CC motif ligand 2 chemokine (CCL2) were measured in PMR patients and in healthy controls.

Results: A total of 25 patients with PMR were studied (32% men; 68% women). The mean age was 72.72 ± 9.32 years (range 65–79). We also included 35 healthy controls. In reference to laboratory parameters and proinflammatory biomarkers at the onset of PMR, the mean values ($\pm$ standard deviation) were as follows: C reactive protein (CRP): 28.4 ± 26.6 mg/dL; erythrocyte sedimentation rate (ESR): 54.3 ± 21.3 mm/h; haemoglobin: 12.6 ± 1.6 g/dL; IL-6: 17.6 ± 30.1 pg/mL; IL-8: 10.1 ± 6.9 pg/mL; CXCL10: 259.4 ± 290.7 pg/mL; CXCL9: 5175 ± 4396.8 pg/mL; CXCL2: 25.9 ± 12.5 pg/mL; CCL2: 471.3 ± 152.9 pg/mL.

IL-6, IL-8 and CXCL9 levels were significantly increased in PMR patients comparing with the healthy control group.

The Health Assessment Questionnaire (HAQ) performed at the onset of the PMR yielded a mean value of 1.6 ± 0.6 . The mean dose of prednisone employed at the onset of PMR was 14.8 ± 5.6 mg/day.

After 6 months, the mean dose of prednisone employed was 6.7 ± 3.9 mg/day, the median [range] value for CRP was 3.1 [1.75 – 7.2] mg/dl and the mean value for ESR was 22 ± 14 mm/h. Regarding relationship between proinflammatory biomarkers studied and doses of prednisone employed, no differences were observed.

Regarding pain perception and disability, we observed higher visual analogic scale values (VAS) in association with higher IL-6 levels ($P=0.03$) and higher HAQ values in association with higher IL-6 and CXCL2 values.

After 6 months, our patients showed an excellent evolution, and no complications were observed.

Conclusion: During the acute onset of PMR, IL-6, IL-8 and CXCL9 levels were significantly increased in our population of PMR patients. No differences were observed between proinflammatory biomarkers studied and doses of prednisone employed.

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2021-eular.3114

POS1366

THE RELATIONSHIP OF MAST CELL ACTIVATION SYNDROME AND HYPERMOBILE EHLERS-DANLOS SYNDROME IN HOSPITALIZED PATIENTS IN THE UNITED STATES

K. Mathias¹, A. Mantha², L. Mathias³, D. Arkfeld³. ¹University of Chicago, Internal Medicine, Chicago, United States of America; ²USC Keck School of Medicine, Internal Medicine, Los Angeles, United States of America; ³USC Keck School of Medicine, Rheumatology, Los Angeles, United States of America

Background: Patients with hypermobile Ehlers-Danlos syndrome (hEDS) often report symptoms of cutaneous flushing, pruritis, vomiting, diarrhea and anaphylactoid responses to environmental, food, and mechanical stimuli. There is a growing body of literature investigating the possible link between disorders of mast cell regulation including mast cell activation syndrome (MCAS) and connective tissue disorders including hEDS. The evidence for such a relationship, however, remains limited¹.

Objectives: We aimed to evaluate the association between hEDS and MCAS among hospitalized patients using a nationally representative cohort.

Methods: Hospitalized patients with a diagnosis of hEDS or MCAS were identified in the 2016–2018 National Inpatient Sample (NIS) using the International Classification of Diseases 10 system (ICD-10). The NIS is an all-payer inpatient database that estimates over 37 million annual U.S. hospitalizations and is maintained by the Healthcare Cost and Utilization Project. The primary outcomes were prevalence of EDS and MCAS among hospitalized patients. Secondary outcomes included cause of admission, mortality, length of stay, and cost of care. Multivariate hierarchical regression analysis was using adjusting for demographics, hospital factors, and comorbid conditions.

