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Background: The ankle is one of the most commonly affected sites in juvenile idiopathic arthritis (JIA). This region has a complex anatomical structure owing to the presence of multiple joint recesses and surrounding tendons. While the prognostic value of ultrasound (US)-detected arthritis has been investigated in recent studies, the role of tenosynovitis in JIA remains still unexplored.

Objectives: To investigate: 1) US features of ankle involvement in JIA at disease onset; 2) the predictive value of US-detected tenosynovitis in ankles with clinically active disease of children with new-onset JIA.

Methods: The clinical charts of all consecutive patients with new-onset JIA between May 2018 and January 2020 at study centres (Policlinico and G.Pini Hospitals of Milan) and with clinically active ankle disease among the joints affected were reviewed retrospectively. Data on ankle US assessment were retrieved and patients were then stratified as follows: 1) patients with detection on US of isolated arthritis in at least one of the joint recesses of the ankle region; 2) patients with detection on US of tenosynovitis in at least one of the tendon compartments of the ankle irrespective of the presence of concomitant arthritis. For each of these two categories, estimation of patients who were able to achieve clinical disease remission at 12 months since disease onset was evaluated.

Results: Twenty-seven new-onset JIA patients were found to have clinical involvement of the ankle among the joints affected. Nine of them (33.3%) showed on US isolated arthritis of the ankle, whereas US-detected tenosynovitis was found in 18 (66.7%) patients. The amount of patients who were able to achieve disease remission at 12-months was the same (66.7%) for both patients with and without US-detected tenosynovitis in the ankle (12/18 and 6/9 patients, respectively). In patients with US-detected tenosynovitis and clinical disease remission at 12 months, the lateral tendon compartment (LTC) was the tendon site more frequently affected by pathology (75.0%). Patients with US-detected tenosynovitis that did not achieve clinical disease remission at follow-up had the highest frequency of tendon pathology on US in the medial tendon compartment (MTC) (83.3%). The anterior tendon compartment was the less frequently affected tendon compartment of the ankle in all patients (33.3% in both patients with and without clinical remission of disease at the 12-months follow-up visit).

Conclusion: US-detected tenosynovitis of the ankle is a common finding in patients with new-onset JIA with clinically ankle disease activity and is more frequent than the detection on US of isolated arthritis. The MTC and LTC are the tendon compartments more commonly affected on US. The detection on US of tenosynovitis at disease onset in ankles with clinical disease activity did not seem to affect the change to achieve the overall clinical disease remission compared to patients without tendon pathology but with joint disease in the ankle region.

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POS1308

PREDICTORS OF DISEASE PROGRESSION IN A MONOCENTRIC COHORT OF 100 PATIENTS WITH CHILDHOOD-ONSET SYSTEMIC LUPUS ERYTHEMATOSUS

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Background: Childhood-onset systemic lupus erythematosus (cSLE) accounts for 10-20% of overall cases, usually presenting in early adolescence. SLE in children is characterized by a severe clinical course.

Objectives: This study describes the evolution of clinical manifestations of cSLE, with the aims of i) identifying biomarkers predictive of disease progression and ii) assessing the effects of treatment on disease course.

Methods: Laboratory and clinical data of cSLE patients followed in our Paediatric Rheumatology Unit were retrospectively collected at diagnosis and during follow-up. Continuous data were expressed as median (interquartile range [IQR]) and categorical data as percentages. The association between categorical variables was assessed by chi-squared test, the correlation between variables was tested by Pearson's test. Univariate linear regression analyses were performed to investigate the relationship between the rate of new disease

manifestations at follow-up and candidate predictors. Statistical analysis was performed using GraphPadPrism v6. P values <0.05 were regarded statistically significant.

Results: One-hundred patients (89% of female gender) with cSLE were included in this study, with a median age at disease onset of 13 years (10.5-15). Clinical details are presented in Table 1. Complement levels were reduced in 71 patients (C3 in 62 and C4 in 65). At a median follow-up of 118 months (57-239), the disease progressed in 93 patients, with a median of two new manifestations per patient (1-3). No correlation emerged between the number of new disease manifestations and both age at diagnosis and disease duration. Among serological biomarkers, a reduction in complement fractions at diagnosis emerged as the only predictor of new clinical manifestations due to cSLE (p=0.013 for low C3 and p=0.0004 for low C4). Among the several different pharmacological tools, hydroxychloroquine (HCQ, p=0.021, 95% confidence interval (CI) 0.0084-0.1007), azathioprine (AZA, p=0.0217, 95%CI 0.013-0.1703) and cyclophosphamide (CTX, p=0.0305, 95%CI 0.013-0.170) were identified as protective.

At follow-up, patients most commonly developed new haematological and cutaneous involvements, which were diagnosed in 43 and 11 patients, respectively. A trend towards statistical significance emerged for low C4 levels to predict new haematological involvement at follow-up (p=0.064, chi-squared: 3.42). Differently, positivity for antibodies against dsDNA emerged as the only predictor of the onset of cutaneous manifestations during follow-up (p=0.022, chi-squared: 7.62). Low C3 levels approached statistical significance in the prediction of skin involvement (p=0.058, chi-squared 5.68).

Conclusion: According to the data from our monocentric cohort of 100 patients, complement and anti-dsDNA antibodies are the most accurate tools to predict disease progression in cSLE. HCQ, AZA and CTX reduce the rate of disease progression.

Table 1. Clinical manifestations, laboratory features and treatment details of recruited patients at diagnosis.

	Number of patients
Skin manifestations	44
Haematological involvement	51
Lupus nephritis	16
Musculoskeletal involvement	72
Serositis	14
Neuropsychiatric involvement	6
Anti-nuclear antibodies	90
Anti-Ro antibodies	11
Anti-Sm antibodies	13
Anti-dsDNA antibodies	67
Anti-phospholipid antibodies	29
Hydroxychloroquine	89
Azathioprine	40
Cyclophosphamide	29
Micophenolate mofetil	13
Cyclosporine	15
Rituximab	1
Belimumab	2
Steroids	91

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POS1309

PREVALENCE AND CORRELATES OF UNDERWEIGHT, OVERWEIGHT AND OBESITY AMONG PATIENTS WITH JUVENILE IDIOPATHIC ARTHRITIS (JIA): EVIDENCE FROM THE NATIONAL PAEDIATRIC RHEUMATOLOGIC DATABASE (NPRD)

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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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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.

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POS1311

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

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