



BACKGROUND

In SSc, the most important rehabilitative problems arise from skin induration and joint and muscle involvement. Skin fibrosis and retraction causes deformations and reduction of functionality of hands and face leading to disability and impairing activity of daily living, quality of life (QoL) and psychological well being. Face involvement also causes important aesthetic changes and loss of the self-image.

Despite this, only few rehabilitative approaches on SSc patients were tried. A rehabilitation program composed by therapist-guided exercises, occupational and physical therapy improved QoL, exercise tolerance and hand mobility in SSc patients [1]. A combination of connective massage, Mc Mennell technique and specific kinesiotherapy was useful for treating hand involvement in SSc patients. [2]. Manual Lymph Drainage (MLD) reduced hand edema and stiffness in oedematous phase and improved quality of movement and daily activities [3].

Microstomia was treated with exercises of mouth-stretching and oral augmentation [4-5] and the combination of Kabat's technique, connective massage and a specific kinesiotherapy program was useful for facial involvement in SSc patients. [6].

AIM

to evaluate the efficacy of a district specific and global rehabilitation program tailored for SSc patients

METHODS

20 SSc patients (7 males and 13 females; age and disease duration 57.1 ± 15.0 years and 9.0 ± 4.1 years, respectively) were enrolled and randomly assigned to 2 groups. Interventional Group (10 pts) underwent a rehabilitation program including hand (connective tissue massage and Mc Mennell joint manipulation (and, for 4 pts with edematous hands, manual lymph drainage-MLD-) and face treatments (Kabat's method, connective tissue massage and kinesiotherapy), both performed 1 hour/session, twice a week, and at least a global rehabilitation technique such as hydrokinesytherapy (7 pts) or land-based kinesiotherapy (3 pts), both executed for 1 hour/session, once a week, and comprising respiratory exercises. Observational Group (10 patients) was only provided with educational advices and medical information about SSc.

REHABILITATION TECHNIQUES

HANDS

• **Connective tissue massage:** manual technique used to treat altered connective tissues, in order to increase local bloodstream and relax involved tissue by stretching applied on the hands and forearms (10 minutes-min./side). If necessary, after Mc Mennell manipulations, 5 more min. of massage might be performed [Fig 1a,b].

• **Mc Mennell joint manipulation:** aims to recover the "joint play", i.e. the involuntary, subtle range of motion displayed by normal joints on multiple planes, necessary for normal voluntary joint mobility. It improves articular movement, reduces pain and stretches articular capsulae and ligaments. It consists in manipulations on wrist frontal and radial side diastasis and on metacarpophalangeal and interphalangeal joints (15 min./side)[Fig 1c,d].

• **Manual lymphatic drainage:** (Vodder method) is based on manual adapted pressure that, by stimulating the musculature of lymphatic vessels, moves fluid in the skin, to increase lymphomotricity, reduce pain and relax muscle tension. It was applied firstly at the lymph nodes of the neck and then at the upper limbs, starting from the less oedematous side, and in a centripete direction [Fig 3g,i].

FACE:

• **Connective tissue massage:** applied at face and neck (10 min.)[Fig 2a,b].

