scleroderma australia

# UNDERSTANDING & MANAGING **SCLERODERMA**

SEVENTH EDITION 2024

This booklet is intended to help people with Scleroderma, their families, and others interested in Scleroderma. The aim is to better understand what Scleroderma is, what effects it may have, and what people with Scleroderma can do to help themselves and their health care team manage the condition. It answers some of the most frequently asked questions 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 and patient advocates for editing this booklet for Australian audiences.

Funding for this booklet was provided by a grant from J&J a business unit of Janssen-Cilag Pty Ltd.

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# What is scleroderma?

**Scleroderma, or systemic sclerosis, is a chronic connective tissue condition 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 condition. It is also often called systemic sclerosis, however, we will use the term scleroderma throughout this booklet. The condition may take several forms which will be explained later. There is also much variability among people with scleroderma.

Scleroderma is a condition where 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 condition 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 minimize the symptoms of scleroderma and lessen the chance for irreversible damage.

## Who develops scleroderma, and when?

It is estimated that there are over 6000 people with systemic scleroderma in Australia. Statistically, approximately three to four times more women than men develop the condition. Scleroderma 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, ethnic background and occupational exposure, 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 conditions.

**"Scleroderma can develop and is found in every age group from infants to the elderly, but it's onset is most frequent between the ages of 25 to 55."**

## What causes scleroderma?

The exact cause or causes of scleroderma are still unknown, but scientists and medical investigator in a wide variety of fields are working hard to make those determinations.

What is known about the 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 holds 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 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 defense 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. Systemic scleroderma (SSc) is further divided into limited and diffuse.

**"About 70% of people with systemic scleroderma have limited scleroderma with the other 30% having the more severe diffuse form."**

## 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 10 and 50 but can also be seen in young children.

People 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 or SSc)*

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 condition can take, limited or diffuse. The extent of skin involvement is used to divide people 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 people the legs are also involved. In the legs, the skin thickening tends to begin on the foot and spread up the leg. People are classified as having limited scleroderma 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. People are classified as having diffuse scleroderma if there is more extensive spread of the skin thickening, that is, the skin of the upper arms, thighs or trunk is involved.

## Table 1

| Manifestation | Limited Scleroderma | Diffuse Scleroderma |
|---|---|---|
| Raynaud's phenomenon | 95% | 80% |
| Skin | 95% | 100% |
| Gastro-oesophageal reflux | 90% | 90% |
| Lung fibrosis | 30% | 30% |
| Heart Less than | 5% | 10% |
| Pulmonary hypertension | 15% | 10% |
| Kidney disease Less than | 5% | 10% |
| Telangiectasia | 91% | 64% |
| Calcinosis | 42% | 17% |

## 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 condition. 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.

CREST is another name sometimes used to describe a subgroup of people 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 outcome. **CREST** is the acronym for the clinical combination of **C**alcinosis, **R**aynaud's phenomena, o**E**sophageal problems, **S**clerodactyly (stiff fingers) and **T**elangiectasia *(small dilated red vessels in the skin of the hands or face)*. Most people with CREST have limited scleroderma.

## 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. People 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 people with this type have a serious disorder. Diffuse scleroderma generally has a fairly rapid onset with the skin thickening spreading rapidly over a few months. However, in diffuse scleroderma the skin thickening can significantly improve after several years with little long-term damage.

Although most people can be classified as having either diffuse or limited scleroderma, different people may have different symptoms and different combinations of symptoms of the condition.

**Table 2**

| Blood tests | 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 | 15% test positive | 30% test positive |
| RNA polymerase antibody | <5% test positive | 15% test positive |

## 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 conditions, especially other autoimmune connective-tissue conditions 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 and RNA polymerase antibodies are more frequently associated with diffuse scleroderma whereas anti-centromere usually occurs in limited scleroderma. (Refer to 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 condition with many possible symptoms that can affect many parts of the body. However, most people only develop a few of the symptoms mentioned. Each person 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 treatments 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."**

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. Hats, earmuffs, 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, 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 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 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 antagonists, e.g. nifedipine, amlopidine, felodipine and diltiazem. The PDE5 inhibitors sildenafil and tadalafil are helpful in some people as are ACE inhibitors. 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 person 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.

**"Keeping the entire body warm helps prevent Raynaud's episodes."**

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 three 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 the Raynaud's began in their teenage years and is often quite mild and rarely needs treatment.

