CHARLES UNIVERSITY

Faculty of Physical Education and Sport

Case Study of Physiotherapeutic Treatment of a Patient with Systemic Sclerosis

Bachelor Thesis

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Prague, April 2024

Declaration

I declare that this thesis is entirely my own. All literary sources and information, used in this thesis are referenced at the end of this document. The practical information practiced was based on knowledge taught at the Faculty of Physical Education and Sports at Charles University. I declare the patient was made aware of all the practical procedures and was in full agreement.

In Prague: April 2024 Author: Eva Bartáková

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Abstract

Author: Eva Bartáková

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Title: Case Study of Physiotherapeutic Treatment of a Patient with Systemic Sclerosis

Objectives: The aim of this bachelor thesis is to study and elaborate the theoretical knowledge about the diagnosis of systemic sclerosis and to elaborate a case report of one patient with this diagnosis.

Methods: All methods applied through the therapy were based on literature and lectures from Charles University Faculty of Physical Education and Sport. The methods mainly consisted of soft tissue techniques, joint mobilizations, Post Isometric Relaxation, Proprioceptive Neuromuscular Facilitation, stretching, and respiratory physiotherapy. The patient additionally received occupational therapy at the hospital.

Results: The patient was independent after 11 therapeutic units. The main improvement was in the increased range of motion of the right shoulder joint and improved soft tissues on the periphery of the upper extremities.

Conclusion: Most of the therapies performed on the patients were effective.

Keywords: systemic sclerosis, Raynaud's phenomenon, rheumatology, physiotherapy

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Název: Kazuistika fyzioterapeutické péče o pacienta s diagnózou systémová sklerodermie

Cíl: Cílem této bakalářské práce je nastudovat a zpracovat teoretické poznatky o diagnóze systémová sklerodermie a zpracovat kazuistiku jednoho pacienta s touto diagnózou.

Metody: Všechny použité metody vycházely z literatury a přednášek z Fakulty tělesné výchovy a sportu Univerzity Karlovy. Především byly použity technik měkkých tkání, kloubní mobilizace, postizometrické relaxace, proprioceptivní neuromuskulární facilitace, strečink a dechová gymnastika. Pacientka byla navíc v Rehabilitačním ústavu v péči ergoterapeuta.

Výsledky: Po 11 terapeutických jednotkách je pacientka samostatná. Hlavní zlepšení spočívalo ve zvýšení rozsahu hybnosti pravého ramenního kloubu a zlepšení měkkých tkání na periferii horních končetin.

Závěr: Většina provedených terapií na pacientovi byla účinná.

Klíčová slova: systémová skleróza, Raynaudův fenomén, revmatologie, fyzioterapie

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List of abbreviations

ADL=activities of daily living

Bilat.=bilaterally

dsSSc= diffuse systemic sclerosis

HRQL =health-related quality of life

MSK =musculoskeletal

RÚ =Revmatologický ústav

SSc = systemic sclerosis

VAS=visual analog scale

1 INTRODUCTION

Systemic sclerosis is a chronic rheumatic disease. The most affected organ is the skin. Skin sclerotisation and contraction causes decreased ROM of joints and limits patient's self-care in daily activities. A serious complication is the involvement of the internal organs by fibrotic changes, which reduce their functionality and significantly endanger the patient's life. There is no clear evidence as to what causes systemic sclerosis and the treatment of this disease remains mainly symptomatic.

The bachelor thesis was written during a month-long bachelor internship at the Institute of Rheumatology in Prague. It is divided into two parts - a theoretical part and a practical part. In the theoretical part, the thesis deals with general information about the diagnosis of systemic scleroderma, its classification, etiology and pathogenesis of the disease. A substantial part of the theory is devoted to the clinical symptoms of the disease and the current treatment modalities, in particular non-pharmacological. In the practical part, a case report of a patient with a diffuse form of systemic scleroderma is presented. It includes the history and initial examination based on which the short-term, long-term plan and treatment proposal was drafted. The individual therapeutic units and final examinations are included. Finally, the effect of the therapy is evaluated.

2 THEORETICAL BACKGROUND

2.1 Definition of systemic sclerosis

Systemic sclerosis (SSc) is a chronic rheumatic disease. The prominent features include progressive fibrosis, vascular wall damage, and disturbances of innate and acquired immune responses with autoantibody production. “Scleroderma” means skin thickening. Thickening of the skin is characteristic for the disease. Internal organs can be affected, such as the lungs, heart, kidneys, musculoskeletal system, and the gastrointestinal tract (Sobolewski et al., 2019).

According to its origin systemic sclerosis is a chronic autoimmune rheumatic disease.

Table No. 1 : General classification of rheumatic disease

Autoimmune	Systemic lupus erythematosus
	Rheumatoid arthritis
	Systemic sclerosis
	Idiopathic inflammatory myopathies
	Antiphospholipid syndrome
	Sjögren's syndrome
	Primary vasculitis
Autoinflammatory	Familial Mediterranean fever
	TRAPS
	Mevalonate kinase deficiencies
	Muckle–Wells syndrome
	Familial cold urticaria
	NOMID syndrome
	NLRP12-associated syndrome
	PAPA syndrome
	Chronic recurrent multifocal osteomyelitis
	Adult-onset Still disease
Metabolic	Osteoporosis
	Parathyroid disorders
	Paget's disease
	Calcium and phosphorus disorders
	Vitamin D deficiency/osteomalacia
	Renal osteodystrophy
	Calcium deposition disease (CPPD, gout, hydroxyapatite)
	Hemochromatosis
	Ochronosis
Degenerative	Osteoarthritis

(Calle & Gómez-Puerta, 2018)

2.2 Classification of Systemic Sclerosis

There are two primary forms of scleroderma: localized and systemic scleroderma, also called systemic sclerosis. Systemic sclerosis is characterized by skin, sub skin, and visceral involvement. Llocalized scleroderma is mainly in the skin and underlying tissues (Careta & Romiti, 2015).

Localized scleroderma, also called morphea, can affect any part of the body. Plaques of varying sizes of thickened, bright, atrophic skin; either hyper or hypopigmented, can be seen(Rodríguez-Salgado & García-Romero, 2023).

This work mainly focuses on the form of systemic scleroderma. Systemic sclerosis can be further classified as limited or diffuse systemic sclerosis.

2.2.1 Diffuse Systemic Sclerosis

Diffuse systemic sclerosis is often linked with high mortality rates due to the rapid progression of the disease and early onset of visceral involvement. Survival for patients with severe organ involvement is markedly reduced, for this reason, patients should be closely monitored. The first signs of the skin being affected are often seen distally, affecting the fingers. This progresses proximally, towards the elbow, the knees and the trunk. Skin becomes itchy and painful during the inflammatory phase of diffuse sclerosis and pigmentary changes may appear. Commonly, tightness of the skin causes contractures, particularly flexion deformities of the fingers, often the elbows and knees (Herrick et al., 2022).

2.2.2 Limited Systemic Sclerosis

Limited systemic sclerosis has a slower progression than the diffuse form. The skin is affected mainly on the extremities (distally from the elbows and knees, face and neck), but is not affected in the trunk as in the diffuse form. The first symptoms usually begin with Raynaud's phenomenon. After this, the fingers swell, and the skin becomes thicker. Internal organs are affected later.

Limited systemic sclerosis is also known as the CREST syndrome. The name is an acronym for typical symptoms of the disease:

- Calcinosis - pathologic calcification of soft tissues
- Raynaud's phenomenon- limited blood flow in the feet or hand due to stress or cold exposure
- Impaired esophageal motility
- Sclerodactyly- thickening of the skin on the fingers due to deposition of mucopolysaccharide, glycoprotein, and collagen
- Telangiectasia- widening of small blood vessels in the skin

(Merlino et al., 2013)

2.3 Diagnosis

The diagnostic criteria for SSc were revised in 2013 by the European League Against Rheumatism (EULAR) and the American College of Rheumatology to include important features of the disease. The final score of this classification is the sum of the maximum point values for each item. Patients with a total score of ≥ 9 meet the criteria of having scleroderma. If skin of the fingers proximal to the metacarpophalangeal joints thickens is sufficient for a patient to be classified as having scleroderma. Other items are considered if this is not present.

Table No. 2: The American College of Rheumatology/European League Against Rheumatism criteria for the classification of systemic sclerosis

Item	**Sub-item(s)**	**Weight/ score**
Skin thickening of the fingers of both hands extending proximal to the metacarpophalangeal joints (sufficient criterion)	–	9
Skin thickening of the fingers (only count the higher score)	Puffy fingers	2
	Sclerodactyly of the fingers (distal to the metacarpophalangeal joints but proximal to the proximal interphalangeal joints)	4
Fingertip lesions *(only count the higher score)*	Digital tip ulcers	2
	Fingertip pitting scars	3
Telangiectasia	–	2
Abnormal nailfold capillaries	–	2
Pulmonary arterial hypertension and/or interstitial lung disease *(maximum score is 2)*	Pulmonary arterial hypertension	2
	Interstitial lung disease	2
Raynaud's phenomenon	–	3
SSc-related autoantibodies (anticentromere, anti–topoisomerase I [anti–Scl-70], anti–RNA polymerase III) *(maximum score is 3)*	Anticentromere	3
	Anti–topoisomerase I	3
	Anti–RNA polymerase III	3

(van den Hoogen et al., 2013)

The criteria do not apply to patients with skin thickening sparing the fingers or to patient who have scleroderma-like disorders (e.g. nephrogenic sclerosing fibrosis, generalized morphea, scleroderma diabeticorum, diabetic cheiroarthropathy) (van den Hoogen et al., 2013).

2.4 Etiology and Pathogenesis of Systemic Sclerosis

2.4.1 Etiology

Althought the etiology of systemic scleroderma remains unclear, findings indicate that it is a multifactorial disease with genetic, epigenetic, and environmental factors (Manetti et al., 2012).

Genetic factors play an important role in the susceptibility to develop systemic sclerosis. Specifically, antigen-class II (HLA-class II) are associated with systemic sclerosis susceptibility. Studies indicated that HLA-DQB1, DQA1, and DRB1 are strongly associated with SSc. HLA-class II affect especially autoantibody markers. Among the most common autoantibodies are anticentromere (ACA), anti-topoisomerase I, and anti-RNA polymerase III (Arnett et al., 2009).

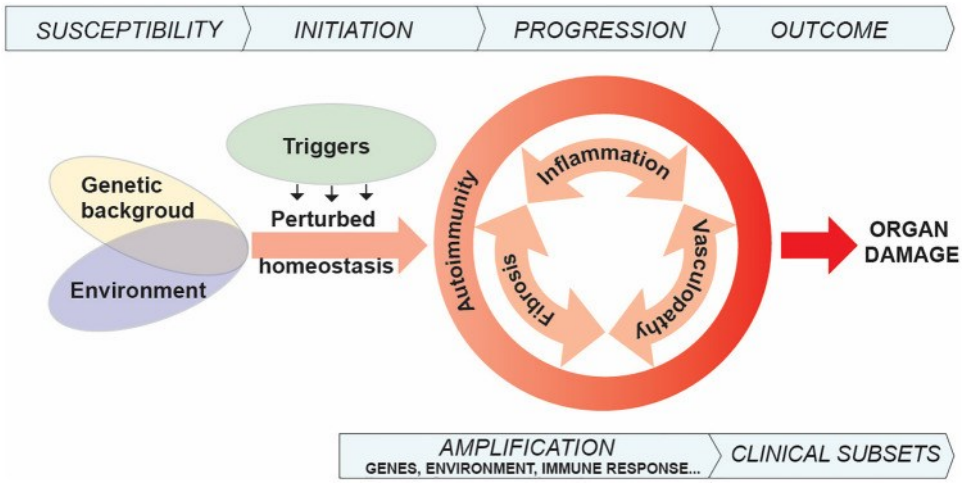
Relatives to individuals with SSc, have an incidence of disease from 1.5 to 1.7%. The probability of developing the disease increases 15–19-fold in siblings and 13–15-fold in first-degree relatives (Pattanaik et al., 2015). In people with a genetic predisposition to SSc environmental factors were also shown to play a role in SSc pathogenesis. Silica or solvents, heavy metals, viral infection, and certain drugs, could lead to the development of SSc (Tsou, 2019). According to a systematic review from 2018 middle age (45-64 years), being female, positive family history, and exposure to silica are risk factors for scleroderma (Abbot et al., 2018).

2.4.2 Pathogenesis

The three most common parts of SSc pathogenesis include immune activation, microvascular dysfunction, and tissue fibrosis. The major cells included in these events are immune cells, endothelial cells, and fibroblasts. The progression of the disease is in sequential order from activation of the immune system, vasculopathy leading to activation of fibroblasts and fibrosis (Truchetet et al., 2021).

The following diagram shows the role of genetic background and environmental factors in triggering the autoimmune response of the body and development of SSc.

Image No. 1: The role of genetic background and environmental factors in triggering the autoimmune response of the body and development of SSc



(Truchetet et al., 2021)

2.5 Clinical Manifestations of Systemic Sclerosis

SSc is a disease that affects multiple systems in the body. Signs and symptoms vary in patients but the most common are vascular manifestations, skin involvement, musculoskeletal manifestations, gastrointestinal manifestations, pulmonary manifestations, cardiac manifestations, and renal manifestations.

2.5.1 Cardiovascular Complications

Autoimmune rheumatic diseases increase the risk of cardiovascular complications. Almost all SSc patients experience vascular changes. Vascular complications occur due to microtrauma of the endothelium leading to an immune response followed by fibroblast proliferation. Several factors including endothelial injury, the imbalance between endothelial vasoconstrictors and vasodilators, and abnormal angiogenesis lead to abnormal reactivity of the vessels and in the end fibrosis. Fibrosis affects not only the cardiovascular system but also other organs (Mani et al., 2019).

Many of SSc manifestations are connected due to their underlying vasculopathy. Amongst them are Raynaud's phenomenon, digital ulceration, scleroderma renal crisis, and pulmonary arterial hypertension.

2.5.1.1 Cardiac complications

Cardiac diseases linked with SSc are myocarditis, myocardial fibrosis, conduction system abnormalities, coronary artery disease, pericardial disease, and valvular disease. A reversible spasm of capillaries leads to the deterioration of myocardial microcirculation, inflammation, and myocardial fibrosis. Fibrosis can affect both the right and the left ventricle and influences both systole and diastole. Atherosclerosis and coronary artery disease can also occur (Nowakowska-Płaza et al., 2022).

2.5.1.2 Raynaud's phenomenon

Raynaud's phenomenon is one of the earliest manifestations of SSc. 90 to 98% of SSc patients experience this symptom (Cappelli & Wigley, 2015). Raynaud's phenomenon is caused by vasospasm of the peripheral microvasculature, usually in the digits. It occurs as a response to a cold environment, emotional stress, or from other physical or medication exposures. The response to this exposure in Raynaud's phenomenon is in three phases: initial white or pallor (cessation of blood flow), then blue or cyanosis (desaturation of blood), followed by red or erythema (vasodilatation) (Pauling et al., 2019). Raynaud's phenomenon can be very painful. Paresthesia, numbness, and impaired hand function commonly occur during episodes.

There are two types of Raynaud's phenomenon, primary and secondary. If there is no underlying disease we speak of primary Raynaud's phenomenon. Specifically, secondary Raynaud's phenomenon is associated with autoimmune diseases, such as SSc (Nawaz et al., 2022).

2.5.1.3 Digital ulcerations

Digital ulcerations reduce the quality of life of patients with SSc significantly, limiting their ability to perform activities of daily living. Ulcers affect around 50% of patients with SSc and usually appear on the tips of fingers, phalangeal joints, elbows, ankles, and toes. Microtrauma can lead to ulcerations on body prominences which are usually difficult to heal. Ulcers can be very painful and heal slowly. Due to digital ulcerations being very prone to infections, there is an increased need for antibiotic treatment of SSc patients (Strange & Nash, 2009).

2.5.2 Renal Complications

SSc patients are susceptible to vasculopathy, fibrosis, and autoimmunity which leads to damage in the kidneys (Cole et al., 2022). Scleroderma renal crisis used to be the primary cause of death in SSc. Whilst the outcome has improved after the use of angiotensin-converting enzymes started it remains a life-threatening complication of SSc. One-year outcomes indicate 30% mortality and 25% of patients remain dependent on dialysis (Hudson et al., 2021).

2.5.3 Respiratory Complications

Lung disease is the leading cause of SSc patient's death. 70 to 90 % of SSc individuals have pulmonary complications. Most deaths are due to pulmonary hypertension and interstitial lung disease as they lead to issues in other areas of the respiratory system, including the parenchyma, muscle, and vasculature. This is why the respiratory system should be screened in SSc patients (Syed Gaggatur et al., 2021;Denton et al., 2018;Doskaliuk et al., 2020).

The most common clinical manifestations are dyspnoea (at first after exertion and eventually at rest), non-productive cough, and severe fatigue. Upon auscultation crackling sounds are heard during breathing (Perelas et al., 2020).

Severe respiratory issues occur due to skin fibrosis and contraction or muscle shortening in all extremities and the thorax. Stiffness of the chest wall and weakness of respiratory muscles cause shortness of breath, non-productive cough, and respiratory failure. The main causes of death are upper airway obstruction, pulmonary

hypertension, hypoxemia, and acute respiratory failure. Respiratory complications affect exercise capacity and physical function (Leelarungrayub et al., 2016).

2.5.4 Gastrointestinal tract manifestations

SSc can affect any part of the gastrointestinal tract, from the oral cavity to the anorectum. Approximately 98.9% of SSc patients suffer from GI symptoms such as meteorism, dysmotility of the esophagus, heartburn, dysphagia, nausea, vomiting, diarrhea, and constipation. The quality of life of patients is negatively affected by these manifestations (Nassar et al., 2022).

Soft tissue fibrosis in the face causes wrinkling and mouth width (microcheilia) and opening (microstomia) is reduced which interferes with eating, speaking, oral hygiene, and dental treatment (Maddali-Bongi et al., 2010). The feeling of dry mouth (xerostomia) is experienced in up to 80% of patients with SSc. Oral dryness favors tooth decay, atrophy, and mouth infections. (Zimmermann et al., 2022) Xerostomia is one of the manifestations of Sjögren's syndrome along with dryness of the eyes (sicca syndrome). Patients with SSc experience secondary Sjögren's syndrome (Balint et al., 2023).

From upper gastrointestinal tract involvement, patients may experience laryngeal-esophageal or gastroesophageal reflux disease, lower esophageal sphincter dysfunction, and esophageal dysmotility. Constipation and diarrhea are major symptoms of lower gastrointestinal tract involvement (Volkmann & McMahan, 2022).

2.5.5 Skin Involvement

Skin involvement is a characteristic manifestation of SSc, except in approximately 10% of SSc cases, which we refer to as SSc sine scleroderma. Skin thickening is caused by increased collagen, intercellular matrix formation in the dermis and microvascular injury and inflammation are most probably the causes of edema. One of the earliest indications of SSc is edema of the fingers and Raynaud’s phenomenon (Czirjak et al., 2008).

The skin thickening of the fingers of both hands extending proximal to the metacarpophalangeal joints is sufficient criteria for the diagnosis of systemic sclerosis.

At present the most used technique for measuring dermal skin thickness is the modified Rodnan skin score. The skin score is evaluated by palpating 17 different anatomical areas (Ferreli et al., 2017).

The skin of affected digits, extremities, and in some cases the trunk eventually thickens. Thickening of the skin always begins in the fingers (sclerodactyly-thickening and tightness of the skin of the fingers or toes), extending proximally. Flexion contractures develop as a result and digital ulcers appear. Flexion deformity of the fingers follows with the loss of flexion at the metacarpophalangeal joints, the loss of extension of the proximal interphalangeal joints, and the loss of thumb abduction. Furthermore, the distal interphalangeal joint may become fixed in midrange flexion. These changes cause a claw hand deformity of metacarpophalangeal extension, proximal interphalangeal joint flexion, and thumb adduction. ADLs are limited due to grasp impairments and for this reason, it is crucial to prevent the formation of deformities of the hands. Other symptoms include telangiectasias (small, widened blood vessels on the skin), calcinosis cutis, and depigmentation of the skin (Zhu et al., 2021; Mugii et al., 2018).

Soft tissue is also affected in the face. The face loses its expressivity and appears mask-like. The skin hardens, mimic folds disappear, lips become thinner, the nose sharper, and facial hair falls. Deep wrinkles appear on the upper and lower lip (Ferreli et al., 2017).

Changes in the skin of the hands and face impact interpersonal relationships, self-esteem, and psychosocial functions. Consequently, skin impairment is one of the components leading to image discontent of individuals affected with SSc (Mugii et al., 2018).

2.5.6 Musculoskeletal Manifestations

Although skin changes are a hallmark of SSc musculoskeletal manifestations are common. Several musculoskeletal pain syndromes can be identified in SSc:

- Tendonitis
- Rheumatoid arthritis
- Polyarthritis
- Myopathies
- Other MSK syndromes

Tendonitis can occur in many locations in SSc. A unique form is tendon friction rubs. Fibrosis appears on the surface of tendon sheaths and around the tendon. Most commonly on the triceps, extensor and flexor tendons of the wrist, patellar tendons, and the posterior and anterior tendons of the ankles. Subscapular, lateral trochanter, and paraspinal regions can also be affected (Clements, 2016).

Some patients may complain about pain but are not aware of the tendon friction rubs. If the pain is not restricted to the nearby joints but goes along the tendon it can also be a sign of tenosynovitis.

Systemic sclerosis also often overlaps with rheumatoid arthritis and polyarthritis. Joint involvemt can be present. Common signs include arthralgia and joint contractures. Arthritis can be also present in SSc but less frequently. Synovitis and periarticular calcification are also associated with SSc (Lóránd et al., 2014).

Muscle involvement in SSc affects both smooth muscle (leading to manifestations for example in the gastrointestinal tract and vessels), and striated muscle. In skeletal muscles non-progressive non-inflammatory myopathy may appear due to digestive disturbance, malnutrition, a sedentary lifestyle, or contractures of fibrotic skin. SSc can also overlap with inflammatory myopathy. SSc patients often have significant muscle weakness in the upper and lower limbs and reduced range of motion in the shoulders (Bratoiu et al., 2022). Furthermore, clinical assessment of joint range of motion might be challenging because of the skin fibrosis or edema (Volkmann et al., 2023).

Other musculoskeletal symptoms involve carpal tunnel syndrome, trochanteric and gluteus medius bursitis, anserine bursitis, olecranon bursitis, epicondylitis (lateral more than medial), and rotator cuff tendonitis (Clements, 2016).

Table No. 3 :Common Musculoskeletal Manifestations of SSc patients

Site	**Manifestations**
Joints	Ranges from symmetrical polyarthralgia to polyarthritis (Similar distribution as in rheumatoid arthritis). Radiographic findings include juxta-articular osteoporosis, erosions, joint space narrowing, pencil in cup deformity and/or subchondral sclerosis.
Bone	Distal phalangeal resorption in hands or feet, osteoporosis, resorption of bone (carpal bones, distal radius and ulna, clavicles, ribs, spine, mandible) and thickening of periodontal membrane.
Tendons	Inflammatory proliferative tenosynovitis, tendon rupture. Fibrous deposits within tendon sheaths → tendon friction rubs in wrists, elbows, knees and shoulders are associated with more severe disease. Contracture of flexor tendons.
Muscle	Myopathy is present to some degree in most patients. In dsSSc, proximal muscle weakness in upper and lower extremities can have overlap with polymyositis or have piecemeal infarction from SSc vasculopathy, fibrous myopathy or ill defined myopathies. Skeletal myopathy is often related to myocardial disease in SSc.

