Concise Report

Health-related quality of life in children and adolescents with juvenile localized scleroderma

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Objectives. To examine the health-related quality of life (HRQOL) of children with juvenile localized scleroderma (JLS) and to compare them with patients with atopic dermatitis (AD) and healthy controls.

Methods. The cohorts were identified through a diagnostic index and were seen between January 1996 and December 2006. We identified 81 JLS patients to whom we age- and sex-matched 75 AD patients and 75 healthy controls. All patients were mailed a survey containing the English-language version of the German Revised Children's Quality of Life Questionnaire (KINDL) and the Children's Dermatology Life Quality Index (CDLQI). Linear regression models, adjusted for age and sex, examined differences in the KINDL and CDLQI scores.

Results. Survey completion rates in the JLS, AD and healthy control groups were 40, 28 and 44%, respectively. There was no difference in KINDL scores between JLS vs AD (73 vs 74, P = 0.3) and JLS vs healthy controls (73 vs 74, P = 0.8). However, CDLQI scores showed some impairment in HRQOL in JLS patients as compared with a healthy reference population, but not to the degree seen in AD (2 vs 4, P = 0.05). An exploratory analysis showed that HRQOL did not differ among the types of JLS with either measure.

Conclusion. JLS patients have some impairment in skin disease-specific HRQOL when compared with a healthy reference population, but not as severe as that seen in AD patients. Overall HRQOL in this JLS cohort was as good as healthy controls, a reassuring finding for patients, families and healthcare providers.

Key words: Juvenile localized scleroderma, Morphoea, Health-related quality of life.

Introduction

Juvenile localized scleroderma (JLS), or morphoea, is a rare non-pruritic, inflammatory autoimmune disease predominantly affecting the skin and subcutaneous tissue resulting in fibrosis [1]. JLS encompasses a spectrum of disorders with a wide range in severity for which a classification scheme was first described in 1995 and is still commonly accepted today [2].

Physicians must be aware of psychological aspects of the disease, including patients’ self-esteem and quality of life (QOL), factors that affect psychosocial development in other chronic skin disorders [3–5]. JLS has the potential to be disfiguring, particularly if the face and scalp are affected, and linear lesions traversing a joint can lead to disabling contractures or differential growth of limbs. The rarity of JLS makes health-related quality of life (HRQOL) of these patients relatively unknown.

We examined the HRQOL of JLS patients and compared them with healthy controls and with patients with atopic dermatitis (AD). We intentionally chose AD for its chronicity, early development in life, highly pruritic nature and potential to be cosmetically bothersome, all of which contribute to significant impairment in HRQOL [6]. We hypothesized that JLS patients would have better HRQOL as compared with AD patients, but poorer than healthy controls. As an exploratory analysis, we also analysed whether HRQOL differed across types of JLS and hypothesized that more severe forms affecting the face or limbs led to poorer HRQOL.

Patients and methods

This cross-sectional study was conducted at Mayo Clinic in Rochester, Minnesota, a referral centre for pediatric rheumatology and dermatology. Patients were identified through a central diagnostic index and had evaluations between 1 January 1996 and 31 December 2006. Patients were considered to have JLS if the diagnosis was made by a paediatric rheumatologist or paediatric dermatologist with or without biopsy confirmation. All consecutive patients within the study period with a JLS diagnosis were included. AD patients were chosen consecutively starting backwards from 31 December 2006 until an equal number of patients with the correct age and sex distribution were assembled. The diagnosis of AD must have been made by a pediatric dermatologist. The same patient selection process was used to select healthy controls. Exclusion criteria for all groups included patients <8 or >18 years old as of 1 January 2008; the presence of a chronic medical condition or skin disease other than those being studied; and patients without research authorization. Written consent was obtained from patients or their parents. This study was approved by and complied with the regulations in place by the institutional review board and the Pediatric and Adolescent Research Committee.

The patients’ medical records were reviewed to obtain demographic, laboratory, clinical, radiological and pathological data. We classified patients with JLS according to the Mayo Clinic criteria [2]. If patients had more than one type of morphoea, we used the predominant type for classification.

