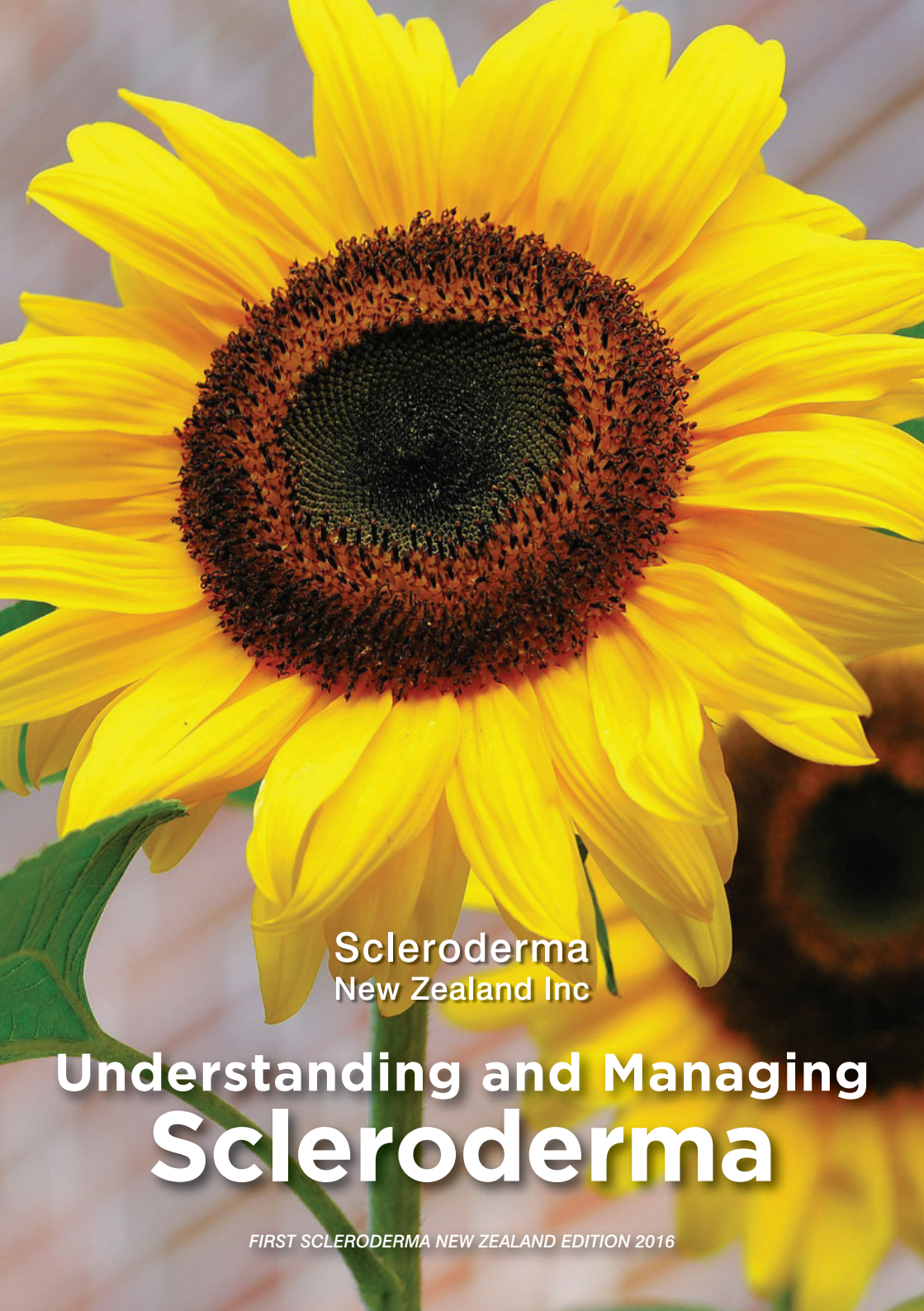
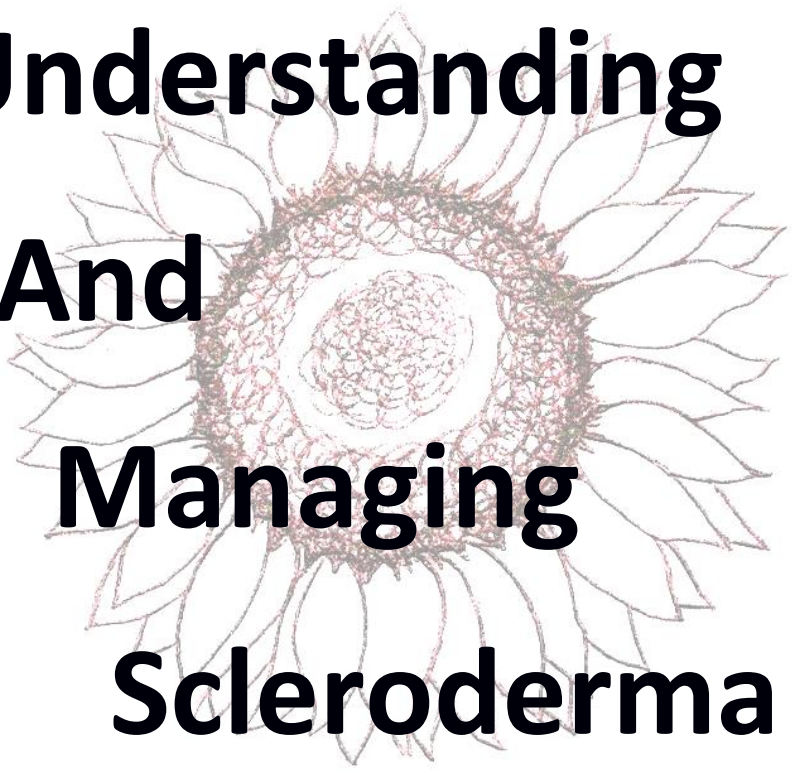


