reported global disease damage (30 vs 20, p=0.011), were significantly higher in djSSc patients.
Conclusion: In the largest jSSc cohort in the world, djSSc patients have significantly more severe disease according to patient and physician related outcomes than ljSSc patients. Patients with djSSc also had more cutaneous, vascular, and musculoskeletal involvements and patients with ljSSc had more cardiac involvement. Interestingly, we found no significant differences regarding interstitial lung disease, pulmonary hypertension or gastrointestinal involvement, although the number of patients with decreased BMI ≤-2 z score was significantly higher in the djSSc patients.
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Patient Consent
Not applicable (there are no patient data)

Disclosure of Interest
None declared

P136
The pattern of medication use significantly changed over 36 months observation period. result from the juvenile scleroderma inception cohort
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Introduction: Juvenile systemic sclerosis (jSSc) is an orphan disease with a prevalence of 3 in 1,000,000 children. Currently no medications are licensed for the treatment of jSSc. Due to its rarity, only recently the first management and treatment guidelines have been published, the jSSc SHARE (Single Hub and Access point for paediatric Rheumatology in Europe) recommendations, reflecting consensus opinion upon pediatric rheumatologists(1). We reviewed the applied medication in the treatment of the patients in the juvenile systemic scleroderma inception cohort (jSScC) up to April 2023.
Objectives: To better understand the international practices of treatment for jSSc, both at baseline and over 36 months observation period and to compare if real world therapies are congruent with the recent SHARE recommendations.
Methods: We reviewed the change of the applied medication in the treatment of jSSc patients over 36 months in the jSScC. The frequency of medications was calculated across the cohort at timepoint 0 (enrollment), and 36 months. 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: We extracted data from 71 patients from the jSScC who were followed for 36 months, 75% had diffuse subtype. At the time of inclusion in the cohort the median disease duration was 2.4 years, median age of the first non-Raynaud symptom was 10.3 years. We captured the recorded medications at 0 months and 36 months. 64/71(90%) received any kind of Disease modifying drug (DMARD).
The glucocorticoid use decreased from month 0 to 36 months from 56% to 31% (p=0.004). The methotrexate use decreased from 53% to 25% (p=0.001), in opposite the mycophenolate use increased from 22% to 67% (p<0.001). The cyclophosphamide use decreased from 14% to 0% (p=0.002). Tocilizumab use increased from 0% to 17% (p=0.001). All other medication use showed no significant changes. Endothelin receptor antagonist was used in 17% patients at time point 0 and 22% at 36 months. PDE-5 blocker use increased from 3% to 9%.
Conclusion: At baseline half of the patients were on glucocorticoids. This is more frequent than typical adult SSc practice but coincides with jSSc SHARE treatment recommendations(#1)(1). After 36 months observation in the cohort over 90% of patients received a DMARD therapy. Methotrexate and mycophenolate mofetil were the most commonly prescribed DMARDs, which also reflects the SHARE treatment recommendations(#2, #3). At 36 months the use of glucocorticoids, methotrexate and cyclophosphamide decreased, and the use of mycophenolate and tocilizumab increased. In general, biological DMARDs are typically considered in severe or refractory disease (SHARE recommendation #7), reflecting the lower percentage compared to csDMARDs. Endothelial receptor antagonists, such as bosentan, were used over time in approximately 20% of the patients, reflecting SHARE recommendation #6 for pulmonary hypertension and/or digital tip ulcers. This is the first evaluation looking at clinical medication practice pattern in jSSc over 36 months, and its comparison to recently published consensus guidelines.
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Trial registration identifying number: 1. Foeldvari I, Culpo R, Sperotto F, Anton J, Avcin T, Baildam E, et al. Consensus-based recommendations for the management of juvenile systemic sclerosis. Rheumatology (Oxford). 2021;60(4):1651-8.

Patient Consent
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Disclosure of Interest
None declared

P137
Anti-topoisomerase positivity is not associated with baseline organ severity in juvenile systemic sclerosis
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Introduction: Juvenile systemic sclerosis (jSSc) is an orphan disease with a prevalence of 3 in 1 000 000 children. In adult patients anti-topoisomerase (anti-Scl70) positivity is a risk factor for diffuse cutaneous subtype and increased rate of interstitial lung disease (ILD), but data in children are scarce. Juvenile systemic scleroderma inception cohort (jSScC) is a prospective cohort of jSSc patients, who developed the first non-Raynaud´s symptom before the age of 16 years and were under the age of 18 years at the time of inclusion.
Objectives: To study and compare clinical presentation of jSSc patients, who are anti-Scl-70 positive or negative at the time of inclusion in the (jSScC).
Methods: We reviewed the baseline clinical characteristics of the patients, who were recruited to the jSScC prior to the 30th of April 2023 and compared their clinical characteristics based on anti-Scl 70 antibody positivity.
Results: 225 patients with jSSc were included in the cohort, 33% had anti-Scl70 antibodies (n=74). 68% (n=155) of patients had diffuse cutaneous subtype. There were no significant differences regarding diffuse subtype in the group with or without Scl70 antibodies (77% vs

