Concise report

United Kingdom survey of current management of juvenile localized scleroderma

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Abstract

Objectives. Juvenile localized scleroderma (JLS) is a rare condition that is often difficult to assess and for which a variety of monitoring tools have been described. We aimed to describe how monitoring tools are used and perceived by clinicians in the UK, to ascertain treatments used for JLS and to provide a description of transition arrangements to adult care.

Methods. An e-survey of UK paediatric rheumatologists and dermatologists managing children and young people (CYP) with JLS was distributed using the national organisations representing these clinician groups. We asked respondents for their views and experience using 15 JLS monitoring tools, about transition services and about treatments used.

Results. Thirty-five dermatologists and 13 paediatric rheumatologists responded. Paediatric rheumatologists managed more CYP with JLS than dermatologists (median 16–20 and 3, respectively). Transition arrangements were reported by 43% of dermatologists and 91% of paediatric rheumatologists. Medical photography was the most frequently regularly used monitoring tool (73% respondents). The modified Rodnan skin score was the skin score used most commonly: 33% of paediatric rheumatologists and 3% of dermatologists reported using this tool frequently. Topical treatments and ultraviolet light were used by 49–80% of dermatologists and 0–8% paediatric rheumatologists. Biologic drugs and CYC were used by 0–3% of dermatologists and 31–46% of paediatric rheumatologists.

Conclusion. How monitoring tools are accessed, used and perceived by paediatric rheumatologists and dermatologists in the UK varies between and within clinician groups, as do treatment prescribing patterns and transition arrangements. These differences will impact on the feasibility of conducting multicentre clinical trials and on standardising clinical care.

Key words: paediatric, localized scleroderma, thermography, ultrasound, photography, access to care, skin, magnetic resonance imaging, skin score, laser Doppler.

Introduction

Localized scleroderma is a rare group of conditions (estimated incidence 3.4 per million [1]) often starting in childhood [2], when it is termed juvenile localized scleroderma (JLS). Diagnosis and classification are essentially clinical [3]. A spectrum of disease activity occurs, ranging from mild self-limiting skin lesions of plaque morphea, through locally significant linear scleroderma, to progressive widespread disease causing disfigurement, functional disability and internal organ involvement, especially of the brain and eye [4, 5]. Although evidence exists supporting the use of various therapeutic agents, optimal treatment strategies are unknown and opinion and clinical practice vary [6, 7].

Significant challenges face researchers and clinicians seeking to improve care for children and young people (CYP) with JLS. The lack of a universal consensus regarding JLS classification criteria for use in clinical practice or trials is a major problem. The Mayo Clinic classification [3]...
published in 1995 is probably the most widely used. Barriers to CYP accessing timely care is also a key area needing to be addressed if early treatment is to occur. We, along with others, have shown prolonged time periods between the onset of first symptom and diagnosis and treatment [2, 8–10]. In addition, these conditions are cared for in a variety of services, from dermatology to specialist paediatric scleroderma clinics in paediatric rheumatology centres, with the rarity of the more severe varieties arguing for care in specialist clinics.

JLS is often difficult to assess, both at disease onset and later when differentiating active skin lesions from changes associated with damage caused by previous inflammation (burnt-out disease) [11]. These challenges can be accentuated in the growing child, as the appearance of lesions may change related to changes in body habitus associated with age and pubertal status which are unrelated to the level of inflammation within a lesion. A study of the modified Rodnan skin score in healthy children showed that many had an abnormal score that was related to age and pubertal status [12]. It is difficult to differentiate multilayer localized scleroderma that will progress to scarring and disabling disease from very superficial lesions that will only cause minor cosmetic changes in the long term. Skin scores [13, 14] have been used in an attempt to objectively quantify disease activity and damage. Recent advances in technology have allowed the development of more sensitive tools to aid in the assessment and monitoring of skin lesions, including infrared thermography [15], laser Doppler flowmetry [16], Doppler US [17] and MRI [18]. Often clinically obvious lesions do not appear on investigations, but there are no comprehensive cohort studies or long-term outcome studies evaluating monitoring tools with clinical impressions.

The primary aim of this survey of clinicians in the UK managing CYP with JLS is to elucidate which monitoring tools are currently used and how these are perceived by clinicians and accessed by patients. Secondary aims of the survey are to better understand which treatments are used and document transition arrangements for CYP with JLS moving from paediatric to adult services.

Methods

We surveyed clinicians in the UK who manage CYP with JLS. Evidence suggests CYP with JLS in the UK are managed either by paediatric rheumatologists or dermatologists [1]. The majority of practising paediatric rheumatologist in the UK are members of the British Society for Paediatric and Adolescent Rheumatology (BSPAR) and many dermatologists overseeing the care of significant paediatric populations are members of the British Society for Paediatric Dermatology (BSPD). By disseminating the e-survey to members of these two groups facilitated e-survey dissemination to group members.

