while 87.5% and 70% of patients had ANA positivity and dsDNA antibodies at the onset. The comparison between the groups with increased and normal IFN I-signature is presented in Table 1.

Table 1.

Parameters	Group 1 (High IFN-s)	Group 2 (Normal IFN-s)	p-value
Girls, n (%)	25 (80.7)	8 (88.9)	0.567
The onset age, years	12.0(10.0; 14.0)	11.0 (9.0; 13.0)	0.353
Time to IFN-signature study, months from onset	18.3 (7.0; 26.5)	0.97 (0.87;1.73)	0.987
Skin involvement, n (%)	12 (38.7)	4 (44.4)	0.837
CNS involvement, n(%)	8 (25.8)	1 (11.1)	0.353
Arthritis, n(%)	11 (35.5)	2 (22.2)	0.455
Anemia, n(%)	9 (29.0)	2 (22.20	0.687
Leucopenia, n(%)	9 (29.0)	1 (11.1)	0.274
ANA-positivity, n (%)	27 (87.1)	5 (55.6)	0.037
anti dsDNA antibodies, n(%)	12 (38.7)	2 (22.2)	0.361
Rheumatoid factor, n (%)	11 (35.5)	0 (0.0)	0.036
Hypocomplementemia, n (%)	18/28 (64.3)	2/6 (33.3)	0.162
Ferritin level, mkg/l	112.0 (39.0; 271.0)	21.0 (5.3; 23.7)	0.0008
Hematuria, n (%)	10 (32.3)	0 (0.0)	0.049
Proteinuria, n (%)	11 (35.5)	0 (0.0)	0.036
SELENA-SLEDAI, points	9 (2;15)	1 (0; 4)	0.073
ECLAM, points	3.0 (1.0; 6.0)	1.0 (0.0; 1.5)	0.048
Treatment with Rituximab or Cyclophosphamide, n (%)	22 (71.0)	3 (33.3)	0.040
GCS dose 0,2 mg/kg achievement for 6 months, n (%)	9/21 (42.9)	5/6 (83.3)	0.080

Conclusion: high IFN I-signature correlated with kidney involvement, ANA and RF-positivity, ferritinemia, proteinuria and hematuria. Patients with high IFN I-signature received more aggressive treatment and needed longer glucocorticosteroid (GCS) treatment. More meticulous dynamic evaluation of IFN-signature is needed to clarify its role as a predictive and prognostic marker.

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POS0079 PATIENTS WITH JUVENILE SYSTEMIC SCLEROSIS HAVE A DISTINCT PATTERN OF ORGAN INVOLVEMENT.RESULTS FROM THE JUVENILE SYSTEMIC SCLEROSIS INCEPTION COHORT. WWW.JUVENILE-SCLERODERMA.COM

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Background: Juvenile systemic sclerosis (jSSc) is a rare disease with a prevalence of around 3 in 1,000,000 children. To better capture the clinical manifestations of jSSc the juvenile systemic sclerosis inception cohort (jSScC) has been prospectively enrolling patients with predetermined clinical variables over the past 12 years. One of the goals is to study the demographic, clinical features, and physician and patient reported outcome differences between those with juvenile limited cutaneous (lc) compared to diffuse cutaneous (dc) disease subtypes, to determine if characteristics are similar or different between dc and lc jSSc.

Objectives: Evaluation of the baseline clinical characteristics of jSSc patients in the jSScC. Compare clinical phenotype between diffuse (dcjSSc) and limited cutaneous (lcjSSc) subtypes.

Methods: Demographic, physical examination, organ system evaluation, autoantibody profile, treatment, and patient and physician reported outcome variables were evaluated from the jSSc Inception cohort and summary statistics applied using chi-square test and Mann Whitney U-test comparing lcjSSc and dcjSSc subtypes.

