Scleroderma Canada | Sclérodermie Canada

EDUCATIONAL HANDBOOK

This booklet contains key information, tips and guidelines to help you better understand scleroderma and its effects.

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WHAT IS SCLERODERMA?

WHAT IS SCLERODERMA?

Literally, the term scleroderma means “hard skin”. It is a progressive disease of the vascular and immune systems and a severe connective tissue disorder. In scleroderma, some unknown factor triggers the over production of collagen (body protein), causing thickening, hardening and scarring of the skin and other organs. This often affects the small blood vessels that carry blood to many parts of the body.

Scleroderma is also known as systemic sclerosis.

TYPES OF SCLERODERMA

Localized Scleroderma
- Morphea
- Linear Scleroderma

Systemic Scleroderma
- Limited*
- Diffuse
- Sine

LOCALIZED SCLERODERMA

This form of scleroderma affects some areas of the skin, but does not affect the internal organs.

Morphea is the most common form of localized scleroderma and is characterized by oval patches of inflamed, often discoloured skin. The trunk, face, and extremities may be involved.

In linear scleroderma, a band or bands of skin hardens or thickens on the trunk and/or extremities.

SYSTEMIC SCLERODERMA

This form of scleroderma involves not only the skin but also internal organs, most commonly the digestive, circulatory, pulmonary, and muscular systems. Systemic scleroderma is divided into three forms: limited, diffuse and sine.

Sine may resemble either limited or diffuse systemic sclerosis causing damage in lungs, kidneys and/or blood vessels. However, unlike other forms of systemic sclerosis, the skin in sine is not usually involved.

*Limited scleroderma is often referred to as CREST. This is an acronym that stands for a combination of symptoms:

C – Calcinosis – small white calcium lumps forming under the skin

R – Raynaud’s Phenomenon – poor circulation in the fingers and/or toes. Small blood vessels in the fingers tend to narrow and decrease blood flow causing patients to be unduly sensitive to cool temperatures. This narrowing can also be a response to stress or emotion. Fingers often turn white, then blue

E – Esophageal dysfunction - difficulty swallowing, heartburn, or regurgitation

S – Sclerodactyly – skin of fingers and sometimes toes become thick and shiny. Affected digits may be difficult to move and may become fixed in a bent position

T – Telangiectasia – small clusters of dilated blood vessels in the skin especially on the face and fingers and palms of hands

The CREST syndrome commonly manifests itself slowly over a period of ten to twenty years. Usually it involves the skin first, then the esophagus, lungs, and bowels.

Diffuse scleroderma is the most serious internally involved form of the disease. It involves many of the internal organs of the body: esophagus, the digestive tract, kidney, heart, and/or lungs. The skin involvement includes the face, neck, torso, and both hands, arms, feet and legs.

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SYMPTOMS

Scleroderma is highly individualized, so it affects patients in different ways. Symptoms and severity of the symptoms differ greatly.

SKIN:

The most commonly identified symptom of scleroderma is the gradual thickening and tightening of the skin. Ulcers, especially on the fingertips, are common. They can be slow to heal because of poor circulation.

MUSCLE WEAKNESS:

Muscles weaken and can become painful.

DIGESTIVE SYSTEM AND GASTROINTESTINAL TRACT:

Swallowing difficulties can result from the narrowing of the esophagus. Digestive difficulties range from poor absorption of nutrients to slow movement of food. The weakened muscles of the gastrointestinal tract can lead to a "backwash of stomach acid."

DENTAL:

Because of tightening of facial skin, dental health may be compromised. Dry mouth can cause difficulties in swallowing, which in turn may lead to an increase in tooth decay.

KIDNEY:

Early signs of kidney damage may include high blood pressure and an excess of protein in the urine (detected by a urine test). Renal crisis is a severe complication of scleroderma and unless treated promptly may lead to kidney failure.

RAYNAUD'S PHENOMENON:

This is a very common symptom and is generally believed that 98% of scleroderma patients have Raynaud's Phenomenon. (See the information on CREST).

JOINTS:

Joints can become stiff and sore, similar in many respects to arthritis.

SJOGREN'S SYNDROME:

Sjogren's Syndrome is dry eyes and mouth due to a decrease in secretions of the tear ducts and salivary glands.

LUNGS:

A build up of fibrosis (scarring) in the lungs and/or weakened respiratory muscles leads to shortness of breath and persistent coughing. The fibrosis affects oxygen absorption and may lead to Pulmonary Arterial Hypertension.

HEART:

The muscles that surround the heart may become thickened and scarred, decreasing heart contractions. This can cause chest pains and irregular heartbeats.

NON–SPECIFIC SYMPTOMS:

These include extreme fatigue, general weakness, weight loss, hair loss and vague aches in muscles, bones and joints.

* Report any change in symptoms or new systems to your doctor.

CAUSES

While the cause of scleroderma is not known, there are several theories being studied which involve different systems of the body: the immune system, the vascular system, and the connective tissues. Some research indicates an infectious component with a virus like organism acting as a trigger. It is generally believed that scleroderma is neither contagious nor inherited. There has been some indication of a genetic predisposition to the disease with an environmental trigger.

WHO DEVELOPS SCLERODERMA?

