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The Localized Scleroderma Assessment Tool (LoSCAT): Responsiveness to Change in a Pediatric Clinical Population

Christina Kelsey, MEd and
Children's Hospital of Pittsburgh of UPMC

Kathryn Torok, MD
Children's Hospital of Pittsburgh of UPMC

Introduction

Localized scleroderma (LS) typically presents in childhood with an estimated annual age and sex adjusted incidence rate of 1–3 per 100,000 children and prevalence of 2 per 1,000 children.¹ It can cause severe physical deformity and corresponding emotional effects.^{2,3} Despite being called a 'localized' disease, one quarter of LS patients have a comorbidity, including orthopedic, neurologic, ocular, and other autoimmune conditions.^{4,5} Efforts have been made by the Childhood Arthritis and Research Alliance (CARRA) group to establish standardized treatment protocols,⁶ but research has been hindered by the lack of agreement on how to accurately capture LS treatment outcomes.

Imaging modalities, such as thermography,^{7,8} magnetic resonance imaging (MRI),⁹ Doppler flowmetry,¹⁰ and ultrasonography,^{11,12} have been proposed as potentially useful in LS. Researchers have also recommended the use of skin scores such as the Rodnan Skin Score,¹³ the Localized Scleroderma Assessment Tool (LoSCAT),^{14–16} and a computerized skin score (CSS).¹⁷ Only a few of these measurement tools - the CSS, ultrasonography, MRI, and the LoSCAT - have been validated in LS. However, all except the LoSCAT require additional, often expensive equipment and observations can be subjective even with extensive training.

The LoSCAT is easy to use and includes two LS domains: the modified Localized Skin Severity Index (mLoSSI), which measures disease activity, and the Localized Scleroderma Damage Index (LoSDI), which measures damage. Both were found to have excellent reliability and validity in prior LS studies.^{14–16} The mLoSSI was able to detect change in a small number of patients whose clinical status improved after treatment ($n = 5$, median period of 3.5 months¹⁵) and was used as an outcome measure in initial studies of systemic LS treatment.¹⁸ The goal of our study was to further evaluate the LoSCAT in pediatric LS patients to provide additional evidence for its value in clinical trials.

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Correspondent: Christina Kelsey, Pediatric Rheumatology – FP Floor 3, 4401 Penn Ave, Pittsburgh, PA 15224, christina.kelsey@chp.edu, P (412) 692-6478, F (412) 692-5054.

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The authors have no conflict of interest to declare.

Methods

To be included in the analysis, patients were required to have pediatric-onset LS, to be treated at the Children's Hospital of Pittsburgh, and be willing to complete the necessary study forms at two clinic visits. There were no restrictions on sex, age, stage of treatment, or disease status (active or inactive). An Institutional Review Board (IRB) approved consent form was signed by all patients prior to enrollment. The secondary study visit (v2), which took place during a routine follow-up clinic appointment, was required to be between 2 to 6 months from the initial visit (v1). This time frame is consistent with expected treatment response for active LS.⁶

At both study visits, the same physician (KT) completed all measures. If systemic therapy was warranted, the University of Pittsburgh (UPitt) treatment regimen for LS was administered.¹⁸ This includes high initial doses of oral prednisone (2 mg/kg/day, maximum 60 mg/day) with subsequent taper, and longer-term methotrexate by subcutaneous injection (1 mg/kg/week, maximum 25 mg/week). This regimen serves as the basis for one of the four jLS consensus treatment plans (CTP) created by the CARRA LS group⁶ and found to be effective and well tolerated in pediatric LS patients.¹⁸

Measures

The LoSCAT assesses 18 cutaneous anatomic sites, capturing both disease activity (mLoSSI) and damage (LoSDI) parameters. Scores for each site are based on the most severe score for each parameter. In order to minimize inter-subject variability, all skin changes are compared with the contralateral or ipsilateral skin area.

