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The website for the answer to all your Orthopaedic Questions

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Scleroderma

Link - <http://www.arc.org.uk/arthritis/patpubs/6036/6036.asp>

Key words: scleroderma, connective tissue disease, systemic sclerosis, vasculitis

What is scleroderma?

'Scleroderma' means 'hard skin'. However, many people with scleroderma have problems not only with their skin but with other parts of their bodies as well. This is the reason why it has a second name, 'systemic sclerosis'.

Scleroderma is an uncommon, chronic (persistent) disease. It affects the connective tissues which surround the joints, blood vessels and internal organs beneath the affected area of skin. Women are affected 3-4 times more often than men. The disease usually starts between the ages of 25 and 50. It only occasionally begins in children or in the elderly. Although there is no cure, proper treatment and care can make it possible for people with scleroderma to lead full, productive lives. There may however be some limitation on your activity.

What are the two types of scleroderma?

There are two major types of scleroderma:

- **Localised scleroderma (sometimes called 'morphoea')** In this condition changes only occur in isolated areas of the skin and the tissues beneath it. It is relatively mild and does not affect internal organs.
- **Systemic sclerosis** With this condition changes may occur in the skin and also in a number of internal organs. These might include blood vessels, joints, the digestive system (oesophagus, stomach and bowel), and occasionally the lungs, heart, kidneys and muscles. Changes in the connective tissue may affect the function of any of these organs.

Scleroderma differs from person to person. It is hard to predict how the disease will develop in each individual, but it usually affects only a few parts of the body, and it often stabilises after a few years. How badly affected you are depends on which

organs are affected. People who have the localised disease do not develop generalised scleroderma.

What is the cause of scleroderma?

The cause of scleroderma is unknown. It is not contagious and it is not passed on directly from one generation to another. However, as is the case with many other diseases, some families seem more likely to get it than others. The connective tissue cells of people who have scleroderma produce too much of a protein called collagen. Collagen is essential for holding the body together, but if there is too much the body becomes stiff and is unable to function properly.

The small blood vessels are also damaged in scleroderma. There may be a link between the build-up of excess collagen and blood vessel changes. The excess collagen is like scar tissue. It causes thickening and stiffening of those parts of the body it affects. The immune system may be important in causing collagen to be deposited. It occasionally seems to be triggered by some unusual chemical exposure. Scleroderma is probably caused by a combination of different genetic and environmental factors.

How does it start and what effects does it have?

Scleroderma is a variable, slow, and long-term disorder. It usually starts slowly, gradually gets worse over a few years, and then stabilises. Sometimes it progresses faster. Occasionally it seems to cure itself. Its severity and effects vary. However, severe disability with scleroderma is uncommon.

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