raised urinary ca creat ratio, raised urinary NAG/RBP) — 4/15 on USS and 9/15 in blood/urine tests, 2/15 on both USS and blood/urine tests. Lungs: 7 (13.5%): 6 abnormal lung function test, 1abnormal CXR, lung function& CT chest - severe interstitial lung disease & presented- progressive shortness of breath. Skin: 10(19.2%), eczema-like/non-specific skin Parotid/glandular: 1 (1.9%) on USS Splenomegaly: 4 (7.7%) on USS Sensorineural hearing loss: 2 (3.8%)
Medications to treat uveitis and/or extra-ocular manifestations were: Methotrexate 13 (25%), Methotrexate + Adalimumab 11 (21.2%), MMF 6 (11.5%), topical steroids 8 (15.4%), systemic steroids 2 (3.8%). 10 patients no medication
Conclusion: Most of the sarcoid-like *uveitis* patients had at least one extra-ocular involvement. ACE does not appear to be a sensitive biomarker. Some patients (19.2%) have a mild phenotype and require no treatment. Our data demonstrate the importance of close monitoring for extra-ocular manifestations and highlight good clinical response to steroids, MTX, MMF and anti-TNF.

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THU0525 DEVELOPMENT OF A PREDICTIVE TOOL FOR RESPONSE TO ANTI-TNF-ALPHA THERAPY IN JIA USING GENE EXPRESSION PROFILES IN PERIPHERAL DERIVED MONONUCLEAR CELLS

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Background: Heterogeneity in response to biological medication is a major challenge in the management of Juvenile Idiopathic Arthritis (JIA) (1). Biomarkers such as cytokines or whole blood RNA expression profiles that predict therapy efficacy prior to treatment pose a solution and could enable precision medicine. It was previously demonstrated that whole blood gene expression profiles can predict response to anti-TNF-α therapy in patients with JIA (2). These findings currently await validation in a multicenter prospective cohort study. Harmonization of data from different cohorts, however, is often hampered by site-dependent differences in biological sample collection and processing (3). For instance, the biological material that is retrospectively available from biobanks depends on what is stored exactly, e.g. whole blood versus peripheral blood mononuclear cells (PBMC). Furthermore, different blood collection tubes containing different additives can affect sample quality at an early stage and make interchangeability or comparability of data impossible.
Objectives: We evaluated the comparability of gene expression profiles when whole blood from patients with JIA is collected in different RNA collection tubes and processed accordingly. Next, we will investigate the predictive capacity for therapy response of RNA expression profiles from frozen PBMC of 63 JIA patients.
Methods: Peripheral blood from 11 children with non-systemic JIA with active disease was collected in PAXgene (PreAnalytiX) and Tempus (Applied Biosystems) tubes. All tubes were subsequently stored in the freezer at -80 °C. After thawing, RNA from the PAXgene tubes was extracted with use of the PAXgene Blood RNA Kit and RNA from the Tempus tubes was isolated by using the Tempus Preserved Blood RNA Purification Kit I. Gene expression of *CSNK1D*, *C1D*, *ASAP2*, *SRRD*, *PPP1R3B*, *HLA-DQA1*, *PDZK1IP1* and *MZB1* was determined with qPCR.
For our next experiment, frozen PBMC derived from patients with non-systemic JIA who are included in our longitudinal Pharmachild registry biobank will be used. Of the 63 patients, 28 (44.4%) children were diagnosed with oligo articular JIA, 19 (30.2%) with poly articular JIA, 9 (14.3%) with enthesitis-related JIA, 4 (6.3%) with psoriatic JIA and 3 (4.8%) with undifferentiated JIA. The average age at sampling was 11.5 years ± 4.0 and 65% of the patients was female. These samples will be run on a 96-analyte NanoString panel and associated with clinical response at time points 3 and 6 months after start of TNF-α blockade.
Results: Both RNA blood collection systems yielded high-quality RNA, with overall higher RNA concentrations for blood collected in Tempus tubes in comparison to PAXgene tubes. qPCR data showed that gene expression of all measured genes is affected by the method of blood collection and processing. However, the inter-individual variation was similar between both collection tubes, indicating that similar RNA profiles were observed.
Conclusion: Gene expression profiles derived from whole blood collected in PAXgene or Tempus tubes are comparable, but are not interchangeable.

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THU0526 LONG-TERM OUTCOME IN JUVENILE IDIOPATHIC ARTHRITIS – A POPULATION-BASED STUDY FROM SWEDEN

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Background: Juvenile idiopathic arthritis (JIA) is the most common rheumatic disease in children. The incidence rate among Caucasians is reported to be 8-15/100 000/year (1, 2). The long-term prognosis is insufficiently studied in JIA, although it is known that the disease often proceeds into adulthood and may cause joint damage leading to significant morbidity and physical disability. There are many challenges in elucidating the long-term prognosis. Amongst others, frequent occurrence of disease misclassification in diagnostic registers and the heterogeneity of JIA have made it difficult to interpret results (3, 4).
Objectives: To study the epidemiology, incidence and long-term outcome of JIA in southern Sweden using a population-based cohort of children with a validated diagnosis of JIA collected over nine years.
Methods: Potential cases of JIA, diagnosed between 2002 and 2010 were collected from a local search at the Department of Rheumatology in Lund and at the National Board for Health and Welfare, using the ICD-codes M08-M09. The study area is Skåne, the southernmost county of Sweden (population 1.24 million; 19.1% aged < 16 years) and the median follow-up time was eight years. The JIA-diagnosis was validated and subcategorised through medical record review based on criteria defined by The International League of Associations for Rheumatism. Parameters on disease activity and pharmacologic treatment were recorded annually until the end of the study period.
Results: 251 cases of JIA were confirmed. The mean annual incidence rate for JIA was estimated to be 12.8/100 000/year. The highest age specific annual incidence is at the age of two years (36/100 000/year). This peak is consistent in the female group, but the incidence peak among males is at twelve years of age.
Almost all patients are at some point during their disease course prescribed non-steroidal anti-inflammatory drugs (98%). Intra-articular steroid injections are also frequently used (78.9% in the total cohort). Methotrexate is the most common disease modifying anti-rheumatic drug prescribed (60.6%). Tumor necrosis factor alpha-inhibitors are used as treatment in 23.9% of the children.
Oligoarthritis was the largest subgroup (44.7%), followed by undifferentiated JIA (16.3%), polyarticular rheumatoid factor negative JIA (13.9%), enthesitis-related arthritis (8.8%), polyarticular rheumatoid factor positive JIA (6.8%), juvenile psoriatic arthritis (6.8%) and systemic JIA as the smallest subgroup (2.8%).

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 (UI/L)	>35	1
LDH (UI/l)	>550	1
gammaGT (UI/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