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ANSWER KEY

Clinical Manifestations of Scleroderma


Scleroderma is a connective tissue disease of unknown etiology. Its most characteristic feature is thickening of the skin due to increased collagen deposition. However, the disease may involve multiple other organ systems. Two broad categories of scleroderma have been defined: localized and systemic. Although all forms of scleroderma are rare, localized scleroderma occurs more frequently than systemic scleroderma and has a more favorable prognosis.

Several types of localized scleroderma exist. Morphea is characterized by the presence of one or more patches of hard, ivory-colored skin lesions. They begin with erythema and progress to nonpitting edema before becoming sclerotic. The margins of active lesions often have a violaceous hue. Underlying muscle fibrosis and atrophy may occur. Linear scleroderma presents with streaks of induration that progress to sclerosis and then may become atrophic. These occur most commonly on the extremities, face, and scalp and often involve underlying subcutaneous tissue, muscle, and bone. This may result in contractures, deformity, and arrested growth of involved bone. Eosinophilic fasciitis is a rare variant of localized scleroderma. It is associated with a pucked cobblestone appearance to the skin overlying the involved area. Increased numbers of eosinophils are found in both blood and involved tissue.

The natural history of localized scleroderma is variable. Some patients' disease becomes inactive after a few years. The lesions of many patients who have morphea regress spontaneously. Linear lesions are less likely to regress. Progression to systemic sclerosis has been reported but is rare.

Systemic sclerosis is a serious disease with reported 10-year survival rates of 35% to 90%. Fibrosis occurs in multiple organ systems. Virtually all patients have cutaneous involvement, with sclerodactyly being the most common and earliest skin finding. This is followed by involvement of the face and then the trunk and proximal extremities. Raynaud phenomenon is found in more than 90% of patients and may precede the onset of skin changes. Joint stiffness and mild pain are common, but actual arthritis is uncommon. Muscle disease is found in about one third of patients. Most patients have problems with gastrointestinal motility, but because many children are asymptomatic, radiologic studies need to be done to document abnormalities. This also is true for pulmonary problems. Although many children are asymptomatic, Garty et al found that 92% of children who have systemic sclerosis have abnormal pulmonary function tests. Pulmonary and cardiac complications are the most common causes of death in children who have systemic sclerosis. Renal disease is less common, but also is associated with significant mortality.

Scleroderma often can be diagnosed by recognizing the characteristic skin findings. A skin biopsy may be useful for confirmation. Antinuclear antibodies are found in most children who have systemic sclerosis and in many whose scleroderma is localized. Pulmonary function tests and radiographic studies of the gastrointestinal tract should be done to document systemic involvement.

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SUGGESTED READINGS

POINT OF CLARIFICATION

Teething—Is It Painful?

A reader writes, regarding the article on Dental Problems in the August issue of Pediatrics in Review (1994; 15:311–318), "...do teething babies really experience pain?"

Dr. Shusterman responds:

Clearly, the process of tooth emergence irritates the oral mucosa, since there is, no doubt, pressure exerted on the tissue and a resultant remodeling of the cellular architecture. Many reliable observers (parents) report an increase in salivation and transient negative effects on personality. The process, however, is unlikely to cause a febrile state, and poor mastication or temporary dietary change may account for loose stools.

I have concluded that there is intermittent discomfort as a result of this process, but it is not the explanation for any concurrent severe metabolic disturbance or elevated fever.

There is no specific treatment for teething symptoms, aside from the palliative of allowing the child to chew on hard or cold food. Topical anesthetic preparations are sold over-the-counter, but these may be ingested, and alcohol, either in these preparations or alone, is inappropriate. Specific proprietary remedies should not be encouraged.
Winicott D. Transitional objects and transitional phenomena. *Int J Psychoanal.* 1956;34:89

**DEPARTMENT OF CORRECTIONS**

**Erratum**

The following reference was omitted inadvertently from the abstract on Munchausen syndrome (*Pediatrics in Review* 1994;15:320):

**Munchausen Syndrome by Proxy.**

Richardson GF. *Am Fam Physician.* 1987;36:119–123
normal values (reference ranges) for each test.

Summary
The interaction between clinicians and microbiology laboratory staff has to be one of mutual benefit. The more the laboratory personnel know about your patients, the more meaningful and thorough will be the results. Communication is the key to success. Visit the microbiology laboratory and get to know the staff. The clinician also needs to be familiar with and use the most commonly used diagnostic tests for individual bacterial pathogens appropriately.

SUGGESTED READING

Pediatrics in Review to Hold 1996 Cover Art Contest: Works by Children!

