Evidence-Based Management of Hand and Mouth Disability in a Woman Living with Diffuse Systemic Sclerosis (Scleroderma)

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ABSTRACT

This paper uses a case study to highlight the evidence for rehabilitation interventions designed to reduce disability of the hand and mouth and manage fatigue in persons with diffuse systemic sclerosis to inform clinical practice and guide future research.

Key Words: activities of daily living; evidence-based practice; hand deformities; mouth rehabilitation; self-care; skin and connective tissue diseases; scleroderma, diffuse.

RÉSUMÉ

Dans ce document, on utilise une étude de cas pour présenter les données probantes à l'appui d'interventions de réadaptation conçues pour atténuer l'incapacité de la main et de la bouche et gérer la fatigue chez les personnes atteintes de sclérodémie systémique diffuse afin d'éclairer la pratique clinique et de guider des recherches futures.

CLINICAL CASE

Andrea is a 48-year-old woman with a recent diagnosis of diffuse systemic sclerosis who has come to a physiotherapy clinic with the goals of improving the functional mobility of her hands and mouth and increasing her energy level, particularly during the workday. Over the past year, Andrea has noted general fatigue, stiff fingers, and episodes of heartburn. She reports that her fingers are curling, which makes it difficult for her to grasp small objects and get items out of her pockets. Five months ago, her dental hygienist told her that her mouth opening had decreased and her oral hygiene was suffering as a result. Three months later, she was evaluated by a rheumatologist who made the diagnosis and started her on a course of oral cyclophosphamide. Her past medical history is uneventful apart from a 10-year history of Raynaud's phenomenon. Andrea is employed as an office manager in a busy family health care centre; she is married, with two daughters who no longer live at home.

CLINICAL QUESTION

What types of evidence-based rehabilitation interventions could address Andrea's hand and mouth disability and fatigue?

THE EVIDENCE

Systemic sclerosis (also known as scleroderma) is an autoimmune disease with a prevalence of 2–3 per 10,000 people.¹ The ratio of women to men is 4 to 1, and most people are diagnosed between the ages of 30 and 50.¹ Diffuse systemic sclerosis, the most severe subtype, is characterized by dermal fibrosis in the face, neck, and trunk and, symmetrically, in the fingers, hands, arms, and legs.¹ Thick and shiny fingers and restricted range of motion (ROM) in the hands and face are common manifestations.¹ Raynaud's phenomenon is very common (86%–98%) in people with systemic sclerosis and may cause pain and/or contribute to the development of skin ulcers.^{1,2} Fibrotic changes in the vascular system and internal organs (gastrointestinal tract, lungs, heart, kidneys) occur early in diffuse systemic sclerosis.¹

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Figure 1 Specific tools such as adapted handles on cutlery, button hooks to assist in dressing, jar-opening devices, and toothbrushes have been designed to help people with systemic sclerosis remain independent.

Andrea's heartburn is characteristic of the gastrointestinal involvement that affects 66%–90% of people with systemic sclerosis^{2,3}; fatigue is another commonly reported problem and may result in difficulty sleeping, increased effort needed to perform daily tasks, or depression.^{1,2}

Management of hand disability

Because of Andrea's hand involvement, ROM exercises emphasizing flexion at the metacarpophalangeal joints, extension of the proximal interphalangeal joints, and flexion and abduction of the thumbs are indicated. To perform these exercises, Andrea simply applies pressure with one hand to stretch the joints in the other hand to the point of skin blanching.

Heat modalities such as paraffin wax or hot packs before ROM exercises may reduce pain and increase the extensibility of collagen tissue. Several studies, including two randomized controlled trials, have shown that paraffin treatment followed by hand ROM exercises increased both joint motion and grip strength beyond im-

provements observed with stretching alone.^{4–6} A recent randomized controlled trial reported that connective tissue massage of the forearm and hand in conjunction with Mc Mennell joint manipulation of the wrist and fingers and a home programme of active hand, wrist, and forearm ROM exercises resulted in significant improvements in joint motion, hand function, and quality of life compared to the daily home-exercise programme alone.⁷

Ideally, rehabilitation will allow Andrea to regain and maintain the ability to perform daily tasks with her hands; however, several assistive devices are also available to maintain independence and make household tasks and personal care manageable (see Figure 1).

