

Outcome

The best treatment can be administered if the patient comes to hospital as soon as possible after the crisis begins. If the kidneys are only slightly damaged by the time the patient is referred, dialysis can be avoided and the kidneys can recover back towards normal. If the kidneys fail (in the case of around 50% of those who develop a crisis) then dialysis can be done. However dialysis is often technically difficult because of the other problems the patients have.

Happily, some patients' kidneys (about 50% of those who need dialysis) recover even after needing dialysis. This recovery occurs up to two years after the crisis. Hence in this period it is not recommended to have a kidney transplant, as there is a possibility it might not be needed.

For those who do not recover, kidney transplantation is possible as long as the scleroderma has not affected other organs badly enough to make the operation and anaesthetic unsafe. It is unusual for the scleroderma renal crisis to come back in the transplanted kidney.

Unfortunately, about 10% of patients who develop a kidney crisis are so ill by the time they get to hospital or their other organs tolerate high blood pressure so poorly that they do not survive the kidney crisis. Those who do get over the renal crisis however are very unlikely to have it happen again. However, these figures are improving all the time with the development of new drugs and better monitoring methods.



The **Scleroderma Society** supports people with scleroderma and their families by providing:

- educational literature
- a telephone helpline
- a comprehensive website & forum
- a newsletter with research information
- member contact
- informal group meetings
- an annual conference

We also work to promote awareness of scleroderma among the medical profession and general public in order to improve early diagnosis and prognosis. We fund medical & scientific research in the UK and are a founder member of FESCA (Federation of European Scleroderma Associations), working to forward the cause of people with scleroderma throughout Europe.

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Scleroderma and the Kidney

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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.

Scleroderma and the kidney

Systemic sclerosis can affect many organs in the body. The skin changes are the most obvious but other organs such as the lungs and oesophagus can be involved. If these organs are affected, it usually leads to symptoms such as breathlessness and difficulty swallowing, which alert the patient and doctor that these organs are involved. The kidneys can also be involved in scleroderma but very often the patient is unaware of a problem.

The kidneys

Each kidney weighs about 150g and is located at the back of the abdomen protected in part by our ribs from behind. They are very hard working organs taking about one fifth of the blood pumped by the heart every minute and cleaning it of wastes and water. Although this cleansing function is the best known job of the kidneys they also perform many other very important functions. These include controlling the production of red blood cells, the strength of bones, the acidity of the blood and, very importantly, the kidneys help to control blood pressure.

Controlling blood pressure

The blood supply to the kidneys is crucial in this task. If the kidneys are not getting enough blood they set in motion a train of events which raise the body's blood pressure to increase blood supply to the kidneys.

A small proportion of scleroderma patients suffer from

slightly high blood pressure which is easily controlled with medication and they may have some protein leaking into their urine. These patients do not worry us so much, as everything is under control.

However, about 5-10% of patients with scleroderma (usually, but not only, those whose skin is worsening fast) develop severe uncontrolled high blood pressure which results from blood vessel changes within the kidney (something like Raynaud's phenomenon) which over a short period of time starves both kidneys of blood and oxygen and sets in motion the "normal" response of raised blood pressure. In this case, the high blood pressure does not return to normal and continues to rise and rise, eventually destroying the kidneys and damaging the heart and lungs of the patient if it is not treated. This is called a scleroderma renal crisis and needs expert medical attention as soon as possible.

Symptoms

The beginnings of a crisis may be noticed when blood pressure is checked at the patients' GP surgery or at a hospital clinic. If the doctor finds protein leaking in the urine, which was not present before, he/she may also become suspicious that all is not well with the kidneys.

The high blood pressure often causes the patient to have a severe headache or blurred vision. Breathlessness, nausea and vomiting may also ensue. Often urine output remains much the same until the problem is quite advanced. A feeling of palpitations or fast beating heart may also occur. Some patients have seizures caused by the very high blood pressure.

Investigations

A patient undergoing a scleroderma kidney crisis needs urgent admission to hospital.

First the diagnosis needs to be confirmed and the severity assessed. This involves blood tests to measure the level of wastes in the bloodstream and to see how badly the kidneys have been damaged. Chest X-rays, cardiographs and urine tests need to be done. The back of the patient's eyes need to be examined with a special instrument (an ophthalmoscope: similar to that used for looking into patient's ears) to see if there are signs of very high blood pressure.

Later, when blood pressure has been brought under control, a kidney biopsy (taking a tiny piece of kidney with a needle through the back) may need to be done to see how much, if any, recovery of kidney function can be expected.

Treatment

The mainstay of treatment is to lower the patient's blood pressure. This is achieved by using a combination of drugs by mouth and into the vein. Patients will need to take the blood pressure tablets for many years.

Sometimes, in special circumstances, blood pressure needs to be lowered quickly but, generally, slower reductions in blood pressure over 10-14 days are aimed for.

Some of the newer medicines used are very effective at lowering blood pressure but they may make the patient feel flushed. Some painkilling drugs are harmful to the kidneys and need to be stopped if the patient is taking them. The patient's kidney function and blood pressure needs to be checked daily and dialysis (artificial removal of wastes and water from the body) can be started if the kidneys fail completely.