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**Case Study of the
Physiotherapeutic Treatment
of a Patient with Scleroderma**

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April 2014

Abstract

Title

EN: Case Study of the Physiotherapeutic Treatment of a Patient with Scleroderma

CZ: Kazuistika fyzioterapeutické péče o pacienta s diagnózou sklerodermie

Summary

The thesis comprises theoretical knowledge about scleroderma and a practical study of a patient with this pathology.

Keywords

Scleroderma, Systemic Sclerosis, Raynaud's Phenomenon, Microstomia, Sclerodactyly, Sjogren's Syndrome, Telangiectasia.

Declaration

I declare that I worked on my thesis separately, under the guidance of a supervisor Doc. PaedDr. Dagmar Pavlů, CSc.

I used only cited professional and literary sources and, no information has been misused, all were authorized and adequately documented.

Julien Barange

Prague, the 16th of April 2014

Acknowledgement

I would like to thank my family, friends and classmates for all their supports and motivation. It was a wonderful experience to study in Prague, and I am definitely happy to have made the choice to come and study here.

Great compliments to my Supervisor Doc. PaedDr. Dagmar Pavlů, CSc. for her wise guidance and advices.

Many thanks to Mgr. Maja Špiritovič for all the help, energy and mentorship she provided during my clinical placement.

Thank you all.

Julien Barange

Prague, the 16th of April 2014

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1. Introduction

The Scleroderma is in an autoimmune, rheumatic and chronic disease that affects the body by hardening connectives tissue. It mainly affects the skin and, in certain forms of scleroderma, can spread to the internal organs.

In the first part of this thesis we will start by giving complete information about Scleroderma: the causes, diagnosis, clinical picture, symptoms and, the treatment of this disease with an emphasis on the physiotherapeutic methods. All this theoretical part is the result of my investigations on the subject and relates the most recent studies about Scleroderma.

In the second part, I will describe in detail the course of my therapy during 8 sessions with a sclerodermic patient. This part includes anamnesis, initial kinesiologic examination, therapy sessions, final kinesiologic examination and, the evaluation of the therapy.

2. General Part

Systemic sclerosis (SSc) or scleroderma is a connective tissue disease characterized by thickening of the skin, fibrosis, and vascular and internal organ involvement.

Derived from the greek words “*sklerosis*”, meaning hardness, and “*derma*”, meaning skin, scleroderma literally means “hard skin”.

2.1 Prevalence and Incidence

Scleroderma is considered to be a rare disease, with an estimated prevalence of 300 cases per million Females are four times more affected than men.

It rarely occurs in children, and the common onset of the disease is between 45 to 60 years old (Mayes, 2003, pp. 2246-2255).

2.2 Aetiology and Pathology

The cause of scleroderma is still not clearly known. A recent study (Zhou, 2009, pp. 3807-3814) suggest that the origine is from a complex polygenetic association. However, multicases in family remains uncommon, even if the occurence is slightly higher than for the normal population.

Some environmental circumstances can act like triggers, including silica (Mccormic, 2010, pp. 763-769), solvents (Kettaneh, 2007). These precipitating events causes cells to produce excess collagen as if some injury has ocured. Once this production has started, the cells does not turn off.

2.3 Diagnosis

Diagnosis of scleroderma may be difficult especially in early stage because the symptoms are similar to other diseases.

Furthermore, there is any single blood test that is enough specific to diagnose scleroderma. However, some tests can give indication to diagnose the type of scleroderma: anti-Scl-70 is frequently associated with diffuse scleroderma and the Anti-centromere antibody is more associated to limited scleroderma (Clements, 1996).

The diagnosis of scleroderma is usually made by physicians who have the experience of this disease and are able to recognize the various visible symptoms (according to clinical picture). They determine the type of scleroderma and its severity. Then the blood test can confirm the diagnostic.

2.4 Clinical Picture

The two main form of scleroderma are localized scleroderma and systemic scleroderma:

- In localized scleroderma, the skin changes are limited to specifics areas and the internal organs are not involved. There are two subsets of localized scleroderma: Morphea and Linear scleroderma.

- In systemic scleroderma, the internal organs are involved and the skin involvement is less localized. There are two subsets of SSc: diffuse and limited cutaneous scleroderma.

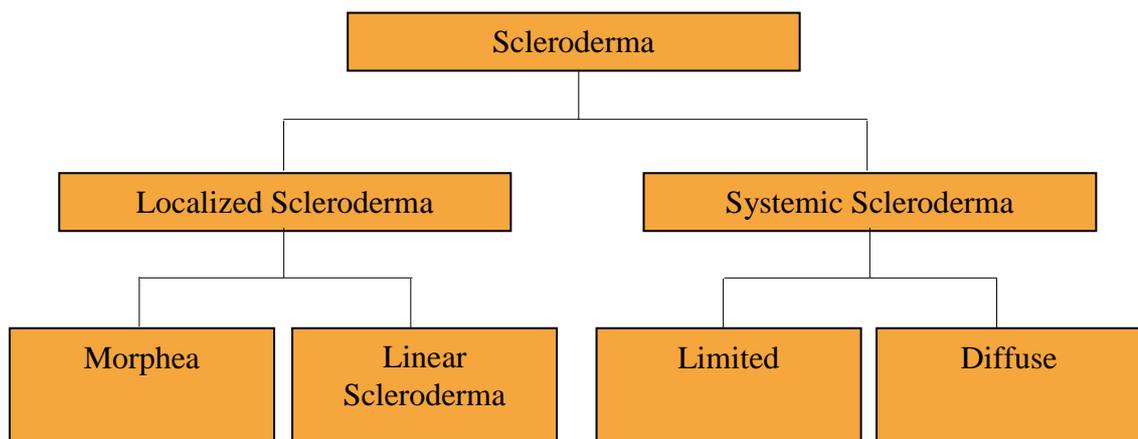


Figure 1- Different subsets of Scleroderma
(adapted from National Institutes of Health, 2010)

2.4.1 Localized scleroderma

The changes which occur in localized scleroderma are found in few places on the skin or muscles and rarely spread in other areas. This type of scleroderma doesn't involve the internal organs and usually doesn't evolve to SSc.

Morphea is a form characterized with patches of thickened skin with different shapes or colors. These patches may change in size and shapes, and also disappear spontaneously. The onset of morphea is common between the ages of 20 or 50, but is often seen in young children.

Linear Scleroderma is a form of localized scleroderma which frequently starts with a line of hardened skin on an arm leg or forehead. It is referred to as “en coup de sabre” because of its resemblance to a saber. Linear scleroderma usually appears in childhood and can affect the growth or the ROM of the affected limbs.

2.4.1 Systemic Scleroderma

The changes in systemic scleroderma may affect many connective tissue in many parts of the body. It can involve the skin, esophagus, gastro-intestinal tract, lungs kidneys, heart and other internal organs. It also affects the blood vessels, muscles and joints.

One half of the SSc patients develops the diffuse scleroderma. This form is the most aggressive type of scleroderma. It may involve most of the internal organs previously cited and also a wide area of skin thickening. The evolution of the disease is fast with an early involvement of the internal organ which leads to a worse prognosis compared to other forms of scleroderma.

The other half of patients develops the limited scleroderma, which is more benign compare to the diffuse form. The skin thickening is less widespread and typically confined to the hands, fingers, face and develops slowly over years. The internal organ involvement is also less generalized and may only affect the lungs, esophagus. This disease used to be called the CREST syndrome which means:

- Calcinosis,
- Raynaud’s phenomenon,
- Esophageal dysfunction,
- Sclerodactyly,
- Telangiectasias.

2.5 Symptoms

2.5.1 Raynaud's Phenomenon

Raynaud's phenomenon (RP) is the most common early symptom of SSc. However the RP is not specific to SSc, it can be either called "Primary Raynaud's" when not associated to other disease or "Secondary Raynaud's" when linked to a disease like SSc. This phenomenon is present with 90% of SSc patients and affect more obviously the fingers and toes but, can also involve the ears and nose.

It is the result of an impaired blood circulation in the vessels of the extremities which become white and dead looking. They may turn blue as the tissues are without oxygen and then red or flushed when the blood flow returns. This is often accompanied with by a burning feeling, pain and numbness.

The most common trigger of RP is the exposure to the cold but can also be a response to an emotional stress.



Figure 2 - Raynaud's phenomenon (dxline, 2010)

2.5.2 Swelling of the Hands

Swelling is another typical early symptom of scleroderma, especially in the morning because of the inactivity of the muscles overnight. The skin of the hand may look full and fingers sausage-like, making it difficult to close the hand into a fist.

2.5.3 Pain and Stiffness of the Joints

Symptoms of pain stiffness, swelling, warmth or tenderness may accompany the joint inflammation which occurs frequently in scleroderma. Muscle pain and weakness are other important symptoms.

2.5.4 Skin Disorders

Skin thickening

Hardening and thickening of the skin, resulting from excess collagen production, give the name of scleroderma (“hard skin”).

Dry skin

Excessive dryness of the skin may lead to ulcerations.

Skin ulcerations

Ulcerations affect commonly the fingertips and, can also occur on the knuckles, elbows, toes or other sites where the skin is hard, dry and with poor blood circulation. These ulcerations may be very slow to heal and cause increased inflammation in the surrounding areas.

Calcinosis

It is characterized by deposit of calcium below the surface of the skin in the form of hard white nodules, which may be painful. These deposits results from tissue hypoxia and they may break into the skin causing infection.

Telangiectasia

It consists of the dilatation of small blood vessels near the surface of the skin, which becomes visible as red spots, usually on fingers, palms, face and lips. These spots are usually not harmful.

2.5.5 Sclerodactyly and limited ROM in Joints

Sclerodactyly literally means “hard skin of the digits”, which affects the fingers or toes. It is characterized by shiny, tight skin of the fingers. Affected digits may be difficult to move and stay in a flexed position in the joint.

This tightening and hardening of the skin may also limit the ROM in wrists or elbows.

