

impaired, giving difficulty in swallowing, a number of drugs are helpful; again the specialist treating you will advise. Very occasionally, if the muscles become inflamed or weak, it will be necessary to consider corticosteroids, either giving them as a trial intravenously, or by mouth.

All drugs do have side effects and because of this they have got a poor image. However, they are extremely useful when prescribed carefully by clinicians understanding how to monitor this disease.

#### **Other Problems**

There may be a considerable time before the correct diagnosis is made and referral to a specialist who knows about scleroderma in young people is achieved. This is because it is so rare.

It is very unlikely that any other young person in your doctor's practice has scleroderma; similarly there is probably no one else in your school with it or who knows anything about it!

It is not always easy to have something odd the matter with you while you are still at school or college. Frequently there is a period of frustration until the diagnosis has been made and obviously

there is a certain amount of anxiety and depression when a young person is not able to join in sport or has disabilities that are obvious; these can be quite upsetting. Adjusting to the disease is very personal, between the patient and their families; they learn to cope in their own way. Sometimes it is shock or denial, then anger before realising that they have to collaborate in treatment. Depression is also a natural reaction and it is important that you seek appropriate support for this. There is no special diet which will cure scleroderma, but it is important that the diet is balanced with all the proper nutrients in it.

It is important to beware of unproven remedies and to discuss anything of this nature with the consultant who is looking after you.

There is no known drug which "cures scleroderma", but early diagnosis and appropriate care along the lines outlined here will do a great deal to help in controlling the disease and keeping you in the normal stream of school and college life.



# **Scleroderma Society**

Supporting people with Scleroderma for over 25 years

# **Scleroderma in Young People**

You can get more information about Scleroderma from:

**The Scleroderma Society**  
**PO Box 581, Chichester, PO19 9EW Tel: 020 7000 1925**

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## Introduction

Scleroderma, which literally means “hardening of the skin”, is a rare disease and is very uncommon in children and teenagers. Because of this it may take some time to be diagnosed.

The purpose of this leaflet is to tell you and your parents something about it. If you go and look it up in your local library, what you read may sound horrific and probably doesn't apply to you anyway!

There are several forms; sometimes it is localised, but it can be generalised and affect not only the skin and some of the other tissues under the skin. Blood tests will be important in sorting out the pattern of illness and the risk of complications.

Who gets Scleroderma? Anyone can, there is no familial incidence, however it is more common in girls than boys; it can occur in any race.

## Localised Scleroderma

This is the commonest form under the age of 16 years, two major types are recognised:

### 1) Morphoea

This is the name given to patches of skin change which may be rounded or oval. The inflammation in the skin makes the patch feel firm and it has a violet colour. These patches are often on the trunk but can occur on the arms or legs and very occasionally on the face. As time passes the mauveness will fade, as long as there is a mauve edge it means that inflammation is still present. Ultimately these patches pass through a brown phase and become white over 2 to 3 years. Usually there are only a few patches, but sometimes there are many, when it is called generalised.

In young patients aged 3 to 10 the skin lesion can be complicated by little nodules in the tendons over joints making them stiff to

move, so sometimes this is mistaken for arthritis.

### 2) Linear Scleroderma

This is the name given to a band-like thickening of the skin which affects particularly the thigh or the lower leg, upper arm or forearm, occasionally the whole arm or leg is affected and rarely both the arm and the leg on the same side. The skin has a firm band on it often with a surrounding violet border.

This type of scleroderma sometimes involves the face or scalp when it is referred to as “Scleroderma en coup de Sabre”, as the scar resembles that caused by a knife wound or a sword wound.

When the band is very firm and occurs over a joint, it can cause bending of the wrists, fingers, ankle or foot as well as an elbow or knee. Very occasionally near the skin lesion on the arm or leg, a joint may become swollen not necessarily where there is a skin lesion.

Linear scleroderma may seem to start after a minor accident, and all forms have been described as developing after viral infections such as chicken pox, otherwise the cause is unknown. To date, efforts to incriminate other germs or infections have so far proved negative. Occasionally the limb does not grow at the same speed as the other side so that leg length and shoe size or hand size may be different on the two sides.

## Management

In the management of local disease it is important for all lesions to be carefully noted and also at times to measure the size of skin lesion as well as the length of the affected limb and the opposite limb. Monitoring in this way is very important to assess the state of the illness and its response to treatment.

For just one or two small patches of morphoea all that will be needed will be a local cream. If

it becomes more generalised other treatment may need to be considered, but this is rare. As linear scleroderma affects not only the skin but also the subcutaneous tissue and fat under the skin lesion, there is a risk of it causing serious deformities, particularly when it causes a joint to bend and failure of the whole limb to grow. Sometimes it will be necessary to have a splint, particularly for night use to prevent the knee bending or to hold a wrist in a good position.

Physiotherapy is very important in maintaining muscle strength and range of joint movement. If there is a very localised tight area of skin, massaging with an oil such as coconut or olive may improve the elasticity.

Research using a machine called a thermograph has shown that these lesions are often hot, that means they are inflamed, and therefore the question of starting therapy with intravenous Methylprednisolone might need to be considered.

If there is an arthritis or joint contracture, an immunosuppressive drug such as Methotrexate may be recommended. Sometimes injections of corticosteroid may be helpful.

All drugs have side effects and therefore have to be monitored by regular blood testing. Because of the alteration in growth, shoe size and leg length may be different, which may need an insole in the shoe or a raise on the shoe on the affected side.

Immediately before and during puberty when the overall growth spurt occurs, the good leg can become considerably longer than the affected one and then the question of whether to slow the growth of the good leg for a period may arise. This is very rare, but must be done before growth is completed. It will require discussions between rheumatologist and the

paediatric orthopaedic surgeon.

## Raynaud's Phenomenon

Some children with Localised Scleroderma develop Raynaud's phenomenon, that is constriction of blood vessels in fingers and toes, causing them to turn blue. This does not mean that they have developed a systemic form of scleroderma, although some additional tests may be necessary.

## Adult type systemic sclerosis

This affects girls more frequently than boys and is characterised by involvement of the blood vessels causing Raynaud's phenomenon as described previously, difficulty in swallowing, pain in muscles and joints and sometimes shortness of breath due to lung involvement occurs. It requires careful monitoring to assess the function of the lungs, kidneys and heart, to detect complications as early as possible.

## Management

If Raynaud's phenomenon is present vasodilator drugs such as nifedipine should be given. If there are tendon contractures, physiotherapy and non-steroidal anti-inflammatory drugs will be required. Immunosuppressive drugs such as Methotrexate may be necessary to treat the skin or internal organs.

In those few adolescents who develop a high blood pressure, the kidneys may be at risk, and drugs to reduce the blood pressure known as anti-hypertensives will have to be used. Should kidney function start to deteriorate, other drugs including Iloprost or ACE inhibitors will be used. Lung involvement may need corticosteroids on alternate days and immunosuppressive drugs.

Assessments for these drugs must be undertaken by people knowing about scleroderma and how to monitor severe cases. When movements of the oesophagus are