Understanding & Managing Scleroderma

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Actelion Pharmaceuticals UK and Encysive Pharmaceuticals

Understanding & Managing Scleroderma

This booklet is intended to help people with scleroderma, their families, and others interested in scleroderma to better understand what scleroderma is, what effects it may have, and what those with scleroderma can do to help themselves and their doctors manage the disease. It answers some of the questions most frequently asked about scleroderma.

The Scleroderma Society wishes to thank Scleroderma Australia and the Scleroderma Foundation in the USA for allowing us to reprint this booklet. We would like to thank the Scleroderma Unit at the Royal Free Hospital and scleroderma specialists for helping to edit this book for a UK readership and all the people with scleroderma who have given us permission to use their photographs.

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Disclaimer

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Contents

What is scleroderma?

Scleroderma, or systemic sclerosis, is a chronic connective tissue disease generally classified as one of the autoimmune rheumatic diseases.

The word "scleroderma" comes from two Greek words: "sclero" meaning hard, and "derma" meaning skin. Hardening of the skin is one of the most visible manifestations of the disease. The disease may take several forms and internal organs are also frequently affected with the systemic form. The manifestations and effects of the disease may vary considerably between one patient and another. For example, scleroderma may be visible, as is the case when the skin is affected, or may be invisible, as when internal organs are affected.

What scleroderma is not

Scleroderma is not contagious, it is not infectious, it is not cancerous or malignant, and it is usually not hereditary.

How serious is scleroderma?

Any chronic disease can be serious. The symptoms of scleroderma vary greatly from individual to individual, and the effects of scleroderma can range from very mild to life-threatening. The seriousness will depend on what parts of the body are affected and the extent to which they are affected. A mild case can become more serious if not properly treated. Prompt and proper diagnosis and treatment by qualified doctors may minimize the symptoms of scleroderma and lessen the chance for irreversible damage.

Who develops scleroderma and when?

Approximately 1 in 10,000 people in the UK suffer from scleroderma, with women affected four times as often as men. This means there are approximately 6,000 – 7,000 persons with scleroderma in the UK. The complex nature of this illness can often lead to misdiagnosis so the true number of people with the disease may be much higher. The onset of scleroderma is most frequent between the ages of 25 to 55 but it may affect any age group, from infants to the elderly. Factors other than gender, such as race and ethnic background, may influence the risk of getting scleroderma, as well as the age of onset, and the pattern or severity of internal organ involvement. The reasons for this are not clear. Although scleroderma is not directly inherited, some

scientists feel there is a slight predisposition to it in families with a history of rheumatic diseases.

What causes scleroderma?

The exact cause or causes of scleroderma are still unknown, but scientists and medical investigators in a wide variety of fields are working hard to try and answer this question.

What is known about the disease process in scleroderma is that it involves three features:

1. An autoimmune process;
2. An overproduction of collagen;
3. Blood vessel damage.

Collagen is the major protein portion of the connective tissue of the body, which includes the skin, joints, tendons, and parts of internal organs. The structure of collagen is made up of tiny fibres, which are woven together much like the threads forming a piece of cloth. When there is an overproduction of collagen, thickening and hardening of the affected areas takes place, often interfering with the normal functioning of those parts.

There are several theories about how collagen is overproduced. The "autoimmune theory" suggests that the body's own immune system plays a part. Normally, the body's immune system produces chemical signals in the blood called cytokines, which coordinate the body's defence against bacteria, viruses, and other foreign invaders. In addition, some cytokines help to repair wounds by stimulating collagen production that forms a scar. There are a number of theories on the way in which the immune system is activated inappropriately, causing abnormal levels of cytokines to be produced. These, in turn, mount an attack not against a foreign invader but against the body's own healthy tissues, stimulating an overproduction of collagen.

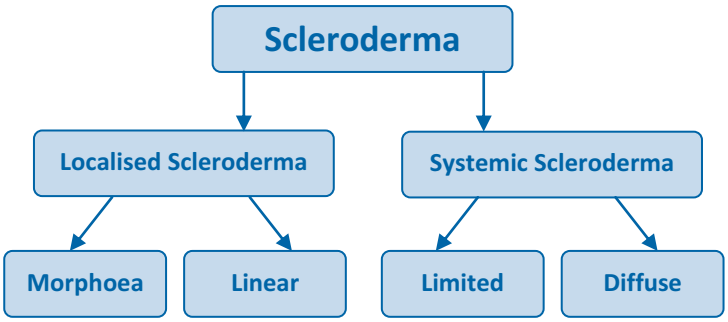
Another theory, the "vascular theory," concerns blood vessels. Damage to the blood vessels, especially the small ones, is typical in scleroderma. Injury to blood vessels causes them to constrict and stiffen and leads them to overreact to cold or stress. These reactions can cause further damage to the vessels themselves and to the organs, which they supply. There may also

The exact cause or causes of scleroderma are still unknown, but scientists and medical investigators in a wide variety of fields are working hard to try and answer this question

be a connection between the build up of excess collagen and blood vessels, the processes which take place, and their significance for prevention and treatment. Research is being done to study these and other theories. It is hoped that a better understanding of what causes scleroderma will lead to better treatment methods and, ultimately, to a cure.

Are there different forms of scleroderma?

There are two major classifications of scleroderma: localised scleroderma (morphoea) and systemic scleroderma (systemic sclerosis, SSc). Systemic sclerosis is further divided into limited systemic sclerosis and diffuse systemic sclerosis. Throughout this booklet, systemic sclerosis is referred to as systemic scleroderma. There is a third subset used by some specialists called “scleroderma sine scleroderma”, which means systemic scleroderma (scleroderma affecting the internal organs) without hard skin.



Morphoea or localised scleroderma

In this condition there are localised patches of thickened skin. The skin affected often appears waxy and may be red or brown in colour. These changes are usually found in only a few places on the skin or in the muscles, and only rarely spread elsewhere. The patches may enlarge or shrink, and often disappear spontaneously. Morphoea usually appears between the ages of 20 and 50, but can also be seen in young children.

Patients with this condition do not have Raynaud's phenomenon (see page 13) and very rarely have any internal organ involvement. The long-term outlook is excellent. People with morphoea rarely develop systemic scleroderma. Antinuclear antibodies that are normally found in the blood of people with systemic scleroderma are generally absent in people with morphoea or localised scleroderma.

Linear morphoea is a form of localised scleroderma which frequently starts as a streak or line of hardened, waxy skin on an arm or leg or on the forehead. Sometimes it forms a long

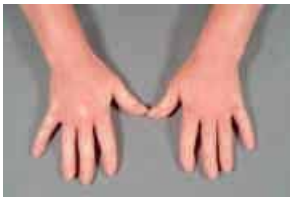
crease on the head or neck, referred to as “en coup de sabre” because of its resemblance to a sabre or sword wound. Linear scleroderma tends to involve deeper layers of the skin as well as the surface layers, and sometimes restricts the movement of the joints that lie underneath. Linear scleroderma usually develops in childhood. In children the growth of the involved limb may be affected.

Systemic scleroderma (systemic sclerosis)

The changes occurring in systemic scleroderma may affect the connective tissue in many parts of the body. Systemic scleroderma can involve the skin, gastrointestinal tract (oesophagus, stomach and bowels), lungs, kidneys, heart and other internal organs. It can also affect blood vessels, muscles and joints. The tissues of involved organs become hard and fibrous, causing them to function less efficiently. The term systemic sclerosis indicates that “sclerosis” (hardening, scarring) may occur in the internal organs of the body. There are two major subsets of systemic scleroderma - limited and diffuse. The difference between the two subsets is determined by the extent of skin involvement. Patients are classified as having limited cutaneous systemic sclerosis, (limited scleroderma), if the skin tightness affects only the face, the arms up to the elbows and legs up to the knees. Patients are classified as having diffuse cutaneous systemic sclerosis (diffuse scleroderma) if the skin thickening also involves the upper arms, thighs or trunk.

