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 (22.2)	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.

Acknowledgements: This work was supported by the RSF grant № 20-45-01005.

Disclosure of Interests: None declared.

DOI: 10.1136/annrheumdis-2021-eular.1956

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 jSScC 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)	9.8 (± 3.6)	10.6 (± 4.3)	0.219
Mean age of onset of non-Raynaud's (years)	17 non-Raynaud 10.2 (± 3.9)	10 non-Raynaud 10.0 (± 3.7)	7 non-Raynaud 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				
Elevated CK	25% (30/122)	30% (26/88)	12% (4/34)	0.041
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				
Cardiac Involvement	6% (10)	2% (3)	15% (7)	0.002
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.

Supported by the "Joachim Herz Stiftung"

Disclosure of Interests: None declared.

DOI: 10.1136/annrheumdis-2021-eular.799

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

Table 1.

#	Diagnosis	Indication	Previous biologics	Current treatment	TOF, dose, mg/kg	Duration of TOF treatment, months	Genetic variants detected	Efficacy
1	AID	Severe inflammation, aortitis, colitis	INX, TCZ, ADA	CS, TOF	0.5	7	NOD2 c.2578G>A (p.A860T); NOD2 c.2722G>C (p.G908R)	PR
2	JDM	recurrent skin rash		CS, TOF, MTX	0.5	5	ADA2 c.927G>A (p.M309I)	PR
3	JDM	skin involvement, ulceration		CS, MTX, TOF	0.7	7	NLRP12c.154G>A (p.G52S)	CR
4	AID,	skin rash, recurrent inflammation,	CAN, TCZ	CS, TOF, CAN	0.5	32	RNASEH2B c.916dupA (p.I309Nfs*7)	CR
5	IFP	failure to thrive						
6	JIA, poly	severe arthritis	ETA, TCZ, ADA	TOF, TCZ	0.27	22		PR
7	soJIA	resistant to CS and biologic systemic inflammation, arthritis	TCZ, ABC, CAN	TOF, TCZ	0.4	23		PR
8	JIA, poly (RF+)	alopecia	ETA, ABC, TCZ, ADA	TOF, MTX, CS	0.3	37	IL1RN c.10G>C (p.A4P); NLRP3 c.2113C>A (p.Q705L); MEFV c.1105C>T (p.P369S)	CR
9	IFP, CANDLE-like	recurrent inflammation, digital ischemia, ulcers, CS-dependency	ETA, RTX, CAN	TOF, CS	0.25	43	MDA5	CR
10	AID, IFP	systemic inflammation, ulcers	ETA	TOF, CS	0.5	13	NLRP3	CR
11	JIA, ERA	arthritis	ETA	TOF, MTX	0.2	44		CR
12	JIA, poly	arthritis	ABC, ETA	TOF, MTX	0.25	38		CR
13	JIA, poly	Arthritis, alopecia	ETA	TOF	0.15	31		PR
14	soJIA + MAS	systemic inflammation, arthritis	TCZ, CAN, ETA	TOF, CS	0.5	38		NR
15	JIA, poly	arthritis	INX, ETA, ADA, TCZ	TOF, MTX	0.2	24		PR
16	AID	Systemic inflammation, CS-dependency	TCZ	TOF, CS	0.5	21	STAT3, c1343A>C	PR
17	JIA, ERA	arthritis	ETA	TOF	0.25	21		PR
18	JIA, poly	arthritis	ADA, TCZ, ETA	TOF	0.15	39		CR

improvement of symptoms and disease activity or no response (NR) - no changes in disease activity.

Results: Mean duration of TOF treatment was 25.4 ± 18.9 months. TOF was used as monotherapy in 3 cases, in combination with methotrexate (MTX) in 6, and in combination with other biologics in 3 children: tocilizumab (n=2) and canakinumab (n=1). Nine patients received CS. (Table 1). In two JIA patients with alopecia TOF induced intensive hair growth and controlled joint inflammation. 9 patients had CR: AID (n=3), JIA (n=4) and JDM (n=1); 7 patients had PR and 1 was NR. 13 patients had a previous history of several subsequent failed biologic: 4 biologics (n=1), 3 biologics (n=6), 2 biologics (n=1), 1 biologic (n=5). TOF treatment allowed discontinuation of CS in patient#6 and reducing the CS in 8/10 patients from 0.4 ± 0.27 mg/kg to 0.15 ± 0.1 mg/kg in 3.7 ± 3.4 times: in 2 cases the tapering of steroids failed (Figure 1). 4 patients had side effects not requiring treatment discontinuation: liver enzymes elevation (n=2), hypercholesterolemia (n=1), lymphadenitis (n=1). In pt#6 after achievement of the remission the TOF dosage was decreased up to 2 times and tocilizumab intervals were increased up to 6 weeks.

Figure 1.

Conclusion: Tofacitinib is a promising agent in treating pediatric rheumatic diseases. In our study the best results were in AID patients with rare alleles in

interferon pathway genes, patients with arthritis and alopecia and in children with JDM. Future studies are needed to identify clear indications for treatment with JAK-inhibitors.

Acknowledgements: This work was supported by the RSF grant №20-45-01005.

Disclosure of Interests: None declared.

DOI: 10.1136/annrheumdis-2021-eular.1913

POS0081 CLINICAL FEATURES OF PEDIATRIC BEHÇET'S DISEASE IN CHINA

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Background: Behçet's disease (BD) is a systemic inflammatory disease involving primarily the oral and genital mucosa, skin and eyes. Some patients exhibit broader involvement with multi-organ system inflammation, causing significant morbidity and mortality. With much of the literature focused on adult-onset BD, the clinical spectrum of BD in children is not well studied. The diagnosis of pediatric BD is difficult to make due to its rarity in childhood, the paucity of experience among pediatricians and the lack of validated diagnostic criteria.

Objectives: We aim to describe the clinical characteristic of children with Behçet's disease in China.

Methods: We performed a cross-sectional cohort analysis of patients with BD recruited between June 2017 and May 2020. We enrolled a total of 848 patients with pediatric BD (onset prior to age 18 years) and 1531 patients with adult-onset BD. Clinical data were collected by retrospective chart review.

Results: From a cohort of 848 patients with pediatric BD (412 males, 436 females), the average age of disease onset was 11.59 ± 4.11 years and the earliest case was appeared the first symptom at 7 months of age. Clinical manifestations in this group included recurrent oral aphthosis (100%), genital ulcers (77.71%), ocular involvement (30.19%), skin involvement (63.56%), neurological involvement (4.60%), gastrointestinal involvement (20.05%), vascular involvement (9.79%), heart valve involvement (3.66%), arthritis / arthralgia (32.43%), fever (0.35%) and positive pathergy test (21.11%; Table 1). The prevalence of these clinical findings was largely similar to that of adult BD patients with the exception of mild differences in the frequency of gastrointestinal, heart valve, joint involvement and fever. However, the time from symptom onset to diagnosis was significantly longer in children with BD compared to adults (13.34 ± 9.13 years vs. 6.73 ± 33.21 years, $P < 0.001$). The most common initial symptom in pediatric BD was recurrent oral aphthosis (91.57% vs. 86.28% in adults, $P < 0.001$).

In the pediatric group, clinical manifestations also differed between males and females. In addition to oral ulcers, the most common clinical manifestations in male patients were ocular involvement (42.23%), vascular involvement (15.53%), heart