

# Scleroderma and the kidney

## What is scleroderma?

Scleroderma means 'hard skin', but it is a disease that can affect many other organs, when it may be called systemic sclerosis or systemic scleroderma. It is a rare disease, but it is much more common in women than men, and most often develops between the ages of 30 and 60. Rarely it occurs in children.

Sometimes scleroderma affects only one area of the skin – localized scleroderma. But usually it affects many areas, when it is particularly visible as tight skin over the face and fingers.

**Systemic sclerosis** may also affect gut, lung, and other organs including kidneys.

**CREST** is a condition that overlaps scleroderma, but it is more limited. It stands for **C**alcinosis (calcium deposits in skin), **R**aynaud phenomenon (fingers etc go cold and painful), **E**osophageal dysmotility (swallowing difficulties), **S**clerodactyly (tightening of skin over the fingers), **T**elangiectasia (visible blood vessels on skin).

## How are the kidneys affected in scleroderma?

Scleroderma mainly affects the blood vessels of kidneys. In **scleroderma renal crisis**, there is a sudden and severe rise in blood pressure and decline in kidney function over days or weeks. It may lead to complete kidney failure. It is not known what causes the crisis to develop. It occurs in about 5% of patients with scleroderma

Renal crisis used to be a leading cause of death in scleroderma, but this has been changed by use of Angiotension Converting Enzyme Inhibitors (ACE inhibitors), drugs which are commonly used now to treat high blood pressure.

Less acute kidney disease can also occur sometimes in scleroderma. Some patients may only have some protein in their urine and slight impairment of their kidney function.

## How is the diagnosis made?

Scleroderma is often suspected from the symptoms combined with the skin

changes. Blood tests to look for certain antibodies (for instance anti-Scl-70 antibodies, anti-centromere antibodies) may help, and can help predict prognosis.

Scleroderma kidney disease usually has very few early signs, and sometimes it occurs in people who are not known to have scleroderma, and who don't have all of the classic symptoms and signs of systemic scleroderma. Urine tests are often normal, and losing a little kidney function does not cause symptoms. A rise in blood pressure is probably the first sign. The blood pressure rise can be severe enough to cause severe damage in the kidneys and elsewhere too - including the eyes and sometimes the brain. This is called **malignant or accelerated phase hypertension**. [More info on malignant hypertension.](#)

Proving the diagnosis, and showing how severe the damage is, may require examination of a piece of kidney - a kidney biopsy. [More information about kidney biopsies.](#)

It is important to have your blood pressure checked regularly if you have scleroderma.

## **What causes scleroderma?**

It is not known what causes scleroderma but it is believed to be an autoimmune disease, in which the body's immune system attacks itself. That kind of process causes a number of important diseases - thyroid diseases, juvenile diabetes, rheumatoid arthritis, and many kidney diseases.

## **What is the treatment?**

Treatments for scleroderma vary according to the problems you have. Various drugs (as well as warm gloves) may help Raynaud's syndrome, for instance. Lung disease may require specific treatments, and this and other severe disease may be treated with immunosuppressive drugs to prevent severe scarring and lung damage. [More information about immunosuppressive drugs used in kidney diseases.](#)

**Treatment for kidney disease** focuses on blood pressure control using ACE inhibitors. These have transformed the outlook for people with a 'renal crisis', which often used to lead to the death of those who got it. One study has shown that up to 75% of patients with scleroderma renal crisis will survive at least one year with ACE inhibitor treatment. Angiotensin receptor blockers (ARBs) are an

alternative for some people who develop side effects from ACE inhibitors. Dialysis is often required – temporarily in about 20% of cases and long-term in about 40% of cases.

For people with severe kidney damage, the information below about chronic kidney disease, and treatment options should dialysis become necessary, will be important.

### **Further information**

Very useful information is available from the [Scleroderma Society \(UK\)](#)

There is also excellent information on the University of Maryland Medical Center (USA) website:

[What is scleroderma](#) gives general information and links to very full information on causes, treatment, and many other things. More information on renal scleroderma is on their page on [How serious is scleroderma](#) – go down the page to ‘Kidney Complications’. This page also has extensive information on lung, gut and other complications.

Relevant information on other pages of this website:

More information about [chronic kidney disease and its treatment](#)

More information about [End stage renal failure and its treatment](#)

More information on [high blood pressure and the kidney](#).

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