In 1996, we plan to display a piece of art by children on the covers of our 1996 issues. Four pictures will be chosen, and the cover artwork will be changed quarterly.

Rules of the Contest
1. The contest will run from January through July 1995. (Winners will be chosen in August 1995 for display in 1996. PRIZES will be awarded to each winner!)
2. The theme of each submission: Draw a picture of you (ie, the child/adolescent artist) doing your favorite thing.
3. Qualification: The artist must be either between the ages of a) 5 and 10 years or b) 11 and 15 years. (There will be two categories, by age, for submission and judging.)
4. Requirements: The picture must be in color and be reproducible to a size of 3 inches by 4 inches. FREE HINT TO ARTISTS: Think Big! Small details don’t show up as well.

Pediatricians: Please have your patients send art they would like considered to:
Sydney Sutherland,
Editorial Assistant
Pediatrics in Review
c/o The Department of Pediatrics, Box 777
University of Rochester Medical Center
601 Elmwood Avenue
Rochester, NY 14642
(716) 275-0170
a closer look at the child and family's psychosocial functioning.

Maris D. Rosenberg, MD
Montefiore Medical Center
Albert Einstein College of Medicine
Bronx, NY

Comment: For pediatricians, the most important paragraph of this abstract is the last one. Prolonged thumb sucking as a potential expression of underlying frustration, emotional stress, anxiety, or other phenomena should be a red flag for the pediatrician. Children express their insecurities in a number of different ways; the degree of this insecurity potentially is reflected in this excessive need to suck the thumb. Clearly pediatricians must identify these insecurities and develop means for intervention. Simply removing the symptom by aversive therapies or other suggested interventions will not deal satisfactorily with what may be causing the need to suck non-nutritively. We merely may be substituting symptoms if we do not pay serious attention to those underlying anxieties. Time well spent uncovering the insecurities may lead to healthier outcomes down the road.

Steven P. Shelov, MD
Editor, In Brief

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PIR Quiz-CME Credit

The American Academy of Pediatrics is accredited by the Accreditation Council for Continuing Medical Education to sponsor continuing medical education for physicians.

The American Academy of Pediatrics designates this continuing medical education activity for 56 credit hours in Category 1 of the Physician’s Recognition Award of the American Medical Association.

This program has been reviewed and is acceptable for 56 Prescribed hours by the American Academy of Family Physicians. (Term of approval: beginning date January 1995. Enduring materials are approved for 1 year, with option to request renewal.) For specific information, please consult with the AAFP Office of Continuing Medical Education.

This program has been reviewed and is acceptable for 32 AOA Category 2-B CME hours by the American Osteopathic Association. For specific information, please consult with the AOA Department of Education.

In addition, this course has been approved for 56 NAPNAP contact hours. An individual requesting contact hours should submit proof of participation and verification of PREP accreditation to the NAP-NAP National Office.

The questions for the PIR quiz are located at the end of each article in this issue. Each question has a SINGLE BEST ANSWER. To obtain credit, record your answers on the PIR Quiz Card found in the January issue and return the card to the Academy. PREP group participants will receive the PIR Quiz Card and Self-Assessment Credit Reply Sheet under separate cover.

To receive CME credit on the 1995 annual credit summary, you must be enrolled in PREP or subscribe to Pediatrics in Review and return the PIR Quiz Card by February 28, 1996. PIR Quiz Cards received after this deadline will be recorded in the year they are received, with cards from the 1995 PIR journals accepted through December 31, 1997.

The PIR Quiz Card is bound into the January issue. Complete the quizzes in each issue and send to: American Academy of Pediatrics, PREP Office, PO Box 927, Elk Grove Village, IL 60009-0927.

PREP EDUCATION AWARD:
The PREP Education Award provides recognition and support for those Academy Fellows and Candidate Fellows who participate in PREP. Individuals who qualify for the PREP Education Award will receive their award automatically. To be eligible for this award, a Fellow or Candidate Fellow of the American Academy of Pediatrics must receive, over a 3-year period, 150 hours of Category 1 CME credits from the following sources:

- 75 hours must be obtained from participation in PREP (the Self-Assessment Exercise and/or Pediatrics in Review) or PREP: The Course.
- The balance (75 hours) of the 150 CME credits may be obtained through other programs sponsored or approved by the Academy. These include: the AAP Spring Session or Annual Meeting, CME courses, Academy-approved courses, the Pediatric UPDATE audiocassette tape program, or AQUIP.

The correct answers to the questions in this issue appear on the inside front cover.