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

## 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, which can contribute to reduced mobility and function, and to disability. 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 people. Sometimes your doctor may recommend use of an immunosuppressive agent such as methotrexate, hydroxychloroquine, mycophenolate mofetil or leflunomide if the arthritis is severe. Occasionally if the arthritis is severe and unresponsive to these medications, one of the newer biological drugs used for rheumatoid arthritis might be considered by your treating rheumatologist.

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

**"Hardening and thickening of the skin gives scleroderma its name ("hard skin")... It is, however, important to note that some people with scleroderma often experience a dramatic softening of their skin after several years."**

## Skin disorders

### *Skin thickening*

Hardening and thickening of the skin gives 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 people with rapidly progressing diffuse scleroderma the doctor may recommend a trial of an immunosuppressant such as methotrexate, mycophenolate with cyclophosphamide or rituximab. It is, however, important to note that some people with diffuse condition often experience a dramatic softening of their skin after several years. There are several new "antifibrotic" medications being investigated in clinical trials around the world. Some of the major hospitals in Australia with scleroderma clinics are participating in these trials. If you have early (<3 years) diffuse scleroderma you may be suitable for one of these trials. It is worth speaking to your rheumatologist about possible trials in your area.

### *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. The symptoms that suggest an ulcer might be infected include sudden increase in local pain, redness around the ulcer and smelly discharge from the ulcer. Appropriate dressings can aid healing and relieve pain. Nowadays many hospitals have clinics or nurses who specialize in management of wounds and ulcers with specialized dressings.

If you have a wound or ulcer that is not healing quickly it may be worthwhile talking to your doctor about a referral to one of these specialized centres to assist 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 and dressings to soften the calcium deposits may be helpful. Antibiotics may be prescribed to treat secondary infection. In severe cases, surgery to remove calcium deposits may be required.

**"Excessive bathing and hand washing should be avoided, and rubber gloves be worn to avoid direct contact with household detergents... Frequent use of moisturising creams containing lanolin or sorbolene is advisable."**

### *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 telangiectasia 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 moisturising soaps such as Neutrogena® and Dove™ are preferable to harsh soaps which dry out the skin. Frequent use of moisturising skin creams containing lanolin or sorbolene is advisable. A room humidifier may be helpful during the winter months.

### *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. Antihistamine 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 characterized 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, causing mobility impairment and disability.

"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. They may also provide devices to help perform common personal care and household tasks more easily.

**"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 15 to 20cms with wooden blocks."**

## 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 oesophagus.

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 esomeprazole, omeprazole pantoprazole, lansoprazole and rabeprazole. The H2 receptor antagonists can also decrease acid production and are sometimes used, example of this type of drug is nizatidine. 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 drugs 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 15 to 20cms 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 digestion are made easier by eating softer foods such as slow cooked casseroles, soups, fish and egg dishes 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.

**"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."**

### *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 (SIB0), 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 dietitian.

### *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, can 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 psyllium husk, Metamucil® or Fybogel.

## Sjögren's syndrome

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 occasionally 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 K-Y Jelly 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 size (*"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 decay.

Your dentist can also recommend a program of good oral hygiene. Floss holders, water flossers, pump toothpaste tubes, electric toothbrushes and built- up handles on toothbrushes can help people with hand impairment. *(These measures are especially important for those with Sjögren's syndrome.)*

### *Kidney involvement*

Although kidney or renal involvement in systemic scleroderma is uncommon it can be quite a serious problem. 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 likely to occur in the first four years of their disease. It is very uncommon to see renal crisis in people with limited scleroderma.

**"Renal crisis occurs in about 10% of people with diffuse scleroderma and is most likely to occur in the first four 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 stabilising and even improving kidney function. In cases of severe kidney failure, dialysis may be required. People with early diffuse scleroderma are advised to have their blood pressure and kidney function monitored at regular intervals, and if you have early diffuse scleroderma your doctor may recommend that you check your blood pressure at home every week.

People can recover successfully from renal crisis, especially 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 effect 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 four years of their scleroderma illness. Recent trials have shown that people with severe and progressive lung fibrosis may gain some benefit from immunosuppressive drugs such as cyclophosphamide. Treatment with cyclophosphamide is generally given for 6–12 months and then followed with other immunosuppressive drugs, most commonly azathioprine or mycophenolate. A large study comparing cyclophosphamide to mycophenolate as initial treatment for early severe lung disease has recently been completed in the USA. The results of this study showed that both cyclophosphamide and mycophenolate mofetil slowed the progression of the ILD in about 2/3 of people in the trial. In the other 1/3 the lung disease progressed despite these medications.