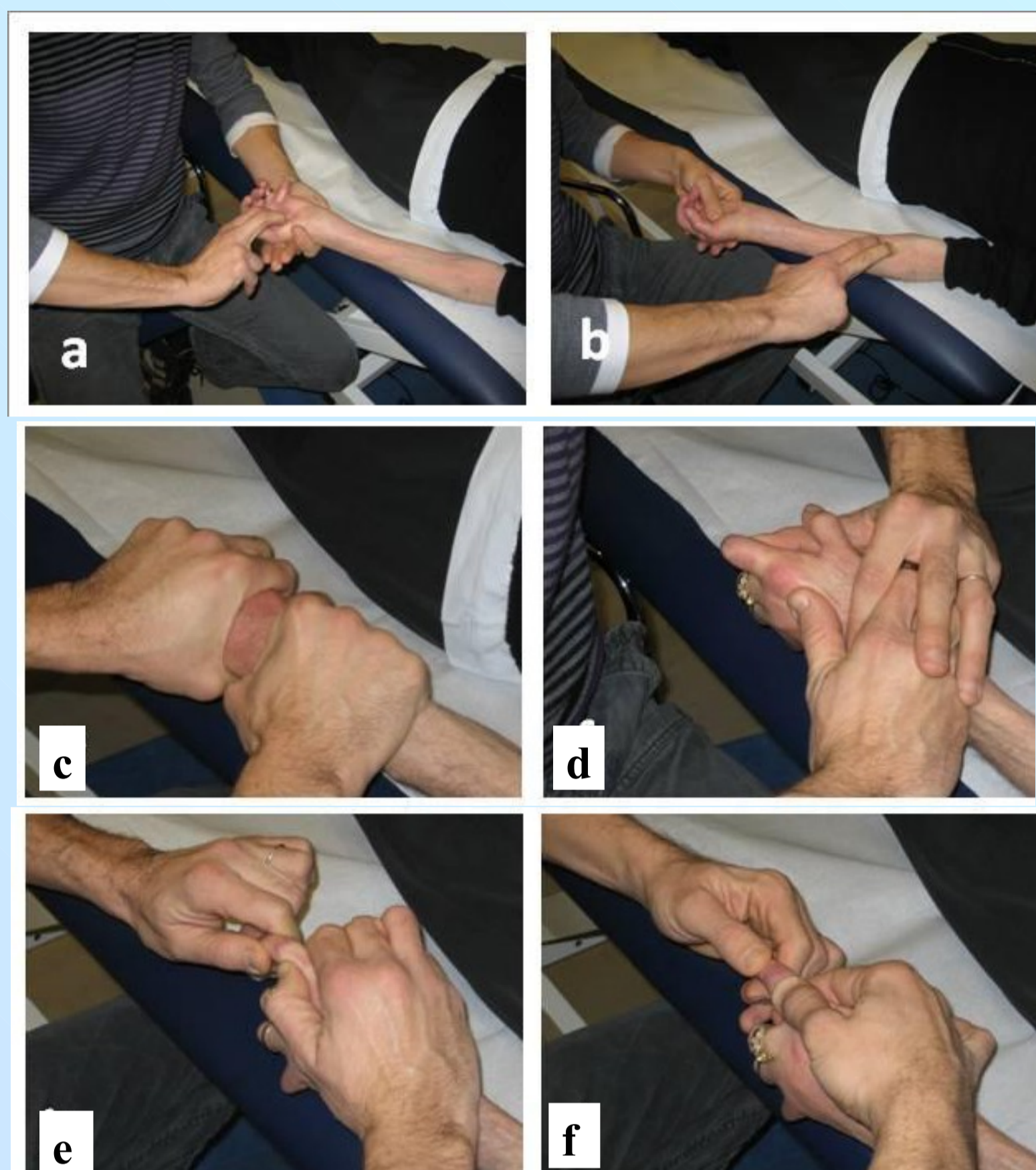
• **Kabat's method:** neurorehabilitation technique using spiral and diagonal movement patterns in conjunction with stretch, resistance and other proprioceptive facilitation techniques to reinforce neuromuscular recruitment applied to mimic face muscles (15 min.) [Fig 2c,d].

• **Kinesiotherapy:** specific passive, active or assisted exercises to improve mouth opening by mouth-stretching and exercises to increase mimic muscles motility and to recover motions of temporo-mandibular joints (15 min. of active, 20 min. of relaxing exercises) [Fig 2e,f].

