About Juvenile Localized Scleroderma

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"On Call" excerpt from *Scleroderma Voice*, summer 2014

What is juvenile localized scleroderma (jLS)?

Scleroderma means hard skin. In jLS, the hardening affects focal areas, called lesions that are usually only on one side of the body. Tissues beneath the skin are also often affected in jLS, including muscle and bone. Early on, there is inflammation (activity) in the lesions, which triggers excessive collagen to be made, leading to harder skin and other affected tissues.

What are the different types of jLS?

The most common form of jLS is linear scleroderma. Lesions appear as bands or streaks, often extending longitudinally along an arm or leg, or vertically on the forehead or scalp. Another common form is circumscribed or plaque morphea, which consists of oval lesions. In generalized morphea, several plaque lesions are present, often bilaterally, that merge to form large lesions. The most severe and rarest form is pansclerotic morphea, which causes widespread, circumferential, and deep tissue involvement. Many children have mixed morphea, meaning they have two or more different tpes of lesions, commonly linear and circumscribed morphea.

What are common problems associated with jLS?

The hardening of the skin can lead to discomfort or limited movement of the affected site, especially if it crosses a joint. Many children also have other tissues besides the skin affected (extracutaneous involvement). Extracutaneous involvement can cause a range of problems including arthritis, joint contractures, undergrowth growth of affected site, vision problems, dental or jaw issues, neuromuscular spasms, headaches, and seizures. Unlike systemic sclerosis, jLS is not associated with severe involvement of internal organs such s the heart, lungs, kidney, or gut. Lesions often become darker (hyperpigmentation) and depressed or sunken (atrophy),with color often improving over time, along with softening of the skin.

How is jLS diagnosed and how is it evaluated/monitored?

jLS is usually diagnosed by a rheumatologist or dermatologist based on the patient's history and an examination. There are no specific laboratory tests for diagnosis; skin biopsies are sometimes done if the diagnosis is not clear. Careful monitoring of the lesions is needed since signs of disease activity can be subtle, and treatment is directed at controlling activity. Other evaluations, including imaging (ultrasound, magnetic resonance imaging) and thermography may be done to help assess disease state and/or tissue involvement. Because active disease can persist for many years, and disease flares occur in up to 40 percent of patients off treatment, patients should have regular follow-ups.

How is jLS treated?

Patients with superficial lesions may not need treatment or may be treated with topical agents. Those with linear scleroderma, facial lesions, and/or moderate to severe disease need systemic treatment to control disease activity and minimize risk for developing serious problems such as joint contractures, disfigurement and growth defects. Methotrexate has been shown to be effective at controlling activity in most patients.

Phototherapy has been used to treat patients with widespread superficial disease. More study is needed to identify optimal regimens and to evaluate potential side effects from prolonged ultraviolet light exposure.

Physical and occupational therapy are important for improving function and strength, especially for those with linear or deep limb lesions. Surgery can improve the appearance and function of some patients, but has the potential risk of triggering a disease flare-up, so it is best to delay surgery until the disease is in remission.

What is the long-term prognosis for jLS?

Outcomes are generally good, and best for those who are diagnosed and begin an effective treatment plan early. Delays increase the risk for developing more damage or severe problems. Mortality is extremely rare in jLS, and jLS does not develop into systemic sclerosis.