2.5.6 Digestive and Gastrointestinal Tract Disorders

Esophageal dysfunction

In SSc, the lower sphincter of esophagus, which opens for allowing food going to the stomach and close to prevent food to go up, doesn't close properly. It results in burning sensation in esophagus coming from the going back up of the food and the acid of the stomach. The acid may also injure the esophagus causing scars and narrowing of the tube.

Swallowing difficulties

The slow movement of the food and the narrowing of the esophagus causes swallowing difficulties. The dryness of the mouth (Sjogren's syndrome) increases the swallowing difficulties.

Diarrhea

In SSc, there can be damages in the muscles of the small intestine, the movement and absorption of food is slower and it can cause an abnormal development of bacteria which causes diarrhea.

2.5.7 Sjogren's Syndrome

It is characterized by decreased secretion of the tear and salivary glands, which provides lubrication to the eyes and mouth. Dryness of the mouth leads to difficulties for swallowing the food, SSc patients require often water when they are eating. Dry eyes may be lubricated by artificial tears drop.

2.5.8 Oral, Facial and Dental Problems

Persons with scleroderma have usually a tightening of the skin over the face. The opening of the mouth may be decreased in size (microstomia), making lips, mouth movement and oral hygiene difficult. These oral changes may also lead to dental problems



Figure 3 - Microstomia in Scleroderma (Dziedzic, 2010, p.309)

2.5.9 Internal Organs Involvement

Lungs

Involvement of the lungs is caused by thickens lung tissues resulting in sclerosis and making the transport of oxygen more difficult. Also, the damage of blood vessels may increase the resistance of blood flow and causes pulmonary arterial hypertension (PAH).

Kidney

The involvement of kidneys in SSc may be mild or very serious. Early signs of kidney involvement may include mild hypertension, protein in the urine and blood test abnormalities. The worst complication can be a “renal crisis” characterized by an abrupt rise of the blood pressure. Other symptoms can be headache, visual disturbances, shortness of breath, chest pain or mental confusion.

Heart

If the heart muscles becomes thickened and fibrotic, the force of heart contraction is decreased which may result ultimately to heart failure.

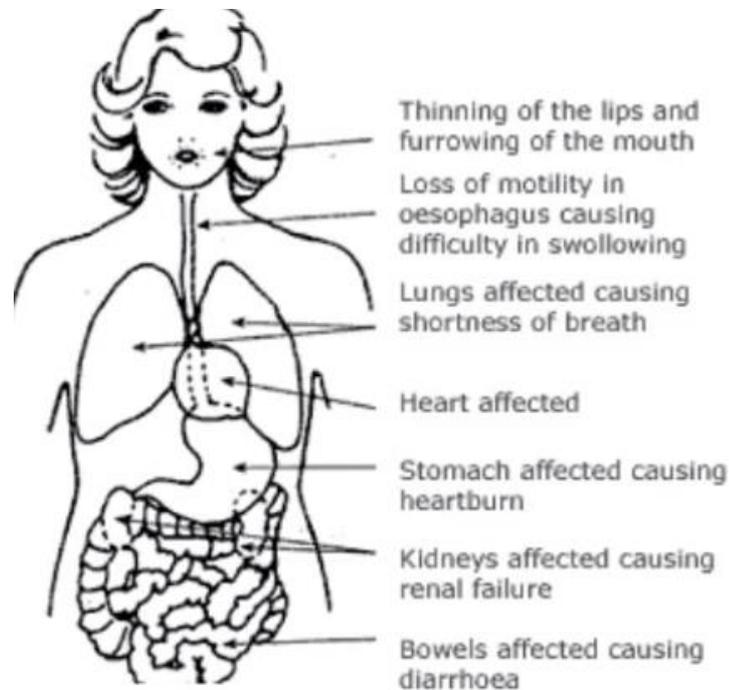


Figure 4 - Internal Organs Involvement in SSc (Doctor Decides, 2013)

2.6 Prognosis

At this time there is no cure for scleroderma, but there are many means available to control its symptoms and, an early diagnosis gives the opportunity to manage the disease before damages and fibrosis of internal organs lead to poor outcomes.

The disease course varies with each individual. The life expectancy in patients with SSc depends on the extents of the complications involving the internal organs. Therefore, mortality figures vary enormously. The ten-year rate survival is around to 60-70 % for SSc patients (Altman, 1991).

2.7 Rehabilitation

2.7.1 Physiotherapeutic Treatment

Before starting the rehabilitation, the physiotherapist must realize a full assessment of the patient in order to build an adapted therapeutic program. In this program, he should include ROM exercises, strengthening of the weak muscle, soft tissues techniques and others techniques that are adapted to this patient. He has also to educate the patient for self-therapy exercises and advices for ADL to have the maximum results in the therapy. In the following chapter we will study the actual most effectives method used in the rehabilitation of sclerodermic patients.

Hands therapy

The hands are the more common part of the body involve in scleroderma with symptoms such swelling, Raynaud's phenomenon, sclerodactyly causing pain and reduction in the joints ROM. The normal function of the hand becomes altered and the patient has problems for performing his ADL. That's why it is a priority for the physiotherapist to work on improving the hand function.

The following table summarizes the results of studies focusing on the hand rehabilitation for SSc patients.

Type of rehabilitation	Authors	Description of intervention	Results
Hand ROM exercises	Mugii et al. (2006)	Stretching exercises for the joints of the hand	Passive motion improved at 1 month after the intervention and improved further or was maintained at 1 year
Manual lymphatic drainage	Maddali Bongi et al. (2011)	Manual lymphatic drainage once a week for 5 weeks	Reduction of edematous in hand (less volume) and reduction of pain and improvement in hand function.

(continued)

Type of rehabilitation	Authors	Description of intervention	Results
Paraffin wax treatment and hand ROM exercises	Askew et al. (1983)	Combination of paraffin wax, friction massage and ROM in 1 session	Joint motion, skin compliance and hand function improved
	Sandqvist et al. (2004)	Paraffin wax on one hand and hand exercises once a day for one hand, the other served as control	Significant improved of ROM, skin thickness and elasticity in the hand receiving the intervention
Splints	Seeger and Furst (1987)	Dorsal splint with dynamic PIP extension 8h/day for 2 month	No significant improvement
Overall rehabilitation consisting of specific and global techniques	Maddali Bongi et al. (2009)	Specific tech.: hand and face exercises, connective tissue massage. Lymphatic drainage. Global tech.: respiratory exercises and land water based adapted program.	Functional ability hand mobility and function, mouth opening and face mobility were all significantly increased at the end of the 9 week program. At the 9 week follow-up only improvement in mouth opening and hand mobility were maintained.

Table 1 – Summary of Studies on Rehabilitation Techniques in Systemic Sclerosis focusing on the Hands (modified from Bijlsma, 2012, pp. 279-280)

In the previous studies, the use of the following techniques (alone or combined), improved significantly the condition of the patient's hands:

- ROM exercises,
- Hand massages,
- Lymphatic drainage,
- Paraffin wax,
- Joint mobilization.

On the contrary, the wear of splints on dorsal side of the hand didn't show significant improvement.

The paraffin wax treatment has shown good results but, it is contraindicated in case of skin ulceration. It is also important to add that the hand's massage should be accompanied with moisturizing cream in order to give hydration to the skin and this way, preventing the dryness and skin ulceration.

Facial exercises

Patient with scleroderma may experience a general tightening of skin over the face. The opening of the mouth may be decreased in size (microstomia), making lip and mouth movements as well as oral hygiene difficult.

The best approach consists of facial stretching exercises with exaggerated facial movements, manual opening of the mouth with fingers and oral augmentation exercises. A recent study examined the effectiveness of a connective tissue massage to the face Kabat's technique (PNF for the face) and face exercises demonstrate significant improvements in mouth opening and decreases in facial skin scores were observed in the in the intervention group compared to a group who received a home exercise program for the mouth and face (Johnson, 2013, pp. 106-106).

General physical exercise

In the study by Antonioni et al (2009), the intervention consisted of breathing exercises, aerobic training on a treadmill and, finger stretching. The participants had 30 minutes sessions during 2 weeks. The results show significant increase in hand function, decrease in heart rate and, increase in physical functioning and health. There was no significant change in walking distance nor in lung function.

Another study from Oliveira et al. (2009) consisted of an 8-week aerobic training program. The results show an improvement in VO₂ peak and in exercise intensity. Although we have to notice that the participants didn't have lung involvement.

2.7.2 Measures for Daily Living

Self-therapy exercises

Mugii et al. (2006) evaluated the effectiveness of self-administered stretching program for individual fingers. Each exercise was held for 10 seconds and repeated 3 to 10 times. The three exercises were MCP flexion, IP extension and IP extension. The total ROM measures in each finger joint improved 1 month after intervention as well as the hand function. These improvements were maintained or improved 1 year after the first visit.

Raynaud's phenomenon (RP)

Many common-sense preventive measures can be taken to prevent the occurrence of the RP. The most obvious is to minimize exposure to cold, such as outdoor weather, air conditioning or reaching into a refrigerator. Gloves should be worn as much as possible. It is also important to keep warm the body extremities with for example wearing hat, ear muffs, heavy socks and warm layered clothing. Keeping the whole body warm help prevent the Raynaud's episodes.

Coping with stress

The onset of Scleroderma is a mixed between genetics factors and environmental factors, of which one is stress. A study by Hui et al. (2009) has shown that 72% of sclerodermic patients self-reported having an increased psychological stress before the onset of the disease. Among these people, 80% had undergone some kind of surgery and 41% had history of neck tension. The stress not only plays a role on the onset but also in continuation and exacerbation of the disease.

That's why it is important for the patients to be aware of this reality and to adopt new attitudes in their life to cope with stress. There are many possibilities for helping individuals managing their stress, it could be learning some relaxing techniques (yoga, meditation...), having some sport activities or having more social interactions.

Common advices

SSc patients should absolutely avoid smoking because tobacco use increases vascular changes and hyperkeratosis, complicating scleroderma symptoms as well as increasing periodontal and lung disease risk (Alantar, 2011, pp. 1126-1133). It also worsens the RP.

