

# **The Role of the Nurse**

**(in Raynaud's and Scleroderma)**



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# SUPPORT FOR SUFFERERS

The Raynaud's & Scleroderma Association (RSA) offers support and practical advice to people 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.

## AIMS OF THE RSA

- Creating a greater awareness of Raynaud's and scleroderma.
- Promoting better communication between doctors and patients.
- Putting patients in touch with each other in order to exchange ideas and information.
- Offering support, advice, and information to sufferers, their families and carers.
- Raising funds for research and welfare projects, as research is absolutely essential if we are to determine the cause and find better treatments in the future.

Patient leaflets on Raynaud's and all aspects of scleroderma are available from:

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# THE ROLE OF THE NURSE

Raynaud's phenomenon and scleroderma are two conditions about which many nurses know very little. Scleroderma is an uncommon connective tissue disease, which few nurses will have dealt with on a regular basis. Raynaud's phenomenon is a much more common condition but is often not diagnosed and treated. This booklet is designed for nurses who want to learn more about both conditions. It covers the basic medical aspects and the role of the nurse in caring for patients with Raynaud's and scleroderma.

Nurses looking after a patient with Raynaud's or scleroderma have a responsibility to ensure that they have a basic understanding of the health problems which the patient may currently be suffering from or may develop in the future. This is in order to allow them to assist the patient in planning care.

## WHAT IS RAYNAUD'S?

There has been much confusion over the name and more recently the word 'Disease' has been dropped, the condition simply being called Raynaud's or Raynaud's phenomenon. Raynaud's can be subdivided into primary and secondary.

Raynaud's phenomenon is a common, episodic circulatory disorder in which the small blood vessels in the extremities are over-sensitive to changes in temperature. It affects between 3-20% of the adult population worldwide and there may be as many as ten million sufferers in the UK. Raynaud's is most commonly found in females and approximately 10% of women suffer from Raynaud's to some degree. The condition can affect children, adolescents and adults. Many sufferers have never seen a doctor as they are unaware that their condition has a name or that there is anything that can be done to help. However, for a small number of people, Raynaud's is one of the earliest symptoms of impending scleroderma and for this reason Raynaud's patients require prompt assessment and diagnosis.

The hallmark of Raynaud's is biphasic or triphasic colour changes (white and/or blue and red) of the extremities on exposure to the cold, or to sudden but slight temperature changes or stress. The symptoms usually occur in the fingers and toes but ears, lips, nose, nipples and penis may also be affected. The patient may also complain of pain, numbness or tingling.

The symptoms of Raynaud's, whether isolated (primary Raynaud's phenomenon) or secondary to another condition, may cause severe pain, discomfort and problems with hand function. For the vast majority of sufferers, Raynaud's is a benign primary condition which may interfere with patients' daily activities but does not cause any long term damage to the extremities. However, patients who have Raynaud's phenomenon secondary to an underlying disease such as scleroderma, will often suffer more acute symptoms and in severe cases may develop persistent finger and/or toe ulcers, infection and ultimately gangrene.

## TREATMENT FOR RAYNAUD'S

Many treatments are available for Raynaud's phenomenon. Patients should be advised that practical measures such as using hand warmers, thermal gloves and hats often help to alleviate symptoms. An even ambient temperature is as important as keeping warm because often it is not the absolute temperature, but a small change in the ambient environment, which precipitates an attack. Cold draughty places should therefore be avoided where possible.

All patients should be strongly encouraged to stop smoking as it causes narrowing of the blood vessels, and to avoid excess caffeine, which may also cause peripheral vasoconstriction. Many patients with Raynaud's like to try natural products such as vitamins and fish oil or evening primrose oil, ginkgo biloba and ginger. These simple measures seem to help some patients and are popular as they can be purchased without prescriptions and do not have side effects. Many conventional vasodilator drugs are also available but patients often have to try several before they find one that works.

## WHAT IS SCLERODERMA?

Confusion often surrounds the name of the disease which is also called systemic sclerosis. The word scleroderma is derived from the Greek 'sclero' meaning hard and 'derma' meaning skin. In fact, although the term scleroderma is often used as if it were a single disease it is really a generic or umbrella term covering a number of related diseases that involve the overproduction of collagen. Collagen is the major protein of the connective tissue which binds the body together. It is found in the skin, blood vessels, joints and internal organs. In this condition the skin, usually of the hands and feet becomes stiff, tight and shiny. This is because of swelling and then thickening of the connective tissue which becomes fibrotic or scarred. Internal organs can also be affected in a similar way.