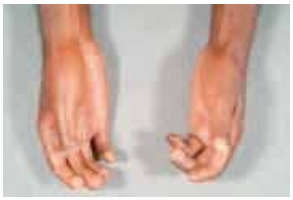
Limited scleroderma (limited cutaneous systemic sclerosis)

Limited scleroderma usually presents with Raynaud's phenomenon and hardening of the skin over the hands and sometimes over forearms, feet and lower legs. Almost always there are changes in the facial skin and facial appearance. Gastrointestinal problems are common and other internal organs can be involved. Although this involvement is often mild and may occur after many years of the disease, it is sometimes severe and life threatening. For that reason it is important to undergo regular internal organ tests, which are usually organised by your doctor. The onset of limited scleroderma is often very slow and may remain unnoticed until a specific internal organ complication occurs. Many patients have symptoms of Raynaud’s phenomenon for months or years before they develop any skin changes. Skin is generally mildly involved and usually does not change much over the years. About 70% of people with systemic scleroderma have limited scleroderma with the other 30% having the diffuse form.



The term CREST, or CREST syndrome, is sometimes used instead of limited scleroderma. It is the acronym for the clinical combination of **C**alcinosis (calcium deposits under the skin), **R**aynaud's phenomenon, o**E**sophageal problems, **S**clerodactyly (stiff fingers) and **T**elangiectasia (small dilated blood vessels in the skin). The usage of this term is now generally avoided because patients with this condition can have involvement of their internal organs and do not necessarily have all the components of CREST.

Diffuse scleroderma (diffuse cutaneous systemic sclerosis)



Diffuse scleroderma affects the skin not only on the hands, forearms, feet and lower legs, but it also affects the skin on the upper arms, thighs and trunk. Patients with this condition often have systemic involvement with the scleroderma process potentially affecting different internal organs, which can be life-threatening. This type of scleroderma often requires more intensive treatment with immunosuppressive drugs, even if there is no internal organ involvement. Diffuse scleroderma generally has a fairly rapid onset with the skin thickening and Raynaud's phenomenon occurring at the same time or one shortly after the other. The skin tightness and thickening can spread over the body within a few months of disease onset. It can remit after several years with softening of the skin and significant improvement in mobility.

It is important to remember that both limited and diffuse scleroderma may affect the internal organs and that severity of skin changes does not necessarily represent severity of internal organ involvement.

Although most patients can be classified as having either diffuse or limited disease, different people may have different symptoms and a different combination of symptoms of the illness.

How often does scleroderma affect different parts of the body?

Scleroderma is very variable but some features such as Raynaud's phenomenon or gastro-oesophageal reflux are much more common than others such as pulmonary hypertension or serious kidney disease. The approximate frequency of major features of scleroderma is summarised in the table below. For some complications of scleroderma there is a clear difference in frequency between limited or diffuse disease. It should be remembered that some of these features may be quite mild in some cases and that specific treatment may

not be required. In all cases it is important that regular tests are performed to check whether new complications have developed and our recommendation is for heart, lung and kidney tests to be performed at least once a year, even in cases where these tests have previously been normal.

Manifestation	Limited Scleroderma	Diffuse Scleroderma
Raynaud's phenomenon	95%	80%
Gastro-oesophageal reflux	75%	90%
Lung fibrosis	30%	30%
Heart	<5%	10%
Pulmonary hypertension	15%	5%
Kidney disease	<5%	20%
Telangiectasia	91%	64%
Calcinosis	42%	17%

Table 1 Approximate frequency of major features of scleroderma

How is scleroderma diagnosed?

Diagnosis of scleroderma may be very difficult, particularly in its early stages. Many of its symptoms are common to, or may overlap with, those of other diseases, especially other autoimmune connective tissue diseases such as rheumatoid arthritis and systemic lupus erythematosus (SLE or lupus). Different symptoms may develop in stages over a very long period of time and few people with scleroderma experience exactly the same set of symptoms and effects.

While scleroderma can often be suspected from its more visible symptoms, no single test can prove its presence. The diagnosis is usually made by your doctor through a combination of the following: the medical history, including past and present symptoms; a thorough physical examination; and findings from a variety of laboratory tests including capillaroscopy, blood tests and other investigations. In making the diagnosis, it is important not only to confirm the presence of scleroderma, but also to determine its extent and severity, particularly with regard to the involvement of internal organs.

While scleroderma can often be suspected from its more visible symptoms, no single test can prove its presence

VEDOSS the Very Early Diagnoses clinics for Systemic Sclerosis is a new initiative from EUSTAR (The EULAR Scleroderma Trials and Research Group) to detect early systemic scleroderma throughout Europe. The launch of VEDOSS clinics will be on the 29th June 2009, which coincides with the first ever European scleroderma awareness day. The basic idea is that once a person is diagnosed with Raynaud's and puffy fingers they will undergo capillaroscopy and anti-nuclear antibody tests. If tests are found to be positive then appropriate education and a referral to a connective tissue clinic will follow.

Anti-nuclear antibody and extractable nuclear antigen testing

Specific blood tests can be very useful in determining the pattern of disease and the potential risk of specific internal organ problems. These are immunological tests for anti-nuclear antibodies (ANA) and extractable nuclear antigens (ENA). These are markers of autoimmune disease and the different types are associated with different patterns of scleroderma:

Anti-topoisomerase-1 (Scl 70) antibody – strongly associated with lung fibrosis in both subsets of systemic sclerosis and with renal disease;

Anti-centromere antibody (ACA) – seen almost only in patients with limited scleroderma and is associated with increased risk of pulmonary hypertension (high blood pressure in the lung arteries), but infers relative protection from lung fibrosis and kidney involvement;

Anti-RNA polymerase I and III antibody - associated with diffuse scleroderma and especially with kidney involvement;

Anti-fibrillarin (U3RNP) antibody - associated with heart involvement, pulmonary hypertension, kidney involvement and myositis (inflammation of the muscles);

Anti-PM-Scl antibody – strongly associated with the combination of myositis and scleroderma;

Anti-U1RNP (nRNP) antibody - associated with joint involvement and overlap syndromes.

Nailfold capillaroscopy

The systemic forms of scleroderma commonly cause changes to the size and number of capillary blood vessels in the skin. These changes can sometimes even precede the symptoms of a connective tissue disease, so checking the capillaries of patients with symptoms such as Raynaud's phenomenon can help doctors ascertain if there is any risk of scleroderma developing over time. Capillaroscopy uses a microscope to view the capillaries in the area of skin at the base of the fingernail. Figure 1 shows how, in scleroderma (bottom panel), these capillaries can become enlarged and disorganised compared to normal (top panel).

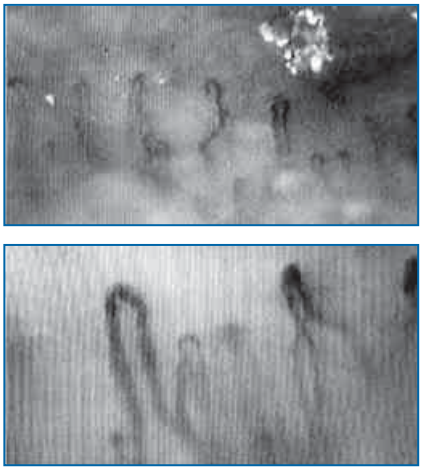


Figure 1

Treatment for localised scleroderma

Plaque morphoea does not always need treatment, as it may get better by itself. Strong steroid creams or ointments are sometimes used as they may help to reduce inflammation and prevent progression. They are applied thinly once a day and can be used safely for many weeks. They may also be used under dressings or be injected into the edges of plaques. Other creams including a group of drugs called the Vitamin D analogues have been described to have some effect on plaque morphoea. Immunosuppressive ointments including tacrolimus are also sometimes recommended.

To suppress immune activity within the skin affected by morphoea, and in rare or severe cases, oral or intravenous steroids may be given as treatment. Other treatments such as hydroxychloroquine, cyclosporin, methotrexate or mycophenolate mofetil have also been described as useful in the treatment of morphoea. Light therapy such as PUVA (using a **p**soralen medicine followed by **u**ltra**v**iolet **A** light) may also be used as a treatment in generalised morphoea. UVA1 is another form of light therapy but it is not widely available and its value in the treatment of morphoea is still not established.

