

The Role of the Physiotherapist



Raynaud's & Scleroderma

Published by

Raynaud's & Scleroderma Association

Charity Reg 326306



Support for sufferers

The Raynaud's & Scleroderma Association can give emotional support and practical advice to sufferers 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.

Our aims are to promote a greater awareness of Raynaud's & scleroderma, to raise funds for research and for the welfare of patients. As with all self help groups, members usually find great comfort in contacting others who understand their problems.

We have the full support of a group of doctors who act as medical and scientific advisors to the Association.

Anne H Mawdsley MBE, Director of The Raynaud's & Scleroderma Association would like to thank Jill Lloyd for her support in the preparation of this publication.

For further reading: *Connective Tissue Disorders in Rheumatological Physiotherapy*, Chapter 13. Eds: Carol David and Jill Lloyd. Mosby, 1999.

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

Raynaud's & Scleroderma Association

112 Crewe Road

Alsager

Cheshire

ST7 2JA

Tel: 01270 872776

Fax: 01270 883556

e-mail: info@raynauds.org.uk

website: <http://www.raynauds.org.uk>

*This booklet has been sponsored by
an educational grant from*

Actelion Pharmaceuticals UK

The role of the physiotherapist

This booklet has been written as a brief guide for the physiotherapist who may be involved in the management of patients with Raynaud's and scleroderma.

Raynaud's is very common but is closely linked to scleroderma. It is therefore important to understand that Raynaud's can be of two types. It can be a phenomenon in itself, when it is usually benign or can be secondary to other conditions such as scleroderma and systemic lupus erythematosus.

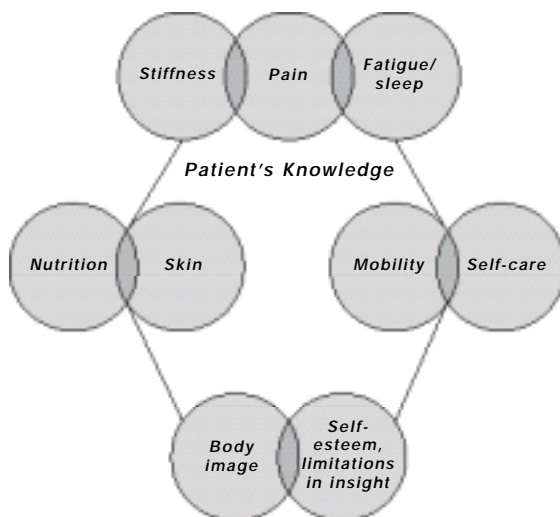
Scleroderma is a generalised disorder of the connective tissue. Fibrotic and degenerative changes occur in the skin, synovium, digital arteries and internal organs including the oesophagus, intestinal tract, heart, lungs and kidneys. It affects about 20 per one million of the population per year and is three times more common in women than in men.

Scleroderma is usually managed as an out patient with referral to specialist centres. Some patients, however, may be admitted to wards for therapeutic drugs and for complications of their condition. It is essential that this rare condition is recognised and understood by the physiotherapist.

The physiotherapist has a very important role in encouraging the patient to remain positive and in control of their condition. Through the provision of knowledge about the disease and the judicious use of heat, exercise and stretching etc. the patient will be able to be active in self care and retain an internal focus of control.

