Learning what scleroderma means

My mum has been pretty much my rock throughout. She did all the research for me. My family are very positive, and I think as a whole, we just focus on the positive things. They’re all just happy, and I’m happy, and that’s how we get through.

Jessica, a person living with scleroderma

A guide for caregivers and loved ones of a person living with scleroderma

“"My mum has been pretty much my rock throughout. She did all the research for me. My family are very positive, and I think as a whole, we just focus on the positive things. They’re all just happy, and I’m happy, and that’s how we get through.”

Jessica, a person living with scleroderma
Scleroderma (also known as systemic sclerosis) may be a complicated disease to understand. But it’s worth taking the time to get your head around it. The more you know and understand, the more confident you will feel in the care you can provide, and the better support you can give.

Current scientific knowledge and real-life experiences have been summarised in this booklet to help you learn about scleroderma.

You will also find more information and handy resources at morethanscleroderma.com

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What is scleroderma?

Scleroderma is a condition that results in hard, thickened areas of skin and sometimes problems with internal organs and blood vessels.¹

Scleroderma is caused by the immune system attacking the connective tissue under the skin and around internal organs and blood vessels. This causes scarring and thickening of the tissue in these areas.¹

There are several different types of scleroderma that can vary in severity. Some types are relatively mild and may eventually improve on their own, while others can lead to severe and life-threatening problems.¹

There’s no cure for scleroderma, but most people with the condition can lead a full, productive life.¹

The symptoms of scleroderma can usually be controlled by a range of different treatments.¹

Scleroderma is part of a family of diseases that affect connective tissue. Connective tissue is in almost every part of your body. It is what helps to hold your body together. It supports, connects and separates different parts of your body. Because scleroderma affects the connective tissue, symptoms can occur in any part of the body, including the skin, muscles, blood vessels and internal organs.⁵,⁶

In scleroderma, the immune system causes the natural healing process to go into overdrive and produce too much collagen.⁵,⁷,⁸

Scleroderma is a rare disease.² In Europe, a rare disease is defined as affecting fewer than 5 people per 10,000.³ In the United States, a rare disease is defined as one that affects fewer than 200,000 people in the whole population.⁴
Connective tissue is like a sponge cake with cells sitting in it like pieces of fruit. It is made up of a mesh of fibres that supports and holds cells.

Collagen is one of the fibres that make up the mesh.

Fibroblasts are one type of cell held within the mesh that helps us heal, keeps us healthy, repairs damaged tissues and forms scars.

Understanding connective tissue

Connective tissue is in almost every part of your body. It helps to hold your body together. It supports, connects and separates different parts of your body.

What happens in the body?
When a part of your body gets damaged (for example when you are injured), it sets off a natural healing cycle to repair the damage.\(^5\,7\,10\)

1. **When you are injured, the body's natural defence system (immune system) gets involved. The area then becomes inflamed.**

2. The immune system signals to the fibroblasts to repair the damage.

3. Fibroblasts produce collagen and other substances to repair any damage to the connective tissue; this in turn forms a scar.

4. The damage is repaired. The scar will soften over time as the normal surrounding tissues recover. In fibrosis, this normal softening of the scar does not occur enough.

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Your immune system has a memory. Your immune system protects you against threats from bacteria, viruses and danger from damage to tissues. Your body's immediate response to a threat is to produce inflammation. Inflammation is a normal response from your body's defences. It surrounds, contains and then gets rid of whatever is causing the problem (infection, splinter, thorn, etc).

Once the danger has been removed, your immune system produces special substances called antibodies. These are programmed to recognise bacteria, viruses or other dangers you have already been exposed to.

Antibodies find them and let the immune system know, so that they can deal with them more quickly.
In scleroderma, the immune system causes the natural healing process to go into overdrive and produce too much collagen. The immune system sends the wrong messages to the fibroblasts, telling them to produce lots of collagen. This is because it thinks your own cells are a danger and tries to defend your body against itself.

Fibroblasts then produce too much collagen.

The extra, unneeded collagen, gathers to form thick and rigid areas like a scar.

Extra scarring can make the problem worse. The scar tissue (fibrotic tissue) itself can cause damage, setting off the inflammation, collagen and scarring response; and the cycle goes on and on.

