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.

Scleroderma Australia wishes to thank the Scleroderma Foundation in the United States for allowing us to reprint this booklet. We owe a great deal of gratitude to Dr. Wendy Stevens for editing this booklet for Australian audiences.

Funding for this booklet was provided by an educational grant from Actelion Pharmaceuticals Pty Ltd.

Funding for this booklet was provided by an unrestricted educational grant from Actelion Pharmaceuticals Pty Ltd.

Understanding & Managing Scleroderma

contents



scleroderma australia
Supporting the Scleroderma Community in Australia

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 has been called "progressive systemic sclerosis," but the use of that term has been discouraged since it has been found that Scleroderma is not necessarily progressive. The disease may take several forms which will be explained later. There is also much variability among patients.

Scleroderma is a disease whose symptoms may be visible, as is the case when the skin is affected, or the symptoms 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. Prompt and proper diagnosis and treatment by qualified doctors may minimise the symptoms of Scleroderma and lessen the chance for irreversible damage.

Who develops Scleroderma, and when?

It is estimated that there are over 5000 persons with Systemic Scleroderma in Australia. . Statistically, approximately three to four times more women than men develop the disease. Scleroderma can develop and is found in every age group from infants to the elderly, but its onset is most frequent between the ages of 25 to 55.

Several factors including gender, race and ethnic background, may influence the risk of getting Scleroderma, 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 make those determinations.

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

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

Collagen is the major protein portion of the connective tissue of the body, which is the tissue that hold the cells together. Collagen is found in the skin, joints, tendons, and in parts of internal organs. 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.

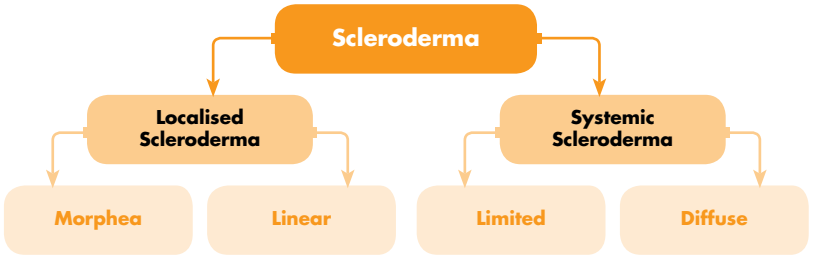
There are several theories about how collagen is overproduced. The *"autoimmune theory"* suggests that the body's own immune system plays a part. Normally, the body's immune system produces chemical signals in the blood called cytokines, which coordinate the body's defence against bacteria, viruses, and other foreign invaders. In addition, some cytokines help to repair wounds by stimulating collagen production to form a scar. There are a number of theories on the way in which the immune system is inappropriately overactivated resulting in excessive amounts of cytokines being produced. These cytokines cause damage to the body's own healthy tissues and can also stimulate 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 be a connection between the build up of excess collagen and the damaged blood vessels, . It has been suggested that the damaged blood vessels may allow increased cytokine leakage into the surrounding tissues which in turn may stimulate the excess collagen production

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: Morphea or Localised Scleroderma and Systemic Scleroderma *(SSc)* is further divided into Limited and Diffuse.



Morphea or Localised Scleroderma

In this condition there are localised patches of thickened skin. The skin affected often appears waxy and may have red or brown colour. These changes are usually found in only a few places on the skin and occasionally in the underlying muscles. They rarely spread elsewhere. The patches may enlarge or shrink, and often disappear spontaneously. Morphea 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 and very rarely have any internal organ involvement. The long-term outlook is excellent. People with morphea rarely develop systemic Scleroderma. Antinuclear antibodies that are normally found in the blood of people with Systemic Scleroderma are generally absent in people with Morphea or localised Scleroderma.

