Childrens' and parents' beliefs about childhood onset scleroderma are influenced by child age and physical function impairment

Sr., Childhood scleroderma is a rare and potentially debilitating condition occurring as part of the multisystem disease SSc or (more commonly) localized and confined to the skin and subcutaneous tissues [1, 2]. Assessments of quality of life have thus far focused on localized scleroderma and its impact on self-perception [3] and the physical appearance of skin lesions [4]. Empirical literature suggests, however, that patients construct their own common sense cognitive model of their medical condition [5]. These patient-held beliefs are of fundamental importance in adjustment and influence psychological outcomes such as distress, coping and functional disability [6]. Previous studies in adult scleroderma have illustrated that illness beliefs are an important factor in patients' emotional responses [7], but to date, no attempt has been made to assess beliefs about the illness experience of childhood scleroderma or correlate these beliefs with demographic and clinical factors.

Within a single cross-sectional study of physical function and quality of life in childhood scleroderma [8] we sought to describe childrens' and parents' beliefs using the Revised Illness Perceptions Questionnaire (IPQ-R) [9], a validated measure to assess illness representations that has been widely used in rheumatology, including a study of adult scleroderma [7]. As per the IPQ-R instructions, participants (children over 11 years or if under, parents or guardians) completed the measure and were asked to reflect upon their experiences during the previous 2 weeks. The IPQ-R consists of a set of multiple choice questions and is designed to assess the cognitive representations of illness around the following dimensions: (i) illness identity; (ii) chronicity; (iii) consequences of the condition; (iv) personal and treatment control; (v) illness coherence; (vi) emotional response; and (vii) causes of the condition. Full demographic and clinical data, including Child Health Assessment Questionnaire (CHAQ) scores [10], were available for the cohort and are shown in Table 1. Data were not normally distributed and Spearman’s correlation coefficient and Wilcoxon rank sum tests were used to examine the relationships between variables. The study was approved by the UK North West Research Ethics Committee.

Seventeen children and 11 parents (data from 28 children, 68% female, median age 13 years, 86% localized scleroderma and 14% SSc) participated in the study. Twenty-two (79%) of the children were receiving MTX or (more commonly) parenteral steroids. Of the 24 cases of localized scleroderma, 9 had face or head lesions, 14 trunk or limb lesions and 1 lesion to the face, trunk and limbs. The most common symptoms reported in the 2-weeks prior to the IPQ-R assessment were tiredness (50%), stiff joints (43%), feeling unwell (39%) and weight loss or gain (39%). More than 23 (82%) participants believed

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**Table 1** Demographic characteristics of the sample

<table>
<thead>
<tr>
<th></th>
<th>Total sample (n=28)</th>
<th>Localized (n=24)</th>
<th>SSc (n=4)</th>
<th>Arat et al. [11] (n=217)</th>
<th>Richards et al. [7] (n=49)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Gender, female, n (%)</td>
<td>19 (68)</td>
<td>15 (63)</td>
<td>4 (100)</td>
<td>169 (78)</td>
<td>42 (86)</td>
</tr>
<tr>
<td>Ethnicity, white British, %</td>
<td>24 (86)</td>
<td>20 (87)</td>
<td>4 (100)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Age at assessment, median (range), years</td>
<td>13 (5-17)</td>
<td>13 (5-17)</td>
<td>11 (7-14)</td>
<td>54 (46-64)</td>
<td>53 (12)^a</td>
</tr>
<tr>
<td>Disease duration since diagnosis, median (range), months</td>
<td>30 (2-135)</td>
<td>22 (2-135)</td>
<td>68 (15-83)</td>
<td>5 (2-10)</td>
<td>9 (6)^a</td>
</tr>
<tr>
<td>CHAQ score, median (range), 0-3</td>
<td>0.1 (0-1.6)</td>
<td>0 (0-1.6)</td>
<td>0.6 (0-1-2)</td>
<td>0.50 (0.12-1.25)</td>
<td>1.12 (0.72)^a</td>
</tr>
<tr>
<td>IPQ-R chronicity</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Timeline</td>
<td>21 (13-30)</td>
<td>20 (13-30)</td>
<td>22 (20-29)</td>
<td>21</td>
<td>25^a</td>
</tr>
<tr>
<td>Timeline cyclical</td>
<td>11 (5-18)</td>
<td>11 (5-18)</td>
<td>13 (11-16)</td>
<td>15</td>
<td>14^a</td>
</tr>
<tr>
<td>IPQ-R consequences</td>
<td>18 (12-26)</td>
<td>17 (12-26)</td>
<td>23 (19-23)</td>
<td>21</td>
<td>23^a</td>
</tr>
<tr>
<td>IPQ-R control</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Personal</td>
<td>16 (9-23)</td>
<td>16 (9-23)</td>
<td>17 (11-23)</td>
<td>17</td>
<td>17^a</td>
</tr>
<tr>
<td>Treatment</td>
<td>18 (13-21)</td>
<td>18 (15-21)</td>
<td>15 (13-18)</td>
<td>16</td>
<td>15^a</td>
</tr>
<tr>
<td>IPQ-R illness coherence</td>
<td>16 (5-24)</td>
<td>17 (5-24)</td>
<td>16 (10-19)</td>
<td>15</td>
<td>16^a</td>
</tr>
<tr>
<td>IPQ-R emotional response</td>
<td>18 (6-28)</td>
<td>18 (6-28)</td>
<td>21 (15-28)</td>
<td>19</td>
<td>18^a</td>
</tr>
</tbody>
</table>