Management of mouth disability

Skin tightening due to subcutaneous and ligamentous collagen deposits associated with diffuse systemic sclerosis also results in a mechanical inability to open the mouth.⁸ Limited range of mouth opening, along with

Figure 2 A template for monitoring changes in finger extension.

other symptoms such as dry mouth, can lead to difficulties with oral hygiene and eating.⁸ Several studies have shown that ROM exercises are effective in increasing mouth opening,⁹ with subsequent improvements in speaking and eating ability¹⁰ and in oral hygiene.^{11,12} The exercises consist of exaggerated facial movements, manual stretching of the mouth with the thumbs, and oral augmentation exercises using tongue depressors.^{9–13} Significant improvements in mouth opening and decreases in the thickness of facial skin were reported following a multicomponent intervention for the face that included connective tissue massage; proprioceptive neuromuscular facilitation techniques; and passive, active, and active-assisted exercises for the temporomandibular joints, in addition to a daily home-exercise programme consisting of two passive mouth-opening exercises and a series of active exaggerated facial expressions to exercise facial and orofacial muscles.¹³

Oral hygiene can also be maintained by using tools that are easier to grip and manoeuvre in a smaller mouth opening, such as electric toothbrushes and flossers, water jets, children's toothbrushes, and built-up handles. By performing mouth ROM exercises and using such devices, Andrea should be able to improve her oral hygiene.¹²

Management of fatigue

The four P's of energy conservation are prioritize, plan, pace and posture.¹⁴ Andrea should set priorities and delegate additional tasks to others. Careful planning will also allow her to perform more tasks over a longer time frame. By analyzing her pace, Andrea can reduce fatigue by balancing rest and activity. Finally, maximizing time spent promoting correct postures and body mechanics will conserve energy.¹⁵

Work situations are more difficult for people with systemic sclerosis due to fatigue, pain, temperature in-

tolerance, stiffness, and impaired movement.¹⁶ By dressing in layers, Andrea can stay warm in her temperature-controlled office. To conserve energy, she could work fewer and more flexible hours. Asking for help and disclosing challenges to co-workers has been shown to generate support for working people with systemic sclerosis.¹⁶ Andrea could consider accommodations such as voice-activated software for the computer and a telephone headset.

Outcome measures

Andrea should self-monitor improvements in hand and mouth ROM as she performs her home exercises. She can create templates of the flexibility in the joints of her hands. Figure 2 shows a template she can use to monitor finger extension; an additional template can be created by tracing the metacarpophalangeal joint position for each finger. Thumb abduction can be monitored by taking note of the largest object Andrea can grasp while maintaining contact with the object along the entire web space (between the thumb and index finger). By referring to these templates, Andrea can note changes in ROM; when ROM extends beyond the templates, this indicates an improvement. Although mouth ROM is more difficult to track independently, a tool used by dentists, called a Boley gauge, may be used to accurately measure the opening of the mouth.

LIMITATIONS OF THE EVIDENCE AND FUTURE DIRECTIONS

Recent research has shown improved functioning of people with systemic sclerosis, but gaps in the research remain. Published evidence suggests that exercises for the hands and face, paraffin wax, massage, and joint manipulation improve joint motion.^{4–7,9–11,13} However, sample sizes for these studies were small, and, apart from one study with outcomes assessed at 12 months,¹¹ the follow-up was usually 3 months^{5,9} or less.^{4,6,7,10,13} Adequately powered studies with longer follow-up are needed to determine optimal dosages of modalities and volume, intensity, and progression of exercises.

By the time someone is diagnosed and referred for physiotherapy, joint motion can be significantly decreased. For example, Andrea's dental hygienist observed her difficulty with oral care before she was diagnosed with diffuse systemic sclerosis. Further studies should explore the effect of early physiotherapy interventions for people with systemic sclerosis.

CONCLUSIONS

Although systemic sclerosis is a devastating diagnosis, addressing impairments in mouth and hand ROM and fatigue may contribute to a life that is more independent and fulfilling. Andrea's situation is common to many people with diffuse systemic sclerosis. Clearly, a variety of exercises is important to restore and maintain the ROM necessary for activities of daily living. Andrea should

promptly begin physiotherapy to minimize hand and mouth disability and prevent functional limitations elsewhere. She needs support to perform stretches at sufficient intensity and to implement the four P's to reduce her fatigue so that she can continue to participate in meaningful activities. Through education, multicomponent treatment, and self-management, Andrea may preserve her quality of life and enjoy activities of daily living, mobility, and participation.

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