



[Authoritative facts](#) about the skin from the [New Zealand Dermatological Society Incorporated](#).

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Systemic sclerosis

Systemic sclerosis (systemic scleroderma) is a multisystem disease that results in fibrosis and vascular abnormalities in association with autoimmune changes. These lead to breakdown of the skin, subcutaneous tissue, muscles and internal organs (e.g. digestive tract, heart, lungs and kidneys). The skin becomes thickened and tightly bound to underlying structures.

Localised scleroderma (also known as [morphoea](#)) is an unrelated skin disease and is confined to the skin.

Features of systemic sclerosis



Calcinosis



Calcinosis X-ray



Sclerodactyly



CREST syndrome



Ulcerated and resorbing fingertips



Vasculitis

Who gets it and what is the cause?

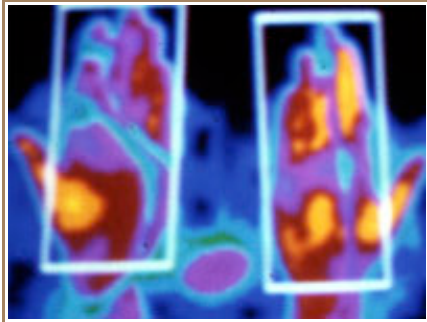
Systemic sclerosis is a rare condition that may occur in people of any race, although it is less common in people of Asian descent. It appears to be three to four times more common in women than men and is comparatively rare in children. It usually starts between 30–40 years in women and later in men.

It is classified as an autoimmune disease of an unknown cause. This means the immune system is reacting against one's own tissues. It appears to involve some injury to the cells that line blood vessels (endothelial cells) and this results in excessive activation of the dermal connective tissue cells, the fibroblasts. Fibroblasts normally produce collagen and other glycosamine proteins. Certain factors have been identified that may trigger the disease. These include injury, drugs (e.g. vitamin K, cocaine, penicillamine, appetite suppressants and some chemotherapeutic agents), and chemicals (e.g. silica, organic solvents, pesticides, aliphatic hydrocarbons and epoxy resin).

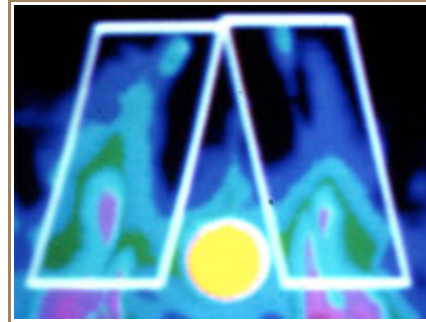
What are the signs and symptoms of systemic sclerosis?

Raynaud phenomenon is usually the first symptom of systemic sclerosis. Patients experience episodes of vasospasm, which causes blood vessels in the fingers and toes to constrict. As less blood is reaching these extremities the skin changes colour to white and the fingers and toes may feel cold and numb. As they warm up, they go blue and then red before returning to normal again.

Thermal imaging (heat sensing)



Normal hand



Raynaud phenomenon

Other skin changes include:

- Itchy skin
- Thickening of the skin of the fingers, then atrophy (thinned) and sclerosis (scarring). The fingers become spindle-shaped (sclerodactyly) from resorption of the fingertips.
- Fragile nails become smaller with ragged cuticles
- Taut, shiny skin that may have dark or pale patches (hyper- or hypopigmentation). The tight skin may affect most parts of the body, including the face, resulting in loss of expression and difficulty opening the mouth properly.
- Visibly dilated blood vessels (telangiectases) appear on the fingers, palms, face, lips, tongue and chest.
- **Calcinosis** (calcium deposits) develops in the skin, particularly the fingers, hands and other bony areas. These can breakdown and discharge chalky material.
- Ulcers may follow minor injuries over the joints, or on the tips of fingers and toes where the circulation is poor. Ulceration can lead to dry gangrene and eventual loss of the tips of the fingers (like frost bite).
- Ulcers may also arise over calcinosis and on the lower legs.

