up of 3 years [1 – 11.7]. Apart typical cutaneo-muscular presentation, seven developed calcinosis, and six presented with subcutaneous oedema (of which 5 were anti-NXP2 positive). Median CMAS was 24/52 [1 – 47], and median MMT was 52/80 [2 – 76]. Gastro-intestinal involvement was found in 10 patients (91%), 4 presented a pulmonary impairment, and 3 girls had psychiatric symptoms. Six patients had a severe form (54%), one of them led to death (after 2.8 years of evolution).

MSA were found in 9 patients (anti-NXP2 = 1, anti-fibrillarine = 2, anti-MDA5 = 1 and anti-TIF1gamma = 1). IFN-signature was positive in 9 patients. Complement exploration was normal in all patients, and median positive rate of Anti-Factor H Ab was 260 UA/L [115 – 2800]. Muscle biopsy was performed in all patients but one with a median total score severity of 20. All patients received corticosteroids, and one to six other lines of treatment to achieve remission, obtained in 8 patients, with a median time to remission of 2 years [0.5; 4.8]. Use of plasmapheresis/immunoadsorption and/or Ruxolitinib was necessary in 3 of them (27%).

Conclusion: Presence of anti-factor H auto-antibodies in JDM or overlap myositis patients seems associated with more frequent gastro-intestinal involvement, and a more severe phenotype, requiring more aggressive treatment.

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SAT0496 THE FREQUENCY OF JOINT HYPERMOBILITY IN TURKISH SCHOOLCHILDREN: EFFECTS TO PHYSICAL ACTIVITY, BALANCE, PAIN AND QUALITY OF LIFE

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Background: Recent studies have focused on the joint hypermobility (JH) to show the association with musculoskeletal pain, functional disability, motor development and psychological distress (1). In contrast, in some publications, the negative effects of JH in childhood were not observed (2).

Objectives: The aim of this study was mainly to determine the frequency of JH in Turkish schoolchildren and to investigate whether relationship between JH and pain, physical activity level and the balance. In addition, the study aimed whether JH has an impact on the quality of life.

Methods: This cross-sectional school-based study evaluated 737 children (52.5% girls) from 8 to 14 years of age, and the data were collected in 2018 in the city of Denizli, Turkey. Firstly, each of the participants was individual assessed by a clinician on the Flamingo Balance Test for stability and Beighton for the diagnosis of JH. According to Beighton, children who scored ≥ 5 were accepted as hypermobile. Secondly, all participants completed the self-reported measures for the screening of physical activity level and quality of life. The Physical Activity Questionnaire (PAQ) and the Pediatric Quality of Life Inventory (PedsQL) tests were used to determine the level of physical activity and the quality of life. Also, pain severity was quantified by the Visual Analogue Scale (VAS) that is ranging from no pain (score: 0 mm) to worst pain (score: 100 mm) in the last month.

Results: The prevalence of JH in schoolchildren was 19.7% in Turkish population (Table 1). The mean pain severity was 1.29 ± 2.024 in all children. Significant differences were found between hypermobile and non-hypermobile groups in social and school functioning ($p < 0.05$), but no significant differences were found in pain, physical activity level ($p > 0.05$) (Table 2). Beighton score was not significant correlated with pain severity, physical activity, quality of life and balance in childhood ($p = 0.515$, $p = 0.986$, $p = 0.512$, $p = 0.362$ respectively).

Conclusion: As a result, the existence of hypermobility in children had an impact on school and social functions. However, it has been observed that hypermobility does not have a negative effect on these children's pain, balance, physical activity level, and physical and emotional functions.

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Table 1. Demographic findings of 737 Turkish Schoolchildren

Age		11.47 $\pm$ 1.304 (8-14)
Sex	Female	387 (52.5%)
	Male	350 (47.5%)
Beighton Score		2.75 $\pm$ 2.191 (0-9)
BS = 5		46 (6.3%)
BS = 6		53 (7.2%)
BS = 7		24 (3.3%)
BS = 8		15 (2.0%)
BS = 9		7 (1.0%)
Hypermobility	Positive	145 (19.7%)
	Negative	592 (80.2%)
Pain severity		1.29 $\pm$ 2.024 (0-10)

Table 2. Comparison of the results of children with and without hypermobility

	With Hypermobility (n= 145)	Without Hypermobility (n=592)	P
Age (year)	11,13 $\pm$ 1,35	11,55 $\pm$ 1,28	0.000
Weight (kg)	42,42 $\pm$ 11,85	45,58 $\pm$ 12,3	0.004
Height (cm)	148,8 $\pm$ 10,7	151,76 $\pm$ 10,19	0.002
Beighton Score	6,2 $\pm$ 1,14	1,9 $\pm$ 1,42	0.000
Pain severity (During Motion)	1,04 $\pm$ 1,8	1,35 $\pm$ 2,07	0.148
Pain severity (During Rest)	0,91 $\pm$ 1,86	1,16 $\pm$ 1,9	0.063
Balance	28,13 $\pm$ 9,44	30,04 $\pm$ 9,45	0.062
PAQ Score	27,13 $\pm$ 7	26,18 $\pm$ 7,4	0.102
PedsQL 4.0			
Physical functioning	85 $\pm$ 12,74	96,21 $\pm$ 331,71	0.068
Emotional functioning	77,05 $\pm$ 18,18	75,37 $\pm$ 20,04	0.533
Social functioning	91,67 $\pm$ 13,1	90,44 $\pm$ 29,14	0.049
School functioning	83,72 $\pm$ 13,71	80,32 $\pm$ 16,09	0.030

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SAT0497 CLINICAL PICTURE OF 7 PAPA PATIENTS FOLLOWED IN A SINGLE PEDIATRIC RHEUMATOLOGIC CENTER

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Background: Pyogenic sterile arthritis, pyoderma and acne (PAPA) syndrome is an autosomal dominant inflammatory disorder caused by mutations in the *PSTPIP1* gene primarily affecting joints and skin. The E250K mutation cause the hyperzincemia/hypercalprotectinemia syndrome termed *PSTPIP1*-associated-related proteinemia inflammatory (PAMI) syndrome in which a bone marrow involvement is reported

Objectives: To describe the clinical presentation of 7 PAPA patients followed at a single pediatric rheumatology center

Methods: For each patient clinical and laboratory data were collected from medical charts. *PSTPIP1* was sequenced through Sanger Sequencing or targeted resequencing using a customized panel, and analyzed with the NextSeq[®] platform (Illumina)

Results: We describe 7 patients from 4 unrelated families with the E250K mutation in a mother and 2 siblings, the A230T variant in a father and his son and the R405C and D266N respectively in the last 2 unrelated patients. Disease onset occurred within the 7th year of life in all patients. Patients 3 and 4 (table) presented since the 1st year of life recurrent episodes of fever without any cutaneous or articular symptoms. In both patients inflammatory markers were elevated during fever episodes, but persistently negative during wellbeing not requiring any therapy. The variants described in these patients were not previously reported. However their pathogenic role is supported by the detection of markedly high serum calprotectin levels (>10.000 microg/ml). The predominant feature of patients 1 and 2 was articular involvement with recurrent episodes of arthritis associated to acne. Patient 1 was initially treated

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.