Although scleroderma strikes every age, sex, and ethnic background, more than 80% of patients are women between the ages of 30 to 50. There is no authoritative incidence rate of scleroderma available, but the most conservative estimate is 9,000 cases and responsible estimates are as high as 40,000 cases.

TREATMENT

Although there is no known cure for scleroderma, symptoms can be moderated with medication and lifestyle changes. Some medications are aimed at specific symptoms, while others are aimed at decreasing the activity of the immune system. Because of the advances in treatment, patient survival has improved a great deal over the past years.

We would like to thank **Dr. Margaret Larché, MBChB, MRCP(UK), PhD** for her assistance with this information pamphlet.

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MANAGING SCLERODERMA

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You have just been diagnosed with scleroderma. Your doctor says that it's a chronic disease that cannot be cured but can be managed. When you are first diagnosed, it is perfectly reasonable to experience a wide range of emotions – fear, sadness, anger. You will feel alone and uncertain. No matter the diagnosis, you can learn to manage.

GET PROFESSIONAL CARE

It is important to have professional care. A chronic condition usually involves contact with one or more medical specialists, but don't abandon your family doctor. That physician probably knows you best and can provide a valuable perspective on what the specialists say. Specialists offer facts, opinions and advice about treatment and products that can make your life easier. Cooperate with medical specialists and work with them to develop a schedule of tests to ensure that scleroderma related complications are diagnosed and treated as early as possible.

BE YOUR OWN ADVOCATE

Learn all you can about scleroderma.

> "Knowledge is power and ignorance is not bliss. You need to know what is happening to you and what could happen to you. The well informed patient may not always do well, but he or she will always do better than those who keep themselves in the dark".
>
> *- Lee Shapiro. MD, FACP, Scleroderma VOICE 2002 #4 pp 10-13.*

Inform yourself about scleroderma and existing treatments, research, and symptom management. It is important to understand your illness so that you are able to:

- Communicate effectively with physicians
- Identify available options
- Alleviate anxieties

It is useful to:

- Keep a notebook for visits with physicians
- Jot down questions, concerns and recent symptoms before visits with physicians
- Write down main points of the discussions
- Keep accurate medical records of:
  - Physicians and specialists
  - Medical conditions
  - Medications – doses and dates
  - Date and description of procedures and tests.

CONSIDER COMPLEMENTARY THERAPIES

Depending on your condition, some complementary therapies may be helpful. Massage therapy, relaxation therapy, and acupuncture are just a few possibilities. Both conventional and alternative (complementary) health practitioners should be made aware of all medical treatments.

LEARN TO CONTROL AND MINIMIZE STRESS

Stress and anxiety tend to exacerbate many symptoms, particularly pain. There are many paths to stress management: exercise, meditation, biofeedback, yoga, tai chi, prayer, and social support from family, friends or a support group. Choose the approaches that appeal to you. Ask your physician, social worker, family, friends or community organizations for referrals.

GET REGULAR, MODERATE EXERCISE AND THERAPY

Exercise is important for many reasons: it improves cardiovascular health, strength, stamina and flexibility, helps to control weight, improves the quality of sleep, and is a natural antidepressant. “Use it or lose it” does apply to scleroderma patients. Exercises may include range of motion, stretching, strengthening and conditioning and/or aerobic exercises. For most people, the easiest exercise program to adopt is walking. Exercises should be performed gently and with due care. It is always important to protect joints to prevent further pain and swelling. Consult your physician or physiotherapist about the types of exercise you can do.

FOLLOW A HEALTHY DIET

It is important for everyone to eat well-balanced meals. In the case of scleroderma patients, antioxidants are essential to help prevent cell damage and to support the immune system. When scleroderma impacts the digestive system, the following may be considered:

- Drink plenty of fluids
- Eat smaller more frequent meals
- Eat slowly and chew foods well
- Eat soft or pureed foods
- Decrease the intake of alcohol, carbonated soft drinks, chocolate and caffeine
- Avoid highly acidic foods
- Avoid fatty and greasy foods
- Sit upright for 1-2 hours after eating so gravity can help move food down
- Raise the head of your bed 4-6 inches (allows gravity to help the digestive process)
- Consider taking a multivitamin, especially if several foods are eliminated from a healthy diet
- Eat food with probiotics

If there are dietary concerns, contact your physician or a registered dietician.

RETHINK YOUR LIMITATIONS WHILE MAINTAINING A BALANCE OF ACTIVITIES

Scleroderma will cause limitations in your life. Changes may mean limitations in diet or activities. It’s perfectly normal to miss the things you can’t do anymore. Look for alternatives.

ADAPT TO CHANGE

Having scleroderma means that changes in lifestyle will be necessary. Adapting to this is important. You may have to treat pain in different ways (consult your physician). It may become necessary to avoid being chilled. More rest and sleep may be required as energy levels diminish. You must learn to pace yourself.

FIND A SUPPORT GROUP

Most organizations that focus on chronic medical conditions sponsor support groups. Support groups can provide emotional comfort, background information on your disease, opinions about treatments, recommendations about specialists, and support at a time when other friends and family members might not be able to meet your needs. If your condition limits mobility, you can still participate in a support group by joining one online.

REMAIN HOPEFUL

Maintain a positive attitude, but don’t expect miracles. It’s quite possible that in the not too distant future, a treatment advance may make your life considerably easier. Remember, your life is worth the fight.

