

Systemic Sclerosis (Scleroderma)

What is Systemic Sclerosis?

Scleroderma is one of the more uncommon of the connective tissue diseases. These diseases affect the connective tissue of the joints, tendons, muscles, skin, blood vessels, heart, lungs, and many other organs. Although it is a chronic illness, proper treatment and management can make it possible for people with scleroderma to lead full and productive lives.

What causes systemic sclerosis?

The cause of systemic sclerosis is still unknown. It is not contagious, so you can't catch it from someone or give it to anybody else. It is believed that people with systemic sclerosis have a genetic predisposition for the illness and that other factors act as triggers for the disease onset. Most people with systemic sclerosis will not have other family members with the condition.

There are 3 aspects of the disease processes: small blood vessel damage, autoimmune inflammation and tissue scarring (fibrosis). The blood vessel damage leads to Raynaud's phenomenon and may cause ulcers. The immune system is your body's natural defence against illness. Unfortunately, it can sometimes be defective and attack the healthy body, as is the case in Systemic Sclerosis. We also know that the tissue cells of people who have systemic sclerosis produce too much of a protein called collagen, which results in scar tissue development in the skin or internal organs.

How many kinds of scleroderma are there?

Scleroderma is an umbrella term used to indicate a spectrum of disorders. There are two main types of scleroderma, localised and systemic. Often medical professionals use the term "scleroderma" when they talk about systemic sclerosis. Localised scleroderma (See our Understanding Localised booklet) only affects the skin and sometimes the underlying soft tissues, while systemic sclerosis may affect the internal organs. With systemic sclerosis, distinction can be made between the limited and diffuse form based upon the extent of skin involvement. In the limited cutaneous systemic sclerosis, it can affect the whole body. The face is usually affected in both forms of the disease. The internal organs, including the blood vessels, digestive system, lungs, heart, kidneys, muscles, and joints may become involved.

These two major forms are different but should not be regarded as mild or severe types of the same disease as they can both have serious internal organ complications or conversely patients can have a mild course in either subset.

Systemic sclerosis varies from person to person. People with the illness may have one or more parts of the body affected to different degrees. Systemic sclerosis should not be confused with multiple sclerosis. There is no connection between the two diseases.

How does systemic sclerosis start?

One of the early problems people with systemic sclerosis get is sensitivity to cold. This is called Raynaud's phenomenon and it affects most people with the condition at some point. Raynaud's phenomenon occurs when the blood vessels over-react to cold, causing the fingers and toes to turn white, blue and then go red when warmed up. This causes numbness and tingling, and sometimes even pain. Swelling and puffiness of the hands and feet is also a common early feature. The fingers may look and feel swollen, making the skin lose its normal lines and look shiny.

Who does systemic sclerosis affect?

The disease affects four times more women than men. The disease usually starts between the ages of 25 and 50 but it can affect children and the elderly.

How does systemic sclerosis develop?

Most people with systemic sclerosis have some thickening and hardening of the skin, especially of the hands, arms and face. The joints can tighten into a bent position due to tightening of the skin or tendons. It can also cause joint inflammation and symptoms of pain, stiffness, swelling, warmth and tenderness. You may also get tired quickly and the muscles may be weak. Ulcers are not uncommon and some people may develop calcium deposits in various parts of the body (calcinosis).

What can be done to help?

Although at the moment there is no proven cure for systemic sclerosis, there are multiple treatments for the different aspects of the disease. Medications (including tablets, infusions and creams) are too numerous to mention here. Special exercise programs have proved very useful in the treatment of systemic sclerosis by keeping the skin flexible, maintaining better blood flow, and keeping the joints supple. Social and psychological support is also very important.

Most people with scleroderma will experience pain at some point during their disease and some may have chronic pain. In those cases, a referral from your GP or consultant to local pain management services is important.

Take sensible preventive action

It is sensible to keep skin healthy to prevent cracking, peeling and ulcerating. This can be done by regular moisturising. A good blood supply is essential and can be helped by keeping warm. The head, hands, and feet are often the worst affected areas and should be covered up when in the cold. Hats, thermal clothing, hand warmers, electrically heated gloves/socks, and many others can help this and stopping smoking is very important. Irritants such as detergents and certain perfumed toiletries can worsen the damage to the skin while some soaps, creams and bath oils can help keep the skin supple and moist. This requires a degree of experimentation to find the products that suit you.

The disease may also affect the tissue of the internal organs. For example, if the digestive system is involved, the muscles in the gullet may become weak leading to difficulties swallowing. Heartburn can also be a frequent problem. Sometimes the middle and lower bowel are affected and this affects bowel motion. The lungs, heart and kidneys can also be affected. The limited subset of systemic sclerosis is more common than the diffuse. The disease outcome and organ complications can vary between patients from extremely mild to severe. As diffuse and limited systemic sclerosis are long-term conditions, regular tests and specialist reviews should be continuous.

The disease can change the way you look to some degree, but it does not have to change who you are!

Social and psychological implications of scleroderma

No less important are social and psychological aspects of the disease. This involves many of the health care team that are looking after you. There are often lots of things that can be done to help people cope at home despite their disabilities. A range of aids might help you do the things you can't manage. The "hidden problem" is often the emotional aspect of having a condition like systemic sclerosis. It is difficult to emphasise enough how important it is to treat this part of the disease.

BE POSITIVE

There is a lot of support available both from professionals (medication, counselling and other psychological support) and other people affected by systemic sclerosis (patient support organisations). A positive outlook is important. Open discussions can help family and friends understand the physical problems and feelings you are experiencing. Often this can help them to help you when you need it most.

Scleroderma & Raynaud's UK

We are here for you, to support you through your diagnosis and to help you understand and manage your condition.

We know how important it is to join a community that understands your condition, provides expert information and support, works for you to improve access to treatment and care whilst driving research into more effective treatments as we work to find a cure.

Ways we can support you

Our informative website has up to date, specialist information on Scleroderma and Raynaud's, personal stories from the community on managing the conditions and details on services and events that are available in your local community. You can find information, support and friendship through our online community: www.sruk.co.uk

Our helpline operates 365 days a year providing support and information to everyone affected by or interested in Scleroderma and Raynaud's: **0800 311 2756**. Our experienced helpline volunteers understand the challenges and difficulties you face and provide a confidential space to talk through worries or concerns.

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

Find out more about the work of the charity and how we can support you by visiting our website: www.sruk.co.uk or calling our friendly team on: **020 3893 5998**.

Interested in joining our community?

We have a variety of ways that you can join the community. Our most popular is to become a member. For a small subscription fee you will receive 4 magazines per year, free attendance at annual conference, priority booking for educational events and member only discounts for our shop products.

Find out more about all the ways you can join the community by visiting our website: www.sruk.co.uk or calling our friendly team on: **020 3893 5998**.

We are here for you,
when you need us.

0800 311 2756

Our helpline operates
365 days a year

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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

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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