As mycophenolate mofetil generally causes less serious side effects than cyclophosphamide, most people with severe or progressive lung disease are treated with mycophenolate first. Cyclophosphamide is reserved for people who do not respond or progress on mycophenolate therapy.

In addition there is a small amount of evidence that an immunosuppressant used sometimes to treat Rheumatoid Arthritis, rituximab may be helpful in people with progressive lung disease.

Recently in Australia a new antifibrotic therapy has been approved for treatment of ILD that has progressed despite other treatments. This medication nintedanib has previously been approved to treat Idiopathic interstitial lung disease but is now approved to treat progressive ILD in scleroderma. It can only be prescribed by specialists with experience in these conditions and can only be used if there is documented decline in lung function of >10% over 12 months or progression of the ILD on a lung scan over the same time period.

Regular breathing tests (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 CT scan 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. It is important to note that lots of people with scleroderma have very mild non-progressive ILD which does not require treatment.

**"It is increasingly being recommended that the person with scleroderma has regular pulmonary function tests and an ultrasound of the heart called an echocardiogram."**

### *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 in 10–15% of people with systemic scleroderma. PAH often occurs after some years of scleroderma. 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 seven medications approved for the treatment of PAH associated with scleroderma.

There are three different classes of drugs used to treat PAH in scleroderma. The most frequently used are the endothelin receptor antagonists.

These medications block the action of endothelin, a naturally occurring cytokine that is increased in people 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 three different endothelin antagonists; bosentan, macitentan and ambrisentan. These are all tablets taken once or twice a day. The second class of drugs that are used in PAH are the PDE5 inhibitors. There are two different PDE5 inhibitors; sildenafil and tadalafil. The third class of drugs works via the prostaglandin pathway.

Epoprostenol is the oldest prostaglandin available to treat PAH in scleroderma. It is given by a continuous intravenous infusion and so it is reserved for the rare person with severe PAH who is no longer responding to these other treatments.

More recently a new oral drug called selexipag has become available for people with PAH who are not responding to a combination of an endothelin receptor antagonist and a PDE5 inhibitor. Whilst not actually a prostaglandin, selexipag works by stimulating the receptor on cells for prostaglandin, so is very similar to epoprostenol in its action and side effects, but can be given by a tablet twice per day.

All of these drugs have been shown to improve 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 follow up.

Early detection and treatment of PAH seems to improve outcomes 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 either a blood test called ProNT BNP or an ultrasound of the heart called an echocardiogram.

These tests can be used to detect PAH in its early stages. A new blood test called NT-proBNP with pulmonary function tests may in the future become an easier way to screen for PAH, although it is unlikely to completely replace echocardiograms for screening for PAH. Whichever method is used it is important that people with scleroderma do have annual screening as PAH can gradually develop even after many years of scleroderma.

### *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)* or blood test with ProNT BNP, and 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. However, as indicated above, because of new therapies for lung complications of scleroderma doctors are increasingly recommending regular screening for these complications with at least annual pulmonary function tests and echocardiogram even in people without symptoms of lung problems. The blood test ProNT BNP is only useful for screening for PAH in symptomatic people and is now reimbursed by Medicare once or twice per year.

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 as much as possible.

Your doctor 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, causing mobility restriction and disability. Treatments or medications recommended by your doctor will depend on their evaluation of the causes of these symptoms.

# Managing scleroderma

**"It is helpful to keep scleroderma in perspective. Many people with Scleroderma have few or minimal symptoms and can continue with their usual daily living activities."**

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 condition 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. Therefore, you and your doctor must often make decisions about treatment based on incomplete information. With everyone weighing up the possible benefits against the potential risks or side effects. Further investigation will ultimately determine which treatments are beneficial and which treatments are not. There are also an increasing number of clinical trials of new therapies for early diffuse scleroderma and for scleroderma related ILD.

These trials are held at specialized hospital-based clinics in Australia. The ASIG website will soon have a list of all these trials https://asigresearch.com.au

## The course of scleroderma

Scleroderma has many forms and a number of different symptoms that may present themselves alone or in combinations at various times throughout the course of the condition. Some symptoms develop with relative suddenness; others take years to develop. The exact course that scleroderma may take is unpredictable, and the prognosis will vary from person to person.

Systemic scleroderma is a chronic, life-long condition. At present there is no known cure. As with other chronic conditions, there are many ways to control or manage its symptoms. It is helpful to keep scleroderma in perspective. Many people with scleroderma have few or minimal symptoms and are able to lead their 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, they may feel quite ill. Spontaneous improvements may occur.