(Postlethwaite et al., 2010)

Patients with SSc are limited in their activities due to skin retraction, stiffness, pain, dysfunction in joints, bursas, and tendons. This limits SSc patient's activity, life expectancy and productivity (Frade et al., 2022).

2.6 Treatment of Systemic Sclerosis

SSc is a multisystem connective tissue disease with varying manifestations between individual patients. For this reason, it is crucial to tailor the treatment to the goal of the patient and requires a multifaceted approach (Pope et al., 2023). A multidisciplinary team of physicians, rheumatologists, pulmonologists, cardiologists, and dermatologists is vital. Impaired skin, musculoskeletal system, and respiratory system require physiotherapy to be carried out by a team of physicians, physiotherapists, podiatrists, and occupational therapists (Mugii et al., 2018).

2.6.1 Pharmacotherapy

Pharmacotherapy focuses on treating the symptoms of SSc. Amongst the most prominent are Raynaud's phenomenon, digital ulcers, scleroderma renal crisis, and pulmonary arterial hypertension. The following table describes medication for the most common SSc manifestations (Postlethwaite et al., 2010).

Table No. 4: Most Common SSc Manifestation and Pharmacotherapy

Disease Manifestation	**Pharmacotherapy**
Raynaud's Phenomenon	Calcium Channel Blockers, Sildenafil and related phosphodiesterase type-5 inhibitors, intravenous iloprost
Digital Ulcers	Bosentan, statins, intravenous iloprost, aspirin
Scleroderma Renal Crisis	ACE inhibitors plus calcium channel blockers and addition of beta adrenergic blockers or nitrate infusions as warranted to control hypertension
Esophageal Dysmotility	Proton pump inhibitors
Delayed Gastric Emptying	Pro-motility drugs (metoclopramide domperidone, erythromycin)
Small Bowel Dysmotility (Blind Loops)	Octerotide, Antibiotics
Colinic Dysmotility	Erythromycin, prucalopride, octreotide
Coronary Artery Vasospasm	Calcium channel blockers, ACE inhibitors, dipyridamole
Progressive Pulmonary Fibrosis	Cyclophosphamide, ? Mycophenolate mofetil
Pulmonary Arterial Hypertension	Prostacyclin analogs (epoprostenol, treprostinil, iloprost), bosentan, sitaxsentan, ambrisetan, phosphodresterase type-5 inhibitors
Arthritis	DMARDs (hydroxychloroguine, methotrexate, sulfasalazine)
Osteoporosis	Calcium, Vitamin D, intravenous bisphosphonates

(Postlethwaite et al., 2010)

2.6.2 Physiotherapy Methods and Procedures

Apart from medical treatment, physiotherapy is recommended to SSc patients. Studies indicated rehabilitation was effective in decreasing local and systemic disabilities, leading to an improved quality of life. Rehabilitation and occupational therapy are important for systemic sclerosis patients (Liem et al., 2019;Mugii et al., 2018).

SSc patients are affected by fibrosis which restricts physical activities. Due to the involvement of skin, subcutaneous tissues, and the musculoskeletal system, the hand and face functions, breathing and postural maintenance is impaired. Rehabilitation of SSc can be divided into two attitudes: local and global rehabilitation. Local rehabilitation is focused on hand and mouth function and global rehabilitation consists of aerobic and resistance exercises. (Mugii et al., 2018)

The commonly used physiotherapeutic interventions include connective tissue massage, joint manipulation, manual lymphatic drainage, Kabat's method, paraffin application, laser therapy, transcutaneous electric nerve stimulation (TENS), ultrasound therapy, soft tissue techniques, massage, various exercises, such as hand and mouth exercises, range of motion exercises, kinesiotherapy, recreational exercises, resistance, and aerobic training. Physical therapy and exercise are crucial to sustain range of motion and to meet the demands of activities of daily living. SSc patients may benefit from structured exercise. Muscle strength and function, cardiovascular function, and fatigue may be improved by exercise in SSc patients (Frade et al., 2022).

De Oliviera et al. reviewed the effectiveness of aerobic and aerobic with resistance exercise. A majority of the studies involved patients with and without pulmonary involvement. Exercise tolerance, aerobic capacity, walking distance, muscle strength, and muscle function as well as health-related quality of life (HRQL) were improved after participation in the programs. SSc patients both with and without pulmonary involvement benefited from aerobic exercises. SSc patients without pulmonary involvement could be as physically active as the general population and patients with pulmonary involvement should perform exercise in moderate intensity (de Oliveira et al., 2017).

Alexanderson et al. performed a 6-week non-interventional baseline period program an 8-week exercise intervention period program with four SSc patients. Patients completed aerobic exercise and muscular endurance training three times a week. Oxygen uptake was improved in two patients and muscle endurance improved in three patients. This exercise program showed an overall positive outcome with an increased aerobic capacity and muscle endurance of participants with SSc (Alexanderson et al., 2014).

According to Pettersson et al., patients experienced benefits after exercise including improved blood circulation, particularly in hands and feet, prolonged core-warming, breathing, fatigue, pain, sleep, vitality, and musculoskeletal function. Aerobic and muscle-strengthening exercises seem to improve cardiovascular function as well as lung mechanics.

Pulmonary rehabilitation is effective in improving breathing, exercise tolerance, fatigue, and cough. Techniques such as strengthening of the diaphragm improve respiratory capacity as well as core strength and proper postural alignment. Other practices that combine movement and breath are beneficial. They include yoga, dance, Tai Ch'I, or singing (Pettersson et al., 2021).

Local rehabilitation mainly focuses on the function of the mouth and hands.

A 2021 randomized controlled trial by Roque et al. evaluated the how effective was of Maitland's joint mobilization and therapeutic exercises on hand function in SSc individuals. 12 of the 24 patients were part of the physical therapy group and 12 patients were the control group. The trial lasted 12 weeks. The physical therapy group performed joint mobilization protocol and therapeutic exercises, applied to the hands. Twice a week for approximately an hour physical therapy took place. This program reduced pain in the hands and wrists, improved the functionality of the hands and the range of motion (Santos Cardozo Roque et al., 2021).

Waszczykowski et al. performed a one-year follow-up study with 51 participants. 27 patients underwent a 4-week supervised rehabilitation program. The control group exercised alone at home. Improvements in hand function were obtained after the rehabilitation program was completed and after 3 and 6 months of follow-up. Nevertheless, between 6 and 12 months after the conclusion of the program, gradual reductions in hand function and the recurrence of pain developed occasionally. In comparison, in the control group, the improvements only lasted until the one-month follow-up (Waszczykowski et al., 2021).

Gregory et al. carried out a randomized controlled trial aimed at comparing the effects of daily hand exercises with or without daily home wax baths on patients with SSc. Participants were divided into wax bath versus no wax bath groups. Every participant continued performing hand exercises. This trial concluded that regular wax

bath treatment had no added effect in comparison to only daily home exercises (Gregory et al., 2019).

A 2023 study by Scaturro et al. looked into the treatment of ischemic digital ulcers by immersion ultrasound therapy in combination with manual therapy. The treatment was 20 sessions daily, 5 days a week, for 4 weeks. SSc patients in the treatment group also received ultrasound. The ultrasound was used with a frequency of 1 MHz, an intensity of 1 W/cm^2, a duty cycle of 60%, for 15 min per session. The patient's hands were immersed in a container with 4 L of water at a temperature of 37–37.5 °C. The treatment group showed improvement in the functional capacity of the hand, pain, and Pressure Sore Status Tool scale. In the standard group, there was an improvement only for the functional capacity of the hand. The finding included an improvement in pain and greater healing of ulcers than in the treatment group (Scaturro et al., 2023).

In comparison, Saito et al. used extracorporeal shockwave therapy to treat digital skin ulcers. An electromagnetic lithotripter was used for the application. 1 session per week for 9 weeks was applied and there were 9 participants. After one treatment digital ulcers seemed to be healing. The number of ulcers decreased from 5.4 to 1.1 in 9 weeks. A large number of ulcers disappeared and became smaller, namely, the mean size decreased from 10.9 mm to 2.5 mm at 20 weeks (Saito et al., 2016).

Fibrosis in the orofacial area causes problems in many areas of life such as difficulty eating and speaking.

A 2021 randomized controlled trial with 60 participants evaluated the effect of a home-based orofacial exercise on the oral aperture of SSc patients. The first group was prescribed an exercise regime and received oral hygiene care advice for the first month, followed by oral hygiene care advice without exercise for the next four weeks. The control group received oral hygiene care advice for the first one month, followed by the exercise regimen twice a day along with oral hygiene care advice for the next four weeks. The oral aperture of the first group increased significantly, while the control group showed no statistically significant increase (Cuzdan et al., 2021).

According to Maddali Bongi et al., personalized exercises for the orofacial muscles, cervical muscles, and temporomandibular joint may induce better therapeutic

effects than a randomly assigned protocol. 22 patients in the trial were assigned to a random protocol with home exercises for temporomandibular joint, mimic, masticatory, and cervical spine muscles. 25 patients were randomized to protocol 2 with home exercises and combined physiotherapeutic procedures performed by a physiotherapist (intra- and extra-oral manipulations of TMJ, exercises of neuromuscular coordination for the temporomandibular joint, stretching, and mobilization of the cervical region, tongue muscles, hyoid, and tracheal muscles, Kabat technique) The protocol spanned 12 weeks (Maddali Bongi et al., 2021).

2.6.3 Physical Therapy

Physical therapy is nowadays an essential part of rehabilitation and physiotherapy. The principle consists of the use of almost all physical energies for application to the tissues of the human body. The action of these agents can directly affect the tissue biochemical processes in the body, indirectly, through the nervous system or hormones to influence afferent signals or act as a placebo. Especially positive thermotherapy seems to have a beneficial effect on microcirculation, myorelaxation, and analgesia (Poděbradský & Poděbradská, 2009).

Heat leads to the improvement of the trophic function of cells and tissues, decreases the viscosity of connective tissue, and increases its elasticity and distensibility. This is beneficial for systemic autoimmune diseases such as SSc. Before individual physiotherapy, it is advantageous to use prewarming procedures. That is, before mobilizations, manipulations. and soft tissue techniques (Jandová, 2009).

A frequently used method in physiotherapy of rheumatic diseases is paraffin treatment. At 52–62 °C paraffin solidifies and gives latent heat (Špiritović & Tomčík, 2017). In a 2004 trial, SSc patients dipped their hands five to six times into a 50°C warm paraffin bath. Insertion of the hand in a plastic bag and then mittens for 10 minutes was performed (Sandqvist et al., 2004).

Phototherapy uses specific wavelengths of the electromagnetic spectrum to disrupt the dysfunctional and pathologic tissue that has developed in some patients with skin disease. Ultraviolet A1 (UVA1) radiation has been studied the most in the context of SSc. 20–50 J/cm^2 of UVA1 therapy 3–4 times a week for 30 treatments is

recommended. This treatment seems to reduce sclerosis. However, the anti-sclerotic effects of an UVA1 treatment typically last less than 1 week (Hassani & Feldman, 2016).

Lasers can be also used to treat SSc symptoms. Pulsed dye laser at wavelengths 585 and 595 nm is used primarily for the treatment of cutaneous vascular lesions, such as port-wine stains, hemangiomas, spider angiomas, and telangiectasias. Treatment can also improve pigmentation. Authors suggest the heat of the laser causes collagen bundles to shrink and the density of the collagen decreases (Szczepanik-Kułak et al., 2021).

2.6.4 Spa Care

In Czechia, SSc can be also treated in spas. Sulfurous mineral waters type B have vasodilating, antiseptic, antisclerotic, and keratolytic effects and have a significant therapeutic effect in all rheumatic diseases. In Czechia, this type of mineral waters are found in Velké Losiny and in Hungary Hevíz and Harkányi are popular.

Iodine-bromine waters are also used for external balneation. Iodine affects the cardiovascular system and causes mild vasodilatation. It also contributes to the increase of elasticity and distensibility of the connective tissue and at the same time to the increase of its mechanical resistance. Combining the vasodilating effect on the connective tissue and thermotherapy enables the resorption of edema and infiltrates and relaxes the shortened soft tissues (Jandová, 2009).

2.6.5 Occupational Therapy

Work and functional disability is very high in SSc patients. Occupational therapy can help with the structural and functional changes of the hands and face in SSc. Occupational therapy typically addresses limitations in function. Along with physical therapy occupational therapy aims to preserve mobility of SSc patients and aid them in participation in ADL (Frech et al., 2023).

2.6.6 Regimen Measures

Of the regime measures, minimizing exposure to influences that induce or exacerbate attacks is crucial. The effort should be to reduce the frequency of attacks,

their intensity, and duration. The patient should be advised to protect both the periphery (warm gloves, electric gloves and boots, gel heaters, caps, etc.) and the core of the body and to not be exposed to sudden changes in temperature and wind chill. Movement is important to increase flow through the peripheral circulation (hand rubbing, circular arm movements, immersion in warm water) and reducing stress and anxiety. It is necessary to quit smoking and avoid vibrations and substances with a vasoconstrictive effect (serotonin agonists, alpha-sympathomimetics, beta-blockers, caffeine, weight loss drugs, and addictive substances) (Bečvář et al., 2017).

2.6.7 Prognosis

SSc prognosis depends mainly on the skin lesion size. The size of skin lesion correlates with the severity of the cardiovascular, pulmonary, and renal manifestations (Meyer, 2006).

Cardiovascular, renal, and pulmonary involvement are amongst the main causes of death of individuals with SSc. The primary cause are pulmonary manifestations (Rubio-Rivas et al., 2014). According to a French cohort study survival rates of SSc at 1, 3, 5, and 10 years from diagnosis were 98.0%, 92.5%, 85.9%, and 71.7% respectively (Pokeerbux et al., 2019).

3 PRACTICAL PART

3.1 Methodology

The practical part of this thesis was carried out during compulsory winter practice, which took place between 8.1.2024 and 2.2.2024 in Revmatologický ústav in Prague. An adult patient with the diagnosis of systemic scleroderma was familiarized with the course, the aim of the bachelor thesis, and the wording of the informed consent, which she signed. The topic of the bachelor thesis and the wording of the informed consent werre approved by the Ethics Committee of UK FTVS Head of the Department on the 10.1.2024 on the basis of fulfilled conditions given by the Ethics Commitee of FTVS UK. The original Ethics committee approval with the Informed consent form are in Appendix 1 and 2 of the thesis.

There were a total of 12 therapy units carried between 10.1 and 25.1. During the first unit, the medical anamnesis was taken and an initial kinesiological examination was performed. In the last unit, a final kinesiological examination was done. Most of the therapy sessions lasted 60 minutes. A few therapies were shortened to 30 minutes due to medical examinations.

The patient was admitted to the inpatient ward at Revmatologický ústav and attended daily therapy in the inpatient gym. The methods used for therapy were those with which we were familiar during our undergraduate studies: soft tissue techniques, soft ball techniques, PIR techniques, mobilization and manipulation techniques according to Lewit, passive stretching, and proprioceptive neuromuscular facilitation (PNF) according to Kabat. A tape measure was used to examine anthropometry and spinal distances. A neurological hammer was used to examine deep tendon reflexes. To examine the ranges of motion of the upper and lower limbs and trunk a two-arm goniometer and SFTR method were used. Special systemic scleroderma questionnaires were used to assess the function of the patient's affected areas. In therapy, I used a foam ball to relax the soft tissues and a roller to support the head or lower limbs.

3.2 Anamnesis

Examined person: D.Š.

Gender: female

Year of birth, age: 1956, 67 years

Diagnosis, other diagnosis: M.34 systemic sclerosis- limited form with interstitial lung involvement

Status Praesens-objective: Alert at rest, without problems, responsive, clear speech

Dominant limb: left-handed

Glasses: yes- multifocal

Height: 158 cm

Weight: 56 kg

BMI, somatotype: 22.4, ectomorph

Status Praesens-subjective:

Chief complaint/problem: The patient is momentarily most focused on the pain of her digits on the upper extremities, especially II. and III on her LUE and II on the RUE, accentuated Raynaud's syndrome and painful digital ulcerations. The pain is usually dull, at the moment a 5 on the VAS. She has some issues with buttoning her clothes and drinking with a straw. She is not feeling stiffening of the skin, does not have problems with swallowing, no abdominal pain, diarrhea, or disuria. She cannot fully open her mouth and has some problems with microstomia such as during the visit to the dentist. The patient does not feel breathlessness.

Medical history: in 1998 the patient experienced tendonitis on her right hand after a strenuous bike ride. It was treated with fixation for a fortnight and then a sensory impairment appeared. She was treated with rehabilitation without any effect. In 2000 she had a carpal tunnel operation I dx. This was also without effect, the sensory impairment prevailed, the movement of the hand was uncoordinated, the hand was painful and had edema. No other joints were painful. For these issues the patient was sent for examination to ÚVN and then Revmatologický ústav. At ÚVN she had a

capillaroscopy supporting the diagnosis of systemic sclerosis and direct immunofluorescence. With these findings, she was sent for a diagnostic stay at RÚ. Systemic scleroderma was confirmed in 2005. She was treated in ambulance for systemic scleroderma with intersticial lung involvement in hospital, and in the Center for treatment of lung hypertension. She is currently admitted to RÚ for re- examination.

PA (personal anamnesis):

Systemic scleroderma

Mitral regurgitation 2+

Ostheorarthrosis of peripheral joints of LE, beginning of patellofemoral arthrosis bilaterally

Osteopenia

Status post dysplasia of right hip joint

St. P. borreliosis in 1997, medicated with antibiotics

- injuries: 2022- fracture of the right clavicle and right calcaneus after fall- conservative treatment
- past medical and surgical history: strabismus operation at age 3, carpal tunnel operation of RUE in 2000, 2022 endovenous laser varicose vein surgery

Previous physiotherapy: 2001 rehabilitation of carpal tunnel on her RUE with positive effect, summer of 2023 rehabilitation of C spine without any effect

MA (medications, pharmacological anamnesis): Pentomer Retard 400 mg 1-0-1, Loseprazol 20 mg 1-0-0, Vigantol 21 drops 1x a week, Calcichew 500 mg/ 400 IU 0-0-1

AA (allergic anamnesis): no allergies

FA (family anamnesis): no family records of rheumatic diseases

GA (gynecological anamnesis): menarche at age 13, the patient has 2 children, birth was

without complication, menopause at the age of 55

OA (occupation anamnesis, vocation): she is working an office job

SA (social anamnesis): the patient is married living with her husband and dog in a house

Abusus: she exceptionally drinks alcohol, non-smoker

Diet: no specific diet

Sport, regular physical activity (past; actual): past- cycling and skiing, actual- twice weekly circuit training for women

Hobbies: cycling, skiing, enjoys reading

Aids: the patient uses a neck brace for traveling

Functional anamnesis/history: The patient is independent, able to achieve all ADLs, sleep is regular but the patient is able to sleep less than before and wakes up earlier. She has slight difficulties with fine motor movement with her right hand such as buttoning up clothes, opening jars, and locking the door. She started using her left hand for many of these activities including using a computer mouse with her left hand.. Fully opening her mouth is difficult and this causes some issues at the dentist's, she also has difficulty drinking with a large straw.

Indication of rehabilitation:

2001 Complex kinesiological examination

21003 Kinesiological examination

21221 Individual kinesiotherapy I

21225 Individual kinesiotherapy II

21219 Supervised therapeutic physical education on an apparatus

21413 Soft tissue techniques

21415 Mobilization of the spine and peripheral joints

3.3 Initial Kinesiological Examination

Date: 10.1.2024

Status praesens, subjective: The patient is arriving alone, without any aids, she is oriented in time and space and cooperates well, there is a cannula distally in the dorsum of the left forearm

Status praesens, objective: the patient is experiencing slight pain in her digits, she arrived without any aids and used the lift to arrive to therapy.

Goal of therapy unit: Initial kinesiological examination

Examination of standing:

Posterior view

- The base of support: base of support is relatively narrow, feet are less than hip-width apart, feet are slightly externally rotated
- Shape and contours of the heels: right heel is more rounded, left more square
- Shape and position of the ankle joints: left malleolus medialis is more prominent
- Knees: slight external rotation and valgosity of knees
- Symmetry of thoracobrachial triangles: the left seems to be smaller
- Position of the pelvis: right SIPS is higher than the left, pelvic tilt to the left, crista iliaca is higher on the right
- Paravertebral muscles: paravertebral muscle in the left half of the body seem more in hypertone than on the right
- Position of the scapula: angulus infraspinatus as well as the margo medialis of the right scapula is more prominent- scapula alata
- Position of the spine: there are clear signs of scoliosis—dextroconcavity of L spine
- Position of the shoulder girdle: right shoulder is more elevated and protracted

- Position and contour of the nuchal muscles: middle third of right trapezius is about 1 cm more prominent cranially
- Position of the head: slightly tilted to the left

Lateral view

Left

- Weight distribution: left foot more weight distributed on the forefoot
- Weight distribution: left foot more to the back
- Position of the knee joints: straight, slightly hyperextended
- Position and curvature of the L, L/Th and Th spine: slight flattening in the thoracic spine
- Shape of the abdominal muscles : slight prominence of the abdomen
- Position of the shoulder girdle: left shoulder is protracted
- Position and curvature of the Th/C and C spine :thoracic spine is slightly flattened, there is protraction of the head

Right

- Weight distribution: left foot more to the back
- Position of the knee joints: straight, slightly hyperextendeed
- Position and curvature of the L, L/Th and Th spine: slight flattening in the thoracic spine
- Shape of the abdominal muscles : slight prominence of the abdomen
- Position of the shoulder girdle: right shoulder is protracted

Anterior view

- The base of support: feet are less than hip-width apart
- The position of the feet, examination of the arches: left foot is stepped slightly more to the front than the left, vertical arches are collapsed
- Weight distribution: weight more on the heels
- The position and shape of the toes: both on the right and left side the big toes are extended, slight valgosity of toes
- Shape and position of ankle joints: the right malleolus medialis is more prominent
- Position of the pelvis: tilt to the left, the right SIAS is higher than the left
- Symmetry of the muscle tone of abdominal muscles: prominent abdomen
- Position of the navel (belly muscle symmetry): slightly to the right
- Symmetry of thoracobrachial triangles - both thoracobrachial triangles are very small
- Position of the collarbones and supraclavicular holes: prominence of the right clavicle
- Position of the shoulder girdle: right shoulder appears protracted and elevated
- Position of the upper limbs: right upper extremity is more internally rotated than the left
- position of the head : slight tilt to the left

Dynamic spine evaluation

Flexion

The patient is able to perform movement but did not reach the tips of her fingers to the ground. However, less curvature is seen in the cervical spine and the thoracic spine. The movement showed an increased prominence in the left lumbar spine during flexion keeping in line with the scoliotic curve.

Extension

The range of movement is very small and the movement is not fluent.

Lateroflexion

The vertebrae, especially in the thoracic region spread only mildly. Pelvic synkinesis is seen during lateral flexion to both sides. During the movement there is a pause when the pelvic shift appears and in the area of the scoliotic curve. The range of motion to the left was larger.

