SCLERODERMA & RAYNAUD'S UK

SYSTEMIC SCLEROSIS (SSc) AND LUNG INVOLVEMENT

WE ARE DEDICATED TO IMPROVING THE LIVES
OF PEOPLE AFFECTED BY SCLERODERMA AND RAYNAUD'S

WHAT'S INSIDE

ABOUT THIS INFORMATION

Systemic sclerosis (SSc) is a complex condition that affects people in different ways, and the information in this booklet is designed to provide an overview. It is written for people diagnosed with systemic sclerosis and lung involvement and is also useful for family and friends. This guide is backed up by reliable sources and evidence and has been reviewed by healthcare professionals as well as people living with these conditions themselves. Living with a rare condition means that you may need to become something of an expert yourself, so that you can make informed choices about your treatment. This guide can get you started, and we are here to provide information and support along the way.

UNDERSTANDING SYSTEMIC SCLEROSIS

What is systemic sclerosis?

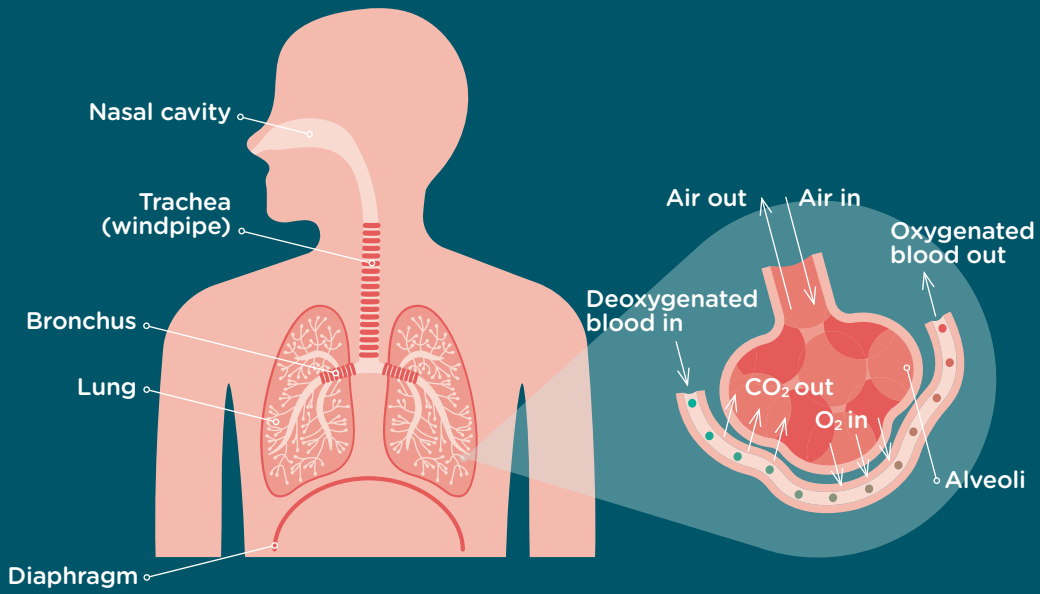
Systemic sclerosis (also referred to as scleroderma) (SSc), is a chronic, autoimmune disease associated with skin thickening and changes to blood vessels, particularly those supplying your fingers and toes. The hardening of the skin is caused by the formation of scar tissue, that occurs because of an increase in collagen, a normal component of tissues that is produced in excess. In SSc, scar tissue or 'fibrosis' can also form within the internal organs including the lungs, the gut or the kidneys.

Excessive scarring is due to the overreaction of the immune system, whereby it attacks the body itself. Why this occurs in individuals with SSc is unknown. One of the important ways that the immune system deals with injury or infection is by causing inflammation. Normally, following injury (e.g., a bad cut) or infection (e.g., pneumonia), inflammation helps the body to repair itself by triggering the process that leads to healing. A necessary part of this process is scarring. In people living with SSc, the immune system remains continuously 'switched on' and is therefore overactive. This means that inflammation is constant, leading to damage within the body through fibrosis (scarring).