Surveys were mailed to each cohort in November 2007. The survey was re-mailed to non-responders 1 month later. They were followed up by phone during January 2008 using standardized scripts. Patients or their guardians were asked if they would like to participate. If so, the survey was re-mailed. No remuneration was offered for survey completion.

Patients with JLS or AD received an age-appropriate survey consisting of two instruments: the English language version of the German Revised Children’s Quality of Life Survey...
(KINDL) [7] and the Children’s Dermatology Life Quality Index (CDLQI) [8]. Healthy controls only received the KINDL as the CDLQI is designed for patients with skin disease and reference values for healthy children had been established during initial validation studies [8]. These surveys were designed to be completed by the patients and not by parent proxy, which was indicated in the instructions.

The KINDL is a generic questionnaire measuring overall HRQOL of children and adolescents [7]. The instrument is short, easily completed, and available for different ages. It can be completed by healthy children or those who are chronically or acutely ill. The questionnaire contains 24 Likert-scaled items ranging from 1 to 5 (‘never’ to ‘all of the time’) among six domains: physical well-being, emotional well-being, self-esteem, family, friends and everyday functioning during the prior week. An additional subscale entitled ‘disease’ is completed in case of prolonged illness or hospitalization. Raw scores are converted to a scale of 0–100, with higher scores correlating to higher HRQOL.

The KINDL questionnaire has been used in studies of over 5000 healthy and ill children and has been proven to be reliable and valid [9]. Cronbach’s-α reached 0.70 for most of the subscales whereas the overall scale displayed a consistency coefficient of 0.80 [7]. The subscales and overall score have also been highly correlated with the Child Health Questionnaire and the Short Form-36 survey [7].

The CDLQI is designed for children aged 5–16 years and measures HRQOL specifically related to skin disease over the last week [8]. Questions relate to six domains: symptoms and feelings, leisure, school or holidays, personal relationships, sleep and treatment. Scores range from 0 to 30, with higher scores indicating more HRQOL impairment. The questionnaire has been widely used and is reliable and valid [8]. Since the CDLQI was specifically developed to measure the impact of skin disease on HRQOL, this instrument is useful in comparing the HRQOL of patients with different skin diseases [8]. In the initial validation studies, children with no skin disease scored an average of 0.38 on this survey and this value was used as the healthy reference population [8].

Descriptive statistics (means, proportions, etc.) were used to summarize the data for each group. The groups had similar sex distributions, but age adjustment was needed. Linear regression models were used to examine differences in the KINDL and CDLQI scores and their subscales between the groups with adjustment for age and sex. A log-transformation was applied as the scores were not normally distributed. Comparisons of patients with different types of JLS (plaque, linear and other) were performed using Wilcoxon rank sum tests and Fisher’s exact tests. Age and sex distributions were similar for the different types of JLS, so no age and sex adjustment was performed. Responders and non-responders among the study groups were compared using chi-square and Wilcoxon rank sum tests. Because both survey instruments were validated for use up to the age of 16 years, we performed a sensitivity analysis by excluding patients aged 17 and 18 years to determine if our results differed. A P-value < 0.05 was considered statistically significant. All analyses were performed using SAS (SAS Corporation, Cary, NC, USA).

Results

We identified 81 JLS patients, 75 AD patients and 75 healthy controls. Survey response rates were 32 (40%), 21 (28%) and 33 (44%) for JLS, AD and healthy control groups, respectively. We compared baseline characteristics of survey responders and non-responders from each group (Table 1). No differences were observed between JLS responders and non-responders, or between the AD responders and non-responders. There were a greater number of females among healthy control responders compared with non-responders.

There were no differences in the median KINDL total or subscale scores between JLS and AD groups and JLS and healthy control groups (data not shown). The JLS group had impairment in skin disease-specific HRQOL as measured by the CDLQI but as compared with AD patients, it was less (median score 2 vs 4; P = 0.045) (Fig. 1). Differences were observed in the subscales ‘symptoms/feelings’ (median score 1 vs 2; P = 0.003), school/holidays (one JLS patient vs 7/21 AD patients with score >0; P = 0.006) and sleep (median score 0 vs 1; P = 0.002).

