Efficacy of a tailored rehabilitation program for systemic sclerosis


INTRODUCTION

Rehabilitation may contribute to the management of Systemic Sclerosis (SSc) dealing with disabilities due to skin and joint involvement. AIM: to evaluate the efficacy of a district specific and global rehabilitation program tailored for SSc patients.

MATERIALS AND METHODS

20 SSc patients were enrolled and randomly assigned to 2 groups. Interventional group (10 pts) was treated that included hand and face specific rehabilitation and at least a global rehabilitation technique such as hydrokinesytherapy or land-based program, also comprising respiratory exercises. Hand lymphatic drainage was added when necessary. Observational group (10 patients) was only provided with educational advices and medical information about SSc. Patients were evaluated at baseline (T0) and after the 9 weeks treatment period (T1). Interventional group was also assessed after a 9 weeks follow-up (T2). Patients were evaluated by SF-36, HAQ and a purpose-built-questionnaire for global health condition and with Hamis test, Duruöz scale, range of motion, water volumetric test, mouth opening and a purpose-built-questionnaire for hand and face involvement.

RESULTS

At the end of the treatment, patients of interventional group improved in all the parameters evaluated. At follow-up, mouth mobility and functionality such as global health status was partially lost, only hand mobility and functionality parameters were maintained. No changes were observed in controls.

CONCLUSION

The association of and district specific and global rehabilitative techniques conceived and tailored for SSc patients improves disability, HRQoL, hand and face disability and functionality, with its effects partially maintained at the follow-up.

Key words: Systemic sclerosis, rehabilitation, physiotherapy, Mc Mennell, Kabat, connective tissue massage, manual lymphatic drainage.

Competing interests: none declared.

ABSTRACT

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Introduction

Systemic sclerosis (SSc) is a multisystem disease that affects the microvascular system and the connective tissue with a highly heterogeneous clinical presentation, extent and severity of organ involvement and immunologic abnormalities that may have a variable and largely unpredictable course. In the latest years, clinical outcomes considerably improved, which may be attributed in part to effective therapies for organ-specific manifestations, as well as to advances in general medical care (1). However, rehabilitation is still an understudied field of research that could have important practical consequences. In SSc, the most important rehabilitative problems arise from skin induration and joint and muscle involvement (2, 5), often insufficiently responsive to a merely pharmacological approach (3, 4). Skin fibrosis and retraction cause deformations and reduction of functionality of hands and face, potentially leading to disability and affecting activity of daily living, quality of life and psychological well being (6). Face involvement also causes important aesthetic changes and loss of the self-image (6).

As in SSc symptoms may differ widely according to disease phases and also from patient to patient, rehabilitation should be approached by a multitasking rehabilitative team including doctors and therapists with different expertise and tailored on specific patient necessities (7). Though most authors agree that a specific rehabilitation program can prevent and reduce disabilities deriving from skin and joint involvement (2, 5, 7), only few data are published (8-11). A specific exercise program was shown to improve mouth opening in SSc patients (8). Previous studies from our group, based on a specific and individualized rehabilitative approach, suggest...
that the association of connective massage and Mc Mennell joint manipulation in treating SSc hand (9) and the association of Kabat’s technique, connective massage and a specific kinesitherapy for the treatment of SSc face (10), are more effective than a non-specific passive and active-assisted mobilization protocol and than a simple home-based exercise program, respectively. Manual lymph drainage reduced hand swelling and stiffness in oedematous phase and caused improvement in quality of movement and in daily activities (11). These encouraging results support the concept that rehabilitation should be individualized and tailored to fit patients needs in order to improve their overall quality of life. In our studies, each rehabilitation program was specifically conceived for SSc and individualized to fit patients necessities (7, 9-11). Recently, it has been shown that an individual rehabilitation program may be efficacious in improving quality of life and hand mobility in a limited number of SSc patients (12). The use of splints, which is effective in other rheumatic diseases, cannot be employed in SSc due to the lack of evidence in literature (13).

