

Evidence-Based Physical Therapy Practice for Patients with Scleroderma

A Case Report

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Matthew W. Attia

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APPROVAL SHEET

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Matthew Wade Attia

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Sharon Irish Bevins, PhD, PT
Committee Chair

Jacqueline van Duijn, DPT, OCS

This case report has been examined by the signatories, and we find that both the content and the form meet acceptable presentation standards of scholarly work in the above mentioned discipline.

Table of Contents

Title Page.....	i
Approval Sheet.....	ii
Table of Contents.....	iii
List of Tables.....	iv
Abstract.....	v
Introduction.....	1
Purpose.....	4
Case Description.....	5
Patient presentation.....	5
Patient history & system review.....	5
Examination.....	5
Objective measurements.....	8
Strength.....	8
Soft tissue mobility.....	8
Posture.....	9
Range of motion (ROM).....	9
Other examination recommendations.....	10
Interventions.....	10
Problems, Goals, and Plan of care.....	18
Outcomes.....	21
Discussion.....	23
Conclusion.....	27
References.....	28

List of Tables

Table 1. ACR/EULAR 2013 Systemic Sclerosis Classification.3

Table 2. Initial evaluation of active cervical ROM.10

Table 3. Interventions Performed Date of Initial Evaluation (October 29th, 2013). ..12

Table 4. Interventions Performed on Last Visit (November 26th, 13).13

Table 5. Plan of Care at Initial Evaluation (October 29th, 2013).20

Table 6. Problem list, Goals, and Assessment.22

Abstract

Scleroderma i.e. systemic sclerosis is a rare rheumatic autoimmune disease characterized by fibroblast dysfunction with excessive collagen deposition leading to vasculopathy and subsequent dysfunction throughout the body. This case report addresses the care provided to a 71 year-old Caucasian female who was treated for cervical dysfunction in an outpatient orthopedic clinic. The symptoms experienced by patients with the diffuse progressive form of the disease are diverse and systemic. Physical therapists should ensure that patient care is multidisciplinary and has an emphasis on prevention. It should also be focused on delaying functional limitations, treating exacerbated symptoms, and gathering baseline information to track disease progression and guide further treatment. Current literature and evidence-based research is very limited regarding physical therapy and our potential interventions to this patient population. This case report identifies the treatment provided to this patient, as well as recommendations on providing current and research-based interventions specific to a patient with a diagnosis of systemic sclerosis.

Introduction

The term “scleroderma” is an umbrella term for multiple sub groups of the connective tissue & vascular disease systemic sclerosis (SSc). Scleroderma can be either localized or systemic; localized scleroderma has a more favorable prognosis than systemic scleroderma as it only affects the integumentary system (“umm.edu”, 2012). Systemic scleroderma can present as either limited or diffuse cutaneous SSc. Limited SSc affects the skin, esophagus, and distal joints, while the much more severe diffuse SSc is progressive and compromises the majority of the internal organs and the integumentary system. Patient presentation and disease characteristics of diffuse SSc, also known as progressive systemic sclerosis (PSSc), includes skin thickening, cutaneous ulcerations, joint pain, contractures, gastrointestinal complications, esophageal dysmobility, a compromised respiratory system, kidney disease, and a decrease in cardiac function (Goodman, 2009). The increased risk of mortality in PSSc has a magnitude of 250% in comparison with an age and sex-matched general population (Elhai, 2011).

Systemic sclerosis is classified as a rheumatic autoimmune disease and has previously been subcategorized as a disease of connective tissue metabolism. Current research is challenging conventional thinking to consider SSc as a vascular disease (Matucci-Cerinic, 2013). Progressive SSc is different from other rheumatic diseases in that patients with the disease display systemic vasculopathy and fibrosis. Universally throughout the body, the intima of blood vessels, the pericapillary space, and the interstitium of the skin are filled with extracellular type 1 collagen (Warrington, Nair, Carbone, Kang, Postlethwaite, 2006.; Goodman, 2009).

The hallmark signs of systemic sclerosis include three physiological characteristics: small vessel vasculopathy, production of autoantibodies, and a hyperactive fibroblast dysfunction (Hoogen et al., 2013). Type I collagen is the major collagen of tendons, skin, ligaments, cornea, and many interstitial connective tissues, with the exception of very few tissues such as hyaline cartilage, brain, and vitreous body. The hallmark fibroblast dysfunction leads to unregulated type I collagen deposition. In healthy individuals, type I collagen provides appropriate tensile stiffness (Gelse, Poschl, & Aigner, 2003). Due to the deposition of collagen within the extracellular space, the intima of blood vessels systemically, and the interstitium of skin; fibrosis occurs resulting in a decline in function throughout the body.

The classification of SSc has been recently updated by the American College of Rheumatology/European League Against Rheumatism (ACR/EULAR) from the original classification by the American College of Rheumatology of 1980. The new classification criterion is more sensitive to patients in the early phase of the disease process and allows for more patients to be classified correctly (Hoogan et al, 2013).

