

Localized Scleroderma (Juvenile)

Fast Facts

- In children, localized scleroderma is more common and less severe than the generalized form of scleroderma, which is also called systemic sclerosis and severely affects internal organs.
- Scleroderma can cause growth and joint problems in children.
- There is no known cure, but treatments can control the disease or reduce associated problems.



Scleroderma means “hard skin.” Children with localized scleroderma often have involvement of the tissues below the skin, including muscle and bone. Besides the skin hardening, there can be changes in skin color and texture, and the underlying tissues may fail to grow normally. Localized scleroderma can occur in several different forms, including linear scleroderma (where the lesion appears as a line or streak) and circumscribed morphea (where the lesion appears as a roundish lesion). Most patients have the disease on just one part or side of their body. Early on, some lesions may have a red or purplish color that may be limited to the lesion border. Others may have a white or waxy appearance and feel hard.

⊕ Who gets localized scleroderma?

Localized scleroderma can occur at any age and in any race, but is more common in Caucasians. Most patients who develop scleroderma are female. Environmental factors, such as trauma, infections, or drug or chemical exposure, may play a role, but not for most patients. The disease is not contagious. The disease is not passed on directly from parent to child by any one gene, though certain genes may make a child more likely to develop localized scleroderma.

Localized scleroderma is a rare disease, and the exact number of patients with this disease is not known. The best estimate is that 50 children out of every 100,000 will develop localized scleroderma.

⊕ What causes localized scleroderma and how is it diagnosed?

Localized scleroderma is an autoimmune disease in which the immune system causes inflammation in the skin. The inflammation can trigger connective tissue cells to produce too much collagen, a fibrous protein that is a major part of many tissues. Excess collagen can lead to fibrosis, which is like scarring.

Scleroderma usually is diagnosed by a rheumatologist or dermatologist based on the patient's history and physical examination. There are no specific laboratory studies to diagnose localized scleroderma, but tests often are done to evaluate the level of inflammation and problems related to localized scleroderma and to make sure the patient does not have another condition. A skin biopsy may be done to confirm the diagnosis.

+ How is localized scleroderma treated?

Treatment varies depending on the patient's disease activity, lesion location and extent, and whether there are related problems. Careful clinical evaluation is the primary method for monitoring scleroderma. X-rays and computerized tomography (CT) scans are used to look at bone abnormalities. Thermography can detect differences in skin temperature between the lesion and normal tissue. Ultrasound and magnetic resonance imaging (MRI) can aid soft tissue assessment.

Current treatment is focused on controlling inflammation, as this decreases the risk of serious problems such as differences in limb length, areas of sunken skin on the face, limited joint movement, and an internal organ problem. Patients with linear scleroderma, lesions on the head, deep lesions, or widespread disease, are usually treated with systemic medications that suppress the immune system. These medicines include methotrexate which is given by injection or taken by mouth once a week, and corticosteroids, which is taken by mouth (prednisone) or given by infusion (intravenous methylprednisolone).

One randomized clinical trial showed that methotrexate was better than placebo at maintaining disease control after initial corticosteroid treatment. Other immunosuppressive medicines include mycophenolate mofetil, cyclosporine, and tacrolimus. More work is needed to determine the best therapy for localized scleroderma. Immunosuppressive medicines can increase a patient's risk for developing an infection and have other possible side effects; these will be reviewed by your doctor.

For patients with mild superficial disease, topical medications often are used to control the inflammation and soften the skin. These medications include corticosteroids, calcipotriene, tacrolimus, pimecrolimus, and imiquimod. Moisturizers may help protect and soften the skin.

Phototherapy has been used to treat patients with widespread, superficial skin disease. Both UVB and UVA have been reported to help. More study is needed to evaluate the potential side effects from exposing children to large amounts of ultraviolet light.

Physical and occupational therapy to improve strength and function is important for patients with muscle weakness, limb length differences, and limited joint movement. Early referral to therapy should be considered in order to prevent loss of joint motion and function.

Surgery is not recommended during treatment of active disease. Surgery may be needed for patients with severe pain or limitation, and can improve the appearance of patients with severe facial lesions. However, because the skin is abnormal there can be poor wound healing, and surgery may trigger a flare of disease in some cases. Cosmetic makeup can be used to make the lesions less noticeable.

There is no known cure for localized scleroderma. The disease can stop on its own (remission), but the timing of this varies. Circumscribed morphea lesions that do not extend into deeper tissues may go into remission within a few years, while linear scleroderma lesions - especially on the head or scalp - can remain active for many years or even throughout childhood. It is important that patients who have lesions involving deeper tissues continue to be monitored at least yearly, even after treatment is stopped, because the disease can come back (relapse).

+ Broader health impacts: Associated problems

The overall prognosis is much better for localized scleroderma than for systemic sclerosis, because life-threatening internal organ involvement is extremely rare. However, localized disease can cause disfigurement, and the skin hardening can cause discomfort and sores as well as problems such as limited joint movement. In addition, many children can have problems of other organs or tissues. Those with linear scleroderma are at a higher risk for severe growth problems, such as developing a deformed, shorter, or smaller limb or portion of their face or scalp. Children with linear scleroderma lesions on their face or head can develop eye inflammation, eyelid or dental problems, headaches, seizures, and other brain problems. They need regular eye examinations and - in some cases - MRI evaluation of the brain and/or eyes. Other problems include arthritis, limited joint movement, muscle atrophy, and reflux of stomach contents. Children with pansclerotic morphea can develop chronic skin ulcers and may be at risk for squamous cell carcinoma.

+ Living with localized scleroderma

Most children with localized scleroderma don't need to make major lifestyle changes. All affected children should go to school. Some accommodations may be needed for children with severe disease affecting their ability to walk or write easily. Encourage affected children to remain active, but some activities - such as contact sports - should be limited in patients at risk for skin breakdown or in those with extremely limited movement in major joints.

When diagnosed, don't forget:

- Treatment for scleroderma should start as soon as possible. Treatment is more effective during the early inflammatory stage as the medicines do not directly target fibrosis.
- Children are at risk for growth problems and internal tissue involvement, so regular follow-up visits with the rheumatologist are essential to ensure that treatment is controlling inflammation and to minimize side effects from treatment.
- Children should see their pediatric rheumatologist at least once per year, because linear scleroderma can persist for many years, or come back after being inactive for several years.