If any contractures of joints occur, surgery and long-term physiotherapy can be of use. Plastic surgery may be a possible form of treatment for deeper forms of morphoea, in particular en coup de sabre.

At all times, the side effects of treatment versus *the effects of the* condition itself must be weighed up by the patient and his or her doctor. If the disease is mild or not progressing then it may be appropriate not to apply or take specific therapy. In extensive morphoea or where there is interference with growth, as can occur in childhood, then treatment should be much more extensive.

In the UK, several centres are investigating new measurement techniques for morphoea that show promise for assessing disease activity. Apart from regular clinical review by a doctor experienced in care of patients with morphoea (usually a dermatologist or rheumatologist) there is no standard way of monitoring the progression of morphoea or its response to treatment.

What are the symptoms of systemic scleroderma and how are they treated?

This section describes the most common symptoms of scleroderma and some of the treatments being used to control them.

Scleroderma is a complex disease that can affect many parts of the body with many possible symptoms. Most people only develop a few of the symptoms mentioned. Each patient is different in terms of symptoms and severity. Typically, the symptoms may also vary over time with periods of improvement and worsening. It is not possible in a booklet of this length to describe all of the symptoms or all of the methods being used in the management of scleroderma. A great variety of treatments and medications have been tried over the years and new ones are constantly being researched and tested. Doctors experienced in scleroderma should be consulted regarding any symptoms or treatments mentioned here, as well as for any other symptoms that may be experienced.

Scleroderma is a complex disease that can affect many parts of the body with many possible symptoms

Even though scleroderma is not curable, many of the symptoms can be improved with medication or lifestyle changes.

Raynaud's phenomenon

Raynaud's phenomenon is the most common early symptom of systemic scleroderma. It is present at one time or another in about 90% of patients. It is most obvious in the fingers and toes but can also involve the ears, nose, and tip of the tongue. In Raynaud's phenomenon, the blood vessels constrict or narrow in response to cold or to emotional upset and stress. The resulting disturbance in circulation of the blood causes a series of colour changes in the skin: white, blanched, or pale when circulation is reduced; blue as the affected part loses oxygen from decreased blood flow; and then red or flushed as blood flow returns and the affected skin re-warms. Finally, as the attack subsides and the circulation returns to normal, usual skin colour is restored. In the "white" or "blue" stages, sensations such as tingling, numbness and coldness may be felt. In the "red" stage, a feeling of warmth, burning or throbbing may be noted. In some people, the Raynaud's attacks are painful.

Although Raynaud's phenomenon is diagnosed by a doctor on the basis of symptoms, there are a number of hospital tests that can be used to assess the severity of Raynaud's and help with decisions about appropriate care and treatment. Perhaps the most widely used test is thermography with cold challenge. This involves cooling the hands in water and measuring their rewarming with an infrared thermal camera. In Raynaud's phenomenon, rewarming is greatly delayed in comparison to healthy subjects.

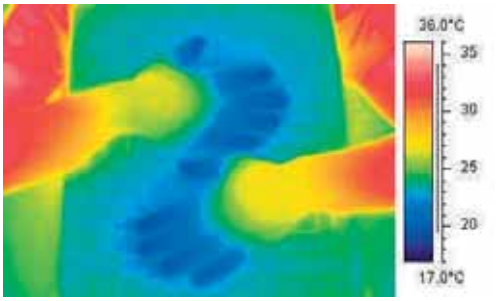


Figure 2

Figure 2 shows a thermal image of the hands of a Raynaud's patient after cooling in water: the blue areas show the coldest parts of the hand.

Many common-sense **preventative measures** can be taken by those susceptible to Raynaud's phenomenon. The most obvious is minimizing exposure to cold, such as outdoor weather, air conditioning, or reaching into a refrigerator or freezer. Keeping warm is very important. It is important to not only protect your hands and feet from cold but to also keep the whole body warm. Gloves or mittens should be worn, and a number of warming devices are available to protect the hands. Hat, ear muffs, heavy socks, and warm layered clothing of fibres such as silk, cotton, wool and down are effective in maintaining body temperature. It is important to protect the hands with gloves when touching refrigerated

or frozen items. Electric heaters and electric blankets can supplement the heat in the home. Keeping the entire body warm helps prevent Raynaud's episodes.

A warm bath or shower, or a heating pad or hot water bottle on the back, may relieve an attack better than just warming the hands. Avoidance of emotional upset and stress may help but is not always possible. Relaxation techniques of various kinds, whether self-taught or learned through training courses, have proven effective for some people in managing stress. One particular technique, biofeedback, has been used to increase finger temperature.

Many common-sense preventative measures can be taken by those susceptible to Raynaud's phenomenon

Smoking definitely worsens Raynaud's phenomenon. For this, amongst other reasons, people with scleroderma should not smoke.

When Raynaud's phenomenon does occur, carefully waving the arms in a circular motion can help to restore blood circulation. Rubbing or massaging the hands and feet may also help. Hands should not be placed on hot radiators or in hot water as the numbness caused by an attack may mean that it is not possible to accurately assess the temperature and burns may occur.

Your doctor may suggest a number of different medications to prevent, reduce the frequency, or minimize the effects of Raynaud's phenomenon. Most of these drugs dilate or open up the blood vessels. The most commonly used and best-tolerated medications are the calcium channel blockers, e.g. nifedipine and diltiazem, as well as the angiotensin II receptor antagonists such as losartan. There have been some reports of Fluoxetine, an antidepressant, being used to treat Raynaud's with good effect. These medications can cause side effects such as palpitations, facial flushing, headaches, light-headedness, swelling of ankles and constipation. There are many medications to improve blood flow not listed here, however through trial and error most people with Raynaud's will manage to find a treatment which helps them.

In severe cases, particularly when there are digital ulcers or infection, treatment with a synthetic prostacyclin analogue (e.g. iloprost) may be recommended. It is given via an intravenous drip over three or five days but can be given continuously over a longer period of time in severe and complicated cases. This treatment can produce an improvement in Raynaud's symptoms for three or more months and has been shown to help the healing of digital ulcers in some people. Iloprost works by opening up the blood vessels and improving blood supply to the fingers.

Raynaud's phenomenon is not confined to people with scleroderma. It is also seen in lupus, myositis, Sjogren's syndrome and other connective tissue diseases. In addition, many healthy people have Raynaud's phenomenon without any other illness. In this situation, it is called "primary Raynaud's phenomenon." People who have Raynaud's due to scleroderma or another condition are described as having "secondary Raynaud's phenomenon".

Swelling or puffiness of the hands

Swelling is another typical early symptom of scleroderma, and this may be especially noticeable upon awakening because of muscle inactivity overnight. The skin of the fingers may look full and sausage-like, making it difficult to close the hand into a fist. Exercising the fingers and toes is helpful. Your doctor may recommend medications to reduce inflammation.

Pain and stiffness of the joints

Symptoms of pain, stiffness, swelling, warmth or tenderness may accompany the arthritis-like joint inflammation that frequently occurs in scleroderma. Muscle pain and weakness are other important symptoms. Anti-inflammatory prescription drugs can be helpful in reducing pain in this situation. Because of their side effects however, they are not suitable for all patients

Other treatments and suggestions are included in the sections on "Physical Therapy and Exercise" and "Protecting the Joints." A physiotherapist can develop an exercise plan after consultation with your doctor.

Skin problems

Skin thickening

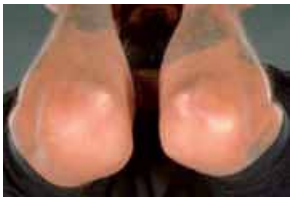
Hardening and thickening of the skin give scleroderma its name ("hard skin"). There are no proven treatments as yet to universally prevent or alter the course of the skin changes in scleroderma. In patients with rapidly progressing diffuse scleroderma, doctors may recommend a trial of an immunosuppressant such as Mycophenolate or Cyclophosphamide. Some patients with diffuse disease experience softening of their skin after several years even without treatment.

