Assessing the impact of childhood scleroderma on physical function and quality of life

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BACKGROUND

• Childhood scleroderma represents a rare and poorly understood spectrum of conditions and can be either localised or systemic.

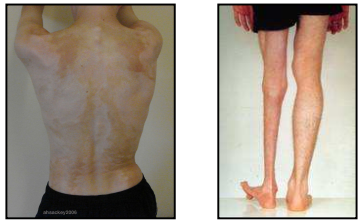


Figure 1. Manifestations of childhood scleroderma. Left: morphoea to trunk and back. Right: Contractures and limb length inequality caused by linear scleroderma.

• Few studies have assessed quality of life and physical function in childhood scleroderma.

• Quality of life measures are likely to be used in the future as part of a composite assessment to measure disease activity and estimate disease impact.

AIMS

• This cross-sectional study aimed to describe quality of life and physical function in childhood scleroderma in relation to clinical and demographic characteristics.

METHOD

• Recruitment at 4 UK hospitals: Alder Hey Children's Hospital, Liverpool; Royal Victoria Infirmary, Newcastle; Booth Hall Children's Hospital, Manchester; Royal National Hospital for Rheumatic Diseases, Bath.

•Children with either localised scleroderma or systemic sclerosis (SSc) attending paediatric rheumatology clinics invited to participate.

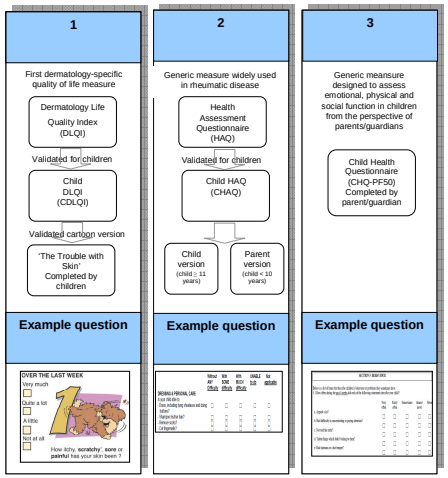
•Children, together with their parents/guardians, completed 3 validated paper-and-pencil measures (outlined in Figure 2):

- Child Health Questionnaire (CHQ-PF50)
- Children's Dermatology Life Quality Index (CDLQI)
- Child Health Assessment Questionnaire (CHAQ).

•Clinical and demographic data were provided by consultant paediatric rheumatologists using modified Paediatric Rheumatology European Society (PRES) forms.

INSTRUMENTS

Figure 2. Background to the quality of life instruments used.



RESULTS

• 28 children participated, together with their parents/guardians. Table 1 shows the demographic profile, CHAQ and CDLQI scores for the sample.

Table 1. Demographic profile of sample with CDLQI and CHAQ scores.

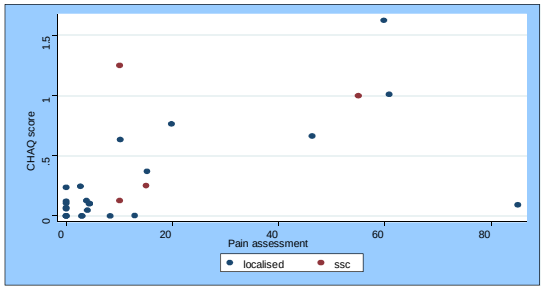
	Total sample n = 28	Localised scleroderma n = 24	SSc n = 4
Female, n(%)	19 (68)	15 (63)	4 (100)
White Caucasian, n(%)	24 (85)	20 (87)	4 (100)
Age at assessment, median (range) years	13 (5-17)	13 (5-17)	11.7 (7-14)
Disease duration, median (range) months	30 (2-135)	22 (2-35)	68 (15-83)
CHAQ physical function score, median (range), 0-3	0.1 (0-1.6)	0 (0-1.6)	0.6 (0.1-1.2)
CHAQ VAS pain score, median (range), 0-100	15 (0-85)	0 (0-85)	12.5 (10-55)
CDLQI total score, median (range), 0-30*	5 (0-10)	5 (0-10)	3 (0-6)

* CDLQI total scores available for 23 children.

• Median CHAQ and CDLQI scores suggest very moderate impairment but the wide range of scores indicates considerable variation.

• As anticipated, CDLQI scores higher for children with localised disease.

Figure 3. CHAQ function scores and VAS pain assessment in localised scleroderma and SSc.



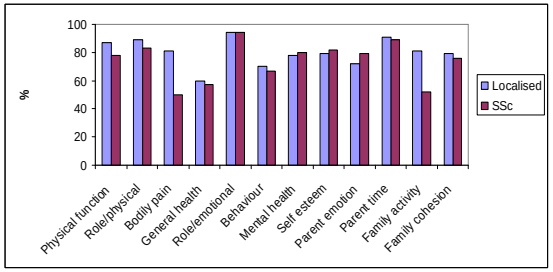
• Strong correlation between CHAQ physical function and VAS pain assessments (shown in Figure 3) p<0.001.

•Median CHQ-PF50 scores suggest only moderate impairment with greater impairment in ratings of general health, behaviour, pain and family activity (shown in Figure 4).

• Children with localised scleroderma had lower median scores in psychosocial (eg self-esteem) domains than physical domains, while the reverse was true for SSc.

•Lower self-esteem assessments significantly associated with CHAQ function and pain scores (p=0.04 and p=0.03 respectively).

Figure 4. CHQ-PF50 scores by domain (0-100 scale, with 100 = no impairment)



CONCLUSIONS

• Scleroderma had a moderate impact on quality of life and physical function within this sample, as measured by 3 validated questionnaires.

• Localised scleroderma had a more detrimental impact on psychosocial than on physical wellbeing.

• A reduction in physical function was associated with impaired self-esteem.

Alder Hey Children's NHS Foundation Trust

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