Fibrotic tissue is stiff and doesn’t work in the same way as normal tissue. Fibrosis and inflammation in the skin and other organs affects how they work and causes the symptoms of scleroderma.

Scleroderma is an 'autoimmune' disease
'Auto' = self
'Immune' = protection against
Why do people get scleroderma?

There is no simple answer to this question. No one really knows why some people get scleroderma, while others don’t.7

We know that scleroderma is not an infection, like chicken pox or flu. It isn’t caught from another person who has it.

Scleroderma is not a disease that can pass directly to children or be inherited from parents. Genetics and genes may play a small part in the risk of developing it.5-7,11 If a family member has scleroderma, there is a slightly higher risk that those closely related to them could develop scleroderma than for everyone else.5

It seems likely that some people have a genetic ‘switch’ for scleroderma that is triggered by something. Nobody really knows what the trigger might be.

It’s likely that events which are very stressful on the body (like a physical injury, exposure to toxic chemicals, or mental stress) can alert the immune system and also change how the person’s genes work. This could be the trigger for scleroderma in some people.7,9,12,13

There isn’t a way to find out who will get scleroderma, or to know how it happened in those who have been diagnosed. Scientists are looking in to how scleroderma works, so we might know more in the future.
Scleroderma is a family of diseases usually characterised by hardening of the skin.\(^6\)

**Scleroderma**
*combines the Greek words ‘sclero’ (hard) and ‘derma’ (skin)*

Different words are used to describe the parts of this disease family, and they all have slightly different meanings:

- **Scleroderma**: used to describe the sclerosis (hardening) of the skin (derma)
- **Systemic**: used when a disease affects a number of different tissues and organs in the body, or the whole body
- **Sclerosis**: used to describe the hardening of tissues in the body

Scleroderma is roughly divided into two forms, morphoea and systemic sclerosis.\(^6\)

Systemic sclerosis is then divided into four subtypes (limited cutaneous, diffuse cutaneous, sine scleroderma and overlap syndrome).\(^2\) The subtypes of systemic sclerosis help doctors to get a better picture of what sort of symptoms and problems the person you care for may experience, and to work out what sort of treatment they might benefit from.

The terms ‘scleroderma’ and ‘systemic sclerosis’ are often both used to mean the type of scleroderma that affects several organs in the body, but the correct term is **systemic sclerosis**.\(^10\)
Because connective tissue is everywhere in the body, there can be lots of different scleroderma symptoms. The person you care for won’t necessarily get them all. Most people start with just one or two symptoms, but they can appear in any order. They can be mild or more severe, and can change as time passes.5,14,15

That’s one of the things about scleroderma – people get different combinations of symptoms, and have different experiences and problems with those symptoms.9,16,17

The majority of these symptoms can be managed, making sure that you and the person you care for can continue doing most of the things you enjoy.

Encourage the person you care for to report all their symptoms to the doctor/nurse, no matter how insignificant they may seem. This can help them to understand the effects of scleroderma better. Don’t be afraid to ask any questions you have about their symptoms.

Don’t count on your hand the things you can’t do anymore, but look forward and think of the things you can do.  

Mandy, a person living with scleroderma
Skin symptoms

Nearly all people with scleroderma (more than 90%) get some skin symptoms.²,³ These symptoms can include:

- **Skin thickening** particularly on the joints of the fingers.²
- **Small red spots** (called telangiectasia) in around 75% of people with scleroderma.⁷ These are little, widened blood vessels, usually on the hands and face.
- **Tight or shiny skin** on the face, arms, or legs. Tightened skin around the joints can prevent proper movement. These are known as contractures. People may also get **itchy skin**.²
- **Hard lumps** (called calcinosis) in around 25% of people with scleroderma.⁷ These are a collection of calcium salts beneath the skin and develop mainly on pressure points, such as the fingertips.

“Simple things like reaching to take her mug from the top shelf and make a cup of hot chocolate is something she can’t do, because the scleroderma tightens her skin.”