Table 1

ACR/EULAR 2013 Systemic Sclerosis Classification

ITEM	SUB-ITEM	SCORE
Skin thickening of the fingers of both hands extending past the level of the metacarpophalangeal joint		9
Skin thickening of the fingers	Puffy fingers	2
	Sclerodactyly of the fingers	4
Fingertip lesions	Digital tip ulcers	2
	Fingertip pitting scars	3
Talangiectasia		2
Abnormal nailfold capillaries		2
Pulmonary arterial hypertension/ Interstitial lung disease		2
Reynaud's phenomenon		3
SSc-related autoantibodies		3
A score of 9 is required to be diagnosed with SSc. More details available (Hoogan et al, 2013)		

Scleroderma has the highest mortality rate of any of the rheumatic diseases. There is currently no cure for PSSc. The prognosis of PSSc is dependent on the level of systemic involvement. The leading causes of mortality found in patients with PSSc include pulmonary hypertension, cardiac dysfunction, and renal disease. The prevalence of PSSc within the United States ranges from 300,000-400,000 or 2 to 10 per 1,000,000. It affects women more than men with a seven to one ratio, and has a slightly higher prevalence in the African-American population (Goodman, 2009; Brandenstein, 1999).

Purpose. Systemic sclerosis is a rare disease that presents differently in affected patients. Based on these factors, available evidence based research literature for physical therapy intervention for patients with PSSc is very limited. Current treatment recommendations and guidelines exist for the medical management of scleroderma (Kowal-Bielecka, 2009); however, they do not exist for physical therapy interventions. The lack of available information makes it difficult for a physical therapist to optimally treat a patient with a diagnosis of systemic sclerosis. The purpose of this case report is to discuss the effects of the disease process relevant to physical therapy, evidence-based physical therapy interventions relative to SSc, and SSc-appropriate patient care.

Due to the variable presentation of the disease, physical therapy intervention will vary on a case-by-case basis. Physical therapists need to administer validated and evidence-based evaluations and interventions based on professional guidelines to deliver quality patient care. The current available literature specific to educating physical therapists concerning scleroderma is limited. Therefore, it is important for

the physical therapist to keep in mind the pathophysiology and disease process of scleroderma when treating the functional mobility of patients diagnosed with PSSc.

Case Description

Patient presentation. The case patient is a 71 year-old Caucasian female with a medical diagnosis of Scleroderma for a duration of 5 years. The patient was referred to physical therapy by her rheumatologist for neck pain. The patient was past the average age of disease onset, which is typically the early to middle forties.

Patient history & system review. The patient was seen in an outpatient physical therapy setting in which the patient's subjective medical history was recorded via a medical history questionnaire. The questionnaire was positive for arthritis, vascular disease, previous deep vein thrombosis (DVT), recent weight loss or gain, headaches, and anxiety. The patient stated that her reason for obtaining her referral was neck dysfunction, and chief complaints secondary to her SSc included GI disturbances, and difficulty using her hands. The patient had degenerative joint disease and osteoarthritis of the bilateral cervical spine confirmed by X ray.

Examination. Based on the patient presentation, objective and functional measures were taken pertaining to the patient's cervical dysfunction. Neck symptoms are noted to be present in nearly 90% of patients diagnosed with SSc (Barnett, 2005). Symptoms typically include tightening of the skin of the neck but may include limitations of ROM and decreased strength. The Neck Disability Index (NDI) was chosen by the physical therapist to evaluate the patient's functional status. This test is considered to have moderately high validity (0.69-0.70) and test-retest reliability (0.89) (Vernon & Moir, 1992). The raw score on the NDI was 13

with a 26% calculated score, which indicates mild to moderate dysfunction. A functional status assessment revealed independence without difficulty in her activities of daily living prior to her chief complaint of neck symptoms. At the time of the initial evaluation, the patient reported that her level of independence was modified independent with moderate restrictions due to pain. The disease process of SSc includes widespread deposition of collagen, and involvement of small vessel vasculopathy. Due to this, all structures of the neck need consideration in addition to an orthopedic cause for the patient's symptoms. Based on the known disease process of SSc, considerations need to include possible dysfunction of the cervical lymphatic system, the thyroid gland, salivary glands and ducts etc., as the dysfunction may be related to a non-orthopedic origin and thus require appropriate referral.

Although not completed in this case, a more thorough evaluation of patients with SSc should be administered due to the global effects of the disease process. The list of available standardized tests is extensive, and choosing a test is highly variable and based on symptom presentation. Scleroderma population-validated tests regarding disability assessment include the Scleroderma Health Assessment Questionnaire (SHAQ), Symptom Burden Index (SBI), and Cochin Hand Function Scale (Pope, 2011; Rannou, 2007). Due to the known pathophysiology of systemic sclerosis and the scope of physical therapy, the physical therapy outpatient facility is an appropriate setting for these objective measures of function to be taken and monitored.

