

## Value of Combined Facial Rehabilitation Techniques and Temporomandibular Osteopathy in a Cohort of Egyptian Patients with Systemic Sclerosis

Enas A. Elattar<sup>1</sup>, Takwa. B. younes<sup>1</sup>, Sherin M. Hosny<sup>2</sup> and Mohammed S. Hassan<sup>3</sup>

<sup>1</sup>Physical Medicine, Rheumatology and Rehabilitation Department, <sup>2</sup>Internal Medicine Department, and <sup>3</sup>Radiology Departments, Faculty of Medicine, Ain Shams University, Cairo. Egypt.

[Enasattar2@yahoo.com](mailto:Enasattar2@yahoo.com)

**Abstract:** Systemic Sclerosis (SSc), is an autoimmune connective tissue disease, with fibrosis of soft tissues  
**Objective:** to evaluate the value of the combined rehabilitation techniques [facial stretching exercises & connective tissue massage, Kabat's method, and kinesiotherapy and manipulative osteopathy] in patients with systemic sclerosis; both clinically and ultrasonographically. **Methods:** 20 patients were subjected to combined rehabilitation technique and home exercise program (group A) for 8 weeks, and 20 patients (group B) were assigned only for home exercise program for 8 weeks. All patients were assessed at baseline (F0), at the end of the treatment (F1) and after 8 weeks of follow-up (F2). They were evaluated with SF-36, health assessment questionnaire (HAQ), modified Rodnan skin score, mouth opening and Mouth Handicap in Systemic Sclerosis scale (MHISS). TMJ ultrasonography was done for the both groups and for 20 normal controls for comparison. **Results:** In both groups, SF-36 and HAQ were not affected by the treatment. At F1, both groups improved in mouth opening ( $p < 0.05$ ), and the improvement was maintained at for both groups. Facial skin score improved in (group A) at F1, and maintained at F2 ( $p < 0.05$  versus F0), while no change was observed in (group B). MHISS scale improved significantly in (group A) at F1 ( $p < 0.001$ ) only, while no change was found in (group B). Temporomandibular joint (TMJ) ultrasonography data didn't show any difference neither from the controls nor the two groups at different assessments. **Conclusion:** The combination of rehabilitation techniques, is more effective than home exercise program alone in the rehabilitative treatment of SSc facial involvement.

[Enas A. Elattar, Takwa. B. younes, Sherin M. Hosny and Mohammed S. Hassan. **Value of Combined Facial Rehabilitation Techniques and Temporomandibular Osteopathy in a Cohort of Egyptian Patients with Systemic Sclerosis.** *Life Sci J* 2014; 11(12):574-585] (ISSN: 1097-8135). <http://www.lifesciencesite.com>. 112

**Key words:** Systemic Sclerosis, combined rehabilitation techniques, & ultrasound of TMJ.

### 1. Introduction:

Systemic sclerosis (SSc) is an autoimmune connective tissue disease characterized by excessive collagen deposition, vascular hyper-reactivity and obliterative micro-vascular phenomenon involving the skin and internal organs.<sup>(1,2)</sup> SSc results in skin, tendon, joint and vessel damage leading to worsening of life's quality.<sup>(3,4,5)</sup> Two main subtypes are distinguished: limited cutaneous SSc (lSSc) and diffuse cutaneous SSc (dSSc).<sup>(6)</sup> The focus of attention when assessing a patient with SSc is often based on physiological measurements<sup>(7)</sup> which do not necessarily relate to the true impact of the disease on patient's lifestyle or functional capacity.<sup>(8)</sup>

The involvement of face and oral tissues is a typical feature of SSc patients, that causes impairment of the self-image.<sup>(9)</sup> Where the cutaneous furrows of the face disappear, due to skin retraction, and the nose becomes sharp. Face changes also include thinning and reduction of mouth width (microcheilia) and opening (microstomia), also favoured by osteolysis of mandibular angles and by fibrosis of soft tissues. These modifications interfere with eating, speaking, oral hygiene measures, and dental treatment, thus deteriorating the health-related quality of life

(HRQoL) of SSc patients.<sup>(10-15)</sup> and in most severe cases leading to surgical interventions, such as bilateral commissurotomies.<sup>(10-15)</sup> Recently a Mouth Handicap in Systemic Sclerosis scale (MHISS) has developed, which is a specific tool quantifying in SSc patients the handicap associated with mouth disability and the oral health-related quality of life (OHRQoL), that explores problems not assessed by HAQ and SF 36.<sup>(16)</sup>

To help patients cope with the consequences of the disease, rehabilitative strategies are often provided, these may include psychoeducational interventions; exercise therapy; the application of physical modalities, assistive devices, and orthoses; joint protection; and energy conservation, dietary interventions, and comprehensive multidisciplinary team care interventions.<sup>(17)</sup> A few studies demonstrated a significant improvement of measures of global health and hand function<sup>(18,19)</sup> as well as mouth function<sup>(18,19)</sup>, in patients participating in a rehabilitation program. Studies on the effect of single interventions, including patient education<sup>(20,21)</sup>, aerobic exercise<sup>(22)</sup>, finger<sup>(23)</sup>, or mouth stretching<sup>(24)</sup>, and hand massage and manipulation showed promising results.<sup>(25)</sup>

Osteopathic manipulative medicine (OMM), also known as osteopathic manipulative treatment (OMT), is the core technique of osteopathic medicine<sup>(26)</sup>. It is based on a philosophy addressed by Andrew Taylor Still, which posits the existence of a myofascial continuity that interlinks all parts of the body. Practitioners believe they are able to diagnose and treat a variety of systemic human ailments by manipulating the bones and muscles of a patient.

Rehabilitation management of microstomia, is mainly based on self administered home-based exercise program including mouth-stretching and oral augmentation exercises. This traditional approach has been shown to have some positive effect on mouth opening in SSc patients.<sup>(10)</sup>

#### **Aim Of The Work:**

The aim of this work is to study the value of the combined rehabilitation techniques including facial stretching exercises & connective tissue massage, Kabat's method, and kinesitherapy and tempromandibular joint (TMJ) manipulative osteopathy in patients with systemic sclerosis; both clinically and ultrasonographically.

## **2. Patients and Methods:**

### **Patients**

The present case control study included forty adult female patients, fulfilling the American Rheumatism Association criteria for the classification and diagnosis for systemic sclerosis (SSc) (scleroderma proximal to the metacarpophalangeal joints, sclerodactyly, digital pitting scars (not pulp loss), and bilateral basilar pulmonary fibrosis) and also the new 2013 ACR/EULAR classification criteria for systemic sclerosis was considered the items are; skin thickening of the fingers, fingertip lesions, telangiectasia, abnormal nailfold capillaries, pulmonary arterial hypertension and/or interstitial lung disease, Raynaud's phenomenon, and SSc-related autoantibodies.<sup>(27)</sup> Moreover, our patients had clinical, skin involvement of the face of ( $\geq 1$ ) according to the modified Rodnan skin score (MRSS)<sup>28</sup>, who attended Internal Medicine and Physical Medicine, Rheumatology & Rehabilitation inpatient departments and outpatient clinics of Ain shams University hospitals, Cairo, Egypt.