Linear Morphea 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, oesophagus, gastrointestinal tract *(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)* may occur in the internal systems of the body. There are two major recognised patterns that the illness can take, limited or diffuse disease. The extent of skin involvement is used to divide the patients into these two groups. In general, the skin involvement in Scleroderma begins at the fingers and spreads up the arms. Some thickening of the skin of the face is very common, and in some patients the legs are also involved. In the legs,

the skin thickening tends to begin on the foot and spread up the leg. Patients are classified as limited if, in addition to the involvement of the face, there is thickening of the skin from the hands only to the elbows, and in the legs, if the thickening extends from the foot only as far as the knee. Patients are classified as diffuse if there is more extensive spread of the skin thickening, that is, the skin of the upper arms, thighs or trunk is involved.

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

Table 1

Limited Scleroderma

Limited Scleroderma usually causes Raynaud's phenomena and hardening of the skin in the hands. There may be some changes in the facial skin and as indicated above, occasionally there is thickening of the skin on the forearm and lower leg. Oesophageal problems are common. Although, as indicated in *Table 1*, occasionally other internal organ involvement does occur, it is important to realise that this involvement is often very mild and may occur only after many years of the disease. The onset of limited Scleroderma is often very slow, and any progression of skin involvement is also very slow occurring only after many years. The outlook for limited Scleroderma is generally very good. About 70% of people with Systemic Scleroderma have Limited Scleroderma with the other 30% having the more severe Diffuse form.

Diffuse Scleroderma

Diffuse Scleroderma affects the skin not only on the hands and forearms but it can also affect the skin on the trunk, upper arms and thighs. Patients with this condition often have a more systemic illness with the Scleroderma process potentially affecting many other organs and tissues. This type of Scleroderma often requires more intensive treatment, and some patients with this type have a serious disorder. Diffuse Scleroderma generally has a fairly rapid onset of disease with the skin thickening spreading rapidly over a few months. However skin thickening can remit after several years with little long-term damage.

CREST is another name sometimes used to describe a subgroup of patients with Scleroderma. This term was more commonly used in the past but nowadays it is used less commonly as classification into Limited and Diffuse has been found more useful in predicting long-term outlook. CREST is the acronym for the clinical combination of Calcinosis, Raynaud's phenomena, oEsophageal problems, Sclerodactyly *(stiff fingers)* and Telangectasia *(small dilated red vessels in the skin of the hands or face)*. Most patients with CREST have Limited Scleroderma.

Although most patients can be classified as having either Diffuse or Limited disease, different people may have different symptoms and different combinations of symptoms of the illness.

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 lupus (SLE). 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 and other studies. 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

Diffuse and limited Scleroderma can sometimes be differentiated by the presence of different antibodies, called anti-nuclear antibodies *(ANA)* in the blood. For example, anti-Scl-70 is more frequently associated with diffuse Scleroderma whereas anti-centromere usually occurs in limited Scleroderma. *(Refer to Table 2)*.

	Limited Patients	Diffuse Patients
Anti-nuclear antibody	98% test positive	98% test positive
Anti-centromere antibody	41% test positive	3% test positive
Anti-Scl-70 antibody	16% test positive	29% test positive

Table 2

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 with many possible symptoms that can affect many parts of the body. 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 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.

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.

Many common-sense preventive 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 toprotect the hands with gloves when touching refrigerated or frozen items . Electric heaters, electric blankets, and comforters 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 is helpful but 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.

Smoking definitely worsens Raynaud's phenomenon. For this and 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.

Your doctor may suggest a number of different medications to prevent, reduce the frequency, or minimise 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 antagonists, e.g. Adalat, Norvasc, Felodur and Diltiazem . The ACE inhibitors can also be helpful and there have been some reports of Fluoxetene an antidepressant being used to treat Raynaud's with good effect. However, these medications can cause side effects such as palpitations, facial flushing, headaches, light-headedness, swelling of ankles and constipation, which might not allow the patient to take a large enough dose to control their Raynaud's. These side effects are all temporary and will go away once the medication is ceased or the dose reduced.There are now over a dozen medications to improve circulation, and it is not possible to list them all here.

In severe cases particularly when there are digital ulcers or infection, treatment with a prostaglandin or prostacyclin infusion may be recommended. These treatments are given via an intravenous drip. The duration of treatment is generally 3 days but can vary depending on the circumstances. This treatment can produce an improvement in Raynaud's for three or more months and has been shown to help the healing of digital ulcers in some people.