The aim of this study was to evaluate the efficacy of an individualized rehabilitation program specifically conceived and tailored for SSc.

Materials and methods
Twenty Caucasian SSc patients (14) (7 males and 13 females; age: 57.1±15.0 years; disease duration 9.0±4.1 years) were consecutively enrolled from the outpatient clinic of the Department of Biomedicine, Division of Rheumatology of the University of Florence. They agreed to participate in the study, approved by the local ethical committee, by a written informed consent. All patients underwent a clinical examination and were assessed according to international guidelines (15). Ten out of 20 patients presented lung involvement (interstitial lung disease and/or pulmonary hypertension) (16). None was suffering from arthritis (ruled out by clinics and imaging) or myositis (normal muscle enzymes and electromyography evaluation). All patients were affected by flexion contractures, caused by retraction of skin, subcutaneous tissues and tendon sheaths. Seven patients presented with hand oedema and seven were affected by fingertip ulcers. At enrollment, all patients were provided with educational recommendation on general measures (nutrition, skin warming and skin and mucosal protection)

Fig. 1. Connective tissue massage applied at the hands (a) and forearms (b, c). Mc Mennell joint manipulation of the wrist on frontal and radial side diastasis (Fig. 1d, e); of the metacarpophalangeal (Fig. 1f, g) and interphalangeal joints (Fig. 1h, i).
and medical information about SSc. All patients continued their pharmacological treatments (alprostadil-α-cycloDEXtrin, calcium channel blockers, topical glyceryl trinitrate, proton pump inhibitors, clebopride, steroids, cyclophosphamide, azathioprine, D-penicillamine, methotrexate) with no changes throughout the period of the study. Patients were assigned, using a random number sequence prepared by a blinded statistician, to interventional or observational group. After randomization, patients in the interventional group underwent a 9-week rehabilitation program, individualized for each patient according to clinical characteristics and patient’s expectations.

Rehabilitation programs

The rehabilitation programmes were tailored according to the clinical characteristics, SSc phase, involved districts and patient’s expectations. Patients in the interventional group were treated with district specific techniques for hands and face involvement and with a global technique:

1. **Hand involvement** was treated with a combination of connective tissue massage (17-19) and Mc Mennell joint manipulation (20) (1 hour/session, twice a week). Patients with edematous hands were also treated with supplementary sessions of manual lymphatic drainage (11, 22) (1 hour/session, twice a week).

2. For the **face involvement** a combination of Kabat’s method (18, 21), connective tissue massage (17-19) and kinesitherapy (8, 10) was used (1 hour/session, twice a week).

3. The global rehabilitation programs include Hydrokinesytherapy, performed by SSc patients without ulcers (no. 7).

The patients with ulcers (no. 3) were assigned to a land-based rehabilitation. In both cases, patients performed respiratory rehabilitation exercises (23) (1 hour/session, once a week).

Patients of the observational group (controls) were followed up and recommended not to start any new physical or pharmacological therapy during the study period.

Rehabilitation techniques for hand involvement

Connective tissue massage (17-19) was applied on the hands (Fig. 1a) and forearms (Fig. 1b, c) (10 minutes per side) after a brief examination of upper limbs to verify the presence of ulcers. If necessary, after Mc Mennell manipulations, 5 supplementary minutes of massage might be performed. Mc Mennell joint manipulation is a...
technique aimed at recovering the “joint play”, i.e. the involuntary, subtle range of motion that normal joints display on multiple planes, necessary for normal voluntary joint mobility. It improves articular movement, reduces pain and stretches articular capsules and ligaments (20). Treatment begins with wrist manipulations on frontal and radial side diastasis (Fig. 1d, e) and continues with manipulations of the metacarpophalangeal (Fig. 1f, g) and interphalangeal joints (Fig. 1h, i).