As well as twenty healthy subjects age and sex matched with the patients (limited to ultrasonography assessment and measurements of mouth opening).

\*\*\*Exclusion criteria included:

- Patients with skin diseases other than scleroderma.
- Patients with previous trauma or dislocation to the TMJ.
- Patients who were involved in similar rehabilitation programmes before.

\* Our 40 SSc patients were subdivided randomly regardless their disease subtype into two groups:

\* (Group A): (20 SSc patients assigned for combined rehabilitation techniques)

They were treated for 8 weeks (trice per week, 45 minutes per session) in the form of:

Facial hot fomentations for 5 minutes, then applying an emoliant cream to the face and neck area, with a combined three steps procedure of connective tissue massage<sup>(29-31)</sup> during the first 10 minutes, Kabat's method<sup>(31,32)</sup> for 15 minutes and kinesitherapy for the following 15 minutes.

The patients were instructed also to perform a daily home exercises program for the whole 16 weeks of the study.

\* (Group B): 20 SSc patients were assigned only for daily home exercise program for 16 weeks.<sup>(10,12,33)</sup>

\*the control group(20 patients), were examined clinically for the measurements of mouth opening, and bilateral U/S of both tempromandibular joint.

**Ethical considerations:** Patients and controls were informed about the nature of the study and consent was taken to participate in the study.

**Methods:** All the patients were subjected to:

### **I. Full history taking and thorough clinical examination:**

according to international guidelines.<sup>(34)</sup> Interstitial lung disease, pulmonary function tests and/or pulmonary arterial hypertension (BAL), Cardiac involvement was defined by the presence of pericarditis, arrhythmia or left ventricular congestive heart failure; oesophageal involvement was defined as hypomotility; muscle-skeletal involvement by the presence of arthralgias, arthritis, flexion contractures; with special attention to TMJ (*opening, side to side movement, jaw retraction & protrusion, click sounds while moving*), with special emphasis on the presence of active digital ulcers, telangiectasia, skin (hypo-hyper) pigmentations and Raynaud phenomenon. Sicca syndrome according to Vitali et al.<sup>(35)</sup>

\* Evaluation of the severity and the extend of the skin involvement was done; using the modified Rodnan skin score (MRSS) of 17 sites with a maximum score of 51.<sup>(28,36)</sup>

### **Assessment of global health condition:**

\*Health Assessment Questionnaire disability index (HAQ-DI)<sup>(37,38)</sup>:

was used to measure global disability; HAQ, a self-report questionnaire, is organised in 20 items divided into 8 categories: dressing and grooming, arising, eating, walking, personal hygiene, reaching, gripping and other activities. Each item is rated from 0 (no difficulty) to 3 (unable to do). A score for each category is the highest score for any question in the category. Then the disability index is calculated by adding the scores from each category and dividing by

the number of categories answered and rated from 0 (less disabled) to 3 (more disabled).<sup>(39)</sup>

\* SF-36, a self-report questionnaire, consists of 36 items organized into 8 domains measuring 8 health concepts: physical functioning, role limitations due to physical problems, bodily pain, general health perceptions, vitality, social functioning, role limitations due to emotional problems and general mental health. In SF-36 domains, scores are rated so that higher values correspond to better conditions and lower scores to worse conditions (range 0–100). The 8 domains, weighted according to normative data, are also combined into a summary physical index (SPI) and a summary mental index (SMI), scored from 0 to 100, with higher values reflecting better HRQoL<sup>(9)</sup>. the Physical Synthetic Index (PSI) and the Mental Synthetic Index (MSI), of SF36 were used to assess quality of life (QoL)<sup>(9,40,41)</sup>

#### \*Face Assessment:

\*Mouth Handicap in Systemic Sclerosis scale (MHISS), it is a specific tool quantifying in SSC patients the handicap associated with mouth disability.<sup>(42)</sup>

It consists of 12 items (each scored 0–4, Never; 0, Rarely; 1, Occasionally; 2, Often; 3, Always; 4) with a total score ranging from [ 0 to 48] divided into 3 subscales:

- \*subscale 1 (5 items: 1, 3, 4, 5 and 6; range 0–20) examines handicap related to reduced mouth opening.
- \* subscale 2 (5 items: 2, 7, 8, 9 and 10; range 0–20) assesses handicap related to sicca syndrome.
- \*subscale 3 (items 11 and 12; range 0–8) examines aesthetic concerns. The total score is obtained by summing the score for all items.<sup>(16)</sup>

Mouth Handicap in Systemic Sclerosis (MHISS) scale:

1. I have difficulties opening my mouth.
2. I have to avoid certain drinks (sparkling, alcohol, acidic).
3. I have difficulties chewing.
4. My dentist has difficulties taking care of my teeth.
5. My dentition has become altered.
6. My lips are retracted and/or my cheeks are sunken.
7. My mouth is dry.
8. I must drink often.
9. My meals consist of what I can eat and not what I would like to eat.
10. I have difficulties speaking clearly.
11. The appearance of my face is modified.
12. I have trouble with the way my face looks.

\*Mouth opening size (microstomia or small mouth) by *measuring tape*. Mouth opening was assessed in centimetres by measuring the distance between the tips of upper and lower right incisive teeth (mean of two consecutive measurements) it is considered as a

clinical profile of TMJ range of motion and the elasticity of the soft tissues of the mouth.

\*Palpation of TMJ (click) & from inside and outside the mouth.

\* Skin involvement of the face was assessed by means of a modified Rodnan skin score separately with a range of (1-3).<sup>(28)</sup>.

#### II. Routine laboratory and specific immunological tests for SSC:

Complete blood count CBC; Erythrocyte sedimentation rate (ESR); C-reactive protein (CRP); Antinuclear antibody (ANA); Anti-Scl-70; Anticentromere; SSa/Ro and SSb/La autoantibodies.

#### III. Radiological tests:

\* Plain X rays to the TMJ (lateral views both: open and closed mouth) (to exclude patients with mandibular osteolysis).

\*Plain chest X ray (posteroanterior and lateral views).

\*Barium swallow X ray.

#### IV. Ultrasonography of tempromandibular joint (U/S):

\*Ultrasonographic assessment of the tempromandibular joint was performed with a high-resolution real-time scanner, (LOGIQ, S 7 Expert GE) (UK). using a 8MHz linear transducer.

All patients and the controls were examined by the same radiologist who had no informations regarding the patients' clinical findings. The transducer was positioned against the skin surface of the TMJ in a transverse direction running parallel to the Camper line (the line intersecting the ala of the nose and the tragus of the ear). The transducer was placed to abut the tragus of the ear and was gradually shifted to obtain optimal visualization of the lateral pole of the mandibular condyle, which was clearly seen to be nearest to the skin surface while the patient or control was in both the closed- and open-mouth position, the distance between the articular capsule and the lateral surface of the mandibular condyle was measured in mm<sup>(43)</sup>, for both right and left sides.