Scleroderma
New Zealand Inc

Understanding and Managing Scleroderma

FIRST SCLERODERMA NEW ZEALAND EDITION 2016

Understanding And Managing Scleroderma



First Scleroderma New Zealand Edition 2016

Acknowledgement

An acknowledgement of gratitude to all of the Scleroderma New Zealand members and their families who have worked hard fundraising for the printing of this publication.

Understanding and Managing Scleroderma

This booklet is intended to help people with scleroderma, their families and others interested in learning more about the disease 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 most frequently asked questions about scleroderma.

Disclaimer

The Scleroderma Foundation and Scleroderma New Zealand do not provide medical advice nor do they endorse any drug or treatment mentioned herein. The material contained in this booklet is presented for general information only. It is not intended to provide medical advice, to answer questions specific to the condition or problems of particular individuals, nor in any way to substitute for the professional advice and care of qualified doctors and nurses. Mention of particular drugs and/or treatments is for information purposes only and does not constitute an endorsement of said drugs and/or treatments.

Thanks!

Scleroderma New Zealand wishes to acknowledge and thank the Scleroderma Foundation in the United States for permission to use their booklet for New Zealand.

The Scleroderma Foundation and Scleroderma New Zealand expresses their deep appreciation to the many physicians whose efforts have led to this booklet.

Special thanks are owed to Maureen D. Mayes, M.D., M.P.H., of the University of Texas Medical School, Health Science Center at Houston; and Khanh T. Ho, M.D., Baton Rouge, La.

Contents

What is scleroderma?

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

The word “scleroderma” comes from the 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 not usually hereditary.

How serious is scleroderma?

Any chronic disease can be serious. The symptoms of scleroderma vary greatly from individual to individual, and the effects of scleroderma can range from very mild to life-threatening. The seriousness will depend on what parts of the body are affected and the extent to which they are affected. A mild case can become more serious if not treated properly. 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 approximately 1,000 persons with scleroderma in New Zealand. Scleroderma has been reported worldwide.

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 25 and 55.

Factors other than sex, such as ethnicity, 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. This suggests that there are some genes that can predispose toward getting scleroderma. In addition, some genes may influence the type and severity of this disease.

What causes scleroderma?

The cause or causes of scleroderma are being studied. Studies have found that both solvent and silica exposure are associated with scleroderma.

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

✦ an overproduction of collagen.

✦ an autoimmune process.

✦ blood vessel damage.

Collagen is the major protein portion of the connective tissue of the body, which includes the skin, joints, tendons and parts of internal organs. Collagen is made up of tiny fibres, which weave together much like the threads forming a piece of cloth. When there is an overproduction of collagen,

thickening and hardening of the affected areas takes place, often interfering with the normal functioning of those parts.