Sharon, supporting a person living with scleroderma
**Hand and foot symptoms**

**Hand symptoms**

- **Raynaud’s phenomenon** occurs in more than 90% of people with scleroderma. It’s one of the most common early symptoms. It is caused by spasming of small blood vessels in the fingertips. This reduces blood supply to the area, creating a colour change (usually white to blue/purple, then to red) and making them cold. It often happens in cold weather or when the person is stressed. It can sometimes cause pain or make their fingers feel numb.

- **Red, swollen areas** usually on the fingers and toes that are painful to touch (sometimes called chilblains) can develop because of Raynaud’s phenomenon.

- **Puffy hands and fingers**

- **Tightening and thickening of the skin on the fingers** (called sclerodactyly). Sometimes this can make it hard to keep the hand flat. The fingers may not move as well as they should or it may make it difficult for the person to hold objects.

- **Ulcers or open sores** occur in around 40% of people with scleroderma, particularly on their fingertips or toes. These usually happen after a cut or knock, but can also happen due to Raynaud’s phenomenon. The person you’re caring for should always see their doctor if they get an ulcer or open sore, no matter how small or mild it may seem. The doctor can help make sure it doesn’t get infected.

- **Pain or stiffness** in the joints of the fingers

**Foot symptoms**

- **Raynaud’s phenomenon** can also affect a person’s toes.

- **Uncomfortable walking** because of changes under the ball of their foot. Some people describe this as feeling as though they are walking on pebbles.
Joint and muscle symptoms

Around 65% of people with scleroderma have problems with their joints or muscles at some time. They can affect any area, from their hip to their fingers, and can make it more difficult for them to stay active. Muscles can become weaker if fitness is lost because of doing less exercise.

Symptoms can include:  

- **Tight skin or swollen joints**
- **Joint pain or tenderness** might also be caused by inflammation
- **Other joint conditions** can sometimes also occur alongside scleroderma, such as rheumatoid arthritis
- **Muscle pain or aching** - this can make the person feel like they’ve done a bit too much exercise
Lung problems

While most people with scleroderma get some fibrosis and scarring in their lungs, only 40 to 75% develop changes that they can notice. Up to half of people who will be affected experience symptoms within the first 3 years of the disease. It is important to find out whether scleroderma is affecting the lungs of the person you care for. If they experience any symptoms related to their lungs, they should tell their doctor as soon as possible. They can then be checked over and can get any recommended treatments.

There are two ways in which scleroderma might affect the lungs:

1. Pulmonary arterial hypertension (or PAH). PAH affects around 15% of people with scleroderma. It happens because the fibrosis narrows the small blood vessels in the lungs. This causes the pressure to rise in the pulmonary artery (the big blood vessel that carries blood to the lungs) because the artery is trying to force blood through narrower blood vessels. The body tries to fix the problem by making the right side of the heart work much harder.

   Symptoms of pulmonary arterial hypertension include:
   - Shortness of breath
   - Feeling tired
   - Dizziness
   - Chest pain
   - Swollen legs

   The doctor will regularly monitor the person you care for. If they do develop pulmonary arterial hypertension, they will help manage it with appropriate treatments.

2. Lung fibrosis (also known as interstitial lung disease [or ILD]).

   Pulmonary (lung) fibrosis can be one of the most serious complications of scleroderma. This affects most people with scleroderma to some extent, but only between 40 and 75% of people will have enough fibrosis to affect their lung function and/or have symptoms.

   Lung fibrosis occurs because the fibrosis and scarring affects the little air sacs in the lungs (the alveoli), making them stiff and less able to inflate and deflate. The alveoli are responsible for transferring oxygen from the air into the blood, and removing carbon dioxide from the blood to be breathed out. The scarring and stiffness mean that they can’t do this job effectively.
Some people with lung fibrosis have mild respiratory symptoms that they may only notice when they really exert themselves. Others might experience more severe shortness of breath that affects their daily activities.\textsuperscript{16}

**Symptoms of lung fibrosis include:**\textsuperscript{16,24,25}

- Feeling very tired when going about normal activities
- Getting out of breath doing simple tasks like climbing stairs
- A dry cough that doesn’t go away
- Feeling tightness in the chest
- Feeling dizzy

Specialist doctors across a number of different countries recommend that lung function should be tested regularly, and at least annually, in people with scleroderma.\textsuperscript{27-30} This is so that doctors can help to manage the symptoms as early as possible.