The skin, in particular, can soften and become more pliable after a number of years. Spontaneous remissions, times when symptoms may actually disappear, can also occur and potentially 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 condition, but to provide them with useful information on what to look for, what may occur during the course of their condition, and some of the things that can be done if symptoms do develop.

Learning to recognise early symptoms of scleroderma related 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 any 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, 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 works out an individually tailored treatment program to meet the specific needs of a person with this condition. Close cooperation with your treating team will help them both 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

Exercise Physiologists, Physiotherapists and hand therapists can help the person with scleroderma develop an appropriate program. The most important focus of physical therapy and exercise for people with scleroderma is to maintain and potentially increase movement and improve mobility, cardiovascular fitness, and strength. This includes both joint range and muscle length. This is performed through a guided exercise program of stretching, strengthening and aerobic training, initially supervised and then performed independently. Hydrotherapy can be a useful medium for exercise but may not be suitable if skin ulcers are present and it may exacerbate Raynaud's phenomenon in some people.

**"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."**

### *Hand Therapy*

Hand therapists are either physiotherapists or occupational therapists that specialize in the treatment of upper limb conditions. Hand therapy can play an important role in maintenance of your upper limb joints and can prevent any unnecessary complications caused by poor positioning, postural guarding and inadequate movement on a regular basis. Hand therapists can fabricate orthoses/splints to prevent these unnecessary contractures or to protect joints with ulcers while they are healing. Additionally, hand therapists can provide a range of exercises, thermal modalities and adaptive aids to increase your independence and improve outcomes in your daily living activities.

To find a local therapist, please visit the Australian Hand Therapy Association website – www.ahta.com.au

### *Some exercise considerations*

- Seek advice from a trained exercise professional for a tailored exercise program.
- Ensure you are at a comfortable temperature when exercising (e.g., warm clothing, heating if needed) to help with Raynauds.
- Avoid eating large meals 1–2 hours before performing exercise (some people may need a light bite to keep their stomach settled) to help reduce reflux.
- Raise your head (~10cm) when performing exercise on your back (you can use pillows or a blanket for elevation) to help reduce reflux.
- Use modified equipment to support you with exercise (e.g., adjustable ankle/wrist weights, hand straps).
- If ulcerations are present, you may wish to avoid water-based exercise.
- Ensure you are practising good hygiene to avoid the risk of infection.
- Pace yourself with exercise. Choose appropriate days and times that suit you and your energy levels.
- Listen to your body!

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 minimize pain and 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 strengthening exercises. A variety of self-help aids and adaptive mechanical devices are available to help protect and to alleviate stress on the joints while maintaining your daily activities.Occupational therapists can demonstrate such devices and give further instruction on joint protection.

## Taking medications

It is essential that the person with scleroderma takes all medicines wisely; takes only those prescribed; reads label warnings and follows instructions carefully; and takes the medications exactly when, for how long, and in the dosages prescribed by their 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 as well as any decisions you make to stop medications.

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, whilst others may not. Furthermore, individual tolerance for the drugs used in treating scleroderma varies greatly. Your doctor may find it necessary to adjust the medication program accordingly. If you are unsure about your medications, talk to your doctor and/ or pharmacist.

## 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 their 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 minimize stress by taking care of your mental well-being.
- 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 people 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 condition such as scleroderma is *"Why me?"*

It is not known why some people develop the condition and others do not. One does not bring scleroderma upon themselves; therefore, one need not feel guilty or responsible for the illness.

Someone newly diagnosed with scleroderma may feel alone and uncertain about where to turn for help. They may experience 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 they are 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.

**"Joining a scleroderma support group, such as one affiliated with Scleroderma Australia, enables a person with scleroderma to meet and exchange information with others who have similar experiences, as well as learn more about scleroderma."**

## 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 condition. 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 them to follow the recommended treatment program, and assist in carrying out activities that they find 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 a 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 condition.

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.

As a result of this laboratory research new drugs that target various new mechanisms have been developed and clinical trials of new therapies for severe skin disease and lung disease are underway around the world. However, it is important to understand that the process of developing new treatments is quite slow. Firstly, the new treatment has to be tested for unexpected side effects and then the actual effect of the treatment in people with scleroderma has to be tested. Many new treatments which have seemed promising in laboratory or animal studies have unfortunately not been found to work in people with scleroderma. However, in the last few years there have been increasing numbers of clinical trials in scleroderma.

