PATIENT FACT SHEET

Localized Scleroderma (Pediatric)

CONDITION DESCRIPTION

Localized scleroderma ("hard skin") is an autoimmune condition that causes hardening of the skin and muscles in one part of the body. There are several types of scleroderma; the localized form is also called "morphea."

Localized scleroderma causes inflammation in the skin, which triggers 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.

Localized scleroderma is more common in Caucasians, and can happen at any age. Most patients are female. Certain genes may increase a child's risk of developing localized scleroderma. It affects about 50 out of every 100,000 children.

SIGNS/ SYMPTOMS

In localized scleroderma, excess collagen leads to fibrosis, or tissue scarring. Skin lesions include linear forms (lines or streaks) and circumscribed morphea (round). Most children have disease in one area or part of their body.

Localized scleroderma may affect muscle and bone, and cause growth and joint problems in affected areas. Early signs include lesions that are red or purplish around the border. Some children have white, waxy, hard lesions. Localized scleroderma could cause disfigurement. Skin hardening may cause discomfort, sores and limited joint movement. Linear lesions on the face could cause eye inflammation, eyelid or dental problems, headaches, seizures or brain problems.

A pediatric rheumatologist or dermatologist may diagnose the disease with a physical exam. Tests can evaluate inflammation and rule out other causes. A skin biopsy will confirm localized scleroderma. Imaging scans can measure bone and tissue abnormalities.

COMMON TREATMENTS

The first goal of treating localized scleroderma is to control inflammation with systemic medications. Inflammation is the cause of the skin lesions and joint problems. Injected or oral methotrexate (Rheumatrex, Trexall, Otrexup, Rasuvo), oral corticosteroids (prednisone) or infused corticosteroids (intravenous methylprednisolone) may suppress the immune system to control inflammation.

Other medications that may be used to treat localized scleroderma are mycophenolate mofetil (CellCept), cyclosporine (Neoral, Sandimmune) and tacrolimus (Prograf, Protopic, Hecoria).

Mild skin lesions may be treated with topical agents, such as corticosteroid creams, as well as phototherapy. Early on, children should have physical and occupational therapy to improve strength and function, and prevent loss of joint flexibility and function. Therapy is especially important for children with limb length differences, limited joint movement or muscle weakness. Surgery is only needed for children with severe lesions, pain or limitation.

CARE/ MANAGEMENT TIPS

Children with localized scleroderma should live as normally as possible. They should continue to go to school. Children with severe disease, who may have impaired ability to walk or write, may need accommodations or therapy to adapt movements. Children should stay active, although those at risk for skin breakdown or with severe joint problems may need to limit contact sports. Cosmetics help make skin lesions on the face or limbs less noticeable.

Patients need yearly check-ups with their pediatric rheumatologist to ensure that treatments are controlling their inflammation. Localized scleroderma can persist for years or recur after years of inactive disease. Regular eye and skin exams can spot serious complications early so treatment can be prescribed.

Updated March 2019 by Elizabeth Roth-Wojcicki, RN, MS, CPNP, and reviewed by the American College of Rheumatology Committee on Communications and Marketing. This information is provided for general education only. Individuals should consult a qualified health care provider for professional medical advice, diagnosis and treatment of a medical or health condition.

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