#### (Figure 1)

#### V. Special investigations:

\* Pulmonary function tests; Ecocardiography; cardiac catheter.

\*The presence of secondary Sjögren syndrome was evaluated by the determination of salivary scintigraphy, salivary minor glands biopsy and the ophthalmologic tests including Schirmer Lissamine green test.<sup>(44)</sup>

#### Combined Rehabilitation Techniques:

##### \* Connective tissue massage with face mobilization:

Which is a manual technique used to treat altered connective tissues, in order to increase local blood flow and the release of involved tissue by connective

tissue stretching<sup>(22-24)</sup>. Massage should include eyebrows, forehead, area around the eyes, cheeks from nose to ears, area around the mouth, chin to ears and neck to collar bones. Facial mobilization carried at the same locations as massage with a technique that affects deeper structures, subcutaneous tissue, fascia & muscles and not only the skin; which is done with finger tips, where the tissues are grasped and stretched in all directions. **(Figure 2) and (Figure 3).**

**\*Kabat's method:**

It 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<sup>(31,32)</sup>. The Kabat's method concern the stimulation of the orbicularis oris, zygomaticus, levator labii, nasalis, buccinator, frontalis, and corrugator muscles **(Figure 4).**

**\*Kinesitherapy and TMJ Osteopathy:**

Which is a specific passive, active, assisted exercises and resisted exercise for temporal-mandibular joint (exercises for improving mouth opening and jaw lateralizing)<sup>(10,12)</sup>. **(Figure 5).**

**\*Program of home daily exercises:**

All patients of the two groups were instructed to perform daily mouth stretching and opening exercises<sup>(10,12)</sup> and grimacing exercises for mimic muscles<sup>(30)</sup>. Home exercises were done all through the 16 weeks of the study & were divided in three steps:

- In the first step (to be done for at least 5 minutes, 3 times/day), the patient placed thumbs into the mouth in order to enlarge the oral angles. The movements were done bilaterally and simultaneously.<sup>(10)</sup>

- In the second exercise step (to be performed once a day) the patient inserted a number of tongue depressors between the premolars of one arch towards the molars of the contralateral one to properly open the mouth. The tongue depressors should be maintained for a minimum of 8 minutes and it could be repeated if possible for additional 8 minutes, by increasing the number of tongue depressors.<sup>(10)</sup>

- The third exercise is a mimic exercises to be performed once a day, based on a series of grimaces to exercise orofacial muscles.<sup>(30)</sup>

\*\*\*The adherence to the exercises programme for (group A) was done in the inpatient and outpatient clinic, as well as for the home-based individual exercises

(group B) which was monitored by attendance lists and records for individual progress.

All patients (both groups) were assessed at baseline (F0), at the end of the 8 weeks of the treatment (F1) and after 8 weeks of follow-up (F2). They were evaluated with HAQ, SF-36, modified Rodnan skin score (for the face), mouth opening in

centimetres, Mouth Handicap in Systemic Sclerosis (MHSS) scale and U/S examination.

**Statistical Analysis:** <sup>(45)</sup>All data were collected, tabulated and statistically analyzed.

Analysis of data was done by an IBM computer using SPSS (statistical program for social science version 18) as follows:

- Description of quantitative variables as mean, SD and range,
- Description of qualitative variables as number and percentage,
- Chi-square test was used to compare groups as regards qualitative variables,
- ANOVA was used to compare data of repeated measures (F0, F1, and F2).
- An unpaired t-test was used to compare two groups as regard quantitative variable,
- p value >0.05 was considered insignificant, p > 0.05 significant and p <0.001 was considered highly significant.

**3. Results:**

This study was conducted on 40 SSc adult females, their age ranged from (45- 61) years, with a mean of  $54.28 \pm 10.91$  years, and a mean disease duration of  $9.4 \pm 4.4$  years, and a mean of total skin score of  $11.7 \pm 5.6$  years. Their subsets were; 22 patients (55 %) of ISSc, and 18 patients (45 %) of dSSc, the other descriptive data of the patients were presented in **(Table 1).**

Antinuclear antibody (ANA) were positive in all the patients (100 %); Anticentromere antibodies were positive in 8 patients (20%); while anti Scl-70 were positive in 20 patients (50%); ESR ranged from (12-30) with a mean of  $17.0 \pm 7.9$  mm/hour, these laboratory findings are shown in **(Table 2).**

\*On studying the clinical mouth opening: the mean  $\pm$  SD for the control group was  $5.90 \pm 2.31$  while that of the patients' was  $3.99 \pm 1.49$  with a high significant difference (p<0.001). **(Table 3)**

\*U/S measurements of the distance between the articular capsule and the lateral surface of the mandibular condyle showed that: the means of right and left Sides in (mm) for the control group were not significantly different, and also the means of right and left Sides in (mm) for the patients group were not significantly different. Thus on comparing the mean of controls  $1.89 \pm 0.41$  and that of patients  $1.94 \pm 0.35$  there was no significant difference (p>0.05). **(Table 4)**

\*The initial assessment F0 of all the parameters [(MSI (SF-36), PSI (SF-36), HAQ-DI, Mouth opening (cm), Facial skin score, MHSS and U/S measuring of distance between the articular capsule and the lateral surface of the mandibular (mm)] for both groups, didn't show any significant difference (p>0.05). **(Table 5)**



**Table 1: Description of the patients' clinical data with comparison between (Group A & Group B).**

Variables	SSc patients	Group A	Group B	P	S
Number	40	20	20	>0.05	NS
Sex	40 females	20	20	>0.05	NS
Age (years)	R (45-61) 54.28 ± 10.91	54.1 ± 11.01	54.01 ± 10.99	>0.05	NS
Subset (ISSc)	22	12	10	>0.05	NS
(dSSc)	18	8	10	>0.05	NS
Disease duration (years)	R(5.5-13 ys) 9.4 ± 4.4	9.1 ± 5.01	9.2 ± 4.9	>0.05	NS
Total skin score	R(12-21)				
MRSS	11.7 ± 5.6	10.7 ± 4.6	10.98 ± 5.7	>0.05	NS
ILD	21/40	10(50%)	11(55%)	>0.05	NS
PAH	14/40	7(35%)	7(35%)	>0.05	NS
Cardiac	19/40	10(50%)	9(45%)	>0.05	NS
Oesophageal	15/40	8(40%)	7(35%)	>0.05	NS
Musculo-skeletal	15/40	8(40%)	7(35%)	>0.05	NS
Sicca syndrome	24/40	11(55%)	13(65%)	>0.05	NS
Active ulcers	11/40	6(30%)	5(25%)	>0.05	NS
Raynaud's	40	20(100%)	20(100%)	>0.05	NS
Hypo-hyper pigmentations	4	2(10%)	2(10%)	>0.05	NS

SSc; systemic sclerosis, ISSc;limited; dSSc; diffuseSSc. ILD interstitial lung disease, PAH pulmonary hypertension, NS; non significance, R; range.