Results: At the time of data extraction, 175 jSSc patients were enrolled in the cohort, 81% were Caucasian and 81% female. Diffuse cutaneous jSSc subtype predominated (73%). Mean disease duration was 3.1 year (±2.7). Mean age at Raynaud´s was 10 years (+3.8) and mean age of first non-Raynaud´s was 10.2 years (±3.8). Significant differences were found between dcjSSc versus lcjSSc, regarding several clinical characteristics. Patients with diffuse cutaneous subtype had significantly higher modified Rodnan skin score (p=0.001), presence of sclerodactyly (p=0.02), presence of Gottron's papules (p=0.003), presence of telangiectasia (p=0.001), history of digital tip ulceration (p=0.01), and frequency of elevated CK value (p=0.04). Cardiac involvement was significantly higher in limited cutaneous jSSc subtype (p=0.02). Diffuse cutaneous jSSc patients had significantly worse scores for Physician Global Assessment of disease activity (38 vs 25; p=0.002) and disease damage (34 vs 19; p=0.008).

Table 1. Comparison of demographic data and significant differences between dcjSSc and lcjSSc at time of inclusion

	Whole Cohort N=175	Diffuse Subtype N=128	Limited Subtype N=47	P value
Female to Male Ratio	4.3:1 (142/33)	4.1:1 (103/25)	4.8:1 (39/8)	0.829
Cutaneous subtype				
Diffuse subtype	73% (128)	128	0	
Limited subtype	27% (47)	0	47	
Mean Disease duration (years)	3.1 (± 2.7)	3.3 (± 2.9)	2.6 (± 2.2)	0.135
Mean age of onset of Raynaud´s (years)	10.0 (± 3.8) 17 non-Raynaud	9.8 (± 3.6) 10 non-Raynaud	10.6 (± 4.3) 7 non-Raynaud	0.219
Mean age of onset of non-Raynaud´s (years)	10.2 (± 3.9)	10.0 (± 3.7)	10.9 (± 4.3)	0.173
Disease modifying drugs	88% (154)	89% (114)	85% (40)	0.446
Cutaneous				
Mean modified Rodnan skin score	14.3 (0-51)	17.4 (0-51)	6.1 (0-24)	0.001
Gottron Papules	27% (46/171)	33% (41/124)	11% (5)	0.003
Sclerodactyly	78% (126/162)	82% (98/119)	65% (28/43)	0.020
Laboratory values	25% (30/122)	30% (26/88)	12% (4/34)	0.041
Elevated CK				
Vascular				
Telangiectasia	36% (56/154)	44% (49/111)	16% (7/43)	0.001
History of ulceration	53% (91/173)	61% (77/127)	30% (14/46)	0.001
Cardiac	6% (10)	2% (3)	15% (7)	0.002
Cardiac Involvement				
Patient Related Outcomes				
Physician global disease activity (0-100) min -max	35 (0-90) n=141	38 (0-90) n=108	25 (0-80) n=33	0.002
Physician global disease damage (0-100) min -max	31 (0-85) n=140	34 (0-85) n=108	19 (0-60) n=32	0.008

Conclusion: Results from this large international cohort of jSSc patients demonstrate significant differences between dcjSSc and lcjSSc patients. According to the general organ involvement and physician global scores, the dcjSSc patients had significantly more severe disease. These observations strengthen our previous findings of the unique organ pattern of pediatric patients.

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POS0080 TOFACITINIB TREATMENT IN CHILDREN WITH RHEUMATIC DISEASES: SINGLE-CENTER EXPERIENCE

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Background: While efficacy of tofacitinib (TOF) has been proven in many adult immune-mediated conditions, the information on its' safety and efficacy in the pediatric population is limited.

Objectives: to evaluate the safety and efficacy of TOF in children with immune-mediated diseases.

Methods: from 23 children whom TOF has been initiated, 17 children with treatment duration of > 6 months were extracted including 16 girls and 1 boy with the following diagnosis: JIA (n=10), autoinflammatory diseases (AID) (n=5) and juvenile dermatomyositis (JDM) (n=2) due to impossibility to taper corticosteroids (CS) or previous biologic treatment failed. The treatment outcome was classified according to the opinion of the attending physicians as complete response (CR) i.e., the absence of disease activity, partial response (PR) – a significant

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