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Letter to the Editor (other)

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

SIR, 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 and 9 (32%) parenteral steroids. Of the 24 cases of localized scleroderma, 9 had face or head lesions, 14 trunk or limb lesions and 1 lesions 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 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 can determine whether my scleroderma gets better or worse), with children scoring significantly higher than parents ($z = -2.31$, $P = 0.03$). There was also a positive relationship between the age of the child and belief in personal control ($\rho = 0.53$, $P < 0.01$).

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 ($\rho = 0.4$, $P < 0.05$).

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

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Table 1 Demographic characteristics of the sample

	Total sample ($n = 28$)	Localized ($n = 24$)	SSc ($n = 4$)	Arat *et al*. [11] ($n = 217$)	Richards *et al.* [7] ($n = 49$)
Gender, female, *n* (%)	19 (68)	15 (63)	4 (100)	169 (78)	42 (86)
Ethnicity, white British, %	24 (86)	20 (87)	4 (100)		
Age at assessment, median (range), years	13 (5–17)	13 (5–17)	11 (7–14)	54 (46–64)	53 (12)[a]
Disease duration since diagnosis, median (range), months	30 (2–135)	22 (2–135)	68 (15–83)	5 (2–10)	9 (6)[a]
CHAQ score, median (range), 0–3	0.1 (0–1.6)	0 (0–1.6)	0.6 (0.1–1.2)	0.50 (0.12–1.25)	1.12 (0.72)[a]
IPQ-R chronicity					
Timeline	21 (13–30)	20 (13–30)	22 (20–29)	21	25[a]
Timeline cyclical	11 (5–18)	11 (5–18)	13 (11–16)	15	14[a]
IPQ-R consequences	18 (12–26)	17 (12–26)	23 (19–23)	21	23[a]
IPQ-R control					
Personal	16 (9–23)	16 (9–23)	17 (11–23)	17	17[a]
Treatment	18 (13–21)	18 (15–21)	15 (13–18)	16	15[a]
IPQ-R illness coherence	16 (5–24)	17 (5–24)	16 (10–19)	15	16[a]
IPQ-R emotional response	18 (6–28)	18 (6–28)	21 (15–28)	19	18[a]

[a]Mean (S.D.).

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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