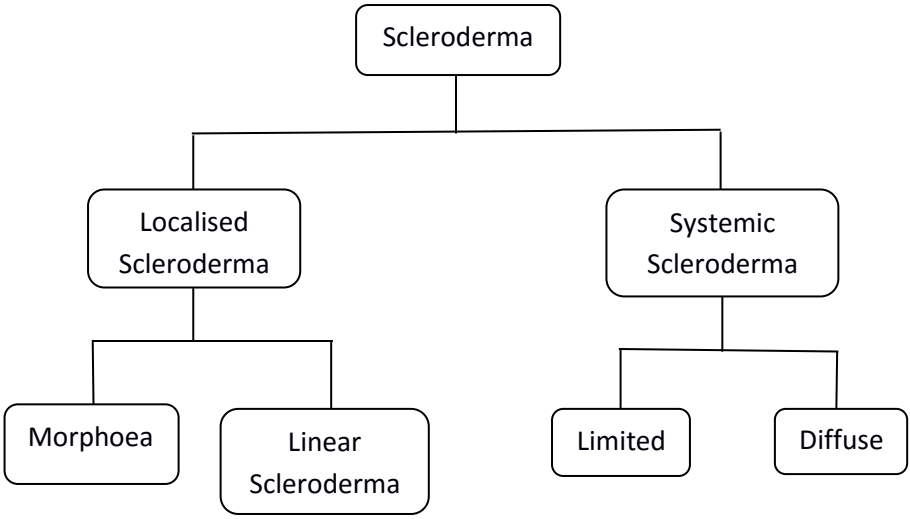
There are several theories about how collagen is overproduced. The "autoimmune theory" suggests that the body's own immune system plays a part. Normally, the body's immune system produces chemical signals in the blood called cytokines, which coordinate the body's defence against bacteria, viruses and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

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 blood vessels, the processes which take place, and their significance for prevention and treatment.

Research is being done to study these and other theories. It is hoped that a better understanding of what causes scleroderma will lead to better treatment methods and, ultimately, to a cure.

Are there different forms of scleroderma?

There are two major classifications of scleroderma: **localised scleroderma** and **systemic sclerosis** (SSc). Other forms or subclassifications, each with its own characteristics and prognosis, may be identified through future research.



Localised scleroderma

The changes, which occur in localised scleroderma, are usually found in only a few places on the skin or muscles, and rarely spread elsewhere. Generally, localised scleroderma is relatively mild. The internal organs are usually not affected, and persons with localised scleroderma rarely develop systemic scleroderma. Some laboratory abnormalities commonly seen in systemic scleroderma are frequently absent in the localised form.

Morphoea is a form of localised scleroderma characterised by waxy patches on the skin of varying sizes, shapes and colour. The skin under the patches may thicken. The patches may enlarge or shrink, and often may disappear spontaneously. Morphoea usually appears between the ages of 20 and 50, but is often seen in young children.

Linear scleroderma 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 it resembles a sabre or sword wound. Linear scleroderma tends to involve deeper layers of the skin as well as the surface layers, and sometimes affects the motion of the joints which lie underneath.

Linear scleroderma usually develops in childhood. In children, the growth of involved limbs 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—**diffuse** or **limited** disease.

Diffuse scleroderma. In **diffuse scleroderma**, skin thickening occurs more rapidly and involves more skin areas than in limited disease. In addition, people with diffuse scleroderma have a higher risk of developing "sclerosis" or fibrous hardening of the internal organs.

Limited scleroderma — CREST syndrome. About 50 percent of patients have a slower and more benign illness called limited scleroderma. In **limited scleroderma**, skin thickening is less widespread, typically confined to the fingers, hands and face, and develops slowly over years. Although internal problems occur, they are less frequent and tend to be less severe than in diffuse scleroderma, and are usually delayed in onset for several years. However, persons with limited scleroderma, and occasionally those with diffuse scleroderma, can develop pulmonary hypertension, a condition in which the lung's blood vessels become narrow, leading to impaired blood flow through the lungs resulting in shortness of breath.

Limited scleroderma is sometimes called **CREST syndrome**. CREST stands for the initial letters of five common features:

- **C**alcinosis
- **R**aynaud's Phenomenon
- **O**esophageal dysfunction
- **S**clerodactyly
- **T**elangiectasia

To further complicate the terminology, some people with diffuse disease will go on to develop calcinosis and telangiectasias so that they also have the features of CREST.

Although most patients can be classified as having either diffuse or limited disease, different people may have different symptoms and different combination 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 and may overlap with those of other diseases, especially other autoimmune connective-tissue diseases such as rheumatoid arthritis and lupus. Different symptoms may develop in stages over a very long period of time, and few people with scleroderma experience exactly the same set of symptoms and effects.

While scleroderma can often be suspected from its more visible symptoms, no single test can prove its presence. Doctors with extensive experience in the treatment of scleroderma usually make a diagnosis by a combination of the following: a patient's 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.