About lung function

The human body uses oxygen to convert fuel from the food we eat into energy. When we breathe in, the muscles of the chest move upwards and outwards, and the diaphragm moves down. This allows space for the lungs to fill with the air we inhale, which moves through the lungs to the air sacs, or alveoli. Oxygen passes into the blood and is then carried to cells throughout the body.

When we exhale, the alveoli absorb the carbon dioxide from the blood, the chest muscles move downwards and inwards and the diaphragm moves up. This forces the air out of the lungs. Carbon dioxide from the alveoli flows though the lungs and out through the mouth or nose.



HOW DOES SSc AFFECT THE LUNGS?

Lung involvement in SSc means that scarring or fibrosis is occurring within the lungs. The most frequent type of lung involvement is **interstitial lung disease (ILD)**, in which the walls of the small air sacs or alveoli that make up the lungs are thickened by a mixture of inflammation and fibrosis. Because of the presence of scarring, this type of lung involvement is also called **pulmonary fibrosis**. A relatively less common presentation of lung involvement in SSc is **systemic sclerosis-associated pulmonary arterial hypertension** (abbreviated to **SSc-PAH**), meaning high blood pressure in the lungs. For more information on SSc-PAH, please see page 14.

When a scarred lung is examined under a microscope, it is possible to recognise a number of different patterns of fibrosis, that will usually relate to different conditions. In scleroderma, the most frequently occurring pattern is called non-specific interstitial pneumonia (or NSIP for short).

How frequent is severe lung involvement in systemic sclerosis?

Not everyone with SSc has lung involvement, however lung disease is found in more than half of patients with SSc, with its degree varying from person-to-person. In most cases this is limited in its extent, and only causes symptoms on exertion. In this group, the condition tends to remain stable over time and does not require specific treatment.

In approximately one-out-of-five cases, interstitial lung disease is more severe and may worsen with time, because of the continued development of scar tissue within the lungs. Significant scarring can result in problems with normal lung function, and treatment to prevent the fibrosis from worsening will be necessary. In only a very small number of individuals, lung disease may progress despite the treatments currently available.

Symptoms

Symptoms of ILD may include:

- Shortness of breath that may be more noticeable on mild exertion
- Dry cough
- Chest pain
- Dizziness or feeling lightheaded
- Persistent tiredness
- Tightness within the chest

Symptoms will often include a cough and shortness of breath, particularly when climbing stairs and walking uphill. However, these will not necessarily mean that lung disease is present: e.g., if you develop a cough this could be a sign of something else, such as an infection or chronic heartburn. If lung involvement is limited in its extent, you may have no symptoms at all.

HOW IS LUNG INVOLVEMENT DIAGNOSED?

Testing is vital to accurately assess whether you have SSc-lung involvement, and if so, to determine its extent and the best course of action.

Annual tests are an essential aspect of managing systemic sclerosis as these will monitor disease progression. Even if you have never had any issues in the past, regular tests are crucial to identify any changes early on so that you can be treated before things get any worse. If you notice any changes in-between appointments, it is important to tell your doctor.

Lung function tests

Lung function testing includes a variety of tests that check how well the lungs work and is the most useful way to detect lung fibrosis and assess whether your condition has progressed. Testing should be repeated every six-to-twelve months and should form part of the annual tests that you will be offered routinely. A steady decline in lung function test results could indicate that fibrosis is progressing and that further tests or treatment may be required.

These tests are not painful and are generally performed by a pulmonary function technician. It is normal to repeat the tests several times to confirm that the results are accurate. Before having these tests, it is helpful to keep the following in mind:

- You should take your daily medications prior to testing unless told otherwise.
- If you are using a short-acting inhaler as required, do not take this for six-to-eight hours prior to testing, if possible.
- Your doctor may give you other instructions regarding medications.

Spirometry

This is the most basic test that is used to measure the amount of air you can breathe out, as well as how forcefully air can be emptied from the lungs. Spirometry is used to screen for diseases that affect lung volumes, and for conditions affecting the airways, such as bronchitis, emphysema or asthma. There are also tests to check if asthma is present when the usual breathing test results are normal.

