

The Role of the Occupational Therapist



Raynaud's & Scleroderma

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Support for sufferers

The aims of the Association are to promote a greater awareness of Raynaud's and scleroderma, to raise funds for research and to offer advice and support to sufferers, their families and friends on the problems of day to day living. On joining the Association, members receive quarterly newsletters giving up-to-date information on research and treatments.

Anne H Mawdsley, MBE, Director of the Raynaud's & Scleroderma Association would like to thank Anne Johnson, Occupational Therapist at the Royal National Hospital for Rheumatic Diseases, NHS Trust, Bath, for her support in the preparation of this publication.

Further information on Health Professional booklets and patient literature is available from:

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The role of the Occupational Therapist

This booklet has been written as a brief guide for Occupational Therapists who may at some time be responsible for treating a person with Raynaud's phenomenon (RP) or scleroderma. The booklet can also be given to patients and other health and social care professionals for information/educational purposes.

The importance of employing a multidisciplinary team approach for people with Raynaud's or scleroderma, needs to be emphasized in order to provide a comprehensive treatment programme. Obtaining other profession specific leaflets published by the Raynaud's & Scleroderma Association is recommended to increase understanding of these conditions and the role of the team.

What is Raynaud's?

Raynaud's is a condition in which the blood supply to the extremities, usually the fingers and toes but sometimes also the ears and nose, is interrupted. During an attack the fingers and/or toes go through a process called triphasic colour change: first they become white/dead looking, second they may turn blue and then finally go red. The person may experience considerable pain, a burning feeling, numbness or tingling.

Raynaud's can range in severity from minor discomfort to the onset of ulcers or even gangrene. Anyone of any age can develop Raynaud's. Children and teenagers can be affected.

Progress may be slow over a period of many years. Females are affected nine times more than males. Triggers include stress and anxiety, touching cold objects, going into a cold atmosphere and more importantly, any slight change in temperature. It is important for the occupational therapist to be aware of these triggers in order to eliminate them during therapy sessions.



Photograph showing a Raynaud's attack

Raynaud's can be subdivided into:

1. Primary Raynaud's which occurs spontaneously without any underlying condition being present (it can be hereditary, in which case it is usually mild).
2. Secondary Raynaud's, which is less common and is associated with underlying diseases such as scleroderma, systemic lupus erythematosus, Sjögren's Syndrome and rheumatoid arthritis. Secondary RP is more serious and early and accurate diagnosis is essential.



What is scleroderma?

Scleroderma, also known as systemic sclerosis is an auto-immune, generalised disorder of the connective tissue. Fibrotic and degenerative changes occur in the skin, synovium and digital arteries and can affect the internal organs including the oesophagus, intestinal tract, heart, lungs and kidneys. It affects about 20 per million of the population per year and is three times more common in women than men. The cause is unknown and it is neither contagious or inherited.



How serious is scleroderma?

Any chronic disease can be serious but the symptoms can vary greatly from one individual to another. The effects of scleroderma can range from mild to very severe. The seriousness will depend on what parts of the body are affected and the extent to which they are properly treated. In its most severe form scleroderma can be fatal. Cardiopulmonary failure has now surpassed renal failure as the leading cause of death in patients with scleroderma.



Classification of the Systemic Sclerosis subsets

Pre-scleroderma

Raynaud's phenomenon plus nailfold capillary changes, disease specific circulating anti-nuclear autoantibodies, (anti-topoisomerase-I, anti-centromere (ACA), or nucleolar), and digital ischaemic changes.

Diffuse cutaneous SSc (dcSSc)

Onset of skin changes (puffy or hidebound) within 1 year of onset of Raynaud's. Truncal and acral skin involvement.

Presence of tendon friction rubs – audible leathery creaking due to fibrinous deposits. Early and significant incidence of interstitial lung disease, oliguric renal failure, diffuse gastrointestinal disease, and myocardial involvement.