Skin ulceration

Sores, especially on the fingertips, are a common symptom of systemic scleroderma and are often called ulcers. They may be very slow or difficult to heal because of poor

circulation. These ulcers may also occur on the knuckles, elbows, toes or other sites of the body where the skin is especially tight or stretched. The affected area should be kept warm to increase blood flow, and scrupulously clean to avoid infection. Hirudoid cream can be bought without prescription and can help if small amounts are applied twice a day around any ulcerated areas. Should these remedies prove unsuccessful in relieving the pain or infection of ulcerated skin, your doctor may prescribe oral antibiotics or take other measures, including in severe cases a hospital admission for treatment with iloprost. It is very important to consult your doctor early if you suspect an ulcer is infected as early treatment can prevent further damage. A clean, dry dressing should be used to protect the ulcer but no dressing has yet been shown to positively aid healing.

Calcinosis



This condition is characterized by deposits of calcium in the skin which may be painful. The calcium deposits may occur just below the skin surface in the form of hard lumps or nodules. They may break through the skin, becoming visible as chalky white material, and they may become infected. Care should be taken not to bump or injure affected areas. Hand-waxing may be helpful. Antibiotics may be prescribed to prevent or control infection. In severe cases, minor surgery to remove calcium deposits may be required.

Telangiectasia

This abnormality consists of the dilation of small blood vessels near the surface of the skin, which become visible as small red spots, usually on the fingers, palms, face and lips. The spots usually fade with pressure, but turn red again when the pressure is released. These spots are generally not harmful. Special cosmetics may be used to mask the spots or to reduce their visibility. In severe cases, or where a person is very affected by the telangiectasia, laser therapy can help to remove the red spots. Treatment, however, can be slightly uncomfortable and may not be permanent.

Dry skin

Excessive dryness of the skin may lead to skin breakdown and ulcerations. Excessive bathing and hand washing should be avoided, and rubber gloves worn to avoid direct contact with household detergents. Keeping the skin moist and well-lubricated is important to avoid complications from dry skin. Bath oils and moisturizing soaps such as Neutrogena®

and Dove™ are preferable to harsh soaps which dry out the skin. Frequent use of moisturising creams is recommended. The list is long however, and it's a matter of finding which product suits you. For further information regarding the skin in scleroderma, please see our leaflet, "The Skin in Scleroderma".

Itchy skin

If moisturising creams do not work, your doctor may prescribe a topical cortisone cream to be rubbed on the skin to relieve itching (although this should not be used for a prolonged period of time). Antihistamine tablets have been effective for some people, particularly at night.

Other skin symptoms

There may be a decrease in hair over affected areas of the skin, as well as a decrease in the ability to perspire. In addition, there may be an increase in pigment (which looks like a skin tan) or a spotty loss of pigment.

Sclerodactyly and joint contractures

Sclerodactyly means simply "hard skin of the digits"; that is, of the fingers and toes. It is characterized by shiny, tight skin of the fingers. Affected digits may be difficult to move, and they may become fixed in a bent (flexed) position called a "flexion contracture." Tightening and hardening of the skin and of the tissues surrounding the joints can cause decreased movement of the wrists, elbows and other joints.

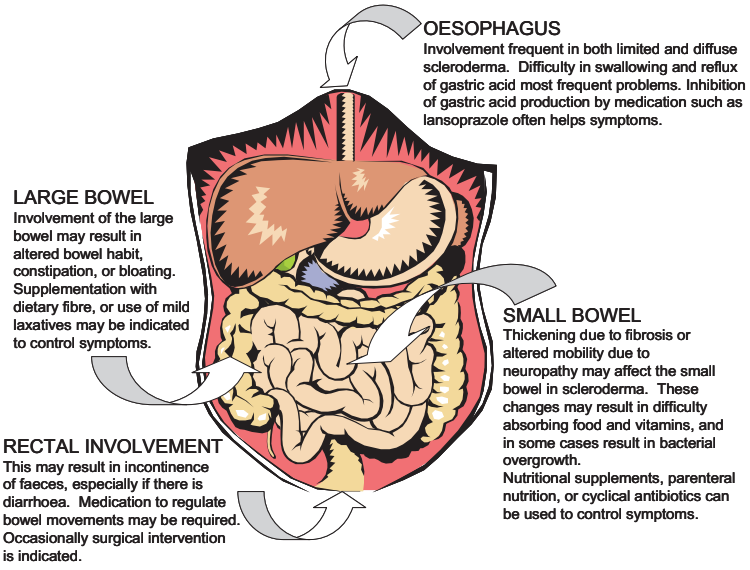
"Range of motion" exercises performed daily are important in preventing or slowing down the development of such contractures and in keeping the joints flexible. They may also help to increase blood supply to the tissues. These exercises are simple to perform and can be done at home. A typical exercise consists of laying the hand as flat as possible on a table, placing the heel of the other hand across the fingers and gently pressing down to straighten the fingers. An occupational therapist can develop an exercise plan after consulting with your doctor. He/she may also provide devices to help perform common personal care and household tasks more easily.

Digestive system and gastrointestinal tract problems

People with systemic scleroderma may develop abnormalities of the digestive system and gastrointestinal tract right from the mouth down to the anal canal. The overproduction of collagen typical of scleroderma can cause thickening and fibrosis (or scarring) of the tissues. This can result in weakened muscles and lead to the abnormally slow movement of food ("dysmotility") in the digestive process.

Oesophageal dysfunction

Food travels from the mouth and throat into the stomach through the oesophagus. Normally, the lower oesophageal sphincter or valve acts as a gate which opens to permit food to enter the stomach and then closes promptly to prevent food from coming back up. In systemic scleroderma, the gate does not close properly and the result is a backwash of acid and a burning sensation ("heartburn") as food and acid return into the oesophagus. The acid may also injure the lining of the lower portion of the oesophagus, causing scarring and a narrowing ("stricture") of the tube. Acid production can be reduced, and the problems of acid reflux and heartburn lessened, by avoiding (as much as possible) alcohol,



greasy or fatty foods, spicy foods, chocolate, tobacco and caffeine. Many people, however, may require a medication to decrease the acidity of the stomach so that any fluid washing back into the oesophagus will not be harmful to it.

There are several drugs that decrease the acidity of the stomach. The most commonly used drugs nowadays are known as proton pump inhibitors. These include lansoprazole, omeprazole and esomeprazole. The H2 receptor antagonists (ranitidine) can also decrease acid production and are sometimes used. In some people with severe reflux, a combination of the two different classes of drugs is used. Your doctor may also occasionally prescribe a drug such as metoclopramide or domperidone to try to stimulate muscular activity in the stomach and oesophagus.

The force of gravity helps to keep food and acid in the stomach; an upright position after meals is therefore helpful. Other common-sense measures to prevent acid from coming up into the oesophagus include eating smaller and more frequent meals, not eating for several hours before bedtime, and elevating the head of the bed six to eight inches with wooden blocks. Being overweight can make reflux worse and wearing girdles or other tight-fitting garments should be avoided.

Abnormally slow movement of food and narrowing of the oesophagus may cause swallowing difficulties. Eating slowly and chewing thoroughly are important. Swallowing and digesting are made easier by eating softer foods (many foods can be prepared in a blender) and avoiding foods which tend to stick in the throat. The medication metoclopramide and domperidone, as described above, may help. If the oesophagus has narrowed significantly, you may need to have the oesophagus dilated periodically to permit easier swallowing.

Small bowel involvement

In systemic scleroderma, there can be damage to the muscles of the small bowel (small intestine). The weakened muscles do not work effectively in pushing food through the bowel. Simply put, things "sit" rather than move well. One consequence can be an overgrowth of natural bacteria, leading to diarrhoea. There may also be a bloated, distended feeling and some pain if the bowel is stretched. Another effect is that the nutrients of food remain in the bowel instead of being absorbed into the body. This condition is called "malabsorption," and it may lead to weight loss and stool abnormalities.

