



## The impact of altered hands function on working ability of patients with systemic sclerosis

### Uticaj izmenjenih funkcija šaka na radnu sposobnost obolelih od sistemske skleroze

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#### Abstract

**Background/Aim.** Hand impairment in systemic sclerosis (SSc) patients is the most frequent manifestation of this progressive disease and often cause problems in daily activities and working ability. The correlation of altered hand functions in patients with SSc and their impact on working ability is not fully explained. The objective of this study is to assess the impact of altered hand functions of patients with SSc on their working ability. **Methods.** We assessed 20 patients with SSc (17 females, 3 males), with mean age of  $45.5 \pm 11.9$  years (min 29, max 69, Med 44.0 years). The movements of fingers, wrist and forearm were examined by measuring active range of motion of the hand and fingers, muscle strength of the fingers, the skin lesions by modified Rodnan score, the function of the fingers, hands and forearms by the Hand Mobility in Scleroderma (HAMIS) test, as well as the condition of the capillaries in the fingers by capillaroscopy and working capacity by Work Ability Index (WAI). **Results.** Reduced strength on at least one tested muscle, was established in all patients, thickening of the skin on the hands in 14/20 (70%) and the fingers in 19/20 (95%), “scleroderma type” capillary changes in 15/17 (85%), decreased range of motion in finger joints in 16/20

(80%) of the patients. Also, 14/20 (70%) of subjects reported problems at work [decrease performance achieved in 8/20 (40%), reduction of working hours in also 8/20 (40%), leaving out some of the work in 14/20 (70%), and investing of extra effort in 15/20 (75%)]. By means of WAI, work ability assessment questionnaire, it was found that the working capacity was reduced in 11/20 (55%) of our patients. None of the patients was in the group of the most serious cases, the “poor” category; in the “moderate” category there were 6/20 (30%), while 5/20 (25%) of the patients were in the group with minimally reduced working capacity (“good” category). There was a statistically significant correlation between the thickened skin on the fingers ( $p < 0.05$ ), reduced muscle strength in the fingers ( $p < 0.002$ ) and limited mobility of the individual finger joints ( $p < 0.05$ ), with information on reduced working capacity obtained by means of WAI questionnaire and with answers to questions about problems at work. **Conclusion.** The thickened skin on the fingers, reduced fingers muscle strength and limited mobility of certain finger joints in patients with SSc cause decreasing working capacity for all jobs that include manual activities.

**Key words:** scleroderma, systemic; work capacity evaluation; hand.

#### Apstrakt

**Uvod/Cilj.** Oštećenje šaka kod obolelih od sistemske skleroze (SSc) je najčešća manifestacija ovog progresivnog oboljenja i često uzrokuje probleme tokom svakodnevnih aktivnosti i radnih obaveza. Povezanost izmenjenih funkcija šaka obolelih od sistemske skleroze i njihov uticaj na radnu sposobnost nije u potpunosti objašnjen. Cilj ovog rada je da se kod obolelih od SSc ispita povezanost izmenjenih funkcija šaka sa njihovom radnom sposobnošću. **Metode.** Ispitano je 20 bolesnika sa SSc (17 ženskog i 3 muškog pola), prosečne starosti  $45,5 \pm 11,9$  godina (min 29, max 69, Med 44,0 godine). Ispitana je pokretljivost prstiju, ručja i podlaktice

kroz merenje aktivnog obima pokreta šake i prstiju, snaga mišića prstiju ruku, rasprostranjenost kožnih promena modifikovanim Rodnanovim kožnim skorom, funkcije prstiju, šaka i podlaktica *Hand Mobility in Scleroderma* (HAMIS) testom, stanje kapilara na prstima ruku kapilaroskopijom i procena radne sposobnosti primenom *Work Ability Index* (WAI). **Rezultati.** Smanjena mišićna snaga na bar jednom ispitivanom mišiću, utvrđena je kod svih ispitanika, zadebljanje kože na šakama kod 14/20 (70%), na prstima kod 19/20 (95%), promene kapilara “sklerodermnog tipa” kod 15/17 (85%), smanjen obim pokreta u zglobovima prstiju kod 16/20 (80%). Na probleme na poslu žalilo se 14/20 (70%) ispitanika [na smanjenje postignutog učinka (8/20)

(40%), na skraćivanje radnih sati, takođe, 8/20–40%, na izostavljanje nekih radnji 14/20 (70%), na ulaganje dodatnog napora 15/20 (75%)]. Procenom radne sposobnosti WAI upitnikom utvrđeno je da je radna sposobnost umanjena kod 11/20 (55%) ispitanika. U grupi najtežih slučajeva, kategoriji „loše“, tj. ispitanika gde je bila izraženo umanjena radna sposobnost, nije bilo nijednog ispitanika; u grupi koja pripada kategoriji „osrednje“ bilo ih je 6/20 (30%) i u grupi sa najmanje umanjenom radnom sposobnošću (kategorija „dobro“) 5/20 (25%) ispitanika. Nađena je statistički značajna povezanost između zadebljale kože na prstima ruku ( $p < 0,05$ ), smanjene mišićne snage prstiju ( $p < 0,002$ ) i

ograničene pokretljivosti pojedinih zglobova prstiju ( $p < 0,05$ ), sa podacima o umanjenoj radnoj sposobnosti dobijenim WAI upitnikom i sa odgovorima na pitanja o problemima na poslu. **Zaključak.** Zadebljala koža na prstima ruku, smanjena mišićna snaga prstiju i ograničena pokretljivost pojedinih zglobova prstiju kod bolesnika sa SSC uzrokuju smanjenje radne sposobnosti za sve poslove vezane za manuelne aktivnosti.