A therapeutic approach

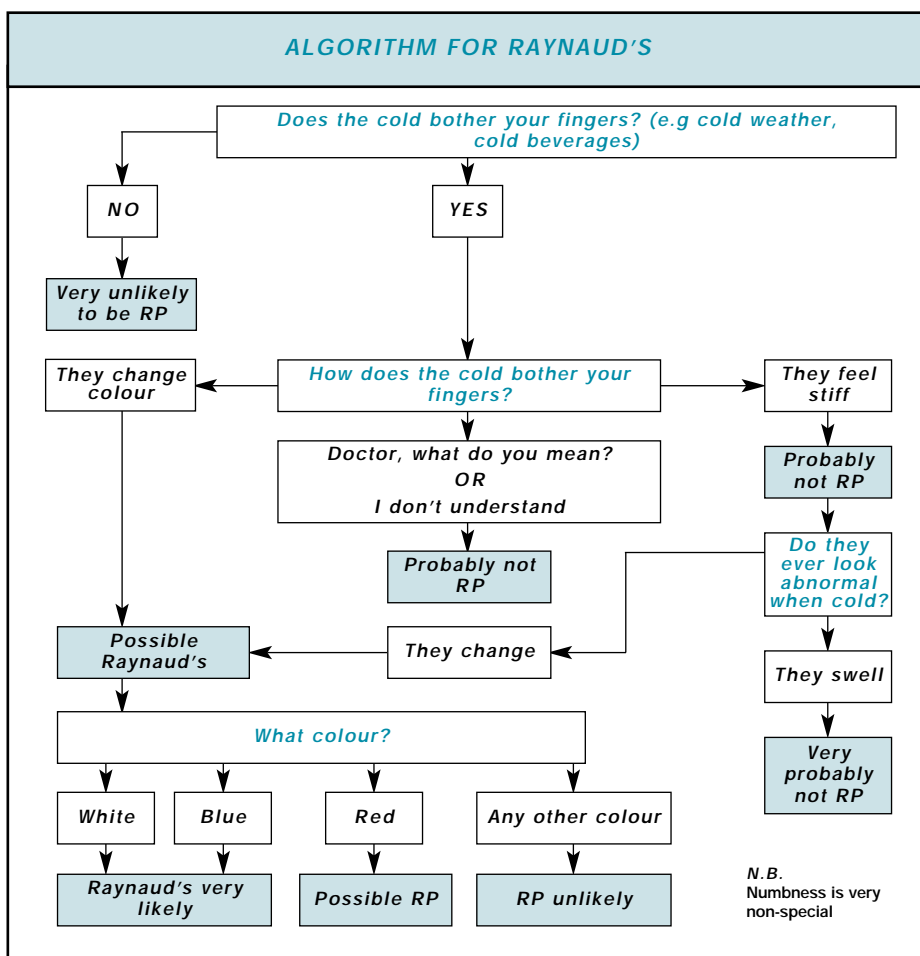
As with all chronic disorders the persons needs must be considered on an individual basis and assessment of physical, psychological and social wellbeing are all important. It is useful, therefore, to refer to the rheumatology nursing forum model. This is based on patient problems and ensures that a shared therapeutic approach is taken to core planning (see model).



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 patients with scleroderma. In Raynaud's, the blood supply to the extremities, usually the fingers and toes but sometimes also the ears and nose, is interrupted. During an attack there is a tightening or constriction of the arteries which normally supply blood to the affected parts causing them to first become white and dead looking. They may then turn blue as the tissues use up the oxygen and finally bright red when the artery relaxes again. This is often accompanied by a burning feeling, considerable pain or numbness.

Having numb fingers, feeling constantly cold or having chilblains does not necessarily mean that the patient has Raynaud's phenomenon.





What triggers a Raynaud's attack?

An attack will often be triggered by touching cold objects, going into a cold atmosphere or more importantly by any slight change in temperature. The physiotherapist must assess the level of sensitivity of the Raynaud's and act accordingly. The administration of therapeutic ice is not indicated.

Stress and anxiety can also play a part. Patients have much to consider when attending hospital and clinic appointments and very often their fears and worries can make their condition worse.



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 very 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.



What are the main symptoms?

Skin change is the most common presenting feature. The skin on the hands, feet and face become 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, this is due to obliteration of the sebaceous glands by the connective tissue. Ulcers, calcinosis (deposits of small amounts of calcium) and pitting of the skin are all important features of scleroderma.

The ulcers may take a long time to heal due to the compromised circulation and are a consideration for the physiotherapist.

Microstomia (or small mouth) is often a common feature in scleroderma and can make eating difficult. It will also reduce the mobility of the mouth, impairing speech and making oral hygiene difficult. Regular dental check ups are recommended.

Dry mouth and dry eyes, known as Sjögren's Syndrome, another auto-immune disease which is related to scleroderma can be a problem.

Patients may feel very tired and exhibit signs of synovitis and myalgia. They may also complain of shortness of breath. These are all areas where the expertise of the physiotherapist is vital.

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

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, telangiectasia and gastrointestinal involvement.

A high incidence of ACA (70-80%)

Dilated nailfold capillary loops, usually without capillary dropout.