For diarrhoea or malabsorption, your doctor may prescribe an antibiotic, and in some cases supplementary vitamins and iron may be necessary. Your doctor may also suggest that the amount of fatty foods in the diet be reduced and the amount of carbohydrates increased. In some cases your doctor may recommend that you see a dietician.

Large bowel involvement

Weak or scarred muscles in the colon wall make it difficult for the bowel to work well, resulting in constipation or other abnormalities of the colon. Maintaining a diet high in fibre, and drinking at least six to eight glasses of fluids daily, especially water, will help reduce constipation. Fresh fruits and vegetables are natural laxatives. Exercise also helps to keep bowel movements regular. Your doctor may also recommend stool softeners and bulking agents like lactulose or Fybogel.

Rectum and anus

Involvement of the last portion of the large bowel can lead to faecal incontinence. This may be aggravated when patients have diarrhoea. There are tablets which sometimes may be helpful by slowing down the bowel movement, and in more severe cases, surgical intervention may be required.

Sjögren's Syndrome

Some people with scleroderma may have features of other connective tissue disorders (overlap syndromes). One such condition is Sjögren's Syndrome. It can exist on its own (primary) or can appear in association with other connective tissue diseases, including scleroderma (secondary).

The mouth should be kept as well-lubricated as possible by sipping fluids throughout the day

The most common features of Sjögren's Syndrome seen in people with scleroderma/Sjögren's Syndrome overlap are dryness of the eyes and mouth (sicca symptoms). These are caused by decreased secretions of the tear and the salivary glands, which provide lubrication for the eyes and mouth. The unusual dryness of the eyes resulting from this condition can lead to serious irritation and inflammation. Excessive dryness of the mouth may lead to difficulties in swallowing and in speaking, a pronounced increase in tooth decay and cavities, and a reduced sense of taste. The lack of secretions may also involve the vagina and other areas of the body.

Dry eyes may be lubricated by the frequent use of artificial tears and ophthalmic ointments, some of which are available without prescription. Regular visits to the ophthalmologist are important. The mouth should be kept as well-lubricated as possible by sipping fluids throughout the day (a plastic squirt bottle filled with water may be useful), and by chewing sugar-free gum or sucking sugar-free sour sweets to stimulate salivary activity. Artificial saliva is also available. (See next section for preventative dental care.)

Lubricants such as KY gel can help to moisten the vagina and facilitate sexual relations. Avoiding tights and other tight-fitting clothing may help to reduce irritation and prevent infection. Choose cotton rather than nylon underwear.

Facial changes

People with systemic scleroderma often develop specific facial changes related to their disease. The nose can appear pinched and thinned and the opening of the mouth may be decreased in size ("microstomia" or "small mouth"). Often the skin over the upper lip can

become lined, forming ridges called rugae. Development of dilated small blood vessels, called telangiectasia, can affect both the face and other parts of the body. The appearance of the eyes can also alter.

Oral and dental problems

The general tightening of skin over the face makes lip and mouth movements, as well as oral hygiene, difficult. Microstomia may make it difficult to open the mouth wide enough for dental procedures therefore preventative dental care through regular flossing and brushing of the teeth and gums after each meal is very important.

The best approach to management is by means of facial grimacing and mouth stretching exercises daily. Regular dental visits are also important to help prevent dental caries. Your dentist can also recommend a programme of good oral hygiene. Floss holders, pump toothpaste tubes, and built-up handles on toothbrushes can help people with hand impairment and an electric toothbrush is strongly recommended.

Kidney involvement

Kidney or renal involvement in systemic scleroderma may be mild or very serious in nature. Early signs of kidney involvement may include mild hypertension (high blood pressure), protein in the urine and blood test abnormalities. “Renal crisis,” (kidney crisis) a highly dangerous complication of systemic scleroderma, may occur quite suddenly. Its most important warning signal is an abrupt rise in blood pressure. Renal crisis occurs in about 10% of people with diffuse scleroderma and is most common in the first four years of their disease. It is less common to see renal crisis in people with limited scleroderma.

People may recover successfully from renal crisis, but only if the problem is recognised and treated quickly

Symptoms include new severe headache, visual disturbances, shortness of breath, chest pain or discomfort, or mental confusion. Unless treated promptly, renal crisis leads to kidney failure, a condition in which the kidneys lose their ability to eliminate waste products from the body. The treatment of choice is ACE inhibitors, which are a group of anti-hypertensive drugs. These medications are quite effective in controlling blood pressure and in stabilizing and even improving kidney function. In cases of severe kidney failure, dialysis may be required. People with systemic scleroderma are advised to have their blood pressure and kidney function monitored at regular intervals. People with systemic scleroderma considered to be at risk of a scleroderma renal crisis are often issued with “renal cards”. They contain information for both patients and physicians about what

to do in an emergency situation. People may recover successfully from renal crisis, but only if the problem is recognised and treated quickly.

Lung involvement

The lung can be affected in scleroderma by three different processes:

1. Build-up of collagen thickens lung tissue and causes fibrosis or scarring, making the transport of oxygen into the bloodstream more difficult. This is called pulmonary (lung) fibrosis or Interstitial Lung Disease (ILD);
2. Pulmonary Arterial Hypertension (PAH), a state of increased blood pressure in the lung arteries, can result from damage to blood vessels, and leads to increased strain on the heart, resulting in heart failure;
3. Respiratory muscle weakness may decrease lung function.

Symptoms of lung involvement include shortness of breath, a decreased tolerance for exercise and a persistent cough. The physician may order a chest x-ray and/or CT scan of the chest, an echocardiogram (ultrasound of the heart), or special breathing tests (pulmonary function tests) to detect or confirm lung involvement and to determine which of these three processes is responsible for the symptoms.

In the very early stages of lung fibrosis, medications may be given to decrease the inflammation which is thought to lead to lung scarring. Recent trials have shown that patients with severe and progressive lung fibrosis may gain some benefit from immunosuppressive drugs such as Cyclophosphamide.

It is important for a person with scleroderma to take whatever measures are within his or her control to avoid further damage to the lungs. It is essential to avoid smoking, a major cause of lung disease. Exposure to air pollutants may worsen breathing problems and should be avoided as much as possible.

It is essential to avoid smoking, a major cause of lung disease

Pulmonary Arterial Hypertension (PAH) can affect approx 10 - 15% of people with scleroderma. PAH can now be treated with special medications targeted at dilating or opening up the blood vessels of the lungs and changing the underlying disease process. This is one complication of scleroderma for which new medications have proven successful. In the UK there are currently several medications licensed for the treatment of PAH. New treatments are under development and may become available in the next few years. Bosentan (brand name Tracleer®) and Sitaxsentan (Thelin®) are endothelin receptor

antagonists, taken in tablet form. Sildenafil (Revatio®) is a strong vasodilator administered orally three times a day. Iloprost (brand name Ventavis) is a prostacylin derivative that is inhaled via a special hand held nebuliser approximately every three hours during the daytime. Epoprostinol (Flolan®) is also a prostacyclin derivative that is given continuously through a Hickman® line.

All these drugs have been shown in clinical trials to improve exercise capacity of people with severe PAH. These medications can only be prescribed by specialists with experience in managing this condition.

Heart involvement

This is an important and potentially life-threatening complication of systemic scleroderma. The heart muscle can become inflamed (myocarditis) and can develop scar tissue. This may ultimately result in heart failure. Inflammation of the outer heart lining (pericardium) is called pericarditis and may cause pain and accumulation of fluid around the heart. An irregular heartbeat may also be experienced. These conditions require careful evaluation and treatment by your specialist.

Non-specific symptoms

The person with systemic scleroderma may experience a variety of non-specific symptoms, including fatigue (ranging from mild to severe), lack of energy, generalized weakness, weight loss, and vague aching of muscles, joints, or bones. Treatments or medications recommended by your doctor will depend on his/her evaluation of the causes of these symptoms.