## WHAT CAUSES SCLERODERMA?

Like many other rheumatic disorders (conditions characterised by inflammation and/or pain in the joints, muscles and tissues) scleroderma is believed to be an auto-immune disease. The immune system becomes dysfunctional and stimulates cells called fibroblasts to produce excess collagen, which is then laid down in the connective tissues causing them to become thickened, hard and fibrosed. The connective tissues are those tissues that provide the supportive framework (musculoskeletal system) and protective covering (skin and mucous membranes and vessel linings) for the body.

The exact cause of scleroderma is unknown but female hormones probably play a role in the development of the condition and genes may also be a risk factor. Research suggests that exposure to some environmental factors and viruses may also trigger the condition.

# DIFFERENT TYPES OF SCLERODERMA

There are several different types of scleroderma – in the localised forms of the disease hard, tight skin is the extent of the problem, while in the systemic forms such as diffuse scleroderma and limited scleroderma, the problems affect the internal organs.

## Scleroderma Spectrum of Disorders

### Raynaud's Phenomenon

Primary Raynaud's Phenomenon  
Auto-immune Raynaud's Phenomenon

### Localised

#### Morphoea

*Localised*  
*Generalised*

#### Linear scleroderma

*En coup de sabre*

### Systemic

Limited cutaneous systemic sclerosis  
Diffuse cutaneous systemic sclerosis  
Scleroderma sine scleroderma  
Overlap syndromes

## Systemic sclerosis

The most common sub-type of scleroderma is the systemic form of the disease which can be sub-divided into diffuse cutaneous systemic sclerosis (dcSSc) and limited cutaneous systemic sclerosis (lcSSc). These two types have different disease progression and organ involvement.

### ***Diffuse Cutaneous Systemic Sclerosis***

Skin changes within 1 year of onset of Raynaud's

Skin thickening and tightening all over body, often causing reduction in range of movement

Significant incidence of lung, kidney, gastrointestinal and myocardial involvement

Joint and muscle pain

## **Limited Cutaneous Systemic Sclerosis (previously known as CREST syndrome)**

Long standing Raynaud's phenomenon

Skin involvement limited to hands, face, feet, forearms and lower part of legs

A significant late (10-15 years) incidence of pulmonary hypertension, skin calcifications, telangiectasia and gastrointestinal involvement. Some lung fibrosis.

## **Localised scleroderma/linear scleroderma/morphoea**

In these conditions only the skin and underlying tissues, muscles and sometimes bones are involved, the organs are not affected. However some blood tests may be positive and some patients may have Raynaud's phenomenon. This is the type more commonly found in children.

# TREATMENT FOR SCLERODERMA

Although there is still no cure for scleroderma, there are treatments available that aim to arrest the disease process, both in the skin and internal organs. This can help to relieve symptoms suffered by the patient.

Immunosuppressive drugs are used for patients with diffuse scleroderma to attempt to slow down the disease process. The main immunosuppressive drugs currently used are mycophenolate mofetil, cyclophosphamide, methotrexate and azathioprine. The choice of immunosuppression usually depends on the patient's organ involvement.

### **Organ-specific therapies are given to reduce symptoms, some examples are:**

- proton pump inhibitors to alleviate gastro oesophageal reflux and motility drugs to help with dysphagia.
- antibiotics to help with bacterial overgrowth in the bowel
- ACE-inhibitors to prevent and treat vascular damage in the kidneys and help reduce symptoms of Raynaud's phenomenon
- Iloprost, a potent intravenous vasodilator to improve blood supply to ulcerated fingers. (A nebulised version is also available)
- Bosentan, Sitaxentan and Sildenafil - oral medications for pulmonary hypertension

However, it is important to realise that not every patient will be affected by every problem and that the course of the disease will vary with each patient.

# ORGAN INVOLVEMENT

In addition to the fibrosis caused by excess collagen, scleroderma also causes damage to both the small and large vasculatures resulting in circulatory problems. This means that potentially any organ of the body can be affected by either fibrosis (scarring) or vascular damage.

## Upper and Lower Gastro-intestinal Tract

Almost all patients with scleroderma suffer from problems in the GI tract and most will be on some medication to help alleviate symptoms. Fibrosis of the GI tract can cause symptoms ranging from difficulty swallowing and acid reflux to diarrhoea, constipation, malabsorption, malnutrition and faecal incontinence.