Scleroderma sine scleroderma

Raynaud's +/-

No skin involvement

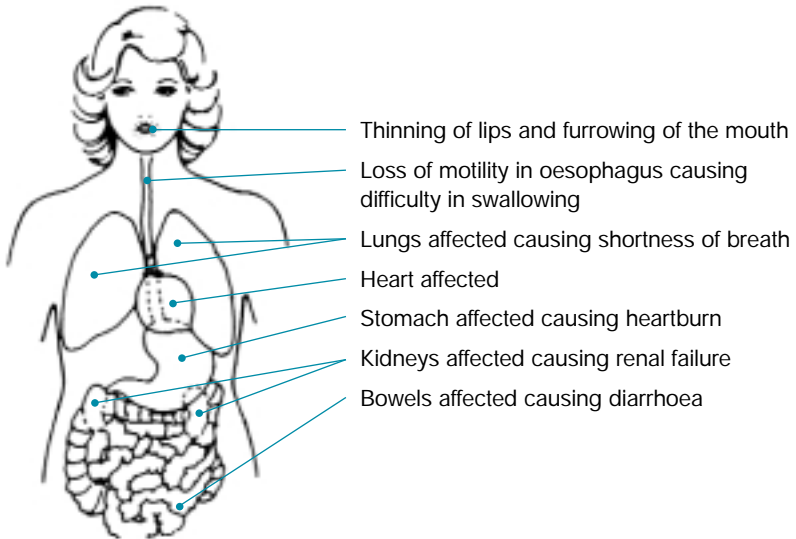
Presentation with pulmonary fibrosis, scleroderma renal crisis, cardiac or gastrointestinal disease. Antinuclear antibodies may be present (Scl-70, ACA, nucleolar).

Organs affected by scleroderma

Lungs

The lungs are very commonly involved in scleroderma but not everybody will develop lung disease which affects them symptomatically. Studies of lung fibrosis in conditions other than systemic sclerosis have suggested that the earlier the disease is detected, the more likely there is to be a good response to treatment. Breathlessness could be indicative of either fibrosis of the lungs or pulmonary hypertension.

Cardiopulmonary failure has now surpassed renal failure as the leading cause of death in patients with scleroderma.



Kidneys

Kidney and heart problems can also be serious. The development of fibrous tissue in the kidneys with reduced local blood supply, may lead to sudden dramatic failure of the kidneys. Treatment for scleroderma kidney has improved greatly over the past few years but patients should be closely observed for any sign that the kidneys are involved.

Heart

Problems with the heart, although rarely affected, occur due to thickening of tissue and reduced blood flow within the heart.

Bowels

Patients find bowel problems an embarrassing subject and do not always tell their doctors that they have a problem. They also find it difficult to discuss this with nursing staff. Both constipation and more often diarrhoea are common, along with the involvement of the duodenum and colon which results in a reduction in the absorption of food. This consequently leads to loss of weight and should be monitored carefully.

Absorption of nutrients

Food passes through the mouth, oesophagus and stomach to the small intestine, whose lining secretes many enzymes and is covered by tiny projections (villi) which enable nutrients to pass into the blood.

Malabsorption

Enzyme action breaks food down into molecules that can be absorbed. Lack of certain enzymes may cause malabsorption of nutrients. Other causes include flattened villi and scars on the intestine.

Limited Cutaneous Systemic Sclerosis

Symptoms of the limited form of scleroderma previously called the CREST syndrome are:-

C

Calcinosis which is an accumulation of calcium below the outer layer of the skin.

R

Raynaud's phenomenon – a condition in which the blood supply to the extremities, usually the fingers and toes, is temporarily interrupted.

E

Esophageal (gullet) involvement, causing difficulty in swallowing or indigestion. (Note the 'E' is from the American spelling of oesophagus).

S

Sclerodactyly is when the skin of the digits becomes thin, shiny and leathery looking. Fingers and toes may become flexed and stiff.

T

Telangiectasia is the appearance of small blood vessels near the surface of the skin. These can be seen on the fingers, palms, lips, face, tongue and chest wall.



Management of scleroderma

Following assessment, management will be discussed with the patient with priority areas highlighted. Maintenance of independent function will be a main priority. Exercises which can be performed independently by the patient should be taught. These should be directed to maintaining and increasing range of motion and muscle power. Stretches for all affected areas are vital and may need to involve a partner or friend to assist. Accurate and thorough teaching is essential. Persistence is vital, particularly when evidence of progress may not always be apparent.

Exercises and stretches for hands, face and feet are essential. All joints and muscle groups should be incorporated.

Exercise for the lungs and the respiratory muscles should also be taught concentrating on lung expansion. Aerobic activities to induce deeper breathing could be suggested, e.g. swimming and walking.

Application of wax to the hands and feet has been found to be beneficial by some patients. Clearly attention to the temperature of the wax is vital. The oils in the wax and the warmth serve as a useful preparation for the exercise. Wax can be applied daily and can be used with care in the home.

Patients find massage soothing and some therapists may use aromatherapy oils to assist. Massage of the skin will help to mobilise stiff connective tissues as well as having a beneficial effect on the circulation and nutrition of the skin. Teaching patients to massage themselves helps to increase independence and control. Alternatively a partner could assist.

