

CHILDHOOD SCLERODERMA

- Abstract** At present there are no data on the occurrence of childhood scleroderma which might inform the level and distribution of expert provision required to manage affected children. A 13-month study of the incidence of childhood scleroderma will examine the UK experience. Childhood scleroderma encompasses a rare and poorly understood spectrum of conditions. This spectrum includes *systemic sclerosis*, which can be life-threatening due to internal organ involvement, and *linear scleroderma* (a form of localised scleroderma), which can be associated with major disability, growth defects, and disfigurement. The primary aim of this study is to ascertain the incidence of childhood systemic sclerosis and childhood linear scleroderma in the UK. In addition, a number of other questions will be addressed in relation to the occurrence of these disorders. By answering these questions, the study will provide data that should be of value in defining the need for supra-regional referral services and in designing future clinical trials.
- Principal Investigators** Dr Ariane Herrick, Senior Lecturer in Rheumatology , Arc Epidemiology Unit, University of Manchester. Tel: 0161 275 5993. E-mail: ariane.l.herrick@manchester.ac.uk and Dr Eileen Baildam, Consultant Paediatric Rheumatologist, Booth Hall Children's Hospital, Manchester. Tel: 0161 220 5597. E-mail: eileen.baildam@cmmc.nhs.uk.
- Co-investigators** Professor Alan Silman, Arc Epidemiology Unit and Dr Monica Bhushan, Consultant Dermatologist, North Manchester General Hospital.
- Coverage** United Kingdom and Republic of Ireland
- Duration** July 2005-July 2006 (13 months in the first instance)
- 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).
- Research Questions**
- 1) What is the incidence of scleroderma in childhood?
 - 2) What are the usual presenting symptoms?
 - 3) What is the delay between symptom onset and diagnosis?
 - 4) What is the pattern of care received by the affected children before and after diagnosis?
 - 5) Which ages are most affected?
 - 6) What is the male: female ratio of affected children, and does this vary with age?
 - 7) Are there any major regional or ethnic variations?
- Methods** For ascertainment of cases paediatricians will be asked on a monthly basis to report all cases meeting the case definition through the orange card system. Paediatricians who have reported a case that meets the case definition will be sent a questionnaire seeking demographic and clinical features. Paediatricians will be asked in a 12-month follow-up questionnaire if the diagnosis has been confirmed by a dermatologist or paediatric rheumatologist. Members of the UK Scleroderma Study Group, The British Society for Paediatric and Adolescent Rheumatology and the British Association of Dermatologists will also receive monthly email notification forms.

Reporting Instructions Please report any new or possible cases meeting the surveillance definition that you have seen in the last month on the BPSU orange card, even if you believe the case may have been reported from elsewhere and whatever the reason of referral to you. A detailed instruction leaflet including clinical photographs will be included with every questionnaire.

If you need any advice regarding the eligibility of a particular case for inclusion into the study, please contact: Dr Eileen Baidam (Tel: 0161 220 5597; E-mail: eileen.baidam@cmmc.nhs.uk) or Dr Ariane Herrick (Tel: 0161 275 5993; E-mail: ariane.l.herrick@manchester.ac.uk).

Funding The study is being funded by the Raynaud's and Scleroderma Association. It is being run by the Arthritis Research Campaign (arc) Epidemiology Unit, University of Manchester

Ethics Approval The South Manchester Research Ethics Committee has approved this study.

The Raynaud's and Scleroderma Association, 112 Crewe Road, Alsager, Cheshire ST7 2JA.

- 1) Uziel Y, Miller ML, Laxer RM. Scleroderma in children. *Paed Clin N America* 1995;**42**:1171-1203.
- 2) Silman AJ, Howard Y, Hicklin AJ, Black C. Geographical clustering of scleroderma in south and west London. *Br J Rheumatol* 1990;**29**:93-6.
- 3) Peterson LS, Nelson AM, Su WP, Mason T, O'Fallon WM, Gabriel SE. The epidemiology of morphea (localised scleroderma) in Olmstead County 1960-1993. *J Rheumatol* 1997;**24**:73-80.
- 4) Steen VD, Oddis CV, Conte CG, Janoski J, Caterline GZ, Medsger TA. Incidence of systemic sclerosis in Allegheny County, Pennsylvania. A twenty-year study of hospital – diagnosed cases, 1963-1982. *Arthritis Rheum* 1997;**40**:441-5.
- 5) Arnett FC, Cho M, Chatterjee S, Aguilar MB, Reveille JD, Mayes MD. Familial occurrence and relative risks for systemic sclerosis (scleroderma) in three United States cohorts. *Arthritis Rheum* 2001;**44**:1359-62.
- 6) Laxer RM. Scleroderma in children and adolescents. In Adolescent Rheumatology. Isenberg DA, Miller JJ,(Eds) Martin Dunitz, London 1999.
- 7) Uziel Y, Laxer RM, Krafchik BR, Yeung RS, Feldman BM. Children with morphea have normal self-perception. *J Paediatr* 2000;**137**:727-30.
- 8) LeRoy EC, Medsger TA. Criteria for the classification of early systemic sclerosis. *J Rheumatol* 2001;**28**:1573-6.
- 9) Korn JH.. Scleroderma and fasciitis in childhood. UpToDate Vol 11,2003.