

The Skin in Systemic Sclerosis (Systemic Scleroderma)

All patients with systemic sclerosis will develop some skin changes. However, the severity and the problems created by the changes will vary greatly between patients.

Problems include:

- Tightening of the skin around the fingers, the face and other areas of the body causing contractures (fixed joints);
- ulceration;
- dryness and irritation;
- broken blood vessels (telangiectasia);
- calcinosis

In this leaflet we aim to tell you what can be done to tackle each of the above.

SKIN TIGHTENING

Tightening of the skin is a fundamental problem in systemic sclerosis. There are several medicines which may be of benefit for this problem, and these would normally be prescribed under the supervision of a hospital specialist. New medicines are constantly being developed, which one day may provide a safe and effective cure for this disease.

In addition to drug treatments, physiotherapy and exercises are important to maintain skin movements around joints. Warm paraffin wax baths can also be useful to relieve tight, painful hands and to enable easier exercising. Soaking in warm water with a few drops of a liquid paraffin-based oil may help to soothe and relax painful hands and feet.

ULCERATION

It is not uncommon for patients with systemic sclerosis to develop an ulcer. These are commonest on the fingers and toes. Ulcers may also occur on the legs, often following trauma such as a knock with a shopping trolley. It goes without saying that one should take care to avoid such knocks, but accidents do happen! However, wearing trousers rather than a skirt, or a thicker pair of tights or socks may be enough to prevent the skin from being broken.

If a leg ulcer does develop it can take longer to heal in a patient with systemic sclerosis because the skin is not as healthy. Ulcers should be dressed regularly; the frequency with which the dressings are changed will vary from patient to patient. Sometimes an ulcer requires daily dressing but often two to three times per week is sufficient. Dressings help in several ways; they alleviate pain, contain any discharge, and prevent dirt and infection from entering the wound. It is also believed that there is an optimum moisture level to help with healing. An ulcer should be neither completely dry, nor too wet and sloughy. There are many different dressings available to try and provide the correct moisture level at the wound bed. Again the choice of dressing will be dictated by the type of ulcer.

For ulcers on the fingers and toes a dry dressing is often all that is needed. Healing may be promoted by helping the blood to circulate freely using drugs such as vasodilators which can be given in oral forms such as Nifedipine, or intravenously e.g. prostacyclin (Iloprost®). The intravenous form is only available from certain specialist rheumatology units, but can have good effects on difficult ulcers on the fingers and toes. Ulcers may sometimes become infected and will require a course of oral antibiotics such as flucloxacillin. Signs that an ulcer has become infected are increasing pain, redness, discharge and smell. By eliminating infection one may be able to speed up healing. Ulcers can also develop eczema (inflamed skin) around them which may require a short course of steroid cream.

DRYNESS, ECZEMA AND IRRITATION

Many people, not just people with systemic sclerosis, suffer from dry and irritable skin. In general terms dry skin is not healthy skin. Regular daily use of moisturisers can solve this problem. Moisturisers are either creams or ointments. Creams are based on water which means they are pleasant to use and soak into the skin well. However, they are not usually as effective as ointments which form a barrier on the skin and help to trap in your skin's natural moisture.

Ointments are greasy and not as nice to use as creams. "Greasy" creams such as Diprobase® offer the best of both worlds and are to be recommended. Whilst there are many cosmetic moisturisers on the market, many containing extra additives, these are often very expensive and there is little evidence they offer any special benefit. Products containing more than 10% urea can be effective but can sometimes be irritating.

Bath emollients can be very helpful in keeping the skin moist, but beware as they can make the bath slippery. It is good practice to apply your chosen moisturiser once you have dried yourself after a bath.

CALCIUM DEPOSITS

If calcium deposits cause you no discomfort, then it is quite safe to leave them alone. However, if they are unsightly, cause discomfort or ulcerate and become infected, then they can be removed by simple surgical techniques. Unfortunately, there is no guarantee that the deposits will not reappear. Occasionally these calcium deposits can discharge from the skin spontaneously. Paraffin wax baths may encourage this.

In summary, skin problems are common in systemic sclerosis but there are simple practical solutions to many of them.

Itch is another common problem for some people with systemic sclerosis. If regular moisturising as outlined above does not help, then other measures can be tried. Several creams are available with specific ingredients aimed to combat itch. These include Eurax cream®, E45 anti-itch cream®, Balneum Plus Cream® and Xepin Cream®.

In addition, antihistamine tablets may be tried. Some antihistamines can cause drowsiness and one needs to avoid driving etc. after they have been taken. However, it tends to be the antihistamines that cause mild sedation such as Atarax® which are the best at helping with itching, especially at night.

During the day, a non-sedating antihistamine such as Clarytin® could be tried. People often need to find which antihistamine suits them by trying several different ones. One further treatment which can be used is ultraviolet light therapy. This is only available in hospital dermatology units.

Self-treatment with home ultraviolet lamps is not advisable as the overall dose would not be monitored and one could cause lasting damage to the skin. An eight-week course of therapy can sometimes help with itching, although benefits may only be temporary.

TELANGIECTASIA

These burst blood vessels visible as small red spots may develop on the face and hands. Understandably, many patients are keen for treatment. Pulsed dye laser treatment can often be helpful. This new technology utilises short bursts of high energy light to obliterate the broken blood vessels. This treatment will cause some stinging sensation on the skin and will leave some bruising for seven to ten days afterwards. It may take three to four treatments to work fully. This form of treatment is generally available from specialist hospital departments, and may require special funding.

Cosmetic camouflage is an effective alternative to cover the telangiectasia. Changing Faces offers tremendous help with this, and your local dermatology department can put you in touch with a trained volunteer.

Useful Products

PHYSICAL PROTECTION

- Wear rubber gloves and/or barrier cream when washing up
- Avoid powerful soaps and detergents;
- Avoid damaging or poking at the cuticles, which help protect the nail folds.

ITCHING

- Balneum Plus Cream[®]
- Xepin[®] (avoid using over large areas of skin)
- Antihistamines e.g. Atarax[®], Neo Clarityn[®] (non-drowsy)
- Ultraviolet light therapy (via hospital dermatology department)
- E45 Itch Relief Cream[®]
- Doublebase[®]
- Lanacane[®] (may cause allergic reactions)

BATHING AND WASHING

- Emulsifying Ointment (very cheap!)
- Balneum[®]
- Oilatum[®]
- Bath E45[®]
- Hydromol or Epaderm ointment[®]
- Aveeno[®]

MOISTURISING

- Diprobase[®]
- E45[®]
- Aqueous cream as a soap substitute
- Vaseline/liquid paraffin 50/50 (you can get this from a pharmacy or mix the ingredients yourself)
- Neutrogena hand cream[®]
- Unguentum M[®]
- Doublebase[®]
- Aveeno[®]
- Eucerin[®]
- Double Base Emollient Shower Gel[®]

Bride House
18-20 Bride Lane
London
EC4Y 8EE
T: 020 7000 1925
E: info@sruk.co.uk

112 Crewe Road
Alsager
Cheshire
ST7 2JA
T: 01270 872776
E: info@sruk.co.uk

Helpline: 0800 311 2756

www.sruk.co.uk

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