Patients who suffer from heartburn should be encouraged to eat little and often, perhaps five small meals each day rather than two or three large ones. Acid production can be reduced and the problems of acid reflux and heartburn alleviated by avoiding alcohol, greasy, fatty or spicy foods, tobacco and coffee. Most patients find it beneficial to ensure that they are sitting upright during meals and for a period afterwards. Raising the head of the bed six inches will also help patients who suffer badly from heartburn overnight. Eating slowly and chewing food well will help to prevent swallowing difficulties, and dry foods, like crackers or bagels, should be avoided. The patient's own experience should always be taken into account and to some extent trial and error is necessary to find which foods individuals can manage best.

The nurse is also well placed to discuss with the patient any problems with diarrhoea, constipation or faecal incontinence. Diarrhoea is often caused by too many bacteria in the bowel which can be dealt with by antibiotics. Constipation is a common feature of scleroderma due to the stiffening of the bowel and although laxatives are often prescribed, the nurse should advise the patient on non-medical ways of lessening constipation. A diet high in fibre, fresh fruit and water will help with constipation as should drinking 6-8 glasses of fluid each day and taking plenty of exercise.

## Lungs

Lung disease in scleroderma can take two forms. The first is pulmonary fibrosis which is replacement of the airspaces in the lung by scar tissue. The second lung complication of scleroderma is pulmonary arterial hypertension (PAH) in which damage to, or stiffening of the vessel walls lead to high pressure in the pulmonary artery. This puts strain on the heart resulting eventually in right sided heart failure. Both complications have a fairly poor prognosis although treatment may slow down progression and reduce symptoms. Fibrosis is treated with immunosuppressive drugs while vascular drugs (such as Iloprost, Bosantan, Sitaxentan and Sildenafil) are used to treat PAH.

## Kidneys

Scleroderma causes vascular damage and when this damage occurs to the small blood vessels in the kidneys they lose their ability to filter the blood and remove waste from the circulation. About 10% of patients with diffuse scleroderma develop severe kidney problems which may result in a renal crisis and dialysis. A renal crisis can develop very quickly so it is important that nurses are aware of the signs. These are high blood pressure (above 150/90) resulting in shortness of breath, headaches and blurred vision. Any patient complaining of these symptoms should have their blood pressure checked immediately and if it is high it should be brought to the attention of a doctor without delay. If this is not possible then the patient should attend A&E and tell nursing staff that they suffer from scleroderma.

## Heart

Problems with the heart although not common, may include cardiomyopathy, myocarditis, and arrhythmias. However these conditions are often hidden and are not picked up until the patient is suffering severe, life-threatening symptoms.

# OTHER COMPLICATIONS

## General

Patients with scleroderma may experience a variety of non-specific symptoms, including fatigue, lack of energy, generalised weakness, weight loss and aching of muscles, joints or bones. The role of the nurse is to help patients to develop managing strategies for these symptoms. This may include discussion about:

- Reducing or completely stopping work
- Reorganising their daily routine and responsibilities to include time to rest
- Developing alternative ways of doing things
- Obtaining outside help with some tasks

It is important that family members are included in this period of discussion and planning, in order to both provide support and motivation to the patient, and to allow the family to understand and be involved in the patient's illness.

## Skin care

Skin changes are the most common presenting feature of scleroderma with the skin becoming tight, shiny, dry, scaly, itchy, tender and thickened. This is usually more of a problem for patients with diffuse scleroderma. Skin thickening can also contribute to loss of some movement, making day-to-day functioning more difficult. All patients should therefore be encouraged to moisturise their skin several times a day using a good moisturiser - oil-based creams are usually the most effective. Putting oils and emollients into bath water is also a good way to keep the skin soft.

Hand and face exercises (available from a physiotherapist, or from specialist scleroderma units) should be encouraged. The patient should be made aware that the earlier they start doing exercises and the more diligently they carry them out, then the less likely they are to have problems with loss of hand function over time. The nurse supporting a scleroderma patient is in an ideal position to help alleviate some of the discomfort which is being experienced.

If the patient has a problem with itchy skin (more common in the first few years of active disease) itch-relief or anti-histamine creams available over the counter can be applied and oral anti-histamines can also be taken if required. The nurse should also take the opportunity to reassure the patient that the early aggressive phase of the disease should settle with time and may improve with medical intervention.