Modification of standing:

Standing on tiptoes-patient is able to maintain some balance on tiptoes with slight difficulty

Standing on heels- she is able to balance

Examination of stability

Single leg stance- does not manage on either side

Trendelenburg- does not manage on either side

Romberg I is negative

Romberg II is negative

Romberg III is positive

Gait examination

Gait is without any assistive device, cadence and velocity is rhythmical. The base of support is narrow.

Stride length: right foot longer step, left shorter

Movement of the foot: first heel strike then flat foot then loading response the heel-off, however toe off is limited in both feet

Movement and position of hip and knee: weight is applied mostly on left leg with initial swing, the right leg is externally rotated

Position and movement of pelvis: sinister tilt

Position and movements of the trunk: shift to the left

Position of spine: flattened thoracic and lumbar spine, slight stiffness in movement

Activity of back muscles: active

Position of shoulder: right protracted forward, minimal arm swing of both hands

Position and movement of the head: tilt to the left

Movement of the upper extremity: the movement is limited on both sides

Stability of walking: relatively stable

Walk with eyes closed –unstable

Walking on tiptoes: unstable

Walking in squat: unstable

Palpation of pelvis

LSIAS lower than the right, LSIAP lower than the right

Pelvic tilt to the left.

Breathing examination

Aspection:

Trunk position: upright, slight scoliotic curve in the lumbar spine

Chest shape: symmetrical

RR, regularity, ratio of inspiration and expiration: 17 breaths/min, eupnea

Location of breath movements: mainly lower thoracic, less movement in abdomen

Breathing wave: normal, during inspiration the abdomen rises and is followed by a lateral costal expansion and then chest rising.

Accessory respiratory muscles: There is no shortness of breath, therefore no difficulty breathing and no need to use accessory respiratory muscles.

Basic movement stereoytypes according to Janda

Hip extension: The same stereotype has been found on both sides. Firstly, the homolateral lumbar extensors initiate the movement even before motion begins, which is then followed by a contraction of the contralateral lumbar extensors. Once this happens, the superior part of the homolateral extensors continues the activation (L-Th region). The same occurs on the contralateral side, where the activation is also visible in the scapular area, reaching almost to the shoulder girdle. Only once this happens, the hip actually begins to extend with initiation of homolateral hamstring activation, causing knee flexion and subsequent bilateral activation of primarily the m. gluteus maximus.

Hip abduction: Bilateral tensor mechanism is present as the hip begins to flex during greater range of motion. In the final range of motion, the lower extremity also begins to externally rotate as the toe starts to point more outwards (this happens bilaterally but is more visible in the right LE).

Shoulder abduction: The right shoulder initiates abduction with the elevation of the whole shoulder girdle. There is a visible activation of the right m. trapezius before

the deltoideus muscle activates. The right scapula is winging. The left shoulder, surprisingly, showed physiological activation of muscles, starting with m. deltoideus, m. trapezius and then the lower fixators of the scapula

Trunk flexion: The patient is unable to achieve a full curl-up. The spine does not curve much in the cervical and upper thoracic region but shows some curvature in the mid-thoracic section.

Neck flexion: The movement is initiated by the protraction of the head rather than by clear flexion but the curvature of the cervical spine does occur once greater range of motion is achieved. The spine does not produce a symmetrical arch but rather breaks at the C5-C6 intervertebral joint.

DSS test according to Kolář

Diaphragma test: there is a slight shoulder elevation present and the patient has difficulty expanding latero-dorsal sections of the abdominal wall.

Intra-abdominal pressure test: the patient has difficulty expanding the lower abdominal wall

Anthropometric measurements

Height in standing: 158 cm

Table No. 5: Initial Length of Lower Extemities

Length of LE		
Length	**Left**	**Right**
Anatomical	82 cm	82 cm
Functional		
SIAS- malleolus medialis	87 cm	87 cm
Umbillicus- malleolus medialis	89 cm	89 cm
Thigh	40 cm	40 cm
Middle leg	37 cm	37 cm
Foot	24 cm	24 cm

Table No. 6: Initial Circumferences of Lower Extremities

Circumference of lower extremities		
	Left	**Right**
Thigh		
15 cm above kneecap	44,5 cm	44,5 cm
10 cm above kneecap	41 cm	41 cm
Knee joint	37 cm	37 cm
Tuberositas tibiae	32 cm	32 cm
Calf	34 cm	35 cm
Ankle joint	32,5 cm	34 cm
Heel	31 cm	31 cm
Foot	22 cm	22 cm

Table No. 7: Initial Length of Upper Extremities

Length of upper extremities		
	Left	**Right**
Whole UE	70 cm	70 cm
Humerus	26 cm	26 cm
Forearm	28 cm	28 cm
Hand	20 cm	20 cm

Table No. 8: Initial Circumferences of Upper Extremities

Circumference of upper extremities		
	Left	**Right**
Upper arm		
Relaxed	25 cm	25 cm
Contracted	26 cm	26 cm
Elbow joint	23 cm	23 cm
Forearm	18 cm	18 cm
Wrist	15 cm	15 cm
Metacarpal heads	19 cm	19 cm

Table No. 9: Other Initial measurements

Circumferences		
Head	56 cm	
Thorax	Max.inspire	Max. expire
Mesosternal (average of 3)	76 cm	73 cm
Xiphosternal (average of 3)	84,5 cm	81 cm
Hips	110 cm	

Spinal distances

Table No. 10: Initial Spinal distances

Spinal distance test	**Result**	**Normal range**
Thomayer's test	13 cm	0 cm
Schober's distance	3,5 cm	5 cm
Stibor's distance	5 cm	7 - 10 cm
Otto's distance	3 cm	≥ 4,5 cm
Cepojev's distance	2 cm	2,5 - 3 cm
Forestier Fleche	1 cm	0 cm
Latero flexion	left side-16 cm	20-25 cm
	right side - 14 cm	
Cervical spine flexion	3 cm	0 cm

Examination of Shortened Muscles

Table No. 11: Initial Shortened Muscles

Length test	**Author**	**Grade**	
		Left	**Right**
Plantar flexors	Janda	1	1
Hip flexors	Janda	1	1
Hip adductors	Janda	0	0
Hamstring muscles	Janda	2	1
Paravertebral muscles	Janda	2	
Quadratus lumborum	Janda	1	1
Piriformis	Janda	1	1
Pectoralis major	Kendall		
sternal part		Shortness	Shortness
clavicular part		Shortness	Shortness
Pectoralis minor	Kendall	Shortness	Shortness
M. teres major, m. latissimus dorsi, mm. rhomboid major and minor	Kendall	Shortness	Shortness
Lateral shoulder rotator	Kendall	Slight	Sligth shortness
Trapezius cranial part	Janda	2	1
M. levator scapulae	Janda	1	2
Sternocleidomastoideus	Janda	1	1

ROM examination according to Janda

Table No. 12: Initial ROM Examination

	AROM		**PROM**	
	Left	**Rigth**	**Left**	**Right**
Shoulder joint	S 40-0-160 F 160-0-0 T 35-0-130 R 70-0-90	S 30-0-130 F 120-0-0 T 20-0-115 R 60-0-80	S 40-0-165 F 160-0-0 T 40-0-135 R 70-0-90	S 35-0-130 F 125-0-0 T 25-0-120 R 65-0-90
Elbow joint	S 0-0-140	S 0-0-140	S 0-0-140	S 0-0-140
Forearm	R 90-0-80	R 90-0-80	R 90-0-80	R 90-0-80
Wrist	Not tested due to cannula	S 60- 0 -75 F 30 -0 -45	Not tested due to cannula	S 70- 0 -80 F 30 -0 -45
Thumb CM joint MP joint IP joint	F 30-0-15 S 30-0-0 Opposition: tip of V S 0-0-50 S 5-0-85	F 30-0-15 S 40-0-0 Opposition: tip of V S 0-0-50 S 5-0-85	F 30-0-15 S 35-0-0 Opposition: tip of V S 0-0-50 S 5-0-85	F 30-0-15 S 40-0-0 Opposition: tip of V S 0-0-50 S 5-0-85
Fingers				
MP joint	II. S 30-0-80 F 20-0-20 III. S 30-0-90 F 20-0-20 VI. S 45-0-90 F 20-0-20 V. II. S 40-0-90 F 20-0-20	II. S 30-0-90 F 20-0-20 III. S 30-0-90 F 20-0-20 VI. S 45-0-90 F 20-0-20 V. II. S 45-0-90 F 20-0-20	II. S 35-0-90 F 20-0-20 III. S 40-0-90 F 20-0-20 VI. S 45-0-90 F 20-0-20 V. II. S 45-0-90 F 20-0-20	II. S 40-0-90 F 20-0-20 III. S 40-0-90 F 20-0-20 VI. S 45-0-90 F 20-0-20 V. II. S 45-0-90 F 20-0-20
IP1 joint	II.S 0-0-80 III. S 0-0-80 VI. S 0-0-90 V. S 0-0-100	II.S 0-0-90 III. S 0-0-90 VI. S 0-0-90 V. S 0-0-100	II.S 0-0-80 III. S 0-0-80 VI. S 0-0-90 V. S 0-0-100	II.S 0-0-90 III. S 0-0-90 VI. S 0-0-90 V. S 0-0-100
IP2 joint	II.S 0-0-55 III. S 0-0-65 VI. S 0-0-70 V. S 0-0-65	II.S 0-0-60 III. S 0-0-60 VI. S 0-0-75 V. S 0-0-70	II.S 0-0-60 III. S 0-0-70 VI. S 0-0-70 V. S 0-0-70	II.S 0-0-60 III. S 0-0-70 VI. S 0-0-80 V. S 0-0-70
Hip joint	S 15-0-125 F 45-0-20 R 25-0-40	S 15-0-125 F 45-0-20 R 25-0-40	S 15-0-125 F 45-0-20 R 45-0-40	S 15-0-125 F 45-0-20 R 45-0-40
Knee joint	S 0-0-130	S 0-0-130	S 0-0-130	S 0-0-130
Ankle	S 15-0-45 R 15-0-40	S 15-0-45 R 20-0-40	S 15-0-45 R 20-0-40	S 20-0-45 R 20-0-40

Examination of Muscle Strength

Facial Muscles

According to Kendall

Grade 0-5

Table No. 13: Initial Muscle Strength of Facial Muscles

Muscle	**Left**	**Right**
Frontalis	5	5
Corrugator supercilii	4	4
Procerus	4	4
Orbicularis oculi	5	5
Nasalis	3	3
Zygomaticus major	4	4
Levator labií superioris	2	2
Levator anguli oris	3	3
Mentalis	3	3
Buccinator	3	3
Risorius	5	5
Depressor labii inferioris and platysma	2	2
Depressor anguli oris	3	3
Pterygoideus medialis and lateralis	3	3

Muscle Strength Test of the Trunk and Neck

According to Kendall

Grade 0-10

Table No. 14: Initial Muscle Strength of the Trunk and Neck

Muscles	**Grade**
Rectus abdominis, external and internal obliques on the opposite side	7
Upper abdominal -serratus anterior, external oblique, rectus abdominis	8
Lower abdominal muscle test -rectus abdominis	8
Anterior neck flexors-longus capitis, longus colli, rectus capitis	8
Anterolateral neck flexors-SCM, scaleni	6
Posterolateral neck extensors –splenius capitis and cervicis, semispinalis capitis and cervicis, cervical erector spinae	8

Muscle Strength Examination of Extremities According to Janda

Grade 0-5

Table No. 15: Initial Muscle Strength of Upper Extremities

Movement	Left	Right
Abduction of shoulder joint	5	3
Flexion of shoulder joint	5	3
Internal rotation of shoulder joint	5	4
External rotation of shoulder joint	4	4
Flexion of shoulder joint	5	5
Extension of shoulder joint	5	5
Elevation of shoulders	4	3
Forearm pronation	5	5
Forearm supination	5	5
Wrist flexion	Not tested	4
Wrist extension	Not tested	4
Radial duction	Not tested	4
Ulnar duction	Not tested	4
Finger flexion	4	3
Finger extension	4	3
Finger abduction	4	4
Finger adduction	4	4
Flexion of hip joint	5	5
Extension of hip joint	4	4
Abduction of hip joint	5	5
Adduction of hip joint	5	5
IR of hip joint	5	5
ER of hip joint	4	4
Flexion of knee joint	5	5
Extension of knee joint	5	5
Dorsiflexion of ankle joint	5	5
Supination of ankle joint with plantar flexion	5	5

Neurological Examination

Table No. 16: Initial Neurological Examination of Facial Nerves

Nerve	**Results**
Olfactory	The patient was able to identify familiar odors
Optic	The patient was able to read different fonts and sizes. Peripheral vision is in norm.
Oculomotor	Pupils seemed to have normal constriction
Trochlear	The patient follows finger with slight movement of head but upon instruction maintains the position of the head.
Trigeminal	Sensory: the patient could feel when she was touched in both sides of the face and could distinguish between a sharp and dull sensation. Motor: the patient can hold mouth open
Abducens	The patient follows finger with slight movement of head but upon instruction maintains the position of the head.
Facial	Motor: patient can smile, wrinkle face and puff cheeks without difficulty. Sensory: Patient can differentiate between sweet and salty taste.
Vestibulocochlear	Hearing: Patient is able to hear snap of fingers close to her ears.
Glossopharyngeal	The patient can swallow and say “Ah” without difficulty.
Vagus	Gag reflex can be elicited.
Spinal Accessory	The patient can shrug shoulders.
Hypoglossal	The patient can stick out tongue and move it from side to side.

Peripheral (Deep Tendon) Reflexes

Table No. 17: Initial Deep Tendon Reflexes

Reflex	Left	Right
Patellar	Normoreflexia	Normoreflexia
Achilles	Normoreflexia	Normoreflexia
Medioplantar	Normoreflexia	Normoreflexia
Biceps	Normoreflexia	Normoreflexia
Radial	Normoreflexia	Not tested
Triceps	Normoreflexia	Normoreflexia

Sensory system examination

Light touch was used to examine dermatomes of upper and lower extremities. On the upper extremities dermatomes C4, C5, C6, C7, C8, T1, and T2. On the lower extremities dermatomes L2, L3, L4, L5, S1, and S2. Bilaterally on both upper and lower extremities the patient felt the sensation simultaneously without any abnormalities. Some sensation was decreased in the area of the C7 dermatome on her right hand.

Cortical sensory function

Tactile discrimination: The patient could distinguish the number of points on her left thigh and right forearm.

Kinesthetic sensation: The movement was carried out on the metacarpophalangeal joint of the index finger of the right upper extremity the movement was carried out to about 30º extension and the patient was able to correctly tell in which direction the movement was and when it ended, however, had difficulty with determining the initiation of the movement.

The movement was also carried out on the metacarphophalangeal joint of the right lower extremity and the movement was carried out to about 30° extension and the patient was able to correctly tell when the movement started, in which direction it was and when it ended.

Graphesthesia: the patient was able to recognize the number drawn on the skin of her right thigh.

N. medianus tests

The tests were performed by the patient's right hand

Circle test- patient manages without difficulty

Fist- patient manages to make a fist, slight difficulty flexing fully the II. and III. digits on both hands

Examination of Grips

Table No. 18: Initial Examination of Grips

Tests	**Left**	**Right**
Lateral grip	Manages well	Manages well
Pincer grip	Manages	Manages but takes longer time
Grip with terminal opposition of the thumb and index	Manages	Manages
Medium wrap	Manages- there are llimitation with larger heavier objects	Manages- there are limitation with larger heavier objects
Power sphere	Manages- there are llimitation with larger heavier objects	Manages- there arellimitation with larger heavier objects
Hook grip	Manages well	Manages well

Examination of reflex changes

The examination was performed by palpation.

Skin

Face: signs of microstomia and radial creases around the mouth, skin is slightly dry in the area of the face and there is a tightness around the temporomandibular joint, and the mouth.

Upper extremities: On both upper extremities Raynaud's syndrome on the periphery of the digits is present. Resistance of the skin on the palms, dorsum of the hands and all fingers is present bilaterally. The second and third fingers of both hands are especially affected. There are ulcerations on the second and third fingers bilaterally. Slight dryness of the skin is present near the left elbow. There is a scar after carpal tunnel surgery on the base of the right palm. There is a slight resistance of the skin around the right shoulder.

Trunk: the skin drag is increased around the pectoral area, especially around the right shoulder and clavicle. There is an increased resistance of the skin in the area of the upper trunk, around the thoracic spine bilaterally and the right scapula.

Lower extremities: the patient has varicose veins on her lower extremities, the skin is resistant on the calves slightly in all directions, on the thigh there are restrictions bilaterally around the tensor fasciae latae. The skin on the periphery is limited slightly dorsally and on the planta, the fingers are colder and slightly whiter, there is a resistance of the skin present here bilaterally. There are ulcerations on the bellies of all fingers and laterally on the left pinkie.

Subskin

Trunk: there was limitation of moving the subskin on the nape of the neck, around the upper ribs, especially on the right side and on the back in all directions.

Upper extremities: there is resistance around the right shoulder joint and periphery of the hands.

Lower extremities: there is resistance on the medial part of the thigh, around the tensor fascia latae especially on the right and the frontal part of the thigh slightly bilaterally.

Fascia

Neck: increased anterior cervical resistance of the fascia in lateral direction both to the right and left side. The resistance was slightly more present on the right

Pectoral fascia : hard barrier on both right and left side

Thoracodorsal fascia: hard barrier bilaterally in all directions

Fascia of the upper extremities: increased resistance of the fascia on the whole upper extremities, especially on the right side.

Fascia on the lower extremities: increased resistance on the thighs laterally

Metacarpal and metatarsal movement is limited bilaterally

Muscles

Trunk: hypertonus of the upper part of m. trapezius, mm. scaleni, m. supraspinatus, m. infraspinatus, m. levator scapulae and short extensors of the craniocervical junction was present bilaterally.

Upper extremities: hypertone of m. biceps brachii and flexors and externsors of the hand was present

Lower extemities: hypertone of mm. adductores was present, m. tensor fasciae latae- more on the right lower extremity, m. rectus femoris and m. triceps surae bilaterally

Periostal points

An unpleasant feeling when palpating the right clavicle and the right acromion was present.

Scars

There is a scar present after carpal tunnel surgery at the base of the right palm. There is limited mobility in the distal part of the scar in lateral directions.

Joint play examination according to Lewit

- Blockage in the sternoclavicular joint on the right was found
- Blockages in the acromioclavicular joint on the right in all directions were found
- Blockages in the first rib bilaterally was found
- Blockages of the right shoulder were found in the dorsal and cranial direction
- Movement of the right scapula was limited
- Blockages of the radiocarpal joints in all directions were found on the right, they were not tested on the left due to the canulla in the dorsal part of the forearm
- Blockages of the peripheral parts of the upper extremities were present bilaterally - the metacarpophalangeal joints, proximal and distal interphalangeal joints in dorsal and lateral directions
- Blockages in the peripheral parts of the lower extremities are present- in the metatarsophalangeal joints in dorsal and lateral direction, the proximal and distal interphalangeal joints bilaterally in dorsal and lateral directions

Scleroderma health assessment questionnaire

Appendix 3

Cochin hand function scale questionnaire

Appendix 5

Mouth handicap in systemic sclerosis questionnaire

Appendix 7

Conclusion of the initial examination

The patient diagnosed with systemic scleroderma has been hospitalized in Revmatologický ústav for re-examination and daily infusions and to improve blood flow in peripheral parts of the body.

Today the patient is well-oriented in time and place and cognitive functions are well-maintained. The patient is can achieve all ADLs and is capable of transfers to her room and the gym but is slightly limited by Raynaud's syndrome on her upper extremities causing her issues with dressing. She is also limited due a to slight loss of sensitivity on her right hand after carpal tunnel operation.

The patient markedly has scoliosis and a protruding right scapula. The abduction stereotype of the right scapula is pathological and limitations are also seen in the joint-play of the right shoulder, acromioclavicular joint, and sternoclavicular joint. The ROM of the right shoulder is limited mainly in flexion and abduction. She is unstable when standing on one leg and the DSS tests showed a weakness of the deep stabilization system. The overall gait of the patient is quite stiff, the left leg is bearing more weight, and the right is externally rotated. There is a limitation in the external rotation of both hip joints.

Limitations in joint play are mainly visible on the small peripheral joints of the upper and lower extremities. The skin, subskin, and fascia are also mainly limited in these areas as well as on the whole back, pectoral area and area around the right shoulder.

The spinal distances showed limitations in the whole spine. Muscle hypertension was palpated on the flexors and extensors of the hands, scaleni, upper trapezius, infraspinatus and supraspinatus.

The body is affected by systemic sclerosis mainly on the periphery of the extremities and the face. Mainly restrictions of the skin, subskin and fascia are present here. Limitations of ROM of the scapula and right shoulder might be due to the injury of the clavicle in 2022.

3.4 Short-Term and long-term Physiotherapy Plan

Short-term rehabilitation plan

- Improve tissue mobility of the orofacial area
- Improve skin, sub skin and fascia mobility in the area of the neck, back, pectorals, upper extremities and lower extremities
- Improve mobility of the scar at the base of the patient's right palm
- Improve ROM of the right shoulder joint and proximal and distal interphalangeal joints
- Release joint blockages in the right acromioclavicular and sternoclavicular joint, the first ribs, the right shoulder
- Improve joint mobility in the radiocarpal joints, the metacarpophalangeal joints and proximal and distal interphalangeal joints
- Release joint blockages in the lower extremities- the talocrural joint, metatarsophalangeal joints, the proximal and distal interphalangeal joints
- Improve muscle tone of hypertonic muscles of the trunk, upper extremities and lower extremities
- Improve breathing stereotype
- Improvement of fine motor function of the hands

Therapy proposal

- Soft tissue techniques in the orofacial area, neck, back, pectorals, upper and lower extremities- fascia stretch, Kibler's fold
- Soft tissue techniques for the scar at the base of the right palm- pressure massage, C- curve, S- curve
- Mobilization techniques for blocked joints of the upper and lower extremities, acromioclavicular, sternoclavicular joints and scapula
- PIR technique according to Lewit to release hypertone of m. trapezius, mm. scaleni, m. supraspinatus, m. infraspinatus, m. levator scapulae,short extensors of the craniocervical junction, m. biceps brachii, flexors and externsors of the hands, mm. adductores was present, m. tensor fasciae latae, m. rectus femoris and m. triceps surae
- PNF according to Kabat for the right scapula and right upper extremity
- Respiratory physiotherapy

Long-term rehabilitation plan

- Improve overall posture
- Improve gait stereotype
- Improve shoulder abduction stereotype
- Improve breathing stereotype
- Strengthen weakened muscles
- Improve ROM of joints on upper and lower extremities
- Improve deep stabilization system activation

Therapy proposal

- Deep stabilization system activation to improve posture
- PNF diagonals according to Kabat for the shoulders
- Strengthening exercises for weakened muscles
- Soft tissue techniques for the trunk, upper and lower extremities-fascia stretch, Kibler's fold
- Mobilization according to Lewit to renew joint ROM
- Respiratory physiotherapy
- Deep stabilization system exercises
- Training of fine motor skills and improving muscle strength of the upper extremities

3.5 Daily Record of Therapy Progress

Therapy unit 1

Date: 11.1.2024

Status praesens, subjective: The patient felt cold during the initial kinesiological examination and is arriving warmly dressed and with gloves on her hands today. She has been feeling slightly tired due to bad sleep. She is complaining about her roommate snoring at night. The patient is feeling pain on the tips of her fingers on both hands, a 5 on the visual analogue scale. It is most present on the second finger of her right hand and second and the third finger on her left hand. She is also experiencing a tingling feeling in her fingers, which is unpleasant. The patient applied scleroderma cream on her face and hand in the morning. Resistance of the skin on the palms, dorsum of the hands and all fingers is present bilaterally.