Encourage the person you care for to report any symptoms to their doctor. The sooner they say something, the sooner their doctor can help do something about it.

_Mandy, a person living with scleroderma_
Heart disease is more common as we get older, so it’s not surprising that people with scleroderma get the same heart problems as everyone else. However, there are some specific heart problems that can occur because of scleroderma.

Heart problems can arise for several reasons, but usually because fibrosis (scarring) can affect the muscle of the heart, or the valves and structures in and around it that help it to work properly. Heart problems can help the doctor or nurse understand a little more about how the person’s condition is progressing.31

The symptoms of heart problems are varied and can be hard to spot. There may be no symptoms at all.31

**Symptoms include:**

- Breathlessness
- Feeling like their heart is pounding, fluttering or beating irregularly (palpitations)
- Light headedness
- Fainting
- Tiredness (fatigue)

Your doctor will check the heart of the person you care for regularly, to look for problems and to make sure nothing is missed. Encourage the person you care for to attend their appointments and tell their doctor if they have had any heart issues. They can then be identified and managed as quickly as possible.
The digestive system involves different organs in the body — the mouth, the stomach, and the intestine (gut). It is involved in lots of activity, so it’s not surprising that most people with scleroderma (around 90%) get some symptoms related to it. There are many different symptoms and they can differ from person to person.14\,16\,17\,32

Symptoms happen because of the way the body digests food. As food enters the body through the mouth, it is swallowed to reach the stomach. The muscles in the wall of the gut then move the food along, allowing it to be digested. In scleroderma, fibrosis (scarring) might make the walls of the gut a bit thicker and then muscles don’t work as effectively. It can also make the valve at the top of the stomach a bit stiff so that it doesn’t close properly. These cause the different symptoms that people experience.14\,16\,17\,32

**Symptoms include:**14\,16\,17\,32

- Heartburn or indigestion
- Changes in appetite
- Stomach pain
- Constipation
- Diarrhoea
- Feeling sick (nausea)
- Finding it hard to swallow

“We don’t go out and eat like we used to, but what we do is we find ways around that. So, we go for coffee, we love going for coffee, and we’ll sit, and chat and we’ll watch the world go by.”

Alison, supporting a person living with scleroderma
Just under half of people with scleroderma get some kidney problems. Scarring and fibrosis in the kidneys can reduce their ability to work properly, but in most cases there aren’t many symptoms and it’s usually not serious.

A few people with scleroderma (5 to 10%) experience a more serious issue, called renal crisis. It happens quickly and needs urgent treatment in hospital.

The symptoms of renal crisis include:

- Changes in vision
- Headache
- Feeling feverish
- Chest pain
- Feeling generally unwell

If the person you care for experiences these symptoms, encourage them to contact their doctor straight away. They may need urgent treatment.
Over 90% of people with scleroderma experience severe tiredness (sometimes called fatigue). It is one of the most common symptoms. There are several ways you can help support the person you care if they experience fatigue. Often, it’s about encouraging them to rest. Planning and pacing of activities can help. They can still do what they want or need to do, but maybe over a slightly longer time, in shorter bursts, or with rest times in between.

Encourage them to tell their doctor if they experience severe fatigue. This is so that the doctor can make sure their fatigue isn’t caused by other conditions.

“...If I’ve spent two hours out in the garden, or two hours out shopping, afterwards I usually do have to come and put my feet up for a while, and recuperate, regain my energy before I do something else.”

Mandy, a person living with scleroderma
Mental and emotional health

While not an actual symptom of scleroderma, some people find that the disease has an impact on their mental and emotional health. Some people find scleroderma changes how they see themselves, or how others see or treat them. It can affect the way the person you care for does their job, or the role they play in their family.

Adjusting to life with a new set of limits can be hard. It may create uncertainty or sometimes loneliness.

If you have any concerns that the person you are supporting has more serious mental health issues, such as severe depression or suicidal thoughts, talk to your nurse or doctor, or get in touch with a patient organisation.

Amongst it all, remember to consider your own health

Your health is as important as the person you care for. Try to remember to take time to look after yourself too.