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PREPARING FOR MEDICAL APPOINTMENTS

Coming prepared is an important step in managing your medical appointments to gain the most from these visits.

COMPLETE PAPERWORK AHEAD OF TIME

Have your caregiver accompany you to your medical appointments when possible. Your primary escort can help you remember details during and after your clinic appointments, and provide moral support. Being present at your clinic visits also helps your caregiver to understand your disease better, empowering them to better advocate for you in emergency situations.

CREATE A RECORD OF YOUR MEDICAL INFORMATION

Call your health care provider and ask them to mail or e-mail any paperwork you need to fill out ahead of time. This will give you time to complete the forms. Make two copies: one for your appointment and the other for your own medical information record.

If you are unable to get the paperwork in advance, bring your record of medical information with you to assist you to fill out the form completely and accurately. Ask the clinic to give you a copy for your records.

Keeping a record of your medical information in one place (file, binder) can help you to keep track of your treatment plan and reduce your stress in preparing for your appointments.

Your record should include:

- A list of all your health care providers and how to contact them: doctors, dentist, therapists, etc.
- Information on all your current medications such as: name of medication, dose, frequency, who prescribed it, why you are taking the medication and when you started (if possible, you should bring all your medications in their original containers to your appointment)
- Information on other treatments you may be trying, including: dietary regiment, vitamins, other non-prescribed supplements, alternative therapies, etc.
- A copy of your medical records
- Copies of tests and lab results

You can add a notebook to jot down notes or list questions.

MAKE A LIST OF QUESTIONS

Writing down your questions to ask your medical team at your next appointment will help ensure you remember them.

Questions you may want to ask:

ABOUT TESTS & PROCEDURES

- What will the results of this test or procedure tell you?
- What does the test or procedure involve?
- What are the potential risks of this test or procedure?

ABOUT MEDICATIONS

- Why are your prescribing this medication to me?
- How does this medication work to improve my health?
- What are the side effects of this medication?

ABOUT APPOINTMENTS

- How often should I see you?
- What changes should I make to my diet, exercise routine or lifestyle?

Write down questions as they occur to you during your day so you do not forget them.

Do not be afraid to ask for more explanation if you don't fully understand the information provided.

YOU ARE YOUR OWN BEST ADVOCATE

The members of your healthcare treatment team are medical experts in scleroderma, but you know your body best.

Don't be afraid to ask for explanations or seek a second opinion.

Don't be afraid to tell your doctor or clinic nurse about how you are feeling.

You are your own best advocate! Being informed and prepared will empower you to take charge of your well-being, communicate effectively with your medical team and ensure that your needs are met.

LEARN MORE

Educate yourself about scleroderma

Knowing more about the disease is key to understanding what's happening to your body. Scleroderma Canada and your local scleroderma group can provide educational resources in print and electronically to help you understand more about scleroderma.

Learn more about your treatment plan

Ask your treatment team about what information is relevant for you. Ask them to provide resources.

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MANAGING FATIGUE

FATIGUE & SCLERODERMA

Everybody gets tired from time to time. Fatigue from scleroderma, however, is different from normal tiredness in that it is often not related to physical exertion and not helped by getting enough rest. For many people with scleroderma, doing their daily activities or putting in a full day at work is exhausting. By the end of the day there is barely enough energy left over for spending time with your family or for enjoying hobbies and other activities.

Fatigue often develops gradually in scleroderma. You may not notice how tired you are until someone else points it out, or you realize you can't do some of your old activities. It is wrong to think of fatigue as a personal weakness or as failing. Your body is less capable of using energy reserved for everyday activities because some of this energy is used in the body's attempt to heal itself.

WHAT IS THE SOURCE OF YOUR FATIGUE?

If fatigue is a problem for you, the first job is to try to determine the cause.
It's important to remember that fatigue can be caused by, and made worse by, things other than scleroderma.
It is sometimes difficult to tell the difference between fatigue due to scleroderma and that due to depression or feelings of hopelessness.

COPING WITH FATIGUE

Fatigue is hard to treat. There aren't any medications that you can take that will return your old energy, but there are some things you can stop doing that might be making your fatigue worse:

BE REALISTIC

Before you had scleroderma, running a home and a full time job may not have been enough to make you tired. You may find now that balancing work and family leaves you exhausted. Even doing a couple of loads of laundry and making lunch can leave some patients very tired.

KNOW YOUR LIMITS AND WORK WITHIN THEM

In this way, you can gradually increase them. You must be especially careful not to overdo it when you are beginning to feel better again.

PACE YOURSELF

If you know that you can be active for 15 minutes before your symptoms worsen, do an activity for only 15 minutes, then rest, and repeat. Don't try to finish an activity before taking a break if needed.

STAY ACTIVE

Inactivity can worsen fatigue. A nap can help you, but staying in bed all day may actually make your fatigue worse. Physical activity or an exercise program may help you to feel more energized. Taking a short walk instead of lying down the next time you're feeling tired may actually help.

IMPROVE YOUR NUTRITION

Food is our basic source of fuel for energy. We can feel fatigued if we are not well nourished. Maintain a healthy weight and improve the quality and quantity of your food if your fatigue is made worse by a poor diet.

BE MINDFUL OF YOUR MOOD

Remember that depression can cause fatigue.