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 frequently associated with diffuse scleroderma whereas **anti-centromere** is usually indicative of limited scleroderma.

	Diffuse patients	**Limited patients**
Anti-nuclear antibody	90 percent test positive	90 percent test positive
Anti-centromere antibody	3 percent test positive	41 percent test positive
Anti-Scl-70 antibody	29 percent test positive	16 percent test positive

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 number 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 every symptom or all of the methods being used in scleroderma management. 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 has no cure, 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 percent 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 emotional upset and stress. The resulting disturbance in blood circulation 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 part 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. Some people find Raynaud's attacks painful.

Raynaud's-like cold-induced constriction and narrowing of blood vessels may occur in the lungs, kidneys, gastrointestinal, and heart circulations in people with systemic scleroderma.

Many commonsense preventive measures can be taken by those susceptible to Raynaud's Phenomenon. Most obvious is minimising exposure to cold, such as outdoor weather, air conditioning, or reaching into a refrigerator or freezer. Keeping your extremities and body warm is very important. Gloves or mittens should be worn, and a number of warming devices are available to protect the hands. Hats, ear muffs, heavy socks and warm, layered clothing of

made from silk, cotton, wool and down feathers can help maintain body temperature. It is important to protect your hands with gloves when touching refrigerated or frozen items. Electric heaters, electric blankets and comforters can supplement the heat in the home or apartment. Keeping the entire body warm helps prevent peripheral Raynaud's episodes as well as internal Raynaud's. A warm bath or shower, or 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 can help but isn't always possible.

Various relaxation techniques, whether self-taught or learned through training courses, prove effective for some people to manage stress. One particular technique, biofeedback, has been used to increase finger temperature.

Smoking definitely worsens Raynaud's Phenomenon. For this and other reasons, persons with scleroderma should not smoke.

When Raynaud's Phenomenon does occur, carefully waving the arms in an underhand, circular motion (like a softball pitcher) can help to restore blood circulation. Rubbing or massaging the hands and feet may also help.

Doctors use 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. Drugs commonly used include vasodilators or calcium channel blockers. Mild blood thinners such as aspirin, or drugs that decrease the stickiness of platelets, may improve circulation. There are now more than a dozen medications to improve circulation, and it is not possible to list them here.

Raynaud's Phenomenon is not confined to people with scleroderma. It is also seen in people with 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.

Swelling or puffiness of the hands

Swelling is another typical early symptom of scleroderma, and this may be especially noticeable upon waking up in the morning due to muscle inactivity at night. 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 can help. 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 which frequently occurs in scleroderma. Muscle pain and weakness are other important symptoms. Aspirin or aspirin-like medications (nonsteroidal, anti-inflammatory prescription drugs) can help.

Be sure to discuss these medications with your doctor if you have digestive problems because they may irritate the stomach. Other treatments and suggestions are included in the sections on “Physical Therapy and Exercise” and “Protecting the Joints.” A physical therapist can develop an exercise plan after consulting with your doctor.

Skin disorders

Skin thickening. Hardening and thickening of the skin gives scleroderma its name (“hard skin.”) There are no proven treatments as yet to prevent or alter the course of the skin changes in scleroderma. Many medications and treatments are being tested. The skin sometimes softens spontaneously over time.

Skin ulcerations. Sores, especially on the fingertips and knuckles, 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 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 carefully cleaned to avoid infection. If an infection develops, it may help to soak the affected area in warm water, apply an antiseptic or use an antibiotic ointment. Should these remedies prove unsuccessful in relieving the pain or infection of ulcerated skin, your doctor may prescribe oral antibiotics or take other measures.

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 make-up may be used to mask the spots or to reduce their visibility.

Dry skin. Excessive dryness of the skin may lead to skin breakdown and ulcerations. Excessive bathing and handwashing 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 are preferable to harsh soaps which dry out the skin. Frequent use of moisturising skin creams containing lanolin is advised. During the winter months, a humidifier may help.

Itchy skin. If moisturising creams do not work, your doctor may prescribe a topical cortisone cream to rub on the skin to relieve itching. Antihistamines 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 sweat. In addition, there may be an increase in pigment (which looks like a skin tan) or a spotty loss of pigment.

Sclerodactyly and joint contractures

Sclerodactyly means “hard skin of the digits,” particularly the fingers and toes. It generally occurs after initial swelling has subsided. It is characterised by shiny, tight skin of the fingers.