The patients should protect their hand from cold and also from strong chemical agents (detergents, acids...) by wearing some plastic gloves when washing the dishes or cleaning for example.

Patients should be reminded to avoid alcohol, excessive intake of caffeine, and secondhand smoke because they cause peripheral vasoconstriction and contribute to xerostomia (dry mouth).

2.7.3 Occupational Therapy

Occupational therapy may also be necessary to address independence in activities of daily living. A home assessment might be indicated to ascertain that the environment is optimal for the individual's needs. Assistive devices might be recommended to aid with common daily tasks.

An ergonomic evaluation to assess the work station may enable individuals with scleroderma to maintain employment.

2.7.3 Medications (Imboden, 2004)

The current therapies use medications that focus on the four main features of the disease: inflammation, autoimmunity, vascular disease, and tissue fibrosis.

Anti-inflammatory medications

The inflammation in scleroderma can affect the joints (arthritis), the muscles (myositis) or the lungs (pleuritis) and the heart (pericarditis). It is treated by common anti-inflammatory drugs: NSAIDs (e.g. ibuprofen) or corticosteroids (e.g. prednisone).

Immunosuppressive therapy

The immunosuppressive therapy is the most popular approach for controlling the inflammatory phase in scleroderma. It is logical as it is the autoimmune process that causes inflammation and result in tissue damage and fibrosis. The immunosuppressing

drugs that are the more used for scleroderma include the methotrexate, cyclosporine, antithymocyte globulin, mycophenolate mofetil and cyclophosphamide.

Drug therapy of vascular disease

The vascular disease in scleroderma is widespread and affects medium and small arteries. It is manifested by Raynaud's phenomenon. There are three major features of the vascular disease that need treatment: vasospasm (spasm of blood vessels), vasculopathy (thickening of blood vessels) and, thrombosis (blood clots). Vasospasm is treated by vasodilator drugs. The most effective are the calcium channel blockers (e.g. nifedipine).

There are no specific agents that can reverse the thickening of the vessels but, the vasodilators can be also effective as they directly affect the tissue fibrosis.

The blood clotting can be prevented by anti-platelet therapy and, in acute states, with anti-coagulants.

Anti-fibrotic agents

Several drugs are used for limiting the collagen production including the colchicine, para-aminobenzoic acid (PABA), dimethyl sulfoxide, and D-penicillamine.

3. Special Part

3.1 Methodology

I underwent my clinical practice at the Revmatologický ústav, Prague, from the 3rd of February to the 14th of February 2014. The institute is specialized in the treatment of arthritis and all forms of rheumatic diseases. I worked in the physiotherapy department which is composed of 6 physiotherapist. The care to the patient is either given in the physiotherapy hall (5 therapy tables) or directly in the patient bedroom when he is less ambulant. The inpatient department provides 50 beds.

The more frequent symptoms of the patients are the Rheumatoid Arthritis, Ankylosing spondylitis, Lupus, Polymyositis, Reactive Arthritis and scleroderma.

The age of the patients is mainly over 45 years old, but some patients are younger with some specific pathologies (Juvenile Idiopathic Arthritis for example).

My clinical practice was under the supervision of Mgr. Maja Špiritovič and, all examinations and therapeutical procedures were done in cooperation with her.

My patient gave her informed consent on participating in the bachelor case study, and the work has been approved by the Ethics committee of the Faculty of Physical Education and Sport at Charles University, Prague.

3.2 Anamnesis

Performed 04.02.14

Person being diagnosed: I.K.

Year of birth: 1962

Female

Diagnosis: Systemic Scleroderma – M 340

Present state:

Height : 167 cm

Weight : 67 Kg

BMI : 24

The patient was admitted to hospital a week ago for a total endoprosthesis of the right hip. As she had an infection of the left hand caused by an ulceration of one fingertip (resulting from the scleroderma), the surgeon postponed the operation until this problem is solved.

She is suffering a lot from her right hip (arthrosis, grade III) and she needs 2 crutches for walking which she left at home.

From the scleroderma symptoms, she reports feeling pain mainly in the left hand, and more globally in the joints of both hands. She has difficulty for eating due to esophageal issues, she eats only small pieces of food and needs water for helping. She also reports difficulties for breathing especially when she has activities such as climbing stairs.

This pathology limits her in her activities for daily living, as she is working on a computer 8 hours a day, the pain in hands and fingers is a big disable for her and she even think that she will not be able to continue this job for a long time.

At home, she can fulfill almost every task by herself like cooking, washing or cleaning, however sometimes she needs help for example for washing her hairs or putting socks.

Her parents and her daughter (who is also affected with scleroderma) are good supports for her.

Family history:

Her daughter was diagnosed with sclerodermia when she was 20 year o (2001) and with Crohn disease when she was 27 year old. She has a son who doesn't have any pathology.

Her father had a stroke and suffered from hemiplegia, he has psoriatic arthrosis and diabetes mellitus type II. Her mother had hysterectomy, scoliosis and hormones deficits.

Her grandfather had a cancer of stomach and her grandmother some heart diseases.

A sister from her mother also suffers from scleroderma.

Medical history:

- 1977 : appendix ablation,
- had angina almost chronic (every month) when she was younger,
- had 2 times colles fracture when she was younger,
- 1980 : arthroscopy of right knee in 1980,
- 1981 : diagnosed with hepatitis A.
- 2004: TEP of left hip in 2004, the TEP of right hip is planned.

History of present problem:

The scleroderma was diagnosed for the first time in september 2008, she is coming for the 3rd time to hospital for this problem.

Allergies:

Penicillin, iodin.

Gynecological:

Menses at 15 yo, menopause since 2006.

Medication:

Prednison 20 mg (2-0-0), Loseprazol 20 mg (1-0-1), Calcichew D3 (1-0-1), Vigantol (2 per day), KCl obden, Ganaton (1-1-1), Coxtral (1-0-1), Novalgin, Tralgit 100 mg (1-0-1).

Social:

She is living in a house (2 stairs) with her parents and her husband. Her parents help her when she needs it but they start get older.

Occupational:

She is working as administrative agent for medical care.

Hobbies:

She likes watching TV and reading books.

Abuses:

Not smoking, drinking socially, coffee 2 times a day.

Prior rehabilitation:

The patient remembered having first rehabilitation in 1980 after the arthroscopy of her right knee, the therapy was mainly focused on gait re-education exercises. She didn't suffer much from her right knee since this time.

She also underwent rehabilitation after the TEP of her left hip in 2004, she had a normal recovery after this operation without complication. But now she is suffering a lot from her right hip and waiting for a TEP, she will be able to have it when the actual problem with scleroderma (inflammation) will be solved.

Excerpt from patient's health care file:

She underwent a bone density test the 30th of January on the lumbar spine, neck of femur and on radius. The results are normal which means that she doesn't have osteoporosis.

Indications to rehabilitation:

The doctor's prescription of physiotherapy dated from the 30th of January 2014 recommends mobilization of joints, soft tissue techniques, PIR on shortened muscles, ergotherapy sessions in group, initial and final kinesiological examination. This prescription concerns exclusively the symptoms of the scleroderma and excludes any therapy on the right hip which is too painful.

Differential balance:

As discussed in theoretical part and according to the diagnosis of SSc, we can expect the following symptoms for this patient:

- The CREST syndrome corresponding to :
 - o Calcinosis,
 - o Raynaud s phenomenon,
 - o Esophagal dysfunction,
 - o Sclerodactyly,
 - o Telangiectasias
- Skin thickening,
- Involvement of the musculoskeletal, pulmonary cardiac and renal system,
- Skin tightening and fibrosis leading to contracture in hands and limited joint ROM,
- Mandibular resorption and thickening of the periodontal membrane leading to microstomia,
- The symptoms of the lungs involvement are shortness of breath and coughing.

3.3. Initial Kinesiologic Examination

Performed 05.02.14

Postural examination according to Kendall (Kendall, 2005)

Posterior view

- Head: tilted to the left,
- Cervical Spine: Lateral flexion to the left,
- Shoulder: Left is lower,
- Scapulae: abducted,
- Thoracic and lumbar spine: thoracolumbar scoliosis convex toward the left,
- Sub-gluteal line: shifted to the right

Lateral view

- Head: forward,
- Cervical Spine: extended,
- Shoulders: protraction
- Lumbar spine: hyperlordosis,
- Trunk: bent forward,
- Pelvis: anteversion and right side is forward,
- Hip Joints: right hip flexed at 30°, left hip more neutral position,
- Knee Joints: right knee flexed at 20°, left knee neutral position,
- Feet: right foot is forward.

Anterior view

- Head: tilted to the left, slightly rotated to the left,
- Shoulders: Left is lower,
- Trunk: rotation to the right side,
- Lower extremities: right leg is forward with flexion in hip and knee,
- Hips Joints: internal rotation of right hip,
- Knee Joints: valgosity,
- Feet: narrow base, inward rotation.

Pelvis examination

- Iliac crest : right crest is lower,
- PSIS: right is lower,
- ASIS: right is lower,
- PSIS/ASIS: PSIS is upper than ASIS.
 - ⇒ Pelvis is tilted to the right side with anteversion.

Gait examination

Normal walking (without crutches)

- Proximal pattern, Hip walking, the main movement comes from the hip,
- Antalgic gait, the patient spend a minimum time on the right foot which is landing in plantar flexion (on tip toes), with barely touching the floor with the heel,
- The right hip and right knee stay in flexion during walking,
- The whole trunk is bent to the right side when the right foot is supporting, the cervical spine compensate left latero-flexion,
- The trunk is almost straight when the left foot is supporting,
- This asymmetry between right and left leg gives the following pattern to the trunk:
 - Up and straight when left foot is supporting,
 - Down and lateral bending to the right when right foot is supporting,
- Increased lordosis in lumbar area during walking,
- No hip extension on both sides.
- The patient is not able to walk on long distance.