**Table 2: Laboratory data of the patients and the used medicine.**

Variant	Group A (N=20)	Group B (N= 20)
ANA	20 (100%)	20 (100%)
Anti-Scl-70	10 (50%)	10 (50%)
Anticentromere	3 (15%)	5 (25%)
ESR, mm/hour	18.0± 6.9	20.5± 8.9
Methotrexate	4 (20%)	2 (10%)
Prednisone	4 (20%)	1 (5%)
Azathioprine	2 (10%)	2 (10%)

ANA; anti nuclear antibody, ESR; erythrocyte sedimentation rate.

**Table 3: comparison between mean ± SD of mouth opening (cm) in both patients and controls.**

Variable	Patients	Controls	P	S
Mouth opening (cm)	Range (3.5–4.5) 3.99 ± 1.49	Range (5.0–6.5) 5.90± 2.31	<0.001	HS

HS; highly significant.

**Table 4: Comparison between US measurement (mean ± SD) of both (right & left) TMJs, as well as comparison between patients and controls.**

	Ultrasound right TMJ (mm)	Ultrasound left TMJ (mm)	P	S	Mean± SD (right & left)	P	S
Patients	Range(2.0–2.9) 1.99 ± 0.11	Range (2.0–2.6) 1.90± 0.39	>0.05	NS	1.94± 0.35		
Controls	Range(2.5–3.0) 1.91 ± 0.50	Range (2.0–2.9) 1.79 ± 0.30	>0.05	NS	1.89± 0.41	>0.05	NS

TMJ; tempromandibular joint, NS; none significant.

**Table 5: Comparison between initial assessments data (F0) of the two groups.**

Variables	Group A	Group B	P	S
MSI (SF-36)	38.18 ± 9.8	40.78 ± 7.99	>0.05	NS
PSI (SF-36)	37.88 ± 8.76	37.23 ± 8.00	>0.05	NS
HAQ-DI	0.49 ± 0.6	0.32 ± 0.03	>0.05	NS
Mouth opening (cm)	3.97 ± 1.09	4.1 ± 1.09	>0.05	NS
Facial skin score	3.99 ± 1.99	3.6 ± 1.4	>0.05	NS
MHISS	17.20 ± 8.99	18.10± 5.34	>0.05	NS
U/S measuring of distance between the articular capsule and the lateral surface of the mandibular condyle mean of right and left sides(mm)	1.90 ± 0.33	1.85 ± 0.34	>0.05	NS

MSI; mental synthetic index, PSI; physical synthetic index, HAQ-DI; health assessment questionnaire disease index, MHISS; mouth handicap in systemic sclerosis scale, US; ultrasound, NS; none significant, S; significant,

**Assessment of global health condition initially F0; and at follow up F1 & F2**

In both groups, neither the combined rehabilitation techniques nor the home exercise program modified the Physical and Mental Summary Indexes of SF36 and HAQ-DI as well, at the end of the study (F1) and at follow-up (F2), (**Table 7 and 8**).

**Face assessment**

**Mouth opening:** Patients of both groups showed improvement from the treatments for mouth opening. In (group A), a significant increase of mouth opening was shown at F1 ( $4.1 \pm 0.04$ ) and still maintained at F2 ( $4.3 \pm 0.9$ ), ( $p < 0.05$  versus F0, respectively). Moreover, In (group B), the home exercise program improved mouth opening at F1 ( $p < 0.05$  versus F0), and F2 ( $p < 0.05$  versus F0). (**Table 7 and 8**).

**Facial skin score:** In (group A), the combined rehabilitation techniques decreased significantly skin score at F1 ( $1.97 \pm 0.91$ ), ( $p < 0.001$  versus F0). The decrease of the skin score was still significant at F2 ( $1.5 \pm 1.09$ ), ( $p < 0.05$  versus F0). In group B; the home exercise program did not modify skin score throughout the study.

( $p > 0.05$ ). (**Table 7 and 8**).

**MHISS scale:** The mean of the initial (F0) MHISS score in the two groups of patients was not significantly different, and also the scores of subscale 1 (handicap related to reduced mouth opening) and scores of subscales 2 (handicap related to sicca syndrome) and 3 (aesthetic concerns) respectively were not significantly different in the two groups ( $p > 0.05$ ). (**Table 6**).

**Table 6: Comparison between mean  $\pm$  SD of MHISS and its subtypes for Group A & Group B.**

Variant	Group A	Group B	P	S
<b>MHISS</b>	17.20 $\pm$ 8.99	18.10 $\pm$ 5.34	>0.05	NS
Subscale 1	6.96 $\pm$ 2.05	6.55 $\pm$ 1.90	>0.05	NS
Subscale 2	7.02 $\pm$ 1.99	6.98 $\pm$ 1.85	>0.05	NS
Subscale 3	3.50 $\pm$ 1.09	2.90 $\pm$ 1.0	>0.05	NS

MHISS; mouth handicap in systemic sclerosis scale, S; significant, NS; none significant.

In (group A), the combined rehabilitation techniques significantly improved the scores of MHISS scale at F1 ( $16.83 \pm 4.9$ ), versus F0 ( $17.20 \pm 8.99$ ) ( $p < 0.001$ ), and what is interesting is that; the improvement was in the subscale 1 which indicates a high benefit of the rehabilitation techniques in improving the mouth opening and decreasing the associated pain and difficulties associated with chewing. At F2 ( $16.15 \pm 5.4$ ) MHISS scores didn't improve versus scores obtained at F0 ( $p > 0.05$ ), which indicates that the home based program did not maintain the effect obtained by the combined program during the previous 8 weeks. Moreover, In group B, the home exercise program did not provide any

significant change in MHISS throughout the study ( $p > 0.05$ ). (**Table 7 and 8**).

\*Using ultrasound measurements of TMJ, the distance between the articular capsule and the mandibular condyle did not show differences between both groups at different stages (F0, F1, & F2) of the study ( $p > 0.05$ ). From these results, we could state that there was no disc displacement diagnosis in the patients group, according to the parameters used by (**Hayashi et al, 2001**) (who stated that whenever the distance between the articular capsule and the lateral surface of the condyle  $\geq 4$  mm it is an indicator of disc displacement)<sup>(46)</sup>. (**Table 7 and 8**).