Nailfold capillary dilatation and drop out.

Anti-topoisomerase-I (Scl-70) antibodies (30% of patients).

Limited cutaneous SSc (lcSSc)

Raynaud's for years (occasionally decades).

Skin involvement limited to hands, face, feet and forearms (acral).

A significant (10–15%) late incidence of pulmonary hypertension, with or without interstitial lung disease, skin calcification, telangiectasiae and gastrointestinal involvement.

A high incidence of ACA (70–80%).

Limited cutaneous SSc previously called the CREST syndrome:

- **C** Calcinosis (calcium deposits under the skin)
- **R** Raynaud's
- **E** Esophageal involvement (American spelling)
- **S** Sclerodactyly – skin of digits becomes thin, shiny and leathery looking. Fingers and toes may become stiff and flexed.
- **T** Telangiectasia – the appearance of small blood vessels near the surface of the skin. These may be seen on the fingers, palms, lips, face, tongue and chest wall. Dilated nailfold capillary loops, usually without capillary drop out.

Scleroderma sine scleroderma

Raynaud's +/-

No skin involvement

Presentation with pulmonary fibrosis, scleroderma renal crisis, cardiac or gastrointestinal disease.

Antinuclear antibodies may be present (Anti-topoisomerase-I, (Scl-70) ACA, nucleolar).



Scleroderma signs and symptoms

Raynaud's phenomenon is usually the first symptom of scleroderma and may occur many years before other symptoms. Raynaud's affects over 95% of people with scleroderma (Ref.1). Skin change is the most common presenting sign. Skin on the hands, feet and face becomes stiff, tight and shiny.

This is due to swelling initially and then thickening of the connective tissue which becomes fibrotic or scarred.

The skin may also appear dry due to obliteration of the sebaceous glands

by the connective tissue. Ulcers, calcinosis and pitting of the skin may also be present.

Microstomia (small mouth) is often a common feature, making eating and oral hygiene a problem at times. If the gastrointestinal system is affected, there may be difficulties with swallowing, acid indigestion, malabsorption, diarrhoea or anal incontinence.

Dry mouth and dry eyes, as a result of Sjögren's syndrome, another auto-immune disease related to scleroderma, can be a problem.

Synovitis, myalgia, fatigue and shortness of breath – due to heart or lung involvement, may further complicate the patient's ability to carry out day to day activities.



Photograph showing tight, shiny skin

How can the Occupational Therapist help?

Liaison with other team members is important. The patient has usually been seen by the rheumatologist and possibly a clinical nurse specialist, before reaching the occupational therapist. Through liaison, valuable background information can be gathered, assisting in rapport building not only with the patient but also with the team. Occupational therapists who work closely with physiotherapists will be able to offer the best therapy input for the patient.

Aim of the Occupational Therapist

To work in partnership with the individual and his or her 'family', in order to achieve and maintain optimum independence in activities of daily living.

Assessment, planning and evaluation

Assessment of the whole person in order to establish baselines and plan intervention is of paramount importance. Because of the possible multi system involvement in scleroderma, thorough assessment of the patient is necessary. Assessment will ascertain the individual's motivations, abilities and needs. The use of an activities of daily living assessment, or checklist in conjunction with a validated, standardised, functional assessment is recommended. The Canadian Occupational Performance Measure (Ref.2) is one such example. Such a tool is useful in planning the individual's personal goals and can be used as an outcome measure/evaluation of intervention.

The use of goniometry to measure range of movement in the hand can be useful, but therapists need to bear in mind problems of inter and intra rater reliability, if this method is used to chart hand changes over time. The use of a validated hand assessment should be considered such as the Arthritis Hand Function Test (Ref.3).

Characteristic Hand Changes in Systemic Sclerosis

- Loss of MCP flexion
- Loss of PIP extension
- Tuft resorption DIP tuft = DIP phalanx shortening
- Tightening of thumb web
- Reduced wrist movement in all planes (Ref.4)



Raynaud's

Based on needs established at assessment and organs involved. Raynaud's is largely helped using behavioural changes.