Anthropometric measurement

Measurement	Left	Right
Biceps Brachii relaxed	27	29,5
Biceps brachii contracted	29,5	31
Elbow	23	25
Forearm	23,5	24
Wrist	16	14
Metacarpals	19	20,5

Table 2 - Anthropometry of the upper extremities (cm), initial examination

Measurement	Inspiration	Expiration
Thorax	78	76

Table 3 - Anthropometry of the thorax (cm), initial examination

Measurement	Left leg	Right leg
Anatomical length	Not performed because the trochanter major of right femur is too painful to be touched	
Functional length (from umbilicus)	94	94
Functional length (from iliac crest)	87	83 (permanent flexion of hip and knee)
Circumference of thigh (15 cm above the patella)	51	50
Patella	37	39 (permanent knee flexion)
Under the knee	34	35
Calf	33,5	34
Malleolus	22,5	23
Metatarsals	22,5	22

Table 4 - Anthropometry of the lower extremity according (cm), initial examination

Breathing Pattern

The breathing pattern is mostly clavicular and very short. The patient report to be out of breath when she has to climb stairs.

Range Of Motion

Passive movement upper extremities	Left				Right			
<u>Shoulder</u> : Flexion	160				160			
Extension	30				30			
Abduction	150				160			
Internal rotation	30				30			
External rotation	90				90			
<u>Elbow</u> : Flexion	140				140			
Extension	0				0			
<u>Forearm</u> : pronation	90				90**			
Supination	90				70**			
<u>Wrist</u> : Radial duction	30				10**			
Ulnar duction	35				20**			
Dorsiflexion	90				60**			
Palmar flexion	80				50**			
<u>MCP</u> :Flexion(II, III, IV, V)	80	80	80	90	80	80	80	80
Extension (II, III, IV, V)	20*	20*	20*	20*	45	45	45	45
Abduction (II, III, IV, V)	20	20	20	20	20	20	20	20
<u>PIP</u> : Flexion (II, III, IV, V)	70	70	70	70	70	70	70	70
Extension (II, III, IV, V)	-10	-10	-10	-10	-10	-10	-10	-10
<u>DIP</u> : Flexion (II, III, IV, V)	10	10	0	10	50	0	0	0
Extension (II, III, IV, V)	0	0	0	0	0	0	0	0
<u>Thumb</u> : MCP Flexion	50				50			
MCP Extension	0				0			
IP Flexion	40				45			
IP Extension	0				0			
Opposition(thumb-5 th digit)	OK				OK			

Table 5 - ROM of upper extremities according to Kendall (Kendall, 2005) in degrees, initial examination

*: Pain

** : Intravenous perfusion in forearm

	Left				Right			
<u>Hip</u> : Flexion	90				70*			
Extension	0				-20*			
Abduction	20				10*			
Adduction	Not tested because of TEP				10*			
Internal rotation	10				Not tested because of pain			
External rotation	15				Not tested because of pain			
<u>Knee</u> : Flexion	120							
Extension	0				-5			
<u>Foot</u> : Plantar Flexion	45				45			
Dorsiflexion	10				10			
Inversion	45				40			
Eversion	20				20			
<u>MTP</u> :Flexion (II, III, IV, V)	20	30	45	10	20	30	45	10
Extension (II, III, IV, V)	30	40	45	20	30	40	45	20
<u>PIP</u> : Flexion (II, III, IV, V)	40	45	40	30	45	40	40	30
Extension(II, III, IV, V)	0	0	0	0	0	0	0	0
<u>DIP</u> : Flexion (II, III, IV, V)	10	10	20	20	10	10	20	10
Extension(II, III, IV, V)	0	0	0	0	0	0	0	0
<u>Hallux</u> : MTP Flexion	30				30			
MTP extension	0				0			
IP Flexion	45				45			
IP Extension	0				0			

Table 6 - ROM of lower extremities according to Kendall (Kendall, 2005) in degrees, initial examination

*: Pain

Muscle strength test

Muscle strength	Left side	Right side
Sternocleidomastoid	5	5
Scaleni	3	3
Trapezius	5	5
Serratus anterior	5	4**
Rhomboids	5	4**
Trapezius upper	5	5
Trapezius lower	3	3
Anterior deltoid	5	4**
Supraspinatus and middle Deltoid middle part	5	4**
Deltoids posterior	5	4**
Pectoralis major	5	4**
Infraspinatus Teres minor	5	4**
Subscapularis Teres major	5	4**
Biceps, Brachialis, brachioradialis	5	Not tested because of intravenous perfusion
Triceps brachii	5	“
Supinator biceps	5	“
Pronator teres Pronator quadratus	5	“
Flexor carpi radialis	5	5
Flexor carpi ulnaris	5	5
Extensor carpi radialis longus and brevis	5	5
Extensor carpi ulnaris	5	5
Lumbricals	5	5
Flexor digit. Superf.	5	5
Flexor digit. Profund.	5	5
Extensor digitorum	4	4
Dorsal interossei	5	5

(continued)

Palmar interossei interossei	5	5
Opponens digiti minimi	5	5
Opponens pollicis	5	5
Flexor pollicis brevis	5	5
Flexor pollicis longus	5	5
Abductor pollicis brevis	5	5
Abductor pollicis longus	5	5
Adductor pollicis	5	5
Rectus abdominis	3	3
Quadratus lumborum	5	4
Iliopsoas	5	3*
Gluteus maximus	4	1*
Gluteus minimus	4	2*
Hip adductors	5	3*
Hamstring	5	4*
Quadriceps femoris	5	4
Soleus	5	4
Triceps surae	5	4
Tibialis anterior	5	4
Tibialis posterior	5	4
Peronei	5	4
Lumbricals	5	5
Flexor digitorum brevis	5	5
Flexor digitorum longus	4	4
Extensor dig longus brevis	4	4
Flexor hallucis brevis	5	5
Flexor hallucis longus	5	5
Extensor halluces brevis	5	5
Extensor hallucis longus	5	5

Table 7 - Muscle Strength Test according to Kendall (Kendall, 2005), initial examination

*: pain

** : Intravenous Perfusion

Measurement of the strength of the hand with a dynamometer

The patient has a perfusion in the left forearm and the left is swollen and painful, it prevents her to give the full strength in this hand.

Measure of the strength	Left hand	Right hand
Whole hand	12	17
Thumb and 2 nd finger	2	2
Thumb and 3 rd finger	2	2
Thumb and 4 th finger	1	2
Thumb and 5 th finger	1	1

Table 8 - Hand Muscles Strength Measurement with a Dynamometer in Kg

Assessment of muscle shortness

0 : no shortness

1 : moderate shortness

2 : marked shortness

Muscles	Left side	Right side
Sternocleidomastoid	0	0
Pectoralis major	1	1
Trapezius upper part	1	1
Levator scapulae	0	0
Quadratus lumborum	0	1 (compensation)
Iliopsoas	Not performed because of pain in right hip	“
Tensor fasciae latae	“	“
Rectus femoris	“	“
Piriformis	0 (TP)	-
Hamstrings	-	-
Gastrocnemius + soleus	0	0
Soleus	0	1

Table 9 - Muscle length test according to Janda (Janda, 1983), initial examination

TP: Trigger Point

Special tests

Thomayer: - 34cm

Shober: 3,5 cm

Stibor: 8 cm

The Romberg, Trendelenburg and scale tests were not performed because of the instability of the patient.

Reflex changes

Head

- The skin is tight, dry and shiny.
- The fascia of the face and cranium are not easily movable,
- The opening space of the mouth is reduced (microstomia),
- Hairs are dry.

Arms

- The skin is dry and thin from the elbow to the wrist,
- Fascia are movable with slight restriction,

Hands

- The skin is dry, thick and shiny,
- Lack of elasticity of the connective tissue,
- Fingers are swollen with red and white colors (Raynaud's phenomenon)
- There is an ulceration on the tips of the 4th digit of the left hand.

Legs

- The skin is dry and thin,
- Fascia are movable with a light restriction

Feet

- The skin is tight and shiny without Raynaud's phenomenon and any swelling.

Joint play examination

N: normal movement of the joint without restriction

Joint	Left side	Right side
Elbow	N	N
Distal Radio-ulnar	N	N
Radio-carpal	N	N
Metacarpo-phalangeal	N	N
PIP (II, III, IV, V) of the hand	Restriction in dorsal and lateral with hard barrier direction of II, III, IV, V	Restriction in dorsal and palmar direction with hard barrier of II, III, IV, V
DIP (II, III, IV, V) of the hand	Restriction in dorsal, palmar and lateral direction with hard barrier of II, III, IV, V	Restriction in dorsal, palmar and lateral direction with hard barrier of III, IV, V
IP pollicis	N	N
Patella	N	N
Tibiofibular	N	N
Tarso-metatarsal	N	N
Metatarso-phalangeal (I, II, III, IV, V)	Restriction in hallux in dorsal, plantar and latero-lateral direction. II, III, IV, V joints are N	Restriction in hallux in dorsal, plantar and latero-lateral direction. II, III, IV, V joints are N
PIP (II, III, IV, V) of the foot	N	N
DIP (II, III, IV, V) of the foot	N	N
Lisfranc	N	N
Schopard	N	N
Subtalar	N	N
Tibiofibular	N	N

Table 10 - Joint Play Examination according to Lewit (Lewit, 1999), initial examination

Neurological examination

Cranial nerve	Test provided	Result
I. olfactorius	Common odor used for testing the smell : coffee, food	No problem with smelling
II. opticus	Test of reaction to light	Normal
III. oculomotorius	Test of extraocular movement	Normal
IV. trochlearis	Test of extraocular movement	Normal
V. trigeminus	Test of temporal masseter muscle Test of 3 divisions for pain	Normal
VI. abducens	Test of extraocular movement	Normal
VII. facialis	Test : raise eyebrows, close eyes, smile, show teeth, puff out cheeks	Any asymmetry during these movements
VIII. vestibulocochlearis	Hearing test	Normal
IX. glossopharyngus	Swallowing test "say Ah"	Normal
X. vagus	Same tests like glossopharyngeal nerve	"
XI. accessories	Asymmetry of trapezius Shrug shoulder against resistance Turn head against resistance and palpation of SCM muscle	Any asymmetry OK OK
XII. hypoglossus	Articulation of the words Protrusion of the tongue Move the tongue from side to side	Good Good Good

Table 11 - Examination of the Cranial Nerves

Sensory system	Test provided	Result
Subjective light touch	Light touch of the skin with fingers on both sides	Patient doesn't feel difference between both sides or different area, results OK
Position test	Grasp big toe and ask patient if it is up or down, similar with fingers (with closed eyes)	OK
Pain	Use of sharp object to test sharp or dull sensation on the whole body	OK
Light touch	Light touch with fingers and ask the patient to respond whenever the touch is felt	OK
Discrimination	- Graphesthesia (draw a number on patient hand and ask) - 2 points discrimination (on the hand)	OK Minimal distance for discrimination is 1cm

Table 12 - Examination of the Sensory System

Superficial and tendon reflexes	Result
Abdominal reflex	Normal
Biceps brachii reflex C5-C6	Normal
Triceps reflex C7	Normal
Palmar reflex	Normal
Knee jerk reflex L3-L4	Hyporeflex
Ankle reflex L5-S2	Hyporeflex
Plantar reflex	Hyporeflex

Table 13 - Examination of the Superficial and Deep Tendon Reflexes

Examination's conclusion:

Postural examination

- Flexion of the right hip and knee in standing position,
- Protraction of the head and tilt to the left,
- Trunk bent forward and rotated to the right,
- Scoliosis thoracic area,
- Hyperlordosis in lumbar area,
- Hallux valgus on left foot.