Managing scleroderma

The reader may be aware of, or learn about, other forms of treatment that have been used or are proposed for use in managing scleroderma in addition to those discussed in this booklet. Scleroderma is a difficult disease to study because of its variable nature, its prolonged course, and the relatively small number of people affected by it. Under these circumstances, it is difficult to conduct scientifically sound studies proving the value of a particular drug or treatment. Your doctor must therefore often make decisions about treatment based on incomplete information. He/she must

weigh the possible benefits against the potential risks or side effects. Further investigation will ultimately determine which treatments are beneficial and which treatments are not.

The course of scleroderma

Scleroderma has many forms and a number of different symptoms that may present themselves singly or in combinations at various times throughout the course of the disease. Some symptoms develop with relative suddenness; others take years to develop. The exact course the disease may take is unpredictable, and the prognosis will vary from individual to individual. Systemic scleroderma is a chronic, life-long disease. At present there is no known cure, but as with other chronic diseases there are many means available to control or manage its symptoms. It is helpful to keep scleroderma in perspective. Many persons with the disease have few or minimal symptoms and are able to lead a normal or nearly normal life. There may be periods of time when the person with scleroderma will be free of troubling symptoms and feel well. At other times, he or she may feel quite ill. Spontaneous improvements may occur. The skin, in particular, sometimes softens and becomes more pliable after a number of years. Spontaneous remissions, times when symptoms may actually disappear, may also occur and may continue for long periods. The person with scleroderma should be cautious about attributing such improvements or remissions to a particular treatment, diet, or so-called “cure.”

Being alert to symptoms

This booklet describes many symptoms, although each person with scleroderma usually develops only a few of them. Its purpose is not to overwhelm people with scleroderma or those who suspect they may have the disease but to provide them with useful information on what to look for, what may occur during the course of the disease and some of the things that can be done if symptoms do develop.

This booklet describes many symptoms although each person with scleroderma usually develops only a few of them

Learning to recognize early symptoms of disease activity can lead to earlier detection and diagnosis of scleroderma and to prompt initiation of treatment. Some of the more promising medications in current use are slow-acting and the sooner treatment is begun, the better the results may be. If one has already been diagnosed as having scleroderma, it is especially important to watch for and report to the

doctor new or changed symptoms. Early treatment may prevent symptoms from worsening and may decrease the chance of permanent tissue or organ damage.

In being alert to symptoms, it would be a mistake to assume that every symptom or condition that develops is necessarily related to, or the result of scleroderma. People with or without scleroderma do suffer accidents, contract infectious diseases and develop other illnesses. Your doctor or specialist nurse can help to distinguish what is related to scleroderma and what is not, and recommend appropriate action and treatment.

Developing an individual treatment programme

While there is no proven cure for scleroderma, much can be done to prevent, minimize, or alleviate its effects and symptoms. The symptoms of scleroderma vary greatly from individual to individual; the manner in which each person responds to treatment also varies greatly and there are many treatment options. It is important, therefore, that a doctor experienced in the management of scleroderma work out an individually-tailored treatment programme to meet the specific needs of a person with this disease. Close cooperation with your doctor will help him or her develop such a programme.

Many forms of treatment have been discussed already in the chapter entitled “What are the symptoms of scleroderma, and how are they treated?” The next six sections will discuss other important elements of a programme for managing scleroderma.

Physical therapy and exercise

Physiotherapists can help the person with scleroderma develop an appropriate programme. Such a programme may consist of “range of motion” exercises (as mentioned in the previous chapter under the section “Sclerodactyly and joint contractures”), paraffin wax baths, hydrotherapy or water therapy, strengthening exercises for muscle weakness and gentle massage. These treatments can be carried out at various locations, including a hospital physiotherapy department and in the home.

Your doctor may recommend an exercise programme involving activities such as stretching, walking or swimming. People with scleroderma may find that their tolerance for activity and movement is below normal, so activities should be carried out in moderation, resting when tired. Individual exercises should be performed gently and with due care, and the exercise programme should be built up gradually.

Protecting the joints

The goals of joint protection are to minimize further damage and to reduce the possibility of skin ulcers and infection. Its basic principles include avoiding or minimising pressure or stress on the joints by ensuring their proper use and maintaining their mobility and function by stretching and performing a "range of motion" exercises. A variety of self-help aids and adaptive mechanical devices are available to help protect and to alleviate stress on the joints while carrying on the activities of daily living. Occupational therapists can demonstrate such devices and give further instruction on joint protection.

Taking medications

It is essential that the person with scleroderma take all medicines wisely; take only those prescribed; read warnings labels on medications and follow instructions carefully; and take the medications exactly when, for how long, and in the dosages prescribed by your doctor. The person with scleroderma should advise their doctor of any drugs being taken for other conditions including over-the-counter preparations, herbal supplements, or vitamins. Any side effects encountered should be promptly reported and discussed.

You should not be concerned if your doctor prescribes different medications for different people. Scleroderma symptoms vary from person to person, requiring different treatment. Some may benefit from certain drugs, while others may not. Furthermore, individual tolerance for the drugs used in scleroderma varies greatly. Your doctor may find it necessary to adjust the drug dosages accordingly.

Common-sense measures

Treatment of specific symptoms has been discussed in other sections of this booklet. There are also a number of general common-sense measures that the person with scleroderma can take to enhance his/her well-being. These measures include:

- Avoiding over-fatigue by "taking it easy" and getting sufficient rest; knowing your own limits does not indicate that you are "lazy";
- Learning to control and minimise stress;
- Eating well-balanced meals and maintaining a sensible weight;
- Practising habits of good hygiene, especially of the skin, teeth, gums, and feet (including the wearing of cushioned and well-fitted shoes).

One key measure, mentioned previously, is avoiding smoking. The health risks of smoking are well known but frequently ignored. It is particularly dangerous to persons with scleroderma because it can have effects on blood circulation and lung function.

The emotional aspects of scleroderma

A common reaction to being told that one has a disease such as scleroderma is, “why me?” It is not known why some people develop the disease and others do not. One does not bring scleroderma upon himself or herself; therefore, one need not feel guilty or responsible for the illness.

A person newly diagnosed with scleroderma may feel alone and uncertain about where to turn for help. He or she may experience a number of other feelings and emotional reactions from time to time, including initial shock or disbelief, fear, anger, denial, self-blame, or guilt, grief, sadness or depression. Family members may have similar feelings.

Feelings in themselves are neither good nor bad; one simply “has” them. Sharing them with family and friends or with others who have had similar experiences can be helpful. Professional counselling can also help people with scleroderma and their family members who are having difficulty coping with their feelings.

Thinking of oneself as a total person with a full life to lead may help to keep scleroderma in perspective

The term “person with scleroderma” has been used throughout this booklet instead of “scleroderma patient.” The person with scleroderma may be a “patient” in the doctor’s office, hospital or clinic, but he or she is much more than that. Thinking of oneself as a total person with a full life to lead may help to keep scleroderma in perspective and enable one to maintain a positive but realistic attitude.

Building a health and support network

Participating actively in one’s own healthcare is of prime importance to the person with scleroderma. It is equally important to cooperate and communicate effectively with the doctor who is managing the disease. While these two,—the person with scleroderma and the doctor, are the focal point of the management “team”, many other people and resources can be enlisted to form a health and support network.

Family and friends can provide emotional support for the person with scleroderma, encourage him/her to follow the recommended treatment programme and assist in carrying out activities that he/she finds difficult.

The health team begins with your doctor, but can include many other health professionals such as other medical specialists, nurses, physiotherapists, occupational therapists and psychologists or others trained in counselling.

Directories of community resources typically list a large number of voluntary and governmental agencies providing health, social and rehabilitation services that may be of benefit to the person with scleroderma. These can be accessed via the Internet or from your local library. Joining a scleroderma support group, such as The Scleroderma Society enables the person with scleroderma to meet and to exchange information with others who have similar problems, as well as to learn more about scleroderma. The extent of the health and support network is limited only by the imagination and resourcefulness of those helping to create it.