TALK WITH YOUR DOCTOR ABOUT YOUR FATIGUE

Your physician may help you identify other causes and ways to best cope with your fatigue. It's not always easy to pinpoint a problematic lack of energy so the best advice is to pay close attention to exactly how it feels so you can describe it to your doctor in detail.

THINK ABOUT YOUR SLEEP HABITS

Not getting enough sleep or having poor quality sleep can add to your fatigue.

GETTING ENOUGH SLEEP

Sleep is important for everyone, and even more so for people who suffer from chronic illness such as scleroderma. Sleep difficulties can make coping with scleroderma more difficult.

If you are not getting 8 hours of sound sleep each night, if you do not wake up feeling refreshed or if you sleep more than 10 hours a day, and if any of these conditions have existed for more than 2 weeks: it is time to see your doctor. Be sure to ask for a sleep clinic evaluation if you have any symptoms of sleep apnea, such as snoring or interrupted breathing.

TIPS TO IMPROVE YOUR SLEEP

Keep a regular sleep schedule. Set a fixed bedtime and waking time.

Invest in a comfortable mattress, bedding and pillows. Make your bed as comfortable as possible.

Minimize distracting light and noise.

Avoid caffeine (commonly found in coffee, tea, chocolate) for at least six hours before bedtime.

Avoid heavy meals close to bedtime. Spicy or fatty foods may be particularly troublesome because they are associated with acid reflux.

Avoid alcohol close to bedtime. Alcohol can make you drowsy but as your body begins to metabolize it, REM sleep, the period where your sleep is most restorative, is reduced.

Raise the head of your bed a few inches to reduce acid reflux/ GERD.

Put away electronics such as laptops, cell phones, TVs and tablets well before bedtime. If you use an electronic reader, turn the light level low.

Ask your doctor whether the time of day you take your medications or supplements may be keeping you awake.

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EATING WELL

EATING WELL WITH SCLERODERMA

Since scleroderma affects each person differently and each person may have differing nutritional needs, there is no specific "scleroderma diet".

In addition to difficulties swallowing, you can experience nutrition-related side effects in other parts of the body, such as kidney problems, that require modifications to what you eat. This brochure offers general advice to help you cope with common complaints. If you need more individual dietary information, please seek professional advice from your doctor or a registered dietitian.

FOOD PREPARATION

This is not always easy, especially if scleroderma has hardened the skin on your fingers and hands or if your joints are stiff. If this problem hinders your food intake:

- Get kitchen equipment and utensils with thick rubber handles to aid in gripping
- Try various types of cups and mugs until you find the easiest one to pick up
- Stock up on frozen meals or prepared food when possible
- Buy pre-cut fruit and vegetables

SWALLOWING & DIGESTION

Overproduction of collagen due to scleroderma can cause thickening and scarring of tissue in your esophagus (the pipe that connects the throat to your stomach) and digestive tract. Weakened muscles can cause slow movement of food which is called "dysmotility." Difficulty in swallowing is called "dysphagia." This can also result from narrowing of the esophagus.

- Eat slowly. Allow more time for eating, due to slower movement of food passing through
- Chew well. Be careful not to take any food into your lungs
- Eat soft or pureed foods (mashed potatoes, apple sauce). Process other food (meat, vegetables) in a blender and add seasonings, broth, or margarine to thin to desired consistency

GASTROINTESTINAL REFLUX DISEASE

Gastro-Esophageal Reflux Disease (GERD), can cause irritation in the esophagus. The muscles at the lower end of the esophagus may weaken and allow stomach acid to backwash into the esophagus, causing irritation or heartburn.

To reduce reflux, try the following:

- Sit upright for 1 to 2 hours after eating, so gravity can help your food move downward. Try not to eat just before lying down, napping or going to bed at night. Use a sleep wedge or raise the top of the bed to elevate your head and torso to prevent regurgitation of stomach contents into airways
- Decrease or eliminate the intake of alcohol, carbonated drinks, chocolate and caffeine
- Avoid highly acidic foods, such as citrus fruit, tomatoes and onions
- Avoid fatty and greasy foods. Foods with high fat content stay in the stomach longer than low fat content foods, increasing the likelihood of acid reflux
- Eat smaller, more frequent meals (4 to 6 small meals per day versus 2 to 3 larger meals per day)
- Keep a food diary to establish the foods that give you problems. If you are eliminating several foods from your diet, consult a dietitian or nutritionist to ensure your nutritional needs are being met
- Ask your physician about prescribed medications to help neutralize the acid

CONSTIPATION

Constipation may occur due to weak and scarred muscles in the colon wall. Suggestions for relieving constipation include drinking lots of water (1.5L – 2L daily) and eating more dietary fibre. Keeping active also helps - exercise stimulates movement of the bowels.

DIARRHEA

Diarrhea may occur due to weakened muscles of the gastrointestinal tract, antibiotics, or poor absorption of food. Foods that have soluble fibre, such as bananas, applesauce, apples, oatmeal, oat bran and prunes may be helpful. Avoid whole wheat bread and wheat germ or large quantities of raw vegetables and raw whole fruits (other than bananas). Avoiding high fat foods, fried foods and rich desserts may also help.

WEIGHT MAINTENANCE & NUTRITION

People who suffer from scleroderma are at increased risk of malnutrition caused by inadequate intake of nutritious foods or from poor absorption of nutrients from the GI tract. It is important for you to have your weight and nutritional status monitored regularly.