Gait

The right hip is very painful (arthrosis) and the patient walk with antalgic gait giving the following pattern: up and straight when left foot is landing and, down and right lateral bending when the right foot is landing. She can walk only on short distance.

Anthropometric measurement

The left wrist is bigger than the right one resulting from ulceration on fingertip causing inflammation.

Breathing pattern

Shortness of breath, mostly clavicular pattern.

ROM

- MCP extension limited to 20° for left hand and painful,
- PIP joints of both hands have a permanent flexion which is at 10° in neutral position, there is no possible extension,
- DIP joints of both hands have a limited flexion comprise between 0 and 10° (except index of right hand).

Reflex changes

- Hands swollen with Raynaud's phenomenon, skin is thick, lacking of elasticity,
- Ulceration on 4th digit of left hand,
- Restriction in fascia of the face, cranium,
- Restriction of the opening space of the mouth.

Joint play examination

There is a restricted joint play in digits of both hands, with hard barrier in PIP and DIP joints.

3.4 Short-term and long-term physiotherapy plan

Following the doctor recommendations for this patient, we will not apply therapy on the right hip that is very painful, but we will focus on the symptoms of the scleroderma which mainly affects the hands, the face and the lungs of this patient.

Short-term physiotherapy plan

- Lymphatic drainage of the upper extremities (with a softball or hand),
- Kibler fold on the arms and face,
- Fascia therapy on the arms, cranium and neck,
- Soft tissue technique on the palmar side of hands and fingers,
- Mobilization by Lewit of all the joints of the hands,
- ROM exercises with passive and active movements for the wrists, hands and fingers,
- Strengthening exercises for the hand,
- Mobilization of connective tissue on the face,
- Active stretching exercises of the face (“mimic” exercises),
- PIR of shorten muscles (levator scapula, upper trapezius and pectoralis major),
- Breathing exercises,
- DVT exercises,
- Education of the patient for reducing the symptoms in ADLs (at work and home).

Long-term physiotherapy plan

- ROM exercises for the hands and fingers,
- Facial stretching exercises,
- “Chin tuck” exercise,
- Self PIR for levator scapula, upper trapezius and pectoralis major,
- Breathing exercises.

Once she will have undergone her right hip TEP:

- Corrective posture exercises, walking exercises,
- Some soft sport activities that could improve her general condition : nordic walking, cycling... She should avoid the swimming pools because the chlorine in the water could be too aggressive for her skin,
- Joining an activity like yoga could be a great benefit for her for the breathing patterns, joint movement and for managing the stress which is an important aggravating factor for sclerodermic patients,
- Some manual activities in group (occupational therapy or art-therapy) could also be of great interest.

3.5 Therapy progress

3.5.1 Therapy session 1

Performed 04.03.14

Current state:

The patient has necrosis on the fingertip of her 4th digit on her left hand. This necrosis causes an inflammation to the whole left hand which is swollen, red and very painful. The patient came to hospital primarily to undergo a TEP of her left hip, but because of this problem, the doctors decided to postpone the operation. She has an intravenous perfusion of antibiotics in her left forearm in order to reduce the inflammation.

She also suffers a lot from her right hip. She was used to walk with crutches, but she forgets it at home, and she can barely walk without it.

She feels really disappointed for the postpone of the operation, but she is in a global good mood and very cooperative.

Goals of today therapy:

It was the first meeting with the patient today, and the session included taking the anamnesis of the patient and starting performing the initial kinesiologic examination (see above).

Any therapy was performed.

3.5.2 Therapy session 2

Performed 05.03.14

Current state:

There is no apparent change in the aspect of the necrosis on her left hand, the hand is still swollen and she has still an intravenous perfusion in her left forearm.

She reported the pain in her body as follow:

- Both hands are very painful with disagreeable feelings on her fingertips (tingling). Both hands are swollen (left is worse) with Raynaud's phenomenon,
- Both wrists are painful, the pain increases when flexed or extended,

- Her neck is painful with more intensity on the right side,
- Right hip is very painful, she avoids moving it as much as possible and keeping a permanent flexion of 20°.

Goals of today therapy:

- Continue the initial kinesiologic examination,
- Decrease the swelling in hands,
- Improve the ROM of IP joints,
- Decrease muscle tone in neck muscles,
- Instruct patient for self-therapy.

Therapy applied:

- Lymphatic drainage on the upper extremities with a soft ball,
- Kibble fold on the upper extremities,
- Fascia stretching on the upper extremities,
- Soft tissue technique on the palmar side of the hand and on fingers,
- Mobilization by Lewit of the joints of the hands and wrists:
 - o Mobilization of IP joints in all directions (dorsopalmar, lateral, rotation)
 - o Mobilization MCP heads in dorsopalmar direction,
 - o Mobilization CMC joints of the thumb in in dorsopalmar direction,
 - o Mobilization of RCJ in dorsal direction direction on radial and ulnar sides,
 - o Mobilization of ICJ in palmar direction,
- Fascia stretching of the neck (rotational movement),
- PIR by Lewit of upper trapezius and levator scapulae on both sides.

Results:

The patient is very cooperative and enjoyed this session.

The ROM of wrist and MCP joints are normal, but limited at PIP and DIP on both hands. The mobilization of these joints showed that they are restricted.

Self-therapy:

- Active and passive (with the other hand) ROM of fingers and wrist joints into flexion and extension,

- Self PIR of upper trapezius and levator scapulae,
- Patient was instructed how to provide exercises for strengthening the deep neck flexors : in sitting position the patient move forward and backward the chin (“chin tuck” exercise), 10 repetitions, 3X/day,
- DVT exercises without hip flexion on the right side.

3.5.3. Therapy session 3

Performed 06.02.14

Current state:

The patient feels fewer pains in the neck today maybe because she did the self PIR for relaxing the muscles and also she slept with one less pillow under her head during the night.

There is any particular evolution in the aspect of the necrosis on her fingertips. Hands are still swollen with Raynaud’s phenomenon. The antibiotic perfusion is today on her right forearm.

Goals of today therapy:

- Decrease the swelling in hands,
- Improve the ROM of IP joints,
- Improve the strength of the hand muscles,
- Stretching exercises for the facial muscles,
- Instruct patient for self-therapy.

Therapy applied:

- Lymphatic drainage on the upper extremities with a soft ball,
- Kibbler fold on the upper extremities,
- Fascia stretching on the upper extremities,
- Soft tissue technique on the hands and fingers: the physiotherapist apply with both thumbs a connective tissue massage on the on the palm of the hand and fingers. We use a moisturizing cream for this massage to provide hydration of the skin that is very dry especially on fingertips. We also avoid carefully the necrosis on the fingertip.

- Mobilization by Lewit of the joints of the hands:
 - o Mobilization of IP joints in all directions (dorsopalmar, lateral, rotation)
 - o Mobilization MCP heads in dorsopalmar direction,
 - o Mobilization CMC joints of the thumb in in dorsopalmar direction,
- Strengthening exercises of the hand:
 - o Pressing against a soft tennis ball (15X),
 - o Exercises with a theraband hand trainer : repeated flexion of the fingers (15 X), extension (15 X), rotation with pronation and supination of the forearm (15X),
- ROM exercises:
 - o Wrist extension stretch : palms together in front of the chest and lower them down the chest at maximum with keeping the contact between the palm (keeping position 20 seconds),
 - o Wrist flexion stretch: dorsal side of the hand together and upper them up the chest to maximum flexion (keeping position 20 seconds).
- Fascia stretching of the neck (rotational movement),
- PIR by Lewit of upper trapezius, levator scapulae and pectoralis major on both sides.

Results:

- The PIR of upper trapezius and pectoralis major gave a good relaxation to this muscles which were shortened. The patient reported feeling less tension after treatment. There was not such improvement after the PIR of levator scapulae which were not shortened.
 - The ROM of IP joints are restricted:
 - o PIP joints slightly restricted into flexion (70°) and restricted into extension (-10°),
 - o DIP joints are very restricted into flexion (0° or 10°).
- ⇒ In order to improve these ROM, I instructed the patient for self-therapy as mentioned below.

Self-therapy:

- Self PIR of upper trapezius and levator scapulae,
- DVT exercises without hip flexion on the right side,

- Finger ROM exercises :
 - Practice making a fist and then stretching all the fingers out like a star,
 - Touching each finger tips with the thumb tip and after, slide the thumb tip down the side of each finger,
 - Placing the palm of the hand on a block (edge of a table, matchbox...) and flex each finger joint (MCP, IPJ, DIP joints) individually and straighten out fully with the help of the other hand if necessary.
- “Chin tuck” exercise.