Using SurveyMonkey software, we produced an e-survey and e-mailed this to the members of BSPPAR and BSPD concurrently. Responses were collected over a 2-month period. The survey captured data regarding respondents’ practising centre, whether they reported their personal activity or the activity of their department, how many CYP were under their management, how many new patients are seen annually, the percentage of patients under sole clinician management (either dermatologist or paediatric rheumatologist), the percentage of patients under joint clinician care, and if seen jointly, who provides joint care and if patients are examined by the same clinician at each visit.

Respondents were asked a series of questions relating to the list of monitoring tools displayed in Table 1. Respondents were asked if formal transitional care arrangements were provided, who was involved and which monitoring tools were available once CYP transferred to local adult health care services. Respondents were also asked which treatments they used for CYP with JLS. Ethics approval was not deemed necessary since the work was a service evaluation and all data collected relating to patients and centres were anonymized.

Results

Thirty-five BSPD and 13 BSPAR members responded from centres in England, Wales, Scotland and Ireland. BSPAR comprises a total membership of 338, 111 of whom are consultants (response rate 12%). BSPD has a non-trainee membership of 184 (response rate 19%). Twenty-eight (80%) BSPD members reported their own activity and 7 (20%) reported on behalf of their department; 4 (31%) BSPAR members reported their own activity and 9 (69%) reported on behalf of their department (1 reported on behalf of the Scottish network covering numerous hospitals, thus covering 9 of the 13 UK tertiary paediatric rheumatology centres. The median total patients managed per BSPAR respondent was 3 (modal average 2) and per BSPD respondent was 16–20 (modal average: three respondents saw none, three respondents saw 26–30). BSPD members reported seeing between zero and six new patients annually, while 22 (63%) respondents saw one or two new patients annually.

Responses from BSPAR members were more varied: 8 (62%) respondents saw between 1–5 new patients annually, 2 saw no new patients annually, 2 saw 10 new patients annually and 1 respondent saw >20 new patients annually.

Nine (26%) BSPD respondents managed all patients under their sole care, 9 (26%) managed all patients in collaboration and the remainder managed varying proportions of their patients solely and in collaboration. Collaboration was as follows: 21 (91%) with a paediatric rheumatologist, 5 (22%) with a paediatrician, 1 (4%) with an adult rheumatologist and 2 (9%) with another dermatologist (shared care between local and tertiary services). Two (15%) BSPAR respondents managed all patients
### Table 1 Monitoring tools—availability and views by clinicians (paediatric rheumatologists and dermatologists)

<table>
<thead>
<tr>
<th>Question asked</th>
<th>Thermography</th>
<th>US</th>
<th>Laser Doppler imaging</th>
<th>Laser Doppler flowmetry</th>
<th>1.5T MRI</th>
<th>3.0T MRI</th>
<th>Professional medical photography</th>
<th>Other photography</th>
<th>Body mapping (pre-printed)</th>
<th>Drawing in notes</th>
<th>LoSSI score</th>
<th>LoSDI score</th>
<th>LoSCAT score</th>
<th>Modified Rodnan score</th>
<th>Durometer</th>
<th>Answered question</th>
<th>Skipped question</th>
</tr>
</thead>
<tbody>
<tr>
<td>Which tools are you familiar with?</td>
<td>21 (75)</td>
<td>20 (71)</td>
<td>17 (61)</td>
<td>6 (21)</td>
<td>18 (64)</td>
<td>2 (7)</td>
<td>23 (82)</td>
<td>12 (43)</td>
<td>21 (75)</td>
<td>18 (64)</td>
<td>3 (11)</td>
<td>1 (4)</td>
<td>1 (4)</td>
<td>3 (11)</td>
<td>3 (11)</td>
<td>28</td>
<td>7</td>
</tr>
<tr>
<td>Which tools do you have in-house access to?</td>
<td>3 (9)</td>
<td>20 (63)</td>
<td>5 (16)</td>
<td>1 (3)</td>
<td>9 (28)</td>
<td>1 (3)</td>
<td>27 (84)</td>
<td>5 (16)</td>
<td>9 (28)</td>
<td>21 (66)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>11</td>
<td>2</td>
</tr>
<tr>
<td>Which tools do you regularly use?</td>
<td>2 (6)</td>
<td>7 (22)</td>
<td>1 (3)</td>
<td>0 (0)</td>
<td>5 (16)</td>
<td>0 (0)</td>
<td>26 (81)</td>
<td>4 (13)</td>
<td>5 (16)</td>
<td>18 (56)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>32</td>
<td>32</td>
</tr>
<tr>
<td>Which tools do you occasionally use?</td>
<td>8 (33)</td>
<td>8 (33)</td>
<td>5 (21)</td>
<td>0 (0)</td>
<td>2 (22)</td>
<td>2 (8)</td>
<td>26 (81)</td>
<td>1 (11)</td>
<td>5 (56)</td>
<td>18 (56)</td>
<td>8 (33)</td>
<td>8 (33)</td>
<td>1 (11)</td>
<td>1 (3)</td>
<td>1 (3)</td>
<td>11</td>
<td>9</td>
</tr>
<tr>
<td>Which tools do you perceive as clinically useful?</td>
<td>19 (95)</td>
<td>9 (45)</td>
<td>11 (55)</td>
<td>3 (15)</td>
<td>8 (40)</td>
<td>13 (65)</td>
<td>13 (65)</td>
<td>3 (15)</td>
<td>7 (35)</td>
<td>6 (30)</td>
<td>6 (30)</td>
<td>6 (30)</td>
<td>6 (30)</td>
<td>5 (25)</td>
<td>5 (25)</td>
<td>20</td>
<td>10</td>
</tr>
</tbody>
</table>