Manual lymphatic drainage (according to Vodder method) is based on the use of manual adapted pressure that, by stimulating the musculature of lymphatic vessels, moves fluid in the skin, thus increasing lymphomotricity, reducing pain and relaxing muscle tension (22). In our patients, it was applied firstly at the lymph nodes of the neck and then at the upper limbs, starting from the less oedematous side (22) (Fig. 3g-i).

Rehabilitation techniques for face involvement
Connective tissue massage is a manual technique used to treat altered connective tissues, in order to increase local bloodstream and relax involved tissue by stretching (17-19). For the facial involvement of SSc it is recommended to treat also the neck (Fig. 2a) and the clavicular regions (Fig. 2b).

Kabat’s method is a neurorehabilitation technique that uses spiral and diagonal movement patterns in conjunction with stretch, resistance and other proprioceptive facilitation techniques to reinforce neuromuscular recruitment (18, 21). The Kabat method comprehends the stimulation of the orbicularis oris (Fig. 2c), zygomaticus (Fig. 2d), levator labii (Fig. 2e), nasalis (Fig. 2f), frontalis (Fig. 2g) and corrugator muscles.

Kinesitherapy consists of specific passive, active or assisted exercises to improve mouth opening by mouth-stretching (8, 10) and specific exercises to increase mimic muscles motility and to recover motions of temporo-mandibular joints (exercises for improving mouth opening and jaw lateralization) (Fig. 2h, i).

Global rehabilitation techniques
Hydrokinesitherapy group exercises were performed by patients without ulcers in a 30° pool (1 hour session, once a week). Each session consisted of an initial 10 minutes warm up (walking front ways, sideways, back ways, swimming), followed by 20 minutes of stretching and pulmonary rehabilitation (specific exercises combining the beneficial ef-

![Fig. 3. Hydrokinesitherapy. Relaxation (a) pulmonary rehabilitation (b) and abdominal reinforcement (c) in flotation. Lower limbs stretching and reinforcement (d-f). Manual lymphatic drainage applied firstly at the lymph nodes of the hands (g, h) and forearms (i).](image-url)
Assessment

The global duration of the study was 18 weeks. SSc patients of both interventional or observational group were assessed at baseline (T0) and at the end of the 9-week rehabilitation period (T1). Patients of interventional group were controlled also after a 9-week follow-up (T2).

Outcome measures were selected to evaluate patients’ global health condition and their hand and face involvement. Participants were also asked, at the end of the treatment period, to fill in a questionnaire about their overall level of satisfaction for the whole treatment on the basis of a Visual Analogue Scale (VAS 0-10).

Assessment of global health condition

As previously reported (24), we chose the Medical Outcomes Study Short Form (SF-36) (6, 25-26) (only the Physical Synthetic Index (PSI) and the Mental Synthetic Index (MSI)), and the Health Assessment Questionnaire Disability index (HAQ-DI) (27, 28).

Assessment of specific body districts

The Hand Mobility in Scleroderma (HAMIS) Test (29), the Duruoz scale (30) and the measurements of range of motion of hand opening and fist closing expressed in centimetres were used to assess hand function. The HAMIS examines the effects of SSc on hand function by evaluating 9 items: finger flexion and extension, abduction of the thumb, pincer grip, finger abduction, wrist flexion and extension, and forearm pronation and supination. Each item is scored on a scale from 0 (no impairment) to 3 (total impairment), which yields a total possible score of 27 (29). The Duruoz scale contains 18 items regarding hand ability in the kitchen, during dressing, while performing personal hygiene, office tasks, and other general items. Persons rate their ability from 0 (no difficulty) to 5 (impossible to do) (30).

A water volumetric test (mean of 2 consecutive measurements of the overflow quantity performed by slowly dipping the hand in a bowl full of water) was used to quantify oedema (31). Mouth opening was evaluated by assessing the distance between the tips of upper and lower right incisive teeth (in cm, mean of two consecutive measurements).