## Telangiectasia

Telangiectasia is the medical term for the dilated red blood vessels on the skin which appear on the face and hands. Other areas that may be involved are the stomach and intestine. Many patients find that telangiectasia although painless cause a great deal of distress as they are unsightly. Green make-up which helps to tone down redness is readily available from most cosmetic companies and cosmetic camouflage make-up is also very effective. Patients can be referred to a dermatology unit which offers pulsed dye laser therapy - short bursts of high energy light which obliterates the burst blood vessels.

## Pigmentation

Hypo (too little) or hyper (too much) pigmentation may be a problem in both diffuse and limited scleroderma and does not usually resolve or improve. Camouflage make-up may help. The Red Cross run clinics in hospitals to teach patients how to apply it correctly and nurses can refer directly to these clinics.

## Calcinosis

Approximately 20% of patients with limited scleroderma develop calcinosis - deposits of calcium salts under the skin mainly occurring in the fingers and over bony prominences such as elbows. The lumps may break through the skin and drain white material. They may also be a cause of infection and ulceration. Patients can be taught to use paraffin wax treatments to bring the calcinosis to the surface.

## Ulcers

Patients sometimes develop ulcers on their digits, due to a combination of vascular damage caused by the scleroderma and periods of deoxygenation of the tissue caused by Raynaud's attacks. Ulcers are usually initially triggered by trauma to the digit. Prevention of ulcers is possible to some degree by keeping the skin on the hands in good condition and by using gloves when necessary. Vasodilator medications (which patients are often taking for Raynaud's) help improve the supply of blood to the fingers.

If a patient does develop an ulcer then firstly any infection should be treated with antibiotics. Flucloxacillin 500mg (1-2g daily) is the medication of choice but if there is any doubt then a swab can be sent for sensitivity. It is not currently recommended that patients soak finger ulcers, or that any dressing is used except a clean dry dressing. Wound healing is achieved by improving the supply of oxygen rich blood to the affected area. Hirudoid cream (a topical heparin preparation available over the counter) can be applied to the skin around the edges of the ulcer (not on the broken skin) to encourage blood flow to the ulcer. The nurse may need to bring the ulcer to the attention of a doctor and instigate a review of the patient's current medication. It may be possible that doses of vasodilators can be increased, or new vasodilators added. If refractory ulcers are present for some months and are resistant to all attempts at healing then the patient may require a hospital admission for intravenous iloprost.

## Joint Problems

Aches and pains in joints are common in scleroderma but are not usually associated with a true arthritis in most patients. Other problems are stiff joints and contractures.

In the early stages of the disease excess tissue inflammation and fluid causes feelings of stiffness and can result in patients being unable to make a fist properly. This stiffness is usually self-limiting and will disappear once the disease has been treated or is less active. Nurses can be instrumental in teaching patients to use paraffin wax treatments and perform regular hand exercises. Soaking the hands in warm water with an aromatherapy oil e.g. marjoram or massage will help to soothe and relax the hands. Simple painkillers like paracetamol may help.

Contractures occur when a joint loses part of its range of movement. In scleroderma they are usually due to swelling, skin thickening and tendon shortening not joint damage. Treatment consists of physiotherapy and daily exercises. Generally, hand splints have not been used for scleroderma patients but recent research has suggested that they may sometimes be helpful.

## Dry mouth and eyes

Good mouth care is essential for patients with scleroderma. Dry mouth and eyes [sicca syndrome] is a common problem and can cause difficulty in speaking and swallowing. Drug treatments for dry mouth [xerostomia] are ineffective and cause side effects but it is worth trying artificial saliva, chewing gum, and mouthwashes. Medication taken for other conditions may also cause a dry mouth so it is worth checking what medications patients are taking. Lack of saliva increases the liability to caries, infections and candida. Patients with scleroderma dental problems may need a referral to a specialist hospital and good mouth hygiene is essential to prevent problems occurring.

Mouth ulcers are often seen in immunosuppressed patients and are a common side-effect of many of the disease modifying drugs such as methotrexate and cyclophosphamide. The best treatments are those with a small amount of steroid such as Corlan®, pellets or Ad-cortyl in Orabase®, ointment available on prescription or over the counter. Anti-fungal therapy may be required to treat candida.

Patients should be advised to protect dry eyes with [sun] glasses, and to avoid draughts e.g. car heater. For the treatment of mild dry eyes, products like Liquifilm Tears®, are useful. Use every 30 minutes initially then reduce usage until the eyes feel comfortable. For severely dry eyes, a liquid gel like Viscotears®, or GelTears®, should be used 3 times a day. If preparations are to be used very regularly they should be preservative free but need to be kept in the fridge.