**Ključne reči:**  
**sklerodermija, sistemska; sposobnost, radna, ocena; ruka.**

## Introduction

Systemic sclerosis (SSc) is a chronic, systemic, progressive, autoimmune disease, characterized by lesions on the small blood vessels, excessive deposition of collagen and other extracellular connective tissues in the skin, locomotor system and some internal organs. Etiology is unknown, but genetic predisposition plays an important role in development of fibroproliferative changes<sup>1-4</sup>.

The correlation of altered hand functions in patients with SSc and their impact on working capacity is not fully explained. The results of a number of previous studies which were conducted both in our country and worldwide, were not coherent, and controversial conclusions are explained by different methodological approaches and by evaluating insufficiently specific and characteristic parameters, stages of the disease in which the studies were performed, effects of various therapeutic procedures at the time of testing, and the individual characteristics of subjects.

There are many indicators of the functioning of the hand: anatomical integrity, mobility, muscular strength, sensitivity, capturing function, accuracy, coordination, unilateral and bilateral tasks, daily activities. None of the methods of assessment includes all the functions of the hand.

Working capacity implies appropriate anatomical and functional condition of organs and systems that enables to fully comply with all duties related to occupational or daily activities. It also means the ability of an organism to maintain the internal physiological balance during operation, and after the termination of work promptly and fully establish all forms of balance that deviated from the physiological values<sup>5</sup>.

The objective of this study was to evaluate the correlation between altered function of the hand in patients with systemic sclerosis with their working capacity.

## Methods

The study was conducted at the Institute of Rheumatology in Belgrade, Serbia and included thorough exam of 20 adult patients (17 women and 3 men) with a confirmed diagnosis of SSc without other systemic connective tissue diseases. All subjects were aware of the aim of this study and signed a voluntary consent to participate, and the two independent Ethic Committees approved its implementation.

Evaluation of health status of the patients was marked from 1 to 5 (1 – excellent, 2 – very good, 3 – good, 4 – average, 5 – bad).

Physical pain was marked also from 1 to 5 (1 – no pain, 2 – very weak pain, 3 – weak pain, 4 – moderate pain, 5 – strong pain).

For the evaluation of the extent and severity of skin lesions, the modified Rodnan skin score (mRSS) was used<sup>6</sup>. It is determining skin involvement by palpation of seventeen anatomic sites and scoring on a 0–3 scale (0 – normal finding, 1 – possible skin thickening, 2 – thickened skin not attached to deeper layers, 3 – thickened skin attached to deeper layers). The scores for all sites were summed to give a total skin score, with a minimum possible score of 0, and maximum possible score of 51 points. Of all estimated anatomic sites, only regional skin indicators of four sites (thickening of the skin on fingers and hands, left and right) were taken for further analyses in this study. They were also scored from 0 to 3 (see above). Minimum possible score was 0, maximum was 12 points.

Condition of the capillaries was evaluated semiquantitatively by the Maricq method of capillaroscopy, with minor modifications according to Damjanov<sup>7</sup>. Marks were from 1 to 4 (1 – unspecific changes, 2 – dilated capillaries with minor avascular areas, 3 – dilated capillaries with large avascular areas, 4 – very large avascular areas). For the classification of observed capillary changes, we also used the classification by Cutolo with marks from 1 to 4 (1 – normal findings, 2 – early changes, 3 – active changes, 4 – late changes).

The mobility of the wrist and fingers were monitored by measuring (on both hands) the flexion and extension of the fingers, thumb abduction and adduction, as well as palmar and dorsal flexion (extension) of the wrist. Mobility of the forearm was determined by measuring pronation and supination. Active range of motion (AROM) was measured on both arms in angle degrees at the level of the wrist radiocarpal joint (RC), in the metacarpophalangeal joints (MCP) and interphalangeal finger joints (IP, distal DIP and proximal PIP) and expressed as a percentage of the maximum possible values. In this study, only the mobility of the first three fingers of both hands, which are the most important for the function of capturing, holding and manipulating of small objects, were taken for further consideration.

Testing the strength of individual muscles or muscle groups was performed using manual muscle test (MMT)<sup>8</sup>.

From all assessed muscles of both hands, seven of them (important for movements during work or daily activities) were selected and analyzed: *extensor digitorum communis* (EDC), *flexor digitorum superficialis* (FDS), *flexor digitorum profundus* (FDP), *abductores pollicis (longus et brevis)* (ABDP), *adductores pollicis* (ADDP), *opponens pollicis et digiti V* (OPP), *flexor pollicis longus* (FPL).

