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Background: In patients with JIA, growth impairment and variance in body composition are well-known long-term complications that may be associated with prolonged drug therapy (e.g. glucocorticoids) as well as impaired physical and psychosocial well-being. An increased accumulation of body fat represents a significant risk factor for metabolic abnormalities and a modifiable variable for a number of comorbidities. Recently, evidence has emerged in favour of the potential negative influence of overweight on the course of the disease and treatment response [1].

Objectives: The study aimed a) to estimate the prevalence of underweight, overweight and obesity in children and adolescents with JIA compared to the general population, and b) to investigate correlates of patients' weight status.

Methods: A cross-sectional analysis of physicians' recorded body weights and heights of patients with JIA enrolled in the NPRD in the year 2019 was performed. Underweight (BMI <10th), overweight (BMI >90th) and obesity (BMI >97th) were defined according to age- and sex-specific percentiles used in the German reference system. For comparison with data from the general population [2], sex- and age-matched pairs of 3-17-year-old patients and controls were generated. A multinomial logistic regression analysis was performed to examine the association between weight status and patients' clinical and self-reported outcomes.

Results: In total, data from 6.515 children and adolescents with JIA (age 11.2 ± 4.1 years, disease duration 4.9 ± 3.8 years, 67% girls, 40% persistent oligoarthritis) were included. Of these, 3.334 (age 5.9 ± 2.1 years, 52.5% girls) could be considered for matched-pair analysis. Compared with the general population, patients underweight, overweight and obesity rates were 10.6% (vs. 8.1%), 8.8% (vs. 8.5%) and 6.1% (vs. 5.7%), respectively. No significant sex differences were found in either group. Largest difference in prevalence was registered for underweight, specifically in the age group 3-6 years (12.9% patients vs. 5.9% controls). Similar to the general population, higher rates of overweight were observed in adolescent patients than in affected children (19.1% age group 11-13 vs. 8.4% age group 3-6). While the highest underweight prevalence was registered in patients with RF+ polyarthritis (16%), patients with Enthesitis-related arthritis (22%), psoriatic arthritis (21%) and systemic JIA (20%) showed the highest overweight rates (including obesity). Younger age (OR = 0.51, 95% CI = 0.31-0.83), more frequent physical activity (OR = 0.92, 95% CI = 0.85-0.99) and high parental vocational education (OR = 0.39, 95% CI = 0.18-0.80) were independently associated with a lower likelihood of being overweight/obese.

Conclusion: The overall prevalence of underweight, overweight and obesity in children and adolescents with JIA is comparable to that found in the general population. Behavioural health promotion, including regular physical activity, as part of the treatment strategy in JIA should preventively already begin at preschool age and necessarily be made accessible to patients of all educational levels.

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Acknowledgements: The National Paediatric Rheumatological Database has been funded by AbbVie, Chugai, Novartis and GSK.

Disclosure of Interests: Florian Milatz: None declared, Jens Klotsche: None declared, Martina Niewerth: None declared, Jana Hörstermann: None declared, Daniel Windschall: None declared, Frank Weller-Heinemann Speakers bureau: Pfizer, AbbVie, SOBI, Roche and Novartis., Frank Dressler: None declared, Rainer Berendes: None declared, Johannes-Peter Haas: None declared, Gerd Horneff: None declared, Kirsten Minden Speakers bureau: Pfizer, AbbVie, Consultant of: Novartis

DOI: 10.1136/annrheumdis-2021-eular.2058

POS1310

CANAKINUMAB IS A RESCUE TREATMENT FOR MACROPHAGE ACTIVATION SYNDROME IN PATIENTS WITH SYSTEMIC ONSET JUVENILE IDIOPATHIC ARTHRITIS: SINGLE CENTER EXPERIENCE

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Background: Macrophage activation syndrome (MAS) is a severe life-threatening complication of the systemic-onset juvenile idiopathic

arthritis (soJIA). The treatment options included high-dose of the corticosteroids (CS), cyclosporine A (CsA), intravenous immunoglobulin (IVIg) and biologics, predominantly IL-1 antagonist – anakinra. In Russia anakinra has not approved yet, so canakinumab (CAN) is a single anti-IL-1 option, available in Russia.

Objectives: To evaluate the safety and efficacy of canakinumab in patients with severe MAS in soJIA, who failed the previous treatment.

Methods: In the retrospective case series study were included 9 soJIA patients (4 boys and 5 girls) with severe MAS, resistant to combination of high-dose CS, IVIg and CsA.