Significant weight loss over a period of 3 months or more may indicate inadequate nutrient and calorie intake. Other symptoms of malnutrition include:

Weakness and muscle wasting	Excessive or new onset fatigue	Increased susceptibility to infection
Delayed wound healing	Brittle nails and excessive hair loss	Overly dry, flaky skin

These symptoms above may also be related to underlying scleroderma and can be difficult to distinguish from malnutrition. If you have concerns, consult your doctor or a registered dietitian.

Inform your physician if you develop any of the above symptoms

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DENTAL CARE IN SCLERODERMA

DENTAL CARE IN SCLERODERMA

Scleroderma poses particular challenges to maintaining oral health. People living with scleroderma are more likely to be affected by conditions such as microstomia (small mouth opening), xerostomia (dry mouth), loosening of teeth, jaw pain and gum disease. Reduced manual dexterity may make flossing and brushing teeth more difficult. Please speak to your dentist about adaptive devices or tools that can help. More frequent dental check-ups are also important in managing dental care and minimizing oral health care problems.

Suggestions for your dental appointments:

- Tell your dentist you have scleroderma. Discuss your condition and how it affects you and your oral health
- Schedule short appointments for one area or one tooth at a time, or longer exam and care appointments with breaks
- Keep your lips lubricated with petroleum jelly
- Do your physiotherapy immediately before your appointment
- Schedule appointments at the best time of day for you
- Wear gloves and bring a blanket in case the office is cold

MICROSTOMIA (SMALL MOUTH)

PROBLEMS

- Small opening makes it difficult for patients and professionals to clean teeth
- Tightness of the mucosa (lining of the mouth) may pull gums away from the teeth
- Can be difficult to insert and remove dentures; upper dentures become more easily dislodged

SOLUTIONS

- Work with dentist and hygienist to develop accommodations for effective brushing and flossing
- Ask your dentist to use children's instruments if necessary
- Exercises to improve flexibility of mouth, lips and jaw muscles
- Periodontal (gum) surgery may improve mobility of the tongue and cheeks
- 2-part dentures to ease their passage into the mouth; osseo-integrated implants (titanium screws within the jaw) to secure dentures and bridges

LOOSENING OF TEETH

No treatment specific to scleroderma: traditional treatment for periodontal disease may include surgery and extraction.

XEROSTOMIA (DRY MOUTH)

PROBLEMS

- Harder to swallow food
- Increased incidence of fungal infections(thrush)
- Increases tooth decay and gum disease

SOLUTIONS

- Pay special time and attention to brushing and flossing
- Stay hydrated: drink plenty of water
- Avoid alcohol and smoking, which can worsen existing dryness
- Use medication to increase saliva production or use artificial saliva
- Sugar free hard candies to increase salivary flow.

JAW PAIN

PROBLEMS

- Affects the ability to chew
- May be confused with tooth ache

SOLUTIONS

- Exercise and massage
- Dental appliances
- Medications: muscle relaxants and/or anti-inflammatories
- Ask your dentist to schedule breaks in your longer dental appointments to allow you to rest your jaw

PREVENTION OF GINGIVITIS AND DENTAL DECAY

Scleroderma can have a significant adverse effect on oral health.

A number of strategies can help lessen the risks of long term consequences of oral disease (such as extractions, loose teeth, tooth loss, gum disease, abscesses):

DIET

- Avoid excess sticky or sweet foods, which increase the accumulation of dental plaque leading to decay
- Avoid soda and carbonated/ fizzy drinks, which cause mild chemical erosion of the tooth surfaces
- Limit spicy and acidic foods which may exacerbate gastrointestinal reflux disease (GERD/acid reflux)

DENTAL HYGIENE

- Clean teeth at least twice daily
- Floss between teeth daily
- Use a toothbrush with a small head and soft nylon bristles that can reach all areas of the mouth
- Use toothpaste and mouthwash containing fluoride

LIFESTYLE

- Use massage and oral exercises to keep your mouth and face more flexible
- Limit smoking and alcohol consumption
- Get assistance from a helper/ caregiver

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GERD: GASTROESOPHAGEAL REFLUX DISEASE

WHAT IS GERD?

GERD is an acronym that stands for gastroesophageal reflux disease. Gastroesophageal reflux is the term used to describe the blackflow of acid from the stomach into the esophagus when the lower esophageal sphincter does not close properly. When the reflux touches the lining of the esophagus, a burning sensation is experienced which is normally referred to as heartburn or acid indigestion. Nearly everyone has an attack of heartburn at some point in his/her life, and in the vast majority of cases, the condition is temporary and mild, causing only transient discomfort. Patients develop gastroesophageal reflux disease (GERD) when there is a persistent and frequent incidence of heartburn. If it remains untreated, serious problems can develop.

GERD AND SCLERODERMA

GERD is very common in systemic sclerosis (scleroderma). All gastrointestinal segments may be affected by scleroderma. One of the abnormalities is the dysmotility (muscles do not work properly) of the gastrointestinal tract. Scleroderma patients often have poor sphincter control, leaving it open and allowing more acid to wash into the esophagus.

CAUSES

The esophageal sphincter is normally closed when a person is not swallowing. Transient relaxations of the sphincter do occur a few times each day with most people. However, this occurs more frequently in patients with GERD, resulting in the reflux associated with this disease. Many lifestyle factors such as overeating, eating certain foods, and consuming caffeine may have an adverse effect on GERD.