The mLoSSI includes the sums of 3 separate activity scores as follows: (1) **Erythema (ER)**: using the color of the lesion's edge. 0: no erythema; 1: slight erythema/pink; 2: red/clearly erythema; and 3: dark red or marked erythema/violaceous. (2) **Skin thickness (ST)**: 0: normal skin thickness and freely mobile; 1: mild increase of thickness, mobile; 2: moderate increase of thickness; impaired skin mobility; 3: marked increase of thickness or no mobility of skin. (3) **New lesion/lesion extension (N/E)**: new lesion development and/or enlargement of an existing lesion within the past month (score of 3).

Three cutaneous damage domains are summated to obtain *the LoSDI*, as follows: (1) **Dermal atrophy (DAT)**: 0: normal skin; 1: mild skin atrophy, i.e. shiny skin; 2: moderate atrophy, i.e. visible blood vessels or mild 'cliff-drop' sign; and 3: severe skin atrophy, i.e. obvious 'cliff-drop' sign. (2) **Subcutaneous atrophy (SAT)**: 0: normal subcutaneous thickness; 1: flattening or 1/3 fat loss; 2: obvious concave surface or 1/3 – 2/3 fat loss; and 3: severe subcutaneous fat loss (>2/3 loss). (3) **Dyspigmentation (DP)**: assessing both hyper- or hypopigmentation, whichever is most prominent: 0: normal skin pigment, 1: mild; 2: moderate; and 3: severe dyspigmentation.

Disease Activity Classification—At both study visits, patients were classified as having 'active' or 'inactive' disease by the treating **physician**. Active disease was defined as the presence of new, enlarging and/or erythematous lesions, as these signs often prompt an increase or change in treatment.¹⁵ Inactive disease was defined as lack of these features. The designated disease classification was compared between visits and, if change was observed, the subject was considered to be in the 'change' group. We expected most patients in the 'change' group to have an initial assessment of active disease (v1), and be designated as inactive at the follow-up visit (v2) after initiation of systemic therapy.

The Physician Global Assessment of Activity (PGA-A)—was developed based upon consensus agreement by pediatric LS experts and is typically used in conjunction with

the LoSCAT. The 100-mm analogue scale is anchored by “inactive” at 0 and “markedly active” at 100. The following cutaneous variables were included when scoring the PGA-A: new lesions within the previous month, enlargement of existing lesion within the previous month, erythema/violaceous color and/or skin thickening/induration at the border of lesion.¹⁵

The Physician Global Assessment of Damage (PGA-D)—is anchored by “no damage” at 0 and “markedly damaged” at 100. Based upon consensus agreement, both cutaneous and extracutaneous manifestations (ECM) are taken into account when scoring the PGA-D.¹⁶ The cutaneous manifestations include hyper/hypopigmentation, subcutaneous and dermal atrophy. The ECMs include musculoskeletal involvement (skeletal muscle atrophy, bone atrophy, facial atrophy, limb length discrepancy, physical disability, joint contracture), neurologic involvement (central nervous system symptoms, abnormal brain MRI, eye involvement), and psychosocial quality of life impairment.

Patients and parents (if applicable) were asked to complete a global assessment of disease severity (*Patient/Parent Global Assessments*) on a 0 to 100 analogue scale (0: no problem, 100: very severe problem) at each study visit.

The Children’s Dermatology Life Quality Index (CDLQI)—is a reliable and valid measurement of the degree to which a dermatologic disease affects one’s quality of life,¹⁹ and is appropriate to use with LS patients.²⁰ Patients answer 10 questions, which are summated to determine an overall score (from 0 to 30). Higher scores indicate that quality of life has been adversely affected.

Data Analysis

All analyses were performed using SPSS v. 20 (SPSS, Chicago, IL, USA). Mean or median was used to describe data where appropriate. Change scores were calculated by subtracting scores at v2 from v1. Clinical responsiveness was defined as the change in the LoSCAT relative to a change in physician activity classification.

Spearman’s correlations were used to examine the relationships between the change in the mLoSSI, LoSDI and clinical parameters (PGA-A/D, Parent/Patient Global Assessments, and CDLQI). Based on prior research, we expected a strongly positive correlation between the change in mLoSSI and PGA-A, and a small to moderate positive correlation between the change in LoSDI and PGA-D due to ECMs which are not included in the LoSDI score.^{15,16} We expected weak or fair correlations between changes in the LoSCAT, Parent/Patient Global Assessments and the CDLQI.