Status praesens, objective: The patient is arriving with a cannula in her left forearm. The tips of her fingers are swollen and start to turn pink and slightly purple on the second and third fingers of both upper extremities as soon as the patient takes her gloves off. The skin of the orofacial area shows signs of microstomia and radial creases around the mouth, but the skin is well hydrated.

Goal of therapeutic unit:

- Decrease swelling of patients fingers
- Improve ROM of IP and MCP joints
- Improve mobility of the scar at the base of the patient's right palm
- Improve skin, sub skin and fascia mobility on the upper extremities and right shoulder
- Improve soft tissue mobility of the orofacial area
- Stretch shortened muscles in the area of the right shoulder joint and elbow joint- m. pectoralis, m. biceps brachii
- Proposed therapy:
- Soft balling on the periphery of the upper extremities

- Kiblers fold on both upper extremities
- Fascia release on the upper extremities and right shoulder
- Soft tissue techniques for the scar on the patient's right palm
- Mobilization by Lewit of the joints of the hands:
- Passive stretch of the right upper extremity in the direction of the first extension diagonal PNF
- Light stretch of soft tissues in the orofacial area

Description of therapeutic unit:

The therapeutic unit was initiated in supine position with soft balling techniques in the area of the hands and right forearm. Kiblers fold was performed on both upper extremities except on the the left forearm, where the cannulla is placed. The skin soft tissues were treated on the patient's hands as well as a pressure massage of the scar below the patient's right palm. Mobilization according to Lewit of the IP joints in the dorsopalmar and lateral direction, and mobilization of the MCP heads in the dorsopalmar direction followed. The right upper extremity was stretched passively to flexion, abduction, and then passively in the PNF first extension diagonal. Lastly, the soft tissues of the orofacial area were softly stretched and the patient was instructed self-therapy for the soft tissues of the orofacial area.

Outcome of the therapeutic unit-objective: The patient managed the therapeutic unit without difficulty. There is a slight improvement in the mobility of the skin on the patient's fingers and upper extremities and a slight decrease of swelling in the tips of her fingers, however the tips of the fingers stay very cold with visible Raynaud's syndrome. The scar at the base of the patient's right palm is slightly more moveable in the cranial direction. Joint-play in the IP and MCP is slightly improved on both upper extremities. During passive stretching of the muscles of the right upper extremity there was a soft resistance at about 120° of abduction and the patient felt increased pain here. But once the movement was repeated a few times the ROM increased by about 10° in abduction. The soft tissues in the orofacial area were less resistant after the therapy.

Outcome of the therapeutic unit-subjective: The patient feels well after the therapeutic unit. Her fingers were feeling cold during therapy, so whilst working with one upper extremity, the second was covered by the patient's jacket. She felt some pain

during the passive movements in her right shoulder, but she felt no pain after the therapy.

Self-therapy: The patient was instructed to stretch the soft tissue in her orofacial area as well as lightly massage the face when applying cream in the morning and at night.

Therapy unit 2

Date: 12.1.2024

Status praesens, subjective: The patient feels well today. She had infusions yesterday that took four hours. She is feeling stiffness in her whole body, especially the neck, after sitting so long in the same position. The patient is still feeling pain on the tips of her fingers on both hands, with no changes from yesterday. The patient applied scleroderma cream on her face and hands in the morning and massaged her face.

Status praesens, objective: The patient is arriving alone, she has a cannula in her left forearm. When walking the right arm is very passive, the arm swing is almost not present. The neck fascia is restricted in the lateral direction to both sides. The upper trapezius muscles are in hypertone. The thoracodorsal fascia and pectoral fascia bilaterally are restricted in all directions.

Goal of therapeutic unit:

- Decrease swelling of patient's fingers
- Improve ROM of IP and MCP joints
- Improve mobility of the scar at the base of the patient’s right palm
- Improve skin, subskin and fascia mobility on the upper extremities and right shoulder
- Improve neck, pectoral and thoracodorsal fascia mobility
- Decrease hypertone of the upper part of the trapezius muscle
- Improve the ROM of the right shoulder joint

Proposed therapy:

- Soft balling on the periphery of the upper extremities
- Kibler's fold on both upper extremities and the upper back
- Fascia release on the upper extremities, right shoulder, the pectoral fascia, and the thoracodorsal fascia
- Soft tissue techniques for the scar on the patient's right palm
- Mobilization by Lewit of the joints of the hands and right scapula
- PIR according to Lewit of upper m. trapezius bilaterally
- PNF for right scapula (repeated contractions)

Description of therapeutic unit:

The therapeutic unit was initiated in seated position with the treatment of the anterior neck fascia. The patient then moved to the supine position with soft balling techniques in the area of the hands and right forearm. Kibler's fold was performed on both upper extremities with the exception of the left forearm, where the cannula is placed. The skin soft tissues were treated on the patient's hands as well as a pressure massage of the scar below the patient's right palm. Mobilization of the IP joints in the dorsopalmar and lateral direction, and mobilization of the MCP heads in the dorsopalmar direction followed. The upper trapezius was treated with PIR according to Lewit bilaterally. The pectoral fascia was stretched. The right acromioclavicular joint was mobilized ventro-dorsally and cranio-caudally as well as the right sternoclavicular joint was mobilized according to Lewit. In prone position we treated the thoracodorsal fascia and then mobilized the right scapula. We then moved on to PNF of the right scapula in sidelying. Anterior elevation, posterior depression, posterior elevation and anterior depression were performed using repeated contraction.

Outcome of the therapeutic unit-objective: The patient managed the therapeutic unit without difficulty. There is a slight improvement in the mobility of the skin on the patients' fingers and upper extremities as well as the thoracodorsal and pectoral fascia bilaterally. The scar at the base of the patient's right palm is more moveable in all directions. Joint-play in the IP and MCP are slightly improved on both upper

extremities but especially the second and third fingers are still limited into flexion (about 80°). The hypertone of trapezius muscles bilaterally was improved, although some trigger points are still present upon palpation at the end of the therapeutic unit. The neck facia, however, has released significantly both to the right and left. The mobilization of the right acromioclavicular joint was slightly painful for the patient, but she was able to tolerate the treatment and the sternoclavicular joint treatment. The patient was able to perform PNF repeated contractions with the scapula, but had most difficulty with posterior depression.

Outcome of the therapeutic unit-subjective: The patient felt relief in the area of her neck and overall feels less stiff in her upper body. She experienced slight pain during the acromioclavicular joint mobilization on the right side but overall feels well after the therapy.

Self-therapy: The patient was instructed to stretch her upper trapezius passively and perform self-therapy PIR of the upper trapezius bilaterally. She was instructed to do PIR of the finger flexors and extensors bilaterally.

Therapy unit 3

Date: 15.1.2024

Status praesens, subjective: The patient feels well today. The patient is still feeling tingling pain on the tips of her fingers on both hands and has not noticed any major changes.

Status praesens, objective: The patient is arriving alone, she has a cannula in her left elbow crease. The swelling in the hands is still present but appears to have slightly decreased, the Raynaud's phenomenon is also less present.

Goal of therapeutic unit:

- Decrease swelling of patient's fingers
- Improve ROM in small finger joints
- Treat blockages in the radiocarpal joints
- Improve mobility of the scar at the base of the patient's right palm

- Improve skin, sub skin and fascia mobility on the upper extremities and right shoulder
- Improve the ROM of the right shoulder joint
- Improve mobility of the orofacial area
- Improve breathing pattern

Proposed therapy:

- Soft balling on the periphery of the upper extremities
- Fascia release on the upper extremities, right shoulder, the pectoral fascia, and the thoracodorsal fascia
- Soft tissue techniques for the scar on the patient's right palm
- Mobilization by Lewit of the joints of the hands and right scapula
- PNF according to Lewit for the right shoulder and scapula
- Exercises for the facial muscles
- Practicing the correct breathing pattern

Description of the therapeutic unit:

The therapeutic unit was initiated in a seated position using soft tissue techniques, specifically balling in the hands and forearms. Then we focused on the treatment of the scar with pressure massage, C- curve, and S-curve. After the soft tissue treatment, the unit continued with respiratory exercises, first focusing on localized breathing with facilitation of touch to improve breathing wave progression, followed by breathing against resistance. The breath work ended with practice of the correct breath wave. Soft tissue techniques were performed in the pectoral area and on the thoracodorsal fascia bilaterally. The IP, MCP, radiocarpal joints bilaterally and right scapula were mobilized according to Lewit. Second flexion and extension PNF diagonal repeated contraction according to Lewit was performed with the right upper extremity. We then sat in front of a mirror and performed facial exercises: smiling widely by turning the mouth upward 10 repetitions, opening the eyes wide open 10 repetitions, lifting the eyebrows 10 repetitions, frowning by squeezing the forehead 10 repetitions, opening the mouth as wide as possible 10 repetitions, pouting the lips 10 repetitions.

Outcome of the therapeutic unit-objective: The moveability of skin and soft tissue of the fingers seems to improve slightly in all directions as well as the thoracodorsal fascia and pactoral fascia mobility, the scar on the right hand is also less restricted in the cranial direction. The ROM of the right shoulder improved to about 135° of flexion and 130° of abduction. The ROM of the IP and CMC joints bilateerally are without any major changes but the swelling seems to be decreasing.

Outcome of the therapeutic unit-subjective: The patient enjoyed breathing exercises and facial exercises. She felt no pain during the therapy and is in a good mood.

Self-therapy: The patient is instructed to perform facial exercises in front of the mirror as stated above- smiling widely by turning the mouth upward 10 repetitions, opening the eyes wide open 10 repetitions, lifting the eyebrows 10 repetitions, frowning by squeezing the forehead 10 repetitions, opening the mouth as wide as possible 10 repetitions, pouting the lips 10 repetitions. To improve the ROM of the IP and CMP joints exercises for self-therapy are shown: making a fist and stretching the fingers out, touching each finger tip with the thumb and sliding the thumb down each finger.

Therapy unit 4

Date: 16.1.2024

Status praesens, subjective: After experiencing severe nausea this morning, the patient took Antalgin for pain. She vomited in the morning and has a migraine, which she suffers from a few times a month since childhood. She is feeling better at the time of the therapy.

Status praesens, objective: The cannula was removed from her hand. The swelling in the hands is still present and there are no visible changes. The patient looks slightly pale but believes she can manage today’s therapy. Due to the patient feeling unwell in the morning, the therapy unit was postponed to the afternoon. The patient has severe hypertone of the neck muscles- the upper trapezius, short neck extensors, scaleni, sternocleidomastoid and levator scapulae bilaterally. The skin, subskin, and fascia of the neck are also restricted on both sides. There are also blockages present in the first rib both on the right and left side.

Goal of therapeutic unit:

- Release soft tissues of the neck and upper thorax
- Decrease swelling of patients fingers
- Improve ROM in IP and MCP joints
- Release blockages of the first rib
- Improve skin, sub skin and fascia mobility on the upper extremities and right shoulder
- Improve right shoulder ROM

Proposed therapy:

- PIR according to Lewit for upper trapezius, m. levator scapulae, mm. scaleni, sternocleidomastoid, short neck extensors bilaterally
- Kibler's fold on upper extremities
- Fascia release on the upper extremities, right shoulder, the pectoral fascia, the thoracodorsal fascia, and neck fascia
- Scar care for the scar on the patient's right palm
- Mobilization by Lewit of the joints of the hands
- Mobilization of the first ribs
- PNF repeated contraction according to Lewit for the right shoulder

Description of the therapeutic unit:

We initiated the therapy unit in seated position by releasing the neck fascia and the skin around the neck, pectorals and upper thorax bilaterally. The first ribs were mobilized bilaterally. Next PIR according to Lewit of the trapezius, levator scapulae, scaleni, sternocleidomastoid and short neck extensors was performed. This was followed by soft tissue techniques on the upper extremities- fascia release, Kibler's fold, scar care on the right palm using pressure massage. Mobilization according to Lewit of the IP and MCP joints bilaterally followed. At the end of the unit we performed first

flexion and extension, and second flexion and extension diagonals with the right upper extremity using repeated contraction technique according Lewit.

Outcome of the therapeutic unit-objective: Hypertonicity of the upper trapezius, scaleni, sternocleidomastoid and levator scapulae was treated and improved bilaterally. The soft tissues of the neck, and upper thorax released slightly in all directions. The moveability of skin and soft tissue of the fingers seems to improve slightly in all directions and the scar on the right hand is more moveable in the cranial and distal directions. The ROM of the right shoulder seems to be without any major changes since the last therapeutic unit.

Outcome of the therapeutic unit-subjective: The patient feels a release in the area of the neck and is feeling better than before the therapy.

Self-therapy: The patient was reminded how to perform self-therapy PIR according to Lewit of upper trapezius. She was instructed how to perform autotherapy PIR for mm. scaleni, and short neck extensors as well as automobilization of the C/Th spine according to Lewit.

Therapy unit 5

Date: 17.1.2024

Status praesens, subjective: The patient is feeling better this morning. The nausea and headache had improved markedly. She does not feel as stiff as yesterday.

Status praesens, objective: The patient is arriving with a cannula in her left elbow crease. Her hands are less cold to the touch but still swollen.

Goal of therapeutic unit:

- Decrease the swelling in hands
- Improve the ROM of IP and MCP joints,
- Optimize tone of soft tissues in the orofacial area
- Stretching of shortened muscles in the shoulder and elbow joint bilaterally - m. pectoralis major, m. biceps brachii

- Optimization of muscle tension of the upper extremity muscles bilat. - m. deltoideus, m. pectoralis major, m. biceps brachii, m. pronator teres, m. pronator quadratus, flexors and extensors of the hand

Proposed therapy:

- Fascia release on the upper extremities
- Soft tissue technique on the hands and the fingers
- Facial exercises
- Scar care for the scar on the patient's right palm
- Mobilization by Lewit of the joints of the hands
- PIR technique according to Lewit on muscles m. pectoralis major, and m. biceps brachii bilat.
- Passive stretching of m. pectoralis major, m. biceps brachii, flexors and extensors of the hand bilat.
- Relaxation technique PNF according to Kabat (contraction-relaxation) on the muscles of m. deltoideus, m. pectoralis major, m. biceps brachii, hand flexors bilat.
- PIR technique according to Lewit on flexors and extensors of the hand bilat

Description of therapeutic unit:

Today's therapy started in supine with application of soft tissue techniques in the upper extremities - dorsal side of the hand, wrist and forearm, pressure massage of the scar on the base of her right hand. The small joints of the hand were mobilized (palmar and dorsal fan). This was followed by passive stretching of the upper extremities in a diagonal pattern in the direction of the movement components of the first extensor diagonal followed by application of the PIR technique according to Lewit with stretching to m. pectoralis major, m. biceps brachii bilaterally. PIR technique was performed to affect the hypertonicity of flexors and extensors of the hands bilaterally. Facial exercises were performed- same as on the 15.1. with the addition of puffing the

cheecks- 10 repetitions, sucking the cheeks inward- 10 repetitions, and strengthening the tongue by pushing up towards the ceiling and pointing the tongue to the floor.

Outcome of therapeutic unit- objective: After the therapy, reflex changes appear - hyperemia in the area of the scapulae bilaterally and on the periphery of the hands bilaterally. Skin in the area of the upper extremities was more moveable in relation to the subcutaneous tissue. The ranges in the right shoulder joint increased by about 10° in flexion and 10° in abduction after therapy.

Outcome of therapeutic unit- subjective: Patient leaves in a stable condition, without acute pain, instructed for self-therapy.

Self therapy: The patient was instructed to practice fine motor skills - connecting the fingers of the hand with the thumb of the hand and making a fist and then extending the fingers into a star shape 3 times a day.

Therapy unit 6

Date: 18.1.2024

Status praesens, subjective: The patient feels well today and feels some improvement with activities of daily living like brushing her hair. She says staying in the same position- half reclined, causes her to feel stiff in the afternoons.

Status praesens, objective: The patient is arriving with a cannula in her left elbow crease.

Goal of therapeutic unit:

- Treatment of soft tissues (especially skin and subcutaneous tissue) in the area, thighs, calves, forearms, shoulders, neck, chest and periphery of the upper and lower extremities bilaterally
- Treatment of soft tissues of the orofacial region
- Increasing the range of motion in the right shoulder and small joints of the hand bilat.

- Stretching of shortened surrounding the shoulder joint, right elbow joint, radiocarpal joint, hip joint, knee joint and ankle joint bilat.
- Relaxation of hypertonic muscles in the neck, shoulder joint, right elbow joint, bilaterally
- Removal of joint blockages in the minor joints of the hand and foot bilaterally
- Correction of posture

Therapy proposal:

- Soft tissue techniques on the face, chest, forearms and periphery of upper extremities bilaterally
- Soft tissue techniques on thighs, calves and periphery of lower extremities bilaterally
- PIR technique according to Lewit on flexors and extensors of the hand bilaterally
- PIR technique for m. pectoralis major and m. biceps brachii bilat.
- Passive stretching of m. pectoralis major, m. biceps brachii, m. supinator, m. pronator teres, m. pronator quadratus, flexors and extensors of the hand bilat.
- Relaxation technique PNF according to Kabat on muscles m. deltoideus, m. pectoralis major,m. biceps brachii, m. supinator, hand flexors bilaterally
- Instructing proper posture

Description of therapeutic unit:

Therapy was initiated on the lower limbs using soft techniques on the periphery of lower extremities. Mobilization techniques were performed on the small joints of the leg. This was followed by stretching of m. iliopsoas, m. rectus femoris and mm. adductores by PIR technique with stretching. The therapy was transferred to the upper limbs and applied similarly to the 5th therapeutic unit. Finally, the patient was instructed correct posture in seated and in standing.

Outcome of therapeutic unit- objective: The patient tolerated the therapy. The skin on the periphery of upper and lower extremities was more moveable, the subcutaneous tissue was more moveable against the skin although still with significant resistance. Ranges of motion increased at the right shoulder joint increased to about 140° in flexion.

Outcome of therapeutic unit- subjective: The patient felt relaxed after the therapy and left in a stable condition.

Self-therapy: The patient was instructed to further perform the existing facial exercises- twice a day two repetitions (smiling widely by turning the mouth upward 10 repetitions, opening the eyes wide open 10 repetitions, lifting the eyebrows 10 repetitions, frowning by squeezing the forehead 10 repetitions, opening the mouth as wide as possible 10 repetitions, pouting the lips 10 repetitions, puffing the cheeks- 10 repetitions, sucking the cheeks inward- 10 repetitions, and strengthening the tongue by pushing up towards the ceiling and pointing the tongue to the floor.

Therapy unit 7

Date: 19.1.2024

Status praesens, subjective: The patient feels physically well today. However, she feels nervous as she is to leave for a medical exam in 30 minutes. She sees some improvements in the mobility of the second and third fingers of her left hand.

Status praesens, objective: The patient is arriving with a cannula on the dorsum of her right hand with dressing around the hand with the cannula. Her hands feel lukewarm to the touch, the left more than the right. The tips of the fingers remain colder than the rest of the hands. The skin movability is restricted on the fingers of both hands.

Goal of therapeutic unit:

- Treatment of soft tissues on the upper extremities bilaterally
- Treatment of soft tissues of the orofacial region
- Increasing the range of motion in the right shoulder and small joints of the hand bilaterally
- Joint mobilization in the minor joints of the hands bilaterally

- Breathing stereotype correction

Therapy proposal:

- Soft tissue techniques on the face, chest, forearms and periphery of upper extremities bilaterally
- Passive stretching of m. pectoralis major, m. biceps brachii, m. supinator, m. pronator teres, m. pronator quadratus, flexors and extensors of the hand bilat
- Contraction- relaxation according to Kabat on muscles m. deltoideus, m. pectoralis major,m. biceps brachii, m. supinator, hand flexors bilaterally
- Mobilization by Lewit of the joints of the hands
- Localized breathing, active exhalation exercises

Description of therapeutic unit:

The therapeutic unit was initiated in supine position with soft balling techniques in the area of the left hand and both forearms. Kibler's fold was performed on both upper extremities with the exception of the right dorsum of the hand, where the cannula is placed. The skin soft tissues were treated on the patient’s hands bilaterally. Mobilization of the IP joints in the dorsopalmar and lateral direction, and mobilization of the MCP heads in the dorsopalmar direction followed bilaterally. The right upper extremity was stretched passively to flexion, abduction. Contraction- relaxation PNF with first extension diagonal followed. In supine, localized breathing and active exhalation exercises were performed. Lastly, the soft tissues of the orofacial area were softly stretched.

Outcome of therapeutic unit- objective: The therapy lasted 25 minutes due to the patient having to leave for a medical examination. After the therapeutic unit the subcutaneous tissue was more moveable against the on the hand skin although some resistance prevailed on the right forearm and hand. The deltoid, pectoralis major, biceps brachii and flexors of the hands were in less hypertone. The range of motion of the right shoulder improved slightly throughout the therapy.

Outcome of therapeutic unit- subjective: The patient tolerated the therapy well and left in a stable condition.

Self-therapy: the patient was instructed to continue in the existing self- therapy-hand and face exercises, PIR of hand flexors and extensors bilaterally

Therapy unit 8

Date: 22.1.2024

Status praesens, subjective: The patient feels stiff after the weekend. She has had infusion for three hours daily and she attributes her neck stiffness and pain to the half-reclined position she stays in during the infusions. The hands and fingers feel painful. The pain is about a 4 on VAS.

Status praesens, objective: The patient is arriving with a cannula on the dorsum of her right hand with dressing around the hand with the cannula. Upon palpation, there are trigger points in the upper trapezius, hypertone of scaleni, sternocleidomastoid, pectoral muscles and biceps brachii bilaterally. The mobility of the cervical fascia is limited in the lateral direction. The thoracodorsal fascia is limited in all directions.

Goal of therapeutic unit:

- Release soft tissues of the neck, hands and upper thorax
- Improve ROM in IP and MCP joints
- Improve skin, sub skin and fascia mobility on the upper extremities and right shoulder
- Improve right shoulder ROM
- Breathing stereotype correction

Proposed therapy:

- PIR according to Lewit for upper trapezius, m. levator scapulae, mm. scaleni, sternocleidomastoid, biceps brachii and pectoral muscles bilaterally
- Kibler’s fold on upper extremities
- Fascia release on the upper extremities, right shoulder, the pectoral fascia, the thoracodorsal fascia, and neck fascia
- Mobilization by Lewit of the joints of the hands:
- PNF repeated contractions and contraction-relaxation for the right shoulde

- Localized breathing, active exhalation exercises

Description of the therapeutic unit:

We initiated the therapy unit in a seated position by releasing the neck fascia and the skin around the neck, pectorals and upper thorax bilaterally. Next PIR of the trapezius, scaleni, sternocleidomastoid, pectoralis minor and minor and biceps brachii was performed. This was followed by soft tissue techniques on the upper extremities-fascia release, Kibler's fold. Mobilization of the IP and MCP joints followed. We then released the thoracodorsal fascia in prone. First and second flexion and extension of upper extremities were performed next. Contraction-relaxation in the first flexion diagonal of upper extremities was also performed.