Keeping Warm

- Wearing gloves and socks and adequate clothing to maintain body temperature.
- Assistive equipment might include use of portable hand warmers or battery heated socks.
- Using gloves when getting items out of the fridge or freezer.

Avoiding Stressful Situations

- Identifying stressors and adopting cognitive behavioural strategies to cope.
- Use of relaxation and/or biofeedback.

Smoking

- Advise the patient to give up smoking as this causes vasoconstriction.

If Hands and Feet Get Cold

- Warm them up gradually.

Occupational Hazards

- Vibration White Finger (VWF) is Raynaud's of occupational origin – seen in people who use vibrating machine or tools such as chain saws, pneumatic drills, hammers or polishers as part of their job. Career/job advice and task analysis may be necessary (Ref.5).

Raynaud's in Children

- School play times – children may need permission to stay in on cold days. However, this should not be an excuse to avoid exercise!.
- Health and safety precautions should be taken during 'risky sessions' for example in the science lab or during cooking sessions – cold and numb hands could get burned.
- Take care when showering or bathing – check the water temperature.

Scleroderma

Occupational therapy intervention may need to be multifaceted. The following suggestions are based on the experience of occupational therapists working in this field. Controlled studies in management of scleroderma are lacking and are urgently needed.

Support

Support is vital not only from professionals on an ongoing and potentially long term basis, but also from other people with the same condition – this is where the Raynaud's & Scleroderma Association can help.

Hand Changes

In nearly all cases of scleroderma, skin thickening begins on the hands and fingers (Ref.6).

Limitations in range of movement are as a result of fibrosis of soft tissues: skin, fascia, muscle, tendon and joint capsule. A common early symptom is swelling of the hands and feet, particularly in the morning.

Intervention might include:

- Hand Therapy (Refs.7 & 8) – including exercises to maintain range of movement, joint protection education, and skin care. Tissue mobilization by a physiotherapist would be complementary and beneficial.
- Splinting (Ref.7) – few research studies have been carried out into the effectiveness of resting or dynamic splints - further studies are needed.
- Thumb web orthoses – as loss of thumb abduction is a commonly occurring change, and as thumb function represents approximately 45% of all hand function, it is important to maintain lateral pinch. A C-bar type splint may be useful in achieving this goal (Ref.7).
Care needs to be taken when splinting in terms of temperature of the material being applied to a potentially numb hand. Written instructions regarding wearing regime and precautions should be issued to remind the patient to be vigilant and take appropriate action if adverse reactions occur.



C-Bar splint

- Calcinosis – the deposition of chalky deposits under the skin, often of the digits but may also occur around other joints, such as the elbow. The use of ring padding type protection may be offered, the use of bi-valved finger gutters, or provision of Coban self adhesive tape may also offer some protection during activities. Digital sleeves, such as the type produced by Silipos which are lined with a medical grade oil, can be helpful in protecting dry and cracked fingers and/or toes.
- Finger Ulcers – manage with protective padding to decrease pain during activity or by wearing thin cotton gloves.
- Telangiectasia – laser treatment or cosmetic camouflage may help.

Elbow

A volar based, resting elbow splint could be constructed if elbow extension is compromised.

Education and Assistive Equipment

Education regarding the condition, pacing of activities of daily living, pain management, joint protection techniques, energy conservation and sleep hygiene, should be included to promote self-efficacy.

Patients with fixed changes of the hand, wrist or upper limb may need help in the form of assistive equipment in order to maximize function. Equipment may also be helpful if other joints are involved, such as those of the feet. After thorough assessment, identification of the most helpful items can be made. The patient should be encouraged to try out the equipment in their home environment before making a final decision on its appropriateness. Assistive equipment can also help with energy conservation and joint protection, for example: the use of elastic shoelaces or levers for keys and taps. It may also help people to function more safely in their home environment – the use of a kettle tipper may reduce the risk of scalds. If the person has oesophageal reflux raising the head of the bed with blocks can help (Ref.9).