65%; p=0.5). The median age at onset of Raynaud phenomenon was similar between two groups (10.3 years [7.6–12.8] in the anti-Scl70+ group and 10.6 years [7.2-13.1] in the anti Scl70 negative group). At the time of inclusion, median disease duration was 2.4 years and 2.5 years, respectively. The female/male ratio did not differ significantly. There were no differences between groups regarding organ involvement, including cutaneous, vascular, cardiac, renal, gastrointestinal and musculoskeletal involvement, sicca symptoms, interstitial lung disease or pulmonary hypertension. The only differences found were in the patient reported outcomes, namely in the patient global disease damage (p=0.024) and the reported Raynaud´s activity (p=0.019), which were significantly higher in the anti-Scl 70+ patients.
Conclusion: This is an intriguing finding that reinforces our previous published results of 80 jSSc patients, where the differences were not significant between anti Scl70 positive and negative patients[1]. In this study, there were differences only in the patient reported global disease damage and the patient reported Raynaud´s activity.
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Patient Consent
Not applicable (there are no patient data)

Disclosure of Interest
None declared

Reference

1. Foeldvari I, Klotsche J, Torok KS, et al. CHARACTERISTICS OF THE FIRST 80 PATIENTS AT TIMEPOINT OF FIRST ASSESSMENT INCLUDED IN THE JUVENILE SYSTEMIC SCLEROSIS INCEPTION COHORT. WWW.JUVENILESCLERODERMA.COM. Journal of Scleroderma and Related Disorders 2018;4(1-13). DOI: 10.1177/2397198318790494.

P138
improvement across organ system, physician and patient reported outcome measures over a 36-time period in the juvenile systemic scleroderma inception cohort
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Introduction: Juvenile systemic sclerosis (jSSc) is an orphan disease with a prevalence of 3 in 1, 000, 000 children. The Juvenile Systemic Scleroderma Inception cohort (jSScC) is the largest cohort of jSSc patients in the world. The jSScC collects longitudinal data prospectively in jSSc, allowing the evaluation of the development of organ involvement and patients and physician reported outcomes in jSSc over time.
Objectives: To review changes in clinical characteristics and patient and physician reported outcomes over the 36 months observation period since enrollment in the cohort.
Methods: The jSScC enrolls 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. We reviewed jSScC patient clinical data and patient and physician reported outcomes of those with 36 months follow up from the time of inclusion until 1st of April 2023.
Results: We could extract data of 74 patients, 74% with diffuse cutaneous subtype. The female/male ratio was 3.6:1. 89% of the patients were Caucasian. Median age of onset of Raynaud symptom was 9.3 years and the median age of onset of non-Raynaud symptom was 10.3 years. Median disease duration was 2.3 years at the time of inclusion in the cohort (T0). Ninety percent of the patients were treated with disease modifying anti-rheumatic drugs at T0 and 90% after 36months (T36). Four clinical parameters improved significantly over time: the median modified Rodnan skin score decreased from 10 to 7 (p=0.041), the number of patients with swollen joints decreased from 16% to 4% (p=0.014), the number of patients with elevated CK value decreased from 25% to 9% (p=0.042) and the number of patients with muscle weakness decreased from 16% to 3% (p=0.009). All other organ involvement did not show any statistically significant change from T0 to T36.
Three of the four patient reported outcomes improved significantly from T0 to T36: patient reported disease activity (VAS 0 – 100) from 40 to 20 (p=0.014), patient reported disease damage (VAS 0 – 100) from 40 to 20 (p=0.005), patient reported Raynaud activity (VAS 0 – 100) from 20 to 10 (p=0.034). One of the three physician reported outcomes improved significantly: the physician global disease activity (VAS 0 – 100) from 30 to 15 (p=0.001).
Conclusion: Skin and musculoskeletal clinical features improved significantly over 36 months. It is reassuring that major internal organ manifestations, such as cardiac, pulmonary and gastrointestinal were stable. No renal crisis occurred over the 36-month time period. The patient and physician-reported outcomes had the most positive impact over the 36 months period in this large international cohort.
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Patient Consent
Not applicable (there are no patient data)

Disclosure of Interest
None declared

P139
An updated overview of juvenile systemic scleroderma in a French cohort
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Introduction: Systemic scleroderma encompasses a range of disorders characterized by vascular and connective tissue abnormalities, leading to varying degrees of skin and internal organ inflammation and fibrosis. Although rare in pediatric patients, juvenile systemic scleroderma (JSSc) is a severe and life-threatening condition that significantly impacts children's development.