

JUVENILE SCLERODERMA



**SCLERODERMA
FOUNDATION**

SUPPORT • EDUCATION • RESEARCH

INTRODUCTION

Every parent will experience a moment of panic when told their child has scleroderma. Often they hear little else the doctor has to say. A quick trip to the Internet may reinforce that sense of panic many times over. But there are actually several different diseases that are called “scleroderma” by some doctors. Certain forms of scleroderma are quite serious, but others are much less serious, and all can be treated by knowledgeable physicians.

The first thing a parent must understand is which form of scleroderma their child has. Doctors divide childhood scleroderma into localized and systemic forms, which have very different features. Localized forms of scleroderma are the most common in childhood, and are addressed in this pamphlet. They can be damaging to the skin, muscles, bones, and/or joints, but are unlikely to cause damage to the internal organs. Systemic forms of scleroderma often cause internal organ damage and may be a much greater problem. Many parents are relieved to learn that their child has a localized form of scleroderma, but become concerned that it might turn into systemic. This occurs rarely, if ever. Once your child has been properly evaluated, you should have a definite answer. If you do not know whether your child has a localized or systemic form of scleroderma, please consult your doctor.

LOCALIZED FORMS

Localized scleroderma is generally divided between morphea, linear scleroderma, and scleroderma en coup de sabre. Each type can be subdivided further and some children have more than one type.

Morphea is the most common form of localized scleroderma. It consists of irregular discolored and thickened patches of skin that can occur on the arms, legs, or body. Often the patches start out small and pinkish in color and aren't noticed until they reach the size of a penny or larger. Since many superficial skin infections or irritants may cause pink lesions, a variety of creams may be tried before the child is referred to a dermatologist. Over time, the center of the lesions may become pale, dry, and hard. This often

prompts a referral to a dermatologist and a diagnosis of scleroderma. The diagnosis may be made from the appearance of the lesions, or it may be necessary to do a skin biopsy to examine the tissue microscopically.

The lesions of morphea can vary significantly in size and shape. Some may be two to three inches or more in diameter and of varying shapes. Some children develop only a single lesion, but several lesions of varied sizes and shapes that develop over a period of several months to a year are not uncommon. Once an appropriate evaluation has been completed and the diagnosis has been made, treatment is started.

In most cases, the lesions are primarily a cosmetic problem. Morphea does not typically appear on the face and most lesions are easily hidden under clothing. As a result, morphea is more of an emotional issue. Even young children will wonder why they have the lesion and worry that it might be the result of something they did wrong. Parents should address this and make sure their child understands that this is false. Children will also need help with coping techniques when situations occur such as when others can see the lesions, or when lesions form in places that cannot always be hidden by clothing.

The long-term prognosis for children with isolated areas of morphea is excellent. Although the lesions may initially continue to enlarge and increase in number, they will soften and darken over a period of years. Pale skinned individuals are often left with large, irregular, light brown spots that follow the outline of the original lesions, but are soft and do not cause problems. Dark skinned individuals may be left with areas of pale skin. The period during which the child has active lesions of morphea is worrisome and unpleasant and may last several years, but most often there are no serious long-term consequences. Although parents would like the doctor to say that there will be no new lesions, or that the lesions will look better by a specific date, this is not possible. Morphea evolves over years, but rarely worsens once it clearly begins to improve.

Linear scleroderma refers to a condition in which areas of skin involvement seem to spread out along lines. Instead of an almost-round area of skin involvement, there may be a streak of involved skin. Most often this

appears on an arm or leg, but it can extend to, or appear on, the hand or foot. Usually, only one arm or one leg is involved. However, there are children with more areas of involvement that may extend onto the chest or abdomen. There are also children who have both areas of linear involvement and morphea.

