

Localised scleroderma

What is localised scleroderma?

Localised scleroderma is an inflammatory disease affecting the skin. Inflammation is normally one of the body's protective responses to infection or injury. In diseases such as localised scleroderma, uncontrolled inflammation leads to hardening and discolouration of the involved skin. Localised scleroderma is frequently referred to as morphoea. Localised scleroderma should not be confused with systemic scleroderma (systemic sclerosis), which is a different disease.

What causes localised scleroderma?

The cause of localised scleroderma is unknown. It is an autoimmune disease. The immune system usually fights germs or viruses to keep us healthy. Sometimes the body's immune system mistakes a normal part of the body for something foreign, and starts attacking the body itself (autoimmune). Inflammation of the skin results in the body producing too much collagen (a normal protein in the body) leading to hardening of that area of skin. Sometimes localised scleroderma appears to occur after an injury but this is not always the case. Localised scleroderma cannot be passed on to others and there is no known genetic cause.

What are the symptoms of localised scleroderma?

There are different types of localised scleroderma based on the appearance of the skin and the extent of the area involved. The affected skin is often red or purplish and can become white in appearance. It can feel smooth, thick and waxy. The appearance of the skin affected by localised scleroderma can change over time, often appearing darker than the surrounding skin.

When localised scleroderma occurs in patches it is known as circumscribed morphoea. Linear scleroderma appears as a line and usually occurs on the arms, legs or face. If linear scleroderma occurs around a joint, the movement of the joint can be affected. When scleroderma occurs on the face, it is called 'en coup de sabre' or Parry-Romberg syndrome.

How is localised scleroderma diagnosed?

The diagnosis is usually made based on the appearance of the skin. Sometimes your child's doctor may arrange blood tests or a skin biopsy. A biopsy is when a small piece of skin is taken and looked at under a microscope. Your doctor may also order a MRI (a scan).

How is localised scleroderma treated?

The aim of treatment is to stop areas of affected skin from getting bigger and stop new lesions from forming. There are a number of medications your child's doctor may prescribe.

The most commonly used treatments are: creams or ointments, steroids or other immunosuppressive medicines. Phototherapy (a type of light based treatment) is sometimes also considered. Your child may benefit from physiotherapy which may be useful when the tight skin from scleroderma is affecting the joints or muscles underneath.

As well as medications, your child may need therapy with other health professionals such as the clinical nurse specialist, other specialist doctors or a physiotherapist.

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How can I help my child with localised scleroderma?

Follow up: The most important aspect of managing your child's localised scleroderma is for them to take their medicines and to attend clinic for checkups.

Make-up: Cosmetic make-up can effectively cover up affected areas to make them look less noticeable.

Exercise: Exercise improves muscle, joint and bone strength.

Diet: There are no specific recommendations regarding diet for children with localised scleroderma.

Vaccinations: It is important to check with your child's doctor or nurse before your child receives vaccinations. If your child is taking immunosuppressive medicines or steroids, live vaccines should be avoided.

Complementary and alternative medicines: At present, there is no evidence to support the use of complementary or alternative medicines in localised scleroderma. It is important that your doctor is aware of any other medicines you are taking.

Living with localised scleroderma

Localised scleroderma may last for many years. Most children recover and lesions improve over time.

What is the outlook for my child?

After recovery, the skin involved may still appear slightly different, such as paler or darker, or feel thicker than surrounding skin.

Where can I find more information?

Useful websites:

- www.printo.it
- www.rheumatology.org

Key points

- Localised scleroderma is an uncommon condition that affects the skin
- Localised scleroderma is treated with creams and medicines
- Localised scleroderma may last years but the outcome is good

CONTACT YOUR LOCAL ARTHRITIS OFFICE FOR MORE INFORMATION SHEETS ON ARTHRITIS.

This sheet was produced in association with the Australian Paediatric Rheumatology Group

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