Clinical features of childhood scleroderma in an incidence cohort
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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.

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.

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.

LOCALISED SCLERODERMA
• 87 cases of localised scleroderma notified (93%).
• Majority (66%) classed as linear scleroderma. Subtypes are shown in Figure 3.

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.

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

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.

Table 1. Demographic profile of incidence cohort of childhood scleroderma

<table>
<thead>
<tr>
<th>Features</th>
<th>Total cases n = 94</th>
<th>Localised n = 87</th>
<th>SSc n = 7</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mean age at onset (years)</td>
<td>6.6 ± 3.8</td>
<td>6.3 ± 3.9</td>
<td>11.3 ± 2.2</td>
</tr>
<tr>
<td>Female (%)</td>
<td>62 (66)</td>
<td>55 (63)</td>
<td>7 (100)</td>
</tr>
<tr>
<td>White British (%)</td>
<td>77 (82)</td>
<td>71 (82)</td>
<td>6 (86)</td>
</tr>
</tbody>
</table>

Figure 1. Definition of childhood scleroderma.

Figure 2. Progression of cases through incidence study.

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

Figure 4. Clinical features of 7 cases of SSc.

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

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