with prednisone with good clinical response but relapse of arthritis at discontinuation followed by the development of a sterile muscle abscess. An anti-TNF drug was started in both patients with complete clinical response. Patient 5 reported severe acne and psoriasis, and recurrent episodes of sterile arthritis. She presented a persistent elevation of acute phase reactants with severe anemia and leukopenia not resolving after splenectomy. His son (pts 6) presented with recurrent episodes of sterile arthritis, hepato-splenomegaly, anemia and neutropenia. Zinc and calprotectin serum levels resulted respectively 728 micromol/l and 2600 microg/ml. IL-1 inhibition determined a complete normalization of inflammatory parameters with no effects on anemia and neutropenia. In patient 6 zincemia decreased to almost normal value after 4 months of therapy. Patient 7 presented at the age of 4 years a sterile lymphnode abscess. She also presented with splenomegaly and neutropenia with persistent elevation of acute phase reactants. Anakinra was proposed but not administered for poor compliance

Conclusion: The clinical picture of patients carrying *PSTPIP1* mutation may be heterogeneous. In our cohort TNF-inhibitors were successfully used in PAPA patients preventing new arthritis episodes and resolving cutaneous manifestation where present. In 2 patients the clinical picture was mild not requiring continuous treatment. One PAMI patient had a good response to IL-1 inhibition, which however, had no effect on hematological manifestations

Pts	Mutation	Clinical features	Laboratory features	Therapy
1	A230T	Recurrent fever, acne, cutaneous abscesses, arthritis	↑CRP, ESR, SAA	Anti-TNF
2	A230T	Acne, arthritis	↑CRP, ESR, SAA	Anti-TNF
3	R405C	Recurrent fever	↑CRP, ESR, SAA, zincemia	/
4	D266N	Recurrent fever	↑CRP, ESR, SAA, zincemia	/
5	E250K	Acne, psoriasis, arthritis, hepatosplenomegaly	↑CRP, ESR, SAA, zincemia, calprotectinemia	/
6	E250K	arthritis, hepatosplenomegaly	↑CRP, ESR, SAA, zincemia, calprotectinemia	Anti IL-1
7	E250K	Lymphnode abscess, hepatosplenomegaly	↑CRP, ESR, SAA	/

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SAT0498 THE IMPACT OF OVERWEIGHT ON THE OUTCOME OF JUVENILE IDIOPATHIC ARTHRITIS

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Background: Overweight and obesity are considered to have a negative impact on Rheumatoid Arthritis in adults and there is less information regarding the correlation in juvenile idiopathic arthritis (JIA).

Objectives: To assess the effect of overweight on the activity of JIA as well as the stability and ability to achieve a remission using the cJADAS-10 score.

Methods: This is a longitudinal retrospective study design. We collected data of 164 patients suffering from JIA from three consecutive visits. Treatment was conducted between 2012 and 2015 at our centre in accordance to current guidelines. Remission was defined by cJADAS10 score ≤0.5 in Oligoarthritis and ≤0.7 in Polyarthritis. Patients were categorized by weight-for-age percentiles as heavily underweight (less than 3rd percentile) underweight (4th up to 10th percentile) healthy (11th up to 90th percentile) overweight (91th up to 96th percentile) and obese (97th up to 100th percentile). We compared the cJADAS10 of normal-weighted children with the cJADAS10 of the overweight and the obese patients, respectively.

Results: Of all patients, 13 were "underweight" (7.9%), 109 were defined as "normal weight" (66.5%) and 42 patients were categorized as "overweight" (25.6%) of which 16 children (9.8%) were "obese". 95 (57.9%) reached a remission during follow-up visits. Overweight was associated with higher disease activity compared to healthy weight children at the first visit (mean 9.5 vs. 8.5) and a wider range of the cJADAS10 score (0-22 vs. 0-20.5). Results from the 3-months-follow-up revealed an overall good response to the prescribed medication. At 6-months-follow-up, overweight children couldn't stabilize the improvement since cJADAS10 range

risks while it stays stable in healthy weight children. At the same time, while interpreting the disease activity of "overweight" and "obese" children separately, obese children show significantly less disease activity than overweight children, especially at the 3- and 6-months-follow-ups.

Conclusion: Overweight seems to have a negative influence on the disease activity and remission of JIA patients but it is most likely not the only influencing factor since obese patients show a better result regarding the cJADAS10 score than overweight patients. In the future, factors like socioeconomic status, BMI of the parents or physical activity level of the patient should be included in the evaluation. Also, the 7 subtypes of JIA should be analyzed individually since they show heterogeneous etiology, phenotype and prognosis.