Lung volume testing

Lung volume testing is another commonly performed lung function test. This measures the volume of air in the lungs, including the air that remains at the end of a normal breath. In addition, a diffusing capacity test (also called Transfer Factor or DLCO), measures how easily oxygen enters the bloodstream. This is an important measure as it is very sensitive for the presence of scarring or high blood pressure in the lungs.

Exercise testing

This helps to evaluate causes of shortness of breath. A frequently used test is the six-minute walk, which consists of walking up and down a set distance (usually in a corridor), to measure the total distance you can walk and whether there is a fall in oxygen levels. If you are asked to perform this test, you do not need to prepare for it in advance, and the technician or nurse specialist will explain how it works. Exercise tests may also be performed on a bike or treadmill, in which case you should plan to wear loose fitting, comfortable clothing and appropriate shoes. You will be attached to a heart monitor and blood pressure machine to monitor your vital signs. You will be given additional instructions about how to prepare for this test when it is ordered.

"I had the usual worries with any new treatment. I felt confident that my specialists knew what they were doing and although aggressive treatments at a young age was very scary – I knew what was needed to try and stabilise my lung fibrosis."

Ashton

CT (computed tomography)

A CT scan is a more detailed X-ray scan of the chest, which picks up even mild degrees of lung involvement. This is non-invasive and does not hurt, although you may need an injection of contrast to allow better images. Together, the lung function tests and the CT scan are used to estimate whether interstitial lung disease, if present, is mild or extensive. If the results of lung function tests are declining, a CT scan may be repeated to help assess any progression.

Echocardiogram

As part of the initial assessment for someone diagnosed with SSc, an echocardiogram is also regularly performed. This is an ultrasound scan that is used to look at the heart and nearby blood vessels, to identify any problems with the heart or high blood pressure in the lungs, called pulmonary hypertension (PH). This is achieved by using a small probe to send out high-frequency sound waves that create echoes when they bounce off different parts of the body. These echoes are picked up by the probe and displayed as a moving image on a monitor during the scan. It is very similar to the ultrasound scan used in pregnancy.

Cardiac catheterisation (also known as right heart catheterisation)

If pulmonary hypertension is suspected by echocardiogram or other clinical findings, your doctor may suggest carrying out a right heart catheterisation. This will accurately measure the pressure in your pulmonary artery and is used to definitively confirm the cause of the pulmonary hypertension. It is performed under local anaesthetic.

Bronchoscopy

Occasionally, a bronchoscopy may be carried out. This involves passing a thin, flexible, fibreoptic camera into the airways to check for any problems and obtain samples of the inflammatory cells within the lungs if necessary. One reason to consider this would be to check for possible infection.

Other tests you will have

Lung function testing is one aspect of the annual tests used for disease monitoring in SSc. You are entitled to have these tests, so do not be afraid to ask if they are not being offered, taking this booklet with you if necessary. Do not miss routine check-ups even if you feel that nothing has changed; and report any new symptoms as soon as you can.

As well as lung function tests, annual testing should include the following:

- **Cardiac function tests (ECG and echocardiogram)**
  These tests detect how well the heart is working, to check whether this has been affected by scleroderma or if your condition has progressed since the last assessment.
- **Kidney function tests**
  A blood test will check how much of a waste substance called creatinine is in the blood, as higher levels may indicate that the kidneys are not working properly.
- **Blood pressure tests**
  These estimate how hard blood is being pushed against the sides of arteries as it is pumped around the body.

TREATMENTS

Making treatment decisions

There are various treatments available for interstitial lung disease, and the best option for you as an individual will depend on your situation. Treatments are aimed at reducing symptoms and living well, but also at preventing the fibrosis from worsening. Usually, the purpose will be to stabilise your condition, although in some cases of early diagnosis there may be an improvement if the disease has not yet had a chance to progress.

Before any treatment decisions are made, you will be assessed by a respiratory physician and/or a rheumatologist. They will ask about your medical history and whether you have been a smoker; as well as carry out a physical examination, lung function and imaging tests. Your doctor will then discuss a treatment plan with you, and there may be several options available.

Consent to treatment

Before treatment starts, your doctor will talk to you about the potential benefits, as well as the possible side effects and any associated risks. You will have the opportunity to discuss anything that you do not understand or any concerns you may have. It is important to understand exactly what the treatment involves before you decide to go ahead, and to feel as confident as possible that you have made the right decision, so do ask for more time to consider the options if necessary.

