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Occupational Therapy and Scleroderma (systemic sclerosis)

Janet L. Poole, PhD, OTR/L, FAOTA
Occupational Therapy Graduate Program, University of New Mexico
jpoole@salud.unm.edu

I. Objectives
a. Identify symptoms of systemic sclerosis (scleroderma)
b. Discuss occupational therapy intervention to increase joint motion, reduce pain, and manage daily tasks, fatigue, and Raynaud’s phenomenon
c. Review the evidence for occupational therapy interventions

II. Systemic sclerosis (SSc)
a. Progressive multi-system autoimmune disease characterized by thickening of the skin, vascular involvement and fibrosis of the internal organs
b. Prevalence: 240/1 million
c. Both prevalence and incidence are 4 times greater in women than in men
d. Onset between 30-40 years of age
e. Average 10-year survival rate is now 70% to 80%

http://scleroderma.org/
http://www.niams.nih.gov/Health_Info/Scleroderma/default.asp

III. Subtypes of SSc
a. Limited cutaneous – gradual skin thickening limited to the distal extremities and face; later involvement of the internal organs
   Includes CREST
   C – Calcinosis
   R – Raynaud’s phenomenon
   E – Esophageal dysfunction
   S – Sclerodactaly
   T – Telangiectasias
b. Diffuse cutaneous – Skin thickening proximal to the elbows and knees (i.e. upper arms, thighs or trunk) and early involvement of the internal organs. More severe subtype.

IV. Typical contractures in the hands
a. Decreased flexion of the Metacarpophalangeal (MCP) joint
b. Decreased extension of the Proximal Interphalangeal (PIP) joint
c. Decreased thumb abduction

continued
V. Other hand symptoms
   a. Calcium deposits
   b. Puffy fingers
   c. Raynaud’s phenomenon
   d. Digital ulcers and scars

VI. Organ Involvement
   a. Raynaud’s phenomenon
   b. Gastroesophageal problems
   c. Arthralgias or arthritis
   d. Pulmonary fibrosis
   e. Cardiac involvement
   f. Renal involvement
   g. Myopathy

VII. Assessment
   a. Joint range of motion
   b. Hand strength
   c. Hand function
   d. ADL/IADL
   e. Observation – sores, scars on fingers/fingertips, calcium deposits, puffy fingers

VIII. Role of Occupational Therapy
   a. Overall goal is to provide individuals with the tools and information to resume participation in activities of daily life
   b. Improve joint motion and strength
   c. Provide assistive devices/adapted equipment or alternative methods to accomplish daily tasks
   d. Client education
IX. Range of motion/stretch exercises
   a. Purpose is to prevent or slow down development of contractures
   b. Do frequently and beyond resistance
   c. Maintain position of stretch 3-5 seconds even if skin blanches or turn white
   d. Probably should be started before there is any noticeable loss of motion

X. Hand Exercises

   Exercises for MCP flexion

   Make a fist – heel of one hand may need to press on dorsum of proximal phalanx of the other hand

   Exercises for PIP Joint

   Push hand flat Press fingers against each other Use three point of pressure
Exercises for the thumb

**Abduction**

[Image of hand with fingers abducted]

**Flexion**

[Image of hand with fingers flexed]

XI. How to monitor the motion in key joints

- Draw hand outline for finger extension
- Templates to monitor MCP flexion
- Find largest object that will fit in web space with all sides of web space touching object

XII. Evidence for stretching exercises

<table>
<thead>
<tr>
<th>Author</th>
<th>Participants</th>
<th>Treatment</th>
<th>Results</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mugii et al, J Rheum, 2006; 33:1586-1592</td>
<td>N = 45 (32 diffuse; 13 limited)</td>
<td>Individual fingers stretched for 10s, 3-10 times once/day; monitored 1/month for 1 year <strong>Exercises</strong>: MCP flexion, PIP and DIP flexion, PIP and DIP extension</td>
<td>Total passive motion significantly ↑ in each finger after 1 mo. Improved or maintained at 1 year. Gripping and eating component scores on HAQ ↓ significantly at 1 year follow-up</td>
</tr>
</tbody>
</table>
XIII. Modalities to increase joint motion (Poole, 2010; Mouthon & Poole, 2012)
   a. Heat – paraffin or hot packs
      a. - 126-130° or less
         - do not use with open sores
   b. Connective tissue massage (Bongi et al., 2009)
   c. Manual lymph drainage (Bongi et al., 2011)