A 16-item questionnaire, based on VAS, was also administered in order to assess the following parameters: face relaxation, mouth opening, temporomandibular joint pain, chewing capacity, tongue mobility, swallowing, oral hygiene, ability to whistle, blow air out of mouth and nose, elasticity, expressiveness, symptoms consequences over relational life and desire to improve personal aesthetics.

Statistical analysis

Data are presented as mean ± standard deviation and as numbers and percentages. χ² test was used to compare the groups for clinical characteristics. For all outcome measures, ANOVA for repeated measures was used to detect effects of treatment (between T0 and T1 and T0 and T2) in interventional group. For control group, the comparison between T0 and T1 was assessed by Wilcoxon test. Data analysis was performed using the SPSS statistical package 12.0 for Windows. Significance was set at p<0.05.

Clinical trial registration

The study was registered with Current Controlled Trials (www.controlled-trials.com) n° ISRCTN54093934

Results

The clinical features of SSc patients in interventional and observational group were not statistically different at the study entry (Table I). No drop-out from the study was registered.

In the interventional group, patients improved significantly at the end of the treatment, in the MSI (p<0.005) and PSI (p<0.05) of the SF-36 and in the HAQ scores (p<0.05). HAMIS test (p<0.005), Duruoz scale (p<0.01), fist closure in centimetres (p<0.05), mouth opening in centimetres (p<0.05) and the 16-Items face questionnaire (VAS-Face) (p<0.002) were also improved after the 9-week rehabilitation period. However, the statistical significance was lost at the 9-week follow-up for almost all the items assessed, excluding the HAMIS test (p<0.01) and mouth opening (p<0.01) (Table II).

Patients with oedematous hands, treated with manual lymphatic drainage, showed a decrease of hand oedema and an improvement in the specific questionnaire. Due to the small number of patients (a total of 4), statistical analysis was not performed.

The overall subjective satisfaction of the treatment, explored by a satisfaction questionnaire, was high (9.4±1.3 VAS scale 0-10).

Patients of the observational group did not show any significant improvement in general health condition, hands and face measures at the end of the study.

Discussion

Patients with SSc present a wide range of symptoms according to disease phases and clinical characteristics. For this reason, the rehabilitative approach should be focused on SSc features, thus avoiding rigid protocols and allowing to design a tailored program on each patient (2, 7). Obviously, this requires a multidisciplinary and multitasking rehabilitation approach with a team including doctors and therapists experienced and skilled in the treatment of SSc.

Recently, an Italian study showed, in a group of SSc patients, that a 4 months individualized rehabilitation program composed by a daily 2-week period of therapist-guided exercises, occupational therapy and physical therapy...
was effective in improving quality of life, exercise tolerance and hand mobility (12). Our approach was based instead on a protocol combining different physiotherapy techniques supposed to act synergically.

Kabat’s method, connective massage, kinesitherapy, Mc Mennell joint manipulations and face and hand massage were the mainstay of our rehabilitation program (17-22). Our data clearly show that their association provides a satisfactory approach to the treatment of hands and face in SSc patients. In fact, loss of function and disfiguring may lead to the development of a poor self-image and quality of life, with limitation in daily activities. Even if few patients were treated with manual lymph drainage, it was effective in reducing distal oedema, thus it should be regarded as a supplementary technique useful in the management of the oedematous phase of SSc.

Land-based kinesitherapy, hydrotherapy and respiratory rehabilitation are important in a rehabilitation program of SSc patients, as musculo-skeletal impairment may affect not only the hands, but also other segments (such as feet and limbs) and may cause compensation strategies, potentially leading to pain and further postural alterations. Despite the fact that the individualization of treatment program does not allow to determine the efficacy of each single technique over SSc symptoms, its flexibility and capability to suit patient needs is of outstanding importance. In our study, this is supported by the satisfaction expressed by patients in the final questionnaire and by the absence of drop-outs during the study period.

Other techniques might be integrated in the program to increase its efficacy. A study on 17 SSc patients showed that paraffin baths improved the results obtained by kinesitherapy as regards hands mobility, perceived stiffness and skin elasticity (32).