Affected digits 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 tissues surrounding the joints can cause decreased motion of the wrists, elbows and other joints.

“Range of motion” exercises performed daily are important to prevent or slow down the development of such contractures and to maintain limber joints. 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.

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 allow 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 narrowing (stricture) of the tube.

Acid production can be reduced, and the problems of acid reflux and heartburn helped, by avoiding alcohol, greasy or fatty foods, spicy foods, chocolate, tobacco and caffeine. Antacids (particularly in liquid form) can help neutralise acids and reduce heartburn. Some antacids cause constipation while others cause diarrhoea. Consult your doctor or pharmacist when choosing over-the-counter products. Your doctor may prescribe antacid medications such as proton pump inhibitors or H-2 blockers to decrease acid production in the stomach. Your doctor also may prescribe a drug which promotes muscular activity and causes the oesophagus to work better.

The force of gravity helps to keep food and acid in the stomach; therefore, an upright position after meals is helpful. Other commonsense 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 fifteen to twenty cm with wooden blocks. Being overweight is harmful, and you should avoid wearing girdles or other tightfitting garments.

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, the doctor may need to dilate the oesophagus periodically to permit easier swallowing.

Diarrhoea. 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 diarrhoea. There also may be a bloated, distended feeling and some pain if the bowel is stretched. Another effect is that the nutrients of food remain in the bowel instead of being absorbed into the body. This condition is called malabsorption, and it may lead to weight loss and stool abnormalities.

For diarrhoea or malabsorption, your doctor may prescribe an antibiotic, or supplementary fat-soluble vitamins, and/or iron. Your doctor also may suggest that you reduce the amount of fatty foods in your diet and increase your carbohydrate intake.

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 prevent constipation. Fresh fruits and vegetables are natural laxatives. Exercise also helps to keep bowel movements regular. Your doctor also may recommend stool softeners and bulking agents.

Sjögren's Syndrome

Sjögren's Syndrome (dry eyes or 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 speaking, a pronounced increase in tooth decay and cavities, and a reduced sense of taste. The lack of secretions in Sjögren's Syndrome also may 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 an ophthalmologist are important. The mouth should be kept as well-lubricated as possible by sipping fluids during the day (a plastic squirt bottle filled with water may help), 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.)

Your doctor may prescribe a propionic acid gel preparation or vaginal cream to lubricate the vagina and facilitate sexual relations. Avoiding pantyhose and other tightfitting 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.

Preventive dental care including regular flossing and brushing of the teeth and gums is very important, as are regular dental visits for oral health and for the early detection and prompt correction of any abnormalities. The dentist also can recommend a good oral hygiene programme. 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 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 quickly. The most important warning sign is a sudden rise in blood pressure. Other symptoms are 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 to control blood pressure and stabilise or improve 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

Multiple factors can cause lung involvement in systemic scleroderma. 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. Pulmonary arterial hypertension (PAH), a state of increased resistance to blood flow through the lungs, can result from damage to blood vessels, and may lead to additional strain on the heart resulting in heart failure. Respiratory muscle weakness may decrease lung function.

Symptoms of lung involvement include shortness of breath, a decreased tolerance for exercise and a persistent cough. Your doctor may order a chest X-ray, an echocardiogram (ultrasound of the heart), special breathing tests (pulmonary function tests) or a CT scan of the lungs to detect or confirm lung involvement.

In the early stages of lung fibrosis, medications may be given to decrease the inflammation which is thought to lead to lung scarring.

Although many investigations are under way, currently there are no proven medications to reverse lung changes once they have occurred. It is important, therefore, for the person with scleroderma to take whatever measures are within their control to avoid further damage to their 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 the extent possible. Your doctor may recommend medications to make breathing easier and may also suggest deep breathing exercises and a graduated aerobic exercise programme.

People with pulmonary arterial hypertension may 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; there are now three different classes of medications which can be used.

Some doctors are increasingly recommending regular screening for lung involvement with at least annual pulmonary function tests and echocardiogram even in people without symptoms of lung problems.

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 occur. These conditions require careful evaluation and treatment by the doctor.

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 a doctor will depend on their evaluation of the causes of these symptoms.

Managing scleroderma

You may know other forms of treatment that have been used or are proposed for use to manage 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, doctors must often make treatment decisions based on incomplete information. They 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 alone 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 for each person. Systemic scleroderma is a chronic, lifelong disease. Currently, there is no known cure, but as with other chronic diseases there are many ways to control or manage its symptoms. It is helpful to keep scleroderma in perspective. Many persons with the disease have few or minimal symptoms and are able to lead a normal or nearly-normal life.