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

# Scleroderma Australia

**Scleroderma Australia was formed in 2005 to facilitate a national representative voice in supporting our scleroderma community across Australia. Volunteer delegates state associations make up our Board to ensure we have representation across Australia.**

The purpose of our association is to:

- Coordinate and promote scleroderma awareness across Australia
- Increase community support and engagement for people with scleroderma and their families
- Advocate for people with scleroderma for improved patient outcomes
- Coordinate funding for scleroderma research across Australia
- Support state associations to provide successful and sustainable outcomes.

## Scleroderma Australia's services include:

- Centralised message bank, with calls returned promptly – 02 9990 5159
- Website with news and valuable resources, www.sclerodermaaustralia.com.au
- Links to state-based associations and their support groups
- eNews sign-up with all the current news, events and research

Scleroderma Australia is here to support you, and we welcome you to reach out to us for local and personal support such as events and support groups. You can find more information on the 'Get Support' page on the Scleroderma Australia website.

We encourage you to join your state association as a financial member to help us not only in our advocacy efforts, but to assist with the operational costs of each organisation. Our commitment is to ensure that your donations go to research, community engagement and awareness. We would sincerely appreciate your generous support to keep us going!

Scleroderma Australia supports local incorporated state associations in New South Wales, Victoria and Queensland.

**Scleroderma Australia**

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# 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 conditions 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, standing 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 the 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 and 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 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 tube connecting the mouth and the stomach. When functioning properly, 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 and 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 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.

***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 autoimmune 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.

# Websites

## Australia

### Scleroderma Australia

www.sclerodermaaustralia.com.au

The national not-for-profit organisation in Australia representing and advocating for people with scleroderma. Includes links to state-based scleroderma organisations.

### Australian Scleroderma Interest Group (ASIG)

https://asigresearch.com.au/

A group of doctors who are dedicated to scleroderma research. Project include the Australian Scleroderma Cohort Study.

### Pulmonary Hypertension Association Australia

www.phaaustralia.com

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

### Lung Foundation Australia

www.lungfoundation.com.au

A national charity and leading peak body that funds life-changing research and delivers support services that give hope to people living with lung disease or lung cancer.

## New Zealand

### Scleroderma New Zealand Inc.

http://scleroderma.org.nz/

## USA & Canada

### Scleroderma Foundation

www.scleroderma.org

The national non-profit organisation in the U.S. representing and advocating for people with scleroderma.

### 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

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

### 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 condition 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.

# State-based groups

## New South Wales

### The Scleroderma Association of New South Wales Inc.

PO Box 227, Ashfield NSW
Tel: 02 9798 7351
Email: sclerodermansw@tpg.com.au
Web: www.sclerodermansw.org

### The Autoimmune Resource and Research Centre

2nd Floor
Hunter Area Pathology Building
John Hunter Hospital Campus
Lookout Road
New Lambton Heights NSW 2305
Tel: 02 4921 4095
Email: HNELHD-arrc@hnehealth.nsw.gov.au
Web: www.autoimmune.org.au

## Northern Territory

Arthritis and Osteoporosis NT
PO Box 452, Nightcliff NT 0814
Email: info@aont.org.au
Alternatively, contact Scleroderma Queensland.

## Queensland

The Scleroderma Association of Queensland Inc.
54 Avocado Lane, Maleny QLD 4552
Phone: 0468 801 021
Email: info@scleroderma.org.au
Web: www.scleroderma.org.au

## South Australia

### The Arthritis Foundation of South Australia

111A Welland Avenue, Welland SA 5007
Tel: 08 8379 5707
or Country 1800 011 041
Email: info@arthritissa.org.au
Web: www.arthritissa.org.au
Alternatively, contact Scleroderma Victoria Inc.

## Tasmania

Contact Scleroderma Victoria Inc.

## Victoria

### Scleroderma Victoria Inc.

St. Vincents Hospital
41 Victoria Parade, Fitzroy VIC 3065
PO Box 57, Melton VIC 3337
Tel: 03 9231 3651
Email: enq@sclerodermavictoria.com.au

## Western Australia

Contact Scleroderma Victoria Inc.

## Australian Capital Territory

Contact Scleroderma NSW

# Notes

# Notes

PHARMACEUTICAL COMPANIES OF

Johnson & Johnson

scleroderma australia

Scleroderma Australia

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**w** www.sclerodermaaustralia.com.au