to account for correlations of repeated measures without filling in the missing data after adjusting for sex, age, and disease duration.
Results: We included 294 eligible individuals (median age at onset, 14.0 years; rheumatoid factor positive in 64.7%). The median age at baseline and disease duration was 33.8 (24.1–47.7) years and 21.0 (11.0–34.0) years, respectively. The J-HAQ was completed in all 294 patients, and 171 (58.1%) had a score of less than 0.5 at baseline, which is defined as functional remission. During the 2-year observation period, the median J-HAQ of all patients remained unchanged. There was a trend toward improvement in disease activity over time in all three composite scores. Some differeces were obsered across the three indeces: a higher proportion of patients with high disease activity and a lower proportion of patients in remission/inactive disease were observed with JADAS-27 versus SDAI and DAS28. A significant increasing trend of the estimated mean ΔJ-HAQ at 2 years after baseline was observed along with an increase in the mean disease activity during the first year measured using DAS28 (p = 0.01) and SDAI (p = 0.018), but not using JADAS-27 as shown in Table 1.

Table 1. Association of the mean disease activity categories during the first year after baseline and mean changes in J-HAQ during the two years after baseline

(n = 294)	SDAI	DAS28	JADAS-27
Remission/inactive disease	0.019 [–0.190, 0.228]	0.053 [0.024, 0.130]	0.081 [–0.072, 0.234]
LDA	0.091 [–0.150, 0.332]	0.102 [–0.013, 0.217]	0.054 [–0.102, 0.210]
MDA or HDA	0.155 [–0.286, 0.596]	0.136 [0.030, 0.242]	0.087 [0.022, 0.152]
p-value for trend	0.019	0.010	0.115

(Data are expressed as J-HAQ [95% confidence interval]. P-value less than 0.05 indicate a significant trend of the mean ΔJ-HAQ during the two years after baseline. LDA low disease activity; MDA moderate disease activity; HDA high disease activity)

Conclusion: Disease activity measured using SDAI and DAS28, but not using JADAS27, was significantly associated with subsequent changes in physical function in transitional and adult patients with JIA. This study support the use of SDAI and DAS28, but not JADAS27, in assessing disease activity in these patients to adjust treatments for preventing future deterioration of physical function.
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JUVENILE SYSTEMIC SCLEROSIS TREATMENT PRACTICES IN AN INTERNATIONAL COHORT AND COMPARISON TO RECENT SHARE CONSENSUS GUIDELINES.

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Background: Juvenile systemic scleroderma (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 have the first management and treatment guidelines been published, the jSSc SHARE (Single Hub and Access point for paediatric Rheumatology in Europe) recommendations, reflecting consensus opinion upon pediatric rheumatologists (1).
Objectives: To better understand treatment practices internationally for jSSc, both at baseline and over 24 months observation period and to compare if real world therapies are congruent with the recent SHARE recommendations.
Methods: The juvenile systemic sclerosis inceptions cohort (jSScC) is a multinational cohort that prospectively collects clinical data, including medications at baseline and subsequent visits. The jSScC enrollment criteria include age of onset of the first non-Raynaud symptom younger than 16 years and age younger than 18 years at cohort entrance. The frequency of medications (general category and specific medication) was calculated across the cohort at timepoint 0 (enrollment), 12 months and 24 months.
Results: We extracted data from the jSScC of patients who were followed for 12 or 24 months. 109 patients were followed at time point 0 (T0) and 12 months (T12), and data was available for 77 of them up at 24 months (T24). The mean age of the patients was 13.2 years at the timepoint 0. 77% were female and 75% had diffuse subtype. Disease duration at baseline visit was 3.1 years. The medications the patients were on recorded by the physician were captured at T0, T12 and T24 listed in Table 1.

Table 1.

MEDICATIONS	Time point 0 N=109	T12 months N=109	T24 months N=77
Any Medication	92% (100)	97% (106)	97% (75)
Vascular medications			
Endothelial receptor antagonist	16% (17)	24% (26)	21% (16)
PDE-5-Blocker	5% (5)	8% (9)	9% (7)
Immunomodulators			
Corticosteroids	52% (57)	44% (48)	44% (21)
All csDMARDs:	81% (88)	93% (101)	92% (71)
csDMARDs monotherapy	61% (67)	66% (72)	60% (46)
csDMARDs combination therapy	17% (18)	15% (16)	14% (11)
Methotrexate	51% (56)	50% (55)	39% (30)
Mycophenolate Mofetil	26% (28)	44% (48)	47% (36)
Hydroxychloriquine	11% (12)	15% (16)	21% (16)
Cyclophosphamide	12% (13)	2% (2)	1% (1)
Azathioprine	2% (2)	2% (2)	3% (2)
All bDMARDs:	5% (5)	14% (15)	18% (14)
bDMARDs monotherapy	2%(2)	2%(2)	1% (1)
bDMARDs combined with csDMARDs	3% (3)	12% (13)	17% (13)
Tocilizumab	2% (2)	10% (11)	14% (11)
Rituximab	2% (2)	4% (4)	4% (3)
Adalimumab	1% (1)	0% (0)	0% (0)
Autologous Stem cell transplantation	0% (0)	1% (1)	0% (0)

csDMARDs: Conventional synthetic disease-modifying antirheumatic drugs b DMARDs: Biological disease-modifying antirheumatic drugs

Conclusion: At baseline half of the patients were on corticosteroids. This is more frequent than typical adult SSc practice but coincides with jSSc SHARE treatment recommendations (#1). After 12 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 12 months the use of glucocorticoid decreased and the use of bDMARDs increased. In general, biological DMARDs are typically considered in severe or refractory (SHARE recommendation #7), reflecting the lower percentage compared to csDMARDs. Autologous stem cell transplantation was observed in one patient at 12 months, reflecting an option in jSSc with progressive and refractory disease (SHARE recommendation #8). 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, and its comparison to recently published consensus guidelines.
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