**Table 7: Comparison between different assessment data for (Group A) at different stage (F0, F1, & F2).**

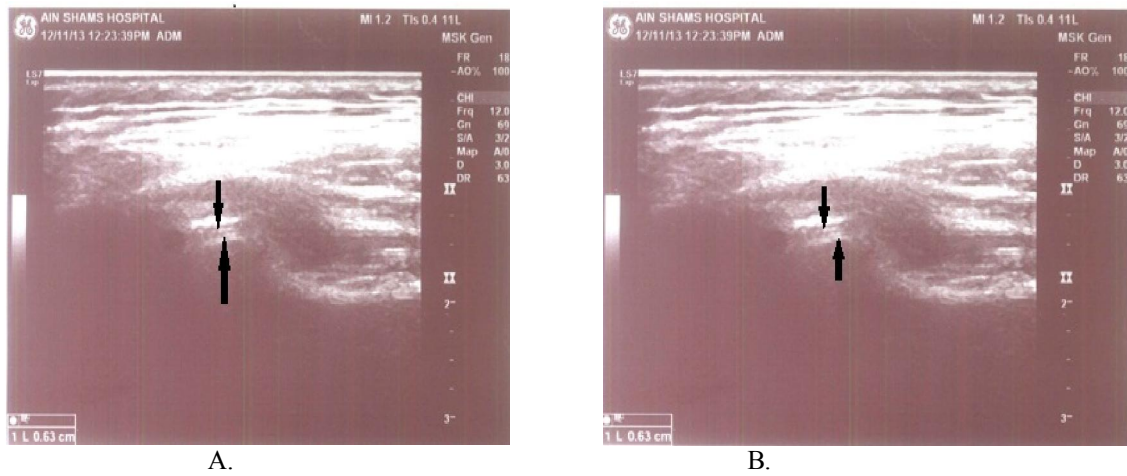
Variables	F0	F1	F2	F0- F1	F0- F2
	Mean $\pm$ SD	Mean $\pm$ SD	Mean $\pm$ SD	P	P
MSI (SF-36)	38.18 $\pm$ 9.8	39.75 $\pm$ 7.01	40.01 $\pm$ 10.01	>0.05 NS	>0.05 NS
PSI (SF-36)	37.88 $\pm$ 8.76	39.9 $\pm$ 7.45	40.01 $\pm$ 7.05	>0.05 NS	>0.05 NS
HAQ-DI	0.49 $\pm$ 0.6	0.31 $\pm$ 0.12	0.33 $\pm$ 0.9	>0.05 NS	>0.05 NS
Mouth opening (cm)	3.97 $\pm$ 1.09	4.1 $\pm$ 0.04	4.3 $\pm$ 0.9	<0.05 S	<0.05 S
Facial skin score	3.99 $\pm$ 1.99	1.97 $\pm$ 0.91	1.5 $\pm$ 1.09	<0.001 HS	<0.05 S
MHISS	17.20 $\pm$ 8.99	16.83 $\pm$ 4.9	16.15 $\pm$ 5.4	<0.001 HS	>0.05 NS
U/S measure of distance between the articular capsule and the lateral surface of the mandibular condyle mean of right and left Sides (mm)	1.90 $\pm$ 0.33	1.93 $\pm$ 1.9	2.01 $\pm$ 1.05	>0.05 NS	>0.05 NS

MSI; mental synthetic index, PSI; physical synthetic index, HAQ-DI; health assessment questionnaire disease index, MHISS; mouth handicap in systemic sclerosis scale, US; ultrasound, NS; none significant, S; significant, HS; highly significant, (F0, F1, F2) data at base line F0, follow up F1 and follow up F2.

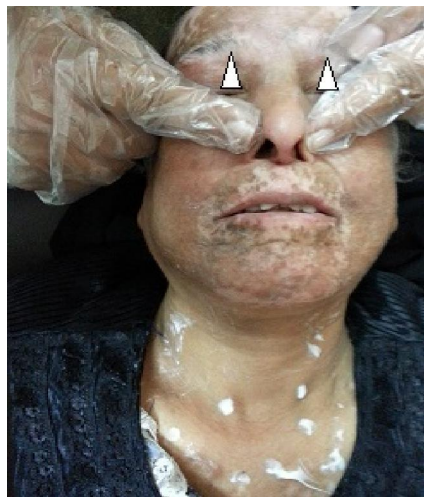
**Table 8: Comparison between different assessment data for (Group B) at different stage (F0, F1, & F2)**

Variables	F0 Mean± SD	F1 Mean± SD	F2 Mean± SD	F0- F1 P	F0- F2 P
MSI (SF-36)	40.78 ± 7.99	40.80 ± 7.87	40.56 ± 7.90	>0.05 NS	>0.05 NS
PSI (SF-36)	37.23 ± 8.00	36.9 ± 8.42	37.01 ± 7.95	>0.05 NS	>0.05 NS
HAQ-DI	0.32 ± 0.03	0.33 ± 0.11	0.32 ± 0.01	>0.05 NS	>0.05 NS
Mouth opening (cm)	4.1 ± 1.09	4.4 ± 1.04	4.3 ± 1.05	< 0.05 S	< 0.05 S
Facial skin score	3.6 ± 1.4	3.6± 1.1	3.5± 1.8	>0.05 NS	>0.05 NS
MHISS	18.10± 5.34	18.83± 4.95	17.93± 4.6	>0.05 NS	>0.05 NS
U/S measuring of distance between the articular capsule and the lateral surface of the mandibular condyle mean of right and left sides(mm)	1.85 ± 0.34	2.03± 1.1	2.1± 1.32	>0.05 NS	>0.05 NS

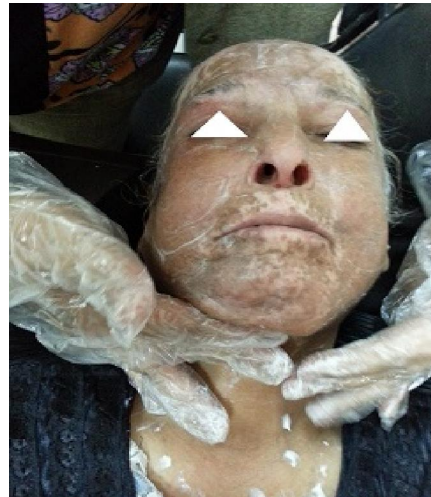
MSI; mental synthetic index, PSI; physical synthetic index, HAQ-DI; health assessment questionnaire disease index, MHISS; mouth handicap in systemic sclerosis scale, US; ultrasound, NS; none significant, S; significant, (F0, F 1,& F2); data at base line F0, follow up F 1 and follow up F 2.



**Figure 1:** Ultrasonography of TMJ joint; arrows showing the distance between the articular capsule and the lateral surface of the mandibular condyle (mm), A; before, B; after rehabilitation.

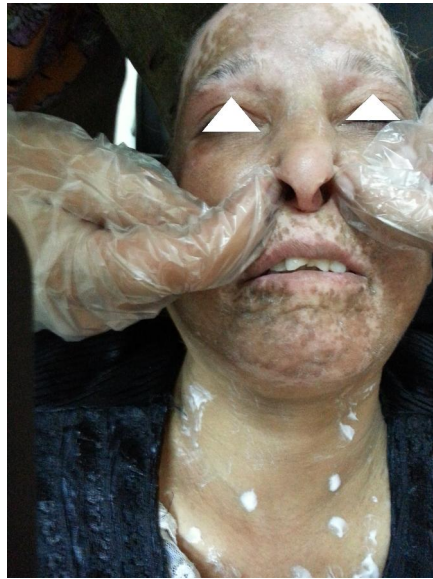


**A. Nasalis**



**B. Neck**





C. Zygomaticus & levator labii



D. Cheeks.

Figure 2: A; massage of nasalis, B; neck & collar area, C; Zygomaticus, D; cheeks.