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SAT0499 ORAL MICROBIOME IN CHILDREN WITH JUVENILE IDIOPATHIC ARTHRITIS IN RELATION TO DISEASE STATUS, TEMPOROMANDIBULAR JOINT ARTHRITIS AND MEDICATION: A NORWEGIAN 2-YEAR PROSPECTIVE STUDY

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Background: Juvenile idiopathic arthritis (JIA) is the most common chronic rheumatic disease in children, with an annual incidence of 1-2 per 1000 children. The temporomandibular joint (TMJ) is involved in 40-70%. The human microbiome might be a potential contributing factor to the development of JIA.

Objectives: To describe the oral salivary microbiome in children with JIA and relate this to disease activity, TMJ arthritis, and systemic medications.

Methods: 93 children; JIA (n=59), Healthy (n=34) were recruited. Demographics, disease activity, presence of TMJ-arthritis and type of medication was collected in this Norwegian prospective study (www.norjia.com). 116 saliva samples were analyzed using Next Generation Sequencing, V1-V3 region of the 16S rRNA gene, coupled with BLASTn-based, species-level taxonomy assignment algorithm. Downstream bioinformatics analysis was performed with QIIME and LEfSe.

Results: Mean age for healthy group (n=34; 27 females) is 12.3 ±3.0 years while for the JIA group (n=59; 43 females) the mean age is 12.6 ±2.7 years. A total of 541 bacterial species belonging to 111 genus and 10 phyla were identified, with Prevotella, Streptococcus, Actinomyces, Rothia Haemophilus and Veillonella accounting for the bulk of the average microbiome. There were no significant difference between JIA and healthy subjects in species richness and alpha diversity. However, differential abundance analysis revealed genera TM7-G1, Solobacterium and Mogibacterium to be associated with JIA, while Haemophilus and Lactococcus to be overabundant in healthy subjects.

Conclusion: It seems that taxa associated with chronic inflammation were found to be enriched in the saliva of JIA patients.

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SAT0500

JUVENILE RECURRENT PAROTITIS: A RARE MANIFESTATION OF RHEUMATIC IMMUNE-MEDIATED DISEASES

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Background: Juvenile recurrent parotitis (JRP) is the second most common childhood disease of the salivary glands after mumps and mainly affects children between the ages of 3 and 6. There is a male predilection, often with spontaneous resolution at puberty. However, in some cases, it can be the first manifestation of a rheumatic immune-mediated disease.

Objectives: To analyze the clinical, laboratory and imaging profile of children with JRP and investigate the prevalence of rheumatic immune-mediated diseases in these patients.

Methods: Retrospective study from 2008 to 2018 including all cases of recurrent parotitis with juvenile onset at our center. Parameters evaluated included gender, age, laterality, number of recurrences, symptoms of presentation, associated conditions, imaging details, blood tests, treatment, outcome and follow-up.

Results: 40 patients were identified over a 10-year period, with a female to male ratio of 5:3; 62.5% females. The youngest child was nine months old and the eldest was 16 years old with median age at presentation of 5 years. The median (min-max) follow-up period was 3 (0.04-33) years. 29 children (72.5%) only ever reported unilateral symptoms, while the rest had both glands affected, although not usually at the same time. Pain and swelling were the most common presenting symptoms, seen in all cases. Fever in 15 (37.5%) and whitish discharge from Stenson's duct in one (2.5%). 14 (35%) had palpable cervical lymphadenopathy and 4 (10%) had dental caries. The average reported frequency was 1.25 episodes per year, with a range of 0.43-5 episodes. Numbers of recurrences were in range of one to 15 with average of 3 times. Younger age at first episode didn't increase the likelihood of recurrences in our sample ($p=0.123$). Sonography was performed in 34 cases as primary investigation. Reports showed multiple hypoechoic lesions with enlarged intraglandular lymph nodes and enlarged gland. MRI was performed in 10, CT in 4, scintigraphy in 2 and biopsy of minor salivary glands in 1. The median level of C-reactive protein and erythrocyte sedimentation rate was 4 (range 0.2-55.5 mg/l, $n=30$) and 7 (0.5-76 mm in 1st hour, $n=20$), respectively. The median blood leukocyte count $8.13 \times 10^9/L$ (range 3.73-25.4 $\times 10^9/L$, $n = 36$). 20 (50%) patients had serum immunoglobulins measured, 6 of them having hypergammaglobulinaemia. One patient had a marginally low serum Immunoglobulin A (50; normal variation 55-210). Antibody screening was performed in 18 patients (45%), with 10 with positivity for antinuclear antibodies, 3 anti-SSA, 2 anti-SSB, 2 anti-RNP and 2 rheumatoid factor. One of the children had sialolithiasis in the parotid gland and other had an abscess with microaerophilic streptococcus isolated in culture of pus. Among all patients, 5 (12.5%) had a final diagnosis of a rheumatic immune-mediated disease: 3 with primary Sjögren Syndrome (SS) and 2 with mixed connective tissue disease. An idiopathic etiology was determined in 33 (82.5%). These were treated with analgesics and nonsteroidal antiinflammatory drugs with complete resolution of symptoms and the previous ones with immunosuppressants/immunomodulators.