Results: Among 37,665 patients identified in this study, 35,115 (95.9%) patients had hEDS and 3,630 (9.6%) patients had MCAS. 1080 patients had concomitant diagnosis of hEDS (3.1%) and MCAS (29.8%), of whom 555 (51.4%) were diagnosed in 2018 compared with 100 (9.3%) patients in 2016 ($P<0.001$). The mean age of patients with both conditions was significantly lower than those diagnosed with hEDS (29.5 vs 36.6 years, $P<0.001$) or MCAS (29.5 vs 39.8 years, $P<0.001$) alone. Those with comorbid hEDS and MCAS were more likely female (95.8% vs 84.5%, $P<0.001$), had private insurance (67.6% vs 52.4%, $P<0.001$) and were among the highest income quartile (47.4% vs 31.9%, $P<0.001$). There were no significant differences in mortality or overall comorbidity burden. Patients with hEDS and MCAS had higher adjusted hospitalization charges (\$64,445 vs \$54,782, $\beta=0.15$, $P<0.05$) and longer adjusted lengths of stay (6.4 vs 4.9 days, IRR 1.35, $P<0.001$).

Conclusion: Nearly 1 in 3 patients diagnosed with MCAS have comorbid diagnosis of hEDS. This association appeared to increase over time with the increasing diagnosis of MCAS. Social determinants of disease such as health insurance status and income appeared to be associated with the identification of the overlap

syndrome. These findings merit further investigation into the co-occurrence of these disease entities and development of consistent clinical diagnostic criteria.

REFERENCES:

- [1] Kohn A, Chang C. The Relationship Between Hypermobile Ehlers-Danlos Syndrome (hEDS), Postural Orthostatic Tachycardia Syndrome (POTS), and Mast Cell Activation Syndrome (MCAS). *Clin Rev Allergy Immunol*. 2020;58(3):273–297. doi:10.1007/s12016-019-08755-8

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2021-eular.3177

POS1367

PROCALCITONIN IN RHEUMATOLOGY

K. Erraoui¹, K. Nassar², S. Janani¹. ¹CHU Ibn Rochd, Department of Rheumatology, Casablanca, Morocco; ²CHU Ibn Rochd, Department of Rheumatology, Casablanca, Morocco

Background: The differentiation between infection and aseptic inflammation is difficult and often a challenge in the daily practice of rheumatology. Procalcitonin (PCT) is a preferred marker for bacterial infections that allows the rheumatologist to make an early diagnosis, choose an appropriate antibiotic therapy for bacterial infection, and avoid unnecessary antibiotic therapy for inflammation.

Objectives: To study the levels of PCT in various inflammatory states observed in a rheumatology department, and to assess its possible discriminating role in the differentiation of bacterial infection from other inflammatory processes.

Methods: This is a preliminary monocentric retrospective study conducted from January 2018 to December 2020 in our rheumatology department. The inclusion criteria were all patients hospitalized in rheumatology for a rheumatic or autoimmune disease or a bone and joint infection. PCT samples from patients were analyzed along with general signs (fever/chills) and with routine assessment (blood count including white blood cell count (WBC), erythrocyte sedimentation rate (ESR) and C reactive protein (CRP)) in patients admitted to hospital for a fever or a biological inflammatory syndrome, or both, with a laboratory cut-off value of 0.5 ng/ml.

Results: 20 patients were included. Their mean age was 46.1 (extremes 19 – 66) and the sex ratio was 1. Three patients had a history of autoimmune disease, 2 of neoplasia, 1 with chronic terminal kidney failure and hemodialysis, and 4 patients took antibiotics before hospitalization. The mean PCT was 100.01 ng/ml (extremes 0.01–200), and Sensitivity at 57.44% and specificity at 100%, PPV: 100%, NPV: 81.25%.

The patients were divided according to their final diagnosis into 3 groups: bacterial or fungal infection; inflammatory abacterial disease (autoimmune or rheumatic flare, neoplasia, metabolic, reactive arthritis); infection + inflammatory abacterial disease. Group 1 included 4 patients, their mean PCT was 6.92 ng/ml (0.16–26.80), mean CRP was 131.725 mg/L, and mean ESR was 50 mm,