A



B



C

Figure 3: A; mobilization of orbicularis oris, B; mobilization of lower lip, C; mobilization of frontalis muscles.





Figure 4: Kabat's method A; frontalis, B; orbicularis oris, C; Zygomaticus.

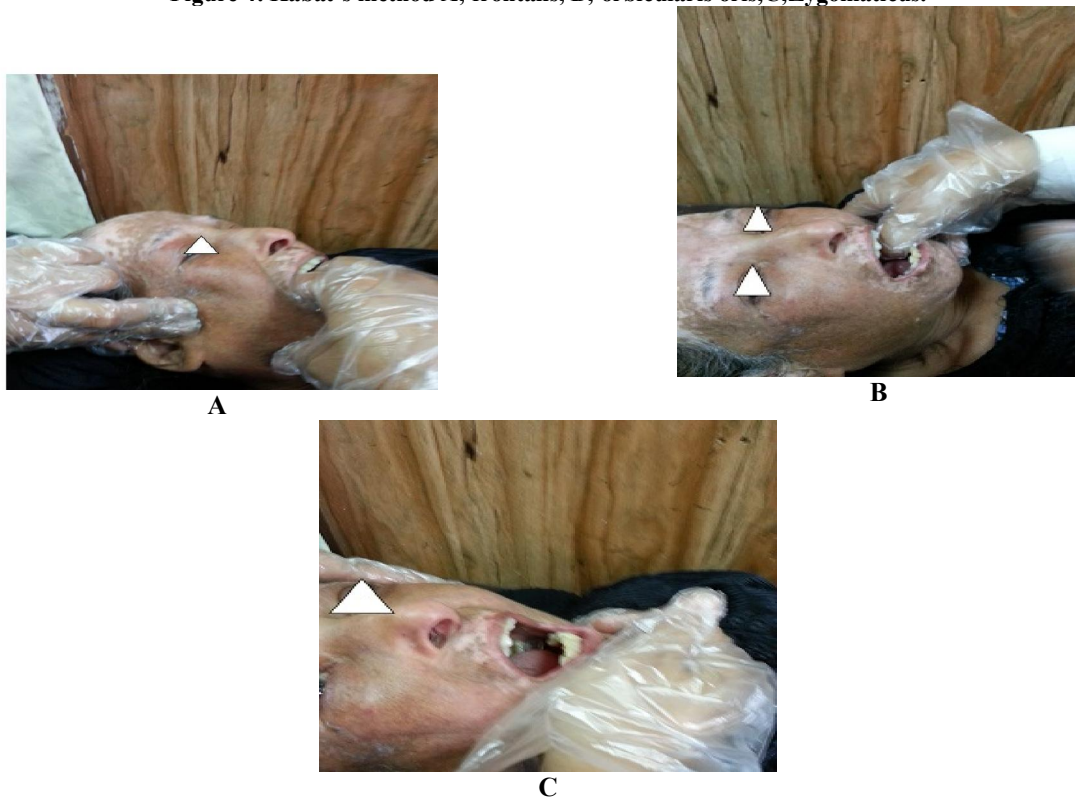


Figure 5: A,B Osteopathy of tempromandibular joint, C; active and resisted exercise.

#### 4. Discussion:

SSc leads to fibrosis of facial soft tissues with disappearance of the cutaneous facial furrows, diminished mouth opening and width, altered dentition, difficulties in dental intervention, with concomitance of sicca syndrome and changes at temporo-mandibular joints often lead to impairment of QoL. In fact, the fibrotic involvement of the face became one of the SSc patients' greatest complaints<sup>(47,48)</sup>, with concern about disfigurement<sup>(49)</sup> and personal self-esteem<sup>(50)</sup> and it was also shown that, in SSc, skin deformities are a core stressor of the disease<sup>(50)</sup>. We have chosen the combination of connective tissue massage and mobilization (able to modify, by stretching, bloodstream and releasing the connective tissues) and Kabat's method (a neurorehabilitation technique, that reinforces neuromuscular recruitment)<sup>(40,41)</sup>, with the traditional kinesiotherapy and manipulation osteopathy of the tempromandibular,<sup>26</sup> joint and home exercises for mouth and mimic muscles, to study their effect on facial rehabilitation in a study of 8 weeks duration and reassessment after another 8 weeks of follow up in a cohort of Egyptian patients with SSc.

It is clear that the treatment of the face did not exert any effect on global function. From our data, it is evident that the treatment of the face had no effect on the values of HAQ DI and SF36 (PI & MI) in both assessments F1 and F2, for both groups; this is probably due to the relatively short period of the treatment and to the fact that the patients were treated with techniques not aimed to the overall body, but specifically conceived for the involvement of the face which was noticed also in a previous study<sup>(42)</sup>, where their designed rehabilitation programme for facial rehabilitation for 9 weeks; in SSc patients didn't improve any of the global health function, although showed a valuable benefits as regards mouth opening and improvement of facial skin score and the MHISS score.

Our data of this combined rehabilitation programme as regards the mouth opening, facial skin score and the MHISS score showed a significant improvement in group A and this effect was maintained even in the follow up where the patients were kept only on the home exercise programme, this can be due to the efficacy of the different techniques acting synergically and to the fact that patients continued, for a 8 week period after the end of the treatment, the program of mimic and stretching exercises in a home self management program. which was supported by a previous study<sup>(42)</sup>. As regards MHISS it is a new scale with an excellent reliability and good construct validity, in the specific assessment of disability involving the mouth in SSc patients<sup>(32)</sup>. Our results showed that; MHISS was helpful in

following-up SSc patients over time for evaluating the outcome of the rehabilitation programmes.

It is interesting to note that the improvement of mouth opening was the only effect of home exercises in group B, which was not lost at follow up and, however no effect was recorded for facial skin thickness or MHISS score although this finding was not supported by the previous study<sup>(42)</sup> as our patients were adherent to daily home exercise programme till the reassessment at F2, which suggests that the continuity of care is mandatory in the rehabilitation of patients affected with chronic rheumatic diseases, such as SSc, in order to maintain its efficacy<sup>(51)</sup>, and patients are encouraged to become part of the health care team and the concept of self-care or self-management plays a central role<sup>(52)</sup> in rehabilitation programmes.

