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.

REFERENCES:

- [1] Thierry S, et al. Joint, bone, spine. 2014;81(2):112-7.
- [2] Berntson L, et al. J Rheumatol. 2003;30:2275-2282.
- [3] Harrold LR, et al. J Rheumatol. 2013;40(7):1218-25.
- [4] Ravelli A, et al. Lancet. 2007;369(9563):767-78.

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2019-eular.1579

THU0527

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

Simone Carbogno¹, Denise Pires Marafon², Giulia Marucci³, Manuela Pardeo³, Antonella Insalaco³, Virginia Messina³, Emanuela Sacco³, Ferhat Demir⁴, Betül Sözeri⁴, Alenka Gagro⁵, Nastasia Kifer⁶, Marija Jelusic⁶, Francesca Minoia⁷, Mikhail Kostik⁸, Olga Vougiouka⁹, Fabrizio De Benedetti³, Claudia Braccaglia³, MAS/sJIA PreS working party. ¹University of Milan, Pediatric Area, Milan, Italy; ²Fondazione IRCCS Ca' Grande Ospedale Maggiore Policlinico, Pediatric Unit, Milan, Italy; ³IRCCS Ospedale Pediatrico Bambino Gesù, Division of Rheumatology, Rome, Italy; ⁴Umraniye Training and Research Hospital, Division of Pediatric Rheumatology, Istanbul, Turkey; ⁵Children's Hospital Zagreb, Zagreb, Croatia; ⁶University of Zagreb, Division of Paediatric Rheumatology and Immunology, Zagreb, Croatia; ⁷Fondazione IRCCS Ca' Grande Ospedale Maggiore Policlinico, Pediatric Rheumatology, Milan, Italy; ⁸Saint-Petersburg State Pediatric Medical University, Saint-Petersburg, Russian Federation; ⁹Athens Medical School, National and Kapodistrian University of Athens, Second Department of Paediatrics, Athens, Greece

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.

REFERENCES:

- [1] Ravelli A et al. Ann Rheum Dis. 2016 Mar;75(3):481-9.

Disclosure of Interests: Simone Carbogno: None declared, Denise Pires Marafon: None declared, Giulia Marucci: None declared, Manuela Pardeo: None declared, Antonella Insalaco: None declared, Virginia Messina: None declared, Emanuela Sacco: None declared, Ferhat Demir: None declared, Betül Sözeri: None declared, Alenka Gagro: None declared, Nastasia Kifer: None declared, Marija Jelusic: None declared, Francesca Minoia: None declared, Mikhail Kostik: None declared, Olga Vougiouka: None declared, Fabrizio De Benedetti Grant/research support from: Abbvie, SOBI, Novimmune, Roche, Novartis, Sanofi, Pfizer, Claudia Braccaglia: None declared

DOI: 10.1136/annrheumdis-2019-eular.5804

THU0528

DISCONTINUATION OF COLCHICINE THERAPY IN CHILDREN WITH FAMILIAL MEDITERRANEAN FEVER

Yonatan Butbul¹, Rawan Sliman², Shafe Fahoum¹, Yackov Berkun³. ¹Rappaport Children's Hospital, Rambam Medical Center, Haifa, Israel, Pediatric Rheumatology Service, Rappaport Faculty of Medicine, Technion-Israel Institute of Technology, Department of Pediatrics B, Haifa, Israel; ²Rappaport Children's Hospital, Rambam Medical Center, Haifa, Israel, Pediatric Rheumatology Service, Rappaport Faculty of Medicine, Technion-Israel Institute of Technology, Haifa, Israel; ³Hadassah-Hebrew University Medical Center, Mount Scopus, Jerusalem, Israel, Department of Pediatrics and FMF clinic, Haifa, Israel

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

colchicine and the group that needed to renew therapy in demographic, genetic and most clinical parameters, including the age (13.4 ± 3.9 vs 11.9 ± 3.7 p=0.26), level of SAA at enrolment (4 ± 3.6 vs 3.3 ± 2.4 p=0.7) and time of last attack prior to enrolment (12.6 ± 9.6 vs 8.6 ± 8.2 months p=0.08). Myalgia and arthritis were more common among children that required to renew therapy compared to the group that didn't (31% vs 6.7% p=0.058 and 31% vs 3% p=0.024 respectively).

