

Study Number:

Please complete **Sections A, C and D** for children with suspected or known **systemic sclerosis**.
If you are a dermatologist or rheumatologist, please complete the PRES form '*Juvenile Systemic Sclerosis*' also.

Please complete **Sections B, C and D** for children with suspected or known **linear scleroderma**.
If you are a dermatologist or rheumatologist, please complete the PRES form '*Juvenile Localised Scleroderma*' also.

SECTION A –If you think the child has SYSTEMIC SCLEROSIS

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

- (a) skin thickening proximal to the metacarpophalangeal joints
- (b) at least two of: sclerodactyly (scleroderma of the fingers), digital pitting (reflecting ischaemic atrophy of the finger pulp), bibasilar pulmonary fibrosis.

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

- (a) limited cutaneous. Skin involvement is confined to distal to elbows, knees and neck
- (b) diffuse cutaneous. Skin involvement extends to proximal limb and/or trunk.

1. Where does the child have skin thickening? (please tick all which apply).

- | | | | |
|--|--------------------------|---------------|--------------------------|
| Fingers ONLY | <input type="checkbox"/> | Trunk | <input type="checkbox"/> |
| Hands or feet | <input type="checkbox"/> | Face and neck | <input type="checkbox"/> |
| Arms or legs
(distal to elbow/knee) | <input type="checkbox"/> | Don't know | <input type="checkbox"/> |
| Arms or legs
(Proximal to elbow/knee) | <input type="checkbox"/> | | |

- | | Yes | No | Don't know |
|---|--------------------------|--------------------------|--------------------------|
| 2. Does the child have proximal scleroderma?
(scleroderma proximal to MCP joints) | <input type="checkbox"/> | <input type="checkbox"/> | <input type="checkbox"/> |
| 3. Does the child have sclerodactyly?
(scleroderma of the fingers) | <input type="checkbox"/> | <input type="checkbox"/> | <input type="checkbox"/> |
| 4. Does the child have digital pitting (finger pulp atrophy)? | <input type="checkbox"/> | <input type="checkbox"/> | <input type="checkbox"/> |
| 5. Does the child have pulmonary fibrosis? | <input type="checkbox"/> | <input type="checkbox"/> | <input type="checkbox"/> |
| 6. Does the child have another connective tissue disease
(e.g. SLE, rheumatoid arthritis)? | <input type="checkbox"/> | <input type="checkbox"/> | <input type="checkbox"/> |

If yes please state features

SECTION B – If you think the child has LINEAR SCLERODERMA.

In linear scleroderma, 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").

7. Area affected (please tick box):

		Right		Left
Face/neck		<input type="checkbox"/>		<input type="checkbox"/>
Upper limb		<input type="checkbox"/>		<input type="checkbox"/>
Lower limb		<input type="checkbox"/>		<input type="checkbox"/>
Trunk	Front	<input type="checkbox"/>		<input type="checkbox"/>
Trunk	Back	<input type="checkbox"/>		<input type="checkbox"/>

SECTION C – MANAGEMENT – LINEAR SCLERODERMA and SYSTEMIC SCLEROSIS

8. Current/previous drug treatment (to include all steroid, immunosuppressant, antifibrotic therapy)

Name of drug	Currently used		Previously used	
	Yes	No	Yes	No
Oral steroids	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Parental steroids	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Methotrexate	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Cyclophosphamide ORAL	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Cyclophosphamide I.V.	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
D-Penicillamine	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Prostanoids	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Calcium channel blockers	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
CE inhibitors	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Vitamin D	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
PUVA	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Topical Steroids	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Cyclosporin A	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>

Other (please specify, including topical treatment)

9. Other previous treatments

Physiotherapy	Yes <input type="checkbox"/>	No <input type="checkbox"/>	Don't Know <input type="checkbox"/>
Occupational therapy	Yes <input type="checkbox"/>	No <input type="checkbox"/>	Don't know <input type="checkbox"/>

10. Has the diagnosis been confirmed by another specialist? If so please tick as appropriate

Dermatologist
Rheumatologist
Other specialist

<input type="checkbox"/>
<input type="checkbox"/>
<input type="checkbox"/>

We are grateful to you for completing this questionnaire. A further follow-up questionnaire will be sent to you in 12 months' time. Thank you for your help with this study.

Ariane Herrick & Eileen Baidam

Please send the completed form in the enclosed SAE to:

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Date: January 2005