It is recommended that pulmonary function testing should take place annually for patients with SSc (Khanna, Gladue, Mclaughlin, et al., 2013). It therefore seems appropriate for the physical therapist to be involved in the evaluation and management of cardio-pulmonary function. The Cambridge Pulmonary Hypertension Outcome Review is reliable and valid for patients with SSc. It contains a symptoms scale, and measures for activity limitations, and quality of life (Gornberg-Maitland, 2008). Pulmonary function testing should take place to establish a baseline, monitor the progression of the disease, and provide grounds for early intervention.

Hand dysfunction, including tightening of the skin, flexion contractures of the joints, digital ulcers, loss of strength, and pain have been found to be some of the most common and easily recognizable symptoms of scleroderma (Goodman, 2009; www.scleroderma.org, 2013). Hand-specific functional disability assessments that are validated for this population include the Cochin Hand Function Scale, the Arthritis Hand Function Test, and the Hand Mobility In Scleroderma Test (Pope, 2011; Rannou, 2007). Joint, tendon, muscle health, and the mobility of the skin are compromised in this population secondary to the deposition of collagen around and within the soft tissue structures. The deposition of the collagen leads to a cascading effect of muscular disuse that leads to atrophy. Inflammatory processes, combined with disuse, also lead to abnormal changes in joint surfaces and joint deformities. Healthy synovial joints require movement in order to nourish the articular cartilage of a joint. The articular cartilage on the contacting surfaces of the bones of a joint is necessary to reduce the coefficient of friction between the articulating surfaces.

When the synovium of a joint and the articular cartilage are disrupted, pain, inflammation, and decreased range of motion will occur (Levangie & Norkin, 2005). Scleroderma affects the joint in multiple ways. The end result is limited range of motion that leads to structural changes and decreased function. It would appear logical that structural changes of the joints will also lead to alterations in muscle length tension relationships that will lead to further strength deficits. Physical therapists need to be aware of this disease process, and understand that aggressive therapy interventions may lead to an exacerbated inflammatory response, and cause detrimental effects, similar to what is seen in patients with rheumatoid arthritis. Physical therapists should assess joint mobility and focus on preventing or maintaining affected joint range of motion.

Objective measurements.

Strength. Manual muscle testing demonstrated decreased strength of the cervical musculature at 3+/5 in flexion and side-bending, as well as shoulder muscle weakness of the shoulder adductors and flexors at 3/5 and 4-/5 respectively. Deep neck flexor muscular strength and endurance was not assessed, although research confirms that reduced performance of this muscle group is associated with increased risk of neck pain (Falla, Gwendolen, & Hodges, 2004). Manual Muscle testing has been shown to be a valid measure of strength with a strong correlation to EMG and dynamometer testing, and has test-retest and inter-rater reliability ranging from 82% to 97% (Cuthbert & Goodheart, 2007).

Soft tissue mobility. Soft tissue mobility was noted to be moderately restricted in the erector spinae, suboccipital, and upper trapezius musculature. A

meta-analysis revealed that the inter-rater reliability of detecting soft tissue changes was strong with a Cohen's Kappa (K value) of 0.03 (Stochkendhal, M., Christensen, H., Hartvigsen, J., Vach, W., Haas, M., Hestbaek, L. et al., 2006).

Posture. Moderate forward head and rounded shoulder posture was noted. There is a lack of agreement regarding the validity, reliability, and significance of forward head posture in the literature. Greater forward head posture was found to be correlated to a greater level of disability (Yip, Chiu, & Poon, 2008). However, Silva, Punt, Sharples, Vilas-Boas & Johnson (2008), state that the reliability and validity of assessing forward head posture by observation is unclear, as well as the therapeutic effect of improving forward head posture on neck pain.

Range of motion (ROM). ROM was measured with a goniometer for rotation and an inclinometer for lateral side bending and flexion. Williams et al (2009) stated, "The single inclinometer and Spin-T goniometer have both good reliability and concurrent validity but require further evaluation." (p. 153). A thorough literature review reported that "The CROM has the advantage over the single inclinometer and goniometer"; however, the author goes on to say there can be no strong recommendation for any tool... and the goniometer is practical, is accepted by clinicians, and has a reliability ICC value of > 0.76 (Jordan, 2000).

Table 2

Initial evaluation of active cervical ROM

Active Motion Tested	Range of Motion in degrees
Flexion	52
Left Rotation	26
Right Rotation	41
Left Side Bending	25
Right Side Bending	20

Other examination recommendations. Considering the disease progression and common manifestations, objective measures should have been taken regarding hand and cardio-pulmonary function. Practical tests and measures include grip strength, finger range of motion assessment, pulmonary function tests, and cardiopulmonary endurance tests such as the six-minute walk test or submaximal exercise testing. Vitals, including heart rate, respiratory rate, and blood pressure, should be monitored throughout patient care. Each of the above would establish baseline information to monitor progress, indicate need for services, and help guide physical therapy interventions.