Having high-quality information will help you feel more confident about your treatment decisions, so always ask for clarification if there is something that you do not understand.

The following questions may be helpful at the time:

- What do you think is the best type of treatment for me?
- What may happen after I start this particular treatment?
- How will this affect my everyday life?
- What are the side effects?
- What clinical trials are available to me?

For consent to treatment to be valid, it must be voluntary and informed, and the person consenting must have the capacity to make the decision.

Consent should be given to the healthcare professional responsible for the person's treatment. Consent is sometimes given in writing, e.g., by signing a consent form for a medical procedure, or it may be verbal, such as confirming to the doctor that you are happy to have an X-ray. It could also be implied, for example, by holding out your arm to have blood pressure taken.

When it comes to giving consent to taking medications, verbal rather than written consent is often used.

"Everything happened so quickly, but I never felt uncomfortable with choices that had to be made. At the time the disease was progressing fast so decisions on the best way to tackle this situation had to be made as quick as possible."

Georgina

Treatment options

There are a number of effective treatments available for lung involvement in systemic sclerosis:

Immunosupressants are the most commonly prescribed drugs for interstitial lung disease. They work by reducing the overactivity of the immune system, slowing the development of irreversible lung scarring. Immunosuppressants come in tablet form and include Mycophenolate and Azathioprine. In some cases, an intravenous treatment (by drip infusion) may be considered.

Low-dose steroids (usually Prednisolone at 10 mg once daily) may be prescribed together with an immunosuppressant drug.

Antibiotics may be given to treat and prevent infections, which is especially important when taking immunosuppressants.

Antacids (e.g. lansoprazole), are often prescribed, since the acid can irritate the lungs and may worsen fibrosis.

Newer treatments such as Tocilizumab that target the immune system and inflammation have been tested and evaluated in clinical trials and shown to be effective, although Tocilizumab has not yet been approved for the treatment of lung involvement in systemic sclerosis in the UK. Rituximab, another treatment that targets the immune system also shows promise and is being evaluated in clinical trials.

Anti-scarring agents such as Nintedanib that slow down the scarring of the lung have also been shown by clinical trials to be effective, although again this has yet to be approved.

Pulmonary rehabilitation (PR) is a programme of exercise and education for people affected by a chronic lung condition. The aim is to help individuals to make the best use of their lungs by breathing more efficiently, and to cope better with breathlessness. A PR course will typically last for eight-to-twelve weeks, with sessions designed to provide information along with a suitable programme of exercise; all delivered by trained healthcare professionals including physiotherapists, occupational therapists and nurses. For more information or to ask for a referral, talk to your doctor.

Treatment should be continued for as long as there is evidence of ongoing inflammatory activity due to scleroderma. For most people, it is usually possible to gradually reduce and stop treatment once the lung disease is no longer active. It is important to realise that this process of waiting for inflammatory activity to subside and then gradually reducing and eventually stopping treatment may take several years.

Side effects during treatment

All drug treatments have side effects. The type and extent of these will vary from person-to-person and will depend upon various factors, including the type of treatment and the dosage prescribed. It is important to be informed and to understand how to deal with them if they happen. This will also help in making treatment decisions.

Side effects can include an increased appetite, mood changes and difficulty sleeping, and occur most commonly with steroid tablets.

Some side effects may develop during treatment and continue for a short time, whereas others can appear later and may be longer-term. These can be unpleasant and uncomfortable, but it is critical that you do not stop the medication without first discussing this with your medical team. Stopping a prescribed medication suddenly may cause further unpleasant side effects in the form of withdrawal symptoms.

Your doctor or respiratory nurse specialist will see you regularly and can answer any questions about managing and reducing the impact of medication side effects.

Follow-up care

Your doctor will work with you to develop a personal, follow-up care plan. This is about how your health will be monitored over the coming months and years and will involve regular check-ups at your hospital or with your GP or community nurse.

Follow-up also provides the opportunity to ask questions and discuss any long-term side effects. Your doctor may help you to manage these, or they may refer you to a service that can. Getting these answers from a professional who understands your individual circumstances can provide reassurance and help you feel more in control of your situation.

