SCLERODERMA & RAYNAUD'S UK

Newly Diagnosed

What is Scleroderma?

Scleroderma is a chronic connective tissue autoimmune condition, involving the over production of collagen. Autoimmune conditions occur when the body's tissues are attacked by its own immune system. In this instance the overproduction of collagen leads to hardening of tissues, which in turn results in scarring and reduced normal function of the affected organs. The exact cause or causes of scleroderma is still unknown and due to this there is currently no cure for scleroderma but treatment and management can make it possible for many people with scleroderma to lead full and productive lives.

There are two main groups of scleroderma, and within are further subsets:

1. **Systemic Sclerosis – the systemic form of scleroderma, which affects the internal organs as well as the skin**
2. **Localised scleroderma (also known as morphoea), – a type of scleroderma that affects specific areas of skin and underlying tissues, but does not involve the internal organs.**

Symptoms can vary widely from person to person, but usually include hardening/ thickening of the skin, swelling of the hands and feet, joint pain and stiffness and blood vessel damage leading to a physical over-reaction to cold or stress. The heart, oesophagus, blood vessels, kidneys, lungs, blood pressure and digestive system can all be affected resulting in a variety of other symptoms, some of which can be life threatening or cause physical disability. According to their severity, the symptoms of scleroderma can be treated with drug therapy, physical therapy, surgery and self-management techniques.

Breakdown of Localised and Systemic

LOCALISED

Affects specific areas of skin and underlying tissues, but does not involve the internal organs.

Linear Morphea

A line of affected tissue. Linear Morphea is common in children and can affect whole limbs. This can lead to poor growth of the muscle and bone.

En Coup de Sabre

Type of linear morphea, a narrow groove runs up one side of the forehead.

SYSTEMIC

Two types of systemic scleroderma: limited and diffuse.

Limited

Skin thickening can affect the face but does not extend above the knees and elbows.

Diffuse

Skin thickening can affect the whole body Internal organs, including blood vessels, digestive system, lungs, heart, kidneys, muscles and joints may be involved in both limited and diffuse.

Diagnosis of Scleroderma

THE YEAR MY LIFE GOT TURNED UPSIDE DOWN

During the summer of 1997, aged 24, I had begun to feel quite unwell. There was not a part of my body, which did not ache, and by the autumn my fingers had become stiff, swollen and shiny. I went to see my GP who arranged for the standard rheumatology blood tests as well as an ANA test.

I was then referred to a Consultant rheumatologist, with by now, typical scleroderma like symptoms with skin thickness covering my entire body, which was not only tight, (I could not stretch my arms out), but very itchy. I also had difficulty with swallowing/ small mouth and stomach cramps, as well as I would turn blue at the slightest of draughts and was constantly tired.

I was relieved that I finally got a diagnosis and I was not going to let it take away my dream of being a barrister. I qualified in 2004 but, reluctantly, had to accept that the physical requirements of the role were too much for me. I now have a disciplined daily routine to keep my symptoms under control, including moisturising my skin (symptom reversal currently at 95%). As well as being respectful of my symptoms – I am always wearing gloves and have changed my diet.

I am extremely grateful for the help and support that the charity has given to myself and my family. In particular, the sharing of up to date treatments, research, survival tips, and gadgets that make the day-to-day living, easier to cope with. As well as continuing to provide hope that a cure will be found in my lifetime.

Nicola

Finding a Specialist

Now you have received a scleroderma diagnosis, it is time to consider seeing a scleroderma specialist to ensure you are receiving the best care. If you received your diagnosis from a specialist, then you are already in the right hands. Your specialist and their team will be able to offer you the best scleroderma management and will refer you to another specialist if you need to be seen by another department.

If you received your diagnosis from your local GP, it is worthwhile looking at our list of specialists, which can be found on our website. Your GP may recommend a specialist to you but if not you can ask your GP to refer you to your chosen specialist. If you need any support with finding the right specialist or receiving a referral please get in touch:
www.sruk.co.uk
020 3893 5998

SRUK Publications

For further information on Scleroderma, Raynaud's, localised scleroderma, lung involvement and gastrointestinal tract please see our publications webpage: **sruk.co.uk/publications**

To order printed copies of our publications, please call: **020 3893 5998** or email: **info@sruk.co.uk**

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**

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**

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Trusted Information Creator

Frequently Asked Questions

We know you probably have lots of questions now you have received a diagnosis but here are some of the frequently asked questions from our community answered by Professor Ariane Herrick – Professor of Rheumatology, University of Manchester; Vice President of the Scleroderma & Raynaud's UK (SRUK).

Can it be cured?

At present, scleroderma cannot be 'cured'. However, there is always something, which can be done to help. This is because there are effective treatments for many of the different manifestations of scleroderma. For example, drugs called

proton pump inhibitors are very effective for the heartburn, which is often experienced by people with scleroderma. Also, several different treatments can help Raynaud's phenomenon and finger ulcers. Ask your doctor or nurse specialist about these different treatments. Remember, 'incurable' does not mean 'untreatable'.

Am I going to die from this?

Some people do die from scleroderma, for example those with severe lung, heart or kidney involvement. However, most people with scleroderma do not die of the disease, and many live a near normal life.

Will the disease spread?

Not necessarily. However, it can do. For example, the extent of skin involvement may progress in people with the diffuse cutaneous type of scleroderma. Also, it is possible that scleroderma can go on to involve what are called the internal organs (including the lungs, heart and kidneys). For this reason it is important to report any new symptoms to your doctor, and to have regular checks (for example of the heart and lungs) at the hospital.

Is it hereditary?

We do not understand why some people but not others develop scleroderma. There is a hereditary component, but this is relatively small. To put this in context, if someone has a first degree relative with scleroderma (for example a parent, brother or sister) then the chances of that person developing scleroderma are in the order of around 1% (one in a hundred).

Will I be able to live a normal life?

This depends on the severity of your scleroderma. Many people with scleroderma lead a normal (or near normal) life, although most have to be careful to avoid cold environments. People who are more severely affected (for example those with lung involvement, or limitation in hand function) are often restricted in what they are able to do. Discuss this with your doctor or nurse specialist who may be able to help. For example, if you have problems with hand function, then it will probably be worthwhile seeing an occupational therapist who will be able to offer advice.

How is this going to affect me?

Everyone is different. It may be that you will be able to lead a normal life. However, if you have significant problems for example with your fingers, or with your breathing, then things may be more difficult. Discuss things with your doctor, so that everything possible can be done to help.

Will my symptoms improve?

Some symptoms may improve. For example, some people find that their Raynaud's improves, perhaps because they become expert in knowing how to avoid situations, which provoke Raynaud's attacks. Also, especially in patients with the diffuse cutaneous type of scleroderma, skin thickening often reduces after a number of years. However, it is possible that symptoms may worsen, and if this happens then you must seek medical advice to check things out further.

Can it prevent me having children?

Many people with scleroderma successfully have children. However, if you would like to plan a family then it is important to discuss this with your doctor. This is because there are certain situations in which it may not be wise to become pregnant (for example if you have severe disease). Also, it may be necessary to make changes in your drug treatment because some drugs should be avoided during pregnancy.

What medication is available?

Many different medications are available. For example, there are medicines available for:

- Raynaud's phenomenon and for finger ulcers
- Gut disease, including for disease of the gullet [oesophagus]
- Kidney involvement
- Muscle inflammation

In addition, there are a number of treatments other than drugs. For example, physiotherapy (with stretching exercises), and camouflage or laser treatment for telangiectases (the red spots which can occur on the face and arms). Always ask your doctor about the different treatments available.

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