3.5.4. Therapy session 4

Performed 07.02.14

Current state:

The patient feels better today. She is experiencing less pain in her hands and wrists during movements. The swelling in hands is still present but appears to have slightly decrease, the Raynauds phenomenon is also less present which means that the irrigation of the soft tissues has increased. The necrosis didn't evaluate in aspect and size.

Goals of today therapy:

- Decrease the swelling in hands,
- Improve the ROM of IP joints,
- Improve the strength of the hand muscles,
- Stretching exercises for the facial muscles,
- Improve the breathing patterns.

Therapy applied:

- Lymphatic drainage on the upper extremities with a soft ball,
- Kibler fold on the upper extremities,
- Fascia stretching on the upper extremities,
- Soft tissue technique on the palmar side of the hand and on fingers,
- Mobilization by Lewit of the joints of the hands:
 - Mobilization of IP joints in all directions (dorsopalmar, lateral, rotation).
- Strengthening exercises of the hand:

- Use of a hand strengthener grip, repeated contractions (20X),
- Exercises with a theraband hand trainer : repeated flexion of the fingers (15 X), extension (15 X), rotation with pronation and supination of the forearm (15X),
- Massaging of the soft connective tissue of the face,
- Exercises for the facial muscles:
 - Raising of the eyebrows (frontalis muscle) and lower them,
 - Squeeze the eyes closed tightly. Wink tightly with either eyes,
 - Flare the nostrils. Close the lips hard,
 - Cover the teeth with the lips, and then open the mouth as wide as possible,
 - Grin as widely as possible, without showing the teeth.
- Breathing exercising.

Results:

We started today for the first time the exercises for the facial muscles. In order to facilitate the stretching of the facial muscles, I first gave a massage to soft connective tissue of the face which allows the skin, fascia and muscles to be more relaxed and warm.

The patient wasn't used to these kind of exercises, and due to the tightness of her skin, the movements concerning the opening of the mouth were very restricted (microstomia).

The patient was then instructed to practice the facial exercises ("mimic" exercises) as self-therapy.

Self-therapy

- Active and passive (with the other hand) ROM of fingers and wrist joints into flexion and extension, at least 3X daily,
- Practice the facial exercises shown during this session, at least 3 times/day, if possible in front of a mirror and to hold each stretch during 10 seconds.
- "Chin tuck" exercise.

3.5.5. Therapy session 5

Performed 10.02.14

Current state:

The patient feels a little bit tired today; she didn't sleep such well last night. She had also esophageal acidity return from the diner the day before.

Goals of today therapy:

The goals of today therapy are the same than the previous session, we will continue to improve the ROM in hands and the facial stretching. As the patient feels a little bit tired, we will avoid the strengthening exercises of the hand.

Therapy applied:

- Lymphatic drainage on the upper extremities with a soft ball,
- Kibler fold on the upper extremities,
- Fascia stretching on the upper extremities,
- Soft tissue technique on the palmar side of the hand and on fingers,
- Mobilization by Lewit of the joints of the hands:
 - o Mobilization of IP joints in all directions (dorsopalmar, lateral, rotation)
- Fascia stretching of the neck (rotational movement),
- Kibler fold provided on whole face,
- Exercises for the facial muscles: same "mimic exercises" as practiced last session,
- ROM exercises of the hand:
 - o Wrist extension and flexion as seen in session 3,
 - o Opposition of thumb with other fingers,
 - o MCP joints extension: start position palm to palm, then palms separate from each other and finger stay in contact. At maximum extension of MCP joints the position is kept during 20 seconds,
 - o MCP joints flexion: making a fist with the help of the other hand to have maximum flexion at MCP joints, this exercise provides also flexion in IP joints,
 - o Extension of IP joints (PIP and DIP): use of 2 fingers acting like a lever on t IP joints of the other hand to provide maximum extension (Photo 3).

- Breathing exercises.

Results:

The patient is responding well to the therapy. She says that she feels better sensations in her hands with less stiffness especially in the morning.

Self-therapy exercises:

- ROM exercises for the hands and fingers,
- Facial stretching exercises,
- “Chin tuck” exercise.

3.5.6. Therapy session 6

Performed 11.02.03

Current state:

The patient is feeling less pain in her right hip and she now can walk (without crutches) with less difficulties on longer distance. The gait improved but is still very antalgic and she bares most of her weight on her left leg. She avoids taking the stairs which is difficult for her.

As she can walk better, she will go today for the first time to an occupational therapy session and the physical therapy session will be shortened. During this occupational therapy session, she practiced different manual activities in group like creating of jewels, silk painting, sewing...

All this activities are managed by an occupational therapist.

Goals of today therapy:

- Decrease the swelling in hands,
- Improve the ROM of IP joints,
- Improve the strength of the hand muscles,
- Stretching exercises for the facial muscles.

Therapy applied:

- Lymphatic drainage on the upper extremities with a soft ball,

- Kibler fold on the upper extremities,
- Fascia stretching on the upper extremities,
- Soft tissue technique on the palmar side of the hand and on fingers,
- Mobilization by Lewit of the joints of the hands:
 - o Mobilization of IP joints in all directions (dorsopalmar, lateral, rotation)
- Strengthening exercises of the hand:
 - o Exercises with a theraband hand trainer : repeated flexion of the fingers (15 X), extension (15 X), rotation with pronation and supination of the forearm (15X),
- ROM exercises:
 - o Wrist extension and flexion as seen in session 3,
 - o Opposition of thumb with other fingers,
 - o MCP joints extension: start position palm to palm, then palms separate from each other and finger stay in contact. At maximum extension of MCP joints the position is kept during 20 seconds,
 - o MCP and IP joints active flexion: making a fist,
 - o Passive extension of PIP and DIP joints,
- Exercises for the facial muscles: same as previous session.

Results:

- Physical therapy session: The ROM with passive movements of IP joints showed some improvement compared to previous sessions. The patient was then encourage to continue the self-therapy exercises for fingers to improve even more the results.
- Occupational therapy session: she enjoyed a lot this moment. That kind of session procures many benefits for the patient at different levels. On the psycho-social level, it allows the patient to have a break from the routine of her bed room: she can relax, establish social contact with other patients, develop her artistic skills... On the functional level, it allows her to re-educate and train her motor functions during pleasurable activities.

Self-therapy:

- ROM exercises for the hands and fingers,
- Facial stretching exercises,
- “Chin tuck” exercise.

3.5.7. Therapy session 7

Performed 12.02.14

Current state:

The patient feels well today, the swelling in hands is significantly reduced and she reported having much less pain in hands and wrists. She also can walk without much difficulties. She is happy because today she will receive the visit of her daughter (sclerodermic too) and they will go outside for eating and have a small walk.

Goals of today therapy:

- Decrease the swelling in hands,
- Improve the ROM of IP joints,
- Improve the strength of the hand muscles,
- Stretching exercises for the facial muscles,
- Improve the breathing patterns.

Therapy applied:

- Lymphatic drainage on the upper extremities with a soft ball,
- Kibbler fold on the upper extremities,
- Fascia stretching on the upper extremities,
- Soft tissue technique on the palmar side of the hand and on fingers,
- Mobilization by Lewit of the joints of the hands:
 - o Mobilization of IP joints in all directions (dorsopalmar, lateral, rotation)
 - o Mobilization MCP heads in dorsopalmar direction,
 - o Mobilization CMC joints of the thumb in in dorsopalmar direction,
- Fascia stretching of the neck (rotational movement),
- PIR by Lewit of upper trapezius and levator scapulae on both sides (Photo 2).
- Strengthening exercises of the hand:
 - o Exercises with a theraband hand trainer : repeated flexion of the fingers (20 X), extension (20 X), rotation with pronation and supination of the forearm (20X),
- ROM exercises: same as previous session with passive and active movements of the wrists and fingers joints,

- Connective tissue massage and Kibler fold on the face (Photo 1),
- Exercises for the facial muscles: same “mimic exercises” as previous sessions,
- Stretching of the cranial fascia.

Results:

The patient feels less pain in hands that are less swollen and RP is less visible (Photo 5). This way, she can exercise more easily. During this session we increased the repetitions for the strengthening exercises of the hand, she did it without difficulties.

The ROM in IP joints continue to improve, the soft tissues around the joints appear to be less stiff. The size of ulceration doesn't appear to be decreasing even slightly bigger (Photo 6).

Self-therapy:

- ROM exercises for the hands and fingers (Photos 3 and 4),
- Connective tissue massage of the face,
- Facial stretching exercises,
- “Chin tuck” exercise,
- Breathing exercises.

3.5.8. Therapy session 8

Performed 13.02.14

Current state:

Today it is the last day with the patient, she is happy going back home even if she feels disappointed not having operated for her right hip. The swelling in hands has been significantly reduced compared to the first days, however there is no particular evolution in the aspect of the necrosis on her fingertip. She reported having less pain in hands and wrists.

Goals of today therapy:

It was the last day with the patient today, and the session consisted in the final kinesiologic examination. I also gave her a complete information about the scleroderma,

the advances of the research in this domain and advices for managing this disease in her ADL. Any therapy was performed.

Results:

It was a real pleasure to work with this patient during these 8 sessions, she was very cooperative and always willing to improve her condition. Even if it is long term disease, she could improve during this short time her condition, and this is very satisfactory. We will discuss about it in the next chapter.

She was also very interested into the information I gave her about the actual researches about scleroderma and how to handle it for the daily living. She will also relate these information to her daughter which is suffering from scleroderma too.

3.6. Final Kinesiologic Examination

Performed 13.02.03

Changes from the initial kinesiologic examination are marked with bold letters.

Postural examination according to Kendall (Kendall, 2005)

Posterior view

- Head: tilted to the left,
- Cervical Spine: Lateral flexion to the left,
- Shoulder: Left is lower,
- Scapulae: abducted,
- Thoracic and lumbar spine: thoracolumbar scoliosis convex toward the left,
- Sub-gluteal line: shifted to the right

Lateral view

- Head: forward,
- Cervical Spine: **slightly less** extended,
- Shoulders: protraction,
- Lumbar spine: hyperlordosis,

- Trunk: bent forward,
- Pelvis: anteversion and right side is forward,
- Hip Joints: right hip flexed at 30°, left hip more neutral position,
- Knee Joints: right knee flexed at 20°, left knee neutral position,
- Feet: right foot is forward.