Interventions. Physical therapy has successfully helped patients with PSSc manage the following symptoms: regression of joint range of motion, respiratory pathology, decreased cardiac function, decreasing skin elasticity, and pain. The above list of manageable symptoms of PSSc are all either directly related to life

expectancy, or directly correlated to functional independence and quality of life (Casale, Buonocore, & Matucci-Cerinic, 1997; Joven, Almodovar, Carmona, Carreira, 2009; Ngian, Stevens, Prior, Gabbay, Roddy, Tran, ... Nikpour, 2012; Pinto, 2011; Tyndall, Bannert, Vonk, Airo, Cozzi, Carreira,...Ulrich, 2010; Yuen, 2012). Skilled physical therapy is essential to ensure patients with scleroderma do not aggravate the inflammation of joints, and that exercise is performed at a safe level considering compromised cardiopulmonary health.

Interventions that were administered to the patient included gentle cervical strengthening and ROM exercise activities, manual interventions of cervical traction and soft tissue mobilization techniques, and modalities that included moist heat and cryotherapy. Interventions performed to the case patient on the day of the initial evaluation, and at the last therapy session administered are provided in Table 3 and Table 4 below. Evidence for the applied interventions to the cervical spine in patients with SSc is limited. However, there is evidence-based literature that supports positive results from relevant interventions to other parts of the body in patients with SSc.

Table 3

Interventions Performed Date of Initial Evaluation (October 29th, 2013)

Interventions	Dosage and Details
Range of Motion: Chest Lift-Chin Tuck	Time Elapsed: 4 Minutes, Repetitions: 10, Sets: 2.
Isotonics: Shoulder Shrugs	Time Elapsed: 4 Minutes, Weight – Pounds: 0, Repetitions: 10, Sets: 2.
Manual Interventions To: <ul style="list-style-type: none"> • Erector Spinae • Suboccipitals • Upper Trapezius 	Time Elapsed: 24 Minutes, Tx Depth: Moderate, Side: Bilateral, Technique: Myofascial Release.
Manual Traction To: Cervical Spine	Time Elapsed: 8 Minutes, Clinical Use: Post Activity Technique 1: Distraction Technique 2: Distraction/R Rotation/Flexion Technique 3: Distraction/ L Rotation/Flexion
Modalities: Moist Hot Pack	Time Elapsed: 10 Minutes, Location: Cervical Region, Clinical Use: Pre Activity, Skin Integrity Post: Intact.

Table 4

Interventions Performed on Last Visit (November 26th, 13).

Interventions	Dosage and Details
Active Range of Motion and Strengthening:	
<ul style="list-style-type: none"> • Chest Lift-Chin Tuck 	Time Elapsed: 4 Minutes, Repetitions: 10, Sets: 2.
<ul style="list-style-type: none"> • Shoulder Flexion 	Time Elapsed: 3 Minutes, Weight – Pounds: 0, Repetitions: 10, Sets: 2, Position: supine, Additional Detail: With Stick.
<ul style="list-style-type: none"> • Shoulder Shrugs 	Time Elapsed: 4 Minutes, Weight – Pounds: 0, Repetitions: 10, Sets: 2.
Flexibility:	
Levator Scapulae Musculature	Time Elapsed: 4 Minutes, Repetitions: 3, Side: Bilateral, Technique: Passive, Body Position, Seated, Hold: 15 Seconds
Manual Interventions To:	
<ul style="list-style-type: none"> • Erector Spinae • Scaleni • Suboccipitals • Upper Trapezius 	Time Elapsed: 32 Minutes, Tx Depth: Moderate, Side: Bilateral, Technique: Myofascial Release.
Manual Traction To:	
Cervical Spine	Time Elapsed: 8 Minutes, Clinical Use: Post Activity Technique 1: Distraction Technique 2: Distraction/R Rotation/Flexion Technique 3: Distraction/ L Rotation/Flexion
Modalities:	
Moist Hot Pack	Time Elapsed: 10 Minutes, Location: Cervical Region, Clinical Use: Pre Activity, Skin Integrity Post: Intact.

It has been shown that a combination of connective tissue massage, joint manipulation, and home exercise significantly improves fist closure, measurements of finger range of motion, and Cochin hand functional disability scale scores of the hands in patients with SSc (Bongi et al. 2009). It is reasonable to assume that similar interventions applied to the cervical spine and neck of a patient with SSc would produce similar results. This study demonstrated improved efficacy of the aforementioned interventions compared to home exercise alone. Oral aperture tightness and decreased size, known as microstomia, is a common side effect of facial soft tissue sclerosis. Collagen deposits in the perioral tissues prevent normal opening. A study conducted by Yuen et al. showed how stretching and exercise could increase lost oral aperture size in patients with SSc. A non-exercise group displayed a slight loss of aperture size at a 3 month follow-up, and the exercise group showed a significant amount of improvement (Yuen, 2012).