The MMT score was also used as an indicator of changes in the peripheral motor neuron, as an indicator of the degree of hypotrophy/atrophy of muscles and as an indicator of damage to the joints in the vicinity of the test muscle. Muscle strength in our work was marked from 1 to 5 (1 – normal muscle strength, 2 – good, 3 – moderate, 4 – low, 5 – absence). For more accurate statistical comparison, the original MMT marking (5 for normal muscle strength, up to 0 for total loss of muscle strength) was shown in reversed order in this work.

The original Hand Mobility in Scleroderma (HAMIS) test which follows 9 tested items, was applied for evaluation of the remaining functions of the fingers, hands and forearms of both hands (for assessing finger flexion, extension and abduction, thumb abduction, pincer grip – thumb and index finger opposition, wrist extension and flexion, pronation and supination)<sup>9</sup>. In addition to analysis of these values, we separately monitored conditions of the fingers regardless of the wrist, then the fingers with the wrist, as well as the total value of HAMIS test, which included forearm mobility in estimation of both hands. Each item was graded on 0–3 scale where 0 corresponds to normal function and 3 denotes that the individual cannot perform the task. For each hand, total HAMIS score was in range from 0 (normal findings) up to 27 (complete inability to perform tasks representing a high degree of dysfunction).

Assessment of disease severity and functional condition of the hands was also carried out by means of the task-induced fatigue (TIFS) questionnaire Scleroderma Assessment Questionnaire (SAQ) which was developed, validated and published by the doctors from the Institute of Rheumatology in Belgrade as an original tool for assessing the severity of illness and functional capability of patients with SSc as a whole, but only questions directly related to the function disturbance of the hands and fingers were covered in our paper<sup>10</sup>. Those questions were related to ability to hold pencil, to unbutton the shirt, to unscrew the tap, to hold the knife and cut meat, to perform household activities, or to detect pain in fingers while holding objects or exposing to cold weather.

In addition, questionnaire for assessing the quality of life were taken into account in respect of the answers to several questions directly showing impact on the disturbed working capacity (questions detecting if the subjects had to reduce time spent on work and other activities, or to leave out some of the work, or to achieve less than expected, or to invest an extra effort in work or other activities).

Work ability was assessed with the Work Ability Index (WAI)<sup>11–13</sup>, which is a self-administered questionnaire comprising seven items considering the demands of work, the worker's health status and resources. The items are grouped according to: (a) estimation of current work ability compared with lifetime best; (b) work ability in relation to physical and

mental demands of the work; (c) number of diagnosed diseases; (d) estimation of work impairment due to diseases; (e) sick leave during the past year; (f) own prognosis of work ability after 2 years; (g) psychological resources. For each item, a single-item score is obtained. The final WAI score is calculated as the sum of all single-item scores. Higher scores on the WAI indicate better work ability. Based on this score the individual's work ability can be classified into four categories: 1 – excellent (44–49 points), 2 – good (37–43 points), 3 – moderate (28–36 points) and 4 – poor (less than 28 points).

#### Statistical analysis

After description of all answers and marks, statistical significance was tested through parametric Student *t*-test, and the nonparametric sum rank test (Mann-Whitney test) for two groups of data. Nonparametric analysis of variance were performed with Kruskal-Wallis test for more than two groups of data. Nonparametric methods were used more often due to quite small sample and the nature of data (scores obtained after point rating of reports, but testing of normal distribution using Kolmogorov-Smirnov test were final judge). In all methods the level of significance was set at 0.05.

#### Results

The study was conducted on patients with SSc treated at the Institute of Rheumatology in Belgrade. The treatment included 17 (85%) females and 3 (15%) males. Their average age was  $45.5 \pm 11.9$  years (min 29, max 69, Med 44 years) and did not significantly differ in relation to sex. Average disease duration was  $6.4 \pm 3.7$  years (min 1, max 17, Med 4.0): females  $6.6 \pm 4.0$  (min 1, max 17, Med 5.0) years and males  $5.0 \pm 3.2$  (min 1, max 16, Med 4.5) years, and the difference in the disease duration between sexes was not statistically significant ( $z = 0.191$ ;  $p > 0.05$ ). Ten of our patients were employed and also ten of them were unemployed.

The diagnosis of SSc was set according to the recommendations of the European League Against Rheumatism (EULAR), which were revised in 2013 by a joint committee of the American College of Rheumatology (ACR) and EULAR<sup>14</sup>.

In assessing their entire health, 19/20 (95%) of our subjects were dissatisfied. The lowest scores for their health gave 9/20 (45%) of the patients.

Moderate to severe pain in the hands and fingers reported up 19/20 (95%) of our subjects (depending on the questionnaire). The pain was provoked by the cold weather, contact with a cold object or emerged spontaneously.

The most common problems were: weakness in the hands and fingers in 11/20 (55%), stiffness in the hands in 14/20 (70%), lack of manual actions – unscrewing taps in 7/20 (35%), dropping objects in 7/20 (35%), inability to hold a pencil in 6/20 (30%) and buttoning buttons in 6/20 (30%) of our patients (Table 1).

Thickening of the skin, as measured by the mRSS was also found in 19/20 (95%) of our subjects on the fingers bilaterally and in 14/20 (70%) on both hands. Thickening of the skin, unattached to the deeper layers (score 2), as well as attached to the

deeper layers (score 3), was found on the fingers in 17/20 (85%), and on the hands in 7/20 (35%) of the patients.