In our study, improvements achieved at the end of the treatment period were only partly retained when patients were assessed several weeks later. In fact, hand mobility and functionality were only retained. This result is not surprising, since in the present study no home self-management program was prescribed to participants after the end of the treatment. This evidence supports the hypothesis that continuity of care is mandatory in the rehabilitation of SSc patients.

Some evidence exists about the effectiveness of self-management in SSc patients. Recently, Mugii et al. (33) studied the efficacy of self-administered stretching of each finger in 45 SSc patients, showing that range of motion gains were present in each finger after 1 month and maintained within 1 year. Finger stretching improved also finger function, since HAQ score for hand functions, such as eating and gripping, was significantly decreased (33).

In the future, it will be necessary to define more specific assessment protocols, including tests for face mobility and mouth and tongue functionality (34-35), in order to reduce the use of self-evaluations. The Mouth Handicap in Systemic Sclerosis (MHISS) scale recently validated by a French group, seems to be promising in evaluating and in following-up the disability related to the involvement of face and mouth in SSc patients (35).

The main limitation of our study is the low number of SSc patients, that did not allow us to divide participants in subsets and subgroups according to disease phase and to analyse the effects of the specific techniques applied.

The usefulness of the single techniques hereby described should be verified in studies aimed to test their effects when applied singularly.

Table I. Patients characteristics (baseline).

<table>
<thead>
<tr>
<th></th>
<th>Interventional group</th>
<th>Control group</th>
<th>p</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sex (F/M)</td>
<td>6/4</td>
<td>7/3</td>
<td>NS</td>
</tr>
<tr>
<td>Age</td>
<td>58.0 ± 15.1</td>
<td>55.7 ± 14.9</td>
<td>NS</td>
</tr>
<tr>
<td>Disease duration (years)</td>
<td>9.2 ± 4.3</td>
<td>8.9 ± 3.9</td>
<td>NS</td>
</tr>
<tr>
<td>Skin Score</td>
<td>11.1 ± 6.5</td>
<td>12.0 ± 7.2</td>
<td>NS</td>
</tr>
<tr>
<td>Active ulcers (yes/no)</td>
<td>3/7</td>
<td>4/6</td>
<td>NS</td>
</tr>
<tr>
<td>Raynaud (yes/no)</td>
<td>10/0</td>
<td>10/0</td>
<td>NS</td>
</tr>
<tr>
<td>Oesophagus (yes/no)</td>
<td>9/1</td>
<td>8/2</td>
<td>NS</td>
</tr>
<tr>
<td>Musculo-skeletal involvement (yes/no)</td>
<td>10/0</td>
<td>10/0</td>
<td>NS</td>
</tr>
<tr>
<td>Edematous hands (yes/no)</td>
<td>4/6</td>
<td>3/7</td>
<td>NS</td>
</tr>
<tr>
<td>Lung involvement (ILD/PAH)</td>
<td>6/4</td>
<td>4/6</td>
<td>NS</td>
</tr>
<tr>
<td>Sjogren (yes/no)</td>
<td>3/7</td>
<td>4/6</td>
<td>NS</td>
</tr>
<tr>
<td>MSI (SF-36)</td>
<td>36.9 ± 6.0</td>
<td>39.8 ± 6.6</td>
<td>NS</td>
</tr>
<tr>
<td>PSI (SF-36)</td>
<td>37.9 ± 7.9</td>
<td>36.2 ± 8.8</td>
<td>NS</td>
</tr>
<tr>
<td>HAQ</td>
<td>1.2 ± 1.2</td>
<td>1.01 ± 1.0</td>
<td>NS</td>
</tr>
<tr>
<td>HAMIS TEST</td>
<td>10.2 ± 4.8</td>
<td>9.6 ± 4.0</td>
<td>NS</td>
</tr>
<tr>
<td>DUROUZ SCALE</td>
<td>23.3 ± 19.9</td>
<td>20.6 ± 21.0</td>
<td>NS</td>
</tr>
<tr>
<td>Hand opening* (cm)</td>
<td>15.7 ± 1.3</td>
<td>14.3 ± 1.8</td>
<td>NS</td>
</tr>
<tr>
<td>Fist closure* (cm)</td>
<td>1.4 ± 2.2</td>
<td>0.92 ± 1.7</td>
<td>NS</td>
</tr>
<tr>
<td>Mouth opening (cm)</td>
<td>3.4 ± 1.1</td>
<td>3.7 ± 2.8</td>
<td>NS</td>
</tr>
<tr>
<td>FACE-VAS</td>
<td>3.7 ± 1.3</td>
<td>4.0 ± 1.6</td>
<td>NS</td>
</tr>
</tbody>
</table>