The extent of the health and support network is limited only by the imagination and resourcefulness of those helping to create it

Progress through research

Is there hope and help for the person with scleroderma?

Emphatically, yes!

As this booklet has discussed, there are many treatments and medications available now to help the person with scleroderma and more and more doctors are becoming interested in the disease.

Researchers throughout the world are intensifying their efforts to understand the nature and discover the cause of scleroderma, to find better means of prevention and treatment and to find a cure. These efforts reflect the increased interest in all of the connective tissue and rheumatic diseases.

Research has already resulted in better laboratory tools to detect the early stages of scleroderma and in improved methods of measurement to evaluate disease progression and the results of treatment. Various animal models of scleroderma have been developed.

Investigators are currently studying the role of the immune system in scleroderma, exploring the relationship between blood vessel changes and fibrosis, and seeking "markers" to identify the various forms and subsets of scleroderma. These are just a few of the many studies in progress.

Scleroderma poses many questions. Answers may come from a variety of medical and scientific fields or from totally unexpected sources… but they will come!

The Scleroderma Society

The Scleroderma Society is a UK-wide Patient Support Group that was formed in 1982. It is a UK registered charity number 286736 and is open to anyone with an interest in scleroderma. The society is managed by a board of four volunteer trustees, assisted by a lay committee of volunteer members. A panel of doctors and scientists provide the trustees with expert advice and guidance.

The Scleroderma Society's mission is education, awareness and research.

The Society's aims are:

- To support anyone who is affected by scleroderma including patients, family members and friends;
- To encourage and facilitate communication between scleroderma patients;
- To create and improve awareness of scleroderma amongst the general public and health professionals at all levels;
- To help patients towards managing their own condition by providing information about scleroderma and progress being made by scientists and doctors;
- To fund medical & scientific research.

The society achieves its aims by providing a range of services to members which include:

- Freephone telephone Advice Line (0800 311 2756) manned by volunteers;
- Website with news and information, www.sclerodermasociety.co.uk , including links to other associations throughout the world and a very popular message board;
- Quarterly Newsletter "Scleroderma News" sent free of charge to all members and other interested health professionals;
- Facilitating member to member contact;
- Promoting and supporting scleroderma research;
- Providing educational literature for people with scleroderma and members of the medical community;
- Organising local groups and meetings;
- Annual Conference with high profile speakers including patients, doctors and scientists.

Contact Information

Write to: The Scleroderma Society
PO Box 581
Chichester
PO19 9EW

Telephone: 020 7000 1925
Advice Line: 0800 311 2756
Web: www.sclerodermasociety.co.uk
email: info@sclerodermasociety.co.uk

Glossary

Here are some useful definitions of medical words and terms:

Acid reflux, heartburn: Stomach acid which abnormally travels up into and irritates the oesophagus. (Acid production is a normal part of digestion in the stomach) Heartburn refers to pain in the centre of the chest caused by acid reflux. (See Oesophagitis)

Analgesic: A medication which reduces or eliminates pain. Example: paracetamol or non-steroidal anti-inflammatory drugs

Antibiotic: Medication used to treat an infection. Each antibiotic kills or inhibits the growth of specific microorganisms, so antibiotics are prescribed based on the type of infection present

Arthralgia: Pain in a joint

Auto-immune: Disease or antibody which acts against the patient's own tissues. (See Immune system)

Biofeedback: A technique used to regulate a body function usually involuntarily controlled, such as a finger temperature or pulse rate. By observing a machine monitoring the function, a person can practise relaxation techniques and learn to control the function. Later, the machine becomes unnecessary (See Relaxation techniques)

Biopsy: The removal and examination of tissue, cells or fluid from the body

Blanched: To become white or pale. In Raynaud's phenomenon, the fingers and toes blanch due to insufficient circulation of blood

Calcinosis: Abnormal accumulation of calcium in the skin

Capillaries: The smallest blood vessels of the body, connecting arteries and veins

Collagen: A normal, fibrous protein found in the connective tissue of the body

Connective tissue: Tissue which pervades, supports and binds together other tissues including mucous, fibrous, reticular, adipose, cartilage, skin and bone. Connective tissue diseases are a group of diseases with similar cellular changes, but with the site where the changes occur determining the specific disease. Included are scleroderma, systemic lupus erythematosus, dermatomyositis, and rheumatoid arthritis

Constrict (vessels), stricture (oesophagus): An abnormal narrowing

Contraction (of intestinal muscles): The rhythmic squeezing action of the muscles of the wall of the intestine which moves food through the system. Also called peristalsis (See Motility)

Coronary arteries: Blood vessels which supply blood to the heart itself

CREST: Form of scleroderma, whose initials stand for **C**alcinosis, **R**aynaud's phenomenon, o**E**sophageal dysmotility, **S**clerodactyly, and **T**elangiectasia. Not used any longer

Cutaneous: Of the skin.

Cyanosis: Blue or purple colour due to lack of blood oxygen. In Raynaud's phenomenon, cyanosis of the fingers and toes may follow blanching

Cytokines: Chemical signals in the blood

Digits: Fingers and toes

Dilate (oesophagus, blood vessels): To widen or enlarge

Diuretic: Medication to increase the flow of urine, thereby decreasing fluid retention in the tissues. Also called, "water tablets" (See Oedema)

Dysfunction: Impaired or abnormal functioning

Dysphagia: Difficulty in swallowing

En coup de sabre: A form of localised scleroderma which forms a long crease of waxy skin, resembling a cut by a sabre or sword wound usually on face or neck

Fatigue: Weariness, a sense of being overwhelmingly tired, or exhaustion

Fibrous: Consisting of, or resembling fibres

Fibrosis: Abnormal formation of excess fibrous tissue

Gastrointestinal tract, bowel, diarrhoea, constipation: The gastrointestinal tract is the digestive system which breaks down food, allows absorption of nutrients, removal of cellular waste products and elimination of solid waste from the body. It begins with the mouth and oesophagus and leads to the stomach. The small intestine consists of the duodenum, jejunum and ileum. Lastly, the large intestine (also called colon) leads to the rectum. The term bowel refers to the intestine. The anal sphincter is the muscle which controls discharge of stool. Diarrhoea is abnormally frequent or excessive passing of stool, usually watery. Constipation is the abnormally delayed or infrequent passage of stool, usually in a dry and hardened state. Normal bowel movements vary from person to person and with diet

Hypertension, anti-hypertensive: Abnormally high blood pressure. An anti-hypertensive medication lowers blood pressure

Immune System: The system of organs, cells and proteins which protect the body from foreign substances by producing immune responses. The immune system organs include the thymus, spleen, lymph nodes and bone marrow. The cells include white blood cells

(neutrophils, eosinophils, basophils, monocytes and lymphocytes (T and B cells)). Immunoglobulins (antibodies) are proteins that can react with and/or neutralize corresponding proteins called antigens (usually damaged or foreign material). The immune system is essentially protective and helpful to the body, but can be the cause of disease and allergy when it attacks parts of the normal body in a process called auto-immunity

Inflammation, anti-inflammatory: Tissue reaction to cell injury marked by redness, heat, pain, swelling and often loss of function. Capillary dilation and white blood cell infiltration help eliminate foreign substances and damaged tissue, so normally; inflammation is a natural part of the healing process. Excessive or inappropriate inflammation can, however, cause further damage. Anti-inflammatory drugs counteract inflammation

Joint contracture, flexion contracture: Fixation of a joint in one position preventing full range of motion. In scleroderma, this frequently affects the fingers due to tightening and hardening of the skin around the joint. In flexion contractures, the fingers become fixed in a bent (flexed) position

Lacrimal glands: Tear-producing glands, also spelled lachrymal

Laxative: A medication which stimulates emptying of the bowels

Lubrication, secretion: Substance which makes a surface slippery or oily, either artificially by applying lubricating fluids, or naturally by secreting fluids made by cells for this purpose. Example: tears