Some studies showed the effect and benefits of the rehabilitation techniques such as paraffin was, massage and stretching and range of motion exercises of the hands.<sup>(8,53)</sup>

Moreover, some evidence exists about the effectiveness of self-management in SSc patients. *Mugii et al* studied the efficacy of self-administered stretching of each finger in SSc patients, showing that amelioration of range of motion was present in each finger after 1 month and maintained within 1 year.<sup>(53)</sup>

We augmented our results by using the U/S study of TMJ, where there was no association between the articular capsule and the lateral condyle surface distances measured by ultrasound using a 8 MHz linear transducer and the clinical state of the patient even in the repeated follow up at F1 and F2 and these results didn't differ from the values of normal controls which highlights; that the initial limitation of the mouth opening was not due to TMJ dislocation<sup>(45)</sup> and whenever the pain and soft tissue mobilization of the facial muscles and TMJ manipulation were improved the mouth opening was improved. Further studies in diagnostic imaging of TMJs with ultrasound should be encouraged, since it has some useful diagnostic applications and does not require special facilities. The limitations of the present findings must be taken into account regarding the signs and symptoms of musculoskeletal disorders in a time dependent context, suggesting the need to carry out repeated clinical records<sup>(54)</sup>. Our data show that, in SSc, a combined rehabilitation approach is significantly more effective than a home exercise program in reducing skin thickness of the face, in recovering mouth opening and in improving self-reported face and mouth related symptoms as well as improved the subjective perceptions and the self perceived disability due to face and mouth involvement.

Due to the complexity and the severity of SSc, the rehabilitation program proposed in the present work should be regarded as a support tool in SSc management. However this combined program should be integrated with global rehabilitation techniques on top of the pharmacological treatments for the disease.<sup>(55,56)</sup>

### Conclusion:

\* The combined rehabilitation techniques, together with home based exercises are effective in the rehabilitative treatment of SSc face. With a main benefits for mouth opening, skin score and subjective perceptions and the self perceived disability due to face and mouth involvement.

\* Further studies are needed to evaluate the long term effects of this combined program.

### References:

1. LeRoy EC, Black C, Fleischmajer R, Jablonska S, Krieg T, Medsger TA Jr, et al. Scleroderma (systemic sclerosis): classification, subsets and pathogenesis. *J Rheumatol* 1988; **15**: 202–5.
2. Medsger TA Jr. Natural history of systemic sclerosis and the assessment of disease activity, severity, functional status, and psychologic well-being. *Rheum Dis Clin North Am* 2003; **29**: 255–73.
3. Sandqvist G, Scheja A, Eklund M. Working ability in relation to disease severity, everyday occupations and well-being in women with limited systemic sclerosis. *Rheumatology (Oxford)*. 2008;47:1708–1711.
4. Hudson M, Thombs BD, Steele R, Panopalis P, Newton E, Baron M, et al. Canadian Scleroderma Research Group. Health-related quality of life in systemic sclerosis: a systematic review. *Arthritis Rheum*. 2009;61(8):1112–1120.
5. Pope JE, Bellamy N. Outcome measures in scleroderma clinical trials. *Semin Arthritis Rheum*. 1993;23:22–33.
6. Medsger TA Jr. Natural history of systemic sclerosis and the assessment of disease activity, severity, functional status, and psychologic well-being. *Rheum Dis Clin North Am* 2003; **29**: 255–73.
7. Smyth AE, MacGregor AJ, Mukerjee D, Brough GM, Black CM, Denton CP. A cross-sectional comparison of three self-report functional indices in scleroderma. *Rheumatology (Oxford)*. 2003;42:732–738.
8. Poole J. Musculoskeletal rehabilitation in the person with scleroderma. *Curr Opin Rheumatol*. 2010;22(2):205.
9. Del Rosso A, Boldrini M, D'Agostino D, Placidi GP, Scarpato A, Pignone A. Health-related quality of life in systemic sclerosis as measured by the Short Form 36: relationship with clinical and biologic markers. *Arthritis Rheum* 2004;51:475-81.
10. Pizzo G, Scardina GA, Messina P. Effects of a non surgical exercise program on the decreased mouth opening in patients with systemic scleroderma. *Clin Oral Investig*. 2003;7:175-8.
11. Nagy G, Kovács J, Zeher M, Czirják L. Analysis of the oral manifestations of systemic sclerosis. *Oral Surg Oral Med Oral Pathol*. 1994;77:141-6.
12. Naylor P, Chester WD, Mix E. The nonsurgical treatment of microstomia in scleroderma: a pilot study. *Oral Surg Oral Med Oral Pathol* 1984; **57**:508–511.
13. Cazol C, Sobral AP, Neves RF, Freire Filho FW, Cardoso AB, da Silveira MM. Oral complaints in progressive systemic sclerosis: two cases report. *Med Oral Patol Oral Cir Bucal*. 2008;13:E114-8.
14. Albilis JB, Lam DK, Blanas N, Clokie CM, Sándor GK. Small mouths. Big problems? A review of scleroderma and its oral health implications. *J Can Dent Assoc*. 2007;73:831-6
15. Shah AA, Wigley FM. Often forgotten manifestations of systemic sclerosis. *Rheum Dis Clin North Am*. 2008;34:221-38.
16. Mouthon L, Rannou F, Berezne A, Pagnoux C, Arene JP, Fois E, et al. Development and validation of a scale for mouth handicap in systemic sclerosis: the mouth handicap in systemic scleroderma scale. *Ann Rheum Dis*. 2007; **66**:1651-5.
17. Antonioli CM, Bua G, Frige A, Prandini K, Radici S, Scarsi M, et al. An individualized rehabilitation program in patients with systemic sclerosis may improve quality of life and hand mobility. *Clin Rheumatol* 2009; **28**: 159–65.
18. Maddali Bongi S, Del Rosso A, Galluccio F, Tai G, Sigismondi F, Passalacqua M, et al. Efficacy of a tailored rehabilitation program for systemic sclerosis. *Clin Exp Rheumatol* 2009; **27** Suppl 54: 44–50.
19. Maddali-Bongi S, Landi G, Galluccio F, Del Rosso A, Miniati I, Conforti ML, et al. The rehabilitation of facial involvement in systemic sclerosis: efficacy of the combination of connective tissue massage, Kabat's technique and kinesitherapy: a randomized controlled trial. *Rheumatol Int* 2010.
20. Brown SJ, Somerset ME, McCabe CS, McHugh NJ. The impact of group education on participants' management of their disease in lupus and scleroderma. *Musculoskeletal Care* 2004;2: 207–17.
21. Samuelson UK, Ahlmen EM. Development and evaluation of a patient education program for persons with systemic sclerosis (scleroderma). *Arthritis Care Res* 2000; **13**: 141–8. Direct Link
22. Oliveira NC, Dos Santos Sabbag LM, de Sa Pinto AL, Borges CL, Lima FR. Aerobic exercise is safe and effective in systemic sclerosis. *Int J Sports Med* 2009; **30**: 728–32.
23. Mugii N, Hasegawa M, Matsushita T, Kondo M, Orito H, Yanaba K, et al. The efficacy of self-administered stretching for finger joint motion in Japanese patients with systemic sclerosis. *J Rheumatol* 2006; **33**: 1586–92.
24. Pizzo G, Scardina GA, Messina P. Effects of a nonsurgical exercise program on the decreased mouth opening in patients with systemic scleroderma. *Clin Oral Investig* 2003; **7**: 175–8.
25. Maddali-Bongi SM, Del Rosso A, Galluccio F, Sigismondi F, Miniati I, Conforti ML, et al. Efficacy of connective tissue massage and Mc Mennell joint manipulation in the rehabilitative treatment of the hands in systemic sclerosis. *Clin Rheumatol* 2009; **28**:1167–73.
26. Howell, Joel D. (1999). "The Paradox of Osteopathy". *New England Journal of Medicine* 341 (19): 1465–8.
27. Van den Hoogen F, Khanna D, Fransen J, Johnson SR, Baron M, Tyndall A, Matucci-Cerinic M, Naden RP, Medsger TA Jr, Carreira PE, Riemekasten G, Clements PJ, Denton CP, Distler O, Allanore Y, Furst DE, Gabrielli A, Mayes MD, van Laar JM, Seibold JR, Czirjak L, Steen