Anterior view

- Head: tilted to the left, slightly rotated to the left,
- Shoulders: Left is lower,
- Trunk: rotation to the right side,
- Lower extremities: right leg is forward with flexion in hip and knee,
- Hips Joints: internal rotation of right hip,
- Knee Joints: valgosity,
- Feet: narrow base, inward rotation.

Pelvis examination

- Iliac crest : right crest is lower,
- PSIS : right is lower,
- ASIS : right is lower,
- PSIS/ASIS : PSIS is upper than ASIS
 - ⇒ Pelvis is tilted to the right side with anteversion.

Gait examination

Normal walking (without crutches)

The gait is still very antalgic and the global pattern didn't change with the following trunk movements (**with decreased amplitude**):

- up and straight when left foot is supporting,
- Down and lateral bending to the right when right foot is supporting,
- The patient is now **able to walk on longer distance**.

Breathing Pattern

The breathing pattern doesn't seem to have improved: still shortness of breathing with clavicular pattern.

Anthropometric measurement

Measurement	Left arm	Right arm
Biceps brachii relaxed	27,5 (+0,5)	29,5
Biceps brachii contracted	30 (+0,5)	31
Elbow	23	24 (-1)
Forearm	23,5	24
Wrist	15 (-1)	14
Metacarpals	19	20 (-0,5)

Table 14 - Anthropometry of the upper extremities in cm, final examination

Measurement	Inspiration	Expiration
Thorax	79 (+1)	76

Table 15 - Anthropometry of the thorax in cm, final examination

- lower extremities final examination : any change compare to the initial examination

Range Of Motion

Passive movement upper extremities	Left	Right
<u>Shoulder</u> : Flexion	160	160
Extension	30	30
Abduction	150	160
Internal rotation	30	30
External rotation	90	90
<u>Elbow</u> : Flexion	140	140
Extension	0	0
<u>Forearm</u> : Pronation	90	90
Supination	90	80 (+10)

(continued)

Passive movement upper extremities	Left				Right			
<u>Shoulder</u> : Flexion	160				160			
Extension	30				30			
Abduction	150				160			
Internal rotation	30				30			
External rotation	90				90			
<u>Elbow</u> : Flexion	140				140			
Extension	0				0			
<u>Forearm</u> : Pronation	90				90			
Supination	90				80 (+10)			
<u>Wrist</u> : Radial duction	30				20 (+10)			
Ulnar duction	35				30 (+10)			
Dorsiflexion	90				80 (+20)			
Palmar flexion	80				70 (+20)			
<u>MCP</u> :Flexion(II,III, IV,V)	80	80	80	90	80	80	80	80
Extension (II, III, IV, V)	25 (+5)	25 (+5)	25 (+5)	25 (+5)	45	45	45	45
Abduction (II, III, IV, V)	20	20	20	20	20	20	20	20
<u>PIP</u> : Flexion (II, III,IV,V)	70	70	70	70	70	70	70	70
Extension (II, III, IV,V)	-5 (+5)	-5 (+5)	-5 (+5)	-5 (+5)	-5 (+5)	-5 (+5)	-5 (+5)	-5 (+5)
<u>DIP</u> : Flexion (II, III,IV,V)	20 (+10)	15 (+5)	10 (+10)	10	50	10 (+10)	10 (+10)	5 (+5)
Extension (II, III, IV, V)	0	0	0	0	0	0	0	0
<u>Thumb</u> : MCP Flexion	50				50			
MCP Extension	0				0			
IP Flexion	40				45			
IP Extension	0				0			
Opposition (thumb to 5 th digit)	OK				OK			

Table 16 - ROM of upper extremities according to Kendall (Kendall, 2005) in degrees, final examination

- ROM of lower extremities: any change compared to initial examination

Muscle strength test

The improvement in muscle strength must be put in perspective because the patient had an intravenous perfusion during first examination that prevents her to give full muscle strength. I didn't test again the muscles of lower extremities which were not the purpose of the therapy.

Muscle strength	Left side	Right side
Sternocleidomastoid	5	5
Scaleni	3	3
Trapezius	5	5
Serratus anterior	5	5 (+1)
Rhomboids	5	5 (+1)
Trapezius upper	5	5
Trapezius lower	3	3
Anterior deltoid	5	5 (+1)
Supraspinatus and middle Deltoid middle part	5	5 (+1)
Deltoids posterior	5	5 (+1)
Pectoralis major	5	5 (+1)
Infraspinatus Teres minor	5	5 (+1)
Subscapularis Teres major	5	5 (+1)
Biceps, Brachialis, Brachioradialis	5	Initially not tested because of intravenous perfusion
Triceps brachii	5	“
Supinator biceps	5	“
Pronator teres	5	“
Pronator quadratus		
Flexor carpi radialis	5	5
Flexor carpi ulnaris	5	5

(continued)

Muscle strength	Left side	Right side
Extensor carpi radialis longus and brevis	5	5
Extensor carpi ulnaris	5	5
Lumbricals	5	5
Flexor digit. Superf.	5	5
Flexor digit. Profund.	5	5
Extensor digitorum	4	4
Dorsal interossei	5	5
Palmar interossei interossei	5	5
Opponens digiti minimi	5	5
Opponens pollicis	5	5
Flexor pollicis brevis	5	5
Flexor pollicis longus	5	5
Abductor pollicis brevis	5	5
Abductor pollicis longus	5	5
Adductor pollicis	5	5

Table 17 - Muscle strength test according to Kendall (Kendall, 2005), final examination

Measurement of the strength of the hand with a dynamometer

During the first evaluation, the patient had a perfusion in the left forearm and left hand was painful and swollen. The perfusion has been removed, hand is less swollen and painful so she can give more strength.

Measure of the strength	Left hand	Right hand
Whole hand	14 (+2)	18 (+1)
Thumb and 2 nd finger	3 (+1)	3 (+1)
Thumb and 3 rd finger	3 (+1)	3 (+1)
Thumb and 4 th finger	1	2
Thumb and 5 th finger	1	1

Table 18 - Hand muscles strength measurement with a dynamometer in Kg, final examination

Assessment of muscle shortness

Any noticeable change in the length of upper trapezius and pectoralis major which are moderately shortened due to a faulty posture and also to long time lying in bed.

0: no shortness

1: moderate shortness

2: marked shortness

Muscles	Left side	Right side
Sternocleidomastoid	0	0
Pectoralis major	1	1
Trapezius upper part	1	1
Levator scapulae	0	0

Table 19 - Muscle length test according to Janda (Janda, 1983), final examination

Special tests

These tests show an improvement in the flexion of the spine which may be due to the diminution of the pain induced by the medicaments.

Thomayer: - 30cm (+4cm)

Shober: 4,5 cm (+1cm)

Stibor: 9 cm (+1cm)

Reflex changes

Head

- The skin is tight, dry and shiny,
- **The fasciae of the face and cranium move more easily but are still restricted,**
- The opening space of the mouth is reduced (microstomia),
- Hairs are dry.

Arms

- The skin is dry and thin from the elbow to the wrist,
- Fascia are movable with slight restriction,

Hands

- The skin is dry, thick and shiny,
- **Better elasticity of the connective tissue,**
- **Fingers are less swollen and the Raynaud's is reduced,**
- **The ulceration on the tips of the 4th digit of left hand is still present and appears slightly bigger.**

Legs

- The skin is dry and thin,
- Fascia are movable with slight restriction

Feet

- The skin is tight and shiny without Raynaud's phenomenon and any swelling.

Joint play examination

N: normal movement of the joint without restriction

Joint	Left side	Right side
Elbow	N	N
Distal Radio-ulnar	N	N
Radio-carpal	N	N
Metacarpo-phalangeal	N	N
PIP (II, III, IV, V) of the hand	Restriction in dorsal and lateral direction with softer barrier of II, III, IV, V	Restriction in dorsal and palmar direction with softer barrier of II, III, IV, V
DIP (II, III, IV, V) of the hand	Restriction in dorsal, palmar and lateral direction with softer barrier of II, III, IV, V	Restriction in dorsal, palmar and lateral with softer barrier direction of III, IV, V
IP pollicis	N	N
Patella	N	N
Tibio-fibular	N	N
Tarso-metatarsal	N	N
Metatarso-phalangeal (I, II, III, IV, V)	Restriction in hallux in dorsal, plantar and latero-lateral direction. II, III, IV, V joints are N	Restriction in hallux in dorsal, plantar and latero-lateral direction. II, III, IV, V joints are N
PIP (II, III, IV, V) of the foot	N	N
DIP (II, III, IV, V) of the foot	N	N
Lisfranc	N	N
Schopard	N	N
Subtalar	N	N
Tibiofibular	N	N

Table 20 - Joint play examination according to Lewit (Lewit, 1999), final examination

Neurological examination: no change from initial examination.

3.7. Evaluation of the effects of therapy

The concern of the physical therapy was to deal with the symptoms of SSc which were mainly affecting the hands and the face of the patient and limited her for her daily living activities. It was very satisfactory to see that after only 8 sessions of physiotherapy some improvements could be observed, especially for a long-term disease.

The main improvements in this patient were as follows:

- **A better ROM in the hands digits**
 - MCP joints : +5° into extension for left hand,
 - PIP joints : +5° into extension for both hands,
 - DIP joints: +5 to 10° into flexion for both hands.

- **A decreased of the swelling on both hands**, which could be visually observed and also seen in anthropometric measurement. This result can be partly attribute to the physiotherapeutic treatment (lymphatic drainage, connective tissue massage), but also to the anti-inflammatory treatment that was administered.

- **A better mobility of the hand and face connective tissues**
 - Hand: better elasticity of the skin, RP less visible,
 - Face: the skin and the fasciae are more easiliy movable,
 - Mouth: any measurment about the mouth opening size evolution, a comparison between initial and final state would have been interesting to see if there was any improvement.