Results: All patients had a MAS during the onset of JIA. The main clinical features of disease onset included: fever 9 (100%), active arthritis 5 (56%), pleuritis 7 (78%), pericarditis – 5 (56%), peritonitis 2 (22%), rash 6 (67%), hepatomegaly 9 (100%), splenomegaly 9 (100%), lymphadenopathy 7 (78%), bleeding 4 (44%), CNS involvement – 3 (33%). Initial treatment included high doses CS 8 (89%), oral CS 9 (100%), methotrexate 5 (56%), tocilizumab 4 (44%), and canakinumab 5 (56%), CsA 4 (44%), IVIg 6 (67%). TCZ was discontinued due to infusion reaction (n=2), TCZ inefficacy (n=3) and presence of MAS in all patients (n=5). In children whom TCZ was switched on CAN we used the standard dose of CAN 4 mg/kg, but if MAS occurred on the CAN we temporally increased the doses since 8 to 25 mg/kg (300 mg). In all cases MAS episodes were successfully resolved during CAN treatment. In 5 (56%) MAS had repeated course during the CAN which lead to temporally increasing the doses of CAN (pt3, pt5, pt8, pt 9) or required to increase immunosuppression with abatacept or tofacitinib. Three patients with repeated MAS developed interstitial lung disease (ILD). Two patients who successfully resolved MAS after CAN had relapses of arthritis and switched CAN to TCZ.

ID	Sex	JIA onset, y	Repeated MAS	Initial bio-logic	MAS on CAN	Experience of increased CAN doses	Outcomes	Current treatment
1	F	7.7	N	TCZ	N	N	Remission soJIA flare, ILD	CAN
2	F	11.5	N	CAN	N	N	Remission	TCZ
3	F	7.9	Y	CAN	Y	Y	Remission	CAN
4	F	3.4	N	TCZ	N	Y	Remission, ILD	CAN, CS, MMF
5	M	0.8	Y	TCZ	Y	Y	Remission, ILD	CAN, abatacept, CsA, CS
6	M	14.2	Y	CAN	N	N	Remission	TCZ
7	M	1.1	Y	CAN	N	N	Remission	N
8	M	1.3	N	CAN	Y	Y	Remission	CAN every 12 months
9	F	9.1	N	TCZ	Y	Y	Minimal disease activity	CAN, tofacitinib

Footnotes: CAN – canakinumab, CS – corticosteroids, CsA – cyclosporine A, ID – identification, ILD – interstitial lung disease, F – females, M – males, N – no, TCZ – tocilizumab, Y – yes.

Conclusion: Canakinumab is an effective rescue treatment either soJIA either MAS. In patients with MAS developed on CAN required the temporal increasing of the doses.

Funding statement: This work was supported by the Russian Foundation for Basic Research (grant № 18-515-57001).

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2021-eular.2141

POS1311

SCLERODERMA PATTERN IN NAILFOLD CAPILLARIES OF (CHILDHOOD-ONSET) SYSTEMIC LUPUS ERYTHEMATOSUS: LESSONS FROM LONGITUDINAL FOLLOW-UP

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Background: It has been suggested that a capillary scleroderma pattern in patients with systemic lupus erythematosus (SLE)-patients predisposes for clinical signs of systemic sclerosis (SSc) or overlap disease (1). However, this was previously shown not to be the case in a cross-sectional study in childhood-onset SLE (cSLE) (2).

Objectives: To assess if nailfold capillary patterns in cSLE change over time and if a capillary scleroderma pattern is associated with prospective development of clinical SSc-features, higher SLE disease activity or -damage.

Methods: Prospective clinical and capillaroscopy data were used from a longitudinal cohort of cSLE patients. Patients with a disease onset before the age of 18 years and diagnosis according to SLICC 2012 criteria were included. Disease activity was defined by SLEDAI score and disease damage by the SLICC damage index. Nailfold images from eight fingers (excluding thumbs) were obtained by videocapillaroscopy. A scleroderma pattern was defined according to the 'fast track algorithm' (3). An abnormal capillary pattern that did not match the criteria for a scleroderma pattern was defined as 'microangiopathy'.