Of the 32 JLS patients that responded to the survey, 11 (34%) had plaque morphoea, 1 (3%) had generalized morphoea, 15 (47%) had linear morphoea including 2 with en coup de sabre and 5 (16%) had deep morphoea. None of the patients had bullous morphoea. Among the entire group there were eight joint contractures: two elbow, two ankle, two wrist, one finger

and one knee. One patient had lower extremity limb length discrepancy. The only significant difference among groups was the use of MTX at last visit, which was used more often in those with deep or generalized morpheaa ($P = 0.0004$). Analysis of the KINDL and CDLQI scores among the different JLS types showed no difference.

**Discussion**

We demonstrated that in patients with JLS, overall HRQOL was no different than patients with AD or healthy controls. Although skin disease-specific HRQOL was impaired in JLS patients, it was better than that observed in AD patients. Our results are reassuring to this patient population that has a potentially disfiguring disease.

This is the first study to directly examine HRQOL of patients with JLS with a skin disease-specific instrument. A previous study by Uziel et al.[10] surveyed 47 children with morphea, primarily examining self-perception using the Harter self-perception profile. A secondary aim was to examine patients’ overall QOL and quality of health using the Quality of My Life Questionnaire [11]. This questionnaire contains two visual analogue scales, one related to overall QOL and one related to HRQOL. Their results suggested that JLS patients had normal self-perception and overall QOL and HRQOL similar to healthy patients. However, their QOL measure was not skin-specific, was determined by a single item and did not have another group for comparing severity of skin disease, as our AD group in this study.

QOL studies have demonstrated the importance of the link between physical appearance and psychological health and development in children with various skin disorders such as psoriasis, acne and AD [12]. While there is some impairment in skin disease-specific HRQOL, it appears that overall HRQOL of patients with JLS is as good as their healthy counterparts without skin disease. An important caveat to this finding is that only two patients with en coup de sabre responded to the survey. Had more patients with this type of morphea responded, we may have found more impairment in HRQOL.

This study has several strengths. The referral nature of our practice allowed us to survey a large number of JLS patients, an extremely rare disease whose estimated annual age- and sex-adjusted incidence is 2.7/100,000 population [13]. Our study design allowed us to gauge the severity of HRQOL impairment that may be seen in JLS patients. We directly compared the JLS group with controls free of skin disease and with children with AD, which is known to significantly impair HRQOL. The surveys used in this study have been validated, were age-appropriate and simple to complete.

Our study has several limitations. Patients were identified using a diagnostic index and cases could have been missed if incorrectly coded. Although the surveys were intended to be completed by patients, it is impossible to ascertain if parents completed them. We minimized this possibility by providing clear instructions, but also by choosing surveys that could be easily completed by children of different age groups. We recognize that our survey response rates were low. However, we demonstrated that the survey responders and non-responders were markedly similar, suggesting that our completed surveys are likely to be representative of the sample on which they were drawn. The healthy group survey responders had a greater number of females than the non-responders. How sex would influence their answers is unknown, but our final models were sex adjusted. Disease duration appears longer in AD responders than non-responders. However, this was not statistically significant in this small cohort of patients. Larger studies are needed to determine if disease duration affects HRQOL. We did not account for disease activity, partly because there is no objective measure of disease activity in JLS. However, patients were being actively treated for their skin conditions, which suggest that the diseases were clinically bothersome to patients. This may have affected our results. We included patients outside the range for which the surveys were designed. Our sensitivity analysis suggested that even after excluding the 17- and 18-year-old patients, our results were not negatively impacted. We had very few patients in each JLS arm, hence needing to group our data as plaque, linear and other. Although our results are informative and descriptive, this analysis was purely exploratory, and would require multicentre efforts to increase sample size to better characterize differences between groups. Finally, total body surface area affected by JLS would be an important piece of information to consider in interpreting the HRQOL scores. This information was not available due to the retrospective nature of this study but should be part of any future prospective studies.

**Conclusion**

Children and adolescents with JLS have similar overall HRQOL compared with healthy children. Impairment in skin disease-specific HRQOL did not reach the level of AD patients, but was worse than healthy controls. Large multicentre studies would be needed to detect HRQOL differences across different types of JLS.

**References**


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