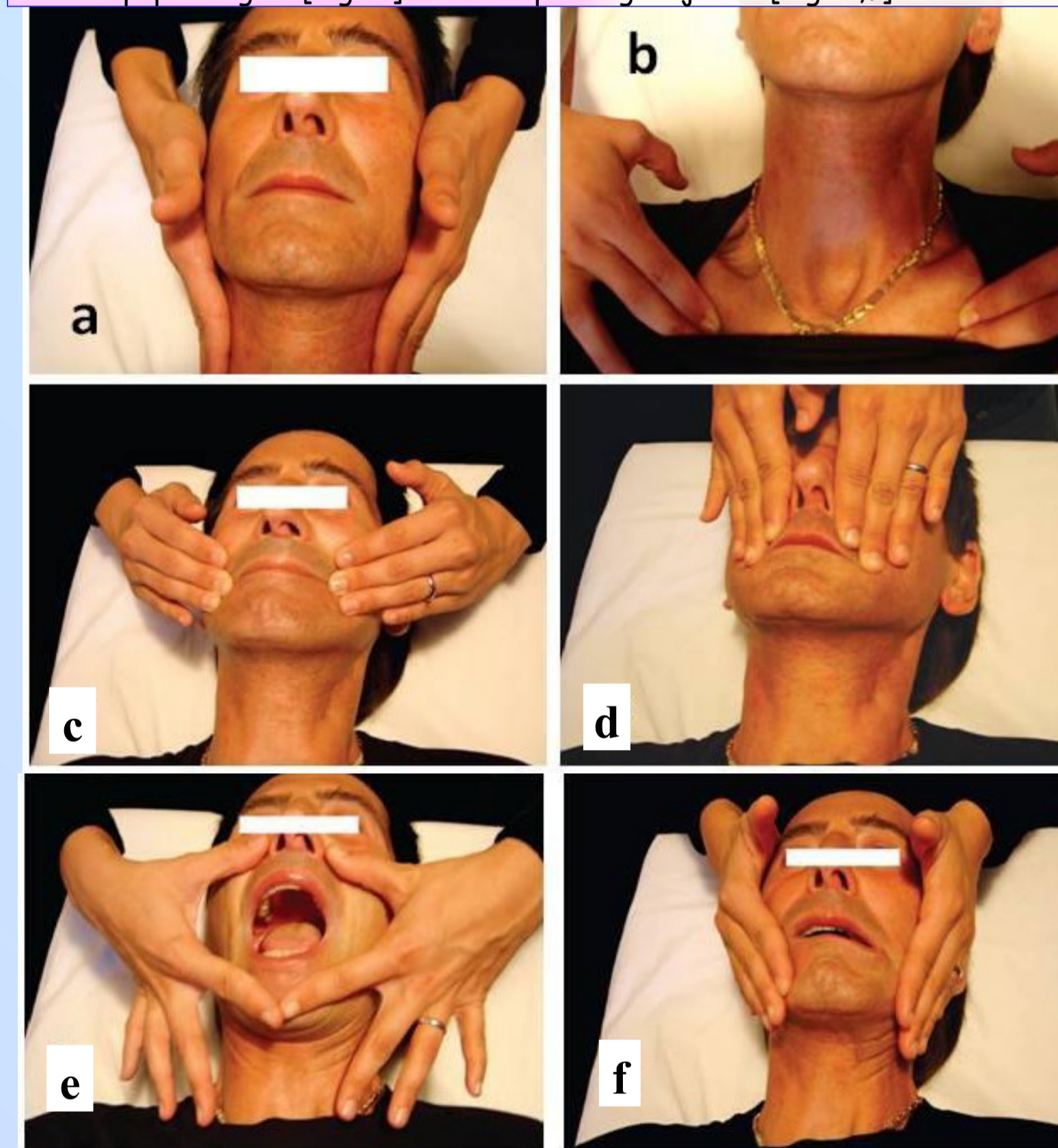
GLOBAL TECHNIQUES

• **Group Hydrokinesytherapy:** performed in a 30° pool (1 hour/session) composed by 10 minutes warm up; 20 min. of stretching and pulmonary rehabilitation; 20 min. for treatment of local and global pain (individualized exercises increasing mobility, muscle strength, body awareness, coordination and balance); at the end: 10 min. relaxing hydro massage bath in a 35° Jacuzzi pool [Fig 3a,f].

• **Group Physical Therapy (1 hour/session):** 10 min. warm up, followed by 30 min. of stretching, body awareness and pulmonary rehabilitation exercises. The final 20 min. exercises treated local and global pain by increasing mobility, muscle strength, flexibility.



Connective tissue massage at the hands (a) and forearms (b). Mc Mennell joint manipulation of the wrist on frontal side diastasis [Fig.1c]; of the metacarpophalangeal [Fig.1d] and interphalangeal joints [Fig.1e,f].



Connective tissue massage of the neck [Fig.2a] and the clavicular regions [Fig.2b]. Kabat method of the zygomaticus [Fig.2c], levator labii [Fig.2d], Kinesiotherapy exercises for mouth opening and jaw lateralization [Fig.2e,fi].