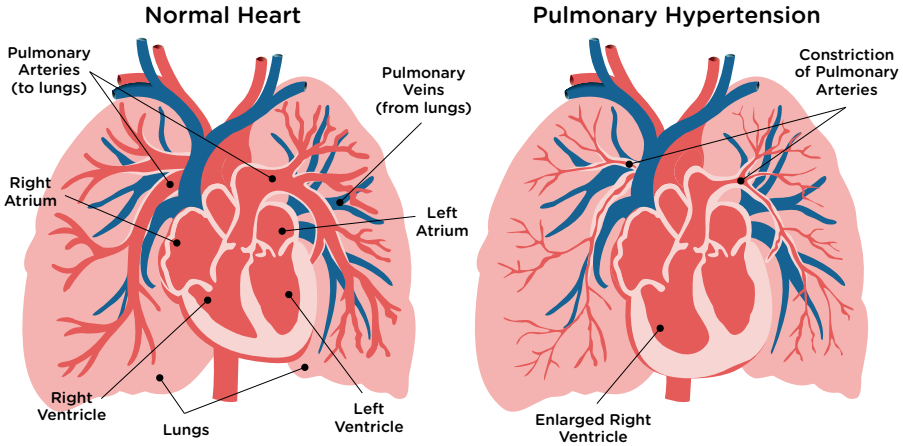
Difficulties between appointments

Your healthcare team can provide details of whom to contact if you have any problems with your treatment or notice any new symptoms between appointments. It is important to get in touch if you have any serious concerns, if you are struggling with side effects or if you notice worsening symptoms such as breathlessness. Remember the first port of call is often your GP or nurse specialist, who can then directly discuss the situation with your doctor.

SYSTEMIC SCLEROSIS-ASSOCIATED PULMONARY ARTERIAL HYPERTENSION

Pulmonary hypertension (PH) means high blood pressure within the pulmonary arteries, which are the main blood vessels that carry blood to the lungs from the heart. This can lead to damage to the right side of the heart, so that it cannot work properly. When the cause is narrowing of the small blood vessels, this is called pulmonary arterial hypertension (PAH), which is another possible complication of systemic sclerosis.

Systemic sclerosis-associated pulmonary arterial hypertension (SSc-PAH) refers to inflammation and scarring within the blood vessels of the lungs. This may lead to the thickening of the walls of the pulmonary artery, to the extent that it can no longer open up easily to relieve the pressure of the blood inside. If extensive, this may lead to a significant build-up of pressure in the vessels feeding into the lungs from the heart, so that blood pressure in the pulmonary artery is higher than normal. This affects the small blood vessels of the lungs, so that there is less blood reaching the lungs and less oxygen carried within the bloodstream. The high pressure then leads to a stretching of the right side of the heart, meaning that it has to work harder.



What causes SSc-PAH?

About one-in-ten people with SSc can go on to develop SSc-PAH. It is thought that this is because SSc thickens and stiffens the walls of the pulmonary artery so that it cannot open up easily to relieve the pressure of the blood inside it. The pressure inside the pulmonary artery is therefore higher than it should be, resulting in extra strain on the heart.

SSc-PAH will often not develop until many years after the first symptoms of systemic sclerosis appear. When it occurs in SSc it may also be called "connective tissue disease-associated pulmonary hypertension." PAH is also associated with various conditions including rheumatoid arthritis, blood clots in the lungs, liver disease and HIV infection, although sometimes there is no known cause.

Symptoms of SSc-PAH

The onset of SSc-PAH can be very slow. For many people, the breathlessness is so slight in the early days that they barely notice it. It is therefore very important to be aware of the symptoms to look out for, and to tell your doctor if you notice any changes, such as increased breathlessness.

Symptoms are caused by low levels of oxygen within the blood stream, or by the heart not working as it should, due to the extra work it is having to do. They include the following:

- Shortness of breath with minimal exertion
- Tiredness
- Fatigue
- Dizzy spells / blackouts
- Chest pain
- Swelling in ankles or legs

Treatments for SSc-PAH

There are a number of effective treatments for SSc-PAH, and the best option will depend on several factors, including the severity of symptoms.