Outcome of the therapeutic unit-objective: The tone of m. trapezius, scaleni, sternocleidomastoid and biceps brachii improved bilaterally. The skin and fascia of the neck and upper thorax released slightly as well. The movability of skin and soft tissue of the fingers seems to improve slightly in all directions. The ROM of the right shoulder seems to be without any major changes since the last therapeutic unit.

Outcome of the therapeutic unit-subjective: The patient feels a release in the area of the neck and is feeling better and less stiff than before the therapy.

Self-therapy: The patient was reminded how to perform autotherapy PIR for mm. scaleni, upper trapezius and short neck extensors accordoing to Lewit.

Therapy unit 9

Date: 23.1.2024

Status praesens, subjective: The patient feels better after yesterday's therapy. She also had occupational therapy for the first time this morning. The occupational therapist performed soft tissue techniques on her hands and forearms and mobilized the joints of her hands. They also performed various exercises to improve the fine motor function of the hands. The doctors are visiting the patient's ward and the patient would like to be in her room for the visit, she is hoping we will manage the therapy before then.

Status praesens, objective: The patient is arriving with a cannula on the dorsum of her right hand with dressing around the hand with the cannula. The hands are lukewarm to the touch with the exclusion of the tips of her fingers. The skin of her hands is relatively well moveable on her hands and forearms and the mobility of the IP and MCP joints has improved since yesterday bilaterally.

Goal of therapeutic unit:

- Treatment of the skin and subcutaneous tissue of the chest and arms bilaterally
- Treatment of the skin and subcutaneous tissue of the calves, thighs and periphery of the lower extremities bilat.
- Stretching of shortened muscles in the shoulder and elbow joint bilat. - m. pectoralis major, m. biceps brachii
- Stretching of shortened muscles in the lower extremities bilaterally - m. iliopsoas, m. rectus femoris, m. tensor fascia latae
- Improve ROM of the right shoulder
- Improve right scapula mobility
- Correction of breathing stereotype
- Activation of the deep stabilization system
- Education of proper posture

Proposed therapy:

- Soft tissue techniques on the chest and upper extremities bilaterally
- Soft tissue techniques on thighs, calves and the periphery of lower extremities bilaterally
- Passive stretching of m. pectoralis major, m. biceps brachii, m. supinator, m. pronator teres, m. pronator quadratus, hand flexors and extensors bilat.
- Relaxation technique (contraction-relaxation) PNF according to Kabat on the muscles of m. deltoideus, m. pectoralis major, m. biceps brachii, m. supinator, flexors of the hand bilat.

- PNF strengthening (repeated contractions) technique for the right scapula
- PIR according to Lewit with stretching on hip flexors, pectoralis major and m. biceps brachii, the flexors and extensors of the hand bilaterally
- Localized breathing
- Third month developmental position in supine
- Postural education and small foot training

Description of the therapeutic unit:

The therapy started with localized breathing with and without resistance. Next, skin and subcutaneous tissue were treated with soft techniques and ball facilitation on the chest and arms. The flexors and extensors of the hand were released using the PIR technique according to Lewit bilaterally. The entire right upper limb was passively stretched in the direction of the first extensor movement components diagonal, relaxed by the PNF contraction-relaxation technique and passively stretched again. Mm. rhomboidei and latissimus dorsi were strengthened in the posterior depression PNF diagonal for the scapula with repeated contractions. We did PIR technique for relaxation of the hip flexors bilaterally. Fascia on the thighs, calves and periphery of lower extremities was treated bilaterally. The patient activated the deep stabilization system in the three- month supine developmental position with her legs placed on a gym ball. The patient was educated on proper posture in sitting and standing in front of a mirror. Finally, also in seated position, small foot was trained.

Outcome of therapeutic unit- objective: The skin on the periphery of upper and lower extremities was looser, the subcutaneous tissue was more moveable against the skin. Ranges of motion increased, especially at the right shoulder joint during the session by about 10° in abduction and 10° in flexion.

Outcome of therapeutic unit- subjective: The patient tolerated the therapy well and left in stable condition.

Self-therapy: the patient was instructed to continue in the existing self-treatment, practicing corrected posture was added

Therapy unit 10

Date: 24.1.2024

Status praesens, subjective: The patient felt nauseous this morning and drank some coffee, which helped. She had a new cannula inserted in the dorsum of her left hand yesterday. The new cannula insertion was very painful. The nurses had to try inserting the cannula in seven places on the hands before finding a suitable one. This caused edema on the left hand. The edema was treated during an occupational therapy session with soft- balling and soft tissue treatment. The joints of the hands were also mobilized. After the cannula insertion yesterday, the patient felt pain in her left hand where a compression bandage was used to help find a suitable place for insertion. The patient was given some pain medication in the afternoon yesterday.

Status praesens, objective: The patient is arriving with a cannula on the dorsum of her left hand. There is visible edema on the whole left hand and the distal part of the forearm. The hands are lukewarm to the touch with the exclusion of the tips of her fingers. But the skin is stiff on the left hand and the patient is unable to make a full fist with her left hand.

Goal of therapeutic unit:

- Decrease edema on the left hand and forearm
- Treatment of soft tissues in the orofacial area
- Treatment of the skin and subcutaneous tissue of the chest, forearms and periphery of the lower extremities
- Optimization of muscle tension on the forearm - flexors and extensors of the hand bilaterally
- Optimization of muscle tension of m. pectoralis major bilat.
- Stretching of shortened muscles in the shoulder and elbow joint bilat. - m. pectoralis major, m. biceps brachii
- Stretching of the shortened flexors and extensors of the arm bilaterally
- Stretching of m. supinator and m. pronator teres, m. pronator quadratus
- Release of blockages in the talocrural and metatarsophalangeal joints bilaterally

Proposed therapy:

- Soft balling of the left hand and forearm
- Soft tissue techniques on the face, chest, forearm and periphery of the lower extremities
- Passive stretching of m. pectoralis major, m. biceps brachii, m. supinator, m. pronator teres,m. pronator quadratus, flexors and extensors of the arm bilat.
- PNF contraction-relaxation of m. deltoideus, m. pectoralis major, m. biceps brachii, m. supinator, flexors of the hand bilaterally
- PIR technique according to Lewit with stretching on m. pectoralis major bilaterally
- PIR technique according to Lewit on flexors and extensors of the hand bilaterally
- Mobilization of the interphalangeal joints of the foot
- Traction of the talocrural joints bilaterally

Description of the therapeutic unit:

Today's therapy started in supine position with soft- balling of the left hand and forearm. Soft tissues around the temporomandibular joint and mouth were lightly stretched. The fascia in the chest, arms and periphery of the lower extremities was treated bilaterally. PIR technique according to Lewit and stretching of shortened muscles of m. biceps brachii, m. supinator, m. pronator teres and m. pronator quadratus followed bilaterally. Passive stretching of the whole right upper limb in the first extension pattern followed. PNF contraxion-relaxation on the muscles m. deltoideus, m. pectoralis major, m. biceps brachii, m. supinator and flexors of the hand in the first extensor diagonal was performed on both upper extremities. The whole therapy was finished on the lower limbs, where fascia was treated. Mobilization of the small joints of the leg (dorsal and plantar fan) and traction of the talocrural joints were performed bilaterally. The patient was also instructed how to correct her gait in front of a mirror.

Outcome of therapeutic unit- objective: The skin on the periphery of upper and lower extremities was more movable, the subcutaneous tissue was more moveable against the skin. The edema slightly decreased on the left forearm. The muscle tone on the upper extremities improved during the therapeutic unit.

Outcome of therapeutic unit- subjective: The patient tolerated the therapy but felt slightly tired and sore when performing PNF.

Self-therapy: the patient was instructed to continue in the existing self- therapy

3.6 Final Kinesiological Examination

Date: 25.1.2024

Status praesens, subjective: The patient feels better than yesterday but is still experiencing some pain and swelling in her left hand. She can make a fist with her left hand better than yesterday.

Status praesens, objective: The patient is arriving with a cannula on the dorsum of her left hand. There is visible edema on the whole left hand and the distal part of the forearm. The hands are lukewarm to the touch with the exclusion of the tips of her fingers, which remain cold.

Examination of standing:

Posterior view

- The base of support: feet are hip-width apart, feet are slightly externally rotated
- Shape and contours of the heels: right heel is more rounded, left more square
- Shape and position of the ankle joints: left malleolus medialis is more prominent
- Knees: slight external rotation and valgosity of knees
- Symmetry of thoracobrachial triangles: the left seems to be smaller
- Position of pelvis: right SIPS is higher than the left, pelvic tilt to the left, crista iliaca is higher on the right
- Paravertebral muscles: paravertebral muscle in the left half of the body seem more in hypertone than on the right
- Position of the scapula: angulus infraspinatus as well as the margo medialis of the right scapula is more prominent- scapula alata
- Position of the spine: there is dextroconcavity of the lumbar spine
- Position of the shoulder girdle: right shoulder is more elevated and protracted
- Position and contour of the nuchal muscles: middle third of right trapezius is about 1 cm more prominent cranially
- Position of the head: slightly tilted to the left

Lateral view

Left

- Weight distribution: left foot more weight distributed on the forefoot
- Position of the knee joints: slightly hyperextended
- Position and curvature of the L, L/Th and Th spine: slight flattening in the thoracic spine
- Shape of the abdominal muscles : slight prominence of the abdomen
- Position of the shoulder girdle: left shoulder is protracted
- Position and curvature of the Th/C and C spine: thoracic spine is slightly flattened, there is protraction of the head

Right

- Weight distribution: left foot more to the back
- Position of the knee joints: straight, slightly hyperextended
- Position and curvature of the L, L/Th and Th spine: slight flattening in the thoracic spine
- Shape of the abdominal muscles : slight prominence of the abdomen
- Position of the shoulder girdle: right shoulder is protracted

Anterior view

- The base of support: feet are hip width apart
- The position of the feet, examination of the arches: feet are standing hip width apart, vertical arches are collapsed
- Weight distribution: weight moreso on the heels
- The position and shape of the toes: both on right and left side the big toes are extended, slight valgosity of toes

- Shape and position of ankle joints: the right malleolus medialis is more prominent
- Position of the pelvis: tilt to the left, the right SIAS is higher than the left
- Symmetry of the muscle tone of abdominal muscles: prominent abdomen
- Position of the navel (belly muscle symmetry): slightly to the right
- Symmetry of thoracobrachial triangles - both thoracobrachial triangles are very small
- Position of the collarbones and superclavicular holes: prominence of the right clavicle
- Position of the shoulder girdle: right shoulder appears protracted and elevated
- Position of the upper limbs : right upper extremity is more internally rotated than the left
- position of the head : slight tilt to the left

Dynamic spine evaluation

Flexion

Patient is able to perform movement but there is about 10 cm distance between her fingers and the ground. Less curvature is seen in the cervical spine and the thoracic spine. The movement showed an increased prominence in the left lumbar spine during flexion keeping in line with the scoliotic curve.

Extension

The range of movement is very small and the movement is not fluent. In extension, the body keeps in line with the scoliotic curve of dextroconcavity in the lumbar spine.

Lateroflexion

The vertebrae, especially in the thoracic region, spread mildly. Slight pelvic synkinesis is seen during lateroflexion to both sides. During the movement there is a

pause when the pelvic shift appears and in the area of the scoliotic curve. The range of motion to the left is slightly larger.

Modification of standing:

Standing on tiptoes-patient is able to balance on tiptoes

Examination of stability

Single leg stance- manages with difficulty on both the right and left leg, but remains very unstable in this position and lasts for a few seconds at most

Trendelenburg- does not manage on either side

Romberg I is negative

Romberg II is negative

Romberg III is positive

Gait examination

Gait is without any assistive device, cadence and velocity is rhythmical. The base of support is hip-width.

Stride length: right foot longer step, left shorter

Movement of the foot: first heel strike then flat foot then loading response the heel-off, toe off follows.

Movement and position of hip and knee: weight is applied mostly on left leg with initial swing, the right leg is slightly externally rotated

Position and movement of pelvis: sinister tilt

Position and movements of the trunk: shift to the left

Position of spine: flattened thoracic and lumbar spine, slight stiffness in movement

Activity of back muscles: active

Position of shoulder: right shoulder is protracted forward, the arm is small on the left and minimal on the right

Position and movement of the head: tilt to the left

Movement of the upper extremity: the movement is very small

Stability of walking: relatively stable

Walking with eyes closed: unstable

Walking on tiptoes: unstable

Walking in squat: unstable

Palpation of pelvis

Left SIAS is lower than the right

Pelvic tilt to the left

Breathing examination

Aspection:

Trunk position: upright, slight scoliotic curve in the lumbar spine

Chest shape: symmetrical

RR, regularity, ratio of inspiration and expiration: 17 breaths/min, eupnea

Location of breath movements: mainly lower thoracic, slight movement in the abdomen is visible

Breathing wave: normal. During inspiration the abdomen rises and is followed by a lateral costal expansion and then chest rising.

Auxiliary respiratory muscles: There is no shortness of breath, therefore no difficulty breathing and no need to use auxiliary respiratory muscles.

Basic movement stereoytypes according to Janda:

Hip extension: The homolateral lumbar extensors initiate the movement even before motion begins, which is then followed by a contraction of the contralateral

lumbar extensors. Once this happens, the superior part of the homolateral extensors continues the activation (L-Th region). The same occurs on the contralateral side, where the activation is also visible in the scapular area, reaching almost to the shoulder girdle. Only once this happens, the hip actually begins to extend with initiation of homolateral hamstring activation, causing knee flexion and subsequent bilateral activation of primarily the m. gluteus maximus. This pattern is seen on both lower extremities.

Hip abduction: Bilateral tensor mechanism is present as the hip begins to flex during greater range of motion. In the final range of motion, the lower extremity also begins to externally rotate as the toe starts to point more outwards, this external rotation is more prominent on the right leg.

Shoulder abduction: The right shoulder initiates abduction with slight elevation of the shoulder girdle. The deltoideus muscle activates and the upper trapezius is simultaneously active. The right scapula is winging. The left shoulder showed physiological activation of muscles, starting with m. deltoideus, m. trapezius and then the lower fixators of the scapula

Trunk flexion: The patient is unable to achieve a full curl up. The spine does not curve much in the cervical and upper thoracic region but shows some curvature in the mid-thoracic section . There is a tendency to lordosis in the lumbar spine.

Neck flexion: The movement is initiated by the protraction of the head rather than by clear flexion but the curvature of the cervical spine does occur once greater range of motion is achieved. The spine does not produce a symmetrical arch but rather breaks at the C5-C6 intervertebral joint.

DSS test according to Kolář

Diaphragm test: the patient has difficulty expanding latero-dorsal sections of the abdominal wall and slight elevation of the right shoulder is present

Intra- abdominal pressure test: the patient has difficulty expanding the lower abdominal wall

Anthropometric measurements

Height in standing: 158 cm

Table No. 19: Final Length of Lower Extemities

Length of LE		
Length	**Left**	**Right**
Anatomical	82 cm	82 cm
Functional		
SIAS- malleolus medialis	87 cm	87 cm
Umbillicus- malleolus medialis	89 cm	89 cm
Thigh	40 cm	40 cm
Middle leg	37 cm	37 cm
Foot	24 cm	24 cm

Table No. 20: Final Circumferences of Lower Extremities

Circumference of lower extremities		
	Left	**Right**
Thigh		
15 cm above kneecap	44,5 cm	44,5 cm
10 cm above kneecap	41 cm	41 cm
Knee joint	37 cm	37 cm
Tuberositas tibiae	32 cm	32 cm
Calf	34 cm	35 cm
Ankle joint	**33** cm	34 cm
Heel	31 cm	31 cm
Foot	22 cm	22 cm

Table No. 21: Final Length of Upper Extremities

Length of upper extremities		
	Left	**Right**
Whole UE	70 cm	70 cm
Humerus	26 cm	26 cm
Forearm	28 cm	28 cm
Hand	20 cm	20 cm

Table No. 22: Final Circumferences of Upper Extremities

Circumference of upper extremities		
	Left	**Right**
Upper arm		
Relaxed	25 cm	25 cm
Contracted	26 cm	26 cm
Elbow joint	23 cm	23 cm
Forearm	18 cm	18 cm
Wrist	**16** cm	15 cm
Metacarpal heads	**20,5** cm	19 cm

Table No. 23: Other Final Measurements

Circumferences		
Head	56 cm	
Thorax	Max.inspire	Max. expire
Mesosternal (average of 3)	**76,5** cm	**72,5** cm
Xiphosternal (average of 3)	**85** cm	**80,5** cm
Hips	110 cm	

Spinal distances

Table No. 24: Final Spinal Distances

Spinal distance test	Result	Normal range
Thomayer's test	**11** cm	0 cm
Schober's distance	**4** cm	5 cm
Stibor's distance	5 cm	7 - 10 cm
Otto's distance	3 cm	≥ 4,5 cm
Cepojev's distance	2 cm	2,5 - 3 cm
Forestier Fleche	1 cm	0 cm
Latero flexion	left side-**17** cm	20-25 cm
	right side – **15** cm	
Cervical spine flexion	3 cm	0 cm

Examination of Shortened Muscles

Table No. 25: Final Shortened Muscles

Length test	**Author**	**Grade**	
		Left	**Right**
Plantar flexors	Janda	1	1
Hip flexors	Janda	1	1
Hip adductors	Janda	0	0
Hamstring muscles	Janda	**1**	1
Paravertebral muscles	Janda	2	
Quadratus lumborum	Janda	1	1
Piriformis	Janda	1	1
Pectoralis major	Kendall		
sternal part		Shortness	Shortness
clavicular part		Shortness	Shortness
Pectoralis minor	Kendall	Shortness	Shortness
M. teres major, m. latissimus dorsi, mm. rhomboid major and minor	Kendall	Shortness	Shortness
Lateral shoulder rotator	Kendall	Slight	Slight
Trapezius cranial part	Janda	**1**	1
M. levator scapulae	Janda	1	**1**
Sternocleidomastoideus	Janda	1	1

ROM examination according to Janda

Table No. 26: Final ROM Examination

	AROM		**PROM**	
	Left	**Rigth**	**Left**	**Right**
Shoulder joint	S 40-0-160 F 160-0-0 T 35-0-130 R 75-0-90	S 35-0-**140** F **140**-0-0 T **25**-0-**120** R **65-0-85**	S 40-0-165 F 160-0-0 T 40-0-135 R 75-0-90	S 35-0-**145** F **145**-0-0 T **30-0-125** R 65-0-90
Elbow joint	S 0-0-140	S 0-0-140	S 0-0-140	S 0-0-140
Forearm	R 90-0-80	R 90-0-80	R 90-0-80	R 90-0-80
Wrist	Not tested due to cannula	S **65**- 0 -75 F 30 -0 -45	Not tested due to cannula	S **75- 0 -85** F 30 -0 -45
Thumb **CM joint** **MP joint** **IP joint**	F 30-0-15 S 30-0-0 Opposition: tip of V S 0-0-50 S 5-0-85	F 30-0-15 S 40-0-0 Opposition: tip of V S 0-0-50 S 5-0-85	F 30-0-15 S 35-0-0 Opposition: tip of V S 0-0-50 S 5-0-85	F 30-0-15 S 40-0-0 Opposition: tip of V S 0-0-50 S 5-0-85
Fingers				
MP joint	II. S 30-0-80 F 20-0-20 III. S 30-0-90 F 20-0-20 VI. S 45-0-90 F 20-0-20 V. II. S 40-0-90 F 20-0-20	II. S 30-0-90 F 25-0-25 III. S 35-0-90 F 20-0-20 VI. S 45-0-90 F 25-0-25 V. II. S 45-0-90 F 20-0-20	II. S 35-0-90 F 20-0-20 III. S 40-0-90 F 20-0-20 VI. S 45-0-90 F 20-0-20 V. II. S 45-0-90 F 20-0-20	II. S 40-0-90 F 30-0-30 III. S 40-0-90 F 25-0-30 VI. S 45-0-90 F 25-0-25 V. II. S 45-0-90 F 20-0-20
IP1 joint	II.S 0-0-80 III. S 0-0-80 VI. S 0-0-90 V. S 0-0-100	II.S **5-0-95** III. S 5-0-**95** VI. S 5-0-**95** V. S 5-0-100	II.S 0-0-80 III. S 0-0-80 VI. S 0-0-90 V. S 0-0-100	II.S **5-0-100** III. S 5-0-**100** VI. S 5-0-**100** V. S 5-0-100
IP2 joint	II.S 0-0-**60** III. S 0-0-65 VI. S 0-0-70 V. S 0-0-65	II.S **5-0-65** III. S **5-0-65** VI. S **5**-0-75 V. S **5**-0-70	II.S 0-0-60 III. S 0-0-70 VI. S 0-0-70 V. S 0-0-70	II.S 5-0-70 III. S 5-0-70 VI. S 10-0-80 V. S 5-0-70
Hip joint	S 15-0-125 F 45-0-20 R 25-0-40	S 15-0-125 F 45-0-20 R 25-0-40	S 15-0-125 F 45-0-20 R 45-0-40	S 15-0-125 F 45-0-20 R 45-0-40
Knee joint	S 0-0-130	S 0-0-130	S 0-0-130	S 0-0-130
Ankle	S **20**-0-45 R 15-0-40	S **20**-0-45 R 20-0-40	S **20**-0-45 R 20-0-40	S 20-0-45 R 20-0-40

Examination of Muscle Strength

Facial muscles

According to Kendall

Grade 0 to 5

Table No. 27: Final Muscle Strength of Facial Muscles

Muscle	**Left**	**Right**
Frontalis	5	5
Corrugator supercilii	4	4
Procerus	4	4
Orbicularis oculi	5	5
Nasalis	3	3
Zygomaticus major	4	4
Levator labií superioris	**3**	**3**
Levator anguli oris	3	3
Mentalis	3	3
Buccinator	3	3
Risorius	5	5
Depressor labii inferioris and platysma	2	2
Depressor anguli oris	**4**	**4**
Pterygoideus medialis and lateralis	3	3

Muscle Strength test of the Trunk and Neck

According to Kendall

Grade 0 to 10

Table No. 28: Final Muscle Strength of the Trunk and Neck

Muscles	Grade
Rectus abdominis, external and internal obliques on the opposite side	7
Upper abdominal -serratus anterior, external oblique, rectus abdominis	8
Lower abdominal muscle test -rectus abdominis	8
Anterior neck flexors-longus capitis, longus colli, rectus capitis	8
Anterolateral neck flexors-SCM, scaleni	**7**
Posterolateral neck extensors –splenius capitis and cervicis, semispinalis capitis and cervicis, cervical erector spinae	8

Muscle Strength Examination of Extremities According to Janda

Table No. 29: Final Muscle Strength of Extremities, Grade 0 to 5

Movement	Left	Right
Abduction of shoulder joint	5	**4**
Flexion of shoulder joint	5	**4**
Internal rotation of shoulder joint	5	4
External rotation of shoulder joint	4	4
Flexion of shoulder joint	5	5
Extension of shoulder joint	5	5
Elevation of shoulders	4	**4**
Forearm pronation	5	5
Forearm supination	5	5
Wrist flexion	Not tested	4
Wrist extension	Not tested	4
Radial duction	Not tested	4
Ulnar duction	Not tested	4
Finger flexion	4	3
Finger extension	4	3
Finger abduction	4	4
Finger adduction	4	4
Flexion of hip joint	5	5
Extension of hip joint	4	4
Abduction of hip joint	5	5
Adduction of hip joint	5	5
IR of hip joint	5	5
ER of hip joint	4	4
Flexion of knee joint	5	5
Extension of knee joint	5	5
Dorsiflexion of ankle joint	5	5
Supination of ankle joint with plantar flexion	**5**	**5**

Neurological Examination of Facial Nerves

Table No. 30: Final Neurological Examination of Facial Nerves

Nerve	Results
Olfactory	The patient was able to identify familiar odors
Optic	The patient was able to read different fonts and sizes. Peripheral vision is in norm.
Oculomotor	Pupils seemed to have normal constriction
Trochlear	The patient follows finger with slight movement of head but upon instruction maintains the position of the head.
Trigeminal	Sensory: the patient could feel when she was touched in both sides of the face and could distinguish between a sharp and dull sensation. Motor: the patient can hold mouth open
Abducens	The patient follows finger with slight movement of head but upon instruction maintains the position of the head.
Facial	Motor: patient can smile, wrinkle face and puff cheeks without difficulty. Sensory: Patient can differentiate between sweet and salty taste.
Vestibulocochlear	Hearing: the patient is able to hear snap of fingers close to her ears.
Glossopharyngeal	Th patient can swallow and say “Ah” without difficulty.
Vagus	Gag reflex can be elicited.
Spinal Accessory	The patient can shrug shoulders.
Hypoglossal	The patient can stick out tongue and move it from side to side.