Muscular strength improved after a 12-week exercise program containing resistance and aerobic training; specifically improving 1RM leg press and bench press, back pull and hand grip strength, and overall muscle function (Pinto, 2011). This indicated that strength gains can be made by patients suffering from systemic sclerosis with the diagnosis of ILD, which will improve their ability to perform functional daily activities. The parameters of the interventions were poorly defined; however, the important take-away presented was that this population achieved the benefits of strengthening exercises and it carried over to improved function.

The cardio-respiratory system can be greatly affected in patients with diffuse SSc. In multiple studies, interstitial lung disease (ILD), pulmonary hypertension, and

cardiac pathology have been identified as leading causes of death (Ngain, et al., Joven, et al., Tyndall, et al., 2010). The involvement of the lungs has a direct effect on both quality of life and life expectancy. A Cochrane review conducted by Holland and Hill (2008) confirmed that physical exercise significantly improved pulmonary function related to ILD. This review demonstrated that physical therapy interventions have directly improved one of the highest causes of mortality and disability in patients with systemic sclerosis. Additionally, exercise training, specifically aerobic and resistance training, has been shown to improve pulmonary hypertension (Arena, 2011), resting heart rate, aerobic threshold, and time-to-exhaustion (Pinto, 2011). Dyspnea, decreased forced vital capacity, decreased lung volume, and fatigue are signs and symptoms experienced by patients with SSc. Also pertaining to patients with SSc and a diagnosis of interstitial lung disease, a case report of a 54-year old female demonstrated that a three-time-per-week cycle-ergometer exercise training program, lasting 6 weeks and progressed from 50% to 80% of peak workload, increased oxygen consumption and workload at anerobic threshold by 4.2 mL·kg⁻¹·min, and six-meter walk test by 101m (Shoemaker, 2009). As mentioned previously in this report, functional ability, pulmonary function, and exercise tolerance, assessed by measures such as the SHAQ and 6-minute walk test, SF-36, and manual muscle testing have all shown to be positively influenced with physical exercise (Chernev et al. 2009; Antolioli et al., 2009; Oliveira, 2009).

It is important for the physical therapist to consider the increased risk of osteoporosis and thus risk of fracture when implementing an exercise program for patients with scleroderma (Avouac, 2012). A two to six-time increase in

osteoporosis was identified in this population with several factors and influences including decreased activity secondary to symptoms, and medications. Physical therapists should be aware of this increased risk for fracture, and educate patients on safety and recommended activities to increase bone density. No studies were found that confirm this population is capable of offsetting their risk of osteoporosis with exercise.

Although not present in the case patient, myositis, dermatomyositis and polymyositis are inflammatory processes of the musculature that are commonly seen in autoimmune diseases such as SSc. An acute case of myositis in a patient diagnosed with diffuse systemic sclerosis who was admitted to a hospital for acute weakness received treatment of physical therapy. Interventions included resistive strengthening exercise, range of motion exercise, and gross and fine-motor training. Interventions were performed in conjunction with steroid treatment. The assessment demonstrated that the patient's strength improved from 1-2/5 to 4-5/5 in the upper and lower extremities. The authors reported that more research was needed to examine and describe the appropriate frequency, intensity, type, and timing of the exercise (Chernev, Gustafson, & Medina-Bravo, 2009).

Thermal modalities such as: superficial heat, paraffin wax, and ultrasound can be used in conjunction with low force prolonged stretching to increase the length of the high collagen content fibrotic tissues (Casale, 1997). Collagen displays plastic deformation to these types of loads, which would promote increased joint range of motion and soft tissue length (Casale, 1997). Paraffin bath treatments in conjunction with hand exercises have been shown to improve joint mobility,

decrease perceived hand stiffness, and improve skin elasticity (Sandqvist, 2004). It is important to remember that there is a highly compromised vascular system in this population. For this reason, burns have a higher potential for occurrence and superficial heating should be monitored accordingly. The available evidence-based literature regarding modalities is almost exclusively concerning paraffin wax bath treatments for the hands. A series of case studies, and a randomized controlled trial demonstrated that paraffin wax hand baths of 20 minutes duration prior to and in conjunction with hand exercises demonstrated statistically significant improvements in measures such as joint range of motion, grip and pinch strength, perceived stiffness, and function items on the Arthritis Hand Function Scale (Sandqvist et al., 2004; Manuso & Poole, 2009). It is reasonable to assume that superficial moist heating applied to the case patient's neck in a fashion similar to that of the above mentioned case study may improve the results of the range of motion and strengthening interventions. All interventions administered to the case patient were centered on the patient's cervical dysfunction. However, the case patient reported dysfunction of her hands during the initial evaluation that interfered with activities of daily living, making it pertinent for the attending physical therapist to include appropriate evaluation and management of hand dysfunction to the plan of care.