Hydrokinesytherapy. 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 hands (g,h) and forearms (i).

ASSESSMENT

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) for:

• **Global health status**, by Physical (PSI) and Mental Synthetic Index (MSI) of SF-36 and HAQ disability index (DI);

• **Hand involvement**, by Hand Mobility in Scleroderma (HAMIS) test, Duruöz scale, fist closure and hand opening (both in cm: mean of 2 consecutive measurement);

• **Hand edema**, by inverse volumetry and edema-related symptoms by a specific 4 VAS questionnaire;

• **Face involvement**, by mouth opening (in cm: mean of 2 consecutive measurement) and a purpose-built-questionnaire to assess mouth functionality (14 VAS).

RESULTS

The clinical features of SSc patients in Interventional and Observational Group were similar and no drop out from the study was registered.

Interventional group (Table 1):

Patients improved significantly, at T1 in MSI ($p < 0.005$), PSI ($p < 0.05$) of SF-36 and in HAQ ($p < 0.05$); in HAMIS test ($p < 0.005$), Duruöz scale ($p < 0.01$), fist closure: ($p < 0.05$), mouth opening in centimetres ($p < 0.05$) and the 16-Items face questionnaire (VAS-Face)($p < 0.002$).

At T2, the statistical significance was lost for almost all the items assessed, excluded HAMIS test ($p < 0.01$) and mouth opening ($p < 0.01$).

Four SSc patients with oedematous hands, treated with MLD, improved in hand oedema and edema-related symptoms (data not shown).

Observational group:

Patients did not show any significant improvement in general health condition, hands and face measures at T1.

Table 1

Global health, hand and face assessment at the study entry (T0), at the end of treatment (T1) and at follow-up (T2) in Interventional Group (mean \pm SD)

	T0	T1	T2	pT0 - T1	p T0 - T2
MSI (SF-36)	36.9 \pm 6.0	44.6 \pm 6.0	34.7 \pm 4.6	<0.005	NS
PSI (SF-36)	37.9 \pm 7.9	44.9 \pm 8.6	39.0 \pm 7.9	<0.05	NS
HAQ	1.2 \pm 1.2	0.9 \pm 1.1	0.8 \pm 1.2	<0.05	NS
HAMIS TEST	10.2 \pm 4.8	6.0 \pm 3.7	6.4 \pm 7.4	<0.005	<0.01
Duruöz scale	23.3 \pm 19.9	14.0 \pm 16.0	17.3 \pm 17.3	<0.01	NS
Hand opening* (cm)	15.7 \pm 1.3	16.4 \pm 1.7	14.7 \pm 3.1	NS	NS
Fist closure* (cm)	1.4 \pm 2.2	0.4 \pm 0.9	0.5 \pm 0.8	<0.05	NS
Mouth opening (cm)	3.4 \pm 1.1	4.0 \pm 1.2	4.8 \pm 1.4	<0.05	<0.01
FACE-VAS (0-10 cm)	3.7 \pm 1.3	3.1 \pm 1.1	4.0 \pm 1.0	<0.002	NS

Legend: MSI =Mental Synthetic Index; PSI =Physical Synthetic Index; HAQ= Health assessment questionnaire; Face-VAS = VAS Questionnaire on Face involvement: *Mean values between right and left hand

CONCLUSIONS

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

To maintain the results, we advice to add a home self management program after the end of the treatment.

References

- 1) Antonielli CM, et al. Clin Rheum 2009 ;28: 159-65;
- 2)Maddali Bongi S et al. Arthr Rheum 2005 52, suppl, abstract 1566, p. 5586.
- 3)Pizzo G, et al. Eclin Oral Investig 2003;7:175-8
- 4) Naylor P, et al. Oral Surg Oral Med Oral Pathol 1984; 57:508-511
- 5) Matucci-Cerinic M, Maddali Bongi S et al. International Workshop on Scleroderma, May 18- 20, Tokyo, Japan, abstr. 091, page 94, 2007
- 6) Sandqvist G, Eklund M. Arthritis Care Res. 2000;13:369-74.
- 7) Brower LM, Poole JL. Arthritis Rheum. 2004 ;51:805-9
- 8) Damstra RJ et al. Res Treat. 2006;99:267-73.

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