Values are n (%) or n. BSPD: British Society for Paediatric Dermatology; BSPAR: British Society for Paediatric and Adolescent Rheumatology; T: Tesla; LoSSI: localized scleroderma severity damage index skin score; LoSDI: localized scleroderma skin damage index skin score; LoSCAT: localized scleroderma cutaneous assessment tool skin score.
under their sole care, 4 (31%) managed all patients in collaboration and the remainder managed a varying proportion of their patients solely and in collaboration. Collaboration was as follows: 7 (78%) with a dermatologist and 8 (89%) with a paediatrician.

Table 1 shows how monitoring tools were perceived and accessed. Ten (91%) BSPAR respondents reported formal transition arrangements involving a paediatric rheumatologist (70%), general paediatrician (20%), dermatologist with paediatric interest (10%) or adult rheumatologist (90%). Twelve (43%) BSPD respondents reported formal transition arrangements involving a paediatric rheumatologist (22%), general paediatrician (6%), dermatologist with paediatric interest (72%), dermatologist (44%) or adult rheumatologist (17%).

Five (46%) BSPAR respondents reported the same monitoring tools being available following transfer of care from a paediatric to adult setting, 2 (18%) reported a greater number of monitoring tools being available following transfer and 4 (36%) were not sure. Seventeen (65%) BSPD respondents reported the same monitoring tools being available, 2 (8%) reported fewer being available and 7 (27%) were not sure. The topic of local transition services attracted the greatest number of free-text comments in this survey. Paediatric rheumatologists comments (total n = 8) varied: five commented that transition services locally were of variable, poor or unknown quality; three commented that transition services locally worked well. Two responses reported varying arrangements depending on where patients lived in relation to services. Discrepancies between multidisciplinary team provision in paediatric and adult services and the need for improved cooperation between paediatric and adult services was commented on. Comments (total n = 12) received from dermatology respondents also varied: six perceived no need for formal transition services (probably reflecting clinician ability to continue providing care into adulthood), one had failed to establish local transition services due to lack of funding and three described varying local transition models in vague terms. Table 2 shows the treatments used by responding clinicians. In addition to the treatments listed, two paediatric rheumatology respondents commented on having used rituximab to treat JLS.

**Discussion**

This is the first UK survey, to our knowledge, of clinical practice in JLS management led by paediatric rheumatology or dermatology. The survey was designed to take no more than 5 min to complete and was disseminated as an e-survey to increase the response rate. The information collected was therefore limited to areas of key interest; information relating to disease severity and associated features, which would have been of interest, were deemed beyond the scope of this survey. We were confident of reaching the vast majority of paediatric rheumatologists and dermatologists providing care to CYP with JLS through the two clinician networks through which the survey was distributed. The response rates (12% BSPAR
clinicians, 19% BSPD clinicians) were reasonable considering the survey was designed to enable a single clinician to respond on behalf of a centre or network where multiple clinician members of either group may have been working.

**Assessment and monitoring**

There is, to our knowledge, no consensus as to the optimal methods to be used in the assessment and monitoring of JLS in CYP. This survey reported that the monitoring tools used most regularly were those deemed to be least complicated and time consuming and those requiring the least technical (and costly) equipment and expertise. The discrepancy between tools regularly used and those perceived as clinically useful is noteworthy: for nearly all monitoring tools, a greater number of clinicians reported perceiving them as useful than reported using them regularly. The lower reported rates of in-house access to monitoring tools compared with the rates of reported perceived usefulness suggest clinicians widely lack access to monitoring tools they perceive as useful. Clinicians may also lack knowledge and training in the use of various monitoring tools. It may be that dermatologists do not have access to certain monitoring tools such as US as readily as paediatric rheumatologists. The wide range of monitoring tools used by clinicians is also noteworthy. While the tools we surveyed have been shown to be useful [11, 13–18], further work is needed comparing these tools in CYP across the severity range of the JLS spectrum.