In addition to the skin changes, the disease affects many other organs. Problems that may occur include:

- Friction rubs over the joints and tendons, particularly the knees.
- Eye changes with tightness of lids, reduced tear secretion, retinopathy
- Joint pain, muscle pain and weakness and limited movement resulting in contractures.
- The digestive tract may be affected throughout its length. Oesophageal reflux is common causing difficulty in swallowing solid and liquid food. This can lead to nausea, vomiting, weight loss, stomach cramps, diarrhoea, constipation and bleeding.
- Lung and heart involvement may manifest as shortness of breath, high blood pressure, chest pain, pleurisy, pneumothorax, pericarditis arrhythmias, general heart enlargement and heart failure.
- Progressive kidney disease resulting in proteinuria, high blood pressure and eventually renal failure.

What is the CREST syndrome?

CREST syndrome (also called CRST syndrome) is a limited form of systemic sclerosis in which there is **C**alcinosis, **R**aynaud's phenomenon, **E**soophageal involvement, **S**clerodactyly and **T**elangiectases. Characteristically, the

telangiectases are well-defined and flat (matt).

What tests should be done?

The diagnosis is generally made from the patient's history and the findings on examination of the skin and other organs. A [skin biopsy](#) is not usually necessary but characteristically shows excessive ground substance and odd-looking endothelial cells in the dermis and later deposits of collagen. The epidermis is usually atrophic.

Up to 90% have elevated antinuclear antibodies (ANA) but these are less frequent than in the more common connective tissue disease, systemic [lupus erythematosus](#). Thyroid antibodies may occur and result in an under-active thyroid gland

Anticentromere antibodies are characteristic of CREST syndrome and may be present in Raynaud phenomenon before systemic sclerosis appears. Scl-70 is unique to systemic sclerosis and is more likely to be associated with severe systemic sclerosis involving the lungs. Many other less specific antibodies have been reported to be associated with different patterns of disease.

Anaemia, raised sedimentation rate (ESR) and increased gamma globulins (hypergammaglobulinaemia) and varying immune abnormalities are quite common especially positive rheumatoid factors.

What is the treatment of systemic sclerosis?

There is no cure for systemic sclerosis and treatment is aimed at controlling symptoms and preventing complications. Because the symptoms of systemic sclerosis are so diverse a team of medical specialists is usually necessary.

It is absolutely essential to discontinue smoking.

Organ	Treatment
Skin	<ul style="list-style-type: none"> • Topical corticosteroids and emollients for itchy skin • Avoid cold temperatures and wear warm clothing including gloves and socks to prevent Raynaud phenomenon. Medical treatment includes calcium channel blockers, aspirin and vasodilating drugs. • Calcinosis may be treated with calcium channel blockers, anticoagulants, colchicine and intralesional steroids and possibly excision of deposits • Research suggests intensive ultraviolet radiation treatment (phototherapy) with UVA1 (340–400nm) can soften fibrosis. This is not yet available in New Zealand • ciclosporin may help some patients
Joint and muscle	<ul style="list-style-type: none"> • Oral corticosteroids • D-penicillamine
Gastrointestinal	<ul style="list-style-type: none"> • Proton pump inhibitor, e.g. omeprazole • Cisapride • Metoclopramide • Surgery for strictures
Kidney	<ul style="list-style-type: none"> • ACE inhibitors • Dialysis

Related information

References:

Textbook of Dermatology. Ed Rook A, Wilkinson DS, Ebling FJB, Champion RH, Burton JL. Fourth edition. Blackwell Scientific Publications.

On DermNet NZ:

- [Systemic diseases](#)
- [Connective tissue diseases](#)
- Localised scleroderma ([morphoea](#))

Other websites:

- [Scleroderma Foundation](#)
- [Scleroderma Research Foundation](#)
- [Arthritis Foundation of New Zealand](#) – Scleroderma Support Group
- [Systemic sclerosis](#) – emedicine dermatology, the online textbook

Books about skin diseases:

See the [DermNet NZ bookstore](#)

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DermNet does not provide an on-line consultation service.
If you have any concerns with your skin or its treatment, see a [dermatologist](#) for advice.

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