Whatever the situation, whatever you’re feeling, you don’t have to go through it alone. Remember to reach out for help.

Help can come from anywhere – your friends and family, your doctor or nurse, or a local support group.

Make sure you speak to someone if you’re finding things difficult.

"I always say to my daughter, ‘I didn’t use to look like this,’ and she says to me, ‘it doesn’t matter, Mum. You are still beautiful,’ and that is nice. That is very encouraging and, you know, it’s not just her saying that."

Doris, a person living with scleroderma
How should symptoms be monitored?

Scleroderma can cause many different symptoms, and affects different people in different ways. The symptoms experienced by a person with scleroderma may change over time, so regular monitoring with the doctor is important. The type of specialist to be seen, and how often to see them, will depend on an individual’s symptoms and how their care is managed.

People recently diagnosed with scleroderma will have check-ups every 3 to 6 months. They may then become less frequent over time (for example, every 6 to 12 months) if your doctor considers the symptoms of the person you care for to be well managed and unlikely to change very much.

As it is important to spot any changes early, your help can be invaluable. You can get involved in helping the person you care for monitor their symptoms. You will know what’s normal for them, and can encourage them to let their doctors know when things change.

The doctor will perform a range of tests routinely, and when things start to change. Symptoms can help predict the course of scleroderma. Serious skin problems, as well as kidney, heart and lung symptoms, may need particular attention.

Let the person you care for know if you have any suggestions for things they might like to ask their doctor/nurse. You might also have your own questions.
There isn’t a specific test for scleroderma. The doctors/nurses might use many different tests to get a clearer picture about what is going on, and to monitor the progress of the person you care for.

The doctor/nurse will decide which tests they need to do and when. It may be that the tests are not all done at the same time.

They will probably perform or arrange lung and heart function tests at least once per year. Depending on the person’s progress and treatment, they may also perform kidney and liver function tests, along with blood counts and blood pressure.

You might be able to accompany the person you care for to their tests, or they may prefer to go alone. Whatever they choose, encourage them to ask for the test results to be explained. If you aren’t there, they can then explain things to you later. This will help you both to gain a better understanding of their symptoms and what you can do to help.
Blood and urine tests

What they are looking for

- Many things that the body uses or produces are carried around in the blood, or are passed out in the urine. These tests look at the levels of different chemicals, proteins, cells, etc, for:
  - Signs that the body’s immune system is active (inflammation, antibodies or autoantibodies)
  - Signs that some organs in the body aren’t working as well as they should

What to expect

To get clear results, the person in your care may be asked to skip breakfast or avoid certain foods just before the test. Sometimes it is important that they do not take their usual medication before blood is collected. The doctor or nurse should let them know these things in advance. If they are not sure of anything, encourage them to ask before going to the test.

The person you care for will usually have to collect their own urine in a small pot.

Understanding the results

Blood tests are commonly used to understand what’s happening in the body and to see if the organs are working properly. There are many types of tests and some could be used by the doctor to exclude other causes of the symptoms.

Often, people with scleroderma have autoantibodies in their blood.

- ‘Antibodies’: These are little proteins that form a key part of the body’s defence against bacteria and viruses. They search for, and attach themselves to these invaders. This helps the immune system find them and get rid of them

- ‘Autoantibodies’: These are antibodies that target the body’s own cells (instead of invaders)

There are many forms of autoantibodies. The most common in people with scleroderma are called anti-topoisomerase I (or Scl-70), anticentromere (or ANA), or anti-RNA polymerase III. These are all antibodies to things inside cells, but the immune system has mistaken them as a danger. Some blood tests looking for autoantibodies are used to help make a diagnosis of scleroderma.
Nail blood vessel test

**What they are looking for**
The health of the little blood vessels.

**What to expect**
The doctor or nurse will use a microscope or a small camera (sometimes a dermatoscope) to look at the skin at the bottom of the fingernails (the nailfold). This process of viewing these blood vessels (capillaries) is called nailfold capillaroscopy. It usually takes place in a doctor’s office. You can reassure the person you care for that this is not a painful test.

