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Background: Juvenile systemic sclerosis (jSSc) is an orphan disease with a prevalence of 3 in 1 000 000 children (1). In adult patients there are significant differences between the clinical presentation of diffuse and limited subtypes (2). We reviewed clinical differences in presentation of subtypes in patients in the juvenile systemic scleroderma inception cohort up to 2021.
Objectives: To study the clinical presentation of jSSc patients with diffuse (djSSc) and limited (ljSSc) subtypes.
Methods: We reviewed the clinical baseline characteristics of the patients, who were recruited to the juvenile scleroderma inception cohort (jSScC) (3, 4) till 1st of December 2021. jSScC is a prospective cohort of jSSc patients, who developed the first non-Raynaud´s symptom before the age of 16 years and are under the age of 18 years at the time of inclusion.
Results: 210 patients with jSSc were included in the cohort, 71% (n=162) had diffuse subtype. The median age at onset of Raynaud phenomenon was 10.4 years (7.3 – 12.9) and the median age at the first non-Raynaud symptom was 10.9 years (7.4 – 13.2). Median disease duration was 2.5 years (1 – 4.4) at the time of inclusion. The female/male ratio was significantly lower in the djSSc subtype (3.7:1 versus 5:1, p<0.001). Antibody profile was quite similar, with the exception of a significantly higher number of anticentromere positive patients in the ljSSc (12% versus 2%, p=0.013). Decreased FVC < 80% was found in approximately 30% and decreased DLCO < 80% was found in around 40% in both subtypes. Pulmonary hypertension assessed by ultrasound was identified in 5% in both groups. Patients with diffuse subtype had significantly higher modified Rodnan Skin Score (mRSS) (16 versus 4.5, p<0.001), sclerodactyly (84% versus 60%, p<0.001), history of digital ulceration (62% versus 31%, p<0.001), decreased Body Mass Index (BMI) < -2 z score (20% versus 4%, p=0.003) and decreased joint range of motion (64% versus 46%, p=0.019). Patients with ljSSc had significantly higher rate of cardiac involvement (13% versus 2%, p=0.001).
Regarding patient related outcomes djSSc patients had more severe disease, looking at patient reported global disease activity (VAS 0 – 100) (40 versus 25, p=0.039), patient reported global disease damage (VAS 0 – 100) (40 versus 25, p=0.021) and patient reported assessment of ulceration activity (10 versus 0, p=0.044). Regarding physician related outcomes the physician reported global disease activity (VAS 0 – 100) (32 versus 20, p<0.001) and physician reported global disease damage (VAS 0 – 100) (30 versus 15, p=0.014) was significantly higher in djSSc.
Conclusion: In this jSSc cohort, the largest in the world, djSSc patients have a significantly more severe disease than ljSSc patients. Interestingly, we found no differences regarding interstitial lung disease and pulmonary hypertension.

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POS0173 DATA-DRIVEN MRI DEFINITIONS FOR ACTIVE AND STRUCTURAL SACROILIAC JOINT LESIONS IN JUVENILE SPONDYLOARTHRITIS TYPICAL OF AXIAL DISEASE