If an adaptation is not commercially available, the occupational therapist may consider constructing a 'one off' adaptation themselves. Alternatively, referral to and working in partnership with local REMAP Panel (Ref.10), may be more appropriate.

Environmental Adaptation

- Home – requires liaison between hospital and community based occupational therapists.
- Work – work based assessment may be required – working in partnership with the patient, employer and possibly a Disability Employment Advisor.
- Leisure – for example – maintenance of driving skills – referral to a specialist mobility centre for assessment regarding car adaptation might be needed.



Facial Involvement

Facial exercises to maintain mobility of the facial muscles (Ref.7) and cosmetics to camouflage telangiectasia can help maintain a patient's self esteem and confidence. Exercises can also help with the ability to maintain good oral hygiene, eating and chewing.

The following is advice on maintaining facial mobility through exercise.

Do exercises in front of a mirror. Massage (firm touch) the entire face using small circular motions with the finger tips, a warm face cloth or vibrator, then massage each specific area again just before exercising that part. The number of repetitions necessary to get maximum mobility, depends upon the individual. One approach, is to do the exercise fast two or three times as a warm up, and then do five repetitions holding each stretch position to the count of five. Sustained stretch is more effective for increasing mobility than rapid motions.

Facial Exercises

Raise the eyebrows as high as possible. Return to the normal position.

Bring the eyebrows down and together as hard as possible as if frowning. Then raise eyebrows as high as possible.

Wrinkle the bridge of the nose by raising the upper lip and then frowning (as if smelling something bad).

Close the eyes very tight. Then release the squeeze slowly and raise the eyebrows as high as possible, before opening the eyes.

Flare the nostrils, then narrow the nostrils down, pushing the upper lip out.

Make an exaggerated tight wink with each eye separately, using the cheek muscles to help close the eye.

Cover the teeth with the lips, then open the mouth as wide as possible without the teeth showing.

Close lips and press hard (as if blotting lipstick).

Open the mouth so that the lips are as wide apart as possible.

Open the mouth so that the teeth are as far apart as possible.

Push the jaw forward to create an under bite (bottom teeth in front of the upper teeth).

Make as wide a grin as possible without showing the teeth.



Hand surgery

Hand Surgery

This may be an option for some patients in the form of:

- Digital sympathectomy
- Revascularisation of the fingers
- PIP fixation
- MCP surgery to excise bone and create a gap at which movement can occur
- Carpal tunnel release
- Darrach's procedure

Purpose of hand surgery: (Refs. 8 & 11)

1. Alleviation of pain
2. Prevention of tissue loss
3. Preservation of function
4. Cosmesis



Tips for Patients

Occupational Therapists are employed in a variety of health and social care settings. If you would like to be assessed and treated by an Occupational Therapist in a Rheumatology Unit, your General Practitioner, Consultant or Clinical Nurse Specialist may be able to refer you to a suitable centre. Occupational Therapists who are community based are usually employed by Social Services. They can advise you if necessary, on major adaptations to your home and also give advice regarding eligibility for grants towards such adaptations. People can self-refer to Social Services Occupational Therapy by contacting their local Social Services office. Local numbers can be found in your telephone book.

Occupational Therapists treat the patient as a 'whole person'. This means, when assessing and treating you they will address physical issues such as hand changes, as well as psychological ones e.g. the stress of coping with scleroderma. They will also work with you to problem solve difficulties in all aspects of your life - your self-care, leisure and productivity roles, which includes paid or unpaid work and management around the home.

For advice on keeping warm, contact the Raynaud's & Scleroderma Association. The Association publishes literature containing details of gadgets and heating aids. Publications on Raynaud's, scleroderma and associated conditions are also available and provide a valuable source of information to patients and health professionals alike.



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