Raynaud's phenomenon is not confined to people with Scleroderma. It is also seen in lupus, rheumatoid arthritis, 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 disease*." It is thought that about 10% of women and 5% of men in the general population have primary Raynaud's disease. For most of these people with primary Raynaud's the Raynaud's began in their teenage years and is often quite mild and rarely needs treatment.

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. However because of their side effects they are not suitable for all patients. Sometimes your doctor may recommend use of an immunosuppressive agent such as methotrexate or plaquenil if the arthritis is severe.

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 disorders

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. However in patients with rapidly progressing Diffuse Scleroderma your doctor may recommend a trail of an immunosuppressant such as Methotrexate or Mycophenolate or Cyclophosphamide. It is however important to note that some patients with diffuse disease often experience a dramatic softening of their skin after several years.

Skin ulcerations

Sores, especially on the fingertips, are a common symptom of systemic Scleroderma. They may be very slow or difficult to heal because of poor circulation. These sores or ulcerations 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. If infection should develop, it may be helpful to soak the affected area in warm water and apply an antiseptic such as betadine. Should these remedies prove unsuccessful in relieving the pain or infection of ulcerated skin, your doctor may prescribe oral antibiotics or take other measures. It is very important to consult your doctor early if you suspect an ulcer is infected as early treatment can prevent further damage. Appropriate dressings can aid healing and relieve pain. Nowadays many hospitals have clinics or nurses who specialise in management of wounds ulcers with specialised dressings. If you have a wound or ulcer that is not healing quickly it may be worthwhile discussing with your doctor referral to one of these specialised centres for assistance with dressings and wound care.

Calcinosis

This condition is characterised 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 may become infected. Care should be taken not to bump or injure affected areas. Warm-water soaks may be helpful. Antibiotics may be

prescribed to prevent or control infection. In severe cases, 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. Laser therapy by a trained doctor can also be used to reduce the number of telangiectasia although it is not uncommon for the telangectasia to regrow in the treated area.

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 moisturizing skin creams containing lanolin or sorbelene is advisable. A room humidifier may be helpful during the winter months.

Itchy skin

If moisturizing creams do not work, your doctor may prescribe a topical cortisone cream to be rubbed on the skin to relieve itching. Antihistamines tablets have been effective for some people.

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 suntan)* 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 generally occurs after initial swelling has subsided. It is characterised by shiny, tight skin of the fingers. Affected fingers may be difficult to move, and they may become fixed in a bent or flexed position called a *"contracture"* or 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 from the mouth 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 a tube called 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 stomach 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 by the stomach can be reduced, and the problems of acid reflux and heartburn lessened, by avoiding *(to the extent possible)* alcohol, greasy or fatty foods, spicy foods, chocolate, tobacco, and caffeine. However many people, perhaps the majority, will 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 will do this. The most commonly used drugs nowadays are known as proton pump inhibitors. These include Nexium, Losec ,Zoton, Pariet and Somac. The H2 receptor antagonists can also decrease acid production and are sometimes used. Examples of type of drug are Zantac, Tagamet and Tazac. In some people with severe reflux a combination of the two different classes of drugs are 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 although unfortunately these drug are not very effective in improving the motility of these organs.

The force of gravity helps to keep food and acid in the stomach; therefore, an upright position after meals is 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.

Swallowing difficulties

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. If the oesophagus has narrowed significantly, you may need to have the oesophagus dilated periodically to permit easier swallowing.

Diarrhœa

In systemic Scleroderma, there can be damage to the muscles of the small bowel *(small intestine)*. The weakened muscles do not work effectively to push food through the bowel. Simply put, things *"sit"* rather than move well. One consequence can be an overgrowth of bacteria, leading to diarrhœa. 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 diarrhœa 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.

Constipation

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 Metamucil® or Fybogel.