There may be periods of time when the person with scleroderma will be free of troubling symptoms and feel well. At other times, they 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, also may occur and 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 living 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 a prompt start of treatment. Some of the more promising medications in current use are slow-acting and the sooner treatment begins, the better the results may be. If one has already been diagnosed with scleroderma, it is especially important to watch for and report to the doctor and/or rheumatology nurse 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.

A doctor can help to distinguish what is related to scleroderma and what is not, and recommend appropriate treatment.

Developing an individual treatment programme

While there is no proven cure for scleroderma, much can be done to prevent, minimise or alleviate its effects and symptoms. Scleroderma symptoms 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 that a doctor experienced in scleroderma management works out an individually-tailored programme to meet the specific needs of a person with this disease. Close cooperation with the doctor will help them develop such a programme.

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 subheads will discuss other important elements of a programme for managing scleroderma.

Physical therapy and exercise

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

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

Protecting the joints

Joint protection helps minimise further damage and to reduce the possibility of skin ulcers and infection. Its basic principles include avoiding or minimising pressure or stress on the joints by 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 alleviate stress on the joints while still completing daily activities. Occupational therapists can demonstrate such devices and give further instruction on joint protection.

Taking medications

It is essential that a person with scleroderma take all medications 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 their doctor. The person with scleroderma should advise the doctor of any drugs taken for other conditions including over-the-counter preparations, herbal supplements or vitamins. Any side effects encountered should be promptly reported and discussed with their doctor.

One should not be concerned if the doctor prescribes different medications for different people. Scleroderma symptoms vary from person to person, requiring different treatments. Some may benefit from certain drugs, while others may not. Furthermore, individual tolerance for the drugs used in scleroderma varies greatly. The doctor may find it necessary to adjust the medication programme accordingly.

Commonsense measures

We have discussed treatment options for specific symptoms in other sections of this booklet. There also are a number of general commonsense measures that a person with scleroderma can take to enhance their well-being. These measures include:

✦ Keep warm, particularly keep your core warm. Keep your living environment warm. If you are going out and it is cold wear gloves, three layers of warm clothing and windproof clothing if needed.

✦ 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 good hygiene habits, especially of the skin, teeth, gums and feet (including the wearing of cushioned and well-fitted shoes).

✦ 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.

✦Although you cannot be certain what caused your scleroderma, a commonsense precaution is to void being exposed to substances that have been linked to scleroderma, such as solvents. Solvents are common in our environments, both at work and at home. You should check the products you intend to use.

The emotional aspects of scleroderma

A person newly diagnosed with scleroderma may feel alone and uncertain about where to turn for help. They may experience a number of other feelings and emotional reactions from time to time, including initial shock or disbelief, fear, anger, denial, self-blame, 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 help. Professional counselling also can 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 health care 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 also 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 programme, and assist in carrying out activities that they find difficult.

The health team begins with the doctor, but can include many other health professionals such as other medical specialists, nurses, physical 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 benefit a person with scleroderma.

Joining a scleroderma support group, such as Scleroderma New Zealand, enables the person with scleroderma to meet and exchange information with others who have similar problems, as well as to learn more about scleroderma. The US based Scleroderma Foundation manages an online

support group community at http://www.inspire.com/groups/scleroderma-foundation.

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 a person with scleroderma?

Emphatically, yes!

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

Investigators throughout the world are intensifying their efforts to understand the nature and discover the causes 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 improved methods of measurement to evaluate disease progression and the results of treatment. Various animal models of scleroderma have been developed.

Investigators currently are 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!

Scleroderma New Zealand

Scleroderma New Zealand is a national society representing and advocating for persons with scleroderma. Scleroderma New Zealand is an incorporated society and a registered charity. The purposes of the Society are support, friendship, education, awareness, advocacy, lobbying, prevention, research and assistance with health care for people, their families and communities with Scleroderma (Systematic Sclerosis) and Undifferentiated Connective Tissue Disease.

Scleroderma New Zealand's website is scleroderma.org.nz

The website has our quarterly newsletter and links to many informative leaflets and websites

Meetings and educational seminars with health experts are held at various localities around New Zealand, see our website for location and contact details for meetings.

There is a phone support network for those who would like personal contact especially useful for people in isolated communities.

Scleroderma New Zealand can put you in touch with a support group near you. If there is no support group nearby, Scleroderma New Zealand can help you start one.