To determine contrasted group validity, patients were placed into groups based on a change in physician classification (‘change’ versus ‘no change’). Wilcoxon rank sum tests were used to compare change scores of each measure between groups. We expected the change scores of the mLoSSI, PGA-A, Parent/Patient Global Assessments and CDLQI to be significantly higher in those who experienced change than in those who did not, expecting the majority of those with active disease to improve from ‘active’ to ‘inactive’ with systemic therapy. Due to the pervasive nature of disease damage, we expected the change scores of the LoSDI and PGA-D to be relatively the same between groups.

Using a modified Jaeschke approach, we produced MCID’s using median changes scores for measures with significant contrasted group validity.^{21,22} The physician activity classification was used to anchor the MCID. To determine if MCIDs were larger than measurement error and thus practically useful, we calculated the standard error of measurement (SEM) for all relevant scores.²³

Results

Twenty-nine pediatric patients were included in the analysis, with LS subtypes and disease onset representative of the general LS population (Table 1).¹ The majority of patients had active disease at the initial study visit (18/29; 62.1 %) and the median number of affected sites was 2 (IQR = 1.5–3.00). The majority of the patients with active disease (17, 58.5%) were not on systemic medication at the time of the initial study visit (this usually corresponded to their initial UPitt rheumatology consultation); of these, 10 had no treatment and 7 were applying topical steroids. Twelve patients were on systemic treatment; 9 on the UPitt treatment regimen and 3 on methotrexate alone.

The second study visit was on average 4 months after study enrollment (median = 4.17, IQR = 3.61 – 4.96). At that time, all patients were being treated with systemic medication: 23 with the UPitt treatment regimen, 4 with methotrexate alone, one with prednisone alone, and one with mycophenolate mofetil. The majority of patients were classified as having inactive disease at their second visit (26; 89.7 %), and overall measurement scores decreased between the initial and secondary study visits.

Clinical Change between Initial and Secondary Study Visit

About half the patients (15, 51.7 %) were classified as having a change in disease activity, from active to inactive, and thus labeled as experiencing ‘change’. The physician classification of disease activity did not change for the other 14 patients (‘no change’). No patient had a negative change in activity classification. For the physician-scored activity measures, patients in the ‘change group’ had higher median change scores than patients in the ‘no change group’ (Table 2). Physician-scored damage measures were similar between groups (Table 2). Since the number of anatomic sites may have an impact on the LoSCAT, a Wilcoxon related samples test was run to determine if there was a significant difference in the number of affected sites between the initial and secondary study visit. No significant difference was found (data not shown).

Relationships with LoSCAT and Clinical Measures

Change in the mLoSSI correlated strongly and significantly with change in the PGA-A ($r_s = 0.783$, $p < 0.01$). As expected, change in the LoSDI correlated weakly with change in the PGA-D ($r_s = 0.361$, $p = 0.05$). There was a weak, negative relationship between changes in the mLoSSI and the Parent Global Assessment ($r_s = -0.391$, $p = 0.04$). There were no other significant relationships between changes in the LoSCAT and clinical parameters.

Contrasted Group Validity

The changes in the mLoSSI and the PGA-A were significantly larger in the group with a change in activity classification (from active to inactive) than the group with no change ($p < 0.01$ for both, Table 2). As expected, there were no significant differences in the changes of the LoSDI and PGA-D between groups ($p = 0.56$, 0.21 ; Table 2). There were no significant differences in the change of the Parent Global Assessment, Patient Global Assessment, or CDLQI between groups ($p = 0.10$, 0.58 , 0.32 ; Table 2).

Minimal Clinically Important Difference

MCIDs were calculated for the mLoSSI and PGA-A, which both exhibited significant contrasted group validity. Using median change scores, a MCID for the mLoSSI was determined to be 6 points (4 – 8; $n = 15$). The standard error of measurement (SEM) for the mLoSSI was determined to be 4.74. For the PGA-A, the MCID was 41 points (34–51) and the SEM was determined to be 8.61.