Results: Our longitudinal study cohort consisted of n=53 cSLE patients with a median disease onset of 14 years (IQR 12.5-15.5 years) and a median SLEDAI score at diagnosis of 11 (IQR 8-15.5). Clinical follow-up data were available from 0.5-16 years after disease onset. Median disease duration at first capillaroscopy was 17 months (IQR 5.5-51.5 months) and 17/53 (32.1%) of patients had Raynaud's phenomenon. In total, n=9 (17%) showed a scleroderma pattern (at some point in time), n=37 (70%) had microangiopathy and n=7 (13%) showed a normal capillary pattern. N=27 patients had follow up with capillaroscopy (1-7 times over 1-5 years). In most patients (23/27) we did not observe any change in capillary pattern during follow-up. Two patients showed changes from microangiopathy to a scleroderma pattern, one patient from a scleroderma pattern to microangiopathy and one patient from microangiopathy to a normal pattern. Raynaud's phenomenon was equally distributed among patients with different capillaroscopy patterns (p=0.487), as was median SLEDAI score at diagnosis (p=0.285). Follow-up patients with a capillary scleroderma pattern did not show any clinical features for SSc over time (follow-up over 1-5 years, Table 1 below. Patients with a capillary scleroderma pattern showed significantly more disease damage (Chi-square, p=0.008), also indicated by a survival analysis for disease damage (Figure 1, p=0.042).

Conclusion: Our study indicates that a capillary scleroderma pattern in cSLE correlates with disease damage but not with clinical SSc features during follow-up period.

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Table 1. Clinical characteristics of cSLE-patients (n=9/53) with a nailfold capillary scleroderma pattern

	Butterfly rash	Photosensitive rash	Lupus nephritis	Autoimmune cytopenia	Positive Coombs	Low C3/C4	Serositis	Damage	Type of auto-antibodies	Follow-up in years
1	+	+	-	+	+	+	-	+	ANA, Anti-ds-DNA, anti-Sm, anti-RNP, anti-Ro52, anti-SS-A	5
2	+	+	-	+	+	+	-	+	ANA, Anti-ds-DNA, anti-Sm, anti-RNP, anti-SS-A	5
3	-	+	+	+	+	+	+	+	ANA, Anti-ds-DNA, anti-Sm, anti-RNP, anti-SS-A	10
4	-	+	+	-	+	+	+	+	ANA, Anti-ds-DNA, anti-Sm, anti-RNP, anti-Ro52, anti-SS-A	9
5	-	+	-	-	+	-	+	-	ANA	1 (lost)
6	-	+	+	+	+	+	+	-	ANA, Anti-ds-DNA, anti-c1q	3
7	-	-	-	+	-	-	-	-	ANA, Anti-ds-DNA, anti-Sm, anti-RNP	1
8	+	+	-	+	-	-	-	-	ANA	9
9	-	+	+	+	+	+	+	+	ANA, Anti-ds-DNA, anti-RNP, anti-SS-A	8

Figure 1. Occurrence of disease damage (with survival as 'no damage') in cSLE patients with microangiopathy (blue) and a capillary scleroderma pattern (red)

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2021-eular.2195

POS1312

DRUG SURVIVAL FOR IL-6 INHIBITOR TOCILIZUMAB: DATA FROM A SINGLE-CENTER OBSERVATION

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Background: The efficacy of tocilizumab for treatment patients with systemic juvenile idiopathic arthritis (sJIA) was demonstrated before. We want to describe tocilizumab drug survival based on data from a single-center observation.

Objectives: To analyze the drug survival of tocilizumab in patients with sJIA treated at the National Medical Research Center of Children's health, Moscow, Russia.

Methods: Medical records from sJIA patients treated with tocilizumab (TOC) were analyzed retrospectively from the National Medical Research Center of Children's health, Moscow, Russia.

Results: One hundred ninety-two patients presenting with sJIA were included in this observation, with a median age at treatment initiation of 7,2 (interquartile range, IQR 3,9-10,8) years and a median disease duration of 1,9 (IQR 0,4-5,9) years. All patients had been bio-naïve. TOC therapy was highly effective in patients with sJIA. At 6 month of follow-up 148/172 (86%) patients achieved inactive disease according the criteria C. Wallace, disease activity persisted in 24/172 (14%) patients. At 1 year of medication 139/150 (92%) patients had inactive disease. We analyzed the reason of TOC withdrawal retrospectively. A total of 82/192 drug withdrawals were performed. TOC was discontinued due to primary ineffectiveness in 4 patients, due to secondary ineffectiveness in 39 patients. 33 patients achieved drug-free remission.