There are certain treatments that are designed to help maintain wellbeing and help you continue living as actively as possible.

These include:

Oxygen therapy: In some cases, continuous oxygen therapy may be suitable for people with very low oxygen levels in the blood. Oxygen is a vital gas that is required for all active bodily processes, such as using muscles and digesting food. It helps to stabilise the level of oxygen in the blood, helping you to breathe more easily. If you are travelling, you may need to have oxygen with you for your flight and in transit; so, try to discuss your plans with your medical team well in advance of your departure date.

Anticoagulants: some forms of PAH may carry an increased risk of developing blood clots. Anticoagulant medicines including warfarin can help to prevent this and enable blood to flow more freely.

Diuretics or water tablets: these work by increasing urine output and are used to remove excess fluid from the body to reduce the strain upon the heart. Removing water from the blood decreases the amount of fluid within the blood vessels, which also reduces pressure. Diuretics also treat symptoms like swollen ankles.

Other therapies are more targeted treatments, that will aim to slow down the progression of your condition and may even reverse some of the damage to the heart and lungs.

These include:

- **Calcium channel blockers:** these are vasodilators that are used to help open up the blood vessels. This group of medications includes nifedipine, diltiazem, nicardipine and amlodipine.
- **Endothelin receptor antagonists (ERAs):** these are a form of targeted therapy used to slow the progression of PAH and even reverse some of the damage to the heart and lungs. They work by reducing the level of endothelin within the blood. Endothelin is a factor in the narrowing of blood vessels that can affect the vessels of the lungs by causing them to narrow, increasing blood pressure within the pulmonary arteries. Reducing the level of endothelin in the blood can limit the damage that it can cause. These medications include bosentan, ambrisentan and macitetan.
- **Phosphodiesterase 5 (PDE 5) inhibitors:** are another targeted therapy that aims to slow progression and limit some of the impact. PDE 5 inhibitors include sildenafil and tadalafil and are widely used as a treatment for erectile dysfunction, because the blood vessels are so similar. They work by inhibiting an enzyme called phosphodiesterase type 5 [PDE5] from working, which helps blood vessels to relax, increasing blood flow and lowering blood pressure.
- **Prostanoids:** are powerful vasodilators that are used to enable blood to flow more freely through the vessels of the lungs, ultimately reducing the strain upon the heart. This group of medications work in a similar way to prostaglandins, that occur naturally within the body. In the longer term, the beneficial effect upon the heart may mean that it is able to work more efficiently. Examples of prostanoids include epoprostenol, iloprost and treprostinil.
- **Soluble guanylate cyclase stimulators:** these are a form of targeted therapy used to slow down disease progression. It is prescribed in the form of riociguat tablets, that work by causing the blood vessels in the lungs to dilate.
- **Inhaled treatments:** these include nebulised iloprost. This is a vasodilator used to open up the blood vessels of the lungs, allowing blood to flow more freely.

At your first treatment session, the process will be explained and you will be told if there is anything you need to do before your treatment starts. Some of these drugs also require regular blood tests for monitoring. There are also some ongoing clinical trials which can be discussed with your PH specialist if this is something that you might consider.

Pulmonary rehabilitation (PR)

This is a specialised physiotherapy programme of exercise and support to help train your muscles and manage breathlessness. PR is suitable for individuals with a variety of lung diseases, including SSc-PAH. For more information, please see page 12.

Surgical procedures

There are certain surgical procedures that may help, but not in all cases. Your medical team will discuss the aims of any potential procedure with you, as well as whether you might be a suitable candidate. You may also be asked to attend a pre-operative assessment clinic. This will involve conducting tests to check how well your lungs are working and your general health, to assess if you are fit enough for surgery.

In severe cases, a lung or heart transplant may be needed, but this is relatively rare.

"The team of doctors and nurses were great. I couldn't fault their care and support. I felt very lucky to have such a good team which took a massive weight of my shoulders knowing I was in good hands."