**Understanding the results**
The results will tell them what the blood vessels in the rest of the body might be like. In scleroderma, they can be abnormally shaped and this can affect how blood reaches areas of the body. It might be one of the reasons for blood flow problems, like Raynaud’s phenomenon.
Blood pressure test

**What they are looking for**
The pressure of the blood in the main blood vessels that supply the body.

**What to expect**
A band will be put around the person’s upper arm. The doctor or nurse will then fill it full of air. This might feel a bit uncomfortable but it’s only temporary. The air will slowly be released and this will tell them the blood pressure. This test is extremely common and usually takes place in a doctor’s or nurse’s office.

**Understanding the results**
It can be used to monitor a person’s general health or determine if other diseases might be causing their symptoms.

Blood pressure comes in waves (with every heartbeat), so two numbers will be recorded; one is the maximum pressure and the other is the minimum. The ideal result is below 120/80 mmHg, however numbers below 140/90 are considered OK.\(^{38}\)
Skin examination

**What they are looking for**

The health of the skin.

**What to expect**

This test is usually one of the first that the doctor will do to assess the skin of the person you care for. The doctor will ask to touch their skin and might use a small microscope, called a dermatoscope, to look at their skin more closely.

**Understanding the results**

This test is used to see if there could be other explanations for any skin symptoms. The person you care for will be referred for further tests, like a biopsy, if the doctor has any concerns.
Skin thickness test

This is often called the modified Rodnan skin thickness score or mRSS.\(^39\)

**What they are looking for**
The thickness of the skin.

**What to expect**
The doctor will feel the thickness of the person’s skin at several areas of their body. Each area is then given a number that shows how thick it is. The higher the number, the thicker the skin is compared with normal skin thickness.

**Understanding the results**
A higher number indicates more thickening of the skin. No thickening (in any areas) has a score of 0. The maximum score depends on the number of areas assessed.
Heart health tests

What are they looking for

• How well the heart is beating. Its rhythm (electrocardiogram [or ECG]).
• The shape of the heart and how well it can pump the blood (echocardiogram [or echo]).

What to expect

In most cases, these tests are done with the person lying down.

For an electrocardiogram, the doctor/nurse will stick sensors to the chest and these will record the electrical signals from the heart.

For an echocardiogram, the doctor/nurse will hold a device against the person’s chest. It uses sound waves to create images of the heart as it moves. From these, the doctor can see the shape of the heart. They can also get an idea of how well the blood is moving through the heart.

The doctor/nurse might ask for some more specialised types of heart tests, but they will tell the person about this before the day. These tests might take place at a specialist heart clinic.

Understanding the results

Heart health is not used to diagnose scleroderma, but the results can indicate if the heart is affected. It can also be used to exclude other diseases that might be causing the symptoms.
Lung health tests

These are also called lung function tests (LFTs) or pulmonary function tests (PFTs).

What they are looking for

How well the lungs are working (how much air is breathed in and out; and how much oxygen gets from the lungs into the body).

What to expect

Lots of different tests are possible but these will all usually involve the person breathing into tubes in certain ways. The doctor may also listen to the sounds inside their lungs, or ask the person to sit in a special room for extra measures and tests.

Common tests

• Measuring the capacity and flow of air into the lungs: spirometry, lung volume tests
• Measuring how effectively the breathed in air is reaching the blood: pulse oximetry, carbon monoxide diffusion test, arterial blood gas test

Most tests will be conducted at a specialist lung clinic, but some can be done by another doctor or nurse.

Understanding the results

This varies depending on the test. The doctor will monitor any changes in the person’s lung health over time.
Imaging and scans

What they are looking for

The health of the organs inside the body.\(^42\)

What to expect

There are lots of different scans and machines. Some include:

- **Ultrasound:** A sensor is placed on the skin and can see inside the body using sound waves. It can see how things inside the person’s body look and move.

- **X-ray:** Creates a picture of the lungs, revealing shadows that can suggest scarring. The doctor will show the person which position they need to sit in. It can help if they wear loose, comfortable clothing. They may need to wear a hospital gown.

- **Computerised Tomography (CT) and High-Resolution CT (HRCT) scan:** A type of X-ray that provides sharper and more detailed pictures than a standard chest X-ray.
  