**Table 2** Respondents’ reported use of treatments in juvenile localised scleroderma in children and young people

<table>
<thead>
<tr>
<th>Treatments used</th>
<th>BSPD (28 responses to question)</th>
<th>BSPAR (10 responses to question)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Topical steroids</td>
<td>28 (100)</td>
<td>1 (10)</td>
</tr>
<tr>
<td>Topical tacrolimus</td>
<td>19 (68)</td>
<td>1 (10)</td>
</tr>
<tr>
<td>Topical calcipotriol</td>
<td>17 (61)</td>
<td>1 (10)</td>
</tr>
<tr>
<td>Systemic steroids</td>
<td>22 (79)</td>
<td>9 (90)</td>
</tr>
<tr>
<td>Ultraviolet light</td>
<td>19 (68)</td>
<td>0 (0)</td>
</tr>
<tr>
<td>MTX</td>
<td>27 (96)</td>
<td>10 (100)</td>
</tr>
<tr>
<td>Ciclosporin</td>
<td>9 (32)</td>
<td>3 (30)</td>
</tr>
<tr>
<td>HCQ</td>
<td>3 (11)</td>
<td>4 (40)</td>
</tr>
<tr>
<td>Etanercept</td>
<td>1 (4)</td>
<td>6 (60)</td>
</tr>
<tr>
<td>Infliximab</td>
<td>0 (0)</td>
<td>4 (40)</td>
</tr>
<tr>
<td>Adalimumab</td>
<td>0 (0)</td>
<td>4 (40)</td>
</tr>
<tr>
<td>MMF</td>
<td>5 (18)</td>
<td>9 (90)</td>
</tr>
<tr>
<td>CYC</td>
<td>0 (0)</td>
<td>3 (30)</td>
</tr>
</tbody>
</table>

Values are n (%). BSPAR: British Society for Paediatric and Adolescent Rheumatology; BSPD: British Society for Paediatric Dermatology.
Responding dermatologists managed fewer CYP compared with paediatric rheumatologists. This may explain the greater reported use of monitoring tools among paediatric rheumatologists (particularly those involving access to specialist equipment). Furthermore, anecdotal evidence suggests paediatric rheumatologists in the UK manage CYP at the more severe end of the disease spectrum than dermatologists. More paediatric rheumatologists than dermatologists manage CYP in clinics jointly with other consultant specialists. The reasons for this are likely to be multifactorial and influenced by the differing service models between the two specialties. Closer collaboration between paediatric rheumatologists and dermatologists will be required to achieve equitable access to monitoring tools and to ensure future high-quality research determining how these tools are used involves CYP managed by both specialist groups.

Transitional care

Differences in transition arrangements between paediatric rheumatologists and dermatologists probably reflect differing service models between the two specialties: within rheumatology, CYP are managed by dedicated paediatric rheumatologists who then transfer patients to adult rheumatologists (usually around the age of 16–18 years). Adult dermatologists (often with a specific interest in paediatrics) manage CYP and often continue managing CYP into adulthood.

Medical management

Table 2 shows that dermatologists use more topical treatments while paediatric rheumatologists use systemic immunosuppressive therapies (biologic therapies and CYC); both clinician groups report using MTX. This could reflect differences in patient case mix—more severe disease managed by paediatric rheumatologists and less severe disease managed by dermatologists—although our survey did not collect data on disease phenotype or severity in individual patients. However, other studies have shown that dermatologists are less likely to use systemic immunosuppression even in patients with linear and generalized lesions compared with paediatric rheumatologists [7]. More detailed work is required to explain the differences in treatment practice reported in this survey. This survey suggests future research into optimal treatment strategies will need to involve patients managed by both specialty groups to avoid bias and to ensure treatment recommendations are applicable across the spectrum of the condition.

Future evolution of service models must place significant emphasis on equitable multidisciplinary provision of care. The coordinating role of the clinical nurse specialist has been highlighted within UK paediatric rheumatology service provision [19], as has the importance of physiotherapist and occupational therapist care. Clinical psychologist input is also important in managing potentially disfiguring skin lesions in CYP with JLS [20].

**Rheumatology key messages**

- Clinicians use a wide range of monitoring tools to assess and monitor children and young people with juvenile localized scleroderma.
- Paediatric rheumatologists and dermatologists use different treatment strategies to manage children and young people with juvenile localized scleroderma.
- Consensus to facilitate research and standardized care requires collaboration between dermatologists and paediatric rheumatologists.

**Acknowledgements**

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12 Foeldvari I, Wierk A. Healthy children have a significantly increased skin score assessed with the modified Rodnan skin score. Rheumatology 2006;45:76–8.


