A study of the incidence of childhood scleroderma in the UK and Ireland

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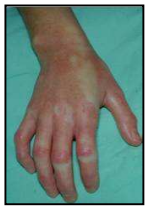
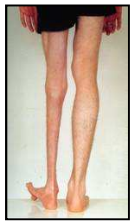
SCLERODERMA

• Localised scleroderma affects skin and subcutaneous tissue and, in children, can be extremely debilitating and cause growth problems

• Children can also suffer from systemic sclerosis which is associated with skin thickening and vascular involvement and can also affect the internal organs



Skin thickening in a case of SSc



Limb length inequality and contractures caused by localised scleroderma

BACKGROUND

•Childhood scleroderma represents a poorly understood and very rare spectrum of conditions

•Few studies have assessed the incidence of childhood scleroderma in the UK and Ireland

•Affected children are seen by different specialists

STUDY QUESTIONS

•What is the incidence of scleroderma in children?

•What is the age, sex, ethnicity and geographical distribution of affected children?

•What is the delay between symptom onset and diagnosis?

•What is the medical management of affected children?

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)

• We also asked clinicians to report morphoea especially when contacting dermatologists

METHOD

•Monthly ascertainment from the British Paediatric Surveillance Unit between July 2005 and July 2007 (25 months)

•Members of the UK Scleroderma Study Group (SSG), the British Society for Paediatric and Adolescent Rheumatology (BSPAR) and the British Association of Dermatologists (BAD) also contacted

•Clinicians reporting a case during the 25 month period were sent a questionnaire

•12 months after notification, clinicians with a valid case were sent a follow-up questionnaire

RESULTS

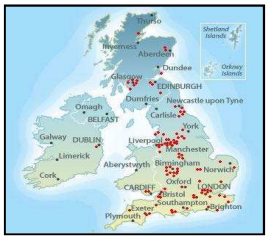
INCIDENCE

•3.4 per million children per year with localised scleroderma

•0.2 per million children per year with SSc

•Extremely rare conditions

GEOGRAPHICAL DISTRIBUTION



•Cases drawn from across the UK

•Only one case from the Republic of Ireland

DEMOGRAPHIC PROFILE

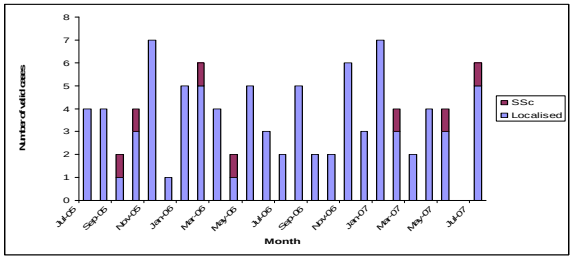
Features	Total cases n = 94	Localised n = 87	SSc n = 7
Mean age at onset (years)	8.6 ± 3.8	8.3 ± 3.9	11.3 ± 2.2
Female (%)	62 (66)	55 (63)	7 (100)
White British (%)	77 (82)	71 (82)	6 (86)

•Majority of cases are female

•Majority are White British

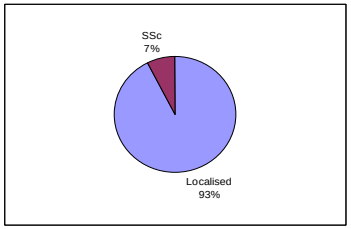
CASE ASCERTAINMENT

Valid cases reported over the 25 month surveillance period



CASES BY SUBTYPE

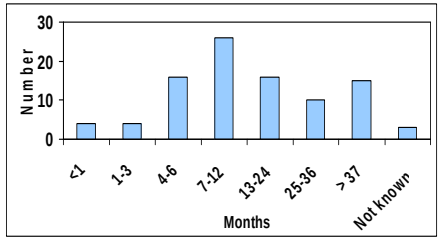
Proportion of cases by subtype



•Majority of cases (93%) were localised scleroderma

ONSET AND DIAGNOSIS DELAY

Delay between onset of symptoms and diagnosis by case



•Mean delay of 19 months for localised scleroderma and 9 months for SSc

12 MONTH FOLLOW-UP

•84 cases (89%) followed up at 12 months

•Majority of cases followed up showed signs of improvement

CONCLUSIONS

•98% of cases are actively treated following diagnosis and 65% reported to be improving 12 months after notification

•Current delays in diagnosis may adversely affect treatment outcome

•Rarity presents a challenge to research

•This study raised awareness of childhood scleroderma and highlighted the need for further research to inform management

Raynaud's & Scleroderma Association

Alder Hey Children's NHS Foundation Trust

www.medicine.manchester.ac.uk/arc