Clinical features of childhood scleroderma in an incidence cohort

Ariane Herrick[1], Holly Ennis[1], Alan J Silman[2], Eileen Baildam[3].

[1]arc Epidemiology Unit, University of Manchester, UK, [2]Arthritis Research Campaign (arc), Chesterfield, UK, [3]Alder Hey Children's NHS Foundation Trust.

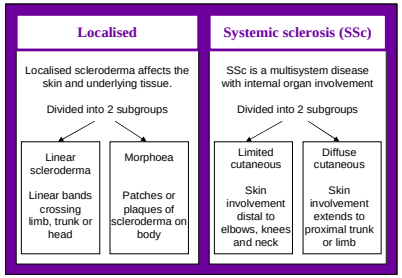
BACKGROUND

• Between 2005 and 2007 we conducted the first nationwide prospective incidence study of newly diagnosed cases of childhood scleroderma (both localised and systemic sclerosis [SSc]) in the UK and Ireland.

• Childhood scleroderma (defined as per Figure 1) can be either localised or occur in association with SSc.

• Previous studies of the clinical features of childhood scleroderma have concentrated on well-established disease.

Figure 1. Definition of childhood scleroderma.



AIMS

• To describe the clinical features and patterns of care in a population of children with newly diagnosed scleroderma notified within an incidence study including;

- age, gender and ethnicity
- delay between symptom onset and diagnosis
- outcome 12 months after diagnosis.

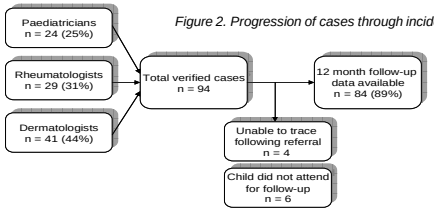
METHOD

• 94 newly diagnosed cases of childhood scleroderma (87 localised scleroderma, 7 SSc) identified in an incidence study (detailed in Figure 2).

• Demographic and clinical details provided for all cases by notifying clinicians using modified Paediatric Rheumatology European Society (PRES) forms.

•12 months after notification, clinicians completed a follow-up form about the child's progress since diagnosis.

Figure 2. Progression of cases through incidence study



RESULTS

DEMOGRAPHIC PROFILE

• Cases notified within the incidence study (n=94) were predominantly female (66%), white British (82%) and the mean age at onset of symptoms was 8.2 years. Details according to subtype are shown in Table 1.

Table 1. Demographic profile of incidence cohort of childhood scleroderma

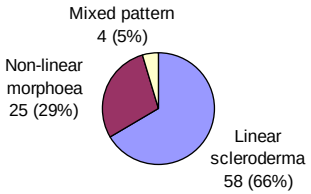
Features	Total cases n = 94	Localised n = 87	SSc n = 7
Mean age at onset (years)	8.6 ± 3.8	8.3 ± 3.9	11.3 ± 2.2
Female (%)	62 (66)	55 (63)	7 (100)
White British (%)	77 (82)	71 (82)	6 (86)

LOCALISED SCLERODERMA

• 87 cases of localised scleroderma notified (93%).

• Majority (66%) classed as linear scleroderma. Subtypes are shown in Figure 3.

Figure 3. Subtypes of localised scleroderma cases, n=87.



• Of the 58 cases with linear scleroderma, lesions were located on;
29 trunk and/or limb (50%)
26 face or head (45%)
3 both face or head and limb (5%)

•Extracutaneous features were reported in 14 (16%) of cases with linear scleroderma including arthritis (4), involvement of the central nervous system (4), teeth and jaw (3) and eye (2).

• ANA testing performed in 37 (43%) of localised cases at diagnosis and was positive in 16 (43%).

• Fewer than half (45%) with localised scleroderma received any treatment before diagnosis. Following diagnosis, 59% were started on methotrexate.

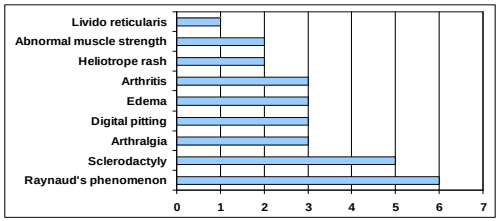
SYSTEMIC SCLEROSIS

• 7 cases of SSc notified (7%).

• 6 (86%) classed as limited cutaneous SSc; 2 of these had a dermatomyositis overlap.

• No single characteristic shared by all 7 cases.

Figure 4. Clinical features of 7 cases of SSc.



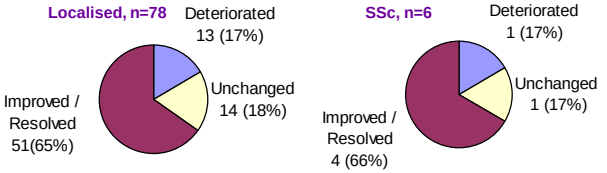
• 6 ANA positive, 2 anti-Scl-70 positive, 1 anti-RNP positive, 1 anticentromere positive.

• 6 (86%) had received treatment prior to diagnosis (5 with methotrexate.

12 MONTH FOLLOW-UP

• Follow-up information obtained for 84 cases (89%) and progress as rated by clinicians shown in Figure 5.

Figure 5. Subtypes of localised scleroderma cases, n=87.



CONCLUSIONS

• There was a high prevalence (50%) of face-head involvement in those with linear disease and a high number of children with extracutaneous disease (16%).

• In contrast to larger studies in well-established disease, most cases in this sample with SSc had limited cutaneous disease.

• The majority (89%) of cases were followed up 12 months after notification and most cases with localised disease had improved according to clinician's opinion.

Raynaud's & Scleroderma Association

www.medicine.manchester.ac.uk/arc