Sjögren's Syndrome

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

Sjögren's Syndrome *(dry eyes, dry mouth)* is characterised by a decrease in secretions of the tear glands 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 in Sjögren's Syndrome 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. 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 candy to stimulate salivary activity. Artificial saliva is also available. *(See next section for preventive dental care.)*

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

Oral, facial, and dental problems

People with Scleroderma may experience a general tightening of skin over the face. The opening of the mouth may be decreased in sise *("microstomia" or "small mouth")*, making lip and mouth movements as well as oral hygiene difficult.

The best approach to treatment is by means of facial grimacing and mouth stretching exercises daily. Good dental care by regular flossing and brushing of the teeth and gums after each meal is very important. Regular dental visits are also important to help prevent dental caries. Your dentist can also recommend a program of good oral hygiene. Floss holders, pump toothpaste tubes, and built-up handles on toothbrushes can help people with hand impairment *(These measures are equally important for those with Sjögren's Syndrome.)*

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"*, 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 4 years of their disease. It is very uncommon to see renal crisis in people with Limited Scleroderma.

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 involves anti-hypertensive drugs that belong to the category of ACE inhibitors. 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 Scleroderma are advised to have their blood pressure and kidney function monitored at regular intervals.

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. Interstitial Lung Disease (ILD)

A 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 fibrosis or Interstitial Lung Disease *(ILD)*. ILD in the person with scleroderma can range from very mild and non progressive to a very severe widespread and progressive condition. Small amounts of fibrosis in the lungs occur in up to 50% of people with

Scleroderma. Fortunately in only a small number of people does the condition progress and have any major affect on their lungs. Studies have shown that people who are going to develop serious lung involvement are likely to have developed significant impairment of lung function in the first 4 years of their Scleroderma illness. Recent trials have shown that patients with severe and progressive lung fibrosis may gain some benefit from immunosuppressive drugs such as Cyclophosphamide. There are also trials of other immunosuppressive therapies underway. Regular breathing (pulmonary function tests), especially in the first few years of the illness, can detect early lung involvement. If any abnormality is detected on these pulmonary function tests your doctor will probably also order a CTscan of the chest to further determine if there is any evidence of ILD. Regular follow-up with repeated pulmonary function tests is used to determine if the condition is progressing and therefore likely to require specific therapy.

2. Pulmonary Arterial Hypertension (PAH)

This is a state of increased resistance to blood flow through the lungs and can result from damage to blood vessels. With time this may lead to increased strain on the heart resulting in heart failure. PAH develops in10-15% of patients with Systemic Scleroderma. PAH often occurs after some years of Scleroderma. Pulmonary Arterial Hypertension *(PAH)* can now be treated with special medications targeted at dilating or opening up the blood vessels of the lungs, and possibly changing the underlying nature of the disease. This is one complication of Scleroderma for which new medications have proven successful. In Australia there are currently five medications approved for the treatment of PAH associated with Scleroderma. There are three different classes of drugs used to treat PAH. The most frequently used are the enodothelin antagonists. These medications block the action of endothelin a naturally occurring cytokine that is increased in patients who have PAH. Endothelin is a very potent constrictor of blood vessels in the lung and blocking its action can improve the blood flow through the lung. Currently there are 3 different endothelin antagonists - Bosentan (brand name Tracleer®), Sitaxentan (brand name Thelin®) and Ambrisentan(brand name Volubris®) . These are all tablets taken once or twice a day. Iloprost (brand name Ventavis®) is a prostacylin derivative that is inhaled via special hand held nebuliser every 2-3 hours during the daytime. The third class of drugs that are used in PAH are the PDE5 inhibitors –the commonest of these is sildenafil(brand name Revatio and Viagra®). All of these drugs have been shown to improved exercise capacity, quality of life and life span of people with PAH. These medications can only be prescribed by specialists with experience in managing PAH and require careful monitoring and regular followup.

Early detection and treatment of PAH seems to improve outcome for those affected by this condition. For this reason it is increasingly being recommended that the person with Scleroderma has regular pulmonary function tests and an ultrasound of the heart called and Echocardigram. These tests can be used to detect PAH init early stages.