Getting help

Healthline

Call Healthline on 0800 611 116 for free advice from trained registered nurses. They are specialists in assessing and advising over the phone. Phone calls are free from within New Zealand. Healthline which is provided by the Ministry of Health is available 24 hours a day, 7 days a week.

Call Healthline if you are:

feeling unwell – but not sure whether you need to see a doctor

needing some urgent advice about a family member or friend who is sick

on holiday and want to know where the nearest doctor or pharmacy is.

If you need to talk to someone in your own language, Healthline can usually arrange this using an interpreting service.

Care Plus

Care Plus is a primary health care funding initiative to support people with high health needs due to chronic conditions, acute medical or mental health needs, or terminal illness. This is administered by the Ministry of Health. Ask your GP if you are eligible.

Hand Therapists

You might require some help with your hands; it may be splints, special exercises, helpful aids for around home and work, and wound prevention. You can get help through a referral from your GP or rheumatologists to a hand therapist. Or you can go privately.

Find one in your area www.nzaht.org.nz/find-a-therapist-2.php

Podiatrists

You may require some help with your feet. You can get help through a referral from your GP or rheumatologists to a hand therapist. Or you can go privately.

Find one in your area
www.podiatry.org.nz/Members/Search/BusinessSearch.aspx

Mobility Parking Permits

You may be eligible for a mobility car parking permit if:

You are unable to walk and always require the use of a wheelchair, or

Your ability to walk distances is severely restricted by a medical condition or disability. If for example, you require the use of mobility aids, experience severe pain, or breathlessness, or

You have a medical condition or disability that requires you to have physical contact or close supervision to safely get around and cannot be left unattended. For example, if you experience disorientation, confusion, or severe anxiety.

Your doctor needs to confirm your eligibility, unless you have a long-term permit.

www.mobilityparking.org.nz

Ministry of Health Home Care Support

The Ministry of Health may be able to help you with support in your own home, with a range of services available if you meet the eligibility criteria, see the links below.

Home and Community Support Services

Home and Community Support Services are services funded by the Ministry to help you live at home. They can help with both household management and personal care.

Household management may include help with:

- meal preparation
- washing, drying or folding clothes

house-cleaning, vacuuming and tidying up.

Personal care may include help with:

eating or drinking

getting dressed or undressed

getting up in the morning or getting ready for bed

showering or going to the toilet

getting around your home.

See web site www.health.govt.nz/your-health/services-and-support/disability-services/types-disability-support/home-and-community-support-services

Community Residential Support Services

Community Residential Support Services are one of a range of support services funded by the Ministry of Health. These services assist disabled people to live in a supported community environment.

See web site www.health.govt.nz/your-health/services-and-support/disability-services/types-disability-support/community-residential-support-services

Equipment and Modification Services

Do you have difficulty doing everyday activities? If you or someone in your family has a physical, intellectual, sensory (vision or hearing) or age-related disability, you may be able to get some equipment or modifications to your home or vehicle to assist you.

See web site www.health.govt.nz/your-health/services-and-support/disability-services/types-disability-support/equipment-and-modification-services

Individualised Funding

Individualised Funding is a way of paying for Home and Community Support Services which lets you directly manage the resources you've been allocated for disability supports.

See web site www.health.govt.nz/your-health/services-and-support/disability-services/types-disability-support/individualised-funding

Work and Income

Scleroderma may make it very difficult for you to work due to a limitation on your physical functioning, mobility, dexterity or stamina.

Work and income may be able to support you while you're not able to work and help you realise your work goals. When it's right for you and your family, they can help you find part-time or full-time work, get training, or become self-employed.

See overview of available services www.workandincome.govt.nz/individuals/how-we-can-help-you/disabled-or-ill/index.html

Help with living expenses

Work and Income may be able to help you with living costs if you're not able to work, or are working fewer hours because:

- you have a health condition, injury or disability
- you're caring for someone with a health condition or a disability

Community Services Card: A card that can help you and your family with the costs of health care. You'll pay less on some health services and prescriptions. See www.workandincome.govt.nz/products/a-z-benefits/community-services-card.html

House Modification Funding: Funding for changes to your home because you or your child is disabled. See www.workandincome.govt.nz/eligibility/health-and-disability/house-modification-funding.html

Residential Support Subsidy: A subsidy for residential care needed because of a physical, intellectual, or psychiatric disability. See www.workandincome.govt.nz/eligibility/health-and-disability/residential-care.html

Rheumatologists

You may be looking for a Rheumatologist. On the New Zealand Rheumatology Association website you will find a list of Rheumatologists in your region. www.rheumatology.org.nz/home/member_list

Once you have decided on a Rheumatologist you will need to be referred to the Rheumatologist by your GP. Many of these Rheumatologists practice in the public hospital and practice privately. When seeing a Rheumatologist it is good to go along with all your concerns written down.