XIV. Evidence for Modalities

**Paraffin + Exercise**

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<tbody>
<tr>
<td>Pils et al. <em>Phys Med Rehabil</em> 1991; 1:19-21</td>
<td>N = 16 RCT Tx = 8 Control = 8</td>
<td>All 16 had 12 paraffin tx Tx continued paraffin for 3 months Control – stopped paraffin</td>
<td>Initial 12 tx → ↑ joint motion, ↓ stiffness. Maintained in Tx group but not controls</td>
</tr>
<tr>
<td>Sandqvist et al., <em>Disability Rehab</em>, 2004; 26: 981-987</td>
<td>N = 17 RCT Tx = one hand Control = other hand</td>
<td>Tx hand received paraffin + exercise for 1 month Control hand received exercise only</td>
<td>Tx hand-finger extension, stiffness and skin elasticity improved significantly more than control hand</td>
</tr>
<tr>
<td>Mancuso &amp; Poole <em>J Hand Ther</em> 2009; 22: 71-77</td>
<td>N = 3 Single case studies</td>
<td>Paraffin + exercise for 8 weeks</td>
<td>↑ ROM, grip, pinch strength ↑ in hand function not as significant as ↑ impairments</td>
</tr>
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</table>

**Connective tissue massage and joint manipulation**

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</thead>
<tbody>
<tr>
<td>Maddali Bongi, et al., <em>Clin Rheum</em> 2009; 28:1167-73.</td>
<td>N = 40 RCT Tx = 20 Control = 20</td>
<td>Tx hand: Combination tissue massage, McMennell joint manipulation for 1 hr, 2X/week for 9 weeks, plus a home exercise program Control: home exercise program</td>
<td>Tx group: fist closure, joint motion, hand and overall QoL increased and maintained at 9 week follow-up. Control group: fist closure improved</td>
</tr>
</tbody>
</table>
Manual Lymph Drainage

<table>
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</tr>
</thead>
<tbody>
<tr>
<td>Maddali Bongi et al.,</td>
<td>N = 35</td>
<td>Tx for edema: manual lymph drainage 1 hr, 1X/week for 5 weeks</td>
<td>Tx group: hand volume, joint motion and perception of hand disability improved. Control group: no improvements</td>
</tr>
<tr>
<td>AC&amp;R 2011; 63:1134-1141</td>
<td>Tx = 20</td>
<td>Control: observation group</td>
<td></td>
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<tr>
<td>Control = 15</td>
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</table>

Dynamic splints

<table>
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</tr>
</thead>
<tbody>
<tr>
<td>Seeger &amp; Furst,</td>
<td>N = 19</td>
<td>Tx hand: Dorsal splint with wrist in 15° extension, MCP 0° and dynamic PIP extension outrigger. Worn 8 hr/day for 2 months. Weekly splint checks. Control hand: unsplinted hand</td>
<td>8/19 completed study. No significant change in PIP extension in either group.</td>
</tr>
<tr>
<td>Am J Occup Ther 1987: 41: 118-121</td>
<td></td>
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</tr>
</tbody>
</table>

XV. Exercises for other upper extremity joints

Shoulder flexion

Shoulder extension

Shoulder external rotation

Supination

Wrist extension

Wrist flexion
XVI. Oral manifestations

1. Oral manifestations
   a. Resorption of the mandible
   b. Reduced mouth opening (microstomia)
   c. Xerostomia (dry mouth)
   d. Difficulty eating and oral hygiene
   e. Increased dental caries and periodontal diseases

2. Why is mouth opening important?

3. How to monitor mouth opening?

XVII. Temporomandibular joint exercises (Naylor et al., 1984)

1. Exaggerated facial movements
   a. Exaggerated smiling

   b. Pursing lips

   c. Blowing up cheeks

   d. Open mouth wide

2. Manual stretch of mouth with fingers

3. Augmentation with tongue depressors
XVIII. Oral hygiene

1. Dental caries and dental problems occur because of Sjögrens syndrome and microstomia (small mouth)

2. Devices that might help include electric tooth brushes, water jets, curved bristle brushes, children’s toothbrushes, large handled flossers or electric flossers