Capillaroscopy demonstrated lesions in the capillaries on fingers of both hands in 15/17 (85%) of our patients („scleroderma type” of capillary lesions, classified as a type II and III according to Maricq). According to Cutolo classification, 3/17 (18%) of the subjects had early changes (early scleroderma sample), 8/17 (47%) active and 3/17 (18%) had late scleroderma.

Reduced hands muscles strength was tested by MMT in all subjects. In relation to the maximum possible maintenance of the strength (100%) the reduction ranged from 10% to 50% and was most frequently registered in the *flexor digitorum profundus* – FDP (R-45%, L-50%), the *flexor digitorum superficialis* – FDS (R-45%, L-50%) and the *flexor pollicis longus* – FPL (R-40%, L-45%) (R-right hand, L-left hand).

Reduced active range of motion in the joints of the fingers of both hands, was registered in 16/20 (80%) of patients.

Most frequently it was the case of the bilaterally reduced flexion of the proximal phalanx of the thumb (75%), flexion of the distal phalanx of the thumb bilaterally (70%), extension of the proximal phalanx of the thumb bilaterally (65%) and flexion of the proximal phalanx of the index finger (R-70%, L-65%). The largest reduction in range of motion was accounted for 89% of the maximum, and was registered for the flexion of the distal phalanx of the middle finger and flexion of the distal phalanx of the thumb bilaterally, as well as 88% of flexion of the proximal phalanx of the right index finger.

Function evaluation by means of the HAMIS test showed that 10/20 (50%) of our patients had damaged function of hands, fingers and forearm, ranging from mild (isolated flexion or extension of each finger failure at 6/20–30%) and severe (complete pronation and supination failure of the forearm, and the limited mobility of the majority of fine joints at 4/20–20%) (Table 2).

**Table 1**

**Frequency of subjective problems**

Subjective problems	T	P	%	Mean ± SD	Min	Max
Dissatisfaction with health status	20	19	95	3.50 ± 0.94	1	5
Pain in the hands	20	19	95	3.40 ± 1.31	1	5
Pain in the fingers	20	11	55	1.80 ± 0.84	1	4
Weakness in the hands	20	11	55	2.65 ± 1.08	1	4
Stiffness in the hands	20	14	70	2.30 ± 1.08	1	4
Difficulties in manual actions						
unscrewing taps	20	7	35	1.55 ± 0.94	1	4
dropping objects	20	7	35	1.50 ± 0.82	1	4
holding a pencil	20	6	30	1.40 ± 0.68	1	3
buttoning buttons	20	6	30	1.40 ± 0.68	1	3

**T – total number of patients; P – number of patients with positive answer; % – percentage of patients with positive answers; SD – standard deviation.**

**Table 2**

**Frequency of objective problems**

Objective problems	T	P	%	Mean ± SD	Min	Max
Thickening of the skin (mRSS) on						
fingers	20	19	95	2.54 ± 0.76	0	3
hands	20	14	70	1.36 ± 0.96	0	3
Capillary changes on hand fingers						
Cutolo method/score	17	14	82	2.65 ± 0.96	1	4
Maricq method/score	17	15	85	2.53 ± 0.80	1	4
Diminished muscle strength						
FDP	20	10	50	2.10 ± 1.33	1	5
FDS	20	10	50	2.05 ± 1.31	1	5
FPL	20	9	45	1.85 ± 1.18	1	5
Diminished range of motion						
flexor of thumb distal phalanx	20	14	70	70.85 ± 31.40	11	100
flexor of thumb proximal phalanx	20	15	75	65.65 ± 30.92	14	100
extensor of thumb proximal phalanx	20	13	65	61.05 ± 34.36	14	100
flexor of III finger proximal phalanx	20	14	70	80.00 ± 29.04	11	100
HAMIS test on						
fingers	20	10	50	1.95 ± 0.77	0	11
fingers + hand	20	10	50	3.70 ± 1.28	0	17
fingers + hand + lower arm	20	10	50	4.00 ± 3.74	0	19

**T – total number of patients; P – number of patients with positive answer; % – percentage of patients with positive answers; SD – standard deviation; mRSS – modified Rodnan skin score; FDP – *flexor digitorum profundus*; FDS – *flexor digitorum superficialis*; FPL – *flexor pollicis longus*; HAMIS – hand mobility in scleroderma.**

Problems at work and difficulties in performing daily activities due to illness, reported 15/20 (75%) of our patients. They tried to solve those problems: by reducing working hours – 8/20 (40%); by failure to meet standards – 8/20 (40%); by leaving out some of the work – 14/20 (70%); by giving an extra effort at work – 15/20 (75%).