Discussion

Change in the mLoSSI and PGA-A were strongly related and both scores exhibited significant contrasted group validity. This is evidence that the mLoSSI and PGA-A are responsive to clinical change in pediatric-onset LS patients. Furthermore, we believe the relatively large MCID for the PGA-A accurately reflects LS disease responsiveness once systemic therapy is initiated.¹⁸ This is the first time an MCID has been associated with an LS measurement tool, and has important implications for the use of these activity measures in future treatment efficacy research. The mLoSSI and PGA-A measure slightly different concepts of LS disease activity, and together they capture 5 of the 7 components used to determine treatment response included in the recently established jLS consensus treatment plans.⁶

As expected, changes in the LoSDI and PGA-D were weakly correlated (mostly likely due to ECMs which are not captured in the LoSDI) and neither score exhibited contrasted group validity. Although the LoSDI and PGA-D were not sensitive to change in this time frame, the extent of damage is still important to discern in the course of normal treatment and clinical trials, as joint contractures, cosmetically disfiguring lesions (especially on the face), and severe subcutaneous atrophy can have a significant impact on the patient's quality of life. At the second study visit, LoSDI scores did decrease significantly when looking at the group as a whole ([median(IQR)]; $v1 = 8(5-12)$, $v2 = 6(4-9.5)$; $p < 0.01$). This is clinically relevant, especially when counseling patients about expectations of treatment; as there may be some mild (but not dramatic) improvement in disease damage.

Parent/Patient Global Assessments and the CDLQI were not found to be responsive to change. Although not the primary focus of this study, it is surprising that the relationships between the parent/patient and the physician scored assessments were found to be minimal, and negative for the mLoSSI and Parent Global Assessment. From a conceptual standpoint this seems unusual, but results in other studies looking at patient/physician agreement were found to be mixed.^{14,15,24} The changes in these measures were highly variable regardless of group (change versus no change; Table 2), as they deal with parallel but conceptually distinct perceptions of LS. Quality of life and severity from the patient's perspective might be more related to ECMs, medication side effects, and social effects (such as teasing and bullying), as opposed to the specific cutaneous manifestations measured by the LoSCAT. Further studies examining the overall constructs captured by the quality of life instruments need to be carried out before these assessments can be used independently as outcome measures in clinical trials. We agree with proposals for creating a specific LS quality of life instrument in order to meet the complexity of parent/patient concerns within this disease.¹⁵

It is possible that given a longer duration between visits, the responsiveness of the damage measures, Parent/Patient Global Assessments, and CDLQI would stabilize or become significant. From the clinical experience of the author (KT), the cutaneous damage features of the LoSDI improve over a longer follow-up period. A future prospective study with a longer duration and/or multiple follow-up visits might determine the responsiveness of these clinical parameters more clearly. In addition, it would be interesting to study if change scores were different for patients with a large number of sites, and to determine if the MCID of the mLoSSI requires modification for patients with more extensive disease (> 4 anatomic sites).

The LoSCAT is easy to learn, inexpensive to produce, and simple to use. The mLoSSI is responsive to change in disease activity, while the LoSDI captures the extent of disease damage. This study provides further evidence that the LoSCAT, along with physician global

assessments, are adequate and important response measures for studies involving treatments for pediatric-onset LS.

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List of Abbreviations

LS	Localized scleroderma
LoSCAT	Localized Scleroderma Cutaneous Assessment Tool
mLoSSI	modified Localized Scleroderma Skin Severity Index
LoSDI	Localized Scleroderma Damage Index
PGA-A	Physician Global Assessment of Disease Activity
PGA-D	Physician Global Assessment of Disease Damage
MCID	Minimal clinically important difference
SEM	standard error of measurement
CARRA	Childhood Arthritis and Research Alliance