Food in general can exacerbate GERD symptoms:

- Food fills the stomach and induces more transient relaxations of the lower esophageal sphincter
- Food stimulates acid production in the stomach to aid digestion which causes an increase of reflux into the esophagus

SYMPTOMS

Persistent heartburn and acid regurgitation are the main symptoms of GERD but there may be other effects:

- Esophageal – patients may experience pain on swallowing or at least have difficulty swallowing (dysphagia). The esophagus may become severely injured resulting in narrowed regions (strictures) that impair swallowing. Stretching procedures or surgery may be required. Paradoxically, strictures may actually improve other GERD symptoms by helping to prevent acid reflux
- Pulmonary – conditions such as asthma, interstitial fibrosis (scarring in the lungs), chest pain, and a dry cough may result from repeated aspiration
- Oral – patients may experience an increase in tooth decay, gingivitis or halitosis
- Throat – patients may damage vocal cords, experience hoarseness or develop laryngitis

LONG TERM COMPLICATIONS

GERD can cause some serious long term complications:

Ulcers of the Esophagus

Reflux can result in ulcers, which lead to bleeding. Persistent bleeding can result in iron deficiency anemia, and in some cases may even require emergency transfusions.

Barrett's Esophagus and Cancer of the Esophagus

A condition called Barrett's Esophagus is thought to result from long-standing GERD in some patients. The normal esophageal lining (epithelium) may be replaced with abnormal (Barrett's) epithelium. Barrett's esophagus is a risk factor for the development of esophageal cancer.

Sleep Apnea

Acid reflux can cause spasms of the vocal cords (larynx), which block the flow of air to the lungs. Such spasms may cause sleep apnea.

Asthma

GERD may cause asthma attacks in patients who have no allergies or history of lung disease. In such cases, some physicians believe that the acid reflux stimulates the vagus nerves, triggering the airways in the lung to constrict, resulting in asthmatic symptoms.

Respiratory Disorders

Patients with GERD appear to have a heightened risk for a number of respiratory disorders, such as chronic bronchitis, emphysema, pulmonary fibrosis and pneumonia.

TREATMENT

Treatment usually begins with lifestyle changes.

- Avoid trigger foods such as acidic foods, chocolate and peppermints
- Eat smaller and more frequent meals
- Lose weight if needed
- Reduce alcohol consumption
- Quit smoking
- Avoid lying down for at least 3 hours after a meal
- Raise the head of the bed 4-6 inches

Note: A journal is a useful tool to use to more accurately identify foods and activities that may trigger GERD symptoms so that lifestyle is not restricted unnecessarily.

MEDICATION

Medications may include:

- H2 blockers, such as Tagamet HB or Pepcid AC, may provide temporary relief
- Proton pump inhibitors (PPI), namely Losec, Prevacid, Pantoloc, and Pariet are probably the best agents for GERD related to scleroderma
- Prokinetics, namely Maxeran (metaclopramide) and Domperidone, help strengthen the sphincter and assist in emptying the stomach faster

ENDOSCOPY

Endoscopy is a diagnostic procedure that is sometimes administered to GERD patients who have swallowing difficulties or to those who do not respond to other therapies. A thin flexible tube is passed down the throat allowing the physician to directly inspect the lining of the upper gastrointestinal tract. This procedure can be used to identify complications of GERD and to take small samples (biopsies) for further analysis.

In most cases, GERD is a chronic condition that can be effectively managed with medications and lifestyle modifications. Patients should review their symptoms with their doctor and establish an appropriate treatment plan.

We would like to thank **Dr. Margaret Larché, MBChB, MRCP(UK), PhD** for her assistance with this information pamphlet.

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WHAT IS PULMONARY HYPERTENSION?

WHAT IS PULMONARY HYPERTENSION?

Pulmonary hypertension is high blood pressure in the arterial system in the lungs and the right side of the heart. There are many causes of pulmonary hypertension and numerous tests are usually required to determine the cause. Problems with the left side of the heart is the most common cause of pulmonary hypertension (pulmonary venous hypertension). Pulmonary arterial hypertension (PAH) is caused by the small arteries in the lungs becoming narrower or blocked. This causes the right side of the heart to work harder to push the blood through the lungs to get to the left side of the heart. Patients with scleroderma may develop PAH either due to interstitial lung disease (ILD) or to the changes in the blood vessels.

SYMPTOMS

Patients may experience the following symptoms:

- No symptoms in early stages
- First symptom is shortness of breath especially when walking or climbing stairs
- Coughing, which worsens upon exertion
- Constant feelings of fatigue
- Shortness of breath at higher elevations
- Shortness of breath even at rest at more advanced stages
- Unusual chest pains
- Swelling of neck veins, belly and feet
- Extreme limitations in daily activities
- Dizziness or fainting

Inform your physician if you develop any of the above symptoms

TESTS

Because of the high prevalence of PAH in scleroderma patients, the World Health Organization recommends that scleroderma patients be screened annually for PAH.