Georgina

MANAGING YOUR WELLBEING

Although living with a chronic illness like SSc can affect your emotional wellbeing as well as your physical health, there are some positive steps you can take to help reduce the overall impact and maintain your quality of life. Treatments will also work best when you play an active role alongside your healthcare team, and there are certain things you can do to help make your treatment as successful as possible and reduce any side effects.

Eating well

Although a healthy diet is always important, living with SSc often leads to some gastrointestinal symptoms that may affect what you are able to eat and digest comfortably. You may also struggle to maintain weight, especially if your breathing is affected, as this may mean that you are using more energy. Your doctor can arrange for a referral to a dietician who will be able to help you optimise your nutrition to help maintain wellbeing.

How will stopping smoking help me?

Smoking cessation can lower blood pressure and improve circulation as well as help your lungs. Smoking can cause emphysema, which when combined with lung scarring can make your breathing worse. In addition, it will increase the risk of lung cancer.

When trying to stop, it is a good idea to have a plan; setting goals such as not smoking at home or in the car and going places where you cannot smoke can help considerably. Ask your friends and family not to offer you a cigarette or smoke near you and tell them that you are trying to stop. Importantly, there are free smoking cessation services to support you and treatments to get you through the cravings. Ask for details at your GP surgery or local pharmacy.

Physical activity

Regular exercise is good for your heart, lungs and muscles, and helps to keep you supple. A common misconception is that physical activity may worsen conditions like PAH, when in fact it can help your body respond better to breathlessness when done in a sensible and controlled manner. Even if you do not feel up to anything strenuous, every little bit helps.

Exercise can help you to:

- Manage fatigue
- Reduce stress and anxiety
- Boost your mood and energy level
- Help you relax
- Improve your sleep

Physical activity can also reduce the risk of depression, increase appetite and boost confidence; and keeping active during treatment is generally safe as well. Whilst it is natural to feel a little nervous about overexerting yourself and getting out of breath, light exercise can have real benefits to your wellbeing. There is no specific exercise that is guaranteed to help, so start with something you enjoy so that you are more likely to stick to it. This can include daily activities such as walking your dog or gardening, or simply moving about more. Examples of aerobic and strengthening exercises can be found on our website, as well as breathing techniques.

The following websites may help you find activities near you:

- NHS – **nhs.uk/live-well/exercise/free-fitness-ideas/**
- NHS inform Scotland – **sinform.scot/healthy-living/keeping-active**
- Sport Wales – **www.sportwales.org.uk**
- Sport Northern Ireland – **www.sportni.net**

Breathing exercises

PURSED-LIPS BREATHING:

This can be used at any time to help control your breathing.

1) Breathe in gently through your nose, then purse your lips (as though you are about to blow out candles).
2) Blow out through your mouth with your lips in this pursed condition.
3) Blow out for as long as comfortable, but do not force your lungs to empty.

BLOW-AS-YOU-GO:

This helps make tasks and activities easier and is effective when doing something that causes breathlessness.

1) Before you make the effort (e.g. lifting something), breathe in.
2) Breathe out while making the effort. Using pursed-lips may also be beneficial.

PACED BREATHING:

Particularly useful when active, such as when walking.

1) Count to yourself as you walk or move, e.g., breathe in for one step and then take one or two steps as you exhale.
2) Adjust your steps to your breathing to find a combination that works best for you.

BELLY BREATHING:

This helps to strengthen the diaphragm muscle, which allows a person to take a deep breath.

1) Rest a hand on your stomach and breathe in slowly through the nose. Note how far your stomach rises.

2) Breathe out through the mouth.

3) Breathe in again through the nose, this time trying to get the stomach to rise slightly higher than it did previously.

4) Breathe out. Try to make each exhalation twice or three-times as long as each inhalation, depending on what feels most comfortable for you.

Other areas to consider when seeking to maintain your quality of life include getting enough sleep and learning relaxation techniques to ease anxiety. Please refer to our *'Understanding Scleroderma'* booklet and our website for more detailed information.

Living with SSc-associated lung involvement

This condition can impact upon various aspects of life and may sometimes mean that you need to take more time to consider certain things, and to ensure that you have as much information as possible.