3.Chest Wall Involvement

Respiratory muscle weakness and tight skin on the chest wall may decrease lung function and cause shortness of breath.

Symptoms of lung involvement include shortness of breath, a decreased tolerance for exercise, and a persistent cough. Should you develop any of these symptoms your doctor may order a chest x-ray and/or CT scan of the chest, an echocardiogram *(ultrasound of the heart)* and special breathing tests *(pulmonary function tests)* to detect or confirm lung involvement and to determine which of these 3 processes is responsible for the symptoms. However as indicated above because of new therapies for lung complications of scleroderma doctors are increasingly recommending regular screening for these complications with pulmonary function tests and echocardiogram even in people without symptoms of lung problems.

It is important for people with Scleroderma to take whatever measures are within their 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 to as much as possible. The physician may recommend medications to make breathing easier and may also suggest deep breathing exercises and a graduated aerobic exercise program.

Heart involvement

If the heart muscle becomes thickened and fibrous scar tissue accumulates, the force of heart contractions may be decreased, which may ultimately result in heart failure. Spasm of the coronary arteries *(the main blood vessels to the heart)*, may cause chest pain and, rarely, lead to a heart attack. The spasm appears similar to that involving the fingers in Raynaud's phenomenon. Inflammation of the outer heart lining *(pericarditis)* 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, generalised 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.

Scleroderma symptoms vary from person to person, requiring different treatment

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 persons affected by it. Under these circumstances, it is difficult to conduct scientifically sound studies proving the value of a particular drug or treatment. Therefore, your doctor must 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.

You may want to ask for a second opinion from another specialist even if you have already started treatment or still want to be treated by your first doctor. A copy of your file can be sent to this doctor.

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.

Learning to recognise 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 your 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 can help to distinguish what is related to Scleroderma and what is not, and recommend appropriate treatment.

Developing an individual treatment program

While there is no proven cure for Scleroderma, much can be done to prevent, minimise, 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 program to meet the specific needs of a person with this disease. Close cooperation with your doctor will help him or her develop such a program.

Many forms of treatment have been discussed already in the chapter titled *"What are the symptoms of Scleroderma, and how are they treated?"* The next six sections will discuss other important elements of a program for managing Scleroderma

Physical therapy and exercise

Physiotherapists can help the person with Scleroderma develop an appropriate program. Such a program 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 program 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 program should be built up gradually.

Protecting the joints

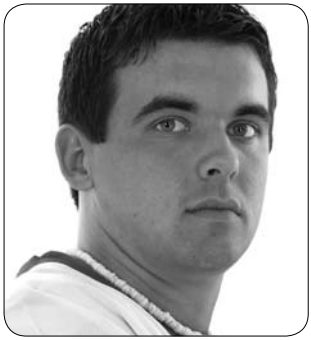
The goals of joint protection are to minimise further damage and to reduce the possibility of skin ulcers and infection. Its basic principles include avoiding or minimizing pressure or stress on the joints by their proper use, and maintaining their mobility and function by stretching and "range of motion" 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.

There are also a number of books, pamphlets, CDs and DVDs that discuss how to apply the principles of joint protection, describe many of the self-help aids and adaptive mechanical devices, and cover other topics of interest to the person with Scleroderma. Two books, written especially for those with arthritis but very helpful to the person with Scleroderma, are *The Arthritis Helpbook* by Kate Lorig and James F. Fries, and *Self-Help Manual for Patients with Arthritis,* prepared by the Arthritis Health Professionals Section of the Arthritis Foundation.

Taking medications

t is essential that the person with Scleroderma take all medicines wisely; take only those prescribed; read label warnings 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 medication program accordingly.

Commonsense measures

Treatment of specific symptoms has been discussed in other sections of this booklet. There are also a number of general commonsense 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 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.

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 program, 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 and 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.

Joining a Scleroderma support group, such as one affiliated with Scleroderma Australia, 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.

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!

There is currently no cure for Scleroderma but, in many cases, it can be maganged

scleroderma australia

Scleroderma Australia Inc. was formed in 2005 and is the national body supporting the Scleroderma community in Australia. Our mission is to support the Scleroderma community in Australia through Awareness, Education and Research.