Malabsorption: The reduced ability of the bowel cells to take nutrients from the digestive tract

Microstomia: Abnormally small mouth opening

Mixed Connective Tissue Disease: Overlap or presence of symptoms of two or more diseases simultaneously (See Collagen and Connective tissue)

Morphoea: A form of localised scleroderma

Motility, dysmotility: Contractions of the digestive-tract muscles occurring in rhythmic waves, propelling food, allowing absorption of nutrients and elimination of wastes (faeces). Dysmotility indicates weakened or absent waves of contraction resulting in abnormally slow movement of food and faeces (See Malabsorption, Gastrointestinal tract, Contraction)

Occupational Therapy (Abbreviated "OT", See also Physical Therapy): Therapy using activity prescribed to promote recovery or rehabilitation. OT is often designed to increase the ability to perform acts of daily living, such as grooming, eating and concentrating on the hands and small muscle control

Oedema: An abnormal excess accumulation of fluid in tissues of the body

Oesophagus, oesophagitis: The muscular swallowing tube connecting the mouth and the stomach. When properly functioning, it contracts in smooth waves to send food to the stomach. At its lower end a sphincter (ring-like muscle) opens to allow food to pass into the stomach, but closes again to prevent stomach acid or partially digested food from backing up into the oesophagus. Oesophagitis is an inflammation or irritation of the oesophagus

Ophthalmic: Related to, or situated near the eye

Overlap syndromes: Presence of the features of more than one connective tissue disease in a patient

Pericardium, Pericarditis: The lining of the heart is called the pericardium and its inflammation is called pericarditis

Peripheral blood circulation: The flow of blood to the arms and legs

Phenomenon: An unusual, significant, or unaccountable fact or occurrence which, when observed, is of scientific interest

Physiotherapy (Abbreviated "PT", See also Occupational Therapy): Treatment of disease and injury by mechanical means such as massage, regulated exercise, water, light, heat and electricity. It is often concerned primarily with joint motion, large muscle groups and activities such as walking and aerobic and isometric exercise

Pleura, Pleurisy: Pleura is the lining of the lungs and its inflammation is called pleurisy

Prognosis of disease: Prediction of the progression and end result of a disease, or estimate of chance of recovery

Pulmonary fibrosis (fibrosing alveolitis): A process of scar tissue development in the lungs, decreasing the transfer of oxygen to the blood

Pulmonary arterial hypertension: Elevated pressure in the arteries of the lungs, decreasing blood oxygen and straining the right side of the heart

Raynaud's phenomenon: A disorder with recurring spasms of the small blood vessels upon exposure to cold; characterized by fingers and toes turning white, blue and red as circulation abnormally overreacts to normal conditions. Emotional stress may also trigger an attack. Named after the French physician (Dr. Maurice Raynaud, pronounced "Ray-no") who first described it. Primary Raynaud's (previously called Raynaud's disease) is a common, benign condition which is not caused by any underlying conditions. When Raynaud's phenomenon is caused by scleroderma or other diseases, it is called secondary Raynaud's phenomenon

Relaxation techniques: Stress-reducing procedures, which can also be used to help regulate body functions such as finger temperature or pulse rate. These include tensing

and relaxing muscles, imagery, breathing techniques, and medication (See also Biofeedback)

Remission, spontaneous remission: A period during which the symptoms of a disease decrease or go away. If the reason for remission is not related to treatment but seems to occur for no apparent reason, it is called spontaneous

Renal: Relating to the kidneys

Respiratory: Pertaining to breathing or the lungs

Rugae: Ridges produced by folding of the skin over the upper lip

Salivary glands: Glands which secrete fluid (saliva) into the mouth

Sclerodactyly: Thick, tight skin of the fingers and/or toes (See Joint contracture)

Sclerosis: An abnormal hardening of tissue

Sicca symptoms: Sicca literally means dry; is often used to describe symptoms of dry eyes, mouth etc. experienced by some people with scleroderma or other connective tissue diseases

Sjögren's Syndrome: Sjögren's syndrome is a chronic autoimmune disease characterised by inflammation affecting the exocrine glands (tear, salivary) leading to decreased tear and saliva secretion. Extraglandular manifestations such as joint pain, fatigue or internal organ complications are less common. It can occur on its own (primary) or in association with other connective tissue diseases (secondary) (Pronounced "show-gren's")

Skin ulceration: A break in the skin with loss of surface tissue. It may also be associated with inflammation, calcium deposits and infection

Spasm: Involuntary and abnormal contraction of muscle

Stasis: A slowing or stoppage of the movement of body fluids or reduced motility of the intestines with retention of faeces

Systemic: Affecting the whole body rather than one of its parts. Opposite of localised

Telangiectasia: An abnormal dilation of skin capillaries causing red spots on the skin

Vascular: Pertaining to, or composed of blood vessels

Vasodilator: A medication (or other substance) which causes widening of blood vessels

VEDOSS: Very Early Diagnoses clinic for Systemic Sclerosis, to be initiated throughout Europe

Additional resources

Here are some more sources of good, reliable information about scleroderma:

Books

The following books may be ordered through your local book shop and are available through www.amazon.co.uk (in the UK) or www.amazon.com (in the USA):

The Scleroderma Book

By Maureen Mayes, M.D., M.P.H.
ISBN 0-19-516940-9
A comprehensive guide to the disease written especially for patients and their families

Voices of Scleroderma Volumes 1 – 3

By ISN
Three volumes of stories by people with scleroderma and carers from around the globe

Scleroderma – The Inside Story

By Anne H Mawdsley MBE
ISBN 0-9538297 15

The Best of the Beacon

Edited by Marie Coyle
A marvellous collection of practical and inspirational articles for those living with scleroderma

Leaflets

The following information leaflets are available to download from our website www.sclerodermasociety.co.uk or by post with a SAE size A5 from The Scleroderma Society. Please state which leaflets you require:

• **The Skin in Scleroderma**	• **Localised Scleroderma**
• **Physiotherapy and Scleroderma**	• **The Gastro-Intestinal Tract**
• **Scleroderma in Pregnancy**	• **Scleroderma & the Fingers**
• **Scleroderma & the Foot**	• **Scleroderma in Kidneys**
• **Scleroderma in Young People**	• **Scleroderma, Not just Hard Skin**
• **Scleroderma of the Lung & Heart**	• **Scleroderma Oral Problems**
• **Sexual Problems in Women with Scleroderma**	• **Pulmonary Hypertension in Scleroderma**

Helpful links

Organisations with a specific interest in scleroderma:

UK & Ireland

The Scleroderma Society
Advice Line: 0800 311 2756
Office: 020 7000 1925
www.sclerodermasociety.co.uk
info@sclerodermasociety.co.uk

Raynaud's & Scleroderma Association
Tel: 01270 872776
www.raynauds.org.uk
info@raynauds.org.uk

Irish Raynaud's & Scleroderma Society
Tel: 00 353 1 2020184
www.irishraynauds.com/raynauds.htm
info@raynauds.com

Europe

FESCA (Federation of European Scleroderma Associations) - includes links to national scleroderma organisations in Europe.

www.fesca-scleroderma.eu

EUSTAR (EULAR scleroderma trials and research group) -lists members and affiliates of EUSTAR - Includes links to European scleroderma Organisations.

www.eustar.org

Australia

Scleroderma Australia - includes links to scleroderma organisations in Australia.

www.sclerodermaaustralia.com.au

USA & Canada

Arthritis Foundation
www.arthritis.org

Scleroderma Foundation
www.scleroderma.org

Scleroderma Research Foundation
www.sclerodermaresearch.org

International Scleroderma Network (ISN)
www.sclero.org

Scleroderma Society of Canada
www.scleroderma.ca

Other related organisations

Pulmonary Hypertension Association
Tel: 01709 761450
www.pha-uk.com
enquires@pha-uk.com

Arthritis Research Campaign (ARC)
www.arc.org.uk

Arthritis Care
www.arthritiscare.org.uk
helplines@arthritiscare.org.uk

Notes

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UK Registered Charity Number 286736

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