Under the microscope, the lesions of linear scleroderma look just like those of morphea. Small areas of linear scleroderma rarely cause trouble and are treated in the same way as morphea. However, larger areas of linear scleroderma that extend over an arm or leg, and areas of linear scleroderma that cross a joint (e.g. across the elbow, wrist, shoulder, knee, ankle, finger), may cause permanent damage. As the skin becomes tight and the lesions harden they can limit the ability of the joint to move. This causes a contracture that may lead to significant problems with use of the arm or leg. When large areas of an arm or leg are involved in a young child, the arm or leg may not grow appropriately. There are many possible explanations for this, but the answer is not yet known. As a result, children with linear scleroderma involving large areas or crossing joints most often are treated more aggressively than children with morphea (see below).

Occasionally, there are children with linear scleroderma on an arm or leg who have elevated muscle enzyme levels. This is referred to as sclerodermatomyositis. These children will require different treatment because of the muscle inflammation, but in general, they do just as well as other children with linear scleroderma. The key is recognizing that children may experience linear scleroderma differently and treatment should be adapted as needed.

The prognosis for children with linear scleroderma who receive appropriate treatment is good. However, there may be long-term damage to involved fingers or toes and/or permanent changes in the size of an arm or leg resulting from linear scleroderma that either was not treated or did not respond to treatment. The skin lesions themselves typically soften and turn light brown over a period of years, as described for morphea.

Linear scleroderma en coup de sabre is the term generally applied when children have linear

scleroderma on the head and/or face. The original use of the term applied to children with a deep furrow along the scalp with tight, hard skin that often extended onto the forehead.

Children with linear scleroderma en coup de sabre are a very diverse group. Some children appear to have the classically described disease with lesions only on the scalp and forehead, while other children may have lesions only on the chin or lip. There is another group of children who are termed as having Parry Romberg syndrome. Children with this condition have similar skin lesions, but may have involvement of the whole side of the face and even involvement of the tongue. Obvious cases of en coup de sabre and Parry Romberg syndrome differ significantly, but many children present with cross-over manifestations, which can make it difficult to determine with certainty which form of scleroderma the child has.

The long-term outcome for children with linear scleroderma en coup de sabre is mixed. If the lesions are confined to the scalp and forehead, they often evolve similarly to linear scleroderma and the effect is primarily cosmetic. The same is true for isolated areas of involvement on the face. Parry Romberg syndrome, in which one side of the face is involved, can present more serious difficulties because the bones may not grow properly. It can be treated by an appropriate team of specialists. The key is to be sure your child has been properly evaluated. Once the evaluation is complete, you can discuss the probable long-term outcome and treatment options with your physician. In the interim, it is important for parents to remember that involvement of the face is going to be obvious to both the child and to others. These children need extra support in understanding that they did not cause their condition and in learning how to deal with others.

TREATMENT OF LOCALIZED FORMS

Appropriate treatment for children with localized scleroderma depends on the area of skin involved. Small areas on the trunk may not require any therapy while lesions on the face or over the joints

often are treated aggressively. For children with mild morphea, topical treatment with calcipotriene cream or ointment during the active phase is often sufficient. Linear scleroderma often responds well to methotrexate, and physicians may use methotrexate for children with widespread or otherwise disturbing morphea as well. Linear scleroderma that does not cross a joint line and is not of sufficient extent to suggest it will cause deformity, probably does not require treatment. However, when large areas of an extremity are involved, or the disease is crossing a joint line, there is a significant risk of permanent damage. In these cases, most specialists will recommend therapy with methotrexate, which is often associated with a slow but steady softening of the involved skin. Like all medicines, methotrexate is not without possible side effects and careful consideration should be given both to the expected risks and benefits.

Discuss the benefits and risks of the medication with your physician. Proper therapy for children with Parry Romberg syndrome is unknown, but the early use of methotrexate may be beneficial. There is no known therapy for advanced lesions. In severe cases of Parry Romberg a consultation with a craniofacial surgeon may be necessary.

Although there are no scientific studies, many families have found the application of commercially available cocoa butter preparations to the hardened skin useful. Many physicians do not recommend food additives or dietary supplements, as they can be, at best, harmless or potentially harmful.