These tests usually include:

- Echocardiogram
- Pulmonary function tests (including DLCO)
- Electrocardiogram

Other tests to establish a diagnosis of PAH may include:

- Six minute walk test
- V/Q scan (a test to see if the lungs have old blood clots)
- CT Scan chest
- Assessment of function in tasks of daily living
- Right heart catheterization is a required test to confirm the diagnosis of PAH

TREATMENT

Treatment of pulmonary artery hypertension (PAH) requires individualized treatment by specialists in this field. Conventional medications such as diuretics, calcium channel blockers and warfarin may be used, but drugs designed specifically to treat the condition are usually required. Drugs that help prevent blood vessels from narrowing through blocking specific receptors in the vessels called endothelin receptor antagonists (ERAs) include: bosentan (Tracleer®), macitentan (Opsumit®) and ambrisentan (Volibris®). Medications that stimulate the lungs to make more of its own natural vessel relaxers (vasodilators) are in a class called Phosphodiesterase inhibitors and include sildenafil (Revatio®) and tadalafil (Adcirca®). Working slightly differently is selexipag (Uptravi®) which activates prostacyclin receptors which help the vessels relax. Combinations of these medications are frequently required and when they are ineffective, epoprostenol (Caripul®) may be prescribed. It is a liquid medication that is delivered by a portable infusion pump into a central vein (large vein leading directly to the heart). Treprostinil (Remodulin®) works similar to epoprostenol in that it is a liquid and the patient wears a pump, it can be delivered like epoprostenol , or the medication is delivered subcutaneously (under the skin) through a fine tip needle.

THE FUTURE

Many research projects are presently being conducted into understanding the causes and mechanisms of PAH. New therapies under investigation may be available in the future.

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RAYNAUD'S PHENOMENON?

WHAT IS RAYNAUD'S PHENOMENON?

Raynaud's Phenomenon is a condition that causes some areas of the body – such as fingers, toes, tips of nose or ears – to feel numb and cool in response to cool temperatures, emotional upset or stress. During a Raynaud's attack, arteries that supply blood to the skin narrow, which results in limited blood circulation to affected areas. The digits go white, then either blue and/or red as they rewarm.

Raynaud's Phenomenon occurs as two main types:

- Primary Raynaud's – When Raynaud's occurs without an underlying disease or associated medical problem that could provoke vasospasm, it is known as primary Raynaud's or Raynaud's Disease. This is the most common form of this disorder and most typically affects both hands and both feet. In Raynaud's Disease, most attacks are not too painful and are reversible
- Secondary Raynaud's – When Raynaud's is caused by another underlying problem it is known as Secondary Raynaud's or Raynaud's Phenomenon. Although secondary Raynaud's is less common than the primary form, it is often a more complex and serious disorder

Raynaud's Phenomenon is probably the most common early symptom of systemic sclerosis (scleroderma), and occurs frequently in both the limited and diffuse forms of scleroderma.

WHO IT AFFECTS

Women are more likely than men to have this disorder, and it is more common in people who live in colder climates. Raynaud's Phenomenon is considered a relatively common disorder occurring in up to 5% of the general population and 10% of young women.

SIGNS AND SYMPTOMS

Raynaud's is more than simply having cold hands and feet. Some of its symptoms may resemble frostbite, however with Raynaud's the numb prickly feeling or stinging pain that comes with warming may also happen with the relief of stress. Signs and symptoms of Raynaud's depend upon the frequency, duration and severity of the blood vessel spasms that underlie the disorder.

During an attack of Raynaud's, affected areas of the skin usually turn white first. The areas then turn blue and feel cold and numb. Sensory perception is dulled. The affected skin may look slightly swollen. As circulation improves, the areas may turn red, throb, tingle and swell. The order of the changes of colour isn't necessarily the same for all people, and not everyone experiences all three colours. An attack may last less than a minute to several hours. Over time, attacks may grow more severe.

Occasionally, an attack affects just one or two fingers or toes. The digits affected are not necessarily always the same digits. Although Raynaud's most commonly affects the fingers and toes, the condition also can affect other areas of the body such as nose, cheeks, ears and even the tongue. The reduction of blood flow often leads to painful ulcers, sores and infection on fingertips and toes. In extreme cases, fingers and toes may develop gangrene and partial amputation may be required.

Symptoms which may suggest Raynaud's Phenomenon (secondary to a connective tissue disease) may include:

- Inflammatory arthritis
- Changes in skin texture
- Shortness of breath
- Rash
- Sun sensitivity
- Severely dry eyes
- Weight loss

CAUSES

When the body is exposed to cool temperatures, extremities are the first to lose heat. The body slows down blood supply to fingers and toes to preserve the body's core temperature. It reduces blood flow by narrowing the small arteries under the skin of the extremities. In people with Raynaud's, this normal response to cold is exaggerated. Stress may also bring about this reaction.

Doctors don't completely understand the cause of Raynaud's attacks, but blood vessels in the hands and feet appear to respond abnormally to nerves that regulate them. The result is that the body overreacts to cold temperatures or stress. Although cold is often the trigger for a Raynaud's attack, changing temperatures is the prime cause even in a warm environment. This is probably caused by an exaggerated response of a normal mechanism, which maintains the central body temperature by shunting blood away from the hands and feet to the core. The cause of this exaggerated response is not known but is thought to be a failure of the normal chemical and neural control of the blood flow. Inflammation or vascular changes associated with scleroderma may also play a role.