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Background: For classification in juvenile spondyloarthritis (JSpA), it is important to develop cut-offs for active and structural lesions typical of axial disease on MRI that are readily and consistently interpreted. Since the maturing sacroiliac joint (SIJ) looks different from the adult SIJ, the criteria developed for positive MRI in adults may not be applicable in JSpA.
Objectives: As part of a study developing classification criteria for axial disease in JSpA, we aimed to determine quantitative SIJ imaging lesion cut-offs for inflammatory and structural lesions typical of axial JSpA using majority imaging expert decision as the reference criterion.
Methods: Subjects were a retrospective cohort of children with SpA who met the provisional Pediatric Rheumatology International Trials Organization criteria for enthesitis/spondylitis-related juvenile idiopathic arthritis or had a rheumatologist JSpA diagnosis. All subjects had symptom onset prior to age 18 years and underwent MRI as part of a diagnostic evaluation for axial disease. To enable SIJ quadrant-based scoring, all MRIs included semi-coronal slices through the cartilaginous part of the joint on fluid sensitive sequences and on T1-weighted sequences for the assessment of inflammation and structural lesions, respectively. MRIs were reviewed by 6 musculoskeletal imaging experts who were blinded to clinical details. MRI evaluation of the SIJ was based on standardized lesion definitions that were decided by consensus of the central imaging team and represented a mix of definitions from ASAS and the Juvenile Arthritis MRI Score Outcome Measures in Rheumatology working group. Using a web-based interface, raters globally assessed the presence or absence of lesions typical of axial SpA and performed SIJ quadrant or joint based scoring. Lesion scores were generated by averaging the scores of all raters. Sensitivity and specificity of lesion cut-offs were calculated using rater majority (≥4/6 raters) on a global assessment of the presence/absence of active or structural lesions typical of axial SpA with high confidence (confidence of ±3 or stronger on confidence scale from -5, "Definitely No", to +5, "Definitely Yes") as the reference standard.
Results: Imaging from 243 subjects, 61% male, median age 14.9 years, had sequences available for detailed MRI scoring. Active inflammatory lesion typical of axial disease in JSpA was defined as bone marrow edema (BME) in at least 3 SIJ quadrants (sensitivity 98.6%, specificity 96.5%). For structural lesion typical of axial JSpA, the optimal cut-off was erosion in at least 3 quadrants or at least one of the following lesions in at least 2 SIJ quadrants: sclerosis, fat lesion, backfill, ankylosis (sensitivity 98.6%, specificity 95.5%).
Conclusion: We propose data-driven cut-offs for active inflammatory and structural lesions on MRI typical of axial disease in JSpA that have high specificity and sensitivity using central imaging global assessment as the reference standard.

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Table 1. Performance of cut-offs for inflammatory and structural lesions of axial disease

Cut-offs for number of SIJ quadrants (any location)	Sensitivity (95% CI)	Specificity (95% CI)
Definite active lesion		
BME score ≥2	100 (95.0-100)	93.5 (88.7-96.7)
BME score ≥3	98.6 (92.5-100)	96.5 (92.5-98.7)
BME, same location on ≥3 consecutive slices	88.6 (78.7-94.9)	98.8 (95.8-99.9)
Definite structural lesion		
Erosion ≥2	95.7 (88-99.1)	96.8 (92.7-99)
Erosion, same location on ≥2 consecutive slices	94.3 (86-98.4)	98.1 (94.5-99.6)
Erosion ≥3	91.4 (82.3-96.8)	98.7 (95.4-99.8)
Sclerosis ≥2	62.9 (50.5-74.1)	98.1 (94.5-99.6)
Fat lesion ≥2	22.9 (13.7-34.4%)	98.7 (95.4-99.8%)
Backfill ≥2	20 (11.4-31.3)	100 (97.7-100)
Ankylosis ≥2	1.3 (0.2-4.7)	100 (94.9-100)
ANY of the following in ≥2 SIJ quadrants: erosion, sclerosis, fat lesion, backfill, ankylosis	98.6 (92.3-100)	93.6 (88.5-96.9)
Erosion ≥3 quadrants *OR* ≥2 quadrants of at least one of the following lesions: sclerosis, fat, backfill, ankylosis	98.6 (92.3-100.0)	95.5 (91.0-98.2)

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New insights into the associations of outcomes and biomarkers in OA