- **PREGNANCY:** pregnancy can be more complicated if you have SSc. The majority of affected women will have normal fertility, however younger women may have a higher risk of infertility than those who have already had children. It is vital to speak to your rheumatologist or PH specialist if you are considering trying to conceive, as they will assess your health and any risks surrounding pregnancy. In addition, many of the drugs used to treat these conditions can affect fertility and pregnancy development.

- **TRAVEL:** travelling with SSc-PAH can be challenging, however many people do manage to take overseas trips successfully. Talk to your medical team about your travel plans and ensure that you have enough medication to cover the entire holiday. Contacting the airline before you travel to discuss any special requirements will also be beneficial.
- **EMPLOYMENT:** by law, you do not have to tell your employer about your diagnosis. Some of our community report that it was more practical to do this, and that their employer was able to better support them after being informed, however everyone's circumstances and preferences are different.

For more detailed information on each of these areas and for further advice on living well with SSc, please visit:
sruk.co.uk/scleroderma/managing-scleroderma/

With thanks to our clinical reviewers:

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Consultant Respiratory Physician at Royal Brompton Hospital and Honorary Senior Lecturer at Imperial College London.

Professor Chris Denton

Professor of Experimental Rheumatology at UCL Medical School and Consultant Rheumatologist and Joint Director of the Centre for Rheumatology, Royal Free Hospital.

Dr Colin Church

Consultant at the Scottish Pulmonary Vascular Unit, Glasgow

"It is difficult to work as you did in the past as you have so many hospital appointments etc., and you can be off sick more than others. This is where I have been very lucky with my employers, they have been very accommodating to my situation."

Diane

HOW SRUK CAN HELP

We are the only UK charity dedicated to improving the lives of people affected by Scleroderma and Raynaud's. We exist to improve awareness and understanding of these conditions, to support those affected, and ultimately to find a cure. We understand that being diagnosed with a condition can be a scary and uncertain time. But don't worry, we are here for you, every step of the way. We provide trusted, reliable and evidence based information on Scleroderma and Raynaud's. We want you to be able to learn more about your condition, feel confident in working with your health professionals and receive the right care for you.

Access to support services

Our online community is a friendly space where you can exchange advice, information and support with others who are affected by Scleroderma and Raynaud's. Many of our community members are living with these conditions themselves, but friends, family and partners are also welcome. We can connect you with your local Scleroderma and Raynaud's Support Group, to connect with others in a similar situation.

You can contact our free Helpline 365-days-a-year on 0800 311 2756.

To find out more and receive all the latest information, please join our community on social media and start building a network of supportive friends today:

WeAreSRUK

Research

We fund scientific and medical research to better understand the causes and progression of these conditions and enable us to find better treatments as we work towards a cure. Our community is at the heart of our research programme. We are committed to addressing your needs to improve life in the here and now, alongside focussing on our long-term aim of discovering a cure. Through our investment in research, we have increased life expectancy for people living with scleroderma and have brought more treatments into clinics.

To find out more about current studies, please visit the central register at **www.clinicaltrials.gov** and search for 'scleroderma'.

HOW TO GET INVOLVED

The work of the charity is funded entirely through donations, fundraising and memberships. We would like to ask you to support our work so we can continue to improve lives.

Become a member

As a member of SRUK, you will be entitled to all the following:

- Four issues of our magazine, received quarterly
- Priority booking for all patient educational events
- Regular member-only discounts in our online shop, where you will find products that are tailored to these conditions
- Invitations to sign up for observation and/or product trials we may be running throughout the year, with key product partners and market research partners

Visit our website to find out more:
www.sruk.co.uk/membership or call our team on 020 3893 5998

Donate to us

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Sources used

We rely on several sources to gather evidence for our information.

All our information is in line with accepted national or international clinical guidelines where possible. Where no guidelines exist, we rely on systematic reviews, published clinical trials data or a consensus review of experts. We also use medical textbooks, journals, and government publications.

If you would like further information on the sources we use on a particular publication, please contact the Information and Support Services team at **info@sruk.co.uk**

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Valuing your feedback

As someone who has received a copy of this booklet, we would very much value your opinion on whether it meets the needs of people affected by Scleroderma and Raynaud's. Please complete the survey online at **sruk.co.uk/publications**

Published: September 2021
Next review: September 2024

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