Myofascial release techniques are another intervention utilized by physical therapists to increase or maintain ROM (Hironobu, 2013). However, there is limited evidence supporting that it is beneficial in decreasing pain and increasing joint ROM (Masi, 2012). There needs to be further research to demonstrate the efficacy of this

intervention in patients with altered inflammatory responses, such as patients with systemic sclerosis. This intervention also could prove to be effective in the treatment of connective tissue adhesions and palpable tendon friction rubs. Tendon friction rubs are described as coarse cracking and crepitus felt on palpation. The pathology of tendon friction rubs in SSc is attributed to non-inflammatory fibrous deposits on the surface of tendon sheaths and the overlying fascia (Rodnan & Medsger, 1968). Tendon friction rubs have been attributed to a more severe disease progression including: increased risk for developing renal, cardiac, and GI involvement as well as a reduced survival rate (Dore, 2013). Understanding that the presentation of this symptom correlates to more severe organ involvement should aid the physical therapist in determining the need for referral or additional organ system monitoring.

Problems, Goals, and Plan of Care. The treating physical therapist indicated that per her clinical judgment, a combination of the patient's age and comorbidities i.e. the diagnosis of scleroderma, the interventions selected would be of low intensity and consist of what was considered to be conservative treatment. The patient's identified problems include difficulty with ADL's, decreased flexibility, pain, range of motion, decreased soft tissue mobility, and decreased strength. The treating physical therapist indicated that slower than typical healing times were anticipated, which was reflected in the documented plan of care.

Based on available evidence-based literature and the known disease process of SSc, goals addressing hand and cardio-pulmonary function should be included, as

those systems are within the physical therapist's scope of practice and directly effect functional independence and quality of life (Casale, 1997; Antonioli, 2009).

The physical therapist responsible for the plan of care determined that the patient would be seen 3 times per week for one month. These recommendations were based on the physical therapist's clinical judgment without availability of clear recommendations regarding this patient population in the literature.

Aforementioned studies identified treatment plans that ranged from single treatment interventions to plans consisting of sixty-minute sessions three times per week. There were, however, no comparisons or conclusions that indicated a frequency or treatment duration recommendation.

It is unclear from the available documentation if the physical therapist modified the plan of care based on the patient's diagnosis of SSc. Administered care led to decreased functional limitations and progressed towards developed goals regarding the patient's cervical dysfunction. However, the systemic nature of the patient's disease, and the known presentation of SSc would make alterations to include addressing problems such as increased risk for osteoporosis, decreased functional endurance, and declining hand function appropriate.

A copy of the plan of care created at the initial evaluation is provided below in Table 5. Goals established as part of the plan of care are detailed in Table 6. The original language and level of detail are preserved in the tables.

Table 5

Plan of Care at Initial Evaluation (October 29th, 2013).

Assessment:	The client tolerated today's treatment/therapeutic activity with minimal complaints of pain and difficulty. In my professional opinion, this client requires skilled physical therapy in conjunction with home exercise program to address the problems and achieve the goals outlined below (Table 5). Overall rehabilitation potential is good. The patient and/or family were educated regarding their diagnosis, prognosis, and related pathology. The client exhibits good understanding and performance of the therapeutic activity and instructions outlines in this skilled rehabilitation session.
Impairments Identified:	ADL's. Flexibility. Pain. Range of Motion. Soft Tissue Mobility. Strength.
Recommendations: Skilled Intervention To:	Decrease Pain. Improve Function. Increase Range of Motion. Increase Strength.
Treatment Emphasis to Focus on:	Range of Motion/Mobility Improvements. Muscle Function Improvements. Pain Relief.
Plan: Amount, Frequency and Duration:	It is recommended that the client attend rehabilitative therapy for 3 visits a week with and expected duration of 1 month. Interventions during the course of treatment will be directed toward addressing the problems and achieving the goals previously outlined (Table 5)
Therapeutic Contents:	Active Range of Motion Activities. Aerobic Conditioning: Upper Body Ergometer. Client Education. Home Exercise Program. Joint Mobilization Techniques. Manual Therapy Techniques. Modalities: As Needed. Neuromuscular Re-education. Self Care/Home Management. Soft Tissue Mobilization Techniques. Stretching/Flexibility Activities. Therapeutic Exercise.

Outcomes. One month after the patient initiated physical therapy, all initial objective measures were re-assessed. Improvements were found in all areas tested with progress made on all goals. The patient was able to increase cervical ROM in left rotation to match that of the ipsilateral side, strength improved in all tested cervical and upper extremity planes, and soft tissue mobility improved. Table 6 documents the initial problem list, goals, progress, and the physical therapists assessment. The language and level of detail used by the treating physical therapist is maintained. The patient subjectively reported a decrease in pain although a formalized scale was not utilized, in addition to reporting decreased limitations in activities such as driving and carrying light objects. Standardized assessment tools such as the SHAQ, and visual pain analog scale are beneficial to document progress more accurately. Based on the progress made, patient compliance and stated understanding of HEP, as well the patient scheduling to go out of town, the patient was discharged from physical therapy.

