

Scleroderma

WHAT IS IT?

Scleroderma is a chronic, often progressive auto-immune disease in which the body's immune system attacks its own tissues. It predominantly affects the skin, but can also affect different parts of the body with symptoms varying enormously from a minor irritation to a life-threatening illness. Symptoms can generally be well managed with appropriate treatment.

WHY DOES IT OCCUR?

The exact cause of scleroderma is unknown. Some research suggests that problems in the connective tissues (tissues that hold together muscle, blood vessels, fat and skin) can result in the tissue becoming damaged and replaced by scar tissue.

WHO DOES IT AFFECT?

Scleroderma affects more women than men. Onset usually occurs between the ages of 20 and 40 years but it can occur at any age. It does not appear to be inherited. Scleroderma may affect the skin alone. This is known as localised scleroderma and occurs either as patches of thickened skin, known as morphea, or as linear scleroderma, a line of thickened skin that may extend along an arm or a leg. Systemic scleroderma affects the internal organs and is generally divided into two types, the first called limited scleroderma, in which only the skin below the elbow and on the face are affected and diffuse scleroderma when skin all over the body is affected.

SYMPTOMS

Symptoms vary greatly and also depend on what part of the body is involved. They may include:

- thickening of the skin, particularly on the fingers, arms and sometimes face
- colour changes in the hands and feet, usually from pale to blue to red, often after exposure to the cold (Raynaud's disease)
- small calcium deposits in the form of nodules on the fingertips and bony prominences
- stiffness in the muscles and joints
- indigestion or heartburn
- diarrhoea or constipation
- lung or kidney impairment.

DIAGNOSIS

There is no single test for scleroderma. Diagnosis will usually involve a physical examination and medical history and may include further tests such as blood tests and sometimes a skin biopsy. About 95 percent of scleroderma sufferers test positive to an antinuclear antibody (ANA) test.

MANAGEMENT

Management will depend on individual symptoms. It may include medication to slow the progression of skin involvement or to decrease the severity of Raynaud's disease. Special medication may be started if the lungs or kidneys are involved. Raynaud's

disease may be reduced by avoiding changes in temperature and by using gloves and other warming devices. Exercise preserves range of motion and improves overall health.

OUTLOOK

For most people scleroderma is not systemic or progressive. For many, scleroderma presents as a mild skin condition. However some people may have involvement throughout the body. Like other forms of arthritis, there is no cure for scleroderma but there are many treatments for specific symptoms.