Peripheral (Deep Tendon) Reflexes

Table No. 31: Final Deep Tendon Reflexes

Reflex	Left	Right
Patellar	Normoreflexia	Normoreflexia
Achilles	Normoreflexia	Normoreflexia
Medioplantar	Normoreflexia	Normoreflexia
Biceps	Normoreflexia	Normoreflexia
Radial	Normoreflexia	Not tested
Triceps	Normoreflexia	Normoreflexia

Sensory system examination

Light touch was used to examine dermatomes of upper and lower extremities. On the upper extremities dermatomes C4, C5, C6, C7, C8, T1, and T2 were examined, on the lower extremities dermatomes L2, L3, L4, L5, S1, and S2. Bilaterally on both upper and lower extremities the patient felt the sensation simultaneously without any abnormalities. Some sensation was decreased in the area of the C7 dermatome on her right hand.

Cortical sensory function

Tactile discrimination: patient could distinguish the number of points on her left thigh and right forearm.

Kinesthetic sensation: The movement was carried out on the metacarpophalangeal joint of the middle finger of the right upper extremity the movement was carried out to about 30º extension and the patient was able to correctly

tell in when the movement initiated, in which direction the movement was carried out, and when it ended.

The movement was also carried out on the metacarphophalangeal joint of the right lower extremity and the movement was carried out to about 30° extension and the patient was able to correctly tell when the movement started, in which direction it was and when it ended.

Graphesthesia: the patient was able to recognize the number drawn on the skin of her left thigh.

N. medianus tests

The tests were performed by the patient's right hand

Circle test- patient manages without difficulty

Fist- patient manages to make a fist, slight difficulty with flexing II. and III. digits

Examination of grips

Table No. 32: Final Examination of Grips

Tests	**Left**	**Right**
Lateral grip	Manages well	Manages well
Pincer grip	Manages	Manages
Grip with terminal opposition of the thumb and index	Manages	Manages
Medium wrap	Manages- there remain limitation with larger heavier objects	Manages- there are limitation with larger heavier objects
Power sphere	Manages- there are limitation with larger heavier objects	Manages- there are limitation with larger heavier objects
Hook grip	Manages well	Manages well

Examination of reflex changes

The examination was performed by palpation.

Skin

Face: signs of microstomia and radial creases around the mouth remain, the skin is well hydrated but there is tightness around the temporomandibular joint and the mouth.

Upper extremities: Slight whiteness of the tips of the fingers and coldness is present on all fingers of the upper extremities, especially on the second and third fingers of the left hand. Resistance of the skin on the palms, dorsum of the hands and all fingers is present, looser and more wrinkled skin appears on the right forearm. Ulcerations on the second and third fingers have decreased in size bilaterally. Slight dryness of the skin near the left elbow remains. There is a scar after carpal tunnel surgery on the base of the right palm. There is a slight resistance of the skin around the right shoulder.

Trunk: the skin drag is increased around the pectoral area, especially around the right shoulder and clavicle. There is an increased resistance of the skin in the area of the thoracic spine bilaterally and the right scapula.

Lower extremities: the patient has varicose veins on her lower extremities, the skin is resistant on the calves slightly in all directions, on the thigh there are restrictions bilaterally around the tensor fasciae latae. The skin on the periphery is limited slightly dorsally and on the planta, the fingers are colder and slightly whiter, there is resistance of the skin present here bilaterally. The ulcerations on the bellies of fingers and laterally on the left pinkie have decreased.

Subskin

Trunk: there was limitation of moving the sub skin on the nape of the neck, on the right side of the back around the right scapula in all directions.

Upper extremities: there is resistance around the right shoulder joint and acrum on of the hands.

Lower extremities: there is resistance on the medial part of the thigh, around the tensor fascia latae especially on the right and the frontal part of the thigh slightly bilaterally.

Fascia

Neck: increased anterior cervical resistance of the fascia in lateral direction both to the right and left side. The resistance was slightly more present on the right

Pectoral fascia : resistance is present on both right and left side

Thoracodorsal fascia: resistance was felt bilaterally in all directions

Fascia of the upper extremities: increased resistance of the fascia on the whole upper extremities, especially on the right side.

Fascia on the lower extremities: increased resistance on the thighs laterally

Metacarpal and metatarsal movement is limited bilaterally

Muscles

Trunk: hypertonus of upper part of m. trapezius, mm. scaleni, m. supraspinatus, m. infraspinatus, m. levator scapulae and short extensors of the craniocervical junction was present bilaterally.

Upper extremities: hypertone of m. biceps brachii and flexors and extensors of the hand was present

Lower extremities: hypertone of mm. adductores was present, m. tensor fasciae latae- more on the right lower extremity, m. rectus femoris and m. triceps surae bilaterally.

Periostal points

Slightly unpleasant feeling when palpating the right clavicle and the right acromion was present.

Scars

The scar at the base of the right palm is relatively well moveable in all directions.

Joint play examination according to Lewit

- Blockage in the sternoclavicular joint on the right was found

- Blockages in the acromioclavicular joint on the right in all directions were found
- Blockages of the right shoulder was found in the dorsal and cranial direction
- Movement of the right scapula was limited
- Blockages of the radiocarpal joints in all directions were found on the right, they were not tested on the left due to the cannula in the dorsal part of the forearm
- Blockages of the peripheral parts of the upper extremities were present bilaterally - the metacarpophalangeal joints, proximal and distal interphalangeal joints in dorsal and lateral directions
- Blockages in the peripheral parts of the lower extremities are present- in the metatarsophalangeal joints in dorsal and lateral direction, the proximal and distal interphalangeal joints bilaterally in dorsal and lateral directions

Scleroderma Health Assessment Questionnaire

Attachment 4

Cochin Hand Function Scale Questionnaire

Attachment 6

Mouth Handicap in Systemic Sclerosis Questionnaire

Attachment 8

Conclusion of the Final Kinesiological Examination

The patient has been hospitalized in Revmatologický ústav for two weeks. Today the patient is well-oriented in time and place and cognitive functions are well maintained. The patient can achieve all ADL and is capable of transfers to her room and the gym. Reduced sensitivity in the C7 dermatome after carpal tunnel operation on her right hand remains, though the scar on the right palm is now more moveable in all directions. Raynaud's phenomenon has been improved on the periphery of the upper extremities, however, the whiteness of the fingertips after cold exposure persists as well as ulcerations, though decreased, remain on the bellies of her fingers on both upper and lower extremities. Currently there is edema in her left forearm and hand most probably causing decreased hand function and reduced ROM.

Movement stereotypes according to Janda remain unchanged, the protrusion of the right scapula remains. However, the range of motion of the right shoulder has improved, most significantly in abduction by about 20°, and during gait the arm swing has also slightly increased in both upper extremities.

The overall gait remains quite stiff, the left leg is bearing more weight and the right is externally rotated. There is a limitation in the external rotation of both hip joints, though she is able to maintain a single leg stance for a few seconds. DSS tests showed a weakness of the deep stabilization system that continues to exist.

Limitations in joint play are mainly visible on the small peripheral joints of the upper and lower extremities. The skin, sub skin and fascia are also mainly limited in these areas as well as on the whole back, pectoral area and area around the right shoulder.

The spinal distances showed limitations in the whole spine, however, slight improvement was seen in spine flexion ROM. Muscle hypertension was palpated on the flexors and extensors of the hands, scaleni, upper trapezius, infraspinatus and supraspinatus.

The entire musculoskeletal system is affected due to structural changes in the skin, limiting physiological movement in individual joints.

3.7 Evaluation of the Effect of Therapy

Evaluation of the Effect of therapy

During the course of the therapy units, improvements in range of motion and overall relaxation of the skin and subcutaneous tissue in the areas targeted by each therapy unit were achieved. The positive outcome depended on the current physical state of the patient, where several therapies were affected by the patient's nausea, edema in the hands after cannula insertions, or stiffness of the whole body after infusions. However, given the diagnosis, it is important to consider the sudden changes and anticipate them.

Changes in examination results are distinguished in the tables by color – bold text.

Evaluation of standing

In standing, there is a change in the position of the lower limbs, where the narrow base has been corrected to a hip-width posture with parallel feet. The position of the shoulders has changed- the shoulders more externally rotated and the protraction of the right shoulder has slightly decreased.

To improve overall posture in the therapies the patient was instructed both verbally and with handling to maintain correct alignment of the body. We also practiced standing in front of a mirror for the patient to have better visual control of how her posture looks. The patient was taught the small foot exercise to improve the position of her feet. This in combination with soft tissue techniques for the skin, sub skin and fascia, relaxation and stretching of muscles, and mobilization of the small joints of the lower extremities might have improved the patient's posture. For the upper extremities, to decrease the protraction of the shoulders, mobilization of the scapula, PNF for upper extremities and scapula as well as soft tissue techniques were used. Verbal instructions affected the patient's posture mostly in the short-term, the gradual effect of local techniques which focused on the individual segments of the patient's body seemed more beneficial for overall posture.

Evaluation of gait

The gait remained relatively unchanged. However, a slightly larger arm swing was seen in the upper extremities and the toe-off phase on both lower extremities is more visible.

The patient was verbally instructed to improve her gait stereotype. This was quite difficult for the patient to do. Improvement in the arm swing is probably correlated with the increase of ROM in the shoulder joints. A large part of the therapy units revolved around treatment of the right shoulder. Techniques such as PNF for strengthening and relaxing muscles of the upper extremities and around the scapula were used along with passive stretching and soft tissue treatment using PIR, soft balling and release of the skin, sub skin and fascia were performed. Improvement of toe- off phase in gait might be due to mobilization of the small foot joints and small foot training. The effect of these techniques was not striking, however slight improvement in toe- off could be seen.

Evaluation of anthropometric measurements and spine distances

Table No. 33: Evaluation of Anthropometric Measurements

Circumferences UE	**Left before**	**Left after**	**Right before**	**Right after**
Wrist	15 cm	**16 cm**	15 cm	15 cm
Metacarpal heads	19 cm	**20,5 cm**	19 cm	**19 cm**
Thorax circumferences	**Max. inspire before**	**Max. inspire after**	**Max. expire before**	**Max. expire after**
Mesosternal (average of 3)	76 cm	**76,5 cm**	73 cm	**72,5 cm**
Xiphosternal (average of 3)	84,5 cm	**85 cm**	81 cm	**80,5 cm**

The circumferences of the left wrist and metacarpal heads have increased, this might be due to the edema after the insertion of cannula for infusions. The therapies used to improve the edema once it appeared was soft balling and stretching the soft tissue- skin and subskin of the hands. These approaches seemed to reduce the edema slightly, yet not fully.

Thorax circumferences have increased by approximately 1 cm in the final examination. During the therapy to improve excursions of the thorax manual techniques such as fascia release (thoracodorsal and pectoral), Kibler's fold for the upper thorax, soft tissue techniques such as passive stretching and PIR to target pectoral muscles, PNF repeated contractions and relaxing techniques (contraction-relaxation) were used. Active breathing exercises were performed, such as localized breathing with and without resistance and active exhalations. The combination of soft tissue techniques and breathing exercises had a positive impact on the results of thorax circumferences.

Table No. 34: Evaluation of Spinal Distances

	Before	**After**
Thomayer's test	13 cm	**11 cm**
Shober's distance	3,5 cm	**4 cm**
Latero-flexion	16 cm	**17 cm**
	14cm	**15 cm**

The spinal distances improved in overall flexion by Thomayer's test, there was 2 cm improvement. Lateral flexion of the spine improved by approximately 1 cm to each side and Shober's distance improved by 0,5 cm.

In the trunk, the treatment of soft tissues was the main focus. The thoracodorsal fascia was stretched and released regularly, as well as the skin was stretched here. This in combination with treating the soft tissue of the lower extremities might have improved the spinal distance results.

Evaluation of ROM examination according to Janda

Table No. 35: Evaluation of ROM Examination

	AROM				**PROM**			
	Left before	**Left after**	**Rigth before**	**Rigth after**	**Left before**	**Left after**	**Right before**	**Right after**
Shoulder joint	S 40-0-160	S 40-0-160	S 30-0-130	S **35**-0-**140**	S 40-0-165	S 40-0-165	S 30-0-130	S **35**-0-**145**
	F 160-0-0	F 160-0-0	F 120-0-0	F **140**-0-0	F 160-0-0	F 160-0-0	F 125-0-0	F **145**-0-0
	T 35-0-130	T 35-0-130	T 20-0-115	T **25**-0-**120**	T 40-0-135	T 40-0-135	T 25-0-120	T 30-0-125
	R 70-0-90	R 75-0-90	R 60-0-80	R **65**-0-**85**	R 70-0-90	R **75**-0-90	R 65-0-90	R 65-0-90
Wrist	Not tested due to cannula	Not tested due to cannula	S 60- 0 -75	S **65**- 0 -75	Not tested due to cannula	Not tested due to cannula	S 70- 0 -80	S **75**- 0 -**85**
			F 30 -0 -45	F 30 -0 -45			F 30 -0 -45	F 30 -0 -45

There were no significant increases in ROM of the small joints of the hands and the ankle. The ROM increased approximately by 5° in the joints of the right hand and stayed the same as during the initial examination in the left hand. This may be due to the oedema after cannula insertion for infusions.

The hands were a big focus of the therapy. IP and MCP joints were mobilized according to Lewit in dorsopalmar and lateral directions, the radiocarpal joint was also mobilized. The skin was treated with soft balling and stretching. The patient was also instructed to do several exercises to improve hand function. PIR of the hand flexors and extensors was done and the patient learned how to perform self-therapy PIR according to Lewit for these muscles. During the last few days of the patient's stay in Revmatologický ústav, she had occupational therapy where they further focused on treating the skin, joints of the hand and overall function. Mostly stretching of the skin and mobilization of the small joints of the hands were effective during therapy.

The ROM of the right shoulder improved most markedly. To improve the ROM of the right shoulder joint the area around the skin, subskin and fascia were treated using Kibler's fold, fascia stretching and soft balling. Mobilization of the first rib and of the scapula, PNF repeated contractions and contraction relaxation diagonals for the

upper extremities and scapula, PIR, as well as passive stretching for pectoral muscles were performed. The biggest improvements in ROM were usually seen after therapies where a combination of stretching and strengthening was performed. Usually a combination of PNF repeated contractions and contraction relaxation was effective. PNF posterior depression of the scapula also seemed to improve shoulder mobility. The muscle strength in the right upper extremity was improved by one grade in abduction and flexion.

Evaluation of reflex changes

Face: signs of microstomia and radial creases around the mouth remain, the skin is slightly more stretchable and is less dry. There is still a resistance of the neck fascia in lateral direction both to the right and left side.

During therapy units the skin of the face was gently stretched, especially around the area of the mouth and temporomandibular joint. The patient was instructed how to massage the face and stretch the skin on her own. She had some difficulty with treating the soft tissues of the face on her own due to Raynaud's phenomenon on her fingers. She however, regularly moisturized her face with creams. She also did facial exercises, at first in the therapy unit, then in front of the mirror as self-therapy. The overall skin texture seemed to have improved but the oral aperture seems to be without change.

Upper extremities: Slight whiteness of the tips of the fingers and coldness is present on all fingers of the upper extremities but Raynaud’s syndrome is less pronounced and the rest of the hands are warmer to the touch. Resistance of the skin on the palms, dorsum of the hands and all fingers is still present bilaterally. But on the right forearm, the skin is more moveable. Ulcerations on the second and third fingers have decreased in size bilaterally. Slight dryness of the skin near the left elbow remains. The scar after carpal tunnel surgery is more moveable in all directions. The resistance of the skin and fascia around the right shoulder remains.

Soft tissue of the upper extremity was treated using Kibler’s fold, soft balling, fascia release, and stretching of the skin. The scar on the base of the right palm was treated with pressure massage. This improved the movability in all directions and

seemed to be effective. During therapy no special technique was used to reduce ulcers on the hands.

Trunk: the skin drag is not as pronounced around the pectoral area, but remains around the right shoulder and clavicle. There are no changes in resistance of the skin in the area of the thoracic spine bilaterally and the right scapula. The thoracodorsal fascia is slightly less resistant in all directions.

Lower extremities: Slightly less resistance was seen on the skin of the calves on the thighs of both lower extremities. The ulcerations on the bellies of fingers and laterally on the left pinkie have decreased slightly.

Both the trunk and lower extremities were treated mainly using fascia release, Kibler's fold, soft balling and stretching of the skin and subskin. The changes in the soft tissues were visible immediately after therapy, however seemed not to last for longer periods of time.

Evaluation of SSc Questionnaires

Scleroderma Health Assessment Questionnaire

This questionnaire assesses the degree of difficulties resulting from systemic scleroderma. The degree of restriction is indicated on a scale of 0 to 100 for 6 questions. After evaluation of the questionnaire before therapy and after therapy, there were no major changes in the patient's responses. The only reply, which showed slight worsening, was to the question How seriously would you rate your illness today?

Cochin Hand Function Scale Questionnaire

The questionnaire evaluates the extent to which impairment of hand function affects everyday life of the patient. The questionnaire consisted of 18 questions. The patient replied mainly that she had no problems in the activities or minor problems. The initial score was 5 and remained the same at the end of the therapy.

Mouth Handicap in Systemic Sclerosis Questionnaire

This questionnaire evaluates the extent to which facial and oral disabilities affect everyday life. There are 12 questions and the patient's score was 12 before the therapy and remained the same after therapy.

4 DISCUSSION

Systemic scleroderma is a rare disease of unknown cause that is currently incurable. Scleroderma affects connective tissue with characteristic stiffening of the skin, Raynaud's phenomenon, or fibrotic changes in internal organs (heart. lungs. gastrointestinal tract,...). The disease is described more in the theoretical part of the thesis

In the theoretical part, I tried to give a simple and brief description of the problems of this disease. In the special part, it was a detailed examination of the patient during the initial kinesiological analysis to determine the appropriate short- and long-term rehabilitation plan and the application of therapy to obtain positive results in the final kinesiological analysis. The final aim of the thesis was to evaluate the effect of therapy based on comparison of the initial and final examinations.

The treated patient was diagnosed with a limited form of systemic sclerosis with interstitial lung involvement. The diagnosis was in 2005 after experiencing issues with healing her right hand after carpal tunnel operation. In the limited form of SSc the progression of the disease is relatively slow and the skin is affected mainly on the extremities (distally from the elbows and knees, face and neck). One of the first symptoms of the disease is Raynaud's phenomenon followed by swelling of the fingers and skin thickening.

A series of vasodilator infusions with prostavasin in a reduced dose was started to reduce Raynaud's phenomenon and digital ulceration of the patient. The digital ulcerations are in mild regression after completion of 15 infusions along with relief of Raynaud's phenomenon. Often in SSC patients ADL are limited due to grasp impairments and for this reason it is crucial to prevent the formation of deformities of the hands. During physiotherapy of the hands I tried to focus on improving the range of motion in the small joints of the hands. My therapies included stretching of the soft tissues of the hand, mobilization of the small joints of the hands according to Lewit, and therapeutic exercises to improve fine motor skills. This approach seemed to increase the ROM of small joints of the hands by about 5° on average. Bongi et al. evaluated the effect of the combination of connective tissue massage, Mc Mennell joint manipulation, and a home exercise program. The outcome was an improvement of hand function and quality of life in patients with SSc (Maddali Bongi et al., 2009). Other studies have

demonstrated adequate results with the use of therapeutic exercises. The combination of joint mobilization with therapeutic exercises may be a beneficial way to treat impairments of the hands (Santos Cardozo Roque et al., 2021). Positive thermotherapy is often used before exercise to decrease pain and improve the flexibility of collagen. In a study by Sandqvist et al., the use of paraffin wax followed by exercise compared to only exercise showed improvement in mobility, stiffness, and skin elasticity in the group with paraffin wax followed by exercise in comparison to the exercise alone group (Sandqvist et al., 2004). During the patient's therapy I did not focus on treating digital ulceration. To reduce digital ulcerations more attentively studies show immersion ultrasound therapy or extracorporeal shockwave therapy to be effective (Scaturro et al., 2023)(Saito et al., 2016). Towards the end of the patient's stay in Revmatologický ústav edema in the left hand appeared after cannula insertion in the dorsum of the hand. Mainly soft balling was used to treat the edema with minor results. Bongi et al. observed manual lymphatic drainage to be effective in reducing hand volume and improvement in hand function after a five-week manual lymphatic drainage program for SSc patients (Bongi et al., 2011).

Other typical symptoms of SSc appear in the orofacial area where the oral opening reduces in size which interferes with eating, speaking, oral hygiene and dental treatment. The skin of the patient showed microstomia and radial creases around the mouth. To assess how SSc affects the patient's cheeks and mouth the questionnaire Mouth Handicap in Systemic Sclerosis was used. This, however, is a subjective way of measuring mouth handicap and mouth opening. To measure maximal mouth opening objectively a caliper could be used to determine the distance between the incisal edge of the upper and lower incisors (Gulia et al., 2018). With my patient we treated the orofacial area with soft tissue massage and facial exercises. During the final examination, the patients' replies in the Mouth Handicap in Systemic Sclerosis Questionnaire were almost identical to the initial examination showing no major improvement in mouth function. Pizzo et al. evaluated the effect of an exercise program in SSc patients with microstomia. The patients performed mouth stretching exercises and oral augmentation exercises with a stick of wood twice daily for 18 weeks. This program improved mouth opening of all participants (Pizzo et al., 2003). According to Maddali- Bongi an intervention program using Kabat's technique,

connective tissue massage and special kinesiotherapy for the face proved to be effective (Maddali-Bongi et al., 2010).

To improve overall ROM of the whole body, hypertone and shortened muscles were treated using PIR and passive stretching, as well as in the case of upper extremities PNF contraction-relaxation technique. The muscle shortness did not improve significantly, but was slightly reduced for example in the left trapezius and right m. levator scapulae by one grade. Stretching increases elasticity of the muscles and tendons, improves joint mobility, helps preserve correct postural alignment, breathing, and efficient movement. It also helps to relax the body, it is a prevention from injuries and reduces fatigue (Špiritović & Tomčík, 2017).