- VD, Inanc M, Kowal-Bielecka O, Müller-Ladner U, Valentini G, Veale DJ, Vonk MC, Walker UA, Chung L, Collier DH, Ellen Csuka M, Fessler BJ, Guiducci S, Herrick A, Hsu VM, Jimenez S, Kahaleh B, Merkel PA, Sierakowski S, Silver RM, Simms RW, Varga J, Pope JE. 2013 classification criteria for systemic sclerosis: an American college of rheumatology/European league against rheumatism collaborative initiative. *Ann Rheum Dis*. 2013 Nov 1;72(11):1747-55.
28. Clements P, Lachenbruch P, Seibold J et al (1995) Inter and intraobserver variability of total skin thickness score (modified Rodnan TSS) in systemic sclerosis. *J Rheumatol* 22:1281–1285.
  29. Goats GC, Keir KA. Connective tissue massage. *Br J Sports Med* 1991;25:131-3.
  30. Alpiner N, Oh TH, Hinderer SR, Brander VA. Rehabilitation in joint and connective tissue diseases. 1. Systemic Diseases. *Arch Phys Med Rehabil*.1995; 75: S32-S40.
  31. Greenman PE. Mobilization with and without impulse technique. 107-112, *Principles of Manual Medicine*. Philadelphia, (3rd edition) Lippincott Williams & Wilkins 2003.
  32. Kabat H, McLeod M, Holt C. The practical application of proprioceptive neuromuscular facilitation. *Physiotherapy* 1959;45:87-92.
  33. Kendall FP, Kendall EM, Provance PG. *Muscles: functions and test*. Lippincott Editor, 2002.
  34. The assessment of the patient with systemic sclerosis. *Clin Exp Rheumatol* (2003) 21(Suppl. 29):S48–S56.
  35. Vitali C, Bombardieri S, Jonsson R et al (2002) Classification criteria for Sjögren's syndrome: a revised version of the European criteria proposed by the American-European Consensus Group. *Ann Rheum Dis* 61:554–558.
  36. Akesson A, Fiori G, Krieg T, van den Hoogen FHJ, Seibold JR. Assessment of skin, joint, tendon and muscle involvement. *Clin Exp Rheumatol*. 2003;21(Suppl.29):S5–S8.
  37. Johnson SR, Lee P. The HAQ disability index in scleroderma trials. *Rheumatology (Oxford)* 2004;43:1200-1.
  38. La Montagna G, Cuomo G, Chiarolanza I, Ruocco L, Valentini G. HAQ-DI Italian version in systemic sclerosis. *Reumatismo* 2006; 58(2):112-5.
  39. Poole J, Steen V (1991). The use of the health assessment questionnaire (HAQ) to determine physical disability in systemic sclerosis. *Arthritis care Res*; 4: 27-31.
  40. Ware JE, Sherbourne CD. The MOS 36-Item Short Form Health Survey (SF-36): I. Conceptual framework and item selection. *Medical Care* 1992; 30:473-83.
  41. Apolone G, Mosconi P: The Italian SF-36 Health Survey: Translation, Validation and Norming. *J Clin Epidemiol* 1998; 51:1025–36.
  42. Maddali Bongi S, Del Rosso A, Miniati I, Galluccio F, Landi G, Tai G, Matucci-Cerinic M. The Italian version of the Mouth Handicap in Systemic Sclerosis scale (MHSS) is valid, reliable and useful in assessing oralhealth-related quality of life (OHRQoL) in systemic sclerosis (SSc) patients *Rheumatol Int* 2012;32:2785–2790.
  43. Kazanoglu A, Unger J. Determining the occlusal plane with the Camper's plane indicator. *J Prosthet Dent* 1992;67:499-501.
  44. HK Yuen' Y Weng, SG Reed, LM Summerlin, RM Silver. Factors associated with gingival inflammation among adults with systemic sclerosis *International journal of dental hygiene* 24 APR 2013
  45. Courtney Taylor. *What is ANOVA. Statistics- about.com. direct link.*
  46. Hayashi T, Ito J, Koyama J, Yamada K. The accuracy of sonography for evaluation of internal derangement of the temporomandibular joint in asymptomatic elementary school children: comparison with MR and CT. *Am J Neuroradiol* 2001; 22:728–734.
  47. Joachim G, Acorn S. Life with a rare chronic disease: the scleroderma experience. *J Adv Nurs* 2003;42:598–606.
  48. Benrud-Larson LM, Heinberg LJ, Boling C et al. Body image dissatisfaction among women with scleroderma: extent and relationship to psychosocial function. *Health Psychol* 2003;22:130–9.
  49. Paquette DL, Falanga V. Cutaneous concerns of scleroderma patients. *J Dermatol* 2003;30:438–43.
  50. van Lankveld W, Vonk MC, Teunissen H, van den Hoogen F. Appearance self-esteem in systemic sclerosis—subjective experience of skin deformity and its relationship with physician-assessed skin involvement, disease status and psychological variables. *Rheumatology* 2007;46:872–876.
  51. Dagfinrud H, Kvien TK, Hagen KB. Physiotherapy interventions for ankylosing spondylitis. *Cochrane Database Syst Rev* 2008 Jan 23;(1).
  52. Bodenheimer T, Lorig K, Holman H., Grumbach K. Patient self management of chronic disease in primary care. *JAMA* 2002 288, 2469-75.
  53. Mugii N, Hasegawa M, Matsushita T, Kondo M, Orito H, Yanaba K, Komura K, et al. The efficacy of self-administered stretching for finger joint motion in Japanese patients with systemic sclerosis. *J Rheumatol* 2006;33:1586-92.
  54. Pereira L, Gavia M, Bonjardim L and Castelo P. *Dentomaxillofacial Radiology* (2007) 36, 402–408.
  55. Allanore Y, Avouac J, Wipff J, Kahan A. New therapeutic strategies in the management of systemic sclerosis. *Expert Opin Pharmacother* 2007;8:607-15.
  56. Matucci Cerinic M, Del Rosso A, Perfetto F, Livi R, Fiori G, Bartoli F et al. Therapeutic challenges for systemic sclerosis: facts and future targets. *Ann NYAcad Sci* 2007;1110:448-54.