity.
Antonioli (2009)	Pretest posttest,	5/week Diaphragmatic breathing and controlled coughing exercises treadmill land free-walking Finger stretching and occupational therapy Home programme after discharge	2 weeks	no rehabilitation Intervention	TG:16 SSc CG:17 SSc	Hand function (HAMIS) Skin score global disability (HAQ) Quality of life (SF36) 6 minutes walk test Lung function tests	TG: at 4 months after treatment end significant increase in hand function (HAMIS), decrease in heart rate and dyspnea (6 minutes walk test), improvement in SF36 CG: no significant improvement in any measure
Maddali Bongi (2009)	RCT	Global rehabilitation techniques: hydrokinesytherapy or land-based exercise program Face: connective tissue massage plus Kabat method plus physiotherapy; Hands: connective tissue massage plus Mc Mennell manipulations (sclerotic phase); MLD (oedematous phase).	9 weeks	no rehabilitation Intervention	TG:10 SSc CG:10 SSc	Hand: (HAMIS, Cochin scale, hand opening, fist closure, hand volumetry) Face and mouth: Mouth opening, purpose-built-questionnaire for face involvement global disability (HAQ Quality of life (SF36)	TG: significant improvement in all the parameters evaluated. CG: no significant improvement in any measure

RCT: randomised controlled trial TG: treatment group; CG: control group; Tx: treatment; N/A: not applicable; ROM: range of motion; HAQ: Health Assessment Questionnaire Disability; HAMIS: Hand Mobility in Scleroderma; VAS: visual analogue scale; SF36: Short Form 36; MHISS: mouth handicap in Systemic Sclerosis scale; TMJ: temporomandibular joint; VO2: oxygen uptake; VCO2: carbon dioxide production; VE: ventilation; AT: anaerobic threshold; RCP: respiratory compensation.

components, allow the patient to improve perception and body awareness and to play a central and active role in global re-education, potentially ameliorating global health, not only the state of musculoskeletal system and the compliance to pharmacological and non-pharmacological treatments (47).

In a pilot study, we showed the efficacy of RM in a small group of SSc patients, who, at the end of an 8-week treatment, improved in QoL and in sleep and movement quality, and referred a reduction of pain. In our experience, RM may be useful in SSc when other rehabilitation methods cannot be used due to the severe involvement of internal organs and to fatigue (48).

The opportunity of the introduction of MBT in rehabilitation of SSc can be corroborated by the lack of improvement of disability and QoL in another multidisciplinary study, in which they executed routine land based exercises, not tailored on patients needs and expectations (1).

The results of the studies of our group (34, 38, 39) demonstrate that the improvements obtained in the experimental groups are higher than those reached in the control groups, who perform home exercises. These better results may be partly due to the supervision of exercises by a skilled therapist.

The need for supervision by physical therapists skilled in SSc rehabilitation is confirmed by a recent Italian study, that compared the results obtained by performing a 3-month programme of home hand exercises with or without the control of therapist, obtained by a telemedicine system (a portable device and a related telemonitoring infrastructure). SSc patients showed an improvement of hand disability, assessed by Dreiser index, in both groups, but the HAQ and the HAMIS test improved significantly only in the patients that were supervised, albeit not personally but by a telemedicine system, by the therapist (49).

In our opinion, in SSc, the ability of a skilled physiotherapist, who treats the patient taking into account the threshold of pain and the onset of fatigue, is preferable to the use of equipment, which could potentially cause injury and possibly severe damage.

Conclusions

- Many studies on the rehabilitation in SSc involve small size samples of patients and sometimes no control group is present. However, their results clearly show that rehabilitation has a remarkable efficacy as a tool to cope with the disability, although there is no prove yet that it could really stop the evolution of the disease.
- In SSc, rehabilitation, when performed by experienced physiotherapists and occupational therapists, is safe and does not cause significant adverse effects.
- Even in SSc, as in the other rheumatic diseases, high-quality randomised controlled trials, that could confirm the efficacy of the rehabilitation, are advocated. However, large rehabilitation RCTs are difficult to conduct due to the lack of information of rheumatologists and SSc patients about the use and the efficacy of rehabilitation (leading to a reduced patient recruitment), the scarcity of physiotherapists and occupational therapists skilled in evaluating and treating rheumatic patients, the difficulties of the patients in reaching rehabilitation centres, and the heterogeneity of SSc patients.
- The reduction of the effects of the rehabilitation after the end of the treatments underlines that rehabilitative therapy in SSc patients should be regularly and constantly carried out, and that the pharmacological therapy should be constantly assumed.
- Thus, it is advisable to perform regularly both global exercises and home face and manual exercises consistently with the phase and the state of the disease, to which planned regular cycles of therapy, alternating individual sessions and group courses, supervised by physiotherapists, should be added (39).

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