MRI	magnetic resonance imaging
CSS	computerized skin score
IRB	Institutional Review Board
jLS	juvenile localized scleroderma
UPitt	University of Pittsburgh
CTP	consensus treatment plans
SA	surface area
ER	erythema
ST	skin thickness
N/E	new lesions or extension of lesions
DAT	dermal atrophy
SAT	subcutaneous atrophy
DP	dyspigmentation
ECM	extracutaneous manifestations
CDLQI	The Children's Dermatology Life Quality Index
V1	visit 1
V2	visit 2
IQR	interquartile range
KT	Kathryn Torok, MD, author and study clinician

Capsule summary

- There is a need for reliable, valid and responsive outcome measures in localized scleroderma (LS).
- The modified Localized Scleroderma Skin Severity Index (mLoSSI), the portion of the Localized Scleroderma Cutaneous Assessment Tool that measures disease activity, was found to be responsive to change in a sample of pediatric LS patients treated with systemic medication.
- The mLoSSI can be used as an outcome measure to determine the effectiveness of LS treatments.

Table 1

Baseline demographics of 29 pediatric patients with localized scleroderma included in this study

	Variable	n (%)
Gender	Female	22 (75.9)
	Male	7 (24.1)
Race	Caucasian	27 (93.1)
	African American	1 (3.5)
	Mixed Race	1 (3.5)
Subtypes	Linear (extremity/trunk)	17 (58.6)
	Generalized morphea	5 (17.2)
	Mixed subtype	3 (10.3)
	Plaque morphea	2 (6.9)
	Linear (head)	1 (3.5)
	Deep morphea	1 (3.5)
	Median (IQR)	
Age (years)	At disease onset	9 (5.5–11)
	At disease diagnosis	11 (8–12)
	At v1 *	12 (10–14.5)
Disease Duration [†] (months)		
	At v1 *	26 (6.5–68.5)

* v1 = initial study visit, corresponding to a normal clinic visit.

[†] Disease duration was calculated as the number of months between onset of the first localized scleroderma symptom (as per patient/parent recall) and the date of the initial study visit.

Table 2

Clinical scores of 29 pediatric localized scleroderma patients over two study visits, by group*. Changes in scores were compared between groups to determine contrasted group validity. Bolded results indicate significant median differences between groups

Score	Group* (by physician classification) Change (n = 15) No change (n = 14)	Median at v1	Median at v2	Change in score [†] (v1 – v2) Median (IQR)	Standardized Test statistic	p
Disease Activity Scores (physician)						
mLoSSI	Change	6	0	6.0 (4 – 8)	3.15	<.01
	No change	0	0	0.0 (0.0 – 1.5)		
PGA-A	Change	45	0	41.0 (34 – 51)	3.24	<.01
	No change	0	0	0.0 (0.0 – 12.5)		
Disease Damage Scores (physician)						
LoSDI	Change	7	5	2.0 (1 – 3)	0.58	.56
	No change	8	6.5	1.5 (0.0 – 3)		
PGA-D	Change	35	18	14.0 (7 – 23)	1.27	.21
	No change	36	26	6.5 (3.8 – 13.5)		
Global Disease Severity Scores (parent/patient)						
Parent Global	Change	33	22	0.0 (–8 – 24)	–1.64	.10
	No change	71	41	22.0 (7.5 – 36)		
Patient Global	Change	11	22	0.0 (–12 – 9)	–0.55	.58
	No change	41	28	10.0 (–14 – 23.5)		
Quality of Life Score (patient)						
CDLQI	Change	3	2	1.0 (–1 – 1)	–1.00	.32
	No change	6.5	6.0	1.0 (–1.5 – 6.5)		

* Physician assessment of disease activity was used to classify patients into two groups; those experiencing a change in disease activity over two time points (change) and those who did not experience a change in disease activity (no change).

[†] The changes in all clinical scores were calculated by subtracting the score at v1 by the score at v2 (v1 – v2). v1 = initial study visit, v2 = secondary study visit, mLoSSI = modified Localized Scleroderma Severity Index, PGA-A = Physician Global Assessment of Activity, LoSDI = Localized Scleroderma Damage Index, PGA-D = Physician Global Assessment of Damage, Parent Global = Parent Global Assessment of Disease Severity, Patient Global = Patient Global Assessment of Disease Severity, CDLQI = Children's Dermatology Life Quality Index.