  During the test, the person will be asked to lie in a donut-shaped machine. A ring will then move up and down their body.

- **Magnetic Resonance Imaging (MRI):** Uses a large magnet and radio waves to look at organs and structures inside the body. Healthcare professionals use MRI scans to diagnose a variety of conditions.
  
  During the scan, the person will be asked to lie on a table that slides inside a tunnel-shaped machine. The scan can take a long time, and the person will be asked to stay still. The scan is painless. The MRI machine can make a lot of noise, so the technician may offer the person earplugs.

None of the scans or images will involve things that hurt

They will all be done by specialist doctors or nurses, and some of them can take a few hours. If you accompany the person you care for to these tests, be prepared for some waiting around.

Understanding the results

The results vary depending on the test used. The doctor will normally be looking for specific things in the images and may monitor the difference against any images taken previously.

The HRCT scan is commonly used to monitor the health of the lungs in people with scleroderma.
How are the symptoms managed?

The management of scleroderma is more far reaching than pharmacological treatment alone, comprising diagnosis of the disease and identifying patients at risk of organ involvement, as well as regular monitoring and use of non-pharmacological approaches. There are also several potential treatments in progress that might be helpful in the management of scleroderma.43,44

**Treatment options**

Currently, there is no cure for scleroderma, but there are several therapies under investigation that aim to alter how the disease progresses.2,7

There is a range of different treatments designed to manage the many symptoms and organ problems associated with scleroderma.2,7,43

The doctor will help find the treatments that work for the person you care for. Encourage them to tell the doctor exactly what they want from their treatment, to help them find the best one.

It is also important to monitor how well you both think the treatment is working and to report any side effects.
What might the future hold?

Everyone is affected by scleroderma in different ways. This can apply to you as a caregiver, as well as the person you care for. It can be difficult to predict how things will progress.

None of us know what the future holds, but by looking at the symptoms a person with scleroderma is experiencing, you can get an idea of what to expect. These are the facts.

- Around half of all organs that will be affected by scleroderma show signs within the first 2 years of disease onset.

Organs that are often affected by scleroderma include:

- The skin (9 out of 10 people)\(^7\)
- The digestive system (9 out of 10 people)\(^7\)
- The lungs (4 to 8 out of 10 people)\(^22\)
  - Less commonly, the kidneys or heart (1 out of 10 people)\(^7\)
- Lung disease can be a common complication of scleroderma and limits breathing function in 40 to 75% of people\(^22\)

- 1 in 4 people may develop noticeable lung disease within 3 years of diagnosis\(^{22,45}\)

- Although some of the information on scleroderma can be quite scary, it is important to know that the condition only shortens the life span in a small number of people who have severe or complicated forms of scleroderma\(^{30}\)

The lifespan of people with systemic sclerosis is extending with ongoing research and as treatments improve\(^{27,46}\).

- More than 8 out of 10 people survive for more than 10 years from diagnosis. And this is increasing every day\(^{46}\)

Encourage the person you care for to report any changes in their symptoms to their doctor or nurse. They will be able to investigate, discuss the options, address any concerns, and put in place any medical support that is appropriate.

Don’t forget to reach out for any help that you need. Caring for another person is a demanding and skilled role, and help is out there if you need it.
Will the person I care for be on medication for life?
It will depend on their symptoms. Different treatments are used to manage the many symptoms of scleroderma. These treatments should lessen the impact the disease has on the person’s activities.

If you have questions or concerns about the medications that are recommended for the person you care for, talk to them about it in the first instance. They might be able to answer your questions. If not, suggest that they ask their doctor/nurse.

If they are having problems with treatment, make sure to contact the doctor who recommended it. They will be able to provide more detailed advice and tips.

Is scleroderma contagious?
No. Scleroderma is not an infection and cannot be passed directly from one person to another like chicken pox or flu. You can’t catch it from the person that you care for.

Will scleroderma change our lives?
While the symptoms of scleroderma can affect life for both you and the person you care for, they don’t have to rule it.
The doctors and nurses will help manage the symptoms of scleroderma and can recommend changes that can help you both continue to do the things you enjoy. If you attend any appointments, don’t be afraid to ask any questions you have. The doctors and nurses will be more than happy to help.