Conclusion: The clinical, laboratory and imaging profile of our sample is in accord with the literature, except for sex predilection. Although recurrent juvenile parotitis is more common, primary juvenile SS should be considered. A high suspicion is crucial mainly because of a lower incidence of xerostomy and xerophthalmia in this population.

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SAT0501

THE IMPACT OF MORNING STIFFNESS ON THE DEFINITION OF INACTIVE DISEASE IN JUVENILE IDIOPATHIC ARTHRITIS

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Background: Juvenile idiopathic arthritis (JIA) is the most common chronic rheumatic disease in childhood. Morning stiffness is a major symptom of JIA, and is usually associated with active disease. The 2004 preliminary criteria for inactive disease (ID) in JIA did not include the assessment of morning stiffness, whereas the 2011 revision of the criteria has allowed the presence of morning stiffness (MS) lasting ≤ 15 minutes. MS was included in 2011 revision based on the consideration that MS of a short duration (i.e., ≤ 15 minutes) can represent residua of previously active disease without current active disease. However, it is unclear whether the disease status of children with ID who have or do not have morning stiffness is comparable.

Objectives: To compare the disease status of children with JIA who met the 2004 and 2011 revised criteria for ID in relation to the presence or absence morning stiffness.

Methods: A database of 1208 Italian children included in 2 multicenter studies (1,2) who underwent a total of 3380 visits was examined to identify all visits in which the patients fulfilled the 2004 or 2011 criteria for ID. In case a patient met the ID criteria in more than 1 visit, only the first visit was retained. For each visit with ID, the duration of morning stiffness was categorized as ≤ 15 min or > 15 min. Clinical assessments included demographic features and parent-reported outcomes

Results: A total of 668 visits in which patients met the criteria for ID were identified. Absence of morning stiffness was reported in 564 (84.4%) visits, whereas in 104 visits (15.5%) there was morning stiffness. Among the visits with morning stiffness, in 55 (8.2%) duration was ≤ 15 min, and in 49 (7.3%) duration was > 15 min. The table shows the comparison of disease duration and parent-reported outcomes between patients with absence or presence of morning stiffness.

	Patients meeting 2004 ID criteria			p-value
	No MS N = 564	MS ≤ 15 min N = 55	MS > 15 min N = 49	
Median [IQR] disease duration	2.2 [0.6, 6.3]	2.0 [0.8, 4.0]	3.4 [0.4, 7.3]	0.859
JAFS total score > 0	107 (19.0)	24 (43.6)	38 (77.6)	< 0.001
PRQL PhS > 1 SD*	61 (10.9)	22 (40.7)	38 (79.2)	< 0.001
PRQL PsS > 1 SD*	32 (5.8)	9 (17.0)	16 (33.3)	< 0.001
VAS DA > 0	170 (30.1)	44 (80.0)	43 (87.8)	< 0.001
VAS well-being > 0	211 (37.4)	42 (76.4)	44 (89.8)	< 0.001
VAS pain > 0	140 (24.8) 484	39 (70.9) 31	35 (71.4) 20	< 0.001
Remission	(87.5)	(59.6)	(41.7)	< 0.001

MS: morning stiffness; IQR: interquartile range, *above the mean of healthy children (2)

Conclusion: Among patients who met the 2011 criteria for ID, those with morning stiffness ≤ 15 min had worse parent-reported outcomes than those without morning stiffness. This finding suggests that parents may not perceive their child's disease state as true remission when lower degrees of morning stiffness are present. Notably, a sizeable proportion (7.3%) of children meeting the 2004 ID criteria had morning stiffness lasting > 15 min. The removal of the criterion "Duration of morning stiffness of ≤ 15 minutes" to "Absence of morning stiffness" in the definition for ID should be considered.

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