Table 6

Problem list, Goals, and Assessment.

Problem	Goal	Level at initial visit 10/29/13	Level at discharge 11/26/13	Physical Therapist's Assessment 11/26/13																										
Questionnaire: Neck Disability Index	Improve by 10%: To drive without increased neck pain.	26.00%	25% improvement documented																											
Posture: Forward head/rounded shoulders	Decrease forward head posture to minimum	Moderate	"Some progress" documented	25% of Goal Met																										
Range of motion: cervical spine	Increase active rotation left & right to 45 degrees: To turn head while driving.	<table border="1"> <thead> <tr> <th>Active Motion Tested</th> <th>Range of Motion in degrees</th> </tr> </thead> <tbody> <tr> <td>Flexion</td> <td>52</td> </tr> <tr> <td>Left Rotation</td> <td>26</td> </tr> <tr> <td>Right Rotation</td> <td>41</td> </tr> <tr> <td>Left Side Bending</td> <td>25</td> </tr> <tr> <td>Right Side Bending</td> <td>20</td> </tr> </tbody> </table>		Active Motion Tested	Range of Motion in degrees	Flexion	52	Left Rotation	26	Right Rotation	41	Left Side Bending	25	Right Side Bending	20	<table border="1"> <thead> <tr> <th>Active Motion Tested</th> <th>Range of Motion in degrees</th> </tr> </thead> <tbody> <tr> <td>Flexion</td> <td>52</td> </tr> <tr> <td>Left Rotation</td> <td>40</td> </tr> <tr> <td>Right Rotation</td> <td>41</td> </tr> <tr> <td>Left Side Bending</td> <td>25</td> </tr> <tr> <td>Right Side Bending</td> <td>20</td> </tr> </tbody> </table>		Active Motion Tested	Range of Motion in degrees	Flexion	52	Left Rotation	40	Right Rotation	41	Left Side Bending	25	Right Side Bending	20	75% of Goal Met
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Muscle Testing: Cervical planes MMT	Improve to +4/5	Flexion L&R +3/5	Flexion L&R -4/5 "good progress" noted	50% of Goal Met																										
Muscle Testing: Upper extremity MMT	To lift/carry light objects pain free Improve Bilaterally: Adduction to +4/5 Flexion to -5/5	Bilaterally: Adduction 3/5 Flexion -4/5	Bilaterally: Adduction -4/5 Flexion 4/5 "good progress" noted	50% of Goal Met																										
Palpation: Decreased mobility	Improve to slight restriction: To sleep through the night	Erector Spinae, Suboccipitals, Upper trapezius: Moderate restriction bilaterally	Erector Spinae, Suboccipitals, Upper trapezius: Mild restriction bilaterally "Excellent progress" noted	75% of Tissue Mobility Goal Met																										

Discussion

The primary goal of the physical therapist regarding patients with systemic sclerosis is symptom management in order to improve function, longevity, and quality of life. Improvements in early diagnosis and early intensive multidisciplinary treatment have led to increased survival length. This places more importance on maintaining and promoting functional ability (Nihtyanova, 2009; Casale, 1997). In a study conducted by Vincent and Wilson (2006), some patients placed more importance on maintaining their functional mobility and what they considered a normal life, than they did on anticipation of a shortened life expectancy. A systematic review with a meta-analysis has shown that the 10-year survival rate among this population has risen from near 60% to slightly above 75% from 1985 to 2002 (Ferri, 2002). Many of the treatments that patients with PSSc undergo are pharmacological and surgically based, including systemic medication trials and immunosuppressive therapy (“umm.edu”, 2012). Regarding the importance of maintaining function, the physical therapist’s role specific to patients with PSSc includes maintaining joint ROM, limiting connective tissue pain and stiffness, and promoting cardio-respiratory fitness. In order to do this, valid and appropriate objective measurements need to be taken. High-level evidence-based practice literature is limited in regards to physical therapy examination and treatments, however the literature that is available supports physical therapy interventions.

Passive and active skin mobilization, strengthening exercises, and joint range of motion helps to prevent contractures, muscular atrophy, degradation of joints and subsequent pain. Joint movement circulates synovial fluid that is compromised

due to decreased vascularity and limited motion. This supports joint nutrition and decreases joint articular surface breakdown (Kisner & Colby, 2007). Additionally, joint movement stimulates mechanoreceptors that are influential in modulating pain (Kisner & Colby, 2007). It is important to remember that each exercise program requires individual customization. A patient with SSc who has significant joint pathology should be placed on an exercise program that does not exacerbate joint pain or swelling. Not doing so may cause degradation of articular cartilage that will in turn increase the inflammatory process and dysfunction associated with the disease. In general, joint ROM and muscular strength should be the focus of interventions while limiting joint inflammation (Casale, 1997). Furthermore, exercise prescription should be based on objective findings and progressed based on patient tolerance and results (ACSM, 2013). The documentation available for this case patient over the one-month plan of care provides few details in regards to the rationale for exercise progression, and only partially addressed objective findings of weakness. It is important for the physical therapist to closely monitor patients with this diagnosis to determine the efficacy of treatment, and adhere to evidence-based exercise prescription parameters.