- **A decreased sensation of pain in joints of the hand and wrists**, at the first meeting she reported having a lot of pain in all the joints of the hands and wrists with more intensity on the left side (inflammation). This pain slowly decreased all along the stay at the hospital, and can be attributed to the combination of the drug and physiotherapeutic treatment.

- **A better strength in the hand.** The dynamometric measurement has shown slight improvement in the hand's strength which result from the daily strength training and from the decreased of the pain and swelling.
- **Softer barrier in joint play of PIP and DIP joints.** The restriction was initially very hard and became softer all along the therapy course.
- **Slight increasing of the ulceration on the fingertips.** Ulceration are usually long to heal for SSc patients, she will have to take care of it once home and protect it at maximum from external contact.
- **Breathing pattern is still short,** any significant improvement even if the anthropometric measurement show an increase of 1cm at inspiration (thoracic circumference).

4. Conclusion

I was introduced to my patient on my second day of practice at Revmatologický ústav. She arrived at hospital 4 days before, to undergo a total endoprosthesis of the right hip but, as she suffered from ulceration on one fingertip causing inflammation, the orthopedist surgeon postponed the operation. Then she was indicated to receive rehabilitation for her Systemic Sclerosis..

We had 8 physiotherapy sessions together during the 2 weeks of practice, and I could follow her from the very first day of therapy until she left, which was very good because I could have a full view on the whole therapy course and the improvement that happened. During this practice, I could experiment my therapeutic skills learned at school like PIR, fascia therapy, joint play, lymphatic drainage or soft tissue techniques and, see the effect of the therapy on my patient which was very satisfactory.

It was a really interesting to work with this patient because she was very cooperative and also very kind and, I learned a lot about scleroderma which is a quite rare disease. It was very rewarding to see that after only 8 therapy sessions we could see some improvement (even small), especially in hands and face which were the focus of the therapy. I was also glad to see, when doing researches aftermath about scleroderma, that the recent studies on the subject were according the best result in the therapy to the same techniques I used with my patient. The only recommended treatment I didn't use with her was the wax bath but, it was contra-indicated in her case because of the fingertip ulceration.

Before she left hospital, I gave her complete information about scleroderma and how to handle it for ADL, with exercises to practice every day and advices to limit the symptoms. I think she should follow these advices as she was motivated to improve her condition and she will relate it to her daughter who is also affected with scleroderma.

5. List of literature

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6. Appendix

Appendix n° 1: Approval of the Ethics Board Committee



CHARLES UNIVERSITY IN PRAGUE
FACULTY OF PHYSICAL EDUCATION AND SPORT
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Application for Ethics Board Review

of the research project, doctoral research, master degree research, undergraduate research, involving human subjects

Project title: Case study of a patient with scleroderma

Nature of the research project: Bachelor thesis

Author Julien Barange

Supervisor (in case of student research): Doc. Paedr. Dagmar Pavlu

Research project description also involves (maximum 10 lines)
Guaranteed safety to be judged by experts: rationale for the use of invasive methodologies, procedures minimizing the risk to subjects
Ethical aspects of the research: special rationale for research involving children, pregnant and nursing women, mentally disabled, prisoners and persons in underdeveloped communities (see the Ethics Board Code, Faculty of Physical Education and Sport, Charles University, and International Ethical Guidelines 5, 6, 7, 8 and 11)
Informed consent (attached)

Date: 13-02-14 Author's signature: Julien Barange

Faculty of Physical Education and Sport, Charles University in Prague
ETHICS BOARD REVIEW

Ethics Board members: Doc. MUDr. Staša Bartůňková, CSc.
Prof. Ing. Václav Bunc, CSc.
Prof. PhDr. Pavel Slepíčka, DrSc.
Doc. MUDr. Jan Heller, CSc.

The Ethics Board at the Faculty of Physical Education and Sport, Charles University, approved the research project.
Approval number: 091/2014
Date: 24.2.2014

The Ethics Board at the Faculty of Physical Education and Sport, Charles University, reviewed the submitted research project and **found no contradictions with valid principles**, regulations and international guidelines for biomedical research involving human subjects.

The chief investigator of the project met the necessary requirements for receiving the Ethics Board approval.

Official school stamp
UNIVERSITA PRAHA v Praze
fakulta tělesné výchovy a sportu
José Martího 31, 162 52, Praha 6
1

Signature, REB Chairman

Appendix n° 2: Informed Consent / Informovaný Souhlas

V souladu se Zákonem o péči o zdraví lidu (§ 23 odst. 2 zákona č.20/1966 Sb.) a Úmluvou o lidských právech a biomedicině č. 96/2001, Vás žádám o souhlas k vyšetření a následné terapii. Dále Vás žádám o souhlas k nahlížení do Vaší dokumentace osobou získávající způsobilost k výkonu zdravotnického povolání v rámci praktické výuky a s uveřejněním výsledků terapie v rámci bakalářské práce na FTVS UK. Osobní data v této studii nebudou uvedena.

Dnešního dne jsem byla odborným pracovníkem poučena o plánovaném vyšetření a následné terapii. Prohlašuji a svým dále uvedeným vlastnoručním podpisem potvrzuji, že odborný pracovník, který mi poskytl poučení, mi osobně vysvětlil vše, co je obsahem tohoto písemného informovaného souhlasu, a měla jsem možnost klást mu otázky, na které mi řádně odpověděl.

Prohlašuji, že jsem shora uvedenému poučení plně porozuměla a výslovně souhlasím s provedením vyšetření a následnou terapií.

Souhlasím s nahlížením níže jmenované osoby do mé dokumentace a s uveřejněním výsledků terapie v rámci studie.

Datum:.....

Osoba, která provedla poučení:.....

Podpis osoby, která provedla poučení:.....

Vlastnoruční podpis pacienta /tky:.....

Appendix n°3: Abbreviations

ADL – Activities for daily living

CMC – Carpometacarpal

DIP – Distal Interphalangeal

DVT – Deep Vein Thrombosis

IC – Intercarpal

IP - Interphalangeal

MCP – Metacarpophalangeal

NSAID – Non-Steroidal Anti-Inflammatory Drug

PIR – Post-Isometric Relaxation

PIP – Proximal Interphalangeal

PNF - Proprioceptive Neuromuscular Facilitation

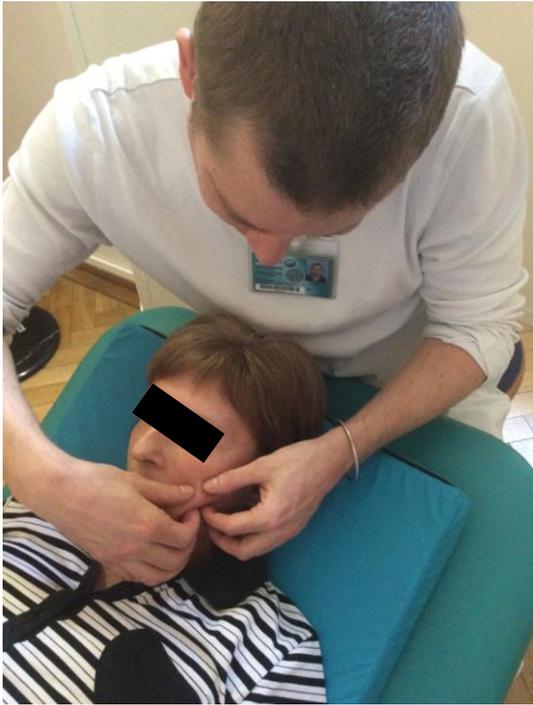
ROM – Range Of Motion

RP - Raynaud's phenomenon

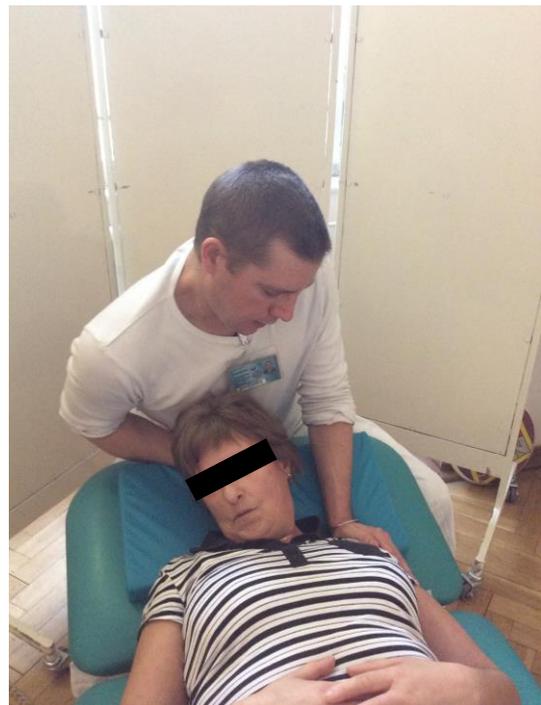
SSc – Systemic Scleroderma

TP – Trigger Point

Appendix n° 5: Photos taken during 7th session



Picture 1 – Kibler fold on the face



Picture 2 – PIR of levator scapula



Picture 3 – Self-stretching of IP joints



Picture 4 – Self-stretching of MCP joints



Picture 5 – Dorsal side of hands with light Raynaud's phenomenon



Picture 6 – Left hand with ulceration on fingertip

Appendix n° 4:

List of tables:

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Table 8: Hand Muscles Strength Measurement with a Dynamometer in Kg

Table 9: Muscle length test according to Janda (Janda, 1983), initial examination

Table 10: Joint Play Examination according to Lewit (Lewit, 1999), initial examination

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Table 20: Joint play examination according to Lewit (Lewit, 1999), final examination

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Figure 2: Raynaud's phenomenon (dxline, 2010)

Figure 3: Microstomia in Scleroderma (Dziedzic, 2010, p.309)

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List of photos:

Photo 1: Kibler fold on the face

Photo 2: PIR of levator scapula

Photo 3: Self-stretching of IP joints

Photo 4: Self-stretching of MCP joints

Photo 5: Dorsal side of the hands with light Raynaud's phenomenon

Photo 6: Left hand with ulceration on fingertip