Having a support person is also a good idea, at least for your first few visits. Your support person can help take notes and help remind you of concerns if need be.

Shop on line for aids

Go online for help in the kitchen, bathroom, bedroom, home living, mobility, and sensory, everything you can imagine:

Mobility Centre www.mobilitycentre.co.nz/shop-daily-living-products-online.html

Independent Living www.ilsnz.org/Products

Mental Health

Having Scleroderma, a chronic disease, can cause depression and anxiety.

See the web site www.health.govt.nz/your-health/conditions-and-treatments/mental-health for getting help with these conditions. If you prefer just to talk to a fellow Scleroderma sufferer, call Dianne Purdie, Scleroderma NZ President, on 04 479 5548

Green Prescription New Zealand.

Physical activity is very important for Scleroderma patients. In New Zealand you can be issued a green prescription to help you obtain your goals.

Here is how it works.

The health professional (GP or practice nurse) issues you with a Green Prescription (GRx), provided your condition is stable.

The script is either written or issued electronically. If you want ongoing support, the script is forwarded through to the nearest GRx Patient Support Person.

The Patient Support Person encourages you to become more active through:

monthly telephone calls for 3-4 months or;

face to face meetings for 3-4 months or;

group support in a community setting for 3-6 months.

Your progress on your path to an active lifestyle is reported back to your referring health professional.

If you feel you would benefit from ongoing support, you are encouraged to ask your health professional for another GRx.

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: aspirin, paracetamol and non-steroidal anti-inflammatory drugs.

Antacid. An agent which neutralises excess stomach acid. This may be liquid/tablets which act immediately in the stomach, or long-acting medications taken regularly and absorbed into the blood in order to suppress acid production. (See **Acid reflux**.)

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.

Autoimmune. Disease or antibody which acts against the person'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 practice relaxation techniques and learn to control the function. Later, the machine becomes unnecessary. (See **Relaxation techniques**.)

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

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

Calcinosis. Abnormal accumulation of calcium in the skin.

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

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

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

Constrict (vessels)**, stricture** (oesophagus)**.** An abnormal narrowing.

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

Coronary arteries. Blood vessels which supply blood to the heart.

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 pills."(See **Oedema**.)

Dysfunction. Impaired or abnormal functioning.

Dysphagia. Difficulty in swallowing.

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

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

Fibrous. Consisting of, or resembling fibres.

Fibrosis. Abnormal formation of excess fibrous tissue.

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

GP. General Practitioner. They diagnose and treat health problems of individuals and families in the community.

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 autoimmunity.

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**.)

Morphoea. A form of localised scleroderma.

Motility, dysmotility. Contractions of the digestive-tract muscles occurring in rhythmic waves, propelling food, allowing absorption of nutrients, and

elimination of wastes (faeces). Dysmotility indicates weakened or absent waves of contraction resulting in abnormally slow movement of food and faeces. (See **Malabsorption, Gastrointestinal tract, Contraction**.)

Occupational therapy. 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 **Physical therapy**.)

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.

Physical therapy. 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. (Abbreviated "PT." 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 "Raynode") 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.")

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

There are more sources of good and reliable information about scleroderma.

Websites

There are links to many informative web sites on the Scleroderma NZ web site

scleroderma.org.nz/links/

And links to many useful leaflets

scleroderma.org.nz/news/useful-leaflets/

Recommended Reading

The Scleroderma Book: A Guide for Patients and Families

Revised and Updated Edition. 2005. Oxford University Press.

By Maureen Mayes, M.D., M.P.H.

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

Scleroderma: From Pathogenesis to Comprehensive Management

Editors: John Varga, Christopher P. Denton and Fredrick M. Wigley.

Springer 2012

A clear and concise synthesis of current concepts in pathogenesis and modern approaches to management, this book is comprised of the authoritative work of international experts. With an integrated multidisciplinary approach to comprehensive care, this book is easily accessible for health care professionals in many fields. It is a valuable resource for rheumatologists, pulmonologists, cardiologists, gastroenterologists, nephrologists and all those involved in the care of scleroderma patients.

Contacting Scleroderma New Zealand Inc

Contacts for Scleroderma New Zealand are on the web site scleroderma.org.nz

Scleroderma
New Zealand Inc
www.scleroderma.org.nz