MUSCULOSKELETAL AUSTRALIA

Scleroderma

Your questions answered

Q. What is scleroderma?
A. Scleroderma is a chronic (or long-term) condition that affects your connective tissue.

Connective tissue is the cells and fibres that hold together the millions of cells that make up your body. It ‘connects’, or supports and holds together your joints, muscles, internal organs, skin and other body tissues. Types of connective tissue include bone, cartilage, fat, blood, and lymphatic tissue.

The connective tissue of people with scleroderma has too much collagen, which causes it to become hard and tight. That’s why it’s called scleroderma – it literally means ‘hard skin’.

Anyone can develop scleroderma, but it’s more common between the ages of 30 and 60 and is more common in females. According to Scleroderma Australian, 6000 Australians have scleroderma.

There’s no cure for scleroderma, but it can be managed effectively.

Q. What causes scleroderma?
A. Scleroderma is an autoimmune disease. That means it occurs as a result of a faulty immune system.

Your immune system is designed to identify foreign bodies (e.g. bacteria, viruses) and attack them to keep you healthy. But in the case of scleroderma, your immune system mistakenly targets healthy tissue. This triggers connective tissue cells, especially a cell type called fibroblasts, to make too much collagen and other proteins. This leads to the hardening and scarring of the skin and other tissues.

We don’t know why this happens. Scientists believe a complex mix of genes and environmental factors may be involved.

Q. Are there different types of scleroderma?
A. There are two broad classifications of scleroderma based on the amount of skin and other organs affected.

Localised scleroderma mainly affects the skin. It can affect both adults and children. There are two types of localised scleroderma, morphea and linear. Both have distinctive signs and symptoms. Localised scleroderma can sometimes restrict normal joint movement due to the hardening of the skin over the joint.

Systemic scleroderma (or systemic sclerosis) affects internal organs like the heart, lungs, kidneys, and skin. *Systemic* means it affects several organs and tissues, or the entire body, rather than a single organ or body part. *Sclerosis* means abnormal hardening of body tissue. Systemic scleroderma can also be broken into sub-types depending on the symptoms. They are limited scleroderma and diffuse scleroderma.

Q. What are the symptoms?
A. The symptoms vary from person to person and will depend on the type of scleroderma you have.

Symptoms can include:
- thickening and hardening of the skin
- skin dryness and ulcers
- sudden constriction of the blood vessels, resulting in pale, blue, or red fingers or toes (Raynaud's phenomenon)
- stiffness and pain in the muscles and/or joints.

If some of your internal organs are involved, you may experience:
- digestive issues, such as heartburn, bloating, constipation, stomach pain
- lungs problems, such as shortness of breath
- heart problems, such as chest pain
- kidney problems may lead to high blood pressure.

While these symptoms sound very scary, your doctor and specialist/s will monitor your condition and symptoms to prevent/treat complications.

Scleroderma Australia has many excellent resources to help you understand and live well with scleroderma. Understanding and managing scleroderma provides info about the types of scleroderma and their symptoms in greater detail.

Q. How is it diagnosed?
A. Scleroderma can be a difficult condition to diagnose. Symptoms vary significantly from one person to another and are similar to those of other conditions. They can also change or fluctuate. So, it may take some time to get a definitive diagnosis of scleroderma.

No single test can diagnose scleroderma, so your doctor will use a combination of tests to confirm your diagnosis. They may include:
- Your medical history.
- A physical examination.
- Blood and urine tests.

Q. How is scleroderma treated?
A. While there's currently no cure for scleroderma, medicines can help ease your symptoms. Your doctor will develop a treatment plan based on your symptoms. You'll generally start on the lowest possible dosage and, if necessary, slowly increase the dose until your symptoms are under control. All medicines have side effects, so you should discuss these with your doctor.

Medicines may include:
- Topical corticosteroid (steroid) creams that you apply to your skin.
- Anti-inflammatory medicines to help manage pain and control inflammation.
- Corticosteroids taken as a tablet, injection, or into the vein (intravenous or IV) act quickly to control or reduce inflammation and may be used in the short term. They aren't used for long periods as they're associated with serious side effects.
- Medicines that suppress your overactive immune system and control symptoms. These medicines may also be in the form of topical creams, tablets, injections, or IV infusions.

As well as seeing your GP, you'll likely see a skin specialist (dermatologist) and a specialist in joint and muscle conditions (rheumatologist). Other healthcare providers, like respiratory physicians and gastroenterologists, may participate in your ongoing treatment.

Q. What else can I do to control my symptoms?
A. There are things you can do to manage your scleroderma, including:

Learn about scleroderma. Knowing as much as possible about your condition means that you can make informed decisions about your healthcare and actively manage it.

Lifestyle changes. Avoid exposure to cold temperatures, dress warmly and don't smoke. This will help you manage Raynaud's phenomenon.

Stay active with regular exercise. Exercise will reduce tightness and help keep your joints moving, especially those areas affected by scleroderma, such as the hands and wrists. Exercise will also strengthen your muscles and improve overall health. Talk with a physiotherapist or exercise physiologist about an exercise program tailored to your condition and symptoms.

See a hand therapist. They're qualified physiotherapists or occupational therapists with extensive knowledge and skill in understanding and treating problems with the fingers, wrists, elbows and shoulders. They can help you keep your hands and wrists flexible and moving well.

Aids and equipment. Scleroderma can make some actions difficult, for example, using door handles or getting dressed. An occupational therapist can advise on aids and other gadgets that may help and provide additional solutions to make life easier.

Eat well. Eating a balanced diet can help provide you with better energy levels, help to maintain your weight, and give you a greater sense of wellbeing.

Get support from others. Research has shown that people with positive social support cope better with pain. Family, friends, colleagues, and health professionals can help you manage. A peer support group may be another option. Contact Scleroderma Australia for details on support options in your state or territory.

Q. Why are sunflowers associated with scleroderma?
A. According to Scleroderma Australia, just as sunflowers follow the sun for warmth, people with scleroderma find the warmer weather more comfortable. That's why sunflowers have become a hopeful symbol that represents scleroderma in many parts of the world, including Australia.

Q. Where can I get more help?
A. Many people and support organisations can help you manage your musculoskeletal condition. They include:

- your doctor
- your rheumatologist
- dermatologist
- physiotherapist
- exercise physiologist
- occupational therapist
- Musculoskeletal Australia | msk.org.au | National Arthritis and Back Pain+ Help Line: 1800 263 265

Q. How can Musculoskeletal Australia help?
A. Our nurses are available for you to speak with about scleroderma, pain or any other musculoskeletal issues you have. You can contact them on weekdays between 9am-5pm. Phone 1800 263 265 or email helpline@msk.org.au. We also have a range of services – including free webinars – you can access on our website.

More to explore

- Scleroderma Australia

- Scleroderma
  American College of Rheumatology
- Scleroderma
  Arthritis Foundation
- Scleroderma
  Healthdirect
- Scleroderma Patient-centered Intervention Network (SPIN)
- Systemic sclerosis (Scleroderma)
  Versus Arthritis

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