The need to cut the time spent at occupational or daily activities had 8/20 (40%) of our subjects. This finding was significantly associated with an impaired function of finger muscles (FPL 2.50 vs. 1.42,  $z = 2.20$ ,  $p = 0.041$ ; ABDP 2.25 vs. 1.08,  $z = 3.74$ ,  $p = 0.002$ ; ADDP 2.25 vs. 1.17,  $z = 2.30$ ,  $p = 0.034$ ), hand muscles (EDC 2.38 vs. 1.16,  $z = 3.31$ ,  $p = 0.004$ ; FDS 3.00 vs. 1.42,  $z = 3.22$ ,  $p = 0.005$ ; FDP 3.00 vs. 1.50,  $z = 2.91$ ,  $p = 0.009$ ), the function of the fingers, hand and forearm checked by means of the HAMIS test (8.25 vs. 1.17,  $z = 3.34$ ,  $p = 0.004$ ), as well as more difficult manual actions (unscrewing the tap, 2.00 vs. 1.25,  $z = 1.85$ ,  $p = 0.050$ ; fastening buttons, 1.75 vs. 1.16,  $z = 2.02$ ,  $p = 0.048$ ).

Also, 8/20 (40%) of our subjects complained to reduction of achieved performance compared to that prior to the disease, which was in statistically significant connection with

pain in the hands (3.25 vs 2.25,  $z = 2.20$ ,  $p = 0.041$ ), weakness in the hands (2.13 vs. 1.42,  $z = 2.10$ ,  $p = 0.050$ ), stiffness of the hands (3.00 vs. 1.83,  $z = 2.74$ ,  $p = 0.013$ ), difficult manual activities (fastening buttons 1.75 vs 1.17,  $z = 2.03$ ,  $p = 0.050$ ), and thickened skin on the hands (3.50 vs 3.10,  $z = 2.05$ ,  $p = 0.050$ ), as well as with the function of the fingers, hands and forearms, which was measured by means of the HAMIS test (3.88 vs. 0.67,  $z = 2.51$ ,  $p = 0.022$ ; 7.00 vs. 1.50,  $z = 2.61$ ,  $p = 0.018$ ; 7.50 vs. 1.67,  $z = 2.51$ ,  $p = 0.022$ ).

Further, 14/20 (70%) of the patients had to exclude some kind of activities, which was in statistically significant relation with a physical pain in the hands (3.00 vs. 1.83,  $z = 2.47$ ,  $p = 0.024$ ) and with reduced range of motion in the joints of the fingers (94.33 vs. 69.57,  $z = 2.39$ ,  $p = 0.028$ ).

Also, 15/20 (75%) of our patients had to make additional efforts in implementing working tasks, with statistically significant connection with the subjective assessment of the current state of health (3.80 vs. 2.60,  $z = 2.89$ ,  $p = 0.010$ ), physical pain in fingers and hands, (3.87 vs. 2.00,  $z = 3.45$ ,  $p = 0.003$ ), as well as with the stiffness of the fingers (2.60 vs. 1.40,  $z = 2.40$ ,  $p = 0.027$ ) (Table 3).

Table 3

Statistically significant correlation of hand changes and working ability

Parameters	Group I	Group II	z	p
	mean ± SD	mean ± SD		
<b>Reducing time spent on the job</b>				
Diminished muscle strength on:				
fingers				
FPL	1.42 ± 0.67	2.50 ± 1.50	2.20	0.041 <sup>#</sup>
ABD	1.08 ± 0.28	2.25 ± 1.03	3.74	0.002 <sup>##</sup>
ADD	1.17 ± 0.38	2.25 ± 1.58	2.30	0.34 <sup>#</sup>
hands				
EDC	1.16 ± 0.38	2.38 ± 1.68	3.31	0.004 <sup>##</sup>
FDS	1.42 ± 0.67	3.00 ± 1.50	3.22	0.005 <sup>##</sup>
FDP	1.50 ± 0.80	3.00 ± 1.50	2.91	0.009 <sup>##</sup>
<b>Difficulties in manual actions</b>				
Unscrewing taps	1.25 ± 0.45	2.00 ± 1.31	1.85	0.050 <sup>#</sup>
Buttoning buttons	1.16 ± 0.39	0.75 ± 0.89	2.02	0.048 <sup>#</sup>
HAMIS test (fingers + hand + lower arm)	1.17 ± 1.08	8.25 ± 7.08	3.34	0.004 <sup>##</sup>
<b>Failure to meet standards</b>				
Pain in the hands	2.25 ± 0.15	3.25 ± 0.89	2.20	0.041 <sup>#</sup>
Weakness in the hands	1.42 ± 0.51	2.13 ± 0.99	2.10	0.050 <sup>#</sup>
Stiffness in the hands	1.83 ± 0.83	3.00 ± 1.06	2.74	0.013 <sup>#</sup>
Difficulties in manual actions				
buttoning buttons	1.17 ± 0.39	1.75 ± 0.88	2.03	0.050 <sup>#</sup>
Thickening of the skin (mRSS) on:				
hands	3.10 ± 1.02	3.50 ± 0.53	2.05	0.050 <sup>#</sup>
HAMIS test on:				
fingers	0.67 ± 0.63	3.88 ± 3.22	2.51	0.022 <sup>#</sup>
fingers + hand	1.50 ± 1.57	7.00 ± 6.67	2.61	0.018 <sup>#</sup>
fingers + hand + lower arm	1.67 ± 1.35	7.50 ± 7.19	2.51	0.022 <sup>#</sup>
<b>Leaving out some of the work</b>				
Pain in the hands	1.83 ± 0.98	3.00 ± 0.96	2.47	0.024 <sup>#</sup>
Diminished range of movement:				
flexor of III finger mid.phal.	94.33 ± 10.13	69.57 ± 27.04	2.39	0.028 <sup>#</sup>
<b>Investing an extra effort</b>				
Dissatisfaction with health status	2.60 ± 0.89	3.80 ± 0.77	2.89	0.010 <sup>##</sup>
Pain in the hands and fingers	2.00 ± 1.00	3.87 ± 1.06	3.45	0.003 <sup>##</sup>
Stiffness in the hands and fingers	1.40 ± 0.54	2.60 ± 1.05	2.40	0.027 <sup>#</sup>