**Evidence for oral interventions**

<table>
<thead>
<tr>
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</tr>
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</table>
Tx = 5  
Control = 4 | Tx: Mouth stretching 3 sets of 5 stretches 1X/day & augmentation 2X/day for 3 months  
Control: facial grimacing, 3 sets of 5 stretches 1X/day | Tx group: ↑mouth opening a  
Control: no significant increase |
| Pizzo et al. Clin Oral Invest 2003; 7:175-178 | N = 10 | Tx: Mouth stretching 15 min, 2X/day and augmentation 1X/day for 8 weeks | Tx group: ↑mouth opening;  
↑speaking and eating abilities |
| Poole, et al. Disabil Rehab 2010; 32: 379-384 | N = 17 | Tx: Dental exam and cleaning, educ on brushing/flossing, individualized adapted dental appliances, Mouth exercises (stretching and augmentation) & hand exercises 1X/day | ↑mouth opening and improved oral hygiene |
| Maddali-Bongi et al. Rheumatol Int 2011; 31: 895-901. | N = 40  
Tx =20  
Control = 20 | Tx: 1hr 2X/wk for 9 weeks: massage to face, PNF stretch and resistive exercise and face exercises  
Control: home exercises for mouth & face | Tx: ↑mouth opening and  
↓facial skin scores  
Control: no change |
<table>
<thead>
<tr>
<th>Study</th>
<th>N</th>
<th>Tx</th>
<th>Control</th>
<th>Intervention</th>
<th>Results</th>
</tr>
</thead>
</table>
Tx: provided rechargeable toothbrush and reach flosser. If oral aperture < 40 mm, face, mouth stretching & oral augmentation exercises 2X/day  
Control: manual toothbrush and floss | Tx: ↑oral hygiene as shown by reduction in Gingival Index scores that was 8% larger and significant at 6 mo. compared to control |
| Yuen et al. Disabil Rehab 2012; 34:84-89 | 48 | 26  | 22      | Tx: If oral aperture < 40 mm, face, mouth stretching & oral augmentation exercises 2X/day. Each exercise held 15-20 s; kept records of daily hygiene. Also issued toothbrush & flosser as described above  
Control: manual toothbrush and floss | Tx: ↑mouth opening at 3 mo. Compared to controls but not at 6 months. Adherence to exercise as low (48.9%) |

IXX. Assistive/ Adaptive devices

1. For manipulation try large handles, universal cuffs, hourglass or stem glasses
2. For dressing for reaching try reachers, sock aids, long shoe horns and for manipulation use button hooks (no buttons) or zipper pulls
3. Numerous devices are available for keys, pills, opening jars, cans, bottles, etc

XX. Proximal weakness

1. Chairs with armrests, pillows on chairs, raisers on chair and bed legs, and raised toilet seats are helpful. Use good body mechanics when coming to standing.
2. Compensate by crossing one leg over the other for lower extremity dressing

XXI. Energy Conservation (the 4 P’s)

1. Prioritize
1. Make a list of priorities and the order by importance; delegate

2. Plan ahead

3. Pace yourself and avoid fatigue; slow down
   a. Balance rest and activity: short frequent rests

4. Posture
   a. Sit when possible but alternate positions too
   b. Use good posture
   c. Use good body mechanics

XXII. Energy management ideas used by working people with scleroderma

1. Ask/accept help from co-workers
2. Modify work hours – 4 day weeks or shorter days (start later)
3. Work from home
4. Keep WARM
5. Switch tasks and take breaks to cut fatigue or stress
6. At home – renegotiate tasks at home, sleep on weekends, prepare for workweek

XXIII. Energy management ideas for leisure activities

1. Always have hand warmers and gloves with you
2. Rest and pace activities
3. Internet shopping instead of mall shopping
4. Be prepared – gloves, sweaters/jackets, and blankets to concerts, theatre, restaurants, etc

XXV. Management of Raynaud’s Phenomenon
1. Education
   a. Insulate hands from cold, strong detergents, irritating chemicals, and bacteria (wear gloves)
   b. Dress warmly/layer to insulate body – always bring sweater, mittens/gloves
   c. Use chemical hand warmers
   d. Avoid cold temperatures
   e. Take care of skin – keep skin moist
   f. Biofeedback

2. Equipment to “warm”
   a. Warm mouse, heated keyboard \textit{www.coldhands.me}

   b. Warm car solutions – remote car starters, heated seats or covers, accessible parking

XXVI. Keyboard and work ideas

1. Voice activated, keyboard aids, “stickykeys”

2. Accommodations
   a. Sick days or extra days can be requested
   b. Work at home, more breaks, light work, equipment can be requested
   c. Explore Family Medical Leave Act
   d. Explore Flextime schedule
   e. Resources
   f. Job Accommodation Network \textit{www.jan.wvu.edu}

\textit{www.disability.gov} \hspace{1cm} \textit{www.dol.gov} \hspace{1cm} \textit{www.eeoc.gov}

XXVII. Leisure ideas

XXVIII. Conclusions

1. Occupational therapy is indicated for persons with scleroderma
2. Range of motion exercises should be **aggressive**, involve **stretching** and be started before obvious deformities

3. Do NOT forget the face

4. Do NOT forget Raynaud’s phenomenon

5. Assistive devices and alternate techniques (i.e. energy conservation) should be investigated to help with independence AND save energy

6. Some evidence available for rehabilitation interventions for persons with scleroderma

References


Poole JL. Musculoskeletal rehabilitation in the person with scleroderma. Curr Opin Rheumatol 2010; 22; 205-212.


