

## Background

This leaflet describes, and shows examples of, the main forms of childhood scleroderma. Scleroderma in childhood is usually due to either localised scleroderma or systemic sclerosis.

## Case definition

All cases of abnormal skin thickening newly diagnosed in the past month (the skin will usually be difficult to pinch normally) suspected by the reporting paediatrician to be linear scleroderma or systemic sclerosis (age up to 16 years).

For confirmation of cases, we shall ask in the follow-up questionnaire if the diagnosis has been confirmed by a dermatologist or paediatric rheumatologist.

## Reporting instructions

Please report any new or suspected case you have seen in the past month, whatever the reason for referral to you and whether or not you are the main clinician responsible for the patient. Further information about the case(s) will be sought from you 1 year after diagnosis. All the information sought will be routinely available from the medical records.

## Localised scleroderma or systemic sclerosis?

Localised scleroderma affects skin and subcutaneous tissues. Deeper tissues including muscle and bone can also be affected, but internal organ involvement does not occur. Raynaud's phenomenon is unusual.

Systemic sclerosis is a multisystem disease with internal organ involvement, and can be life-threatening. Almost all affected children have Raynaud's phenomenon.

## Epidemiology

There are no good estimates of incidence and prevalence but we anticipate approximately 180 children per year in the UK to be diagnosed as having the linear form of localised scleroderma, and approximately 20 per year with systemic sclerosis.

## Research questions

To define the incidence of childhood linear scleroderma, and systemic sclerosis in the UK and Ireland. Also to describe:

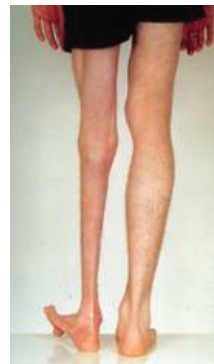
1. The delay between symptom onset and diagnosis.
2. The pattern of care received by affected children before and after diagnosis.
3. Which ages are most affected and the male: female ratio of affected children.
4. Regional differences in incidence rate.
5. Ethnic differences in incidence rate.

The study will provide data which should be of value in defining the need for supraregional referral services and in designing future clinical trials.

## Clinical features of localised scleroderma

Localised scleroderma can be subtyped into:

Linear scleroderma. **This is the form of localised scleroderma which we are asking you to notify.** Here the scleroderma occurs in linear bands on trunk or limbs. It can be very debilitating.



Linear scleroderma can cross joint lines and lead to contractures and limb length inequalities. Scalp and face can be affected ('coup de sabre' variant).



Morphoea. This may occur in patches or plaques. Rarely the plaques cover a large surface area ('generalised' morphoea) **We are not asking you to notify morphoea.**

## Clinical features of systemic sclerosis

This is very rare in children and is similar to the disease in adults. Internal organ involvement often occurs, and can be life threatening. Skin thickening usually begins distally in the fingers, may extend proximally. The ARA (American Rheumatism Association) criteria for diagnosis are:

(a) skin thickening proximal to the metacarpophalangeal joints.



or

(b) at least two of:

- Sclerodactyly (scleroderma of the fingers)
- Digital pitting (reflecting ischaemic atrophy of the finger pulp). Here there is loss of finger pulp.



- Bibasilar pulmonary fibrosis

There are two major subtypes of systemic sclerosis, defined on the basis of the extent of skin involvement:

**Limited cutaneous.** Skin involvement is confined to distal to elbows, knees and neck. Limited cutaneous systemic sclerosis was previously often termed 'CREST' (calcinosis, Raynaud's, esophageal involvement, sclerodactyly, telangiectases)



**Diffuse cutaneous.** Skin involvement extends to proximal limb and/or trunk.

*Overlap syndromes with other connective tissue disorders.*

Both limited cutaneous and diffuse cutaneous systemic sclerosis can occur in overlap with other connective tissue diseases, for example systemic lupus erythematosus, Sjogren's syndrome and inflammatory muscle disease. A significant proportion of children with systemic sclerosis have myositis.

### Differential diagnosis

Other causes of skin thickening in childhood include eosinophilic fasciitis, porphyria cutanea tarda, and lichen sclerosis. Children with early diffuse systemic sclerosis, who may have early swelling of their hands and feet, may initially be thought to have juvenile idiopathic arthritis, or another connective tissue disease.

### Management

There is no data from controlled clinical trials to inform management but the following treatments are used: Physiotherapy (especially for linear scleroderma). Steroids and methotrexate (under specialist supervision). Calcitriol (for linear scleroderma).

## CONTACT DETAILS

### DR EILEEN BAILDAM

Consultant Paediatric Rheumatologist  
Department of Paediatric Rheumatology  
Booth Hall Children's Hospital  
Charlestown, Blackley, Manchester, M9 7AA  
Tel: (0) 161 220 5597 Fax: (0) 161 220 5421  
E-mail: Eileen.baildam@CMMC.nhs.uk

### DR ARIANE HERRICK

Tel (0) 161 275 5993 Fax: (0) 161 275 5043  
E-mail: ariane.l.herrick@manchester.ac.uk

### PROFESSOR ALAN SILMAN

Tel: (0) 161 275 5041 Fax: (0) 161 275 5043  
E-mail: alan.silman@man.ac.uk  
ARC Epidemiology Unit  
Stopford Building, Oxford Road  
Manchester, M13 9PT

### DR MONICA BHUSHAN

Department of Dermatology  
North Manchester General Hospital,  
Delaunay's Road, Crumpsall, Manchester, M8 6RB  
Tel: (0) 161 720 2731 Fax: (0) 161 720 2139  
E-mail: M.Bhushan@mail.nmanhc-tr.mwest.nhs.uk

### British Paediatric Surveillance Unit

Royal College of Paediatrics and Child Health  
50 Hallam Street, London, W1W 6DE  
Tel: (0)20 7323 7911  
E-mail: bpsu@rcpch.ac.uk

### Funded by

### Raynaud's and Scleroderma Association

112 Crewe Road, Alsager, Cheshire ST7 2JA  
Tel: 01270 872776  
E-mail: info@raynauds.org.uk  
Web: www.raynauds.org.uk

## ARC Epidemiology Unit

MANCHESTER  
1824

The University  
of Manchester

## Survey of Childhood Scleroderma in the United Kingdom and Ireland

**The aim of this study** is to ascertain the incidence of childhood scleroderma in the UK and Ireland, focusing on linear scleroderma (a subtype of localised scleroderma) and systemic sclerosis. We shall ascertain age, sex and ethnicity of children affected, and describe current patterns of care.

**Start Date: July 2005**

### Investigators

**Dr Ariane Herrick** (chief investigator)  
Snr Lecturer/ Consultant Rheumatologist,  
University of Manchester/Hope Hospital, Salford  
**Dr Eileen Baidam** Consultant Paediatric  
Rheumatologist, Booth Hall Children's Hospital  
**Prof Alan Silman**, Director, ARC Epidemiology  
Unit, University of Manchester  
**Dr Monica Bhushan**, Consultant Dermatologist,  
North Manchester General Hospital, Manchester