Group I – patients with no problems at work; Group II – patients with problems at work; SD – standard deviation FPL – flexor pollicis longus; ABD – abductor pollicis; ADD – adductor pollicis; EDC – extensor digitorum communis; FDS – flexor digitorum superficialis; FDP – flexor digitorum profundus; HAMIS – hand mobility u scleroderma; mRSS – modified Rodnan skin score. p – statistical significance; z – value of the Mann-Whitney test; # – statistically significant; ## – statistically highly significant.

Table 4

## Statistically significant correlation of hand changes and the WAI score

Hand changes	WAI groups			$\chi^2_{kw}$	<i>p</i>
	excellent (mean ± SD)	good (mean ± SD)	moderate (mean ± SD)		
Stiffness in the hands	1.88 ± 0.92	2.20 ± 0.83	3.00 ± 1.26	2.77	0.050 <sup>#</sup>
Weakness in the hands	1.44 ± 0.52	1.40 ± 0.54	2.33 ± 1.03	3.34	0.050 <sup>#</sup>
Difficulties in manual actions (unscrewing taps)	1.22 ± 0.44	1.22 ± 0.44	2.33 ± 1.36	3.83	0.042 <sup>#</sup>
Thickening of the skin (mRSS) on: fingers	3.44 ± 0.72	2.60 ± 1.14	3.67 ± 0.51	3.74	0.050 <sup>#</sup>
Diminished range of motion in: thumb ABD	1.11 ± 0.33	1.40 ± 0.54	2.33 ± 1.21	4.98	0.020 <sup>#</sup>
thumb ADD	1.22 ± 0.44	1.20 ± 0.44	2.50 ± 1.75	3.31	0.050 <sup>#</sup>

mRSS – modified Rodnan skin score; ABD – *abductor pollicis*; ADD – *adductor pollicis*; SD – standard deviation. *p* – statistical significance;  $\chi^2_{kw}$  – value of the Kruskal-Wallis test; <sup>#</sup> – statistically significant; <sup>##</sup> – statistically highly significant.

WAI questionnaire demonstrated that the working capacity was reduced in 11/20 (55%) of our subjects. In the group of the most serious cases (the “poor” category), there were no subjects; there were 6/20 (30%) in “moderate” category, while 5/20 (25%) of subjects were in the group with minimally reduced working capacity (“good” category).

These findings were in statistically significant correlation with our data regarding stiffness in the hands ( $\chi^2 = 2,77$ ; *p* = 0.050), arms weakness ( $\chi^2 = 3,34$ ; *p* = 0.050), problems with manual activities (opening the tap) ( $\chi^2 = 3,83$ ; *p* = 0.042), followed by a thickening of the skin of the fingers on both hands ( $\chi^2 = 3,74$ ; *p* = 0.050) and reduced mobility of thumb bilaterally (ABD -  $\chi^2 = 4,98$ ; *p* = 0.020 and ADD -  $\chi^2 = 3,31$ ; *p* = 0.050) (Table 4).

The degenerative changes in the small joints of the wrist and fingers (osteoarthritis/ arthrosis on DIP and PIP) were visible at X-ray images of the hands in 4/20 (20%) of our subjects. None of them had ulceration of the fingertips on hands, acroosteolysis, calcinosis or tendon friction.

Comparing other changes on hands with problems at work and with the WAI score, no statistically significant connection was found.

Comparing the capillaries, skin thickness, muscle strength and range of motion of the right and left hands, no statistically significant difference was found.

## Discussion

Impaired function of the hand in SSc is of multifactorial origin and is the result of the pain, increased skin thickness, skin sclerosis, subcutaneous calcinosis, thickening of the connective tissue in the deeper layers of the dermis and muscle fascia, lesions in the bones and joints, the microvascular lesions (paroxysmal vasospasm or permanent ischemia with subsequent digital ulcerations), contractures, tendon shortening, rubs friction on tendons and reduced strength<sup>1-4</sup>. Significant functional disability occurs in excess of 50% of patients in first 18 months from the onset of the disease.

All mentioned elements result in reduced use of damaged hands, affecting the working ability and the quality of life.

The physical pain associated with SSc is present in all patients at different stages of the disease. The first painful

sensations are due to vasoconstriction of the small arteries and arterioles of fingers and manifested as Raynaud's phenomenon (RP). Pain can occur occasionally in the attack, it is mild and lasts up to half an hour, rarely longer. It is followed by pain due to muscle, bone and joint lesions, as well as the lesions of the sensitive nerves, most often in the area of the wrist, at rest or in motion, lasting several hours and mostly medium to high intensity. Later, with the evolution of the disease, the causes of pain are also the lesions of the internal organs<sup>15,16</sup>.