DEALING WITH OTHERS

Dealing with others is one of the most important subjects that many physicians frequently overlook. Children with chronic disease who either have visible lesions such as morphea or linear scleroderma, or appear ill, (as well as their families) need to know what to do when “somebody says something.” While all of us focus on pills and medications for localized forms of scleroderma, the psychosocial impact may be the most severe aspect of the disease. What children and families say when asked about “those spots” on

his/her arm or leg is not as important as the effect that having to formulate an answer and respond to comments can be. Too many people fall into the trap of either trying to give a complete explanation or being defensive. Neither is appropriate. The key is to develop a practiced answer that minimizes the stress for the patient and their family.

COPING AT HOME

Coping at home is an area that families of children with scleroderma must also consider. It can be far more complicated and important than dealing with others. Children with any chronic illness will invariably wonder what they did wrong. Open discussion and reassurance among family members may be sufficient, but professional help may be needed. No one should be ashamed to seek the help they need to deal with the stress of chronic illness in themselves or a family member. If allowed to smolder, the stresses of chronic illness can lead to noncompliance by the patient and often worsen the disease outcome. Divorce, suicide, and other negative behaviors all occur with increased frequency, not only in children with chronic diseases, but in all their family members as well.

CONCLUSION

Juvenile scleroderma can be unsettling for the child and his/her family, but if treated properly by an experienced physician, it is a condition that can be managed. For information on finding a physician, contact the Scleroderma Foundation at 800-722-HOPE, or access the Web site, at www.scleroderma.org.

Please note that this brochure is provided for educational purposes only. It is not intended to substitute for informed medical advice.

The Scleroderma Foundation thanks Thomas J. A. Lehman, M.D., Hospital for Special Surgery, for his assistance in the preparation of this brochure.

BECOME A MEMBER OF THE SCLERODERMA FOUNDATION

When you become a member of the Scleroderma Foundation, you support the organization's mission of support, education and research. Your donation helps pay for programs in each of those three areas, including:



- Funding an average of \$1 million in original research grants awarded to investigators annually.
- Helping patients and their families cope with scleroderma through mutual support groups, physician referrals and the National Patient Education Conference.
- Promoting public education of the disease through publications, seminars, patient education events and publicity campaigns.

As a member of the Scleroderma Foundation, you will receive:

- Our quarterly magazine, the “Scleroderma VOICE.” The magazine includes updates on the latest scleroderma research and treatments, positive and uplifting stories from patients living with the disease; and tips about how to manage living with scleroderma.
- Information and educational offerings from your local chapter.
- Discounted registration fees to the annual National Patient Education Conference.

Please consider joining the Scleroderma Foundation today. A membership form is attached on the reverse side of this panel.

To become a member of the Scleroderma Foundation, fill out this form, tear at perforation and send with your check or credit card information to:

Scleroderma Foundation
Attn: Donations
300 Rosewood Drive, Suite 105
Danvers, MA 01923

I would like to become a member and help support the Scleroderma Foundation's efforts to improve the lives of those with scleroderma, and to assist in the search for a cause and cure. Enclosed please find my check (or credit card information) in the amount of \$_____.

Donations of \$25 or more can be acknowledged as members (\$35 or more for international members).

- I am not interested in member benefits.
- However, I would like to make a contribution in the amount of \$_____.

Name: _____

Address: _____

City: _____

State/Zip: _____

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Email: _____

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Credit Card No.: _____

(Circle One:    )

Exp. Date: _____ CVV Code: _____

Name on Card: _____

OUR THREE-FOLD MISSION IS SUPPORT, EDUCATION AND RESEARCH

Support: To help patients and their families cope with scleroderma through mutual support programs, peer counseling, physician referrals, and educational information.

Education: To promote public awareness and education through patient and health professional seminars, literature, and publicity campaigns.

Research: To stimulate and support research to improve treatment and ultimately find the cause of and cure for scleroderma and related diseases.



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