Scleroderma Australia's services include:

Scleroderma Australia Inc. was formed in 2005 and is the national body in Australia representing and advocating for people with scleroderma. Our mission is to support the scleroderma community in Australia through Awareness, Education and Research.

Scleroderma Australia's services include:

- Centralised message bank, with calls returned promptly, **02 9990 5159**
- Website with news and information, www.sclerodermaaustralia.com.au, including links to state-based associations
- Advocate for and support Scleroderma research
- Providing educational literature for patients and members of the medical community
- Educational meetings and seminars across Australia
- National awareness campaigns
- Advocate for scleroderma research, and for legislation benefiting people with scleroderma.

Scleroderma Australia supports local state associations in New South Wales, Victoria, Queensland, Western Australia and South Australia which have support groups and other services designed to help persons with scleroderma, their families and carers at the local level *(see contact info at the back of this booklet)*.

Scleroderma Australia can put you in touch with the state association or support group nearest to you. If there is no Support Group nearby, Scleroderma Australia can help you start one.

Scleroderma Australia

Scleroderma Australia,
c/- Scleroderma Victoria Inc.,
St. Vincents Hospital, 41 Victoria Parade,
FITZROY Vic 3065

Email. info@sclerodermaaustralia.com.au
www.sclerodermaaustralia.com.au

scleroderma australia
Supporting the Scleroderma Community in Australia

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, and 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 auto-immune diseases. 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 Calcinosis, Raynaud's phenomenon, oEsophageal dysmotility, Sclerodactyly, and Telangiectasia.

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.

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, disfunction. 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 saber or sword wound usually on face or neck.

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

Fibrous. Consisting of, or resembling fibers.

Fibrosis. Abnormal formation of excess fibrous tissue.

Gastrointestinal tract, bowel, diarrhea, 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. Diarrhea 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 cells, lymphocytes, T cells, and B cells. Immunoglobulins *(antibodies)* are proteins that can react with and/or neutralise 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, frequently affecting the fingers, due to tightening and hardening of the skin around the joint. In flexion contractures, the fingers become fixed in a bent or 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 to take nutrients from food into the cells of the body 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.)*

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

Oedema. An abnormal excess accumulation of fluid in tissues or cavities 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.

Pericarditis. Tissue inflammation of the sac enclosing the heart.

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. Treatment of disease and injury by mechanical means such as massage, regulated exercise, water, light, heat, and electricity. Often concerned primarily with joint motion, large muscle groups, and activities such as walking and aerobic and isometric exercise. *(See also Occupational Therapy.)*

Pleurisy. Tissue inflammation of the sac enclosing the lungs.

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

Pulmonary fibrosis. A process in which the lungs are scarred, decreasing the transfer of oxygen to the blood. Also called restrictive lung disease.

Pulmonary hypertension. Elevated pressure in the blood vessels of the lungs, decreasing blood oxygen and straining the right side of the heart.

Raynaud's phenomenon. Also called Raynaud's syndrome. A disorder with recurring spasms of the small blood vessels upon exposure to cold; characterised by fingers and toes turning white, blue, and red as circulation abnormally overreacts to normal conditions. Emotional stress may also trigger an attack. Named for the French physician *(Dr. Maurice Raynaud, pronounced "Ray-node")* who first described it.

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.

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.

Sjögren's Syndrome. A chronic inflammatory disease characterised by decreased secretions, especially dry eyes and dry mouth, named for the Swedish physician who first described it. It may occur alone, or as a part of Scleroderma or other auto-immune diseases. *(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 body fluids as in venous stasis. Also, 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.

additional resources

Here are some more sources of good and reliable information about Scleroderma.

Books

The following books may be ordered through your local bookseller or available through www.amazon.com.au or by calling toll-free 1300 ?

The Scleroderma Book
By Maureen Mayes, M.D., M.P.H.

A comprehensive guide to the disease written especially for patients and their families.

The Best of the Beacon
Edited by Marie Coyle

A marvellous collection of practical and inspirational articles for those living with Scleroderma.