^aMean (s.o.).
that treatment would control their scleroderma, although <6 (22%) believed that treatment would effectively cure scleroderma. Twenty-two participants (79%) either agreed or strongly agreed that it was difficult to predict what scleroderma would do on a day-to-day basis. Fourteen (50%), half of all participants, believed that scleroderma had serious consequences on everyday life and 8 (29%) believed that it also caused difficulties for people close to them. Thirteen (46%) reported that scleroderma contributed to depressed mood. The most commonly reported perceived causes of scleroderma (participants could select more than one) were the immune system (54%), chance or bad luck (46%) and accidents (18%). Other causes (including personality, alcohol, smoking, family problems, stress, poor medical care, diet, pollution, viruses or hereditary factors) were identified by <11% of participants.

IPQ-R dimensions were calculated for the sample as a whole and by disease subtype (Table 1), with the exception of illness identity and causes, which are described above. As the SSc group was too small to perform statistical tests, the following analysis was confined to the localized scleroderma group.

When examining child (n = 15) and parent (n = 9) scores separately, no significant differences were detected between scores in any of the dimensions with the exception of beliefs about personal control (example item: what I do to control scleroderma). Thirteen (46%) reported that scleroderma contributed to depressed mood. The most commonly reported perceived causes of scleroderma (participants could select more than one) were the immune system (54%), chance or bad luck (46%) and accidents (18%). Other causes (including personality, alcohol, smoking, family problems, stress, poor medical care, diet, pollution, viruses or hereditary factors) were identified by <11% of participants.

No relationship was detected between IPQ-R dimensions and any other clinical or demographic parameters, with the exception of CHAQ physical function scores that were positively related to greater belief in the negative consequences of scleroderma (ρ = 0.4, P < 0.05).

There are currently no published studies assessing the IPQ-R in either children or adults with localized scleroderma with which to compare our findings but the measure has been used in two studies of adult SSc [7, 11] (Table 1). Our finding of a relationship between physical function impairment and greater belief in the negative consequences was also found by Richards [7] in an adult population with SSc, and a similar association was found by Arat [11] between the negative consequences subscore and poorer physical health measured by the Short Form (36) Health Survey.

In summary, the stronger belief in personal control of localized scleroderma held by children is an interesting initial finding, although this could be influenced by older children completing their own questionnaires. Clinicians recognize that perceptions of control are important in the adjustment to and management of chronic conditions. Such perceptions are also important for parents and family members. Strategies to eliminate this mismatch may be important in facilitating parental adaptation to their child’s illness. Our findings also suggest that the impact of physical function impairment may influence beliefs about the negative consequences of localized scleroderma among affected children and their families.

**Rheumatology key message**

- Children with localized scleroderma have stronger beliefs in personal control than parents.