Physical exercise directly improves lung function, and is considered the standard of care. (Arena, 2011; Baptista, 2012; Pinto, 2011; Shoemaker, 2009). However, according to one study, only 46% of patients diagnosed with systemic sclerosis participated in physical therapy (Johnson, 2006). It is very important to note that exercise-induced pulmonary hypertension has been regarded as a common finding in patients with SSc (Baptista, 2012). This speaks to the importance of

appropriately trained and informed clinicians monitoring vitals and patient presentation during activity. Physical exercise has also been shown to enhance overall health. It improves quality of life, improves functional capacity, and has been shown to prevent the progression and improve the prognosis of multiple diseases (“CDC.gov”, 2012). In addition to the health benefits, exercise can promote psychological health and provide improved quality to life in the form of goal accomplishment and giving the patient a feeling of control (“CDC.gov”, 2012).

The case patient’s physical therapist stated that she was mildly familiar with the diagnosis of systemic sclerosis. Based on the clinical judgment of the physical therapist, the case patient was treated for cervical neck dysfunction conservatively. The physical therapist anticipated slower than typical gains and increased healing times due to the vascular compromise secondary to the disease process. The administered plan of care and one month of treatment interventions showed 25% efficacy for the correction of posture, 50% effective regarding improvements in strength, and 75% successful in improving the patients cervical neck range of motion.

The APTA states “Physical therapists provide patient/client management in primary care through the processes of screening, examination, evaluation, diagnosis, prognosis, intervention, education, prevention, coordination of care, and referral to other providers to prevent, remediate, decrease, or slow the progression of impairments, activity limitations, and participation restrictions, and lessen the impact of environmental barriers, and optimize cost-effective clinical outcomes. “ (APTA, 2012). Based on this, it is the physical therapist’s responsibility to evaluate

the body systems that are affected by the disease process of SSc and to educate the patient and appropriate health care practitioners. Interventions provided in regards to cervical dysfunction demonstrated progress towards goals; however, assessment of deep neck flexor strength and muscle recruitment during respiration were not mentioned during the examination or course of treatment and could have an influence on long term progress (Falla, 2004). Literature concerning evidence-based assessment and interventions detail the need to address the known systems that are typically affected by the disease process of SSc. Although available literature is limited, the plan of care provided by physical therapists should address morbidities concerning cardiopulmonary health, functional strength and endurance, and hand function.

The extent of patient education was not clearly stated in the available documentation. Patients with SSc should be educated and referred to resources such as the Johns Hopkins Scleroderma Center, or the Scleroderma Foundation, which is a thorough online based information center complete with everything from support groups and forums to the most current information and research. Evidence based practice would have also included a baseline, or a continuation of the monitoring of the major systems that are impacted by the disease process that are within our professions scope of practice. These would include pulmonary tests such as FEV1 and lung capacity, blood pressure and cardiac response to graded exercise, and functional assessments such as the Scleroderma Health Assessment Questionnaire or the Cochin Hand Function Scale. The inclusion of these tests would help form a baseline with which to measure progression, substantiate appropriate

interventions, and provide useful information to clinicians of other disciplines (Follansbee, 2004; Clements, 2009).

Considering other disciplines, it is strongly suggested that patients who either display signs of systemic sclerosis or carry the diagnosis of SSc are under the care of a rheumatologist (www.hopkinsscleroderma.org). The global effects of SSc interfere with the processes of nearly every body-system. Complications and symptoms extend beyond the content of this case report. Notably, SSc has potentially profound effects on the renal, GI, and sexual processes, which require professional monitoring and consultation for appropriate evidence-based management.

Conclusion

After reviewing the literature, it is clear that exercise and other physical therapy interventions are beneficial and produce significant benefit for patients with SSc. However, there is not enough high quality evidence regarding exercise protocols, testing measures, or treatment guidelines. The disease is progressive and variable from patient to patient so interventions need to reflect patient needs; however, there is a lack of guidelines that could help physical therapists in providing evidence-based care. Based on the diagnosis of Systemic Sclerosis, physical therapists should consider evidence-based examination and intervention for all symptoms found to be positively affected by physical therapy. Additionally, physical therapists need to be aware of the systemic nature of the disease, and the recommendations for interdisciplinary delivery of health-care, and early intervention to prolong functional independence and quality of life.

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