The use of heat can assist in preparing muscles for exercise and is a modality much favoured by patients with scleroderma. Hot water bottles and electrically heated pads are useful. However, a hot bath or shower is also beneficial.

Hydrotherapy is to be recommended. Some patients find the temperature of the water is insufficient to prevent an attack of Raynaud's. If a pool of suitable temperature is available, swimming and exercise in water may help to preserve range of motion, as well as preventing decrease in lung volume. The physiotherapist should watch for fatigue in the patient and all activities must be tailored to the patient's exercise tolerance with an eye open for improvement.

Referral to an occupational therapist may be appropriate for some patients. Splinting to prevent deformity has never been proven to be useful, but it may be helpful in assisting activities. The occupational therapist can also advise the patient about pacing of activities to preserve energy and any adaptations and gadgets to preserve independence.

General advice about their condition from other members of the multi-disciplinary team should always be endorsed to the patient.

Hand exercises

The following exercises should be performed at least twice a day, doing ten of each exercise per session. You will find that the exercises are easier if your hands are warm and the skin is made more supple by using moisturising cream or lanolin. In addition to exercises, it is important that you use your hands and arms as much as possible, to perform your everyday tasks.



Bend your fingers into the palm of your hands and make a fist. Keep the thumb out of the palm. Straighten all your fingers out and repeat.



Begin with your fingers straight. Bend all fingers at the knuckles, keeping all other joints straight. Return to starting position and repeat.



Bend your thumb over to the base of your little finger and repeat.



Put your hands together, and make a diamond shape, keeping the wrists together and finger tips together just moving the knuckle joint.

- Stretch your fingers out, separate your fingers and then bring them together again.
- Touch the tip of each finger in turn with your thumb and push them together, then stretch out.
- Support your forearms on your thighs, palms facing the floor and raise your hand backwards from your wrist.
- Regularly stretch your fingers on a flat surface such as a table top.

Breathing exercises

These exercises can be performed sitting, lying on your back or lying on your side.

Place your opposite hand around the opposite part of your chest feeling for the ribs beneath. Breathe out, then take a deep breath pushing your hand away and then relax the breath out.

NB: No more than four deep breaths in any one go.

Elbow exercises

- Keep your elbows tucked into your side, with your forearms parallel to the floor, turn your palm over so that it faces the ceiling and then the floor.
- Bend and straighten your elbows.

Shoulder exercises

These are best done sitting over the edge of the bed.

- With the elbows outstretched raise the arm to brush past the ear.
- Touch your hand behind your neck as low down as possible, but making sure that your elbows are pointing out sideways as far as possible.
- Touch your hand behind your waist, easing your hand as far as possible up your spine. Take your arms up to the side until they are parallel to the ground. Turn your palms upwards and raise your arms up towards the ceiling. Then raise your arms towards your ears.

Leg exercises

These exercises are best performed sitting with legs outstretched on the bed, supported by three or four pillows.

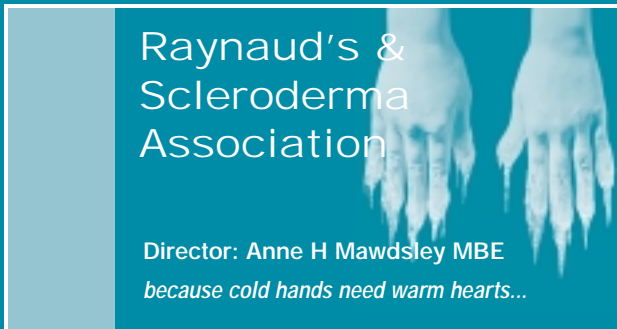
- Bend your knee up towards your armpit and lower again out straight.
- With your knee straight, lift your leg so that your heel is just clear of the bed, then take the leg out to the side and then back in again.
- With your feet a little way apart, roll your legs in and out.
- Pull your ankles up towards you and press your knees into the bed, hold and relax.
- Roll a pillow up and place it behind your knee, straighten your knee over the pillow, hold and relax.
- Pull your ankles up towards you and then point them towards the end of the bed.
- Bend and stretch your toes.
- Circle your ankles.

Check with a physician before starting on any exercise program



Whirling of the arms can be effective in stimulating circulation to the hands

The physiotherapist has a very important role in encouraging the patient to remain positive and in control of their condition.



Published by

Raynaud's & Scleroderma Association

112 Crewe Road

Alsager

Cheshire ST7 2JA

email: info@raynauds.org.uk or info@scleroderma.org.uk

website: www.raynauds.org.uk or www.scleroderma.org.uk

Charity Reg No 326306