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**References**


Joint count reliability in psoriatic arthritis observational trials—an unreported problem

Sir, Multiple observers are a reality of large observational and multicentre studies and introduce the challenge of addressing inter-rater reliability. Long term Outcomes in Psoriatic Arthritis II (LOPAS II) is a multicentre prospective observational study investigating work disability in PsA. The primary endpoint is presenteeism (reduced effectiveness at work), but the secondary endpoints include tender and swollen joint counts. Clinical assessments will be undertaken at multiple sites across the UK. We set out to undertake a reliability exercise to estimate joint count reliability in LOPAS II.

We invited assessors from each centre to an education day at the lead site. A 1-h seminar on the study was followed by a 45-min clinical training session on joint counts led by two trainers, each with >10 years experience in PsA joint assessment. The session was followed by a joint count reliability exercise. Four patients of differing disease duration (1–33 years) and activity (from 22 tender and 9 swollen to 2 tender and 2 swollen joints as assessed by the instructors) were assessed using a modified (asymmetrical) Latin square design. Reliability was measured using Krippendorff’s $\alpha$, a reliability coefficient that accommodates the modified Latin square design [1]. Analyses were undertaken on the group as a whole and then repeated excluding those who self-reported to be unconfident or who had never performed joint assessments before.

Twelve assessors from seven units attended: one doctor, seven rheumatology nurse specialists, one occupational therapist and three research nurses (of whom one had rheumatology experience). Reliability is reported in Table 1. Inter-rater reliability for all was low irrespective of experience, but was higher among those with experience.

There are limited reports of joint count reliability among physicians with an interest in PsA [2–5]. Even among such experts, inter-class correlation coefficients are poor for determining peripheral joint swelling—0.13, 0.55, 0.24—and moderate for tenderness (or activity)—0.73, 0.75, 0.72. It is noteworthy that none of these studies has included the wider multidisciplinary team.

To our knowledge, none of the recently published large observational studies or registry reports has reported on joint count reliability [6–10]. Only the Toronto research group has reported on joint count reliability, and this was at the time of the cohort’s inception [4]. The Toronto study involved three rheumatologists and two trainees assessing five patients in a Latin square design. There was a <1% observer variance, indicating good reliability of assessment. To enable direct comparison, the analysis of variance in our study showed that the proportion of variance attributable to (all) raters was much higher; swollen joints 56% ($P=0.094$) and tender joints 60% ($P=0.004$). It is noteworthy that the Toronto study was undertaken over 20 years ago and since that time the expansion of the multidisciplinary team has meant that clinical assessments are now performed by a wider clinical team including doctors, nurses and extended scope therapists. The general lack of reporting of joint count reliability may reflect a mixture of publication bias, insufficient recognition of the potential problem or misplaced confidence. The poor reliability identified in our study is important to our current study (LOPAS II) and also to assessors from centres in the UK and further afield who collect data for other large observational studies in PsA.

The joint count training offered in the LOPAS II training day was minimal, as we had only anticipated the need for some fine tuning to standardize the assessment techniques. More training is required, as was mentioned by the assessors themselves in the feedback from the training day. We are attempting to standardize assessments and improve our reliability by using an instructional joint count training video as well as offering one-to-one tuition at the lead site. We are also encouraging a period of mentoring within each unit for those with less experience as well as aiming to use the same assessor to perform the joint counts at serial appointments. A repeat assessment day is planned once all centres have completed the training.

To our knowledge, this is the first study investigating the joint count reliability among assessors routinely contributing data in the PsA clinical and research setting. We suggest that future reporting of joint count outcomes should include some assessment of joint count reliability in order to interpret results, particularly negative findings. Furthermore, we suggest that to optimize data collection, individual units document joint count reliability with a view to determining a potential training need.

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**Table 1 Joint count reliability using Krippendorff’s $\alpha$**

<table>
<thead>
<tr>
<th>Raters</th>
<th>Swollen</th>
<th>Tender</th>
</tr>
</thead>
<tbody>
<tr>
<td>All raters</td>
<td>0.19</td>
<td>0.05</td>
</tr>
<tr>
<td>Experienced raters only</td>
<td>0.26</td>
<td>0.12</td>
</tr>
</tbody>
</table>

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Letters to the Editor

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