* indicates significant difference.

Table II. Global health, hand and face conditions at the study entry (T0), at the end of treatment (T1) and at follow-up (T2) in interventional group.

<table>
<thead>
<tr>
<th></th>
<th>T0</th>
<th>T1</th>
<th>T2</th>
<th>p T0-T1</th>
<th>p T0-T2</th>
</tr>
</thead>
<tbody>
<tr>
<td>MSI (SF-36)</td>
<td>36.9 ± 6.0</td>
<td>44.6 ± 6.0</td>
<td>34.7 ± 4.6</td>
<td>&lt;0.005</td>
<td>NS</td>
</tr>
<tr>
<td>PSI (SF-36)</td>
<td>37.9 ± 7.9</td>
<td>44.9 ± 8.6</td>
<td>39.0 ± 7.9</td>
<td>&lt;0.05</td>
<td>NS</td>
</tr>
<tr>
<td>HAQ</td>
<td>1.2 ± 1.2</td>
<td>0.9 ± 1.1</td>
<td>0.8 ± 1.2</td>
<td>&lt;0.05</td>
<td>NS</td>
</tr>
<tr>
<td>HAMIS TEST</td>
<td>10.2 ± 4.8</td>
<td>6.0 ± 3.7</td>
<td>6.4 ± 7.4</td>
<td>&lt;0.005</td>
<td>&lt;0.01</td>
</tr>
<tr>
<td>DUROUZ SCALE</td>
<td>23.3 ± 19.9</td>
<td>14.0 ± 16.0</td>
<td>17.3 ± 17.2</td>
<td>&lt;0.01</td>
<td>NS</td>
</tr>
<tr>
<td>Hand opening* (cm)</td>
<td>16.7 ± 1.3</td>
<td>16.4 ± 1.7</td>
<td>14.7 ± 3.1</td>
<td>NS</td>
<td>NS</td>
</tr>
<tr>
<td>Fist closure* (cm)</td>
<td>1.4 ± 2.2</td>
<td>0.4 ± 0.9</td>
<td>0.5 ± 0.8</td>
<td>&lt;0.05</td>
<td>NS</td>
</tr>
<tr>
<td>Mouth opening (cm)</td>
<td>3.4 ± 1.1</td>
<td>4.0 ± 1.2</td>
<td>4.8 ± 1.4</td>
<td>&lt;0.05</td>
<td>&lt;0.01</td>
</tr>
<tr>
<td>FACE-VAS</td>
<td>3.7 ± 1.3</td>
<td>3.1 ± 1.1</td>
<td>3.8 ± 0.9</td>
<td>&lt;0.0002</td>
<td>NS</td>
</tr>
</tbody>
</table>

* indicates significant difference.
Conclusions
The integration of district-specific and global rehabilitative techniques tailored on SSC was efficacious and partially maintained at follow-up. These encouraging results, obtained in a small number of patients, suggest the employment of the protocol in randomized controlled studies enrolling a higher number of patients.

Acknowledgements
We would like to thank the AMuRR therapists Caterina Di Felice, Giulia Guasparri, Sara Miccino, Elena Fanfani, Fiammetta Orsini and Giovanni Soggio, who participated in the rehabilitation of SSC patients.

References