	Group 1(n=4)	Group 2(n=13)	Group 3(n=3)
Mean age	45	46.77	44.67
Sex ratio M/F	2/2	8/5	0/3
Reason for hospitalization	Phlegmon of the left shoulder and inflammatory gonalgia in a feverish context 25% Multiple vertebral fractures 25% Acute monoarthritis of the left knee 25% Pain in the left hip with functional impairment 25%	Inflammatory polyarthralgia 15.38% Inflammatory back pain 23.07% Activity of his autoimmune disease 15.38% Chronic hip pain 7.69% Acute febrile polyarthralgia 30.77% Biarthritis 7.69%	Acute febrile polyarthralgia 66.67% Flare of polymyalgia rheumatica 33.33%
History	Neoplasia 25%	Autoimmune disease 23.07% Suspicion of arthritis 46.15%	End-stage chronic renal disease with hemodialysis 33.33% Suspicion of sepsis 33.33%
PCT indication	Suspected septic arthritis 75% Exploration of inflammatory syndrome 25%	Exploration of inflammatory syndrome 53.85%	Exploration of inflammatory syndrome 66.67%
Mean PCT value (ng/ml)	6.92	0.025	66.88
fever/chills	75%	30.77%	33.33%
Biological parameters	25%	30.77%	33.33%
Hyperleukocytosis	50	84.38	107.33
ESR (mm)	131.725	132.25	560
CRP (mg/L)			

and 25% had hyperleukocytosis. 2 patients had already taken antibiotics before admission. Group 2 included 13 patients, their mean PCT was 0.025 ng/mL (0.01-0.13), mean CRP was 132.25 mg/L, mean ESR was 84.38 mm, and 30.77% of them had hyperleukocytosis. Group 3 included 3 patients, their mean PCT was 66.88 ng/mL (0.02-200), mean CRP was 560 mg/L, mean ESR was 107.33 mm, and 33.33% had hyperleukocytosis. The patient with the highest PCT was admitted for a sepsis with uremic encephalopathy that improved after hemodialysis. A control PCT was made in 2 patients with infection who received antibiotics, showing a decrease or negatization compared to the initial value of PCT.

Conclusion: Our study showed that PCT levels only increased significantly in bacterial infections. The PCT values were more discriminating than the level of WBC and CRP in differentiating a bacterial infection from another inflammatory process, hence the value of using this marker in case diagnosis doubt.

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2021-eular.3191

POS1368

ANTI-IL-1 THERAPIES IN COLCHICINE-RESISTANT OR INTOLERANT PATIENTS WITH FAMILIAL MEDITERRANEAN FEVER: SINGLE CENTER EXPERIENCE

M. E. Derin¹, B. Karakaş², B. Karataş², N. Çabuk Çelik², İ. Yalçın², A. Şahin².

¹Sivas Numune Hospital, Internal Medicine – Rheumatology, Sivas, Turkey;

²Sivas Cumhuriyet University, Internal Medicine – Rheumatology, Sivas, Turkey

Background: Familial Mediterranean Fever (FMF) is a hereditary auto-inflammatory disease characterized by recurrent fever and serosal inflammation (1). The goal of FMF treatment is to prevent the attacks and to minimize subclinical inflammation between attacks. The main treatment of FMF is colchicine however anti-interleukin-1 treatments are recommended in colchicine resistant and/or intolerant FMF patients (2).

Objectives: The aim of this study is to evaluate the efficacy of anti-interleukin-1 (anti-IL-1) agents in 81 FMF patients with resistant/intolerant to colchicine or complicated with amyloidosis.

Methods: Between January 2014 and December 2020, eighty-one patients who were diagnosed as FMF according to the criteria of Tel-Hashomer that following-up at Cumhuriyet University Medical Faculty Rheumatology-Internal Medicine Department were included in the study.

Results: 45 (55.6%) male and 36 (44.4%) female were included in the study. The median age of the patients was 25 years (min:17-max: 60) and the median age at diagnosis was 15 years (min 3-max 46). 44 patients (54.3%) used Anakinra (100mg/day), and 27 (45.7%) canakinumab (150mg/8month) were used. 49 cases were resistant to colchicine, 16 were intolerant to colchicine, 16 (20%) cases were complicated with amyloidosis. 10 patients had renal transplantation. MEFV gene mutations are shown in Table 1. Median duration of anti-IL-1 agent use was 24 month (min:4-max 52). 9 patients were resistant to anakinra, 18 patients had side effects which anakinra related. After a median follow up 12 months overall clinical response was %95 (frequency of attacks <1/6months). median proteinuria decreased from 3500 mg /day to median 1500 mg /day (p: 0.04) (Table 2). IL-6 treatment was started in 4 patients because of ineffective canakinumab. Five pregnant patients were followed up with anakinra during pregnancy and there were no problems.

