Objectives: To assess quality of life measures in pre-pubertal subjects with SLE and examine the correlation with clinical manifestations.

Methods: In a prospective, multicenter (10 Childhood Arthritis and Rheumatology Research Alliance sites), observational study, subjects with pre-pubertal onset of SLE were enrolled. Subjects either were still in Tanner I or no later than Tanner stage II at enrollment. QOL measures were collected in addition to comprehensive medical history, physical examination findings, laboratory assessments, SLEDAI-2K scores, and SLICC/ACR Damage Index. QOL measures were obtained by PedsQL™ 4.0 Core and PedsQL™ 3.0 Rheumatology Module surveys completed by the child at the enrollment visit. QOL means were compared with disease manifestations by t tests.

Results: QOL data was available for 42 subjects (31 females, 11 males). At enrollment, the mean age of diagnosis was 9.9 years, with a mean disease duration of 1.2 years. Males were older at diagnosis than females (10.9 vs 9.5 years, $p=0.023$) with no difference in enrollment disease duration. The mean SLEDAI-2K score was 7 (range 0-18). Disease activity and individual SLE manifestations did not vary by gender. Laboratory markers of activity were present more frequently on the SLEDAI-2K than clinical exam findings, of which rash (24%), alopecia (12%) and arthritis (12%) were the most common. Seven subjects had disease damage reported on the SLICC/ACR DI.

PedsQL™ Measure	% Affected	Mean Score
PedsQL™ 4.0 Core	86%	83.8
Physical Health	67%	85.4
Emotional Functioning	69%	85.0
Social Functioning	52%	88.3
School Functioning	79%	76.4
PedsQL™ 3.0 Rheumatology Module	81%	86.2
Pain and Hurt	55%	82.6
Daily Activities	24%	95.7
Treatment	60%	86.5
Worry	40%	84.1
Communications	45%	82.1

School functioning domain had the worst QOL measure with the greatest number of children impacted (Table). Active arthritis was associated with lower mean scores in Physical Health ($p=0.007$), PedsQL Core total ($p=0.048$), Pain and Hurt ($p=0.002$), Daily Activities ($p=0.003$), Treatment ($p=0.002$), Worry ($p=0.036$), and PedsQL Rheumatology total ($p=0.001$). There was no association with QOL measures for rash, alopecia, or laboratory markers. Disease damage on SLICC/ACR DI was associated with greater impact on the Worry domain (mean 64.3 vs 88.1, $p=0.23$).

Conclusion: Children with pre-pubertal SLE onset report significant impacts on QOL regarding school. The disease manifestation affecting the most QOL domains was arthritis, whereas skin manifestations, while common, did not demonstrate an impact on QOL. This is the first study to specifically address quality of life for children who develop SLE prior to pubertal development.

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AFTER 24 MONTHS OBSERVATION PERIOD THE PATIENTS RELATED OUTCOMES IMPROVE SIGNIFICANTLY IN THE JUVENILE SCLERODERMA INCEPTIONS COHORT. WWW.JUVENILE-SCLERODERMA.COM

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Background: Juvenile systemic sclerosis (JSSc) is an orphan disease with a prevalence of 3 in a 1 000 000 children(1). Currently there is nearly

no data published about the course of JSSc patients with standardized assessment. We report our data from Juvenile Scleroderma Inception cohort (JSSIC) with a follow up of 24 months.

Objectives: to evaluate the organ involvement and patient reported outcomes (PRO) during the first 24 months in the JSIC

Methods: The JSSIC is a prospective multicenter registry of patients with JSSc, who fulfill the adult classification criteria(2), and presented the first non-Raynaud symptom before 16 years of age and were younger than 18 years at the time of inclusion in the JSSIC. Patients who were followed at least 24 months in the JSSIC, were evaluated.

Results: 52 patients were followed at least 24 months in the registry. 77% were female and 77% had diffuse subtype. 19% had overlap features. Mean disease duration at time of inclusion was 3.2 years. Mean age of at Raynaud's onset was 8.8 years and the first non-Raynaud's symptom 9.4 years. 85% received DMARDs at the time of inclusion and 96% after 24 months. 88% of the patients were ANA positive, 35% anti-Scl70 positive and 3% anticentromere positive. The mean modified skin score decreased from 14.3 to 12.6. The frequency of Raynaud's stayed around 87%. The frequency of the nail-fold capillary changes increased from 56% to 63%, but the frequency of active ulcerations stayed stable around 21%. The number of patients with FVC <80% decreased from 39% to 34% ($p=0.734$). The number of patients with pulmonary hypertension assessed by ultrasound increased from 4% to 8% ($p=0.652$). No patient developed systemic hypertension or renal crisis. Urinary sedimentary changes decreased from 8% to 4%. Gastrointestinal involvement decreased from 33% to 29% ($p=0.829$). Number of joints with decreased range increased from 46% to 63% ($p=0.076$). Total muscle weakness decreased from 8% to 3% ($p=0.237$) and elevated CK from 22% to 9% ($p=0.033$) too.

Several PROs improved significantly. Patient global disease activity (VAS 0-100) changed from 46 to 29 ($p=0.002$), patient global disease damage (VAS 0-100) from 46 to 28 ($p=0.02$) and patient Raynaud activity VAS 0-100) from 27 to 14 ($p=0.009$) as physician global disease activity (VAS 0-100) from 43 to 29 ($p=0.021$) and physician global disease damage from 46 to 28 ($P=0.01$).

Conclusion: Over the 24 months observation period patient and physician related outcomes improved significantly. Regarding organ involvement there was an increase in patients of pulmonary hypertension and joint contractures.

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