With Raynaud's, arteries to the fingers and toes go into what is called a vasospasm. The vessels constrict temporarily, dramatically limiting blood supply. Over time, these same small arteries also may thicken slightly, further limiting blood flow. The result is that affected skin turns pale because of the lack of blood flow to the area. Once the spasms subside and blood returns to the area, the tissue may turn red before returning to a normal colour.

MANAGEMENT

For most people, Raynaud's is more a nuisance than a disability, and there are many common sense preventative measures. Most certainly the best is to minimize exposure to cold. Keep the extremities warm by wearing warm gloves and mitts especially when handling refrigerated or frozen items. Hats in the cold are helpful. It is important to keep the entire body warm as this prevents the onset of the Raynaud's episodes. Warming the body often relieves an attack better than warming the hands or feet.

Smoking and cigarette smoke should be avoided as nicotine decreases blood flow to the fingers and toes.

Learn relaxation and stress management techniques as they have been shown to be effective for many people.

TREATMENT

Treatment of Raynaud's depends on its severity and the presence or absence of associated conditions. While there is no cure for Raynaud's, it can be controlled and symptoms reduced by some medications that have been prescribed by physicians. Most common of these are channel blockers and vasodilators (drugs to dilate the blood vessels). These can be used intermittently or regularly with patients with severe manifestations. Sometimes nitroglycerine paste may be prescribed to open blood vessels. Mild blood thinners or drugs that decrease the stickiness of platelets may help to improve circulation. If gangrene or loss of a finger or toe is a danger, the physician may prescribe prostaglandin infusions. If skin ulcers become infected, they may require antibiotic therapy or local drainage. Many ulcers that develop are painful, so medication may be required.

We would like to thank **Dr. Evelyn Sutton, MD, FRCPC, FACP** for her assistance with this information pamphlet.

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SUPPORT GROUPS

YOUR PERSONAL HEALTH SUPPORT NETWORK

Taking an active role in managing your own health care is important in achieving your optimal wellness level. It is equally important to cooperate and communicate effectively with your primary care physician who is managing your illness. While these two - you and your doctor - are the focal points of your health management team, many other people and resources can form part of your health and support network:

Family and friends can provide emotional support, encouraging you and assisting with activities of daily living that you find more difficult

Other health professionals such as medical specialists, nurses, physical and occupational therapists, psychologists, massage therapists, etc. may form part of your treatment team

Community resources (voluntary and government agencies) offer health, social and rehabilitation services that may benefit a person with scleroderma

Scleroderma support groups will enable you to meet and exchange information with others who have scleroderma

BENEFITS OF SUPPORT GROUPS

One of the biggest advantages of a support group is that you'll meet other people who feel like you do. This can improve your mood and make you feel less alone.

Being in a support group can help you to learn successful tips about coping as well as managing daily choices and challenges.

Problem-solving with your fellow group members may also remind you that you also have knowledge and experience to share. That can prompt you to remember to apply these skills in your own life.

AM I A CANDIDATE?

Support groups can help anyone who needs emotional support, feels isolated or feels that the other people in their lives don't fully understand their struggle with scleroderma.

Peer-led support groups do not replace individual/group therapy or professional counselling, but can be a cost-effective and helpful tool to complement more formal treatment.

JOINING A GROUP - WHAT TO EXPECT

You may be uncomfortable at first sharing your concerns with strangers; however, the fact that the others in the group are facing similar challenges may help you to open up and discuss your feelings. Everything that takes place during the support group will be kept confidential.

Usually a support group is led by a member who has had some training or experience in facilitating group discussions. Unlike group therapy, peer self-help groups are usually not led by a professional therapist (such as a nurse, psychologist or social worker). These groups are not a substitute for more formal professional counselling.

You may get helpful ideas from other members, but don't take their opinions and comments more seriously than those of your doctors and health care team. Always keep your primary care physician informed of new therapies or lifestyle changes.

While groups share a common goal of providing mutual emotional support, all groups are different. Groups vary in size, frequency and format of meetings. Some groups may arrange community activities or guest speakers while others may be more informally organized.

If you have concerns about how your group is doing, you may want to speak privately to the facilitator who leads it to share your feedback, or you may want to find another group or other form of personal support that better suits your needs.

SCLERODERMA CANADA SUPPORT GROUPS

Scleroderma support groups exist across Canada. To find a support group in your area, contact the regional division where you live.

Check http://www.scleroderma.ca

for the most updated information or contact the SC Office at 1-866-279-0632 or info@scleroderma.ca

STARTING A SUPPORT GROUP

If there is no group near you and you are interested in starting one, the following questions can help you decide whether being a support group leader is a good fit for you:

Do you have the time?

Review your schedule and see if realistically you can put aside time to prepare for and run meetings.

How are you feeling?

Determine if your health is adequate at this time to take on this added commitment.

Are you comfortable leading?

Consider your ability to organize, speak in front of groups and facilitate conversations.

How do you approach your own illness?

Support group leaders need to lead with an overall sense of hope and encouragement, focusing on positive solutions vs. negative reactions.

Scleroderma Canada can provide funding, tools and training resources to support you as you support others.
Please contact us!

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DISCLAIMER: This pamphlet is meant to provide information on scleroderma and is not meant to be used as a diagnostic tool or to suggest treatment or medications. Always consult your physician regarding details of symptoms, diagnosis, and treatment.

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