The musculoskeletal system of SSc patients should be treated to reduce or prevent impairments in all other areas of the body (Mugii et al., 2018). Regular exercise improves cardiovascular efficiency and improvement in physical condition. With my patient we did not perform any aerobic training and mainly exercises to improve the activation of the deep stabilization system were used. DeOliviera et al. assessed the effectiveness of aerobic and aerobic with resistance exercise. Exercise tolerance, aerobic capacity, walking distance, muscle strength, and muscle function as well as HRQL results were improved after participation in the programs. SSc patients with and without pulmonary involvement benefited from aerobic exercises (de Oliveira et al., 2017).

70 to 90 % of SSc individuals have pulmonary complications. The main aim of pulmonary rehabilitation is to reduce breathing difficulty and increase physical strength of SSc patients. Maintaining the flexibility of the rib cage, maintaining the flexibility of breathing-related muscles, improving breathing-related muscle strength, strengthening of lower body muscles, aerobic exercise, understanding proper breathing methods, and learning self-care and self-training methods are important to achieve benefits from pulmonary rehabilitation (Mugii et al., 2018). During therapy, we focused on the improvement of the breath wave by using localized breathing with and without resistance and active exhalations. Mobilization of the first rib and relaxation of the pectoral muscles, trapezius, sternocleidomastoid, and scalene muscles were performed. The fascia and skin on the thorax were treated as well leading to an improvement in chest excursions of the patient. However, no aerobic exercises were performed and neither were exercises for strengthening of the lower body from which the patient might

have benefited. Some form of aerobic and resistance training might have also been useful to improve overall physical condition as well as circulation.

Although the number of studies is small and most are not supported by control measurements, it is important to emphasize that the vast majority of studies have concluded that rehabilitation treatment scleroderma is important. Based on the studies evaluated and my own work, it seems that exercise, connective tissue massage and mobilization improves ROM in joints of SSc patients.

5 CONCLUSION

The aim of this bachelor thesis was to introduce the diagnosis systemic scleroderma and to develop a case report of one systemic sclerosis patient. I had the opportunity to work with her during a month-long practical internship at Revmatologický ústav in Prague. The whole experience was very interesting and beneficial. During the practical internship I was able to translate theoretical knowledge into practice. With the help of experienced physiotherapists we create an individual therapy plan for the patient. For the examinations and therapies I used methods and approaches that I was introduced to during my bachelor's degree in physiotherapy at UK FTVS and added the recommended specialized questionnaires. The patient was very cooperative, she was happy to come to the therapies, which was very positive for the patient's progress. On the other hand, the patient experienced some complications after the last cannula insertion causing edema which might have affected the final examination. Of the set therapy goals, we managed to influence the range of motion of the upper limbs, most notably in the right shoulder joint. Using soft tissue techniques, skin and subcutaneous mobility was improved on the limbs, to a greater extent in the upper limbs. The patient subjectively perceived some improvement in personal hygiene. Overall, she felt in better condition after the therapies.

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7. APPENDICES

Appendix 1-Ethics Commitee Approval

Appendix 2- Informed Consent Form

Appendix 3 Scleroderma Health Assessment Questionnaire Initial

Appendix 4- Scleroderma Health Assessment Questionnaire Final

Appendix 5-Cochin Hand Function Scale Questionnaire Initial

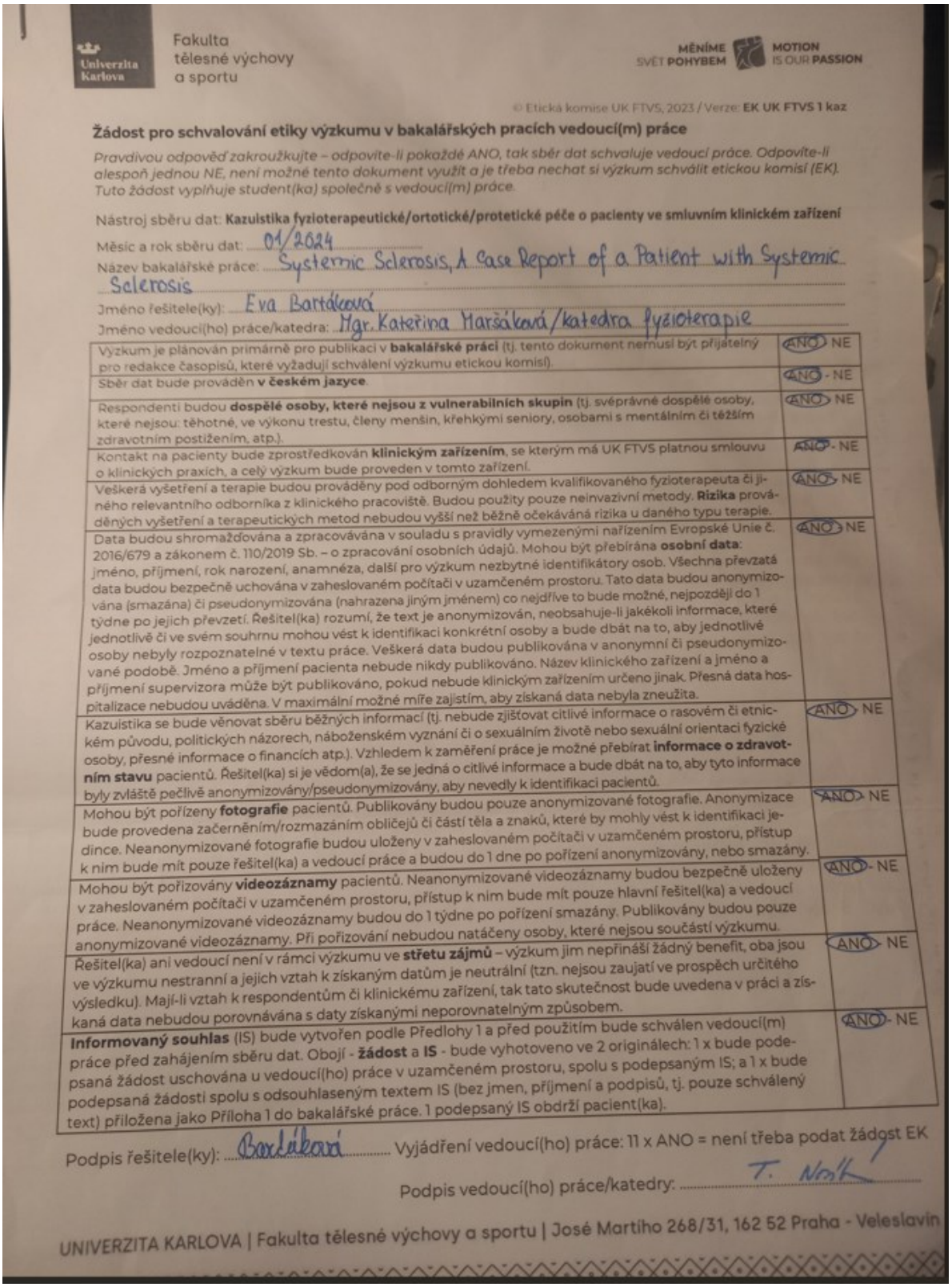
Appendix 7- Mouth Handicap in Systemic Sclerosis Questionnaire Initial

Appendix 8- Mouth Handicap in Systemic Sclerosis Questionnaire Final

Appendix 9 –List of Tables

Appendix 10- List of Images

Appendix 1-Ethics Commitee Approval



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Fakulta tělesné výchovy a sportu

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Žádost pro schvalování etiky výzkumu v bakalářských pracích vedoucí(m) práce

Pravdivou odpověď zakroužkujte – odpovíte-li pokaždé ANO, tak sběr dat schvaluje vedoucí práce. Odpovíte-li alespoň jednou NE, není možné tento dokument využít a je třeba nechat si výzkum schválit etickou komisí (EK). Tuto žádost vyplňuje student(ka) společně s vedoucí(m) práce.

Nástroj sběru dat: **Kazuistika fyzioterapeutické/ortotické/protetické péče o pacienty ve smluvním klinickém zařízení**

Měsíc a rok sběru dat: 01/2024

Název bakalářské práce: Systemic Sclerosis, A Case Report of a Patient with Systemic Sclerosis

Jméno řešitele(ky): Eva Bartáková

Jméno vedoucí(ho) práce/katedra: Mgr. Kateřina Maršáková/katedra fyzioterapie

Výzkum je plánován primárně pro publikaci v **bakalářské práci** (tj. tento dokument nemusí být přijatelný pro redakce časopisů, které vyžadují schválení výzkumu etickou komisí).	(ANO) NE
Sběr dat bude prováděn **v českém jazyce**.	(ANO) - NE
Respondenti budou **dospělé osoby, které nejsou z vulnerabilních skupin** (tj. svéprávné dospělé osoby, které nejsou: těhotné, ve výkonu trestu, členy menšin, křehkými seniory, osobami s mentálním či těžším zdravotním postižením, atp.).	(ANO) NE
Kontakt na pacienty bude zprostředkován **klinickým zařízením**, se kterým má UK FTVS platnou smlouvu o klinických praxích, a celý výzkum bude proveden v tomto zařízení.	(ANO) - NE
Veškerá vyšetření a terapie budou prováděny pod odborným dohledem kvalifikovaného fyzioterapeuta či jiného relevantního odborníka z klinického pracoviště. Budou použity pouze neinvazivní metody. **Rizika** prováděných vyšetření a terapeutických metod nebudou vyšší než běžně očekávána rizika u daného typu terapie.	(ANO) NE
Data budou shromažďována a zpracovávána v souladu s pravidly vymezenými nařízením Evropské Unie č. 2016/679 a zákonem č. 110/2019 Sb. – o zpracování osobních údajů. Mohou být přebírána **osobní data**: jméno, příjmení, rok narození, anamnéza, další pro výzkum nezbytné identifikátory osob. Všechna převzatá data budou bezpečně uchována v zaheslovaném počítači v uzamčeném prostoru. Tato data budou anonymizována (smazána) či pseudonymizována (nahrazena jiným jménem) co nejdříve to bude možné, nejpozději do 1 týdne po jejich převzetí. Řešitel(ka) rozumí, že text je anonymizován, neobsahuje-li jakékoli informace, které jednotlivě či ve svém souhrnu mohou vést k identifikaci konkrétní osoby a bude dbát na to, aby jednotlivé osoby nebyly rozpoznatelné v textu práce. Veškerá data budou publikována v anonymní či pseudonymizované podobě. Jméno a příjmení pacienta nebude nikdy publikováno. Název klinického zařízení a jméno a příjmení supervizora může být publikováno, pokud nebude klinickým zařízením určeno jinak. Přesná data hospitalizace nebudou uváděna. V maximální možné míře zajistím, aby získaná data nebyla zneužita.	(ANO) NE
Kazuistika se bude věnovat sběru běžných informací (tj. nebude zjišťovat citlivé informace o rasovém či etnickém původu, politických názorech, náboženském vyznání či o sexuálním životě nebo sexuální orientaci fyzické osoby, přesné informace o financích atp.). Vzhledem k zaměření práce je možné přebírat **informace o zdravotním stavu** pacientů. Řešitel(ka) si je vědom(a), že se jedná o citlivé informace a bude dbát na to, aby tyto informace byly zvláště pečlivě anonymizovány/pseudonymizovány, aby nevedly k identifikaci pacientů.	(ANO) NE
Mohou být pořízeny **fotografie** pacientů. Publikovány budou pouze anonymizované fotografie. Anonymizace bude provedena začerněním/rozmazáním obličejů či částí těla a znaků, které by mohly vést k identifikaci jedince. Neanonymizované fotografie budou uloženy v zaheslovaném počítači v uzamčeném prostoru, přístup k nim bude mít pouze řešitel(ka) a vedoucí práce a budou do 1 dne po pořízení anonymizovány, nebo smazány.	(ANO) NE
Mohou být pořizovány **videozáznamy** pacientů. Neanonymizované videozáznamy budou bezpečně uloženy v zaheslovaném počítači v uzamčeném prostoru, přístup k nim bude mít pouze hlavní řešitel(ka) a vedoucí práce. Neanonymizované videozáznamy budou do 1 týdne po pořízení smazány. Publikovány budou pouze anonymizované videozáznamy. Při pořizování nebudou natáčeny osoby, které nejsou součástí výzkumu.	(ANO) - NE
Řešitel(ka) ani vedoucí není v rámci výzkumu ve **střetu zájmů** – výzkum jim nepřináší žádný benefit, oba jsou ve výzkumu nestranní a jejich vztah k získaným datům je neutrální (tzn. nejsou zaujatí ve prospěch určitého výsledku). Mají-li vztah k respondentům či klinickému zařízení, tak tato skutečnost bude uvedena v práci a získaná data nebudou porovnávána s daty získanými neporovnatelným způsobem.	(ANO) NE
Informovaný souhlas (IS) bude vytvořen podle Předlohy 1 a před použitím bude schválen vedoucí(m) práce před zahájením sběru dat. Obojí - **žádost** a **IS** - bude vyhotoveno ve 2 originálech: 1 x bude podepsaná žádost uschována u vedoucí(ho) práce v uzamčeném prostoru, spolu s podepsaným IS; a 1 x bude podepsaná žádosti spolu s odsouhlaseným textem IS (bez jmen, příjmení a podpisů, tj. pouze schválený text) přiložena jako Příloha 1 do bakalářské práce. 1 podepsaný IS obdrží pacient(ka).	(ANO) - NE

Podpis řešitele(ky): Bartáková Vyjádření vedoucí(ho) práce: 11 x ANO = není třeba podat žádost EK

Podpis vedoucí(ho) práce/katedry: T. Novák

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Appendix 2- Informed Consent Form

UNIVERZITA KARLOVA
FAKULTA TĚLESNÉ VÝCHOVY A SPORTU
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Předloha 1: Informovaný souhlas

INFORMOVANÝ SOUHLAS

Vážená paní, vážený pane,

v souladu se Všeobecnou deklarací lidských práv, nařízením Evropské Unie č. 2016/679 a zákonem č. 110/2019 Sb. – o zpracování osobních údajů a dalšími obecně závaznými právními předpisy (jakož jsou zejména Helsinská deklarace, přijatá 18. Světovým zdravotnickým shromážděním v roce 1964 ve znění pozdějších změn (Fortaleza, Brazílie, 2013); Zákon o zdravotních službách a podmínkách jejich poskytování (zejména ustanovení § 28 odst. 1 zákona č. 372/2011 Sb.) a Úmluva o lidských právech a biomedicíně č. 96/2001, jsou-li aplikovatelné), Vás žádám o souhlas s prezentováním a uveřejněním výsledků vyšetření a průběhu terapie prováděné v rámci praxe .....,kde Vás příslušně kvalifikovaná osoba seznámila s Vaším vyšetřením a následnou terapií. Výsledky Vašeho vyšetření, průběh Vaší terapie, případně anonymizované relevantní informace Vaší anamnézy budou publikovány v rámci bakalářské práce na UK FTVS, s názvem .....

Cílem této bakalářské práce je .....

Získané údaje, průběh a výsledky terapie, případně fotodokumentace či video, budou uveřejněny v bakalářské práci v anonymizované či pseudonymizované podobě. Osobní data nebudou zveřejněna a budou uchována v anonymní podobě, nebo smazána nejdéle do 1 týdne po jejich převzetí. Budou-li pořízeny fotografie, budou anonymizovány do 1 dne po pořízení; bude-li pořízen videozáznam, bude anonymizován do 1 týdne po pořízení. V maximální možné míře zajistím, aby získaná data nebyla zneužita.

Jméno a příjmení řešitele ..... Podpis:.....

Jméno a příjmení osoby, která provedla poučení[1]..... Podpis:.....

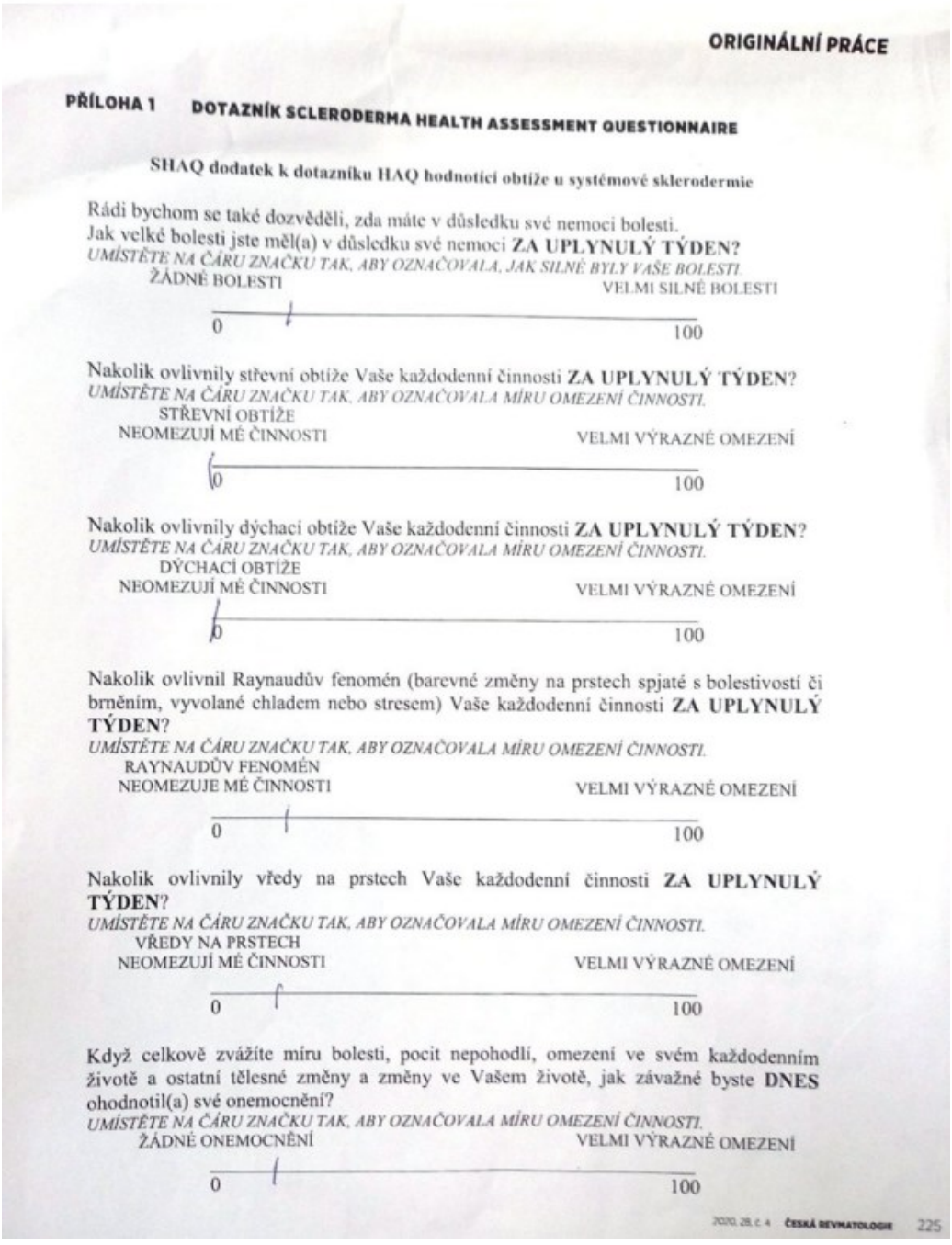
Prohlašuji a svým níže uvedeným vlastnoručním podpisem potvrzuji, že dobrovolně souhlasím s prezentováním a uveřejněním výsledků vyšetření a průběhu terapie ve výše uvedené bakalářské práci, a že mi osoba, která provedla poučení, osobně vše podrobně vysvětlila, a že jsem měl(a) možnost si řádně a v dostatečném čase zvážit všechny relevantní informace, zeptat se na vše podstatné a že jsem dostal(a) jasné a srozumitelné odpovědi na své dotazy. Byl(a) jsem poučen(a) o právu odmítnout prezentování a uveřejnění výsledků vyšetření a průběhu terapie v bakalářské práci nebo svůj souhlas kdykoli odvolat bez represí, a to písemně zasláním Etické komisi UK FTVS, která bude následně informovat řešitele. Dále potvrzuji, že mi byl předán jeden originál vyhotovení tohoto informovaného souhlasu.

Místo, datum .....

Jméno a příjmení pacienta(ky) ..... Podpis pacienta(ky): .....

[1] *Je-li řešitel s pacientem v závislém postavení, poučení provádí jiná příslušně kvalifikovaná osoba*

Appendix 3 Scleroderma Health Assessment Questionnaire Initial



PŘÍLOHA 1 DOTAZNÍK SCLERODERMA HEALTH ASSESSMENT QUESTIONNAIRE

SHAQ dodatek k dotazníku HAQ hodnotící obtíže u systémové sklerodermie

Rádi bychom se také dozvěděli, zda máte v důsledku své nemoci bolesti.
Jak velké bolesti jste měl(a) v důsledku své nemoci **ZA UPLYNULÝ TÝDEN?**
UMÍSTĚTE NA ČÁRU ZNAČKU TAK, ABY OZNAČOVALA, JAK SILNÉ BYLY VAŠE BOLESTI.

ŽÁDNÉ BOLESTI — VELMI SILNÉ BOLESTI

0 — 100

Nakolik ovlivnily střevní obtíže Vaše každodenní činnosti **ZA UPLYNULÝ TÝDEN**?
UMÍSTĚTE NA ČÁRU ZNAČKU TAK, ABY OZNAČOVALA MÍRU OMEZENÍ ČINNOSTI.

STŘEVNÍ OBTÍŽE
NEOMEZUJÍ MĚ ČINNOSTI — VELMI VÝRAZNÉ OMEZENÍ

0 — 100

Nakolik ovlivnily dýchací obtíže Vaše každodenní činnosti **ZA UPLYNULÝ TÝDEN**?
UMÍSTĚTE NA ČÁRU ZNAČKU TAK, ABY OZNAČOVALA MÍRU OMEZENÍ ČINNOSTI.

DÝCHACÍ OBTÍŽE
NEOMEZUJÍ MĚ ČINNOSTI — VELMI VÝRAZNÉ OMEZENÍ

0 — 100

Nakolik ovlivnil Raynaudův fenomén (barevné změny na prstech spjaté s bolestivostí či brněním, vyvolané chladem nebo stresem) Vaše každodenní činnosti **ZA UPLYNULÝ TÝDEN**?
UMÍSTĚTE NA ČÁRU ZNAČKU TAK, ABY OZNAČOVALA MÍRU OMEZENÍ ČINNOSTI.

RAYNAUDŮV FENOMÉN
NEOMEZUJE MĚ ČINNOSTI — VELMI VÝRAZNÉ OMEZENÍ

0 — 100

Nakolik ovlivnily vředy na prstech Vaše každodenní činnosti **ZA UPLYNULÝ TÝDEN**?
UMÍSTĚTE NA ČÁRU ZNAČKU TAK, ABY OZNAČOVALA MÍRU OMEZENÍ ČINNOSTI.