Conclusion: Cessation of colchicine therapy following prolonged remission in selected group of patients who are not homozygous for MEFV mutation could be considered. Patients with arthritis or arthralgia are more likely to have an attack after ceasing colchicine therapy.

REFERENCES:

- [1] Discontinuing colchicine in symptomatic carriers for MEFV (Mediterranean FeVer) variants.
- [2] Sönmez HE, Batu ED, Bilginer Y, Özen S.
- [3] Clin Rheumatol. 2017 Feb;36(2):421-425. d

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2019-eular.2380

THU0529 WHOLE BODY MAGNETIC RESONANCE IMAGING IN JUVENILE DERMATOMYOSITIS. A LONGITUDINAL STUDY

CAPPELLO Carla, Eduardo Bronzatto, Sergio Dertkigil, Roberto Marini, Simone Appenzeller. *University of Campinas, Campinas, Brazil*

Background: Juvenile dermatomyositis (JDM) is a rare multisystem connective tissues disorder of unknown aetiology. Assessment of disease activity is a challenge in clinical practice.

Objectives: To compare whole-body MRI (WB-MRI) with clinical examination in the assessment of disease activity in JDM.

Methods: We included consecutive JDM patients followed in the rheumatology unit. All patients were submitted to clinical and laboratory evaluation. WB-MRI images were obtained using a 1.5 T MRI scanner and short T inversion recovery sequences (STIR). Muscle, peripheral inflammation and subcutaneous inflammations signal abnormalities were scored in 42 muscular groups. Muscle inflammation was classified as: 0 = absent; 1 = Mild to moderate/involvement less than 50% of muscle extension and 2 = Accentuated/greater than 50%. Peripheral and subcutaneous inflammations were classified as: 0 = absent; 1 = present; and on proximal and distal extremities. WB-MRI and clinical assessments were performed concurrently and results compared. Evaluation was repeated after 12 months. Statistics was performed according to the nature of the variable.

Results: WB-MRI revealed muscle inflammation in 6 (31.6%) at study entry. We observed grade 2 muscle inflammation of the right and left scapular girdle (1/19 patients), right and left pelvic girdle (2/19 patients) and right and left thigh (1/19 patients). Grade 1 inflammation was observed in peripheral right and left arm (2/19 patients), peripheral right and left thigh (1/19 patients). Grade 1 subcutaneous inflammation was observed in right and left thigh (1/19 patients) and left leg (1/19 patients). Additionally we observed sacroiliitis (1/19 patients), spinal cord infarction (21%) and osteonecrosis (5.2%). All patients were treated with standardized treatment. After 12 months 13/19 (68.4%) patients repeated the WB-MRI. Five (38.4%) patients had new/worsening of muscle and subcutaneous inflammation, one (7.7%) patient had tibial medullary infarction. Correlations between WB-MRI muscle score and disease activity measures were excellent (Manual Muscle Test: $r = -0.88$, Childhood Myositis Assessment Scale: $r = -0.81$). Patients with subcutaneous inflammation developed clinically evident subcutaneous calcifications during follow-up.

Conclusion: WB-MRI provides additional information to clinical evaluation and represents a promising tool to determine the grade of muscle inflammation to additional peripheral and subcutaneous tissue inflammation.

REFERENCES:

- [1] Malattia C, Damasio MB, Madeo A, et al. Ann Rheum Dis 2014;73:1083-1090

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2019-eular.6037

THU0530 IDIOPATHIC RECURRENT PERICARDITIS: CLINICAL FINDINGS AND TREATMENT APPROACH

Camilla Celani¹, Silvia Federici¹, Anna Tulone¹, Brigitte Bader-Meunier², Virginia Messina¹, Manuela Pardeo¹, Claudia Bracaglia¹, Pierre Quartier², Fabrizio De Benedetti¹, Antonella Insalaco¹. ¹IRCCS Ospedale Pediatrico Bambino Gesù, Division of Rheumatology, Rome, Italy; ²Hôpital Necker Enfants Malades, Unité d'Immunologie-Hématologie et Rhumatologie pédiatrique, Paris, France