Conclusion: Anti-interleukin-1 agents are effectively and safely in the treatment of FMF patients. There are still unanswered questions in FMF treatment such as other factors affecting the frequency of attacks, colchicine resistance is not defined precisely and the importance of some mutations. The effect of anti IL-1 agents on FMF patients with amyloidosis is not clearly. According to our experience, these treatments are effective in patients with glomerular filtration rate > 60 ml/min. For answers to these and similar questions, Large and long follow-up studies are needed for long-term effects.

REFERENCES:

- Özen S, Batu ED, Demir S., Familial Mediterranean Fever: Recent Developments in Pathogenesis and New Recommendations for Management. *Front Immunol.* 2017 Mar 23;8:253. doi: 10.3389/fimmu.2017.00253. eCollection 2017.
- Seza Özen ve ark. EULAR recommendations for the management of familial Mediterranean fever. *Ann Rheum Dis.* 2016 Apr;75(4):644-51.

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2021-eular.3257

POS1369

ADULT-ONSET STILL'S DISEASE: A SINGLE-CENTER EXPERIENCE

B. Egeli¹, S. Ugurlu². ¹Boston Children's Hospital, Immunology, Boston, United States of America; ²Istanbul University-Cerrahpasa, Internal Medicine, Istanbul, Turkey

Background: Adult-Onset Still's disease (AOSD) is an autoinflammatory condition characterized by fever, rash, and arthritis. The diagnosis of AOSD is made by excluding common causes of fever of unknown origin which are infections, malignancies, autoimmune conditions and medication adverse effects. As it is a diagnostic challenge, further data on highlighting clinical and laboratory findings are necessary on guiding clinicians.

Objectives: Our main objective is to present our single tertiary center experience of patients diagnosed with AOSD.

Methods: This retrospective study was conducted at a tertiary rheumatology center. Patients were diagnosed with AOSD using Yamaguchi's criteria and followed between 2007 and 2020. Demographic, clinical and laboratory information was retrieved from the patient charts. Treatment-related and prognostic information were also noted with additional information from phone call interviews.

Results: The study includes 69 patients (23M, 46 F). The mean age of diagnosis was 33.86±14.3. The presenting signs and symptoms of the patients are shown in Figure 1. The laboratory findings supporting the diagnosis at initial encounter are summarized in Table 1. The mean corticosteroid dose at initial diagnosis was 29.7±18 mg. In addition to corticosteroid treatment these patients were followed with different glucocorticoid-sparing agents. Methotrexate was the choice of treatment in 54 patients with the mean dose of 14.5±3.43 mg. Eight patients were treated with leflunomide, seven with anti-TNF agents, seven with tocilizumab, nineteen with anakinra and four with canakinumab.

Figure 1. The presenting signs and symptoms of the patients

Conclusion: In conclusion, the most common presenting symptoms in our AOSD cohort were fever and salmon-colored rash. In the differential diagnosis of fever of unknown origin especially with rash, AOSD should be considered. Corticosteroid was the main treatment modality. In patients who are unresponsive to conventional immunosuppressive treatment, biologic agents can be an alternative.

Table 1. The laboratory findings at initial encounter

	Mean ± Standard Deviation
Ferritin (ng/mL)	3179.46±6503.56
ESR (mm/h)	77.43±28.47
CRP (mg/L)	102.29±70.39
Leukocyte Count (cells/L)	13147.3±4640.9
ESR (mm/h)	80±28.48
CRP (mg/L)	105.15±54.67
Leukocyte Count (cells/L)	12427.14±6530.43

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2021-eular.3280

POS1370

HYDROXYCHLOROQUINE-INDUCED ATRIOVENTRICULAR BLOCK IN IMMUNE-MEDIATED DISEASES. SINGLE UNIVERSITY CENTER STUDY OF 293 PATIENTS

A. Herrero-Morant¹, A. Margarida-de Castro¹, R. Pérez-Barquín¹, J. Zubiaur-Zamacoila¹, M. Á. González-Gay¹, R. Blanco¹. ¹Hospital Universitario Marqués de Valdecilla, Rheumatology and Cardiology, Santander, Spain

Background: Hydroxychloroquine (HCQ) is an extensively used drug in immune-mediated diseases (IMID). Despite its general safety, HCQ can cause serious toxicity such as heart conduction disorders. Atrioventricular block (AVB)