

MORPHOEA (LOCALISED SCLERODERMA)

What are the aims of this leaflet?

This leaflet has been written to help you understand more about morphoea (localised scleroderma). It tells you what it is, what causes it, what can be done about it, and where you can find out more about it.

What is morphoea?

Morphoea is a rare skin disorder where areas of skin become much thicker and firmer than normal. It is sometimes called 'localised scleroderma.' This term originates from two words 'sklero' meaning 'hard' and 'derma' meaning skin. It should not be confused with systemic sclerosis, which is a different disease that affects internal organs and the blood circulation as well as the skin. Morphoea usually affects a few small areas of the skin. Very rarely, it can occur over larger areas of the body in a form called 'generalised morphoea.'

What causes morphoea?

The cause of morphoea is unknown, but it is not contagious. It is not hereditary but seems to be more common in people with autoimmune conditions and these can run in families. Autoimmune disease occurs when the body's natural defence system cannot tell the difference between your own cells and foreign cells, causing your body to mistakenly attack normal cells. Common autoimmune conditions include thyroid disease, diabetes and lupus.

In morphoea the affected areas of skin become hard because of over-production of collagen fibres. These give the skin its normal strength and structure, so when excess collagen is made, the skin feels harder and more rigid than usual. Morphoea usually starts without any apparent trigger, but it has been noted to happen after a skin injury such as an insect bite, repeated friction, surgery and radiotherapy.

Morphoea may occur at any age. It is up to four times more common in women than men. It predominantly affects white people and is less common in skin of colour.

What does morphoea look like?

Morphoea usually appears as bruise-like pink patches of skin that thicken and turn pale and waxy in the middle, leaving a pink/pale purple border. The initial phase may appear as brown-dark patches in skin of colour. The skin feels firmer than normal when gently pinched. Sometimes the affected areas of skin turn slightly darker and beige coloured. Hair is usually lost from the affected areas and the sweat glands stop working so the skin feels dry. In rare cases, fatty tissue and muscle under the morphoea disappears so the skin becomes indented and stuck onto the underlying bone. This is called deep morphoea.

There are several types of morphoea, of which plaque morphoea is the most common in adults. In plaque morphoea, the affected areas usually range from 2 to 15 cm in diameter and are oval in shape (like a thumb print or palm print). In children, morphoea usually appears as a thickened strip of skin called 'linear morphoea'. This usually runs along part of the torso or a limb. It can also affect the forehead to the scalp and cause a grove in the skin with hair loss (sometimes referred to as 'en coup de sabre').

What are the symptoms?

Morphoea usually develops slowly and there may be few symptoms. The affected area(s) can be itchy, uncomfortable or numb. When morphoea affects the skin on a limb or over a joint, it can restrict growth and mobility and in severe cases can cause long-term disability.

Deep morphoea can rarely affect the face, leading to distortion of the natural facial contour and altered facial appearance.

How is morphoea diagnosed?

A dermatologist can usually diagnose morphoea by examining the skin because it has a distinctive appearance. A skin biopsy (a small piece of the skin, removed under a local anaesthetic) can be taken from the affected area if there is uncertainty or to rule out other skin complaints. Blood tests may also be helpful.

Is there a cure for morphoea?

There is no known cure for morphoea and it typically lasts for several years so is considered to be a chronic skin disorder.

How can morphoea be treated?

The treatment for morphoea depends on the type (linear, plaque or generalised), the person's age and whether it has spread deeper underneath the skin. Plaque morphoea does not always need treatment because it usually improves on its own after a few years. However, strong steroid creams or ointments or a non-steroid cream such as Tacrolimus are sometimes used as they may relieve any irritation and stop the patches enlarging. Intralesional injections of steroid (into the affected skin) can also help.

Early treatment is important for linear and generalised morphoea to prevent later problems especially loss of mobility. Options include light therapy, or phototherapy (NB-UVB/PUVA/UVA1), steroids and immunosuppressive therapy (Methotrexate, Mycophenolate Mofetil or Ciclosporin). NB-UVB phototherapy is used to treat superficial areas of morphoea whilst UVA1 phototherapy is better at tackling deep morphoea, owing to its deeper penetration of the skin. Access to UVA1 may be limited by region. Topical or oral PUVA (psoralen plus UVA) may also be helpful.

Treatment is likely to involve a multidisciplinary team of experts including specialists in skin surgery (plastic surgeons), bone surgery (orthopaedic surgeons), joint specialists (rheumatologists) and physiotherapists.

Self care (what can I do?)

Using a moisturiser on the affected areas can help relieve dryness of the skin and irritation. Gentle stretching exercises which do not cause pain (as advised by a physiotherapist if available) may be helpful to maintain mobility in affected areas.

Where can I find out more about morphoea?

Web links to other internet sites:

www.dermnetnz.org/immune/morphoea.html

Links to patient support groups:

Scleroderma & Raynaud's UK 18-20 Bride Lane.

London, EC4Y 8EE

Tel: 020 3893 5998 Email: info@sruk.co.uk

Web: https://www.sruk.co.uk/

For details of source materials used please contact the Clinical Standards Unit (clinicalstandards@bad.org.uk).

This leaflet aims to provide accurate information about the subject and is a consensus of the views held by representatives of the British Association of Dermatologists: individual patient circumstances may differ, which might alter both the advice and course of therapy given to you by your doctor.

This leaflet has been assessed for readability by the British Association of Dermatologists' Patient Information Lay Review Panel

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