Providing care is a vital part of managing life with scleroderma. Your contribution will be invaluable to the person you care for. Caring can also be challenging, so remember to take time out to care for yourself. Consider visiting morethanscleroderma.com for more information.

Will scleroderma affect our relationship?
Becoming a caregiver can alter relationships between you and the person you care for. Your roles might need to change. If you sometimes feel as if you need support, make sure that you reach out to those around you.

My partner has scleroderma. Will it affect our sex life?
People with scleroderma can develop symptoms that have an impact on their relationship. Skin tightening, discomfort, pain, and other symptoms can affect the sex life between partners where one or both has scleroderma. If these symptoms become a problem, talk it over with your partner and consider talking to your doctor.

Some women with scleroderma also have another autoimmune condition called Sjögren’s syndrome. This means that their immune system is attacking the glands that make fluids. Women with Sjögren’s syndrome can experience vaginal dryness, but this can often be overcome by using vaginal lubricants.

Around 8 out of 10 men with scleroderma experience erectile dysfunction. This can be for several reasons, but particularly as scleroderma can cause poor circulation. Erectile dysfunction is a common problem that can be addressed by the doctor or a urology specialist.

With sexual problems, it is a good idea to try and provide support for each other, while seeking help from the doctor/nurse. It might be a bit embarrassing at first, but it is something you can work at together.
Will the person I care for be able to work?
It depends on how they are feeling and what symptoms they develop.
They might be able to talk to their employer and to arrange some support. This might mean changing their role or changing the way they do their job.
You might also be able to provide some extra support that enables them to work - like arranging for lifts or 'outsourcing' some of their household chores.
The person you care for with scleroderma should be aware of their employment rights and benefit entitlements. This can help prepare them when they go and talk with their employer. You could also consider looking into what support you can obtain for your role as a caregiver.

My female partner has scleroderma. Can we have children? 51-53
If you are thinking of having a baby and one/both of you have scleroderma, it’s important to talk to the doctor/nurse. Some medications shouldn’t be taken by women who are trying to get pregnant. Doctors can also do some physical tests to see what risks there might be. 57

Pregnancy in a woman who has scleroderma might be classed as ‘high risk’. While this sounds scary, it means that she will get plenty of extra support and care from the hospital and community. As a carer for a woman with scleroderma, you might be called on to provide a bit of extra support during their pregnancy.

Pregnancy causes changes to the body. In women with scleroderma, pregnancy can cause some symptoms to improve, while some others might get worse. 53-54
Mothers with scleroderma might also need to stay in hospital a bit longer after delivery. 55
This is so the doctors can monitor their symptoms.
Most doctors agree that, with careful planning and monitoring, women who have scleroderma can have a safe and healthy pregnancy. 51-55

Will scleroderma be passed on to our children?
Scleroderma is not a disease that you can pass directly to your children or inherit from your parents. Genetics and genes may play only a small part in the risk of getting scleroderma. 5,7

Is scleroderma a terminal illness?
People are affected by scleroderma in different ways. Every journey will be different.
It can be difficult to predict how the disease will progress. We know that more than 8 out of 10 people survive for more than 10 years following their diagnosis. 46
We also know that the survival of people with scleroderma has been increasing over the last few decades. 46

What will happen depends on many factors, including the type of scleroderma they have. The symptoms and their progression can give an idea of what might happen in the future. Don’t be afraid to ask the person you care for or their doctor/nurse this type of question. It is an important thing to discuss.
Positive actions you can take

You can provide the best support by learning about scleroderma, and how it affects the person you care for. Help them to make a plan for the days when they aren’t feeling so good.

Think about how you can look after yourself too.

Scleroderma can be unpredictable and demanding for you and the person you care for. When considering how you can best care for someone with scleroderma, it’s also important to consider how the condition impacts your own lifestyle. Don’t forget about your own personal goals. These are important too. It might take a while to adjust to a new lifestyle, but you can work at it together.

“Eventually I was able to, kind of, put a positive spin on it, as Michael would say and be that cup half-full person. So, the last couple of years, although they’ve been really challenging, I’ve managed to turn that around and become a lot more positive.”

Alison, supporting a person living with scleroderma