POS0174 **PAIN SEVERITY ACROSS HAND OA PHENOTYPES BASED ON A BIOPSYCHOSOCIAL APPROACH**

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Background: Hand osteoarthritis (OA) pain is heterogeneous and multifactorial, involving biological, psychological and social aspects extending beyond the OA disease process. Different phenotypes may differ in prognosis and treatments requirements, and thus be of clinical importance. The presence of hand OA phenotypes has not previously been examined.
Objectives: To explore classes based on a biopsychosocial framework as possible phenotypes in a hand OA population, and examine whether pain severity differ between the classes.
Methods: Biological, psychological, and social domains were assessed including self-reported questionnaires, clinical assessment and imaging in 300 persons with hand OA. Central pain sensitization was tested by pressure pain thresholds (PPT; kg/cm^2) and temporal summation (TS). Variables used for comparison across the classes included age, sex, hand and overall bodily pain severity (Numeric Rating Scales (NRS) last 24h (0-10)). We used Latent Class Analysis followed by posterior fit statistics. Differences in age, sex and pain between classes were assessed by one-way ANOVAs and chi-squared tests.
Results: We identified 4 classes (Table 1): Class 1 had the lowest hand OA severity, high levels of pain sensitization, comorbidities, sleep problems, psychological distress and the lowest frequencies of working participants. Class 2 had the lowest pain sensitization levels and the highest proportion of participants with university education and living with a partner. Class 3 had the highest level of hand OA severity, and the lowest levels of comorbidity burden and psychological distress. Class 4 was similar to class 1 with high levels of pain sensitization, comorbidities and sleep problems and fewer with university education. Compared with class 1 they had less psychological distress and more severe hand OA.

Table 1.

	Class 1: n=25 (8%)	Class 2: n=53 (18%)	Class 3: n=103 (34%)	Class 4: n=119 (40%)	P-value
Biological domain					
OA severity	22.2 (15.8)	28.4 (15.6)	33.6 (20.1)	29.7 (20.0)	0.05
(Kellgren-Lawrence; 0-128)					
PPT tibialis anterior	3.9 (1.8)	9.4 (1.9)	5.6 (1.9)	4.2 (1.6)	< 0.001
PPT trapezius	2.8 (0.9)	7.2 (1.8)	4.5 (1.4)	3.4 (1.5)	< 0.001
TS change	2.6 (1.7)	0.8 (1.0)	1.4 (1.5)	1.9 (1.6)	< 0.001
BMI	28.4 (5.2)	25.7 (4.5)	24.9 (3.6)	27.9 (5.6)	< 0.001
Comorbidity burden (0-45)	10.7 (4.7)	7.2 (3.5)	4.9 (2.9)	9.8 (3.9)	< 0.001
Poor sleep, n (%)	24 (96%)	30 (57%)	57 (55%)	112 (94%)	< 0.001
Psychological domain					
HADS (0-42)	20.9 (5.9)	5.1 (4.4)	4.6 (4.2)	8.0 (4.5)	< 0.001
ASES (10-100)	47.7 (13.6)	73.8 (9.5)	76.6 (12.7)	63.6 (10.1)	< 0.001
PCS (0-52)	23.5 (8.3)	9.6 (7.0)	6.3 (5.3)	12.9 (7.1)	< 0.001
Social domain					
University education, n (%)	13 (52%)	40 (77%)	72 (70%)	48 (41%)	0.01
Working, n (%)	8 (32%)	38 (72%)	68 (66%)	45 (38%)	< 0.001
Living with partner, n (%)	16 (64%)	34 (71%)	70 (61%)	76 (64%)	0.03

SD=standard deviation; BMI= Body Mass Index; HADS=Hospital Anxiety and Depression Scale; PCS=Pain Catastrophizing Scale; ASES=Arthritis Self Efficacy Scale.

Hand and overall pain severity differed significantly between the classes. Class 3 reported the lowest mean (SD) NRS hand and overall pain (2.7 (1.8) and 2.8 (1.7), p=0.01). The highest mean (SD) for NRS hand and overall pain (5.8 (2.7) and 5.8 (2.4), p=0.05) where reported by Class 1, which also had the lowest mean age of 56.9 (SD 6.5), p< 0.001. Class 2 held the highest proportion of men (n=15; 29%), although not significantly different from the other classes.
Conclusion: Four potential hand OA phenotypes that differed in hand and overall pain severity were identified. The phenotypes with higher levels of pain sensitization, comorbidities, psychological and social burden tended to have more severe hand pain than persons with lower levels. The phenotypes with lowest hand OA severity reported the highest pain severity which may reflect the discordance between OA severity and pain experience. Biopsychosocial factors are likely important in defining hand OA phenotypes and may present a possibility for improved clinical outcomes.
Biopsychosocial characteristics across the proposed hand OA phenotypes. Data presented as mean (SD) unless otherwise indicated.
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