Uveitis, both acute and chronic, was seen in 10.8% of the children. Permanent joint affection was seen in 20.7% of the children and 8.8% have been treated with joint corrective orthopedic surgery.

At five-year follow-up, 57% of the children had disease activity, defined as arthritis and/or uveitis (2.1% with uveitis as active disease).

Conclusion: This is a well-defined, population-based and validated cohort of children diagnosed with JIA investigating long-term outcome in the era of biologics. We found that a considerable part of the children still develop uveitis, permanent joint affection and need joint corrective surgery. Surprisingly, more than 50% of the cohort has active disease five years after diagnosis. In conclusion, we still have long-term challenges in children with JIA, in spite of state-of-the-art treatment.

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THU0527

RISK SCORE OF MACROPHAGE ACTIVATION SYNDROME IN PATIENTS WITH SYSTEMIC JUVENILE IDIOPATHIC ARTHRITIS

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Background: Macrophage Activation Syndrome (MAS) is a severe, life-threatening, complication of rheumatic diseases in childhood, particularly of systemic Juvenile Idiopathic Arthritis (sJIA), occurring in approximately 25% of the patients with sJIA. A score that identifies sJIA patients who are at high risk to develop MAS would be useful in clinical practice.

Objectives: To evaluate whether routine laboratory parameters at disease onset may predict the development of MAS in patients with active sJIA. To define a risk score of MAS for sJIA patients using these parameters.

Methods: Laboratory parameters of disease activity and severity (WBC, N, PLT, Hb, ferritin, AST, ALT, gGT, LDH, TGL, fibrinogen, D-dimer and CRP), were retrospectively evaluated in 86 sJIA patients referred to our Division of Rheumatology from 1998 to 2017 with at least one year of follow-up. Laboratory parameters were evaluated during active sJIA, without MAS, at time of hospitalization (T1) and before treatment for sJIA was started (T2). Patients were divided in two groups: group 1 (patients without history of MAS), group 2 (patients with at least one MAS episode during disease course). To calculate a MAS risk score, laboratory parameters, collected at T2, with a statistical significant difference between the two groups of patients were selected.

Results: Thirty-three patients, who fulfilled the 2016 classification criteria for MAS [1] at time of sampling, were excluded from the analysis. Therefore, we analysed laboratory parameters of 53 patients with sJIA, 33 of whom without history of MAS (group 1) and 20 who developed at least one episode of MAS during disease course (group 2). Levels of ferritin, AST, LDH, gGT and TGL, collected at T2, were statistically significant higher in patients with a history of MAS compared to those without a history of MAS. For each of these parameters an arbitrary cut-off was defined. In order to define the final score an arbitrary rate was attributed to each parameter. Sensitivity (Se), specificity (Sp), positive predictive value (PPV) and negative predictive value (NPV) were calculated to define the best scoring system. The scoring system with the best sensitivity was chosen (Table 1). A MAS risk score >3 identified 19 out of 20 sJIA patients with a history of MAS and 4 out of 33 sJIA patients without history of MAS.

Abstract THU0527 –Table 1.

Table 1. Laboratory parameters and cut-off used to create the MAS risk score in sJIA patients.

Laboratory parameters	Cut-off	Rate
Ferritin (ng/ml)	>900	1
AST (U/L)	>35	1
LDH (U/L)	>550	1
gammaGT (U/L)	>30	2
Triglycerides (mg/dl)	>150	2
Sensitivity (Se)	0.950	CI95% 0.842-0.988
Specificity (Sp)	0.879	CI95% 0.753-0.948
Positive predictive value (PPV)	0.826	CI95% 0.692-0.912
Negative predictive value (NPV)	0.967	CI95% 0.865-0.995

In order to validate the MAS risk score on a different population, we applied it on 53 patients from other paediatric Rheumatologic centres. Thirty-seven of these patients without history of MAS while 16 with at least one episode of MAS. Sensitivity and specificity were 0.750 and 0.784 respectively.

Conclusion: In conclusion we developed a MAS risk score based on routine laboratory parameters that are available worldwide, that can help clinicians to identify patients at higher risk to develop MAS. A validation on a larger population is necessary.

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THU0528

DISCONTINUATION OF COLCHICINE THERAPY IN CHILDREN WITH FAMILIAL MEDITERRANEAN FEVER

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Background: Clinical phenotype of FMF exists in some carriers of MEFV mutation. These patients tend to have a mild disease. Prolonged colchicine free remission was reported in a small group of FMF patients.

Objectives: To describe and characterize a group of children with FMF in whom colchicine was discontinued.

Methods: The study cohort consisted of all children with FMF followed at 2 referral centers in Israel in whom colchicine was discontinued following prolonged attack free period.

Clinical presentation, mutations in MEFV gene and disease outcome of patients who successfully ceased colchicine therapy were compared with patients with relapse of FMF attacks.

We performed a retrospective study in two referral centers in Israel of 43 patients with FMF with 1 or non-mutated MEFV allele who ceased colchicine therapy following prolonged attack free period. The phenotype of the patients was investigated in detail, and the MEFV mutations, laboratory findings, clinical picture and outcome of 30 (70%) patients that successfully ceased colchicine therapy were compared to 13 (30%) patients who failed.

Results: 47 patients (55% males), mean age 6±3.2 years at the diagnosis, were enrolled in the study, of them 4 patients were excluded due to poor follow up. Fever (93%), abdominal pain (79%), arthralgia (19%) and arthritis (12%) were the most common symptom at attack. The average period free of attacks before enrolment was 11.3±9.2 months. The average follow-up after ceasing colchicine was 5.1±2.9 years. Thirteen patients (30.2%) had an attack during follow up with most common symptoms of fever (92%) and abdominal pain (77%) and colchicine therapy was restarted within 10.1 months (1.1-36.4months). There were no differences between the groups of patients that were able to stop