Stisi et al.<sup>17</sup> have analyzed the incidence of pain induced by damage of the musculoskeletal system in 242 patients with SSc. The pain was present in 83% of patients.

According to Clements<sup>18</sup>, pain due to changes in the locomotor system is present in 40–80% of patients with SSc, especially in the initial phase of the SSc.

In our study group, pain was present in 19/20 (95%) of the patients, with 13/20 (65%) of subjects complaining about moderate to severe pain in the hands and fingers.

mRSS is the most suitable method for assessment of the prevalence of skin lesions, which is the basis for distinguishing between limited and diffuse forms of the disease, also to assess the success of the applied therapy, and as an indicator of the disease activity and outcomes<sup>6</sup>. Changes in the skin significantly correlate with mRSS<sup>19</sup>. In the early stages of SSc, extensive skin changes correlate with severe changes on the internal organs observed later in the disease, poor prognosis and increased disability.

Sawy et al.<sup>20</sup> evaluated the functions of hands in 15 patients with SSc and found increased thickness of the skin and *flexor retinaculum*, thickening of inter-metacarpal area II as well as the reduced area of innervation of median nerve, which was significantly associated with reduced grip strength as well as with reduced range of motion of the fingers. They concluded that impaired hand function was mainly linked with reduced mobility and reduced hand strength.

For the purposes of this study, only mRSS values relating to the quality of the skin of fingers and hands were taken. Changes on the hands were found in 14/20 (70%) of our patients, and in more severe forms (thickened skin unattached or attached to the deeper layers) in 7/20 (35%) of them. This finding failed to show a statistically significant correlation with

working capacity. Changes on the fingers were detected in a higher percentage – 19/20 (95%), and were significantly associated with the impairment of working capacity.

Capillaroscopy provides quite accurate assessment of blood vessels condition. On the basis of this finding, it is possible to differentiate primary from secondary Raynaud's phenomenon (RP), to detect a disease before other clinical manifestations, predict the development of the disease, to assess the disease activity and early detect involvement of the internal organs in SSc<sup>7,21</sup>.

“Scleroderma type” capillary lesions, classified as a type II and III in patients with RP can be seen months and even years before the manifestation of other signs of disease, and represents valuable sign for the early detection and prediction of the disease development<sup>22</sup>. Early diagnosis of SSc so far represents the only realistic chance for successful treatment.

Analysing 3,035 capillaroscopy findings in patients with primary RP, scleroderma type lesions were registered even 6 months prior to the manifestation of other signs related to the disease<sup>23</sup>.

The secondary RP is in most cases an introduction to one of systemic autoimmune diseases or the microcirculation disorder. In about 90% of all patients with SSc, RP is the first symptom of the disease. The characteristic of the secondary RP is pathological capillaroscopy pattern, also referred to as scleroderma pattern<sup>21,24</sup>.

Koenig et al.<sup>25</sup> announced that a high percentage of patients with RP and with capillaroscopy finding typical for SSc, with no other signs except the bloated fingers and/or arthritis, will, over time, develop diffuse form of SSc, and suggested the name “early systemic sclerosis” (eSSc).

In our study the pathological lesions of the capillaries were demonstrated in high percentage (15/17, 85%). In 6/17 (35%) of our subjects, it was type II lesion according to Maricq, which was considered to be an indicator of a mild form of SSc and better prognosis. Type III was registered in 8/17 (45%) of the patients, and type IV in a single patient, pointing towards more severe disease form with a poor prognosis. Changes in the capillaries in our patients did not show a statistically significant association with the reduction of working capacity.

Ulcerations of fingertips lead to significant functional impairment, disability and decreased life expectancy. They can be complicated with infections, gangrene and amputation. They are present in 44–60% of patients with SSc, and are associated with early RP, dcSSc, with deterioration in findings on capillaroscopy and decreased survival<sup>26–29</sup>.

Khimidas et al.<sup>29</sup> analyzed the correlation of digital ulcerations with other manifestations of SSc in 938 patients and found that digital ulcers were associated with increased mRSS of hands and fingers, duration of the disease, interstitial lung disease in both dSSc and lSSc.

In our study group no digital ulcer was registered. This can be explained by the timely diagnosis of SSc in our patients, periodic monitoring in a hospital setting in order to check the current status, perform additional diagnostics or adjust medical treatment, as well as less pronounced changes

in the blood vessels, which are normally considered to be a major pathophysiological mechanism for onset of ulcers.

Impaired function of the hands and fingers is manifested mainly by reduced ability of MCP joint flexion, limited extension of PIP joints, and flexion of DIP joints, decreased abduction (and soon the opposition and flexion) of the thumb, and loss of motion in the wrist (RC) in all directions. All that is accompanied by muscle hypotrophy, reduced strength, as well as the pain that can occur even after a few repetitive movements, common in everyday life and work. These changes are rarely isolated, and often present simultaneously, and affect the working activities significantly.

Decrease in muscle strength symmetrically on the extremities was confirmed by MMT in 10% of the patients<sup>18</sup>. Sometimes it is difficult to separate the primary myopathic weakness from the movement restrictions caused by thickening of the skin, changes in the joint near the affected muscle or surrounding tissue fibrosis. Patients with skeletal myopathy do not have worse prognosis compared to those without myopathy.