Background: Recurrent pericarditis affects 15-30% of patients with acute pericarditis. The etiology is poorly understood, with about 80% being idiopathic. Several treatment options are available for recurrences, including NSAIDs, colchicine, glucocorticoids and IL-1 inhibitors (i.e. Anakinra). Standardized guidelines for the management of these patients are still lacking

Objectives: To analyze clinical findings and treatment approach in a cohort of pediatric patients with recurrent pericarditis

Methods: Patients with at least two episodes of idiopathic pericarditis, followed at two Pediatric Rheumatology centers between 2006 and 2018, were included

Results: A total of 42 patients (18 males) were included. Mean age at disease onset was 11.8 years (range 4-17). Chest pain and fever were the presenting symptoms in all patients. In 47% pleural effusion was detected. Laboratory tests showed increased white blood cell count (mean $14.509/\text{mm}^3$), C-reactive protein (mean 18.01 mg/dl) and erythrocyte sedimentation rate (mean 39 mm/h) in all patients. The first episode was variably treated: 18/42 (43%) received NSAIDs alone, 5/42 (11.9%), colchicine alone or associated to NSAIDs and 3/42 patients (7%) received antibiotics alone. 16/42 (38%), not responsive to NSAIDs or colchicine, received glucocorticoids. Patients who received glucocorticoids at the first episode relapsed earlier (median time of 2.1 months range 10 days-5 months), than patients treated with NSAIDs (6.6 months range 10 days -24 months) or with colchicine (5 months range 10 days-5 months) (p<0.05). In our study, initial treatment of the first episode did not affect the number of subsequent flares. To evaluate treatment strategy at relapses, we divided our study population in two groups: Group 1 (20 pts) in which recurrence was treated with NSAIDs, colchicine or glucocorticoid (alone or combined); group 2 (22 patients) in which anakinra was started. Among patients belonging group 2, 9 received anakinra at first relapse, 7 at the second, 2 at the third and 2 at the fourth. Anakinra treatment was followed by a prompt resolution of symptoms and inflammatory signs within 2 days. During daily treatment with full dose anakinra, no relapses were reported over a median of 13.3 months (range 5-24 months). In 13 out of 22 patients, anakinra was gradually tapered reducing the days of administration during the week. Four of these patients relapsed. The mean time from the start of anakinra to tapering was 17 ± 4 months (range 14-23 months) in the 4 patients who experienced a relapse versus 14 ± 4 months (range 7-21 months) in patients who did not flare, with no statistical difference. Among the 22 patients belonging to group 2 anakinra was finally discontinued in 11 after a mean time of 23.4 months (range 12-36). Among these, 8 relapsed after anakinra withdrawal including 2 of the 4 patients already relapsed during tapering). Only 3 patients didn't present any relapse (up to 20.3 months of follow-up). All patients who relapsed responded quickly to the reintroduction of anakinra

Conclusion: Our study confirms the lack of a standardized treatment approach in patients with recurrent pericarditis. Patients treated with glucocorticoid at first episode relapse before than those treated with other drugs. Anakinra is an effective treatment; however, tapering/discontinuation of the drug lead to relapses in several cases. Further experience on larger population is needed to define the best treatment duration and approach to withdrawal of IL-1 inhibitor

Disclosure of Interests: Camilla Celani: None declared, Silvia Federici: None declared, Anna Tulone: None declared, Brigitte Bader-Meunier: None declared, Virginia Messina: None declared, Manuela Pardeo: None declared, Claudia Bracaglia: None declared, Pierre Quartier Consultant for: AbbVie, Chugai-Roche, Lilly, Novartis, Novimmune, Sanofi, and SOBI, Consultant for: AbbVie, Chugai-Roche, Lilly, Novartis, Novimmune, Sanofi, and SOBI, Speakers bureau: AbbVie, BMS, Chugai-Roche, Novartis, Pfizer, and SOBI, Speakers bureau: AbbVie, BMS, Chugai-Roche, Novartis, Pfizer, and SOBI, Fabrizio De Benedetti Grant/research support from: Abbvie, SOBI, Novimmune, Roche, Novartis, Sanofi, Pfizer, Antonella Insalaco: None declared

DOI: 10.1136/annrheumdis-2019-eular.6219