with prednisone with good clinical response but relapse of arthritis at discontinuation followed by the development of a sterile muscle abscess. An average of 6 months (pts 6) were presented with recurrent episodes of sterile arthritis, heptato-splenomegaly, anemia, and neutropenia. Zinc and calprotectin serum levels resolved respectively 729 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 zinc chemera 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


**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 cJADAS10 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 98th percentile) and obese (99th 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 rises while it stays stable in healthy weight children. At the same time, while interpreting the disease activity of “overweight” and “obese” children significantly less children had a “inactive disease” phenotype and prognosis.

**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 some anatomic status and 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.


**SAT0499**

**ORAL MICROBIOME IN CHILDREN WITH JUVENILE IDIOPATHIC ARTHRITIS 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% of patients. 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 12.8 ±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 differences between JIA and healthy subjects in species richness and alpha diversity. However, differences between JIA and healthy subjects in species richness and alpha diversity. However, difference in 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.