# A TOTAL PATIENT APPROACH

## Education

There is an increasing tendency for all patients to want to know more about their diseases and treatment, and part of the nurse's role in scleroderma is to support patients on their journey of discovery. It is particularly important in chronic diseases such as Raynaud's and scleroderma, to educate the sufferers about their disease and ensure that they are best placed to make informed decisions about their current and future care.

Because Raynaud's is often unidentified and scleroderma is uncommon, it is often the case that many doctors and nurses are unused to treating these conditions. It is to be hoped that most patients are given plenty of time with their hospital scleroderma specialists and specialist nurses and given the opportunity to ask all the questions they wish, both when they are first diagnosed and at all follow-up visits. Many patients will ask questions of their practice, community or ward nurse and it is important that these nurses are able to assist the patient. There are an increasing number of resources available to both patients and healthcare professionals, these include:

- Medical and nursing textbooks and journals
- Information leaflets
- Specialist nurses in dedicated scleroderma units
- Internet (many sites have an area dedicated to healthcare professionals)
- Patient support groups

An awareness of these resources will allow the nurse to empower the scleroderma sufferer to be active in the management of their own disease.

## Liaison

Although some patients may prefer to be given their diagnosis and discuss their medication with a doctor, they often find it easier to discuss other aspects of their condition with a nurse. This means that nurses are often the first point of contact for a patient and should be aware of procedures to enable the patient to contact someone who can help with problems. Sufferers of chronic illnesses such as scleroderma can sometimes find that they have difficulty accessing some of the services which they unfortunately find themselves in need of. For that reason some patients may require assistance in gaining access to GP surgeries when required, physiotherapy and occupational therapy, social services, counsellors etc. It is often part of the nurse's role to act as a liaison between hospital and community and to coordinate care between different members of the multi-disciplinary team.

## Patient advocacy and support

Nurses are in an ideal position to act as patient advocates. Healthcare professionals can sometimes be in danger of assuming only they know what is best for patients. This can lead to some patients feeling they have no say in contributing to their own care and that their freedom of choice has been removed. This is particularly the case for sufferers of scleroderma at the beginning of their illness when they are still coming to terms with a diagnosis of an incurable, sometimes fatal disease, about which they often know very little. However, by providing clear information and support, nurses can help patients, particularly those who are vulnerable and without the support of family and friends, to feel confident to make their own decisions. Because of the chronic and deforming nature of many rheumatological conditions, many patients need a lot of emotional support and reassurance and much care should be taken in addressing fully the psychosocial needs of this group. The nurse has a very important role in encouraging the patient to remain positive and in control of their condition.

Patients with a chronic disease also need a point of contact when they feel ill, anxious or depressed. For this reason there is a network of helplines around the country staffed by very knowledgeable nurses. These helplines are useful for healthcare professionals as well as patients.

The nurse must also be aware of the needs of the family of scleroderma sufferers. This is particularly important when the patient is a child. Childhood scleroderma is a very uncommon condition. Scleroderma in childhood is often localised and very rarely life threatening; yet because it is often a chronic illness its morbidity problems may pose additional psychological stresses which both patient and family members find difficult to handle. Children may feel isolated and come to resent the disease. Adolescence can be a particularly difficult time with the challenge of dealing with a disease that may make you appear different from others, at the same time as all of the usual teenage stresses. Any awareness of these psychological aspects must be taken into account whenever one is dealing with the individual patient, as well as a careful explanation of the disease process, its course, treatment and prognosis. The Raynaud's & Scleroderma Association is very keen to help patients and their families and put them in touch with each other if they so wish.

The role of the nurse in Raynaud's and scleroderma is not just restricted to the physical care of patients but it is essential to be able to offer a holistic approach to care, dealing with psychosocial aspects. The important thing to remember when looking after a patient with scleroderma is that not every patient will have the same experiences. They are likely to all suffer different symptoms and to have disease of varying severity. For this reason each patient's needs will be very individual and the nurse has a responsibility to carry out a full assessment of each person's abilities and needs and create a personalised care plan, identifying areas where the patient may require assistance with activities of daily living. Reassessment should be carried out frequently as the disease can develop rapidly and the patient may require additional support. As is the case with all chronic illnesses an effective relationship with a supportive, understanding and well-informed nurse can help the patient maintain dignity, function and a normal life.



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