with prednisone with good clinical response but relapse of arthritis at discontinuation followed by the development of a sterile muscle abscess. An anti-biostatic drug was given 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 729 microg/ml 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.

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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 3 rd percentile) underweight (4 th up to 10 th percentile) healthy (11 th up to 90 th percentile) and obese (91 th up to 96 th percentile) overweight (97 th up to 100 th percentile).

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". 97 (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 separately, obese children reached a remission statistically significantly less often 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 systemic inflammatory 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.