resolution video nailfold capillaroscopy. We compared patients with NF+ and NF- findings from the baseline visit using chi-square test.

Results: 237 patients were included in the analysis, 185 (78%) of them were female. 126 (70%) had diffuse subtype. 183/237 patients (77%) were in the NF+ group. 71% in the NF+ group were Caucasian compared to 85% in the NF- group (p=0.051). Median disease duration was 2.3 years in the NF+ and 3.2 years in the NF- patients. Median age at onset of the first non-Raynaud´s was around 11 years in both groups. More patients in the NF+ group were ANA positive (95% compared to 79%, p< 0.001). There was no difference in the anti-Scl70 or anti-centromere distribution.

NF+ patients had significantly more frequent Raynaud phenomenon (96% compared to 78%, p< 0.001); history of digital ulcerations (59% compared to 27%, p< 0.001); abnormal high resolution CT findings of the lung (49% compared to 30%, p=0.034); overall gastrointestinal involvement (49% compared to 20%, p< 0.001); oesophageal involvement (47% compared to 19%, p< 0.001); musculoskeletal involvement (71% compared to 41%, p=0.003); presence of joints with decreased range (63% versus 45%, p=0.022) and presence of muscle weakness (25% compared to 3%, p=0.002). No significant differences were demonstrated in involvement of other organ systems such as skin, cardiac or renal. (see table 1)

Conclusion: In a jSSc cohort there were significantly more patients affected within various organ systems in those with nailfold capillary changes at enrollment compared to those without. Future studies should assess whether these differences persist over time.

[1] Vanhaecke A, Cutolo M, Distler O, et al. Nailfold capillaroscopy in SSc: innocent bystander or promising biomarker for novel severe organ involvement/progression? Rheumatology (Oxford). 2022 Nov 2;61(11):4384-4396.

[2] Foeldvari I, Klotsche J, Kasapcopur O, et al. Differences Sustained Between Diffuse and Limited Forms of Juvenile Systemic Sclerosis in an Expanded International Cohort. Arthritis Care Res (Hoboken). 2022 Oct;74(10):1575-1584.

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There Is No Difference in Major Organ Involvement Andantibody Pattern Between Diffuse and Limited Subtypejuvenile Onsetsystemic Scleroderma Patients

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SESSION INFORMATION

Session Date: Monday, November 18, 2024
Session Title: Pediatric Rheumatology – Clinical Poster III
Session Type: Poster Session C
Session Time: 10:30AM–12:30PM

Background/Purpose: In adult systemic sclerosis they are significant differences in clinical presentation of diffuse and limited subtype. In juvenile systemic sclerosis (jSSc) are the differences less prominent as we reviewed last time in a publication for the first 150 patients [1] of the juvenile scleroderma inception cohort(jSScC). The differences are changing as the included number of patients is growing in the cohort.

Methods: We extracted date from the jSScC including patients who were enrolled till 1st April 2024 into the cohort [1]. We compared the clinical characteristics, PRO and PhRO of the two subtypes and calculated statistical significance using chi-square test.

Results: 268 patients were included in the study. 70% (n=188) of the patients had diffuse subtype. Around 70% of the patients were Caucasian in both groups. The median age of onset of Raynaud´s were 10.1 in the djSSc and 11.8 years in the ljSSc. The median age at the time of the first non-Raynaud was 10.1 years in the djSSc and 11.8 years in the ljSSc. Looking at antibody we could not show any significant differences. (Table 1.) Modified Rodnan skin score (16 versus 4, p=0.001),

Table 1

Comparison diff/lim at time of inclusion in the cohort	Whole Group N=268	Diffuse Subtype N=188	Limited Subtype N=80	P value
Female to Male Ratio	3.8:1 (212/56)	3.4:1 (145/43)	5.1:1 (67/13)	
Cutaneous subtype				
Diffuse subtype	70% (188)	188	0	
Diffuse overlap	(27)	27	0	
Limited subtype	30% (80)	0	80	
Limited overlap	(30)	0	30	
Median Disease duration (years), IQR	2.5 (1 – 4.8)	2.6 (1.3 – 4.7)	2.0 (0.6 – 4.8)	n.s.
Median age at onset of Raynaud´s (years), IQR	10.4 (7.3 – 13.0)	10.1 (7.5 – 12.5)	11.8 (7 – 13.6)	n.s.
Median age at onset of non-Raynaud´s (years), IQR	10.9 (7.4 – 13.4)	10.5 (7.4 – 12.6)	12.0 (7.3 – 14.4)	n.s.
Cutaneous:				
MRSS, median (IQR)	10 (4 – 21) n=260	16 (8 - 27) n=183	4 (0 – 8) n=77	0.001
Gottron Papules	27% (72/264)	32% (59/184)	16% (13/80)	0.032
Sclerodactylie	74% (187/254)	83% (147/178)	53% (40/76)	0.011
Vascular:				
Telangiectasia	36% (89/245)	43% (73/170)	21% (16/75)	0.011
History of ulceration	52% (139/265)	61% (114/187)	32% (25/78)	0.015
Gastrointestinal Involvement				
BMI ≤ - 2 z score	15% (40/245)	20% (35/171)	6% (5/80)	0.004

Table 2

Physician Reported* (median, IQR)				
Physician global disease activity	30 (20 – 45) n=230	35 (20– 50) n=166	20 (10 – 30) n=64	0.001
Physician global disease damage	30 (15 – 45) n=228	30 (20 – 50) n=166	20 (5 – 30) n=62	0.001
Physician ulceration activity	0 (0 – 15) n=250	5 (0 – 20) n=183	0 (0 – 0.5) n=67	0.012
Patient Reported* (median, IQR)				
Patient global disease activity	40 (20 – 55) n=211	40 (20 – 55) n=156	30 (17.5 – 52.5) n=55	0.001
Patient global disease damage	35 (15 – 60) n=209	40 (20 – 60) n=154	30 (7.5 – 52.5) n=55	0.018
Patient Raynaud activity	30 (10 – 60) n=235	30 (10 – 60) n=172	20 (0 – 52.5) n=63	0.001
Patient ulceration activity	0 (0 – 30) n=236	5 (0 – 30) n=172	0 (0 – 21) n=64	0.028

more frequently Gottron papules (32% versus 16%, p=0.032), with sclerodactyly (83% versus 53%, p < 0.001), with telangiectasia (43% versus 21%, p=0.011), with history of ulceration (61% versus 32%, p< 0.015)), with decreased Body mass Index ≤2 standard deviation (20% versus 6%, p=0.004). None of the patients had renal crisis. There was no significant difference in cardiopulmonary, gastrointestinal involvement and musculoskeletal involvement. Looking at PRO and PhRO in all categories djSSc patients had significantly more severe disease (Table 2.).

Conclusion: These results present a different organ involvement pattern form adult patients. Despite more severe disease according to patient and physician reported outcomes, we found no significant differences in the cardiopulmonary, renal, gastrointestinal involvement and musculoskeletal organ involvement between the subtypes. The antibody profile anti-Scl70, anti-centromere and anti-PMscl was not different between subtypes either. It seems to be that for pediatric patients the subsetting into diffuse and limited does not make so much difference.

[1] Foeldvari I, Klotsche J, Kasapcopur O, et al. Differences Sustained Between Diffuse and Limited Forms of Juvenile Systemic Sclerosis in an Expanded International Cohort. Arthritis Care Res (Hoboken). 2022 Oct;74(10):1575-1584.

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Improvement Across Physician and Patient Reported Outcome Measures over a 24 Months-time Period in the Juvenile Systemic Scleroderma Inception Cohort

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SESSION INFORMATION

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