VŘEDY NA PRSTECH
NEOMEZUJÍ MĚ ČINNOSTI — VELMI VÝRAZNÉ OMEZENÍ

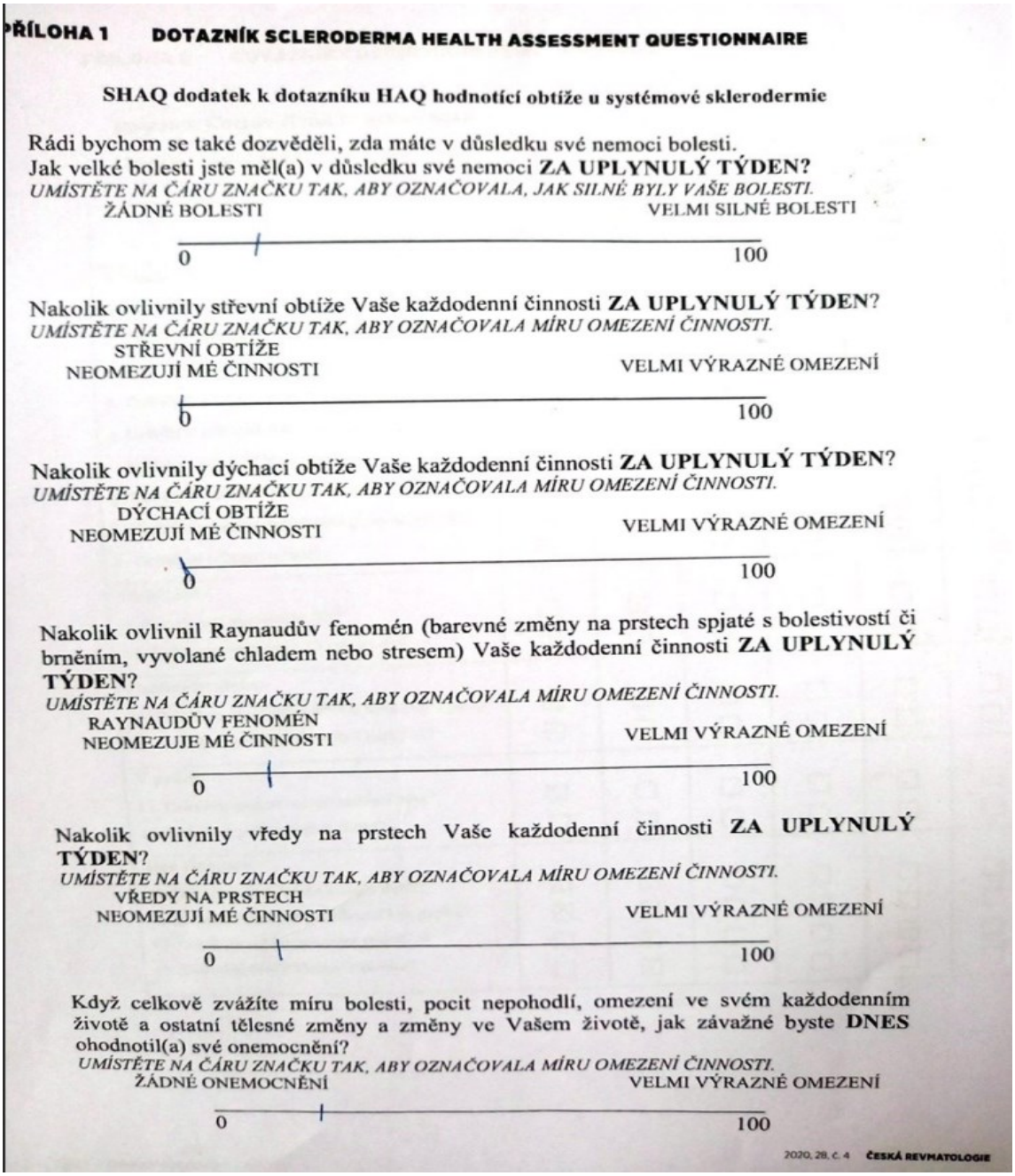
0 — 100

Když celkově zvážíte míru bolesti, pocit nepohodlí, omezení ve svém každodenním životě a ostatní tělesné změny a změny ve Vašem životě, jak závažné byste **DNES** ohodnotil(a) své onemocnění?
UMÍSTĚTE NA ČÁRU ZNAČKU TAK, ABY OZNAČOVALA MÍRU OMEZENÍ ČINNOSTI.

ŽÁDNÉ ONEMOCNĚNÍ — VELMI VÝRAZNÉ OMEZENÍ

0 — 100

Appendix 4- Scleroderma Health Assessment Questionnaire Final



PŘÍLOHA 1 **DOTAZNÍK SCLERODERMA HEALTH ASSESSMENT QUESTIONNAIRE**

SHAQ dodatek k dotazníku HAQ hodnotící obtíže u systémové sklerodermie

Rádi bychom se také dozvěděli, zda máte v důsledku své nemoci bolesti.
Jak velké bolesti jste měl(a) v důsledku své nemoci **ZA UPLYNULÝ TÝDEN?**
UMÍSTĚTE NA ČÁRU ZNAČKU TAK, ABY OZNAČOVALA, JAK SILNÉ BYLY VAŠE BOLESTI.
ŽÁDNÉ BOLESTI — VELMI SILNÉ BOLESTI
0 — 100

Nakolik ovlivnily střevní obtíže Vaše každodenní činnosti **ZA UPLYNULÝ TÝDEN**?
UMÍSTĚTE NA ČÁRU ZNAČKU TAK, ABY OZNAČOVALA MÍRU OMEZENÍ ČINNOSTI.
STŘEVNÍ OBTÍŽE NEOMEZUJÍ MÉ ČINNOSTI — VELMI VÝRAZNÉ OMEZENÍ
0 — 100

Nakolik ovlivnily dýchací obtíže Vaše každodenní činnosti **ZA UPLYNULÝ TÝDEN**?
UMÍSTĚTE NA ČÁRU ZNAČKU TAK, ABY OZNAČOVALA MÍRU OMEZENÍ ČINNOSTI.
DÝCHACÍ OBTÍŽE NEOMEZUJÍ MÉ ČINNOSTI — VELMI VÝRAZNÉ OMEZENÍ
0 — 100

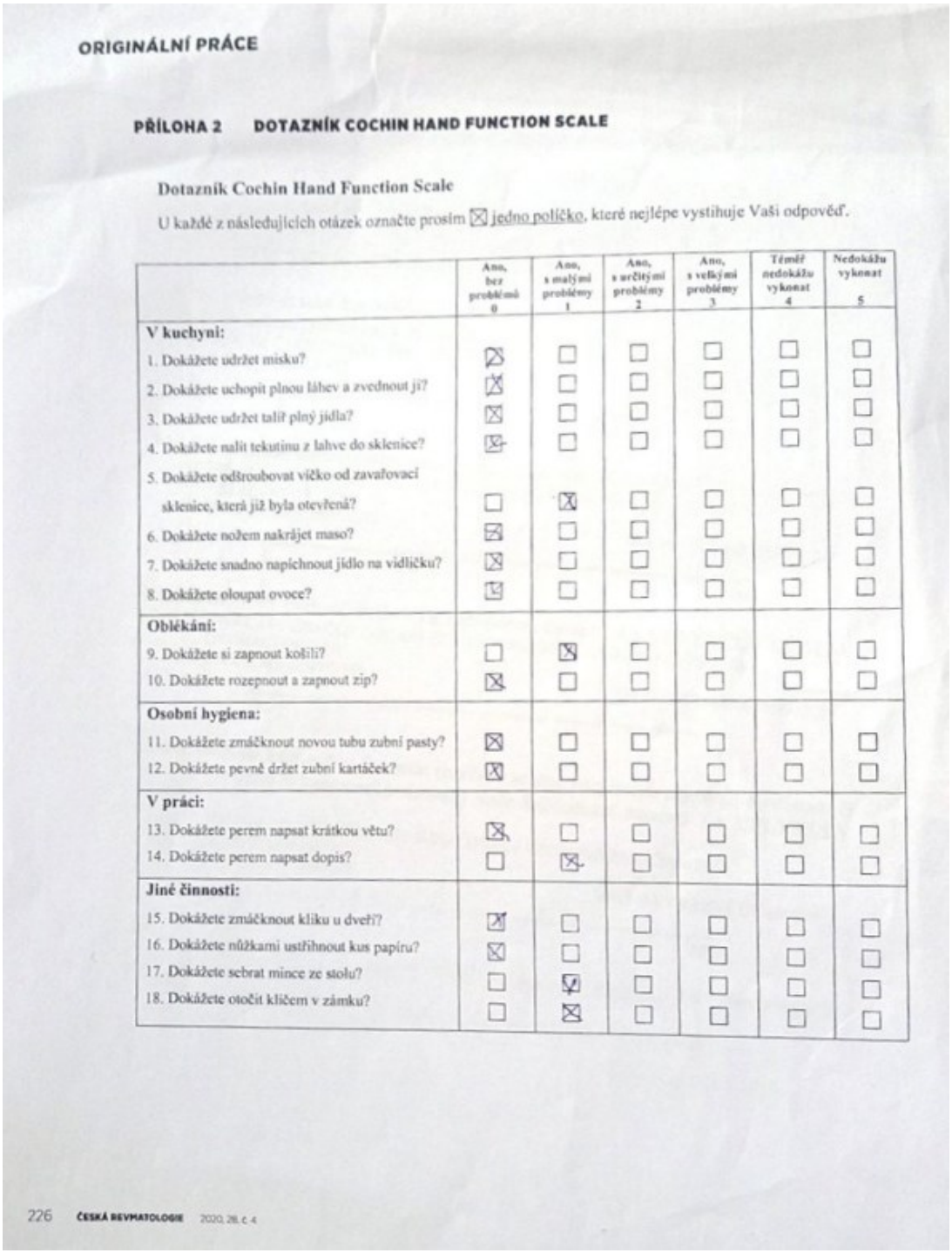
Nakolik ovlivnil Raynaudův fenomén (barevné změny na prstech spjaté s bolestivostí či brněním, vyvolané chladem nebo stresem) Vaše každodenní činnosti **ZA UPLYNULÝ TÝDEN**?
UMÍSTĚTE NA ČÁRU ZNAČKU TAK, ABY OZNAČOVALA MÍRU OMEZENÍ ČINNOSTI.
RAYNAUDŮV FENOMÉN NEOMEZUJE MÉ ČINNOSTI — VELMI VÝRAZNÉ OMEZENÍ
0 — 100

Nakolik ovlivnily vředy na prstech Vaše každodenní činnosti **ZA UPLYNULÝ TÝDEN**?
UMÍSTĚTE NA ČÁRU ZNAČKU TAK, ABY OZNAČOVALA MÍRU OMEZENÍ ČINNOSTI.
VŘEDY NA PRSTECH NEOMEZUJÍ MÉ ČINNOSTI — VELMI VÝRAZNÉ OMEZENÍ
0 — 100

Když celkově zvážíte míru bolesti, pocit nepohodlí, omezení ve svém každodenním životě a ostatní tělesné změny a změny ve Vašem životě, jak závažné byste **DNES** ohodnotil(a) své onemocnění?
UMÍSTĚTE NA ČÁRU ZNAČKU TAK, ABY OZNAČOVALA MÍRU OMEZENÍ ČINNOSTI.
ŽÁDNÉ ONEMOCNĚNÍ — VELMI VÝRAZNÉ OMEZENÍ
0 — 100

2020, 28, č. 4 ČESKÁ REVMATOLOGIE

Appendix 5-Cochin Hand Function Scale Questionnaire Initial



ORIGINÁLNÍ PRÁCE

PŘÍLOHA 2 DOTAZNÍK COCHIN HAND FUNCTION SCALE

Dotazník Cochin Hand Function Scale

U každé z následujících otázek označte prosím ☒ jedno políčko, které nejlépe vystihuje Vaši odpověď.

	Ano, bez problémů 0	Ano, s malými problémy 1	Ano, s určitými problémy 2	Ano, s velkými problémy 3	Téměř nedokážu vykonat 4	Nedokážu vykonat 5
V kuchyni:						
1. Dokážete udržet misku?	☒	☐	☐	☐	☐	☐
2. Dokážete uchopit plnou láhev a zvednout ji?	☒	☐	☐	☐	☐	☐
3. Dokážete udržet talíř plný jídla?	☒	☐	☐	☐	☐	☐
4. Dokážete nalít tekutinu z lahve do sklenice?	☒	☐	☐	☐	☐	☐
5. Dokážete odšroubovat víčko od zavařovací sklenice, která již byla otevřená?	☐	☒	☐	☐	☐	☐
6. Dokážete nožem nakrájet maso?	☒	☐	☐	☐	☐	☐
7. Dokážete snadno napíchnout jídlo na vidličku?	☒	☐	☐	☐	☐	☐
8. Dokážete oloupat ovoce?	☒	☐	☐	☐	☐	☐
Oblékání:						
9. Dokážete si zapnout košili?	☐	☒	☐	☐	☐	☐
10. Dokážete rozepnout a zapnout zip?	☒	☐	☐	☐	☐	☐
Osobní hygiena:						
11. Dokážete zmáčknout novou tubu zubní pasty?	☒	☐	☐	☐	☐	☐
12. Dokážete pevně držet zubní kartáček?	☒	☐	☐	☐	☐	☐
V práci:						
13. Dokážete perem napsat krátkou větu?	☒	☐	☐	☐	☐	☐
14. Dokážete perem napsat dopis?	☐	☒	☐	☐	☐	☐
Jiné činnosti:						
15. Dokážete zmáčknout kliku u dveří?	☒	☐	☐	☐	☐	☐
16. Dokážete nůžkami ustřihnout kus papíru?	☒	☐	☐	☐	☐	☐
17. Dokážete sebrat mince ze stolu?	☐	☒	☐	☐	☐	☐
18. Dokážete otočit klíčem v zámku?	☐	☒	☐	☐	☐	☐

Appendix 6-Cochin Hand Function Scale Questionnaire Final

ORIGINÁLNÍ PRÁCE

PŘÍLOHA 2 DOTAZNÍK COCHIN HAND FUNCTION SCALE

Dotazník Cochin Hand Function Scale

U každé z následujících otázek označte prosím ☒ jedno políčko, které nejlépe vystihuje Vaši odpověď.

	Ano, bez problémů 0	Ano, s malými problémy 1	Ano, s určitými problémy 2	Ano, s velkými problémy 3	Téměř nedokážu vykonat 4	Nedokážu vykonat 5
V kuchyni:						
1. Dokážete udržet misku?	☒	☐	☐	☐	☐	☐
2. Dokážete uchopit plnou láhev a zvednout ji?	☒	☐	☐	☐	☐	☐
3. Dokážete udržet talíř plný jídla?	☒	☐	☐	☐	☐	☐
4. Dokážete nalít tekutinu z lahve do sklenice?	☒	☐	☐	☐	☐	☐
5. Dokážete odšroubovat víčko od zavařovací sklenice, která již byla otevřená?	☐	☒	☐	☐	☐	☐
6. Dokážete nožem nakrájet maso?	☒	☐	☐	☐	☐	☐
7. Dokážete snadno napíchnout jídlo na vidličku?	☒	☐	☐	☐	☐	☐
8. Dokážete oloupat ovoce?	☒	☐	☐	☐	☐	☐
Oblékání:						
9. Dokážete si zapnout košili?	☐	☒	☐	☐	☐	☐
10. Dokážete rozepnout a zapnout zip?	☒	☐	☐	☐	☐	☐
Osobní hygiena:						
11. Dokážete zmáčknout novou tubu zubní pasty?	☒	☐	☐	☐	☐	☐
12. Dokážete pevně držet zubní kartáček?	☒	☐	☐	☐	☐	☐
V práci:						
13. Dokážete perem napsat krátkou větu?	☒	☐	☐	☐	☐	☐
14. Dokážete perem napsat dopis?	☐	☒	☐	☐	☐	☐
Jiné činnosti:						
15. Dokážete zmáčknout kliku u dveří?	☒	☐	☐	☐	☐	☐
16. Dokážete nůžkami ustřihnout kus papíru?	☒	☐	☐	☐	☐	☐
17. Dokážete sebrat mince ze stolu?	☐	☒	☐	☐	☐	☐
18. Dokážete otočit klíčem v zámku?	☐	☒	☐	☐	☐	☐

Appendix 7- Mouth Handicap in Systemic Sclerosis Questionnaire Initial

PŘÍLOHA 3 DOTAZNÍK MOUTH HANDICAP IN SYSTEMIC SCLEROSIS

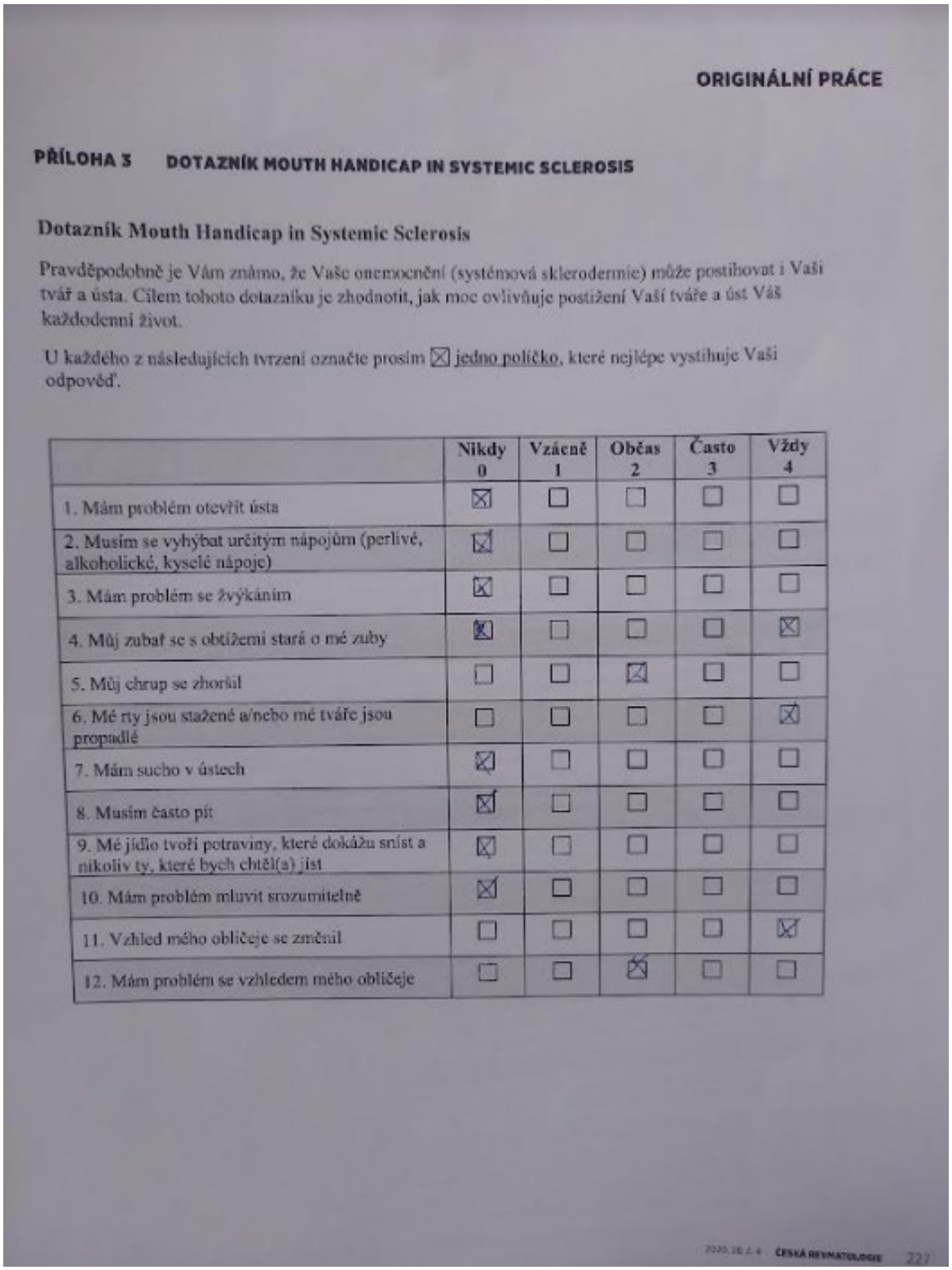
Dotazník Mouth Handicap in Systemic Sclerosis

Pravděpodobně je Vám známo, že Vaše onemocnění (systémová sklerodermie) může postihovat i Vaši tvář a ústa. Cílem tohoto dotazníku je zhodnotit, jak moc ovlivňuje postižení Vaší tváře a úst Váš každodenní život.

U každého z následujících tvrzení označte prosím ☒ jedno políčko, které nejlépe vystihuje Vaši odpověď.

	Nikdy 0	Vzácně 1	Občas 2	Často 3	Vždy 4
1. Mám problém otevřít ústa	☒	☐	☐	☐	☐
2. Musím se vyhýbat určitým nápojům (perlivé, alkoholické, kyselé nápoje)	☒	☐	☐	☐	☐
3. Mám problém se žvýkáním	☒	☐	☐	☐	☐
4. Můj zubař se s obtížemi stará o mé zuby	☐	☐	☐	☐	☒
5. Můj chrup se zhoršil	☐	☐	☒	☐	☐
6. Mé rty jsou stažené a/nebo mé tváře jsou propadlé	☐	☐	☐	☐	☒
7. Mám sucho v ústech	☒	☐	☐	☐	☐
8. Musím často pít	☒	☐	☐	☐	☐
9. Mé jídlo tvoří potraviny, které dokážu sníst a nikoliv ty, které bych chtěl(a) jíst	☒	☐	☐	☐	☐
10. Mám problém mluvit srozumitelně	☒	☐	☐	☐	☐
11. Vzhled mého obličeje se změnil	☐	☐	☐	☐	☒
12. Mám problém se vzhledem mého obličeje	☐	☐	☒	☐	☐

Appendix 8- Mouth Handicap in Systemic Sclerosis Questionnaire Final



PŘÍLOHA 3 DOTAZNÍK MOUTH HANDICAP IN SYSTEMIC SCLEROSIS

Dotazník Mouth Handicap in Systemic Sclerosis

Pravděpodobně je Vám známo, že Vaše onemocnění (systémová sklerodermie) může postihovat i Vaši tvář a ústa. Cílem tohoto dotazníku je zhodnotit, jak moc ovlivňuje postižení Vaší tváře a úst Váš každodenní život.

U každého z následujících tvrzení označte prosím ☒ jedno políčko, které nejlépe vystihuje Vaši odpověď.

	Nikdy 0	Vzácně 1	Občas 2	Často 3	Vždy 4
1. Mám problém otevřít ústa	☒	☐	☐	☐	☐
2. Musím se vyhýbat určitým nápojům (perlivé, alkoholické, kyselé nápoje)	☒	☐	☐	☐	☐
3. Mám problém se žvýkáním	☒	☐	☐	☐	☐
4. Můj zubař se s obtížemi stará o mé zuby	☒	☐	☐	☐	☒
5. Můj chrup se zhoršil	☐	☐	☒	☐	☐
6. Mé rty jsou stažené a/nebo mé tváře jsou propadlé	☐	☐	☐	☐	☒
7. Mám sucho v ústech	☒	☐	☐	☐	☐
8. Musím často pít	☒	☐	☐	☐	☐
9. Mé jídlo tvoří potraviny, které dokážu sníst a nikoliv ty, které bych chtěl(a) jíst	☒	☐	☐	☐	☐
10. Mám problém mluvit srozumitelně	☒	☐	☐	☐	☐
11. Vzhled mého obličeje se změnil	☐	☐	☐	☐	☒
12. Mám problém se vzhledem mého obličeje	☐	☐	☒	☐	☐

Appendix 9 –List of Tables

Appendix 10- List of Images