Reduced hand muscle strength was confirmed in all subjects in our study. In relation to the maximum possible maintenance of the strength (100%) the reduction ranged from 10% to 50%.

Bone and joint damages (arthralgia, arthritis, arthrosis, contractures, tendon friction rubs, tenosynovitis) are very common, they are among the first manifestations of the disease, causing considerable disability and often indicating the involvement of the internal organs<sup>2,18,30–32</sup>.

Lesions in the MCP, PIP and DIP joints seen on X-ray images, are in the form of joint space narrowing, erosions, intra-articular calcification, juxta articular osteoporosis/osteopenia and subluxation<sup>2,33,34</sup>.

Avouac et al.<sup>34</sup> analyzed X-ray images of hands in 120 patients with SSc and found the presence of erosion in 21%, narrowing of the joint space in 28%, arthritis in 18%, demineralization in 23%, acroosteolysis in 22%, flexion contracture in 27%, and calcinosis in 23% of the cases.

In our subjects, X-ray images revealed degenerative lesions (osteoarthritis-arthrosis) in 4/20 (20%) of the patients. There were no signs of calcinosis, acroosteolysis, demineralization or arthritis.

Contractures of small finger joints are the most common joint lesions in SSc [31% according to the European Scleroderma Trials and Research (EUSTAR) Group] and are the major cause of functional disability<sup>35</sup>.

There is no consensus on what degree of motion reduction in joints may be called the joint contracture, and therefore the prevalence of contractures found by physical examinations in the various studies varies from 26% to 56%. In spite of the slow and gradual onset, contractures may occur even in the early stages of the disease. They are result from thickening of the skin, only in the shallow layers at first, followed by connective tissue thickening in the deeper layers of the dermis and muscle fascias, peritendinous sclerosis causing tendons shortening, as well as the destruction of the joints which leads to ankylosis. Contractures of small joints of hands (MCP, DIP and PIP) are mostly flexorial (PIP),

sometimes in combination with the extension contracture (MCP) of joint (claw fist). DIP joints are less frequently affected by flexion contracture and thumbs are mainly affected by adduction contractures. They are more common in dcSSc on the dominant hand. Involvement of more than 4 joints on the same hand is a sign of poor prognosis. It is associated with interstitial lung disease, changes in the esophagus and cardiac involvement<sup>2</sup>.

In our work, decreased range of motion in the joints of the fingers of both hands was registered in 16/20 (80%) of the patients. Most often it was in the small joints of the thumb, index finger and middle finger of both hands. The largest reduction in range of motion was 89% of the maximum.

For estimation of work ability, the primary version of WAI is used. It was established and used at the end of twentieth century by researchers from Finland<sup>36</sup>. The WAI is used today in Finland as a standardized questionnaire of the Finish Institute of Occupational Health (FIOH). In more than 25 countries, the WAI is accepted as a questionnaire for self-estimation of working ability, taking into consideration physical and psychological capacities of an individual<sup>37-40</sup>.

Sandqvist et al.<sup>41</sup> examined the WAI in 48 patients with SSc. Thirteen patients had good or excellent WAI, 15 had moderate, and 20 had poor WAI. Patients with good WAI had less severe symptoms (pain, fatigue, impaired hand functions), better ability to adapt to tasks and needs of the workplace in relation to the patients with worse scores on the WAI questionnaire. Fatigue and decreased functionality of the hand had the greatest impact on the WAI questionnaire.

The WAI questionnaire demonstrated that the working capacity was reduced in 11/20 (55%) of our patients. Those were the patients classified in the category "moderate" and "good". There were no subjects in the group of the most serious cases, the "poor" category, i.e. where respondents expressed extremely diminished ability to work.

Lesions on the hands, which are present even in the early years of the development of SSc, are responsible for strik-

ing disability. The milder disorders present in the beginning of the disease, include occasional and brief spasms of blood vessels, mild thickening of the skin, arthralgia and occasional mild to moderate pain. At that stage of the disease, there is a limited ability to work, especially for manual jobs.

The progression of the diseases results in serious disturbances which include more frequent vascular spasms with longer duration, ulceration of the fingertips, expressed thickening of the skin, acroosteolysis, ankylosis and deformity of the joints, calcinosis, contractures, and frequent medium and severe pain. Associated with damage to internal organs, these disorders lead to complete loss of ability to work.

Our study included relatively small number of patients, so the results cannot be generalized to the whole population of patients with systemic sclerosis. However, our findings are providing solid evidence of significant connection between reduced hands function and performing working and daily activities. Further investigation on a larger population is needed to confirm our results.

### Conclusion

The results obtained by examining hands impairment in patients with systemic sclerosis, showed that there were: thickened skin on the hands and fingers, reduced muscle strength and decreased joint mobility in the fingers and changes in the finger capillaries.

The results of the assessment of working ability in patients with systemic sclerosis, showed that the subjects solved their problems at work by reducing time spent on the job, by leaving out some of the work, by failure to meet standards and by investing an extra effort in work or other activities. Working capacity was reduced (through the WAI evaluation) and it belonged to categories "good" or "moderate".

There was a statistically significant correlation between altered hand functions and diminished working capacity.

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