Perspectives on Living with Scleroderma
By Mark Flapan, Ph.D.

Dr. Flapan's insightful articles on coping, including his personal story.

Scleroderma: A New Role for Patients and Families
By Michael Brown

Offers resources to help you take charge.

Scleroderma: Surviving a 17-Year Itch
By Dana Lovvorn

Self-help book for newly diagnosed patients, emphasising exercise and lifestyle adaptations.

Successful Living with Scleroderma: Guidebook
By Robert Phillips, Ph.D.

Websites

Australia

Scleroderma Australia
www.sclerodermaaustralia.com.au

The national nonprofit organisation in Australia representing and advocating for persons with Scleroderma. Includes links to state-based Scleroderma organisations.

Arthritis Australia
www.arthritisaustralia.com.au

National arthritis organisation in Australia. It is supported by affiliate offices in every state and territory.

Pulmonary Arterial Hypertension
www.pah.com.au

An online resource for people, including patients as well as healthcare professionals, seeking information on pulmonary arterial hypertension *(PAH)*.

USA & Canada

Arthritis Foundation
www.arthritis.org

U.S. arthritis foundation supporting research and community services, helping to find a cure for arthritis. Serving Americans with arthritis, their families and health professionals who help them.

Scleroderma Society of Canada
www.scleroderma.ca

International Scleroderma Network (ISN)
www.sclero.org

Nonprofit international patient and medical organisation. Dedicated to research, support, education and awareness for scleroderma and related illnesses.

Scleroderma Foundation
www.scleroderma.org

The national nonprofit organisation in the U.S. representing and advocating for persons with Scleroderma.

Scleroderma Research Foundation
www.sclerodermaresearch.org

A non-profit foundation to fund basic and clinical research for better treatments and eventually, a cure, for scleroderma.

UK

Raynaud's and Scleroderma Association
www.raynauds.org.uk

UK based national charity and self help organisation, committed to supporting patients and carers who have these conditions.

Scleroderma Society
www.sclerodermasociety.co.uk

The Society aims to offer support to patients who often feel isolated, to increase awareness of the disease and to raise money for vital research.

Pulmonary Hypertension Association
www.phassociation.org

Pulmonary Hypertension Association *(PHA)* seeks to find a cure for pulmonary hypertension and provides hope for the pulmonary hypertension community through support, education, advocacy and awareness.

Information on the Internet

You may be interested in looking for information about scleroderma on the Internet. While there are some very good websites you need to be aware that some websites provide misleading or biased information and may also be outdated.

state-based groups

New South Wales

The Scleroderma Association of New South Wales Inc.
PO Box 227, Ashfield NSW 1800.
Tel. 02 9798 7351 Fax. 02 9798 7985 Email. scleronsw@aol.com

Newcastle/Hunter Valley

Scleroderma/Lupus Support Society
PO Box 373, Warner's Bay NSW 2282. Tel. 02 4921 4095
Email. Renae.Morgan@hnehealth.nsw.gov.au

Northern Territory

Arthritis and Osteoporosis NT
PO Box 452, Nightcliff NT 0814.
Email. health.educator@aont.org.au

Queensland

Scleroderma Queensland
PO Box 154, Mansfield QLD 4122.
Tel. 07 3411 3558 Fax. 07 3411 3558
Email. sclero@pacific.net.au www.scleroderma.org.au

South Australia

Lupus/Scleroderma/Sjogren's Support Group,
1/202 Glen Osmond Road, Fullarton SA 5063
Tel. 08 8263 0513

Tasmania

Contact the Scleroderma Foundation of Victoria.

Victoria

Scleroderma Foundation of Victoria Inc.
St. Vincents Hospital. 41 Victoria Parade, Fitzroy VIC 3065.
Tel. 03 9288 3651 Email. enq@sclerodermavic.org

Western Australia

Scleroderma WA
Arthritis Foundation of WA
17 Lemnos Street, Shenton Park WA 6008. Tel. 08 9388 2199
Email. general@arthritiswa.org.au

notes